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ABSTRACTS

Abstracts of the 54th ESPN Annual Meeting, Ljubljana, Slovenia

ORAL PRESENTATIONS

OP-1 LANDSCAPE OF SUBCLINICAL REJECTION IN A LARGE INTERNATIONAL COHORT OF PEDIATRIC KIDNEY TRANSPLANT (KTX) RECIPIENTS

Julien Hogan[11], Rouba Garro[2], Gillian Divard[1], Olivia Boyer[3], Jodi Smith[4], Katherine Twombley[5], Brad Warady[6], Patricia Weng[7], Rima Zahr[8], Rachel Patzer[9], Alexandre Loupy[1], Alton Brad Farris[10]

[1]Paris Transplant Group, University Of Paris, Parcc, Inserm, France, [2]Pediatric Nephrology, Children Healthcare Of Atlanta, Emory University, Atlanta, GA, USA, [3]Pediatric Nephrology, Necker Hospital, Aphp, Paris, France, [4]Pediatric Nephrology, Seattle Children, Seattle, NY, USA, [5]Pediatric Nephrology, Medical University Of South Carolina, Charleston, SC, USA, [6]Pediatric Nephrology, Children's Mercy, Kansas City, MI, USA, [7]Pediatric Nephrology, David Geffen School Of Medicine At Ucla, Los Angeles, CA, USA, [8]Pediatric Nephrology, Le Bonheur Childrens Hospital, Memphis, TN, USA, [9]Emory Transplant Center, Department Of Surgery, Emory University, Atlanta, GA, USA, [10]Department Of Pathology, Emory University School Of Medicine, Atlanta, GA, USA, [11]Pediatric Nephrology, Robert Debre Hospital, Aphp, Paris, France

Introduction: Kidney allograft rejection can occur in clinically stable patients, but long-term significance in pediatric kTx recipients is unknown. Previous single-center studies demonstrated that subclinical borderline (SC-Borderline) or T-cell mediated rejection (SC-TCMR) are associated with an increased risk of acute rejection. However, the prevalence and significance of subclinical antibody-mediated rejection (SC-AMR) and the impact of subclinical rejection phenotypes on graft survival remained to be assessed.
Material and methods: We used data from pediatric (<21) patients transplanted between 2005 and 2017 from 8 institutions in France and the United States performing surveillance biopsies. Biopsies were identified as surveillance if they were recorded as such in the medical record with no significant increase in serum creatinine or proteinuria. Biopsies were graded according to the Banff 2019 criteria. DSA screening was performed according to each center protocol. Kaplan Meier method and log-rank test were used to compare the risk of acute rejection, transplant glomerulopathy and graft loss stratified on the surveillance biopsies' findings.
Results: 1390 surveillance biopsies were performed in 763 kTx recipients including 135 (9,7%) SC-borderline, 46 (3,3%) SC-TCMR, 54 (3,9%) SC-ABMR, 8 (0,6%) subclinical mixte rejections. Subclinical rejection was associated with acute rejection with 5-year rejection-free survival of 88%, 78%, 68% and 63% in the no rejection, SC-borderline, SC-TCMR and SC-AMR groups, respectively (p<0,0001). SC-TCMR and SC-AMR were associated with the development of transplant glomerulopathy, p<0,0001. Subclinical AMR only was associated with a lower 5-year graft survival (79% vs. 93% (SC-TCMR), 95% (SC-Borderline), 94% (no rejection)), p=0,002.
Conclusions: Subclinical rejection is prevalent in pediatric kidney recipients without clinical dysfunction and is associated with acute rejection. Subclinical AMR is associated with the development of transplant glomerulopathy and with an increased risk of allograft failure.

OP-2 TIME-DEPENDENT EFFECTS OF DONOR AND RECIPIENT CHARACTERISTCS ASSOCIATED WITH GRAFT SURVIVAL AFTER FIRST PAEDIATRIC KIDNEY TRANSPLANTATION: EUROTRANSPLANT REGISTRY ANALYSIS

Ferran Coens[1], Noël Knops[2], Ineke Tieken[4], Serge Vogelaar[4], Jon Jin Kim[3], Agnieszka Prytula[1]

[1]Department Of Pediatric Nephrology And Rheumatology, Ghent University Hospital, Ghent, Belgium, [2]Department Of Pediatric Nephrology, University Hospital Leuven, Leuven, Belgium, [3]Nottingham University Hospital Nhs Trust, United Kingdom, [4]Eurotransplant, Leiden, The Netherlands

Introduction: The aim of this study was to identify time-dependent effects of factors associated with time to death-censored graft survival after paediatric kidney transplantation (KTx).
Material and methods: Data on patients younger than 18 years at primary KTx between 1990 and 2020 were provided by the Eurotransplant Registry. A piecewise-exponential additive mixed model with country level frailty was applied to explore time-varying variables associated with time until death-censored graft loss (DCGL) after first paediatric kidney transplantation.
Results: We included 4528 KTx recipients with median age at KTx 11 years (interquartile range [IQR] 6-14) and median follow-up of 10 years (IQR 4-17). During the follow-up 1688 patients (37.3%) experienced graft failure (GF) and 159 (3.5%) died with functioning graft. The respective 5-year GF after live (LD) and deceased donor (DD) KTx performed in 1990-2000 were 14% and 25% as compared to 4% and 12% in KTx between 2011-2020. There was a time-varying association between time to DCGL (P<0.05) and donor source, recipient and donor age and panel reactive antibodies (PRA). The benefit of LD was most pronounced in the first months after KTx, then decreased steadily and was no longer significant from 130 months on. Increasing recipient age corresponded with a decline in adjusted hazard ratio (aHR) for DCGL in the first months post KTx, but had the opposite effect between 50 and 100 months. Donor age of 20 years corresponded with the lowest aHR for DCGL. Panel reactive antibodies (PRA) 1-15% was associated with a higher aHR than PRA 0% up until 50 months post KTx. The aHR for DCGL decreased with ascending year of KTx up until 2000 and remained stable afterwards.
Conclusions: The effects of factors associated with DCGL vary over time and should be considered at clinical decision making.

OP-3 DETERMINANTS OF PATIENT SURVIVAL AFTER PAEDIATRIC KIDNEY TRANSPLANTATION: THE EUROTRANSPLANT REGISTRY ANALYSIS

Ferran Coens[1], Noël Knops[2], Ineke Tieken[3], Serge Vogelaar[3], Jon Jin Kim[4], Agnieszka Prytula[1]

[1]Department Of Pediatric Nephrology And Rheumatology, Ghent University Hospital, Ghent, Belgium
[2]Department Of Pediatric Nephrology, University Hospital Leuven, Leuven, Belgium
[3]Eurotransplant, Leiden, The Netherlands
[4]Nottingham University Hospital Nhs Trust, United Kingdom

Introduction: We report patient survival and factors associated with time to death after first and sequential paediatric kidney transplantation (KTx).
Material and methods: Data on patients younger than 18 years at first KTx between 1990 and 2020 were provided by the Eurotransplant Registry. Multivariable Cox-regression models with country-level clustering and time-dependent covariates were applied to identify factors associated with overall patient survival.
Results: We included 4528 patients with 5987 grafts with median age at primary KTx 11 years (interquartile range [IQR] 6-14) and median follow-up of 10 years (IQR 4-17). The 10- year mortality after live donor (LD) and deceased donor (DD) KTx performed in 1990-2000 was 4% and 7% as compared to 2% and 4% in KTx performed in 2011-2020. The 15-year mortality risk after the 1st, 2nd, 3rd and 4th KTx was 9%, 12%, 16% and 26%. The risk of death was lower after live donor (LD) KTx (unadjusted hazard ratio [uHR] 0.53, 95% confidence intervals [CI] 0.40-0.69, log-rank P < 0.001). Time to death was associated with: LD (adjusted hazard ratio [aHR] 0.64, CI 0.46-0.89, P=0.007), ascending year of KTx (aHR 0.98, CI 0.97-0.99, P< 0.001), dialysis after 1st (aHR 4.31, CI 3.35-5.55, P< 0.001), 2nd (aHR 8.37, CI 6.75-10.40, P< 0.001) and 3rd graft failure (aHR 7.51, CI 4.23-13.35, P< 0.001), donor age above 50 years (aHR 1.70, CI 1.12-2.60, P = 0.014), HLA-mismatch equal to 5 (aHR 2.69, CI 1.77-4.10, P<0.001) or 6 (aHR 3.90, CI 3.25-4.68, P<0.001), male recipient-female donor (aHR 1.38, CI 1.20-1.58, P<0.001), female recipient-male donor (aHR 1.16, CI 1.07-1.26, P<0.001) and glomerular kidney disease (aHR 1.20, CI 1.06-1.34, P=0.002).
Conclusions: Recipient and donor characteristics are subject to clinical decision making and should be considered at patient counselling and acceptance of organs for KTx.

OP-4 ACUTE KIDNEY INJURY ASSOCIATED WITH COVID-19 AND MULTISYSTEM INFLAMMATORY SYNDROME IN CHILDREN (MIS-C)

Tugba Tastemel Ozturk[1], Ali Duzova[1], Pembe Derin Oygar[2], Demet Baltu[1], Pelin Ozcilingir[2], Sibel Lacinel Gurlevik[2], Eda Didem Kurtsukur[1], Hayrettin Hakan Aykan[4], Seza Ozen[5], Ilker Ertugrul[4], Selman Kesici[5], Bora Gulhan[1], Fatih Ozaltin[1], Yasemin Ozsurekci[2], Ali Bulent Cengiz[2], Rezan Topaloglu[1]

[1]Division Of Pediatric Nephrology, Hacettepe University Faculty Of Medicine, Ankara, Turkey, [2]Division Of Pediatric Infectious Diseases, Hacettepe University Faculty Of Medicine, Ankara, Turkey, [3]Department Of Pediatrics, Hacettepe University Faculty Of Medicine, Ankara, Turkey, [4]Division Of Pediatric Cardiology, Hacettepe University Faculty Of Medicine, Ankara, Turkey, [5]Division Of Pediatric Rheumatology, Hacettepe University Faculty Of Medicine, Ankara, Turkey, [6]Division Of Pediatric Intensive Care, Hacettepe University Faculty Of Medicine, Ankara, Turkey

Introduction: Data on the characteristics of acute kidney injury (AKI) in pediatric COVID-19 and MIS-C are limited. We aimed to define the frequency, associated factors and early outcome of AKI in COVID-19 and MIS-C.
Material and methods: Hospitalized patients ≤18 years of age with confirmed COVID-19 or MIS-C at a tertiary center, between March 2020 - December 2021 were enrolled. AKI was defined and staged according to KDIGO criteria. The characteristics of AKI in the COVID-19 group were investigated in moderate, severe and critically ill patients; outpatients and mild cases who do not have shortness of breath, dyspnea, or abnormal chest imaging were excluded.
Results: The study included 66 moderate-severe-critically ill patients with COVID-19 (9.71 ± 6.08 years) and 111 MIS-C patients (8.72 ± 4.72 years). The frequency of AKI was 22.7% in COVID-19 and 15.3% in MIS-C; among them AKI was present on admission in 73.3% and 88.2% of COVID-19 and MIS-C groups, respectively. In univariate analyzes, presentation with vomiting/diarrhea, high LDH, D-dimer, troponin and procalcitonin on admission were associated with AKI in COVID-19 patients; whereas older age, low albumin, hemoglobin, thrombocyte, and high CRP, procalcitonin, ferritin, D-dimer, troponin, and BNP levels and low ejection fraction on echocardiography on admission were associated with AKI in MIS-C group. Length of hospital stay was significantly longer in both COVID-19 and MIS-C patients with AKI, compared to those without AKI. Mortality was 9.1% in COVID-19 group; there was no mortality in MIS-C patients. AKI was associated with mortality in COVID-19 patients (p=0.021). Serum creatinine returned to normal level in 96% of survivors before discharge.
Conclusions: AKI was seen in 15% of moderate-severe-critically ill COVID-19 group and 23% of MIS-C; it was associated with mortality in COVID-19. Clinical and laboratory parameters associated with AKI were different in COVID-19 and MIS-C. Early outcome was excellent among survivors.

OP-5 SODIUM BALANCE IN CHILDREN ON DIALYSIS: A MULTICENTER PROSPECTIVE STUDY FROM THE EUROPEAN PEDIATRIC DIALYSIS WORKING GROUP

Fabio Paglialonga[1], Rukshana Shroff[2], Ilona Zagozdzon[3], Sevcan Bakkaloglu[4], Ariane Zaloszyc[5], Augustina Jankauskiene[6], Alejandro Cruz[7], Silvia Consolo[1], Alberto Edefonti[1]

[1]Pediatric Nephrology, Dialysis And Transplant Unit, Fondazione Irccs Ca Granda Ospedale Maggiore Policlinico, Milan, Italy, [2]University College London, Great Ormond Street Hospital For Children And Institute Of Child Health, London, Uk, [3]Department Paediatrics, Nephrology And Hypertension, Medical University Of Gdansk, Gdansk, Poland, [4]Gazi University Hospital, Ankara, Turkey, [5]Department Of Pediatric Nephrology, Hopital De Hautepierre, Strasbourg, France, [6]Pediatric Center, Institute Of Clinical Medicine, Vilnius University, Vilnius, Lithuania, [7]Department Of Pediatric Nephrology, Hospital Vall D'hebron, Barcelona, Spain

Introduction: We aimed to assess the sodium balance (NaB) and its clinical correlates in children receiving maintenance dialysis.
Material and methods: We recruited patients < 18 years undergoing thrice weekly hemodialysis (HD) or automated peritoneal dialysis (PD) in the European Pediatric Dialysis Working Group (EPDWG) centers. NaB was calculated from enteral Na intake (eNa), obtained by a three-day dietary diary, medication-related Na intake, and urinary Na losses (uNa), measured by 24-h urine collection. Blood pressure (BP) was measured by 24h-ABPM in children >5 years and median office BP in younger patients. Percentage interdialytic weight gain (IDWG), pre-HD office BP, and first-hour refill index (an index of pre-HD fluid overload based on BVM) were assessed in HD patients, ultrafiltration (UF/kg/session) in PD children.
Results: 41 patients (31 HD, 10 PD), median age 13.3 (10.7-15.8) years, were recruited. Median eNa was 1.3 (1.0-2.0) mEq/kg/day; 12 patients (29.3%) received Na-containing drugs, accounting for 0.6 (0.5-1.1) mEq/kg/day (=40% of the total Na intake in this group). Median total Na intake was 1.5 (1.2-2.3) mEq/kg/day, corresponding to 60.6% (40.9-80.2%) of the maximum RDI for healthy children, and uNa 0.6 (0.1-1.8) mEq/kg/

day. Median NaB was +0.88 (-0.05 to +1.7) mEq/kg/day. NaB negatively correlated with age, urine output and plasma Na; neither systolic nor diastolic BP SDS significantly correlated with NaB. In HD patients, NaB significantly correlated with IDWG (r^2=0.59; p<0.0001), preHD diastolic BP (r^2= 0.30; p=0.001), first-hour refill index (r^2= 0.57; p=0.002). A multivariable analysis including age and urine output demonstrated that NaB was the only significant predictor of IDWG (b=0.85, p<0.0001). A tendency towards a positive correlation between NaB and UF/kg was found in PD patients.
Conclusions: NaB is a significant and independent predictor of IDWG in children on HD. The lack of correlation between NaB and blood pressure deserves further investigation.

OP-6 GLOBAL TRENDS IN PERITONEAL DIALYSIS-ASSOCIATED PERITONITIS IN CHILDREN: FINDINGS FROM THE INTERNATIONAL PEDIATRIC PERITONEAL DIALYSIS NETWORK (IPPN).

Dagmara Borzych-duzalka[1], Lisa Sartz[1], Bruno Ranchin[1], Nikoleta Printza[2], Dorota Drozdz[3], Sevcan Bakkaloglu[4], Tuula Holtta[5], Heiko Billing[6], Koen Van Hoeck[7], Ovidiu Brumariu[8], Lisa Sartz[9], Nakysa Hooman[10], Rayner Loza Munarriz[11], William Wong[12], Bradley Warady[13], Franz Schaefer[14]

[1]Medical University Of Gdansk, Gdansk, Poland, [2]Hippokration General Hospital, Aristotle University, Thessaloniki, Greece., [3]Jagiellonian University Medical College, Krakow, Poland, [4]Gazi University Faculty Of Medicine, Ankara, Turkey, [5]Helsinki University Hospital, Helsinki, Finland., [6]Children University Hospital, Tuebingen, Germany, [7]University Hospital Antwerp, Belgium, [8]St.maria Childrens Hospital, Iasi, Romania, [9]Barnkliniken, Lund, Sweden, [10]Iran University Of Medical Sciences, Teharan, Iran, [11]Cayetano Heredia Hospital, Lima, Peru, [12]Starship Childrens Hospital, Auckland, New Zealand,, [13]Childrens Mercy Hospital, Kansas City, USA, [14]Center For Pediatrics And Adolescent Medicine, Heidelberg University, Heidelberg, Germany.

Introduction:
Peritonitis remains a frequent complication of peritoneal dialysis in children. We described the peritonitis rate, etiology, clinical presentation, and outcome experienced by patients followed in the IPPN registry.
Material and methods: Review of prospectively collected data submitted to the IPPN registry (www.pedpd.org) at 137 pediatric dialysis centers in 44 countries between 04/2007 and 01/2022.
Results: A total of 2107 peritonitis episodes and 170 relapses were reported in 1179 out of 4289 patients during 5498 patient years follow-up, i.e. an annualized peritonitis rate of 0.38 per patient. Peritonitis rates varied by region and country and were lowest in Asia (0.23) and highest in ANZAC and Eastern Europe (0.81, 0.5). Peritonitis rates steadily decreased in all world regions, from 0.5 per patient year prior to 2012 to 0.28 after 2016. 46% of the episodes were caused by gram-positive, 25% by gram-negative, 4% by fungal and 1% by multiple organisms, whereas 24% were culture negative, without significant variation over time. S.aureus and S.epidermidis were the most frequently isolated organisms and culture negative peritonitis was most common in Turkey and Latin America. Multivariate mixed model analysis identified the following risk factors for peritonitis: younger age (OR 0.97, CI 0.95-0.98, p<0.0001), presence of an ostomy (OR 1.44, CI 1.17-1.76, p=0.0004), impaired cognitive development (OR 1.48, CI 1.22-1.79, p<0.0001), low serum albumin level (0.98, CI 0.97-0.99, p=0.005), and exposure to high dialysate glucose (OR 1.23, CI 1.08-1.41, p=0.03). 15% of the total variability was attributable to center effects and 6% to region-specific effects. Full functional recovery was achieved in 82% of cases, without regional variation.
Conclusions: The prevention and management of peritonitis resulted in their worldwide reduction over the last 15 years, with no significant variation in microbiology. Modifiable risk factors include malnutrition, exposure to high dialysate glucose and center experience.

OP-7 A VARIANT OF ASIC2 MEDIATES SODIUM RETENTION IN NEPHROTIC SYNDROME

Marc Fila[1], Ali Sassi[2], Gaelle Brideau[2], Lydie Cheval[2], Luciana Morla[2], Michel Peuchmaur[1], Georges Deschenes[3], Gilles Crambert[2], Alain Douvcet[1]

[1]Pediatric Nephrology Department Chu Arnaud De Villeneuve - montpellier University, [2]Cordeliers Research Center, Sorbonne University Inserm Laboratory Of Renal Metabolism And Physiology-cnrs Erl 8228 - Paris Paris, [3]Pediatric Nephrology Department - Chu Robert Debré Aphp Paris

Introduction: Idiopathic nephrotic syndrome (INS) is characterized by proteinuria and renal Na retention leading to oedema. This Na retention is usually attributed to epithelial sodium channel (ENaC) activation following plasma aldosterone increase. However, most nephrotic patients show normal aldosterone levels.
Material and methods: We used a corticosteroid-clamped version of the PAN nephrotic rat model (CC-PAN) to attempt to identify the sodium entry pathway in these patients.
Emphasis Type="Bold">Results: CC-PAN rat showed electrogenic and amiloride-sensitive sodium reabsorption in cortical collecting duct similar to that of PAN rats but no increase in expression or activation of ENaC. We isolated from CC-PAN rat kidney a cDNA encoding a variant of acid sensing ion channel 2b (ASIC2b) lacking most of its N-ter intracellular domain. Co-expression of truncated ASIC2b with ASIC2a in X. laevis oocytes induced acid-stimulated sodium currents which were not transient but lasted as long as the stimulus remained. Interestingly, in ASIC2b-null rats generated by CRISPR-Cas9 technology, the injection of PAN did not induce sodium retention. Expression of ASIC2a and of truncated ASIC2b was increased in the ASDN of CC-PAN rats. ASIC2a and truncated-ASIC2b thus constitute channels supporting epithelial sodium reabsorption. Expression of truncated ASIC2b was abolished in albumin deficient rats and in rats treated with ERK kinase inhibitor. ASIC2 mRNA was also detected in kidney biopsies from patients with idiopathic nephrotic syndrome but in any of the patients with other renal diseases. In ASIC2-positive nephrotic patients, ASIC2 was expressed in ASDN.
Conclusions: We have, therefore, identified a novel variant of ASIC2b responsible for the renal Na retention in the pathological context of INS.

OP-8 SHIGA TOXIN TARGETS THE PODOCYTE IN HAEMOLYTIC URAEMIC SYNDROME (HUS) RESULTING IN GLOMERULAR ENDOTHELIAL CELL COMPLEMENT DYSREGULATION

Emily Bowen, Jenny Hurcombe, Fern Barrington, Lindsay Keir, Louise Farmer, Abigail Lay, Eva Larkai, Gavin Welsh, Moin Saleem, Richard Coward

University Of Bristol

Introduction: Haemolytic uraemic syndrome (HUS) is a thrombotic microangiopathy (TMA) that has a predilection to present in the kidney. It is a triad of microangiopathic haemolytic anaemia, thrombocytopenia, and acute kidney injury. In 90% of cases, HUS follows gastroenteritis secondary to infection with Shiga toxin (Stx) producing bacteria such as Escherichia coli. Stx HUS is the leading cause of acute kidney injury in

children with a mortality of 5%. However, the precise pathophysiological mechanisms following Stx infection leading to TMA remain poorly understood. Here we show that the podocyte is a key initiator in Shiga toxin HUS, which could explain why the glomerulus is the prime target of systemic Shiga toxin driven infection.
Material and methods: To demonstrate that the podocyte Shiga toxin receptor (Gb3) is sufficient to trigger the development of HUS, we used conditional gene targeting to engineer human Gb3 expression specifically in the podocytes of adult mice (PodGb3).
Results: PodGb3 mice developed HUS (thrombocytopenia, haemolytic anaemia and uraemia $p<0.05$) at day 10 following intraperitoneal Stx. Renal histology demonstrated glomerular TMA; with intracapillary thrombus formation seen on electron microscopy. Immunofluorescence demonstrated an increase in glomerular fibrinogen deposition and C3b vs. controls. Additionally, glomerular expression of complement regulator Factor H was significantly reduced in PodGb3 mice, rendering them more susceptible to complement attack. Furthermore, C5 inhibition was found to rescue the Shiga toxin HUS phenotype.
Conclusions: Together, these observations provide compelling evidence for the importance of podocyte to glomerular endothelial cell cross-talk within the kidney in the development of Shiga toxin associated HUS and suggest a possible therapeutic role for complement inhibition in patients with this devastating disease.

OP-9 X-LINKED HYPOPHOSPHATEMIA: NOT ONLY A PHOSPHATE/CALCIUM DISORDER BUT ALSO AN INFLAMMATORY DISEASE

Meaux Marie-noelle[1], Alioli Candide[2], Linglart Agnes[3], Lemoine Sandrine[4], Vignot Emmanuelle[4], Bertholet-thomas Aurélia[4], Peyruchaud Olivier[4], Flammier Sacha[4], Machuca-gayet Irma[4], Bacchetta Justine[1]

[1]Centre De Référence Des Maladies Rares Du Calcium Et Du Phosphore, Centre De Référence Des Maladies Rénales Rares Filières Maladies Rares Orkid, Oscar Et Ern Erk-net Et Bond, Chu De Lyon, Bron, France, [2]Inserm, Umr 1033, Faculté De Médecine Lyon Est, Université Claude Bernard Lyon 1, Lyon, France, [3]Paris, [4]Lyon

Introduction: X-linked hypophosphatemia (XLH) is a rare genetic disease caused by a primary excess of FGF23. Treatment consists either in Standard of care (SOC) using phosphore plus active vitamin D, either in biotherapy: burosumab. FGF23 has been associated with inflammation and impaired osteoclastogenesis, however these pathways have not yet been studied in XLH. The aim of our study was to evaluate whether XLH patients display peculiar inflammatory and osteoclastic profile.
Material and methods: We performed a prospective multicenter cross-sectional study analyzing transcript expression of 8 inflammatory markers (Il6, Il8, Il1β, CXCL1, CCL2, CXCR3, Il1R, Il6R) by RT-qPCR on PBMCs (peripheral blood mononuclear cells) extracted from total blood samples from 28 XLH patients (17 children, 11 burosumab) and 19 healthy controls. We also differentiated PBMCs into mature osteoclasts using RANKL/MCSF in the presence/absence of native/active vitamin D, osteoclasts were TRAP-stained and counted at the end of the differentiation process.
Results: Expression of all inflammatory markers (except Il6R) were significantly increased in PBMCs from XLH patients as compared to controls. No differences were observed between the two sub-groups of patients, namely SOC and burosumab. Osteoclastogenesis was significantly impaired in XLH patients as compared to controls, and osteoclasts derived from XLH patients also expressed more inflammatory markers. The pro-inflammatory-like profile at the end of the osteoclastic differentiation process decreased in cells derived from patients receiving burosumab. Burosumab treatment restored the capacity of PBMCs-derived osteoclasts to respond to native vitamin D. The adjunction of active vitamin D in culture during the osteoclastic differentiation decreased the expression of inflammation markers.
Conclusions: We describe for the first time a pro-inflammatory-like profile in XLH. XLH patients have a propensity to develop arterial hypertension and obesity, and since inflammation can worsen these clinical outcomes, we hypothesize that inflammation might play a crucial role in extra-skeletal complications of XLH.

OP-10 URINARY HER2: A NEW BIOMARKER OF PEDIATRIC LUPUS NEPHRITIS ACTIVITY

Patricia Costa-reis[1], Kelly Maurer[2], Michelle A. Petri[3], Jon M. Burnham[2], Kathleen O'neil[4], Marisa Klein-gitelman[5], Emily Von Scheven[6], Laura E. Schanberg[7], Kathleen E. Sullivan[1]

[1]Hospital De Santa Maria, Faculdade De Medicina, Universidade De Lisboa, Lisbon, Portugal, [2]Children's Hospital Of Philadelphia, University Of Pennsylvania, Philadelphia, USA, [3]Johns Hopkins University School Of Medicine, Baltimore, USA, [4]Riley Hospital For Children, Indiana University School Of Medicine, Indianapolis, USA, [5]Children's Hospital Of Chicago, Northwestern Feinberg School Of Medicine, Chicago, USA, [6]University Of California, San Francisco, USA, [7]Duke Children's Health Center, Durham, USA

Introduction: Detecting active lupus nephritis in a background of pre-existing renal damage is challenging, so new biomarkers are much needed to guide clinical practice. Recently, we showed that HER2 (*Human Epidermal Growth Factor Receptor 2*) is highly expressed in the glomeruli and in the tubular compartment of patients with lupus nephritis and in a lupus mouse model (NZM2410), but not in healthy individuals or patients with other mesangioproliferative glomerulonephritides. Furthermore, we showed, in vitro, that HER2 controls mesangial cell proliferation through miR-26a and miR-30b. In this study, we explored the clinical utility of urinary HER2 (uHER2) as a biomarker of lupus nephritis activity.
Material and methods: Prospective, multicenter, study of children and adolescents with biopsy-proven lupus nephritis. Clinical data and urine were collected periodically and uHER2 was quantified by ELISA. The control groups were: healthy individuals, patients with polyarticular juvenile idiopathic arthritis (JIA) and patients submitted to bone marrow transplant with acute kidney injury (AKI). A validation study was performed in an adult cohort.
Results: We studied 771 samples of 113 children and adolescents with lupus nephritis (81% female; median age 15; 41% class IV). uHER2 was significantly increased in patients with active lupus nephritis (renal-SLEDAI≥4; $p=0.006$) and not in children with AKI (n=50), JIA (n=20) or healthy controls (n=40). uHER2 levels were associated with casts, hematuria and new onset proteinuria ($p<0.05$). Furthermore, uHER2 was significantly elevated in clinical visits prior to a renal SLEDAI≥4 ($p<0.05$).
In adults with SLE (n=189; 126 with lupus nephritis) uHER2 was significantly increased when nephritis was active when compared with patients with renal-SLEDAI 0 or healthy controls.
Conclusions: In this prospective study of a large cohort of pediatric patients with lupus nephritis we showed that uHER2 is a biomarker of disease activity. This might be a new useful tool for clinical practice.

OP-11 PREVALENCE OF SHIGATOXIN PRODUCING E. COLI IN HOUSEHOLD CONTACTS OF CHILDREN WITH HEMOLYTIC AND UREMIC SYNDROME

Theresa Kwon[1], Florian Manca Barayre[1], Claire Dossier[1], Veronique Baudouin[1], Aurelie Cointe[2], Stephane Bonacorsi[2], Patricia Mariani[2], Julien Hogan[1]

[1]Pediatric Nephrology - Robert Debré Hospital - Aphp, Paris France, [2]Microbiology - Centre National De Référence Escherichia Coli - Robert Debré Hospital - Aphp, Paris France

Introduction: Typical hemolytic uremic syndrome (HUS) is the main cause of acute kidney injury in children under 5 years and occurs after ingestion of Shigatoxin (Stx) producing Escherichia coli (STEC). Prevalence of STEC carriage is unknown in household contacts in France, and has been scarcely studied in other countries.
Objectives: To assess the prevalence of STEC carriage around HUS cases and study the characteristics associated with STEC carriage in the household.
Material and methods: Stool samples from patients with diarrhea-associated HUS that presented to our institution between 2007 to 2020. Stool samples were analyzed by Stx RT-PCR (Stx-PCR) to establish the frequency in HUS patients and their contacts.
Results: 179 household contacts of 151 HUS patients were analyzed. The prevalence of Stx detection by PCR in household HUS contacts was 22.9%. 40% of HUS patients had a least one STEC positive household contact. The prevalence was higher in siblings (34.3%), especially in those with little age-difference (<5 years) with the HUS case (37.8%). Thus, age difference < 5 years (versus > 5 years of age difference) and siblings (versus parents) were statistically associated with an increased risk of STEC detection in household contacts. Contacts with diarrhea or with positive Stx-PCR were prescribed azithromycin for 3 days. None of the 179 household contacts developed HUS symptoms.
Conclusions: Prevalence of STEC carriage in HUS household contacts is higher than in the general population. This population need to be further studied to assess potential protective factors. Further studies are needed to assess whether azithromycin may prevent the development of HUS in household contacts.

OP-12 SAFETY OF THERAPEUTIC APHERESIS IN CHILDREN AND ADOLESCENTS

Julia Thumfart[1], Anne Schaaf[1], Corina Dorn[2], Claus Peter Schmitt[3], Sebastian Loos[4], Nele Kanzelmeyer[5], Lars Pape[6], Dominik MÜller[1], Lutz T Weber[2], Christina Taylan[2]

[1]Charité Universitätsmedizin Berlin, [2]University Hospital Of Cologne, [3]University Hospital For Paediatric And Adolescent Medicine, Heidelberg, [4]University Medical Centre Hamburg-eppendorf, [5]Hannover Medical School, [6]University Hospital Of Essen

Introduction: Therapeutic apheresis (TA) is based on the principles of either removing dissolved pathogenic substances (e.g. antibodies) from the blood plasma or replacing plasma factors. It expands the therapeutic scope for a variety of diseases. Safety analysis in the pediatric field are scant. The aim of this analysis was to analyze specific complications of TA modalities - plasma exchange (PE) and immunoadsorption (IA) - in children and adolescents.
Material and methods: Children and adolescents (n=298) who had received TA from 2008 to 2018 in five pediatric nephrology centers were analyzed retrospectively. In total, 4.004 treatments (2.287 PE and 1.717 IA) were evaluated.
Results: Indications for TA were mainly nephrological and neurological diseases. The three main indications were antibody-mediated graft rejection (13.4%), hemolytic uremic syndrome mainly with neurological involvement (12.8%), and AB0-incompatible transplantation (11.7%).
Complications developed in 440 of the 4004 sessions (11%), of which one third were nonspecific (nausea, headache). IA was better tolerated than PE. Complications were reported in 9.5% (n=163) of the IA versus 12.1% (277) of the PE sessions (p<0.001). When considering different types of complications, significantly more non-specific/non-allergic events (p=0.02) and allergic reactions occurred in PE sessions (p<0.001). More complications occurred with PE, when using fresh frozen plasma (16.2%; n=145) in comparison to human albumin (14.5%; n=115) (p<0.001).
Conclusions: TA in childhood and adolescence is a safe treatment procedure. IA showed a lower complication rate than PE. Therefore, IA may be preferably provided if the underlying disease pathomechanisms do not require PE.

OP-13 URINARY HSP70 IMPROVES DIAGNOSTIC ACCURACY FOR URINARY TRACT INFECTION IN CHILDREN: UTILISE STUDY

Alev Yilmaz[1], Alberto Caldas Afonso[3], Ipek Akil[4], Bagdagul Aksu[2], Harika Alpay[5], Bahriye Atmis[6], Ozlem Aydog[8], Aysun Karabay Bayazit[7], Meral Torun Bayram[9], Ilmay Bilge[10], Ipek Kaplan Bulut[11], Bahar Buyukkaragoz[12], Elif Comak[13], Belde Kasap Demir[14], Nida Dincel[15], Osman Donmez[16], Mehmet Akif Durmus[17], Hasan Dursun[18], Ruhan Dusunsel[19], Ali Duzova[20], Pelin Ertan[4], Asuman Gedikbasi[21], Nilufer Goknar[22], Sercin Guven[5], Duygu Hacihamdioglu[23], Augustina Jankauskiene[24], Mukaddes Kalyoncu[45], Salih Kavukcu[9], Bahriye Uzun Kenan[12], Nuran Kucuk[25], Bahar Kural[26], Mieczysław Litwin[27], Giovanni Montini[28], William Morello[28], Ahmet Nayir[1], Lukasz Obrycki[27], Beyhan Omer[29], Ebru Misirli Ozdemir[30], Nese Ozkayin[31], Dusan Paripovic[32], Cemile Pehlivanoglu[33], Seha Saygili[34], Susanne Schaefer[35], Ferah Sonmez[36], Yilmaz Tabel[37], Nesrin Tas[20], Mehmet Tasdemir[10], Ana Teixeira[3], Demet Tekcan[8], Sebahat Tulpar[38], Ozde Nisa Turkkan[5], Berfin Uysal[39], Metin Uysalol[40], Daiva Vaiciuniene[24], Sevgi Yavuz[41], Sibel Yel[19], Tarik Yildirim[42], Zeynep Yuruk Yildirim[1], Nurdan Yildiz[5], Selcuk Yuksel[43], Eray Yurtseven[44], Franz Schaefer[35], Rezan Topaloglu[20]

[1]Division Of Pediatric Nephrology, Istanbul Faculty Of Medicine, Istanbul University, [2]Department Of Pediatrics Basic Sciences, Institute Of Child Health, Istanbul University, [3]Division Of Pediatric Nephrology, Centro Materno Infantil Do Norte, Centro Hospitalar Universitário Do Porto, [4]Division Of Pediatric Nephrology, Faculty Of Medicine, Celal Bayar University, [5]Division Of Pediatric Nephrology, Faculty Of Medicine, Marmara University, [6]Division Of Pediatric Nephrology, Erzurum Training And Research Hospital, [7]Division Of Pediatric Nephrology, Faculty Of Medicine, Cukurova University, [8]Division Of Pediatric Nephrology, Faculty Of Medicine, Ondokuz Mayis University, [9]Division Of Pediatric Nephrology, Faculty Of Medicine, Dokuz Eylul University, [10]Division Of Pediatric Nephrology, Department Of Pediatrics, School Of Medicine, Koc University, [11]Division Of Pediatric Nephrology, Faculty Of Medicine, Ege University, [12]Division Of Pediatric Nephrology, Faculty Of Medicine, Gazi University, [13]Division Of Pediatric Nephrology, Faculty Of Medicine, Akdeniz University, [14]Division Of Pediatric Nephrology, Tepecik Training And Research Hospital, University Of Health Sciences, [15]Division Of Pediatric Nephrology, Dr. Behcet Uz Children Diseases Training And Research Hospital, University Of Health Sciences, [16]Division Of Pediatric Nephrology, Faculty Of Medicine, Uludag University, [17]Department Of Medical Microbiology, Istanbul Faculty Of Medicine, Istanbul University, [18]Division Of Pediatric Nephrology, Okmeydani Training And Research Hospital, University Of Health Sciences, [19]Division Of Pediatric Nephrology, Erciyes University, Faculty Of Medicine, [20]Division Of Pediatric Nephrology, Faculty Of Medicine, Hacettepe University, [21]Department Of Rare Diseases, Institute Of Child Health, Istanbul University, [22]Division Of Pediatric Nephrology, Bagcilar Training And Research Hospital, University Of Health Sciences, [23]Division Of Pediatric Nephrology, Faculty Of Medicine, Bahcesehir University, [24]Clinic Of Pediatrics, Institute Of Experimental

And Clinical Medicine, Vilnius University, Vilnius, [25]Division Of Pediatric Nephrology, Kartal Training And Research Hospital, University Of Health Sciences, [26]Department Of Pediatrics, BakirkÖy Sadi Konuk Training And Research Hospital, University Of Health Sciences, [27]Division Of Nephrology, Kidney Transplantation And Hypertension, The Childrens Memorial Health Institute, [28]Pediatric Nephrology, Dialysis And Transplant Unit, Fondazione Irccs Ca' Granda - Ospedale Maggiore Policlinico, [29]Department Of Biochemistry, Istanbul Faculty Of Medicine, Istanbul University, [30]Department Of Pediatrics, Okmeydani Training And Research Hospital, University Of Health Sciences, [31]Division Of Pediatric Nephrology, Faculty Of Medicine, Trakya University, [32]Division Of Pediatric Nephrology, University Childrens Hospital, [33]Division Of Pediatric Nephrology, Umraniye Training And Research Hospital, University Of Health Sciences, [34]Division Of Pediatric Nephrology, Cerrahpasa Faculty Of Medicine, Istanbul University-cerrahpasa, [35]Division Of Pediatric Nephrology, Center For Pediatrics And Adolescent Medicine, Heidelberg University, [36]Division Of Pediatric Nephrology, Faculty Of Medicine, Adnan Menderes University, [37]Division Of Pediatric Nephrology, Faculty Of Medicine, Inonu University, [38]Division Of Pediatric Nephrology, Bakirkoy Dr. Sadi Konuk Training And Research Hospital, University Of Health Sciences, [39]Division Of Pediatric Nephrology, Dortcelik Childrens Hospital, [40]Division Of Pediatric Emergency, Istanbul Faculty Of Medicine, Istanbul University, [41]Division Of Pediatric Nephrology, Kanuni Sultan Suleyman Research And Training Hospital, University Of Health Sciences, [42]Department Of Pediatrics, Kanuni Sultan Suleyman Research And Training Hospital, University Of Health Sciences, [43]Division Of Pediatric Nephrology, Faculty Of Medicine, Pamukkale University, [44]Department Of Biostatistics, Istanbul Faculty Of Medicine, Istanbul University, [45]Division Of Pediatric Nephrology, Faculty Of Medicine, Karadeniz Technic University

Introduction: The accuracy of conventional urinalysis in diagnosing urinary tract infection (UTI) in children is limited, leading to unnecessary antibiotic exposure in a large fraction of patients. Urinary Heat Shock Protein 70 (uHSP70) is a novel marker of acute urinary tract inflammation. We explored the added value of uHSP70 in discriminating UTI from other infections and conditions confused with UTI.
Material and methods: 802 children from 37 pediatric centers in seven countries participated in the study. Patients diagnosed with UTI (n=191), non-UTI infections (n=178), contaminated urine samples (n=50), asymptomatic bacteriuria (n=26), and healthy controls (n=75) were enrolled. Urine and serum levels of HSP70 were measured at presentation in all patients and after resolution of the infection in patients with confirmed UTI.
Results: Urinary (u)HSP70 was selectively elevated in children with UTI as compared to all other conditions ($p<0.0001$). uHSP70 predicted UTI with 88.5% sensitivity and 82.2% specificity (AUC=0.934). Among the 265 patients with suspected UTI, the uHSP70 >48 ng/mL criterion identified the 172 children with subsequently confirmed UTI with 90.1% sensitivity and 82.4% specificity (AUC=0.862), exceeding the individual diagnostic accuracy of leukocyturia, nitrite and leukocyte esterase positivity. uHSP70 had completely normalized by the end of antibiotic therapy in the UTI patients. Serum HSP70 was not predictive.
Conclusions: Urine HSP70 is a novel non-invasive marker of UTI that improves the diagnostic accuracy of conventional urinalysis. We estimate that rapid urine HSP70 screening could spare empiric antibiotic administration in up to 80% of children with suspected UTI.

OP-14 A GENOME-WIDE ASSOCIATION STUDY OF THE AETIOLOGY OF SOLITARY FUNCTIONING KIDNEY

Sander Groen In T Woud[1], Carlo Maj[2], Nel Roeleveld[1], Wout Feitz[3], Michiel Schreuder[4], Loes Van Der Zanden[1]

[1]Radboud University Medical Center, Department For Health Evidence, Radboud Institute For Health Sciences, Nijmegen, The Netherlands, [2]Institute Of Genomic Statistics And Bioinformatics, University Hospital Bonn, Medical Faculty University Of Bonn, Bonn, Germany, [3]Radboudumc Amalia Children's Hospital, Department Of Urology, Radboud Institute For Molecular Life Sciences, Nijmegen, The Netherlands, [4]Radboudumc Amalia Children's Hospital, Department Of Pediatric Nephrology, Radboud Institute For Molecular Life Sciences, Nijmegen, The Netherlands

Introduction: Solitary functioning kidney (SFK) is a relatively common condition predisposing to kidney injury. Although several monogenic causes were identified, most cases cannot yet be explained. Therefore, its aetiology is likely multifactorial and several environmental risk factors have been reported, while common genetic variation may also contribute to genetic susceptibility. To identify common variants involved in the aetiology of SFK, we performed a genome-wide association study (GWAS).
Material and methods: GWAS was performed using 702 patients with congenital SFK from the SOFIA study and 660 geographically matched controls from the AGORA data- and biobank. Phenotypic subgroups were created a priori. Association tests were performed in PLINK 2.0 on an imputed dataset under the assumption of an additive model. Principal component analyses were used to exclude ancestral outliers and control for population stratification. Results were analysed on individual SNP level with a fixed genome-wide significance threshold of $5 \times 10\text{-}8$ and on gene level using MAGMA with a significance threshold of 0.05/number of genes tested.
Results: In both the overall and subgroup analyses, no SNP reached the genome-wide significance threshold, with the top SNP at $6.3 \times 10\text{-}7$ in overall analyses and $9.3 \times 10\text{-}8$ in the MCDK subgroup. Analyses on gene level did not yield results above the significance threshold either, but candidate genes were identified that will be investigated further and results will be shared during the conference.
Conclusions: Our GWAS on the aetiology of SFK did not identify variants reaching genome-wide significance, most likely because of a lack of power. However, some of the identified variants may contribute to the genetic understanding of SFK and will be topic of further study.

OP-15 ASSESSMENT OF CAKUT PREGNANCY OUTCOME USING MULTIPLE ULTRASOUND FEATURES

Bénédicte Buffin-meyer[2], Camille Fedou[2], Guylène Feuillet[2], Jacqueline Aziza[3], Jean-sébastien Saulnier-blache[2], Julie Klein[2], Joost P Schanstra[2], Stéphane Decramer[1]

[1]Toulouse Hospital/toulouse University, [2]Inserm-i2mc/toulouse University, [3]Crct/toulouse Hospital

Introduction: Anomalies of the kidney and the urinary tract (CAKUT) have a wide spectrum of outcomes ranging from normal postnatal kidney function to fetal death. Although often already discovered in utero, the ultrasound workup does not allow an accurate assessment of outcome. Our study aimed to clarify the relationship between ultrasound data and outcome.
Material and methods: For a total of 136 fetuses with bilateral CAKUT (except bilateral agenesis) and a 2-year postnatal follow-up, we collected data on amniotic fluid (AF) volume, number of functional kidneys, and presence of renal dysplasia or cysts using a standardized ultrasound-based observation protocol. Cramers V was used to measure the strength of 2-to-2 dependence of changes. Logistic regression analysis was conducted to study the association of ultrasound features with postnatal renal survival. The AF proteome related to dysplasia or cysts was characterized by LC–MS/MS using 22 healthy fetuses and a subset of 36 CAKUT fetuses.

Results: Fourty-two CAKUT patients had oligohydramnios whereas 12 displayed anyhydramnios; 9 had a single kidney; unilateral renal dysplasia was found in 24 fetuses while bilateral dysplasia occurred in 65; cysts were detected in one or both kidneys in 22 and 76 cases, respectively. Dysplasia was significantly associated with both AF volume (Cramer's V:0.24) and the presence of cysts (Cramer's V:0.44). Reduced AF volume (OR=132 [95%CI:21-2666]), dysplasia occurrence (OR=31 [95%CI:3-785]) and presence of cysts (OR=16 [95%CI:1-460]) were identified as independent risk factors of poor prognosis. Combination of the 3 features yielded an area under the receiver operator characteristic curve of 0.92 for prediction of renal postnatal outcome in leave-one-out cross validation. Functional enrichment analysis of the AF proteome led to association of dysplasia with the term extracellular matrix organization and cysts with immunity.
Conclusions: A more detailed exploitation of the antenatal ultrasound-based changes could improve prenatal clinical advice in CAKUT pregnancies.

OP-16 DISPARITIES IN TREATMENT AND OUTCOME OF KIDNEY REPLACEMENT THERAPY IN CHILDREN WITH COMORBIDITIES: AN ESPN/ERA-EDTA REGISTRY STUDY

Raphael Schild[1], Simeon Dupont[1], Jérôme Harambat[2], Enrico Vidal[3], AyŞe Balat[4], Csaba Bereczki[5], Beata Bieniaś[6], Per BrandstrÖm[7], Francoise Boux[8], Silvia Consolo[9], Ivana Gojkovic[10], Jaap W. Groothoff[11], Kristine Hommel[12], Holger Hubmann[13], Fiona E. M. Braddon[14], Tatiana E. Pankratenko[15], Fotios Papachristou[16], Lucy A. Plumb[17], Ludmila Podracka[18], Sylwester Prokurat[19], Anna Bjerre[20], Carolina Cordinhã[21], Juuso Tainio[22], Enkelejda Shkurti[23], Giuseppina Sparta[24], Karel Vondrak[25], Kitty J. Jager[26], Marjolein Bonthuis[26]

[1]University Childrens Hospital, University Medical Center Hamburg, [2]Department Of Pediatrics, Bordeaux University Hospital, [3]Division Of Pediatrics, Department Of Medicine, University Of Udine, [4]Department Of Pediatric Nephrology, Gaziantep University Medical Faculty, [5]Department Of Pediatrics, University Of Szeged, [6]Department Of Paediatric Nephrology, Medical University Of Lublin, [7]The Queen Silvia Children's Hospital, The Sahlgrenska Academy At The University Of Gothenburg, [8]Department Of Pediatrics, Rouen University Hospital, [9]Dialysis And Transplant Unit, Fondazione Ircss Ca' Grande Ospedale Maggiore Policlinico, [10]University Children's Hospital, University Of Belgrade, [11]Emma Children's Hospital, Amsterdam University Medical Center, [12]Department Of Medicine, Holbæk Hospital, [13]Department Of Pediatrics, Medical University Graz, [14]Uk Renal Registry, Bristol, [15]Moscow Regional Research And Clinical Institute Named After M.f. Vladimirskiy, Moscow, [16]1st Department Of Pediatrics, 37782 Aristotle University Of Thessaloniki, [17]Population Health Sciences, University Of Bristol Medical School, [18]National Institute Of Children's Health, Comenius University, Bratislava, [19]Department Of Nephrology & Kidney Transplantation, The Children's Memorial Health Institute, Warsaw, [20]Department Of Specialised Medicine And Transplantation, Oslo University Hospital, [21]Hospital Pediátrico - Centro Hospitalar Universitário De Coimbra, [22]New Children's Hospital, University Of Helsinki, [23]Univeristy Of Medicine Of Tirana, Public Health, Tirana, [24]Pediatric Nephrology Unit, University Children's Hospital Zurich, [25]Department Of Pediatric Nephrology, University Hospital Motol, Prague, [26]Espn/era-edta Registry, Amsterdam Umc, University Of Amsterdam, Department Of Medical Informatics, Amsterdam Public Health Research Institute

Introduction: Data on extra-renal comorbidities in children on kidney replacement therapy (KRT) is scarce. Considering its high relevance for prognosis and clinical decision-making, this study aims to analyse the prevalence and implications of comorbidities in European children on KRT.
Material and methods: We included data from 4127 patients aged <20 years when commencing KRT from 2007 to 2017 from 22 European countries included in the ESPN/ERA-EDTA Registry. Comorbidities were registered at the start of KRT. Differences in access to kidney transplantation (KT), patient and graft survival were estimated using Cox proportional hazard regression.
Results: At least one comorbidity was present in one third (33%) of children commencing KRT, and acquired cardiovascular diseases occurred most frequently. The comorbidity prevalence has steadily increased by 5% per year since 2007. Comorbidities were most frequent in patients from high-income countries (43% vs. 24% in low-income and 32.9 in middle-income countries). Patients with comorbidities had a lower access to transplantation (aHR 0.67, 95% CI: 0.61 - 0.74), and a higher risk of death (aHR 1.79; 95% CI: 1.38–2.32). The increased risk of death was only seen in dialysis patients (aHR 1.60; 95% CI: 1.21-2.13), and not after kidney transplantation. For both outcomes, the impact of comorbidities was stronger in low-income countries. However, once transplanted, 5-year graft survival was not affected by the presence of comorbidities (aHR for graft failure: 1.18, 95% CI: 0.84-1.65).
Conclusions: Extra-renal diseases have become more frequent in children and adolescents on KRT and reduce their access to kidney transplantation as well as survival, especially when remaining on dialysis. Kidney transplantation should be considered as treatment of choice in all pediatric KRT patients and efforts should be made to identify modifiable barriers to KT for children with comorbidities.

OP-17 MANAGEMENT OF DIALYSIS ASSOCIATED PERITONITIS: DOES GUIDELINE ADHERENCE PAY OFF?

Dagmara Borzych-duzalka[1], Karel Vondrak[2], Telma Francisco[3], Charlotte Samaille[4], Maria Szczepanska[5], Julia Thumfart[6], Renata Vitkevic[7], Elena Codina[8], Divina Kruscic[9], Klaus Arbeiter[1], Hee Gung Kang[10], Zenaida L Antonio[11], Bradley A Warady[12], Franz Schaefer[13]

[1]Medical University Of Gdansk, Gdansk, Poland, [2]University Hospital Motol, Prague, Czech Republic, [3]Centro Hospitalar E Universitário De Lisboa Central, Lisbon, Portugal, [4]Service De Néphrologie Pédiatrique, Hôpital Jeanne De Flandre, Lille, France., [5]Medical University Of Silesia, Zabrze, Poland, [6]Virchow-klinikum, Charite, Berlin, Germany, [7]Vilnius University Hospital, Vilnius, Lithuania, [8]Hospital Sant Joan De Deu, Barcelona, Spain, [9]University Childrens Hospital, Belgrade, Serbia, [10]Seoul National University Childrens Hospital, Seoul, South Korea., [11]National Kidney And Transplant Institute, Quezon City, Philippines, [12]Childrens Mercy Hospital, Kansas City, USA, [13]Center For Pediatrics And Adolescent Medicine, Heidelberg University, Heidelberg, Germany.

Introduction: Peritonitis is a significant risk factor for technique failure in children receiving chronic peritoneal dialysis (PD). The aim of the study was describe management and outcome peritonitis experienced by patients followed in the International Pediatric Peritoneal Dialysis Network (IPPN) Registry.
Material and methods: Review of peritonitis episodes submitted to the IPPN registry (www.pedpd.org) between 04/2007 and 01/2022.
Results: A total of 2107 peritonitis episodes were reported to the IPDN registry between 2007 and 2022. Within 3 days of empiric antibiotic therapy 76% of patients were asymptomatic with clear dialysis effluent. The clinical response rate was significantly ($p<0.0001$) better in gram positive (97%) and culture negative (95%) as compared to gram negative episodes (77%). Response rates were identical (89%) in patients receiving glycopeptide/ceftazidime and those treated with cefazoline/

ceftazidime. The resistance rate of gram-positive bacteria to methicillin was high with significant regional variability ranging from 19% in Western Europe to 49% in Central Europe, while glycopeptide sensitivity was 92-100%. Gram negative sensitivity to ceftazidime ranged from 59% in Latin America to 100% in North America, aminoglycoside sensitivity was 76-100%. The overall sensitivity of causative organisms to cefepime was 71-100%. 79% of peritonitis episodes were treated empirically according to the pediatric ISPD guidelines: 37% of episodes were treated empirically with glycopeptide/ceftazidime, 28% with cefazoline/ceftazidime, 7% with glycopeptide/cefepime, 4% with glycopeptide/ aminoglycoside, 3% with cefazoline/cefepime and 4 episodes (<1%) with cefepime monotherapy. The rate of adherence to the empiric therapy treatment schedule recommended by ISPD varied regionally, ranging from 58% in Asia to 83% in Western Europe (p<0.0001). Permanent or temporary discontinuation of PD was significantly more common in patients treated empirically by other protocols than recommended in the ISPD guidelines (23% v. 16%, p<0.001), without significant regional variation.

Conclusions: Treatment of PD-associated peritonitis according to international consensus guidelines is results in superior clinical outcomes in children.

OP-18 TRANSCRIPTOMIC AND PROTEOMIC PROFILES OF CHRONIC KIDNEY DISEASE IN PEDIATRIC ARTERIOLAR TISSUES: WHICH PATHWAYS ARE INVOLVED IN PRO-CALCIFYING PROCESSES?

Julie Bernardor[1], Maria Bartosova[1], Conghui Zhan[1], Iva Marinovic[1], Betti Schaefer[1], Rebecca Herzog[2], Anette Melk[3], Guenter Klaus[4], Klaus Arbeiter[5], Rimante Cerkauskiene[6], Dorota Drozdz[7], Delphine Farlay[8], Arianeb Mehrabi[9], Jun Oh[10], Klaus Kratochwill[2], Claus Peter Schmitt[1]

[1]Division Of Pediatric Nephrology, Center For Pediatric And Adolescent Medicine, University Of Heidelberg, 69120 Heidelberg, Germany, [2]Christian Doppler Laboratory For Molecular Stress Research In Peritoneal Dialysis, Division Of Pediatric Nephrology And Gastroenterology, Department Of Pediatrics And Adolescent Medicine, Comprehensive Center For Pediatrics, Medical University Of Vienna, Vienna, Austria, [3]Department Of Pediatric Nephrology, Hepatology And Metabolic Diseases, Childrens Hospital, Hannover Medical School, Germany, [4]Kfh Pediatric Kidney Center, Marburg, Germany, [5]Department Of Pediatrics And Adolescent Medicine, Medical University Vienna, Austria, [6]Pediatric Center, Vilnius University, Lithuania, [7]Jagiellonian University Medical College, Krakow, Poland, [8]Inserm Umr S1033 Research Unit, Lyon, France, [9]Department Of General, Visceral And Transplantation Surgery, University Hospital Heidelberg, Heidelberg, Germany, [10]Department Of Pediatric Nephrology, University Childrens Medical Clinic, University Medical Center Hamburg-eppendorf, Germany

Introduction: Pediatric patients with chronic kidney disease (CKD) develop major atherosclerosis and vascular calcification until early adulthood; underlying mechanisms are partially understood only. Early mechanisms of vascular calcification in children with CKD devoid of aging and life-style related damage should be particularly sensitive and should allow identification of novel therapeutic targets.

Material and methods: Gene set enrichment and Ingenuity pathway analysis were performed on multi-omics data sets obtained from microdissected omental arterioles from children with normal renal function and with CKD5 (n=7/group; age 7.3 ± 3/6.8 ± 3 years). Based on extensive literature review, we established a vascular calcification pathway library comprising 442 biological processes/molecular functions and extracted linked genes from Gene Ontology database. Histomorphometry was performed in 90 non-CKD and 100 CKD5 children, pathways identified by omics were validated in independent patient cohorts (n=30). Calcium deposits were assessed by Von Kossa staining and electronic microscopy.

Results: Arteriolar lumen to vessel ratio was significantly reduced in CKD5 vs. age matched controls (0.75 (IQR 0.1) vs. 0.56 (0.1), p<0.001). Von Kossa staining yielded significant arteriolar calcification in CKD5, currently characterized by electron microscopy. Vascular calcification pathway analyses identified 21/442 pathways significantly regulated on arteriolar transcriptome (p<0.05; FDR=0.25) level related to complement activation, Wnt signaling, extracellular matrix organization, endoplasmic reticulum stress, apoptosis, autophagy and ossification. 9/442 calcification related arteriolar proteome pathways were enriched and included complement activation, extracellular matrix organization, fatty acid metabolism (all p<0.0001), calcium ion binding (p<0.005), apoptosis (p<0.05); Fibronectin-1 is a key hub-gene. In independent age-matched cohorts, CKD5 children had shorter endothelial telomeres and less endothelial methylated histone 3. Endothelial complement system was activated, C1q and terminal complement complex deposition increased.

Conclusions: Vascular calcifications are already present in young children with CKD5. Arteriolar multi-omics analyses and independent protein validation identified specific molecular pathomechanisms; of which several represent potential therapeutic targets.

OP-19 FGF23, SYSTEMIC INFLAMMATION, MUSCLE AND PROTEIN ENERGY WASTING IN CHILDREN WITH CHRONIC KIDNEY DISEASE

Vasiliki Karava[1], John Dotis[1], Antonia Kondou[1], Athanasios Christoforidis[2], Anna Taparkou[3], Evangelia Farmaki[3], Nikoleta Printza[1]

[1]Pediatric Nephrology Unit, 1st Department Of Pediatrics, Hippokratio General Hospital, Aristotle University Of Thessaloniki, Greece, [2]Pediatric Endocrinology Unit, 1st Department Of Pediatrics, Hippokratio General Hospital, Aristotle University Of Thessaloniki, Greece, [3]Pediatric Immunology And Rheumatology Referral Center, 1st Department Of Pediatrics, Hippokratio General Hospital, Aristotle University Of Thessaloniki, Greece

Introduction: This cross-sectional study investigates the association of fibroblast growth-factor 23 (FGF23) with systemic inflammation, muscle and protein energy wasting (PEW) in children with chronic kidney disease (CKD).

Material and methods: Serum calcium, phosphorus, 25(OH)D, intact parathormone, c-terminal FGF23, a-Klotho, albumin and IL-6 were measured in 53 patients from 5 to 19 years old with GFR<60 ml/min/1,73m^2. PEW was defined as muscle wasting [lean tissue index adjusted to height age (LTI HA) z-score <-1.65 SD), measured by bioimpedance analysis spectroscopy, and at least two of the following: poor growth [height z-score <-1.88 SD], questionnaire based reduced appetite, serum albumin ≤ 3.8 g/dl.

Results: LnFGF23 but not lnKlotho was correlated to LTI HA z-score (rs=-0.320, p=0.021) and lnIL-6 (rs=0.360, p=0.009) after adjustment for CKD stage. We performed logistic regression analysis of PEW criteria, including lnFGF23, lnIL-6, CKD stage and bone mineral parameters as covariates. LnFGF23 was significantly associated with muscle wasting (18 patients) (OR 2.763, 95% CI 1.246-6.124), while CKD stage and lnIL6 were significantly associated with poor growth (14 patients) (OR 9.178, 95% CI 1.346-62.582) and reduced albumin (13 patients) (OR 2.257, 95% CI 1.170-4.352) respectively. In backward logistic regression analysis, lnFGF23 was associated with PEW (8 patients) after adjustment for bone mineral parameters, lnIL-6 and CKD stage (OR 4.910, 95% CI 1.957-12.320).

Conclusions: Increased FGF23 may be associated with the muscle and PEW process in pediatric moderate and advanced CKD, independently of

Klotho. FGF23 association with systemic inflammatory may only in part explain our findings.

OP-20 THE ITALIAN REGISTRY OF THROMBOSIS IN CHILDREN (RITI): POTENTIAL AND PERSPECTIVES.

Davide Meneghesso[1], Alessia Cicogna[1], Chiara Guariento[1], Jacopo Norberto Pin[1], Clarissa Tona[1], Maria Federica Pelizza[1], Andrea Francavilla[3], Giulia Lorenzoni[3], Matteo Martinato[3], Rossana Bagna[4], Laura Ilardi[5], Donatella Lasagni[6], Matteo Luciani[7], Margherita Nosadini[2], Anna Rosati[6], Paola Saracco[4], Agnese Suppiej[8], Maria Caterina Piutti[9], Dario Gregori[3], Stefano Sartori[2], Paolo Simioni[10]

[1]Pediatric Nephrology - University Of Padua, [2]Pediatric Neurology - University Of Padua, [3]Department Of Cardio-thoraco-vascular Sciences And Public Health, University Of Padua, [4]University Of Turin, [5]Neonatology, Milan Niguarda Hospital, [6]Meyer, Pediatric Hospital, [7]Pediatric Onco-hematology, Pediatric Hospital Bambino Gesù, Rome, [8]Pediatric Neurology, University Of Ferrara, [9]Pediatric Onco-hematology, University Of Padua, [10]Thrombotic And Haemorrhagic Diseases Unit, Department Of Medicine - Dimed, University Of Padua

Introduction: Patients with kidney disease (KD) may present an increased risk of thrombotic events due to renal failure, vascular catheterization and medical therapies. In the last decades national and international registries have been created to implement the knowledge around thrombosis in children. The Italian registry of thrombosis in children (R.I.T.I.) was founded in 2007 with the contribution of hematologists, pediatricians, pediatric neurologists and emergency care physicians. Its aim is to collect information nationwide about pediatric and neonatal thrombosis. Starting in 2017, an observational study was designed with the following objectives: i) multiparametric definition of thrombosis in children; ii) development of diagnostic and therapeutic protocols; iii) implementation of a multidisciplinary network (including pediatric nephrologists and oncohemalogists).
Material and methods: Since 2017 clinical, instrumental and laboratoristic findings of pediatric patients affected by thrombosis were systematically collected together with the characteristics of the thrombotic events and follow up data.
Results: To date, 49 italian hospitals and 121 physicians are contributing to the registry. 727 patients have been included(57%males). Of them, 502(69%) had cerebral thrombosis (68%arterial) while 225(31%) had systemic thrombosis (85%venous). The majority of the included patients(51%) had an event at pre-school age. In the evaluated time period, recurrencies were most frequently observed in pediatric patients(9%) than in newborns(3.5%). 66(9%) patients included in the registry were affected by KD. Of them, 31% had cerebral thrombosis and 69% systemic thrombosis. The nephrologic diagnosis was: AKI(n=22); nephrotic syndrome(n=5); CKD(n=5); dialysis(n=23); miscellaneous(n=18).
Conclusions: The progressive implementation of R.I.T.I. from 2007 to date makes it one of the largest registries about children thrombosis internationally. Further collaborations with national and international nephrologic centres are warrented in order to enlarge the knowledge around pediatric thrombosis in patients affected by KD.

OP-22 DO SPHINGOMYELIN PHOSPHODIESTERASE ACID-LIKE 3B (SMPDL-3B) LEVELS IN KIDNEY BIOPSY SPECIMENS PREDICT RESPONSE TO IMMUNOSUPPRESSIVE THERAPY IN CHILDREN WITH NEPHROTIC SYNDROME?

Muhammet Kaya[1], İlknur GİrİŞgen[1], Nagihan YalÇin[3], TÜlay BecerİR[1], Hande Şenol[4], GÜlsÜn GÜlten[3], SelÇuk YÜksel[2]

[1]Department Of Pediatric Nephrology, Faculty Of Medicine, Pamukkale University, [2]Department Of Pediatric Nephrology And Rheumatology, Faculty Of Medicine, Pamukkale University, [3]Department Of Pathology, Pamukkale University, Faculty Of Medicine, [4]Department Of Biostatistics, Pamukkale University, Faculty Of Medicine

Introduction: Idiopathic nephrotic syndrome (INS) is a common glomerular disease in children. Some patients who develop steroid-resistant nephrotic syndrome (SRNS) are also resistant to steroids and other immunosuppressants and can progress to chronic kidney disease (CKD). The treatment of SRNS remains a challenge for pediatric nephrologists**.** Sphingomyelin phosphodiesterase acid-like 3b (SMPDL-3b) contributes to the regulation of the podocyte cytoskeleton by enabling ceramide synthesis in the cell membrane. Recent studies suggest that rituximab prevents proteinuria independent of B-cell depletion by inhibiting the downregulation of SMPDL-3b expression or binding to SMPDL-3b. However, the question remains whether the low amount of SMPDL-3b for rituximab to bind in treatment-resistant NS patients with advanced podocyte injury could be the cause of treatment resistance. The present study was conducted to evaluate whether SMPDL-3b level in pre-treatment kidney biopsy is predictive of the clinical effectiveness of immunosuppressive drugs, especially rituximab, in patients with SRNS and FSGS.
Material and methods: Kidney biopsy specimens from 48 patients diagnosed with INS were analyzed by immunohistochemical staining with anti-SMPDL3B and real-time polymerase chain reaction (PCR) for SMPDL-3b mRNA expression. The results were compared according to treatment response. Real time PCR evaluation was done with $2^{-\Delta\Delta CT}$ method. Fold change (FC)>1.5 was considered stastically significant.
Results: *SMPDL-3b* expression levels were higher in patients responding to all immunosuppressive treatments, including rituximab and steroids, but it was not considered significant because the fold change (FC) was less than 1.5. *SMPDL-3b* expression was 1.788 times greater in patients who achieved remission with treatment compared to patients with CKD. When SMPDL-3b expression was compared between patients with SRNS who achieved remission and those who developed CKD, expression was 2.18 times higher and significant in remitted patients (FC>1.5).
Conclusions: In our study, we sought to determine whether low SMPDL-3b levels in pre-treatment kidney biopsies were associated with resistance to immunosuppressive therapies, especially rituximab. We showed that SMPDL-3b mRNA expression and anti-SMPDL3B staining did not differ significantly between patient groups with different responses to immunosuppressive therapies. Our results indicate that SMPDL-3b-expression is higher in patients with complete remission, both among all patients and within the SRNS subgroup, when compared with patients who progressed to CKD. These results suggest that SMPDL-3b may actually be an indicator of disease progression rather than a marker predicting response to a particular immunosuppressive agent. Our study is the first in the literature to examine SMPDL-3b levels in human kidney biopsy specimens using both immunohistochemical and molecular PCR methods.

OP-23 BASELINE FACTORS ASSOCIATED WITH THE RISK OF RELAPSE FOLLOWING ANTI-CD20 TREATMENT IN CHILDREN WITH FREQUENTLY-RELAPSING/STEROID-DEPENDENT NEPHROTIC SYNDROME

Manuela Colucci[1], Andrea Angeletti[2], Armando Di Donato[2], Michela Cioni[2], Gianluca Caridi[2], Francesca Lugani[2], Pietro Ravani[3], Paolo Cravedi[4], Francesco Emma[1], Gian Marco Ghiggeri[2], Marina Vivarelli[1]

[1]Ospedale Pediatrico Bambino Gesù - Irccs, Rome, Italy, [2]Istituto Giannina Gaslini Irccs, Genoa, Italy, [3]University Of Calgary, Calgary, Alberta, Canada, [4]Icahn School Of Medicine At Mount Sinai, New York, New York

Introduction: B-cell depleting anti-CD20 monoclonal antibodies have prolonged efficacy in children with frequently-relapsing/steroid-dependent nephrotic syndrome (FR/SDNS). However, response to this therapy is variable and treatment-related side effects can occur. We studied the association of several clinical characteristics and laboratory parameters at the baseline with the risk of relapse following anti-CD20 treatment.
Material and methods: In this retrospective study, we included 102 FR/SDNS pediatric patients treated with anti-CD20 monoclonal antibodies (rituximab or ofatumumab). Time to first relapse during a 24-month follow-up and tapering of concomitant immunosuppression was registered for each patient. We used Cox regression to estimate the association of age, sex, previous immunosuppressive treatment (anti-CD20, prednisone, mycophenolate mofetil, calcineurin inhibitors), duration of concomitant immunosuppression (prednisone, mycophenolate mofetil, calcineurin inhibitors), circulating B-cell subset levels (total CD19+, transitional, mature-naïve and memory B cells) at baseline and time to relapse.
Results: Univariate analysis showed that age > 9 years at time of anti-CD20 infusion (p=0.012), previous treatment with anti-CD20 (p=0.056) and maintenance immunosuppression with mycophenolate mofetil (p=0.008) were protective against relapse. In contrast, among all the evaluated B-cell subsets at baseline, only high circulating levels of memory B cells (p<0.001) were significantly associated with relapse. By multivariate analysis, age > 9 years (p=0.017), maintenance of mycophenolate mofetil treatment (p=0.045) and levels of memory B cells at baseline (p=0.003) retained their significant association with time to relapse, whilst previous treatment with anti-CD20 did not (p=0.906).
Conclusions: Younger age and high circulating levels of memory B cells at time of anti-CD20 infusion are associated with a higher risk of relapse following anti-CD20 administration. The maintenance of mycophenolate mofetil treatment, more than other immunosuppressors, may prolong remission following anti-CD20 infusion.

OP-25 INSULIN SENSITIVITY IN CHILDREN WITH AUTOSOMAL DOMINANT POLYCYSTIC KIDNEY DISEASE

Dachy Angélique[1], De Rechter Stéphanie[2], Breysem Luc[3], Vennekens Rudi[4], Mathieu Chantal[5], Casteels Kristina[6], Van Hoorenbeeck Kim[7], Jouret FranÇois[8], Mekahli Djalila[2]

[1]Ku Leuven, Department Of Development And Regeneration, Pkd Research Group, Laboratory Of Pediatric Nephrology, Ku Leuven, Leuven, Belgium, [2]Department Of Pediatric Nephrology, University Hospitals Leuven, Leuven, Belgium, [3]Department Of Pediatric Radiology, University Hospitals Leuven, Leuven, Belgium, [4]Laboratory Of Ion Channel Research, Department Of Cellular And Molecular Medicine, Vib Center For Brain And Disease Research, Ku Leuven, Leuven, Belgium, [5]Department Of Endocrinology, University Hospitals Leuven, Leuven, Belgium, [6]Department Of Pediatrics, University Hospitals Leuven, Leuven, Belgium, [7]Department Of Pediatrics, University Hospital Of Antwerp, Antwerp, Belgium, [8]Division Of Nephrology, Department Of Internal Medicine, Uliège Academic Hospital, Liège, Belgiu

Introduction: Autosomal Dominant Polycystic Kidney Disease (ADPKD) is the most common inherited kidney disorder. Defective glucose metabolism was identified as a key feature in ADPKD, and several 'metabolic' approaches are currently under evaluation in adults with ADPKD. Whether this defective glucose metabolism could be an early primary event and a potential therapeutic option in the early disease stages is still unknown. In this study, we evaluated the insulin sensitivity profile in genotyped children with ADPKD.
Material and methods: We performed a cross-sectional study to evaluate the insulin sensitivity profile in a genotyped cohort of ADPKD children (<19 years) with preserved renal function (eGFR>60 ml/min/1.73m^2). Overweight/obese children were respectively defined as body mass index (BMI) 25-30 and >30 kg/m^2. The Homeostasis Model Assessment Index (HOMA-IR) was calculated: fasting insulin (μIU/ml − 1) x fasting glucose (mmol/l− 1)/22.5. The Quantitative Insulin Sensitivity Check Index (QUICKI) was calculated: 1/(log (fasting insulin μU/mL) + log(fasting glucose mg/dl)).
Results: 37 ADPKD patients (22 boys) were included with a mean (+/-SD) age at diagnosis of 10.3 ±4.2 years. 36 patients had PKD1 mutation (one GANAB mutation). Median BMI was 16.8 ±4.3 kg/m^2. Median serum fasting glucose: 86.0 ± 9.3 mg/dl, median fasting insulin: 6.1 ±7.2 μU/ml and median serum C-peptide: 0.4 ±0.3 nmol/l. Median HOMA-IR was 1.4 ±1.7 and median QUICKI was 0.4 ±0.1. 16 patients presented a HOMA-IR>1.6 and 6 normal-weight children had a HOMA-IR >2.3. No patient displayed glucosuria. An oral glucose tolerance test was performed on 5 overweight patients, 4 of them showed insulin resistance and were treated with metformin.
Conclusions: Even with normal BMI, ADPKD children displayed high index of insulin resistance. Further clinical studies are needed to determine whether ADPKD could be an additional risk factor for insulin resistance.

OP-26 UNATTENDED AUTOMATED OFFICE BLOOD PRESSURE MEASUREMENT IN CHILDREN

Tomas Seeman[4], Krystof Stanek[1], Jakub Slizek[1], Jan Filipovsky[2], Janusz Feber[3]

[1]Charles University Prague, 2nd School Of Medicine, Czech Republic, [2]Department Of Internal Medicine Ii, Charles University, Medical Faculty In Pilsen, Czech Republic, [3]Division Of Nephrology, Department Of Pediatrics, Children's Hospital Of Eastern Ontario, University Of Ottawa, Ottawa, Canada, [4]Dpt. Of Pediatrics, Ludwig-maximillian University Munich, Germany

Introduction: We studied the performance of unattended automated office blood pressure (uAOBP) measurement in children, in relation to oscillometric office BP (OBP) and ambulatory blood pressure monitoring (ABPM).
Material and methods: 111 stable treated and untreated outpatients investigated for hypertension underwent uAOBP measurements (seated unattended in a quiet room separate from the renal clinic room, six times after a 5 min rest with the BpTRU device). Ambulatory 24h blood pressure monitoring (ABPM) was performed on the same day in a subgroup of 42 children.
Results: UAOBP measurements were successful in 106 children (95%), 5 preschool children did not tolerate to be alone in the room. The mean ±SD systolic/diastolic uAOBP, OBP and daytime ABP were 109.1 ± 14.0/70.8 ± 10.7 mmHg, 121.6 ± 16.5/77.6 ± 10.5 mmHg and 123.5 ± 11.3/73.7 ± 6.8 mmHg, respectively. Systolic/diastolic uAOBP was significantly lower than OBP by 13.6/7.6 mmHg (p<0.0001) and lower than daytime ABP by 14.4 ± 0.5/2.9 ± 0.3 mmHg (p<0.0001). The heart rate was not significantly different during uAOBP than during OBP measurements. On Bland Altman analysis the uAOBP underestimated OBP by a mean of 15.6 mmHg for systolic BP and by 8.6 mmHg for diastolic BP. In all 9 children with white-coat systolic hypertension uAOBP was within the normal range (<95th pc for OBP), in six of nine children with white-coat diastolic hypertension uAOBP was within the normal range however, in three of them it was elevated despite normal ABP.
Conclusions: uAOBP measurement is feasible in school-aged children, its values are considerably lower than OBP as well as daytime ABP and it could help with detection of white-coat systolic hypertension. The clinical applicability of uAOBP in children should be confirmed in further studies.

OP-27 CLINICAL COURSE OF ADOLESCENT ONSET ATYPICAL HEMOLYTIC UREMIC SYNDROME: A STUDY OF TURKISH AHUS REGISTRY

Kubra Celegen[1], Bora Gulhan[2], Kibriya Fidan[3], Selcuk Yuksel[4], Neslihan Yilmaz[4], Aysun Çaltik Yilmaz[5], Beltinge DemircioĞlu KiliÇ[6], Ibrahim Gokce[7], Asli Kavaz Tufan[8], Mukaddes Kalyoncu[9], Hulya Nalcacioglu[10], Sare Gulfem Ozlu[11], Eda Didem Kurt Sukur[2], Nur Canpolat[12], Aysun K. Bayazit[13], Mustafa Koyun[14], Yilmaz Tabel[15], Sebahat Tulpar[16], Mehtap Celakil[17], Kenan Bek[18], Cengiz Zeybek[19], Ali Duzova[2], Zeynep Birsin Ozcakar[20], Rezan Topaloglu[2], Oguz Soylemezoglu[3], Fatih Ozaltin[2]

[1]Division Of Pediatric Nephrology, Afyonkarahisar Health Sciences University, Afyonkarahisar, Turkey, [2]Division Of Pediatric Nephrology, Hacettepe University Faculty Of Medicine, Ankara, Turkey, [3]Division Of Pediatric Nephrology, Gazi University Faculty Of Medicine, Ankara, Turkey, [4]Division Of Pediatric Nephrology, Pamukkale University Faculty Of Medicine, Denizli, Turkey, [5]Division Of Pediatric Nephrology, Baskent University Faculty Of Medicine, Ankara, Turkey, [6]Division Of Pediatric Nephrology, Gaziantep University Faculty Of Medicine, Gaziantep, Turkey, [7]Division Of Pediatric Nephrology, Marmara University Faculty Of Medicine, Istanbul, Turkey, [8]Division Of Pediatric Nephrology, Osmangazi University Faculty Of Medicine, Eskisehir, Turkey, [9]Division Of Pediatric Nephrology, Karadeniz Technical University, Faculty Of Medicine, Trabzon, Turkey, [10]Division Of Pediatric Nephrology,ondokuz Mayis University Faculty Of Medicine, Samsun, Turkey, [11]Division Of Pediatric Nephrology, Ankara City Training And Research Hospital, Ankara, Turkey, [12]Division Of Pediatric Nephrology, Istanbul University, Cerrahpasa Faculty Of Medicine, Istanbul, Turkey, [13]Division Of Pediatric Nephrology, Cukurova University Faculty Of Medicine, Adana, Turkey, [14]Division Of Pediatric Nephrology, Akdeniz University Faculty Of Medicine, Antalya, Turkey, [15]Division Of Pediatric Nephrology, Inonu University Faculty Of Medicine, Malatya, Turkey, [16]Division Of Pediatric Nephrology, University Of Health Sciences, Istanbul Bakirkoy Dr. Sadi Konuk Research And Training Hospital, İstanbul, Turkey, [17]Division Of Pediatric Nephrology, Sakarya University Training And Research Hospital, Sakarya, Turkey, [18]Division Of Pediatric Nephrology, Kocaeli University Faculty Of Medicine, Kocaeli, Turkey, [19]Division Of Pediatric Nephrology, Gulhane Training And Research Hospital, Ankara, Turkey, [20]Division Of Pediatric Nephrology, Ankara University Faculty Of Medicine, Ankara, Turkey

Introduction: Atypical hemolytic uremic syndrome (aHUS) is a rare, mostly complement-mediated thrombotic microangiopathy. The majority of patients are infants. Our study aims to describe the clinical manifestations and genetic features of adolescent-onset aHUS.
Material and methods: Patients who were diagnosed as aHUS between the ages of ≥10 years and <18 years were defined as adolescent-onset aHUS, characterized by triad of microangiopathic hemolytic anemia, thrombocytopenia and acute kidney injury. The relevant data of the patients were obtained from the Turkish Pediatric aHUS registry.
Results: A total of 27 patients (20 female, 7 male) from different families were included. The mean age at diagnosis was 12.8±2.3 years. The mean follow-up duration was 4.5±2.1 years. Consanguinity was present in 11 families (48%). A total of 21 patients (77.8%) required acute renal replacement therapy. Extra-renal involvement was noted in 11 patients (40.7%); neurological involvement was the most common (33%). In six patients (22.2%), only plasma therapy was administered, in eight patients (29.6%) eculizumab was administered as first-line therapy. In 13 patients (48.1%), eculizumab was initiated because of unresponsiveness to the plasma therapy. Remission was achieved in 20 patients (74%) in the acute phase. Genetic screening for the mutations in the relevant genes was resulted as the following: CFHR 1-3 deletion (n=4, 14.8%), C3 mutation (n=2, 7.4%), CFH mutation (n=2, 7.4%), CFI mutation (n=1, 3.7%) and no mutation (n=18, 66.6%). Proteinuria and hypertension persisted in 9 (33.3%) and 11 patients (40.7%), respectively. End stage kidney disease was developed in 3 patients (11%). In follow-up, eculizumab was discontinued in 15 patients (55.6%) and relapse was observed in 1 (3.7%) patient.
Conclusions: Adolescent-onset aHUS is a very rare disease and may have different characteristics compared to infants. To our best knowledge, this study is the first mainly focusing on adolescence aHUS providing age-specific clinical and genetic features.

OP-28 PLASMA SOLUBLE TERMINAL COMPLEMENT COMPLEX C5B-9 IS ASSOCIATED WITH ACUTE GLOMERULAR HISTOLOGICAL LESIONS IN CHILDREN WITH KIDNEY DISEASE

Eugénie Fradette[1], Natalie Patey[1], Arnaud Bonnefoy[2], Alexandra Cambier[3], Stéphan Troyanov[4], Anne-laure Lapeyraque[3], Adrien Flahault[5]

[1]Department Of Pathology, Centre Hospitalier Universitaire Sainte-justine, Montreal, Quebec, Canada, [2]Hematology Division, Department Of Pediatrics, Centre Hospitalier Universitaire Sainte-justine, Montreal, Quebec, Canada, [3]Nephrology Division, Department Of Pediatrics, Centre Hospitalier Universitaire Sainte-justine, Montreal, Quebec, Canada, [4]Nephrology Division, Department Of Medicine, Hôpital Du Sacré-coeur De Montréal, Montreal, Quebec, Canada, [5]Nephrology Division, Hôpital Européen Georges Pompidou, Paris, France

Introduction: Complement activation plays a central role in the pathophysiology of glomerulonephritis. This study aimed to determine if urine and plasma levels of soluble terminal complement complex C5b-9 (sC5b-9) reflect complement activity defined by C5b-9 deposits associated with glomerular and tubular histological lesions.
Material and methods: We conducted a retrospective study of 61 kidney biopsy samples in children between 2018 and 2021.Urine and plasma samples were collected just before the kidney biopsy, to assess standard biochemical assays, plasma and urine sC5b-9. Biopsy specimens were examined in a blinded fashion by two operators using indirect immunohistochemistry against C5b-9 in paraffin-embedded sections. We quantified glomerular and tubular deposition of C5b-9 and calculated scores for active and chronic lesions. Chronic lesions were defined by percentage of glomerular sclerosis, segmental hyalin glomerular lesions and atrophic tubules. Active lesions were characterized by all other, non-chronic, lesions.
Results: Median (interquartile range) age of patients at biopsy was 14 (9, 16) years. Seventeen (28%) were follow-up kidney transplant biopsies. Median eGFR (defined the Schwartz formula) was 89 (77, 99) ml/min/1.73 m^2. Median plasma sC5b-9 was 166 (124, 273) ng/ml, and median urine sC5b-9 to creatinine ratio was 1.28 (0.56, 2.69) ng/mmol. Plasma sC5b-9 was significantly associated with active (p=0.019), but not chronic glomerular lesions. Conversely, urinary sC5b-9 to creatinine ratio was significantly associated with chronic (p=0.024), but not active, glomerular lesions. No biomarker was associated with tubular activity. We found no significant association of plasma or urine sC5b-9 with clinical and biological characteristics.
Conclusions: Plasma sC5b-9 is associated with histological acute glomerular activity whereas urinary sC5b-9 is associated with chronic glomerular lesions. Future studies are needed to determine whether plasma and urine sC5b-9 levels may be used as surrogates of renal biopsy to assess progression and treatment of kidney disease.

OP-29 MANNOSE-BINDING LECTIN POLYMORPHISMS IN CHILDREN WITH TYPICAL HEMOLYTIC UREMIC SYNDROME: A COINCIDENCE OR NOT?

Vasiliki Karava[1], Antonia Kondou[1], John Dotis[1], Eleni Gavriilaki[2], Nikoleta Printza[1]

[1]Pediatric Nephrology Unit, 1st Department Of Pediatrics, Hippokratio General Hospital, Aristotle University Of Thessaloniki, Greece, [2]Bone Marrow Transplantation Unit, Hematology Department, G. Papanicolaou Hospital, Thessaloniki, Greece

Introduction: The genetic predisposition for typical hemolytic uremic syndrome (HUS) is largely unknown. Mannose-binding lectin (MBL2) is associated with endothelial dysfunction and its inhibition protects against complement activation and renal injury induced by Stx-2 in mice models. We report 3 cases of typical HUS with presence of MBL2 gene polymorphisms.
Material and methods: DNA next generation sequencing (NGS) analysis was performed on peripheral blood samples from 3 patients with typical HUS secondary to shiga-toxin producing E.Coli (STEC) infection. Amplicons covered exonic regions of TMA- associated genes (CFH, CFI, CFB, CFD, C3, CD55, MCP, thrombomodulin, ADAMTS13, MASP, MBL and FCN).
Results: Two male patients of 8 and 9 months old and one female patient of 5.5 years old presented typical HUS with positive detection of stx by PCR in fecal specimen. Initial soluble C5b-9 levels were increased only in the female patient (920 ng/ml, normal range <245 ng/ml). All three patients required peritoneal dialysis for 14, 15 and 37 days respectively and remained oligo-anuric for 2, 4 and 16 days respectively. The first male patient presented an episode of tonico-clonic seizure at admission, with favorable clinical outcome and normal MRI findings. The female patient presented a severe hemorrhagic colitis, which was gradually resolved. The DNA NGS analysis revealed MBL-2 polymorphisms in the three patients: rs5030737, rs1800450 and rs5030737 respectively.
Conclusions: Three patients with typical HUS presented MBL-2 gene polymorphisms in DNA NGS analysis. The possible causative or triggering role of MBL2 gene polymorphisms on the occurrence of typical HUS needs further evaluation.

OP-30 NOVEL SYNDROME: CRANIOFACIAL AND CENTRAL NERVOUS SYSTEM MALFORMATIONS, AND ATYPICAL HEMOLYTIC UREMIC SYNDROME ASSOCIATED WITH TSEN2 MUTATION

Nur Canpolat[1], Dingxiao Liu[2], Emine Atayar[3], Seha Saygili[1], Nazli Sila Kara[4], Trudi A. Westfall[5], Qiong Ding[2], Bartley J. Brown[6], Terry A. Braun[6], Diane Slusarski[6], Kader Karli Oguz[7], Yasemin Ozluk[8], Beyhan Tuysuz[9], Tugba Tastemel Ozturk[10], Lale Sever[1], Osman Ugur Sezerman[4], Rezan Topaloglu[10], Salim Caliskan[1], Massimo Attanasio[2], Fatih Ozaltin[10]

[1]Department Of Pediatric Nephrology, Istanbul University-cerrahpasa, Cerrahpasa Faculty Of Medicine, Istanbul, Turkey, [2]Carver College Of Medicine, University Of Iowa, Iowa City, IA, [3]Nephrogenetics Laboratory, Department Of Pediatric Nephrology, Hacettepe University, Faculty Of Medicine, Ankara, Turkey, [4]Biostatistics And Medical Informatics Program, Faculty Of Medicine, Graduate School Of Health Sciences, Acibadem Mehmet Ali Aydinlar University, Istanbul, Turkey, [5]Department Of Biology, University Of Iowa, Iowa City, IA, [6]Center For Bioinformatics And Computational Biology, University Of Iowa, Iowa City, IA, [7]Department Of Radiology, Hacettepe University Faculty Of Medicine, Ankara, Turkey, [8]Department Of Pathology, Istanbul University Faculty Of Medicine, Istanbul, Turkey, [9]Department Of Pediatric Genetics, Istanbul University-cerrahpasa, Cerrahpasa Faculty Of Medicine, Istanbul, Turkey, [10]Department Of Pediatric Nephrology, Hacettepe University, Faculty Of Medicine, Ankara, Turkey

Introduction: Atypical hemolytic uremic syndrome (aHUS) is a rare and severe form of thrombotic microangiopathy (TMA). All forms of hemolytic uremic syndromes arise from vascular endothelial cell injury of the microvasculature of the kidney and other organs through complement-dependent or complement-independent mechanisms. The underlying genetic etiology has not been identified in 30% to 40% of all cases of aHUS, which indicates that there are still unknown etiologies.
Material and methods: We described six children from four consanguineous pedigrees, affected with microcephaly and craniofacial malformations, severe growth failure, intellectual retardation, and aHUS that progressed to end stage kidney disease requiring kidney replacement therapy within the first decade of life. Whole exome sequencing (WES) was perfomed to identify underlying genetic etiology.
Results: Our research identified a homozygous intronic variant in the gene TSEN2 (tRNA splicing endonuclease subunit 2) associated with this undescribed new syndrome. Bulk RNA sequencing of peripheral blood cells of four affected individuals revealed abnormal tRNA transcripts, indicating an alteration of the tRNA biogenesis. Morpholino-mediated skipping of exon 10 of tsen2 in zebrafish produced phenotypes similar to human patients.
Conclusions: We have identified a novel syndrome accompanied by aHUS that suggests the existence of a link between tRNA biology and vascular endothelium homeostasis. We propose to name it TRACK syndrome (**T**SEN2 **R**elated **A**typical hemolytic uremic syndrome, **C**raniofacial malformations, **K**idney failure).

OP-31 EFFECTS OF BUROSUMAB TREATMENT ON MINERAL HOMEOSTASIS IN CHILDREN AND ADOLESCENTS WITH X-LINKED HYPOPHOSPHATEMIA: LESSONS FROM THE GERMAN XLH REGISTRY

Ewert Annika Rehberg Mirko[2], Schlingmann Karl Peter[3], Kemper Markus[4], Derichs Ute[5], Patzer Ludwig[6], Staude Hagen[8], John Ulrike[7], Metzing Oliver[7], Weitz Marcus[9], Freiberg Clemens[10], WÜhl Elke[11], Schaefer Franz[11], Hiort Olaf[12], Schnabel Dirk[13], Haffner Dieter[1]

[1]Department Of Pediatric Nephrology, Hannover Medical School, Hannover, Germany, [2]Department Of Pediatrics, University Of Cologne, Cologne, Cologne, Germany, [3]Department Of General Pediatrics, Pediatric Nephrology, University Children's Hospital, MÜnster, Germany, [4]Asklepios Children's Hospital Hamburg-heidberg, Hamburg, Germany, [5]University Children's Hospital, Mainz, Germany, [6]St. Elisabeth And St. Barbara Children's Hospital, Halle/saale, Germany, [7]University Children's Hospital, Jena, Germany, [8]University Children's Hospital, Rostock, Germany, [9]Pediatric Nephrology, University Children's Hospital, TÜbingen, Germany, [10]Department Of Pediatrics, Universitätsmedizin GÖttingen, GÖttingen, Germany, [11]Division Of Pediatric Nephrology, Center For Pediatrics And Adolescent Medicine, Heidelberg University Hospital, Germany, [12]University Children's Hospital LÜbeck, LÜbeck, [13]Center For Chronically Sick Children, Pediatric Endocrinology, University Medicine, Charitè Berlin, Germany

Introduction: Burosumab was approved for treatment of pediatric patients with X-linked hypophosphatemia (XLH). However, data on its efficacy in adolescents (age > 12 years) and in real-world settings are lacking.
Material and methods: Here we assess the effects of 12 months burosumab treatment on mineral homeostasis in 77 pediatric XLH

patients (50 children, 27 adolescents) enrolled in the German XLH Registry. Age and sex related SD scores (SDS) were calculated for serum phosphate and alkaline phosphatase (ALP) levels, and renal tubular reabsorption of phosphate per glomerular filtration rate (TmP/GFR).
Results: At baseline, all patients presented with profound hypophosphatemia (-4.5 SDS), reduced TmP/GFR (-6.5 SDS), and elevated ALP (2.7 SDS, each $p<0.001$ versus healthy children) suggesting persisting rickets despite long-term therapy with oral phosphate and active vitamin D. Burosumab treatment resulted in rapid increases in mean serum phosphate and TmP/GFR by approx. 0.3 mmol/l amounting to -2.2 SDS and -2.5 SDS at 12 months, respectively (each $p<0.001$ versus baseline). This was paralleled by a continuous decrease in serum ALP (1.3 SDS, $p<0.001$ versus baseline). Serum phosphate, TmP/GFR, and ALP values were normalized ind approximately 40%, 30% and 80% of patients, respectively. Two patients had transient hyperphosphatemia due to a dosing error. At 12 months, the median burosumab dosage amounted to 0.8 mg/kg (range 0.6-1.2). Serum phosphate levels at 12 months were comparable between children (-2.3 SDS) and adolescents (-2.1 SDS) and associated with parathyroid hormone (PTH) levels. Serum ALP z-scores were associated with PTH levels in adolescents but not in children.
Conclusions: In this real world setting 12 months burosumab treatment was effective to normalize serum ALP levels in children and adolescents with XLH suggesting healing of rickets despite persisting mild hypophosphatemia in about half of patients. Elevated PTH levels are a risk factor for failure to normalize mineral homeostasis.

OP-32 A RANDOMIZED, PLACEBO -CONTROLLED, PHASE 2/3 STUDY OF GLYCOLATE OXIDASE (GO) INHIBITOR BBP-711 IN CHILDREN AND ADULTS WITH PRIMARY HYPEROXALURIA TYPE 1

Scott Adler, Jia Ma, Ramei Sani-grosso, *Lillian Lee*, *Gustavo Lorente*, *Justin Lafountaine*, *Jonathan Fox*

Cantero Therapeutics, A Bridgebio Company

Introduction: Primary hyperoxaluria (PH) is a group of rare, autosomal recessive, inborn errors of metabolism resulting in excessive production of oxalate that can lead to nephrolithiasis, nephrocalcinosis, renal failure, and systemic oxalosis. PH Type 1 (PH1) is the most common and severe of the subtypes and is caused by a deficiency in the liver-specific peroxisomal enzyme alanine-glyoxylate aminotransferase (AGT) due to mutations in the AGXT gene. Deficiency in AGT impairs the transamination of glyoxylate to glycine in hepatic peroxisomes and the accumulated glyoxylate is subsequently metabolized into oxalate. In the metabolic step preceding the transamination of glyoxylate, glycolate oxidase (GO) catalyzes the conversion of glycolate to glyoxylate.
Material and methods: GO has been shown to be a safe and efficient target for substrate reduction therapy in PH1. BBP-711 is an oral, small molecule inhibitor of GO. Inhibition of GO is expectded to reduce hepatic oxalate production in PH1, reducing hyperoxaluria and its sequelae.
Results: In healthy adult volunteers, BBP-711 had a PK profile to support once daily dosing, and an acceptable safety profile. BBP-711 is a potent inhibitor of GO, resulting in plasma glycolate concentrations comparable to case reports of individuals with germline HAO1 deletions, suggesting near complete GO inhibition.
Conclusions: This global, seamless Phase 2/3 study will evaluate the safety and efficacy of BBP-711 in children and adults with PH1. The study will consist of two parts. Part A will include a dose-finding period to identify a well-tolerated therapeutic dose for Part B. Part B will be a randomized, placebo-controlled, trial in patients with PH1 not requiring renal replacement therapy. The primary endpoint will be a change from baseline ind 24hr urinary oxalate (UOx) excretion corrected for body surface area (BSA). Key secondary endpoints include absolute change from baseline in 24hr UOx excretion corrected for BSA and percentage of participants with 24hr UOx below ULN.

OP-33 HUMORAL RESPONSE TO COVID-19 MRNA VACCINES IN A COHORT OF YOUNG KIDNEY TRANSPLANT RECIPIENTS FROM A SINGLE CENTER IN NORTHERN ITALY

Marta Brambilla[1], Sara Testa[1], Marco Cazzaniga[2], Jessica Serafinelli[1], Chiara Tamburello[1], Viganoni Maria[2], Massimo Oggioni[3], Ferruccio Ceriotti[3], Giovanni Montini[4]

[1]Fondazione Irccs Ca Granda Ospedale Maggiore Policlinico, Pediatric Nephrology, Dialysis And Transplant Unit, Milan, Italy, *[2]University Of Milan, Milan, Italy*, *[3]Fondazione Irccs Ca Granda Ospedale Maggiore Policlinico, Clinical Laboratory, Milan, Italy*, *[4]Fondazione Irccs Ca Granda Ospedale Maggiore Policlinico, Pediatric Nephrology, Dialysis And Transplant Unit; University Of Milan, Department Of Clinical Sciences And Community Health, University Of Milan, Milan, Italy*

Introduction: To investigate immune-response to COVID-19 vaccines in young kidney transplant (KT) recipients from Northern Italy.
Material and methods: We prospectively studied KT patients aged 12 to 25 years, managed in our Center on maintenance IS therapy (corticosteroids, CNI and anti-proliferative agents), who received a complete primary antiSARS-CoV2 vaccination course and an additional dose one month later according to Italian Medicines Agency. From 1st July 2021 to 31st January 2022 we evaluated antiSpike-protein antibody response at T_0 (before vaccine), at T_1 and T_2 (14 ± 3 days after 2nd and 3rd doses respectively) of BNT162b2 (Pfizer/BioNTech) or mRNA-1273 (Moderna). Exclusion criteria were: KT or other IS within 6 months; relapse of primary kidney disease; vaccine before KT; ongoing COVID-19.
Results: 87 patients were eligible; 68 (45M) patients were enrolled. Median age was 19.5 (IQR: 16.3–21.9) years; median time from KT was 61.4 (IQR: 36.7-111.7) months. Anti-spike total Ig titer was considered undetectable if <0.8 U/ml, (Roche® Elecsys Anti-SARS-CoV-2 S). At T_0 14 pts (21.5%; data in 65 pts) had a positive titer (median: 211.5 U/mL IQR:147-829); among them five had a history of SARS-Cov2. Five patients dropped out of study after enrollment.
At T_1 35 pts (63.6%; data in 55 pts) and at T2 44 pts (90.0%; data in 49 pts) seroconverted or enhanced titer, if previously immunized (median:1256 U/mL; IQR:308.00-13000 and 3662 U/mL; IQR: 193–13000, respectively). 55% of non-responders at T1 seroconverted after the third dose. Patients that experienced COVID-19 before vaccination developed significantly higher antibody titer (median 13000 vs 3.7 $p<0.05$). No patient had side effects, including acute rejection or DSAs.
Conclusions: KT pediatric recipients exhibit a satisfactory response after 2 doses of vaccine, that become comparable to that of immunocompetent population after the third. Furthermore, the response after two doses is better if compared with adult KT population (63.6% vs 4-48%).

OP-34 COVID 19 PANDEMIC RESULTED IN SIGNIFICANT WEIGHT GAIN IN IN TEENAGERS AFTER KIDNEY TRANSPLANTATION

Nele Kanzelmeyer[1], Weigel Friederike[2], Boeckenhauer Johannes[3], Drube Jens[1], Haffner Dieter[1], Oh Jun[3], Schild Sebastian[3]

[1]Department Of Pediatric Kidney, Liver And Metabolic Diseases, Hannover Medical School, Hannover, Germany, *[2]Division Of Pediatric Nephrology, University Children´s Hospital, Jena, Germany*, *[3]Department Of Pediatric Nephrology, University Medical Center Hamburg/eppendorf, Hamburg, Germany*

Introduction: The COVID-19 pandemic has led to change of lifestyle, restrictions of social relations, physical activities, modifications in eating habits, and psychological distress. The COVID-19 pandemic leads to significant weight gain in the general population, but its impact on pediatric patients after kidney transplantation (KTx) is unknown.
Material and methods: We retrospectively evaluated body mass index SD scores (BMI-SDS) between September 2019 and September 2021 in 132 pediatric KTx patients followed up at three German pediatric nephrology centers. The patients were categorized according to age (0-11.9 years vs 12-18 years) and sex (female vs. male) in four groups. Data were assessed by a linear mixed model approach.
Results: There was no significant change in BMI-SDS in children (0-11.9 years), irrespectively of sex (boys -0.11 SDS, p=0.22; girls 0.05 SDS, p=0.49). By contrast, a significant increase in BMI-SDS was noted in both male (0.24 SDS) and female (0.20 SDS) teenagers (each p<0.05). In addition, the proportion of obese teenagers tended to increase from 12% to 19% (p=0.08).
Conclusions: The COVID19 pandemic was associated with a significant increase in standardized BMI values in adolescents but not in children after KTx. This may further increase the cardiovascular risk in the former population.

OP-35 HUMORAL AND CELLULAR IMMUNITY TO SARS-COV-2 VACCINATION WITH BNT162B2 MRNA VACCINE IN PEDIATRIC KIDNEY TRANSPLANT AND DIALYSIS PATIENTS

Ruveyda Gulmez[1], Dogukan Ozbey[2], Ayse Agbas[1], Bagdagul Aksu[3], Nurdan Yildiz[4], Diana Uckardes[5], Seha Saygili[1], Zeynep Yuruk Yildirim[3], Mehmet Tasdemir[6], Ayca Kiykim[7], Haluk Cokugras[8], Nur Canpolat[1], Ahmet Nayir[3], Bekir S Kocazeybek[2], Salim Caliskan[1]

[1]Department Of Pediatric Nephrology, CerrahpaŞa Faculty Of Medicine, Istanbul University-cerrahpaŞa, Istanbul, Turkey., [2]Department Of Microbiology, CerrahpaŞa Faculty Of Medicine, Istanbul University-cerrahpaŞa, Istanbul, Turkey., [3]Department Of Pediatric Nephrology, Faculty Of Medicine, Istanbul University, Istanbul, Turkey., [4]Department Of Pediatric Nephrology, Faculty Of Medicine, Marmara University, Istanbul, Turkey., [5]Department Of Pediatric Nephrology, Faculty Of Medicine, Istanbul Medeniyet University, Istanbul, Turkey., [6]Department Of Pediatric Nephrology, Istinye University Liv Hospital BahÇeŞehir, Istanbul, Turkey., [7]Department Of Pediatric Allergy And Immunology, CerrahpaŞa Faculty Of Medicine, Istanbul University-cerrahpaŞa, Istanbul, Turkey., [8]Department Of Pediatric Allergy, Chest Diseases, Infectious Diseases, Department Of Pediatrics, CerrahpaŞa Faculty Of Medicine, Istanbul University-cerrahpaŞa, Istanbul, Turkey.

Introduction: Immune response to SARS-CoV-2 vaccination is lower in kidney transplant (KTx) recipients compared with healthy individuals and dialysis patients. However, few data are available for the pediatric population. We aimed to investigate both humoral and cellular immune response to SARS-CoV-2 vaccination in pediatric KTx and dialysis patients.
Material and methods: In this multicenter study, 61 patients (49 KTx, 12 dialysis) were evaluated for SARS-CoV-2 specific humoral (anti-spike IgG and neutralizing antibody) and cellular (T-cell interferon ɣ release assay; IGRA) immune responses at least one month after two doses of SARS-CoV-2 mRNA vaccine (BNT162b2).
Results: The mean age was 16±2.7 years (54% male), and the median duration after vaccination was 56 (42;77) days. There was no difference between the dialysis and KTx groups considering a positive PCR test for SARS-CoV-2 before the second dose of vaccination. The dialysis group had significantly higher median anti-spike IgG titer than in the KTx group (p=0.007). The prevalence of positive anti-spike IgG, neutralizing antibody, and IGRA were also higher in the dialysis group than in the KTx group [100%, 83.3%, and 100% vs 75.5%, 64.4%, and 71.4%, respectively], but the differences were not statistically significant. In the KTx group, 57% of patients (n=24) had a complete response with both IgG and IGRA positive, but 12 patients had a partial response (Ig G or IGRA positive) and 6 patients had no response (IgG and IGRA negative). The incidence of history of acute rejection was significantly higher in patients with partial and no response than in patients with complete response (5/18 vs 0/24, p=0.018), but there were no differences between the two groups considering age, sex, transplant vintage, induction or maintenance therapy, or SARS-CoV-2 PCR positivity.
Conclusions: In the pediatric population, KTx recipients had a lower immune response than dialysis patients after two doses of SARS-CoV-2 mRNA vaccine.

OP-36 RISK FACTORS FOR KIDNEY SURVIVAL IN PATIENTS WITH AUTOSOMAL RECESSIVE POLYCYSTIC KIDNEY DISEASE (ARPKD)

Kathrin Burgmaier[1], Samuel Kilian[2], Anja BÜscher[3], Ismail Dursun[4], Marc Fila[5], Ibrahim Gokce[6], Nakysa Hooman[7], Matko Marlais[8], Laura Massella[9], Antonio Mastrangelo[10], Djalila Mekahli[11], Lukasz Obrycki[12], Larisa Prikhodina[13], Bruno Ranchin[14], Lutz T. Weber[1], Elke WÜhl[15], Katarzyna Zachwieja[16], JÖrg DÖtsch[1], Franz Schaefer[15], Max Liebau[17]

[1]Department Of Pediatrics, University Hospital Cologne And University Of Cologne, Faculty Of Medicine, Cologne, Germany, [2]Institute Of Medical Biometry, University Of Heidelberg, Heidelberg, Germany, [3]Department Of Pediatrics Ii, University Hospital Essen, Essen, Germany, [4]Department Of Pediatric Nephrology, Erciyes University, Faculty Of Medicine, Kayseri, Turkey, [5]Pediatric Nephrology Unit, Chu Arnaud De Villeneuve-université De Montpellier, Montpellier, France, [6]Research And Training Hospital, Division Of Pediatric Nephrology, Marmara University, Istanbul, Turkey, [7]Department Of Pediatric Nephrology, Ali-asghar Children Hospital, Ali-asghar Clinical Research Development Center (aacrdc), Iran University Of Medical Sciences, Tehran, Iran, [8]Ucl Great Ormond Street Hospital For Children Institute Of Child Health, Ucl, London, Uk, [9]Division Of Nephrology, Department Of Pediatric Subspecialties, Bambino Gesù Children's Hospital, Irccs, Rome, Italy, [10]Pediatric Nephrology, Dialysis And Transplant Unit, Fondazione Irccs Cà Granda, Ospedale Maggiore Policlinico, Milan, Italy, [11]Department Of Pediatric Nephrology, University Hospitals Leuven, Leuven, Belgium And Department Of Development And Regeneration, Pkd Research Group, Ku Leuven, Leuven, Belgium, [12]The Childrens Memorial Health Institute, Warsaw, Poland, [13]Department Of Inherited And Acquired Kidney Diseases, Research Clinical Institute For Pediatrics N.a. Acad. Y. E. Veltishev, Pirogov Russian National Research Medical University, Moscow, Russia, [14]Pediatric Nephrology Unit, Hôpital Femme Mère Enfant, Hospices Civils De Lyon, Centre De Référence Maladies Rénales Rares, Bron, France, [15]Division Of Pediatric Nephrology, Center For Pediatrics And Adolescent Medicine, University Of Heidelberg, Heidelberg, Germany, [16]Department Of Pediatric Nephrology And Hypertension, Faculty Of Medicine, Jagiellonian University Medical College, Krakow, Poland, [17]Department Of Pediatrics, University Hospital Cologne, Center For Molecular Medicine, University Hospital Cologne And University Of Cologne, Faculty Of Medicine, Cologne, Germany:

Introduction: Autosomal recessive polycystic kidney disease (ARPKD) is a rare but severe early-onset disease with pronounced phenotypic variability. It is mainly caused by variants in the PKHD1 gene. We previously elucidated genotype-phenotype correlations and identified prenatal

sonographic risk markers. Here, we aimed to describe further clinical and sonographic risk markers and to weigh their impact on kidney survival.
Material and methods: We analysed clinical datasets of 605 ARPKD patients from the ARegPKD registry study. Kidney survival and yearly eGFR loss were analysed in the overall cohort as well as in subgroup analyses in children with kidney survival >1.0 years.
Results: Ten-year kidney survival differed relevantly according to the factors prematurity (gestational age at birth <37 weeks; 48% vs. 79%), prenatal renal cysts (55% vs. 83%), prenatal renal hyperplasia (50% vs. 83%), oligo-/anhydramnios (50% vs. 86%), age at diagnosis (47% if prenatal diagnosis, 70% if diagnosis within first year of life, 93% if diagnosis ≥1.0 year) and postnatal assisted breathing or ventilation (40% vs. 83%). Type of assisted breathing/ventilation is relevant with 10-year-survival of 63% if CPAP, 35% if conventional ventilation and 26% if high-frequency oscillation was applied postnatally. Genetic risk factors are consistent with previously reported results showing worst kidney survival in patients with two Null PKHD1 variants. Mean yearly eGFR loss in all patients with kidney survival >1.0 years was 1.0 ml/min/1.73m². Type of assisted breathing/ventilation was consistently relevant for yearly eGFR loss, which was highest in patients with reported high-frequency oscillation ventilation (1.9 ml/min/1.73m²) and lowest in patients without perinatal assisted breathing/ventilation (0.8 ml/min/1.73m²).
Conclusions: We identified a number of pre-, peri- and postnatal sonographic, clinical and genetic risk factors associated with poor kidney survival in children and adolescents with ARPKD. We are currently integrating these data into joint models aiming to establish a predictive model for kidney survival.

OP-37 THE POTENTIAL OF CTNS-MRNA BASED GENE REPLACEMENT THERAPY TO TREAT NEPHROPATHIC CYSTINOSIS

Tjessa Bondue[1], Lambertus Van Den Heuvel[1], Rik Gijsbers[2], Roland Brock[3], Elena Levtchenko[1]

[1]Department Of Pediatric Nephrology & Growth And Regeneration, University Hospitals Leuven & Ku Leuven, Uz Herestraat 49-3000, Leuven, Belgium., [2]Department Of Pharmaceutical And Pharmacological Sciences, Ku Leuven, Herestraat 49 - 3000 Leuven, Belgium., [3]Department Of Biochemistry, Radboud Institute For Molecular Life Sciences, Radboud University Medical Center, Geert Grooteplein 28, 6525 Ga Nijmegen, The Netherlands

Introduction: Cystinosis is an autosomal recessive lysosomal storage disorder caused by mutations in CTNS, encoding the cystine transporter, cystinosin. Mutations result in lysosomal cystine accumulation in all cells of the body and the kidneys are the most affected organs. The current standard therapy, cysteamine, reduces cellular cystine levels, but does not cure the disease or improves the renal Fanconi syndrome (a generalized proximal tubular dysfunction).
mRNA has revolutionized the world of molecular therapy and mRNA-based therapeutics have started to emerge in the kidney field**.** Our aim is to investigate mRNA-based gene replacement to treat cystinosis.
Material and methods: Patient derived proximal tubular epithelial cells (CYS PTECs) were transfected with synthetic CTNS-mRNA. A HA-tag was included in the sequence, allowing for immunostaining to assess the stability of the newly expressed cystinosin protein. Co-staining with the lysosomal associated membrane protein 1 (LAMP1) was used to assess the lysosomal localisation and cystine measurement was performed at 24 hours post transfection (hpt).
Results: After transfection, PTECs were evaluated for the cystinosin protein from 12h to 10 days after transfection. Transfection efficiency was 72% (±11%) for the first 48hpt and protein half-life was estimated at 3-4 days, with no protein detectable at 7 days post-transfection. Co-staining with LAMP1 confirmed the lysosomal localisation of the protein at 24hpt. The functionality of the CTNS-mRNA was evaluated by cystine measurement at 24hpt and showed a significant decrease in cystine content after treatment.
Conclusions: Our results shows that mRNA-based gene replacement results in detectable lysosomal cystinosin expression in CYS PTECs for up to 7 days and leads to cystine reduction.

OP-38 GITELMAN-LIKE SYNDROME CAUSED BY PATHOGENIC VARIANTS IN MTDNA

Schlingmann Karl[1], Viering Daan[2], Hureaux Marguerite[3], Nijenhius Tom[4], Klaus Günter[5], Komhoff Martin[5], Beetz Rolf[6], Shenoy Mohan[7], Kleta Robert[8], Houillier Pascal[10], Konrad Martin[1], Vargas-poussou Rosa[9], Knoers Nine[11], Bockenhauer Detlef[8], Debaaij Jeroen[2]

[1]University Childrens Hospital MÜnster, [2]Radboud University Medical Center, Radboud Institute For Molecular Life Sciences, Department Of Physiology, Nijmegen, The Netherlands., [3]Reference Center For Hereditary Kidney And Childhood Diseases (marhea), Paris, France, [4]Radboud University Medical Center, Radboud Institute For Molecular Life Sciences, Department Of Nephrology, Nijmegen, The Netherlands., [5]Kuratorium Für Heimdialyse Pediatric Kidney Center, Marburg, Germany, [6]Johannes Gutenberg Universität Mainz, Zentrum Für Kinder- Und Jugendmedizin, Mainz, Germany, [7]Department Of Paediatric Nephrology, Royal Manchester Children's Hospital, Manchester, United Kingdom, [8]Department Of Paediatric Nephrology, Great Ormond Street Hospital For Children Nhs Foundation Trust, London, United Kingdom, [9]Centre De Recherche Des Cordeliers, Sorbonne Université, Inserm, Université De Paris, Cnrs, Paris, France, [10]Department Of Physiology, Assistance Publique-hôpitaux De Paris, Hôpital Européen Georges Pompidou, Paris, France, [11]Department Of Genetics, University Medical Center Groningen, University Of Groningen, Groningen, The Netherlands

Introduction
Gitelman syndrome (GS) is the most frequent hereditary salt-losing tubulopathy characterized by hypokalemic alkalosis and hypomagnesemia. It is caused by biallelic pathogenic variants in SLC12A3, encoding the Na+-Cl- cotransporter (NCC) expressed in the distal convoluted tubule. However, for approximately 10 percent of GS patients, the genotype still remains unknown.
Material and methods: Mitochondrial DNA variants were identified in three families with Gitelman-like electrolyte abnormalities, then 156 families were investigated for variants in MT-TI and MT-TF encoding the transfer RNAs for phenylalanine and isoleucine. Mitochondrial respiratory chain function was assessed in patient fibroblasts and mitochondrial dysfunction induced in HEK293 cells to assess the effect on NCC-mediated 22Na+ transport.
Results: Genetic investigations revealed four mtDNA variants in near homoplasmic state in 13 families. Importantly, affected members of six families with an MT-TF variant additionally suffered from progressive chronic kidney disease. Dysfunction of oxidative phosphorylation complex IV and reduced maximal mitochondrial respiratory capacity were found in patient fibroblasts. In vitro pharmacological inhibition of complex IV, mimicking the effect of the mtDNA variants, inhibited NCC phosphorylation and NCC-mediated sodium uptake.
Conclusions: Pathogenic mtDNA variants in MT-TF and MT-TI cause a Gitelman-like syndrome. Genetic investigation of mtDNA should be considered in patients with unexplained Gitelman syndrome-like tubulopathies.

OP-39 EFFICACY OF LEVAMISOLE FOR MAINTAINING REMISSION AFTER THE FIRST FLARE OF STEROID SENSITIVE NEPHROTIC SYNDROME IN CHILDREN: THE NEPHROVIR-3 RANDOMIZED CONTROLLED TRIAL

Claire Dossier[1], Theresa Kwon[1], Emeline Chapelon[2], Cyrielle Parmentier[3], Aurelien Galerne[4], Anne Chace[5], Marion Cheminee[6], Sylvie Nathanson[7], Fouad Madhi[8], Ferielle Zenkhri[1], Sebastien Rouget[9], Olivia Boyer[10], Celia Nekrouf[11], Alexandra Rousseau[11], Julien Hogan[1]

[1]Pediatric Nephrology, Robert-debre Hospital, Paris, Aphp, [2]Department Of Pediatrics, Ch Victor Dupouy, Argenteuil, [3]Pediatric Nephrology, Armand-trousseau Hospital, Paris, Aphp, [4]Department Of Pediatrics, Jean-verdier Hospital, Paris, Aphp, [5]Department Of Pediatrics, Chi Villeneuve Saint-georges, [6]Department Of Pediatrics, Gh Nord Essonne, Orsay, [7]Department Of Pediatrics, Ch De Versailles, Le Chesnay, [8]Department Of Pediatrics, Chi Créteil, [9]Department Of Pediatrics, Chsf, Corbeil-essonnes, [10]Pediatric Nephrology, Necker Hospital, Paris, Aphp, [11]Urc-est, Saint-antoine Hospital, Paris, Aphp

Introduction: In children with Steroid Sensitive Nephrotic Syndrome (SSNS), relapse after the first flare occurs in 80% of cases, whatever the dosage or duration of initial steroid therapy. Therefore, there is an unmet need for early interventions to reduce incidence of early relapses. Levamisole is an antihelmintic drug with an immunomodulatory action that reduces relapses in children with Frequent Relapses or Steroid Dependant NS. NEPHROVIR-3 is the first trial to assess the efficacy of levamisole in increasing duration of initial remission after the diagnosis of INS.
Material and methods: NEPHROVIR-3 is a multicentric placebo-controlled randomized trial (1:1), in 38 centers of the Paris area, France. Patients were included at INS diagnosis and randomized, when steroid sensitive within 4 weeks, to receive either levamisole 2.5 mg/kg/48h or placebo for 6 months, in addition to the French steroid protocol (18 weeks-3990mg/m^2). Primary outcome was the relapse-free survival at 1 year. The effect of the study drug was analysed by a Cox proportional hazard model stratified on centre.
Results: Between September 2017 and February 2020, 86 patients were included, median age at INS onset was 5 yrs (IQ 3-7), with 69% of boys. At 4 weeks, 68 of them were randomized. Median time to remission was 8.5 days (IQ 6-12). Relapse-free survival at 12 months was 53.8% (95%IC 34.7-69.5) in the levamisole group versus 20.9% (7.2-39.4) in the placebo group (p=0.007). The risk of relapse associated with levamisole was HR =0.37 (95%IC 0.15-0.89). Treatment was well tolerated with one interruption in each group because of skin rash.
Conclusions: Early treatment with levamisole at the first flare of childhood SSNS is well tolerated and significantly improves relapse-free survival at 1 year.

OP-40 MTOR-ACTIVATING MUTATIONS IN RRAGD CAUSE KIDNEY TUBULOPATHY AND CARDIOMYOPATHY SYNDROME

Schlingmann Karl[1], Jouret Francois[2], Shen Kuang[3], Dafinger Claudia[4], Houillier Pascal[5], Oh Jun[6], Godefroid Nathalie[7], Schermer Bernhard[8], Bergmann Carsten[9], Beck Bodo[10], Sabatini David[1], Liebau Max[4], Vargas-poussou Rosa[11], Knoers Nine[12], De Baaij Jeroen[13], Konrad Martin[1]

[1]University Childrens Hospital MÜnster, [2]Department Of Internal Medicine, University Of Liège, Belgium, [3]Whitehead Institute For Biomedical Research, Cambridge, MA, USA, [4]Department Of Pediatrics, University Hospital Cologne, Germany, [5]Department Of Physiology, Hôpital Européen Georges Pompidou, Paris, France, [6]Department Of Pediatrics, University Medical Center Hamburg, Germany, [7]Division Of Pediatric Nephrology, Uc Louvain, Brussels, Belgium, [8]Department Ii Of Internal Medicine, University Hospital Cologne, Germany, [9]Departmen Of Medicine, Division Of Nephrology, University Hospital Freiburg, Germany, [10]Department Of Human Genetics, University Hospital Cologne, Germany, [11]Department Of Genetics, Hôpital Européen Georges-pompidou, Paris, France, [12]Department Of Genetics, University Medical Center Groningen, [13]Department Of Physiology, Radboud University Medical Center Nijmegen, The Netherlands

Introduction: Over the last decades, advances in genetic techniques have resulted in the identification of rare hereditary disorders of renal magnesium and salt handling. Nevertheless, ±20% of all tubulopathy patients remain without genetic diagnosis. Here, we explore a large multicentric patient cohort with a novel inherited salt-losing tubulopathy, hypomagnesemia and dilated cardiomyopathy (DCM).
Material and methods: Whole exome and genome sequencing was performed with subsequent functional analyses of identified RRAGD variants in vitro.
Results: In 8 children from unrelated families with a tubulopathy characterized by hypomagnesemia, hypokalemia, salt-wasting, and nephrocalcinosis, we identified heterozygous missense variants in RRAGD that mostly occurred de novo. Six of these patients additionally suffered from DCM requiring heart transplantation in 3 of them. An additional dominant variant in RRAGD was simultaneously identified in eight members of a large family with a similar renal phenotype. RRAGD encodes GTPase RagD mediating amino acid signaling to the mechanistic target of rapamycin complex 1 (mTORC1). Identified RRAGD variants were shown to induce an increased interaction with components of mTORC1 and a constitutive activation of mTOR signaling in vitro.
Conclusions Our findings establish a novel disease phenotype combining kidney tubulopathy and cardiomyopathy caused by an activation of mTOR signaling suggesting a critical role of Rag GTPase D for renal electrolyte handling and cardiac function.

OP-41 BONE BIOMARKERS IN RESPONSE TO DIFFERENT DOSING REGIMEN OF CHOLECALCIFEROL THERAPY IN CHILDREN WITH CHRONIC KIDNEY DISEASE

Nivedita Kamath[1], Arpana Iyengar[1], Hamsa Reddy[1], Jyoti Sharma[2], Jyoti Singhal[2], Sudha Ekambaram[3], Susan Uthup[4], Sumithra Selvam[1], Dagmar Christiane-fischer[5], Anja Rahn[5], Mandy Wan[6], Rukshana Shroff[6]

[1]St Johns Medical College Hospital, [2]Kem Hospital, [3]Mehta Multispeciality Hospital, [4]Sat Hospital, [5]University Of Rostock, [6]Great Ormond Street Hospital

Introduction: In a randomised controlled trial we have shown that daily, weekly or monthly dosing regimens of cholecalciferol achieved comparable 25-hydroxyvitamin D (25OHD) concentrations in children with CKD stages 2-4. However, the effect on bone biomarkers is not known. We investigated the effect of cholecalciferol supplementation regimens on markers of bone formation [bone alkaline phosphatase (BAP), N-terminal propeptide-of-type-1-procollagen (PINP)], bone resorption [tartrate resistant acid phosphatase 5b (TRAP), C-terminal telopeptide-of-type 1 collagen (CTX)], osteocyte activity [Fibroblast growth factor 23(iFGF23), klotho and sclerostin].
Material and methods: This is a post-hoc analysis of an open label, multi-centre randomized controlled trial using equivalent doses of cholecalciferol (intensive therapy) administered as

daily(3,000IU), weekly(25,000IU) and monthly(1,00,000IU) therapy in vitamin D deficient (25OHD levels<30 ng/ml) children with CKD. Bone biomarkers were estimated using standard ELISAs at baseline and end of intensive therapy. The change in bone biomarkers was compared between the 3 treatment arms and correlated with change in 25OHD levels.

Results: Bone biomarkers were comparable by age, sex, etiology of CKD and treatment arm at baseline (p>0.05) in 61 children. After intensive phase therapy, there was a significant increase in 25OHD (p<0.001). The change in 25OHD (Δ25OHD) correlated inversely with ΔPTH(r=-0.4, p<0.001) and ΔTRAP(r=-0.26, p=0.04). There was a significant increase in BAP/TRAP ratio(p=0.04) implying bone formation, and in iFGF23 and klotho (p=0.004 and p=0.002 respectively).

Comparing between the therapy arms, BAP z-score was higher with weekly therapy(p=0.01), though other markers were comparable. PTH was lower(p=0.015) and PINP/CTX was higher(p=0.04) in those with 25OHD>30 ng/ml compared to those with 25OHD <30ng/ml after intensive therapy.

Conclusions: Children receiving daily, weekly or monthly cholecalciferol dosing regimens show a comparable bone biomarker profile suggesting that these treatment schedules can be used interchangeably. There was an increase in the ratio of bone formation to resorption markers implying bone formation with 25OHD therapy.

OP-42 ISOLATED KIDNEY TRANSPLANTATION UNDER LUMASIRAN THERAPY IN PRIMARY HYPEROXALURIA TYPE 1 (PH1): A REPORT ON 3 CASES

Anne-laure Sellier-leclerc[1], Charlene Levi[1], Cecile Acquaviva-bourdain[1], Stephanie Clave[2], Justine Bacchetta[1]

[1]Hospices Civils De Lyon, [2]Assitance Publique Hopitaux De Marseille

Introduction: The RNA-interference therapy lumasiran demonstrated its efficacy to decrease urinary (UOx/creat) and circulating (POx) oxalate levels in PH1. Whether combined liver/kidney transplantation (CLKTx) can be replaced by isolated KTx and lumasiran remains debatable.

Material and methods: Three cases of genetically-confirmed PH1 patients receiving isolated KTx are described. They all received post-operatively "standard of care" (SOC), associating hyperhydration (3L/m²/day), potassium citrate (250mg/kg/day), pyridoxine and lumasiran.

Results: Patient 1: diagnosis 1.5 years, dialysis initiation 0.5 years, POx 110µmol/L (N<5) at the beginning of lumasiran at 2.5 years, KTx 13 months after lumasiran (POx 53µmol/L), deceased donor. Post-operative management: 3 early "prophylactic" hemodialysis sessions, then SOC. ARF on JJ obstruction at day 5, 15 hemodialysis sessions. At one month, renal function 125mL/min/1.73 m², POx 14µmol/L, UOx/creat 519µmol/mmol (<100). Follow-up 3 months, stable renal function, POx and UOx/creat.

Patient 2: diagnosis 17 years, dialysis initiation 23 years, POx 20µmol/L at the beginning of lumasiran at 26 years, KTx 10 months after lumasiran (POx 10µmol/L), living donor. No delayed graft function. Post-operative management: SOC. At one month, renal function 48mL/min/1.73 m², POx<5 µmol/L, UOx/creat 67µmol/mmol (<80). Follow-up 3 months, stable renal function and normal UOx/creat.

Patient 3: diagnosis 6 years, dialysis initiation 12 years, POx 128µmol/L at the beginning of lumasiran at 17 years, KTx 17 months after lumasiran (POx 23µmol/L), deceased donor. No delayed graft function. Post-operative management: SOC. At one month, renal function 50mL/min/1.73 m², POx 28µmol/L, UOx/creat 245µmol/mmol (<80). Arterial thrombosis post-lymphocele 41 days post KTx, requiring 13 daily hemodialysis sessions. Follow-up 3 months, renal function 50mL/min/1.73m² and stable UOx/creat.

Conclusions: We report the first successful isolated KTx in PH1 patients under lumasiran. Long-term data are obviously required. As described in CLKTx, post-operative hyperhydration and alkalinization is crucial, as long as urinary oxalate remains elevated from bone release.

OP-43 FIRST INTERIM ANALYSIS OF THE INTERNATIONAL X-LINKED HYPOPHOSPHATAEMIA (XLH) REGISTRY: PAEDIATRIC POPULATION BASELINE CHARACTERISTICS

Dieter Haffner[1], Jonathan Liu[2], Angela Williams[2], Sue Wood[2], Elena Levtchenko[3]

[1]Hannover Medical School, [2]Kyowa Kirin International, [3]University Of Leuven

Introduction: X-linked hypophosphataemia (XLH) is a rare, progressive, hereditary phosphate-wasting disorder characterised by excessive activity of fibroblast growth factor 23. The International XLH patient Registry was established to provide information on the natural history of XLH and impact of treatment on patient outcomes. Data are from the first interim analysis conducted on baseline data from paediatric subjects (age <18y).

Material and methods: The XLH Registry (NCT03193476) was initiated in August 2017, aims to recruit 1,200 children and adults with XLH, and will run for 10 years. At time of analysis (Last Patient In: 30/11/2020; database lock: 29/03/2021), subjects diagnosed with XLH were enrolled from 81 hospital sites in 16 countries. Parameters collected at baseline included demographics, medical and treatment history, and clinical presentation data.

Results: Overall, 360 children were included in this analysis; 61.7% were female. Mean (SD) age was 9.5y (±4.5). Treatment data at entry were available for 281 subjects: among those 58.7% were receiving burosumab (165/281); 40.6% conventional therapy (phosphate salts and active vitamin D) (114/281); 0.7% had no treatment reported (2/281). Among 330 paediatric subjects with reported genetic data, 72.4% had a genetic test, of whom 88.7% had a confirmed PHEX mutation. Data on XLH family history were available for 319 subjects; biological mother was affected in 164 (51.4%); biological father was affected in 49/317 (15.5%). Across all age groups males and females with XLH presented with decreased standardized height (z-score: -1.63±1.22; n=307) yet 'normal' standardized weight (z-score: -0.02±1.35; n=326), resulting in an elevated body mass index (z-score: 1.37±2.285; n=249) suggesting obesity.

Conclusions: This is the largest data set of children with XLH collected to date. Short stature and obesity appear as frequent complications throughout childhood. Information collected over the 10-year Registry duration will generate real-world evidence to help inform clinical practice.

Authors acknowledge the contribution of all XLH Registry Steering Committee members.

OP-44 DECIPHERING THE IMMUNOLOGICAL MECHANISMS UNDERLYING PEDIATRIC IDIOPATHIC NEPHROTIC SYNDROME

Giulia Cricri[1], Linda Bellucci[2], Stefania Bruno[3], Federico Caicci[4], Stefano Turolo[5], William Morello[5], Giovanni Montini[5], Federica Collino[1]

[1]Department Of Clinical Sciences And Community Health, University Of Milano, Milan., [2]Laboratory Of Translational Research In Paediatric Nephro-urology, Fondazione Ca Granda Irccs Ospedale Maggiore Policlinico, Milan, [3]Department Of Medical Sciences, University Of Turin, Turin, [4]Department Of Biomedical Sciences, University Of

Padova, Padua., [5]*Pediatric Nephrology, Dialysis And Transplant Unit, Fondazione Ca Granda Irccs Ospedale Maggiore Policlinico, Milan.*

Introduction: Idiopathic Nephrotic Syndrome (NS) is the most common glomerular disease in children, with immune-related pathogenesis. Corticosteroids are the first-line treatment, but patients not responsive may progress to end-stage renal disease. The identification of a biomarker signature to predict treatment efficacy will be useful to redefine INS patients' classification based on individual immune profiles, rather than on response to treatment.
Material and methods: PBMCs, sera and urines were obtained from thirty INS children (steroid-sensitive and resistant) at different time-points (onset, remission, relapse) and seven young healthy donors (HS). The dynamics of the changes in the immune repertoire were detected by flow cytometry. Extracellular vesicles (EVs) concentration and morphology were analysed by Nanoparticle Tracking Analysis and Transmission Electron Microscopy. Molecular analyses (lipidomic and proteomics) were conducted on Size Exclusion Chromatography isolated EVs.
Results: No significant differences were observed in the total CD19+ B lymphocytes in INS children with active proteinuria in respect to the HS, whereas their levels were significantly enhanced compared with remission state. INS patients show higher levels of naïve and switched memory B cells compared to HS (p <0.05), that were maintained elevated during remission. Focused on the steroid-sensitive INS group, transitional B cells decreased and plasmablasts and plasma cells increased in these patients compared to the same group in remission (p <0.05). In the T cell compartment, we detected a significant increase of Th2 and Th17/Th1 ratio in proteinuric INS in respect to HS, and higher levels of Foxp3+ T reg during remission (p <0.05). NTA analysis revealed no differences in EV concentration. Biofluid INS-EVs expressed tetraspanins with a heterogeneous distribution among the patients' subgroups; the highest expression was observed during remission. Serum INS-EVs showed an increase in lymphocytic markers, that were already detected in EVs at the disease onset. Urine INS-EVs were enriched of adhesion, epithelial and leucocyte molecules, and their number positively correlated with proteinuria levels.
Conclusions: Changes in the biofluid EV profile embody the bloodstream immune dysfunctions, supporting their possible participation in INS pathophysiology.

OP-45 HIGH PHOSPHATE LOAD-INDUCED PROXIMAL TUBULAR INJURY IS ASSOCIATED WITH ACTIVATED STAT3/KIM-1 SIGNALING AND MACROPHAGE RECRUITMENT

Beatrice Richter[1], Tamar Kapanadze[2], Nina Weingärtner[1], Stefanie Walter[1], Isabel Vogt[1], Andrea Grund[1], Jessica Schmitz[3], Jan Hinrich Bräsen[3], Florian P. Limbourg[2], Dieter Haffner[1], Maren Leifheit-nestler[1]

[1]*Department Of Pediatric Kidney, Liver And Metabolic Diseases, Pediatric Research Center, Hannover Medical School, Hannover, Germany*, [2]*Department Of Nephrology And Hypertension, Hannover Medical School, Hannover, Germany*, [3]*Institute Of Pathology, Nephropathology Unit, Hannover Medical School, Hannover, Germany*

Introduction: High phosphate is linked to enhanced all-cause mortality and responsible for the progression of kidney damage in patients with chronic kidney disease. However, it is unclear whether a high phosphate diet (HPD) causes kidney injury in healthy individuals.
Material and methods: C57BL/6N mice were either fed with a 2% HPD or a 0.8% normal phosphate diet (NPD) for one up to six months and investigated for parameters of phosphate homeostasis and development of kidney injury. The specific impact of phosphate and its phosphaturic hormones fibroblast growth factor (FGF) 23 and parathyroid hormone (PTH) on tubular cell damage was investigated using HK-2 human proximal tubular (PT) cells.
Results: HPD in mice caused hyperphosphatemia, hyperphosphaturia, increased FGF23, PTH and creatinine levels and albuminuria. Histopathological analysis revealed progressive PT injury in HPD-fed mice from two months onwards followed by a rapid progression from month 5 to 6. This was accompanied by increased tubulointerstitial fibrosis. The kidney injury marker Kim-1 increased in PT of HPD-fed mice, which was positively associated with tubular injury score and tubulointerstitial fibrosis. Histological staining revealed increased number of $pStat3^+$ cells that was positively associated with Kim-1 synthesis. Concomittantly, the chemokine MCP-1 was upregulated in PT of HPD-fed mice, which was associated with Kim-1 expression indicating an interaction between pStat3/Kim-1 signaling and MCP-1. HPD caused enhanced macrophage recruitment to injured PT that was associated with increased MCP-1 synthesis and increased tubular injury score. Stimulation of HK-2 cells with FGF23 or high phosphate, but not PTH, induced the phosphorylation of Stat3. Interestingly, only high phosphate significantly upregulated Kim-1 and MCP-1 expression.
Conclusions: Chronic high phosphate load leads to progressive PT damage caused by Stat3/Kim-1 signaling activation mediating MCP-1-dependent macrophage recruitment. Our data indicate that high phosphate intake may cause a global health problem and the demand for clinical studies on this issue.

OP-46 IS THER ANY ROLE OF THE URINE DICKOPPF-3/ CREATININE RATIO IN EARLY DETECTION OF ACUTE KIDNEY INJURY IN PEDIATRIC INTENSIVE CARE UNIT?

Sefa Armağan Gökçeli[1], Neslihan Gunay[1], Inayet Güntürk[2], Mehmet Akif Dündar[3], Başak Nur Akyıldız[3], Cevat Yazıcı[4], Sibel Yel[1], M.hakan Poyrazoğlu[1], Ismail Dursun[1]

[1]*Erciyes University Faculty Of Medicine, Department Of Pediatrics, Division Of Nephrology, Kayserİ, Turkiye*, [2]*Department Of Midwifery, School Of Health, Niğde Omer Halisdemir University*, [3]*Erciyes University Faculty Of Medicine, Department Of Pediatrics, Division Of Intensive Care*, [4]*Erciyes University Faculty Of Medicine, Department Of Biochemistry*

Introduction: Acute kidney injury (AKI) is a common complication in the pediatric intensive care unit (PICU) and there are many studies on new biomarkers to predict AKI earlier than serum creatinine (SCr). Recently, few studies have focused on urinary DKK-3, which may be an early biomarker of AKI. In this study, we investigate the role of urinary DDK-3 in early prediction of AKI in patients hospitalized in PICUAcute kidney injury (AKI) is a common complication in the pediatric intensive care unit (PICU) and there are many studies on new biomarkers to predict AKI earlier than serum creatinine (SCr). Recently, few studies have focused on urinary DKK-3, which may be an early biomarker of AKI. In this study, we investigate the role of urinary DDK-3 in early prediction of AKI in patients hospitalized in PICU
Material and methods: In this prospective study, between June 2020 and April 2021, 117 patients who stayed in PICU for at least 48 hours were included. On admission, PRISM, PELOD, "Vasoactive inotrope Score" of patients using vasopressors were also noted. We measured urine DKK-3, creatinine, micro protein, serum creatinine and calculated micro protein to creatinine ratio (PCR), eGFR and urine DKK3/cre ratio. From admission to 10^{th} day of hospitalization or death, patients were followed-up with serum creatinine. AKI was defined based on KDIGO 2012 criteria. The impact of the urine DKK3/Cr on the development of AKI and mortality, and its sensitivity and specificity was investigated.

Results: AKI developed in 42 of the patients. The most common reasons were malignancy and cardiac disease for AKI. Respiratory failure and postoperative cardiac surgery were the most common reasons for hospitalization in PICU. Urine DKK3/cre ratio was found to be higher in patients who developed AKI compared to those who did not develop KDIGO SCr. Urine DKK3/cre ratio was found to be higher in patients with stage 3 AKI than those with stage 1 and stage 2 AKI. The power of urinary DKK3/crea ratio to predict the development of AKI in patients hospitalized in the pediatric intensive care unit was 73.2% by AUROC analysis. The urine DKK3/cre cut-off value for the detection of AKI was 63311 pg/mg, the sensitivity was 23.8% and the specificity was 94.6%. AKI was observed more frequently in patients with nephrotic proteinuria on the first day of hospitalization in the pediatric intensive care unit compared to those without. ***Urine DKK3 was found to be a risk factor both AKI and mortality in univariate and multivariate logistic regression analysis (Table 1,2). The DKK3 value above the cut-off point increases the risk of developing AKI by 5.547 times and mortality by 5.569 times (Table3,4).***

Table 1: Logistic regression analysis of risk factors for AKI

Variables	Univariate regression			Multivariate regression		
	OR	95% CI	p	AOR	95% CI	p
Age (months)	0.998	0.992-1.004	0.606			
Gender	1.190	0.552-2.564	0.658			
Comorbidite	1.986	0.868-4.541	0.104			
Hospital stay duration	0.777	0.652-0.927	**0.005**	0.758	0.610-0.944	**0.013**
MV duration time	1.090	0.988-1.202	0.084			
GKS	0.898	0.799-1.010	0.073			
PRISM	1.070	1.019-1.123	**0.007**	1.082	0.990-1.183	0.081
PELOD	1.130	1.049-1.217	**0.001**	1.021	0.960-1.085	0.510
YIS	5.190	0.000-4.815	0.990			
Log10 Dkk3/cr	4.970	2.186-11.302	**<0.001**	4.367	1.762-10.823	**0.001**

PRISM, *Pediatric Risk of Mortality*, **PELOD**, *Pediatric Logistic Organ Dysfunction*, **MV**; *Mechanic ventilation*, **GSK**, *Glasgow Coma Scale*, **YIS**; *Vasoactive-Inotropic Score*, **AKI**, *Acute kidney injury*

Table 2: Logistic regression analysis of risk factors for mortality

Variables	Univariate regression			Multivariate regression		
	OR	95% CI	p	AOR	95% CI	p
Age (months)	0.998	0.992-1.004	0.546			
Gender	1.236	0.567-2.698	0.594			
Co-morbidity	3.721	1.466-9.446	**0.006**	3.690	0.827-16.456	0.087
Hospital stay duration	0.743	0.620-0.890	**0.001**	0.631	0.446-0.892	**0.009**
MV duration time	1.240	1.114-1.380	**<0.001**	1.196	1.002-1.429	**0.048**
GKS	0.842	0.743-0.954	**0.007**	1.210	0.879-1.666	0.242
PRISM	1.121	1.043-1.208	**0.002**	1.036	0.910-1.178	0.594
PELOD	1.116	1.056-1.179	**<0.001**	1.091	0.980-1.215	0.113
YIS	1.085	1.037-1.136	**<0.001**	1.055	1.010-1.101	**0.016**
Log10 Dkk3/cr	6.423	2.283-16.917	**<0.001**	3.615	1.041-12.550	**0.043**

PRISM, *Pediatric Risk of Mortality*, **PELOD**; *Pediatric Logistic Organ Dysfunction*, **MV**; *Mechanic ventilation*, **GSK**; *Glasgow Coma Scale*, **YIS**; *Vasoactive-Inotropic Score*

Table 3. Urine DKK3/Cr and AKI

Variables	Univariate regression		
	OR	95% CI	p
DKK3/Cr (cut of 63311)	5.547	1.618-19.022	0.008

Table 4. Urine DKK3/Cr and mortality

Variables	Univariate regression		
	OR	95% CI	p
DKK3/Cr (cut of 88963)	5.569	1.329-22.499	0.019

Conclusions

Urine DKK3/cre ratio is a clinically useful biomarker in predicting the development of AKI according to KDIGO SCr in patients hospitalized in PICU. In patients with nephrotic level proteinuria in spot urine, AKI develops more frequently and the urinary DKK3/cre ratio is found to be higher. ***A high level of urine DKK3 is a risk factor for both AKI and mortality in children in PICU.***

OP-47 PERSISTENT MARKERS OF RENAL INJURY FOLLOWING CARDIAC SURGERY-RELATED ACUTE KIDNEY INJURY IN CHILDHOOD: A PROSPECTIVE COHORT STUDY

Jef Van Den Eynde[1], Thomas Salaets[2], Jacoba Louw[3], Jean Herman[4], Luc Breysem[5], Dirk Vlasselaers[6], Lars Desmet[6], Bart Meyns[7], Werner Budts[8], Marc Gewillig[2], Djalila Mekahli[9]

[1]*Department Of Cardiovascular Sciences, Ku Leuven, Leuven, Belgium.*, [2]*Pediatric Cardiology, University Hospitals Leuven, Leuven, Belgium.*, [3]*Pediatric Cardiology, Maastricht University Medical Centre, Maastricht, The Netherlands.*, [4]*Department Of Pediatric Nephrology, University Hospitals Leuven, Leuven, Belgium.*, [5]*Department Of Radiology, University Hospitals Leuven, Leuven, Belgium.*, [6]*Department Of Intensive Care Medicine, University Hospitals Leuven, Leuven, Belgium.*, [7]*Department Of Cardiovascular Diseases, Unit Of Cardiac Surgery, University Hospitals Leuven, Leuven, Belgium.*, [8]*Congenital And Structural Cardiology, University Hospitals Leuven, Leuven, Belgium.*, [9]*Pkd Research Group, Gpure, Department Of Development And Regeneration, Ku Leuven, Leuven, Belgium.*

Introduction: This prospective cohort study investigated the long-term renal consequences and prevalence of chronic kidney disease (CKD) following acute kidney injury (AKI) after pediatric cardiac surgery for congenital heart disease (CHD).

Material and methods: All eligible children (<16 years) who had developed AKI following cardiac surgery at our center between December 2004 and December 2008 were prospectively invited for a formal renal assessment 5 years after AKI. Longer follow-up data on renal function (>10 years after AKI) were collected at latest available visit.

Results: Among 571 patients operated over a 4-year period, AKI occurred in 113 (19.7%). Fifteen of these (13.3%) died at a median of 31 days (interquartile range, IQR 9-57) after surgery. A total of 66 patients participated in the kidney assessment at a median of 4.8 years (IQR 3.9-5.7) after the index AKI episode. Thirty-nine patients (59.1%) had at least one marker of kidney injury, including estimated glomerular filtration rate (eGFR) <90mL/min/1.73m² in 9 (13.6%), proteinuria in 27 (40.9%), alpha-1-microglobinuria in 5 (7.6%), hypertension in 13 (19.7%), and abnormalities on kidney ultrasound in 9 (13.6%). CKD stages 1-5 was present in 18 (27.3%). CKD was associated with syndromes (55.6% vs 20.8%, p=0.015). At 13.1 years (IQR 11.2-14.0) follow-up, eGFR <90mL/min/1.73m² was present in 18/49 patients (36.7%), suggesting an average eGFR decline rate of -1.81 mL/min/1.73m² per year.

Conclusions: Children who developed AKI after pediatric cardiac surgery show persistent markers of renal injury. Detection and treatment of CKD, proteinuria, and hypertension in children is critical because these are risk factors for cardiovascular diseases and progressive renal damage in adults. Prevention of renal disease should start in childhood to ensure optimal outcomes in the growing population of adults with CHD.

OP-48 EFFICACY AND SAFETY OF REGIONAL CITRATE ANTICOAGULATION IN PCKRT

Andrea Cappoli Raffaella Labbadia[1], Emanuele Rossetti[2], Gabriella Bottari[2], Isabella Guzzo[1]

[1]*Division Of Nephrology And Dialysis, Department Of Pediatric Subspecialties, Irccs Bambino Gesù Pediatric Hospital, Rome Italy*, [2]*Department Of Pediatric Intensive Care, Ospedale Pediatrico Bambino Gesù-ircc, Rome, Italy*

Introduction: Acute kidney injury (AKI) is highly prevalent in hospitalized children, especially those in pediatric intensive care unit (PICU). Continuous kidney replacement therapy (CKRT) is the treatment of choice in critically ill children with AKI. One of the great challenges with CKRT is early coagulation of the circuit.

Material and methods: Since December 2018, we have treated fifty critically ill children admitted in pediatric intensive care units with CKRT using regional citrate anticoagulation. Diagnosis for these patients were several, including septic shock, acute kidney injury, liver failure and hematological disorders. Patients were mainly male, mean age was 75.4 ± 77.5 months with mean body weight of 22.7 ± 22.8 kg. Twenty-seven patients were below 15 kg and fourteen patients were below 10 kg. Eleven patients had severe hepatic impairment and received a reduced dose of citrate.

Results: The mean filter lifetime was 54.5 ± 18.2 hours with 70.6% of circuits lasting more than 48 hours and 45.2% of circuits lasting more than 70 hours. The most frequent cause of CKRT interruption was scheduled changes. Interestingly, similar circuit lifetime was also confirmed in the low weight sub-population (≤ 10 kg), 54.3 ± 19.2 hours. In our experience among metabolic complication, in contrast with the adult population metabolic acidosis was more common than metabolic alkalosis. We found no case of citrate accumulation, even in those patient with hepatic failure.

Conclusions: In our experience regional citrate anticoagulation with commercially available solutions was easy to apply, safe and effective in preventing circuit clotting. We conclude that regional citrate anticoagulation can be used for CKRT in children, even in the ones with very low body weight or with liver failure.

PITCH POSTERS

PI-1 PRETERM BIRTH, HYPERTENSION AND ARTERIAL STIFFNESS IN CHILDREN AND ADOLESCENTS

Athanasia Chainoglou[1], Kosmas Sarafidis[2], Katerina Chrysaidou[1], Evangelia Farmaki[1], Konstantinos Kollios[3], Marina Economou[1], Vasilios Kotsis[4], Stella Stabouli[1]

[1]1st Department Of Pediatrics, General Hospital Of Thessaloniki "hippokratio", Greece, [2]1st Department Of Neonatology, Genaral Hospital Of Thessaloniki "hippokratio", Greece, [3]3rd Department Of Pediatrics, General Hospital Of Thessaloniki "hippokratio", Greece, [4]3rd Department Of Internal Medicine, General Hospital Of Thessaloniki "papageorgiou", Greece

Introduction: According to the recent literature, preterm birth is associated with increased risk of developing chronic kidney disease and arterial hypertension in future life. The aim of the current study was to investigate the association between preterm birth and ambulatory blood pressure (BP) levels and arterial stiffness during childhood and adolescence.
Material and methods: The sample of this prospective case control study included 52 children and adolescents born preterm and 26 children born full-term. All of the participants underwent measurement of somatometric characteristics, office BP, central systolic BP, ambulatory BP monitoring (ABPM) and assessment of carotid-femoral pulse wave velocity (PWV).
Results: Overall, in the study participated 52 preterm with mean age 10,4 ±3,8 years and 22 full-term children with mean age 11,2±3,2 years. Preterm children presented significantly higher night systolic blood pressure (SBP) z score values compared to controls ($p<0.05$). Preterm children who were overweight/obese presented significantly higher values of night SBP compared to other subgroups. Preterm children also presented marginally higher PWV z score values ($p=0.06$). Predictors of night SBP z score were preterm birth and BMI z score, whereas prematurity was an independent predictor of PWV z score.
Conclusions: Our study's findings support that preterm birth is associated with higher nocturnal BP and increased arterial stiffness in childhood and adolescence. The early diagnosis of hypertension and the prevention of obesity during childhood are important in the prevention of future adverse health outcomes in individuals born preterm.

PI-2 PREVALENCE OF LEFT VENTRICULAR HYPERTROPHY IN CHILDREN AND YOUNG ADULTS WITH CHRONIC KIDNEY DISEASE AND MASKED HYPERTENSION: A SYSTEMATIC REVIEW AND META-ANALYSIS

Ioannis Goulas[1], Kleo Evripidou[1], Ioannis Doundoulakis[2], Athanasia Chainoglou[1], Thomai Nika[1], Stella Stabouli[1]

[1]1st Department Of Pediatrics, School Of Medicine, Faculty Of Health Sciences, Aristotle University Thessaloniki, Hippokratio Hospital, Thessaloniki, Greece, [2]Department Of Cardiology, 424 General Military Training Hospital, Thessaloniki, Greece

Introduction: Undetected hypertension (HTN) in children with chronic kidney disease (CKD) may result in undertreatment of patients with subclinical TOD. In this meta-analysis we aimed to investigate the prevalence of left ventricular hypertrophy (LVH) in children and young adults with CKD and hypertensive ambulatory BP phenotypes, sustained and masked HTN.
Material and methods: We assessed studies that included children, adolescents, and young adults with CKD and reported data on echocardiography as well as BP levels by both office and ambulatory BP monitoring. Only studies using ambulatory BP monitoring were included, in order to prevent misclassification of BP status.
Results: The systematic literature search retrieved 1283 studies. We finally included 3 studies with a mean participants' age 13.79 ± 4.35 years. Among three studies that reported data on the overall prevalence of LVH in a total of 238 CKD patients with ambulatory hypertension, the prevalence of LVH was found 0.28 (95% CI, 0.19, 0.39) . In four studies (including the three studies mentioned) including 172 CKD patients with masked hypertension, LVH was detected in 49 patients, with the estimated prevalence being 0.23 (95%CI, 0.15, 0.32).
Conclusions: Our data emphasize the growing evidence on the important role of echocardiography and ambulatory BP monitoring to evaluate cardiovascular risk in children with CKD . Diagnosis of masked hypertension carries an adverse prognosis, with increased risk of LVH and associated future cardiovascular events.

PI-3 MUCH TENSION WITHOUT THE ADRENALINE: ORTHOSTATIC HYPERTENSION MIMICS PHEOCHROMOCYTOMA

Marina Avramescu, Olivia Boyer

Ap-hp Hopital Necker

Introduction: In the presence of hypertension in children, physicians should strive to find a diagnosis. Orthostatic hypertension (OHT) and postural tachycardia syndrome (PoTS) signal an abnormality in cardiovascular autonomic control mechanisms and might mimic pheochromocytoma because of the paroxysms of hyperadrenergic symptoms.
Material and methods: A 14-year old boy with a congenital solitary kidney presented with fatigue and paroxysmal headache, lightheadedness and palpitations for the past 6 months. Blood pressure (BP) was 160/80 mmHg.
Results: Complete blood count, serum electrolytes and serum creatinine were within normal reference range. Renal Doppler ultrasound was normal. He had no target-organ damage: heart ultrasound and fundoscopy were normal, proteinuria was negative. 24-h ambulatory BP monitoring found normal daytime and nighttime averages and revealed diurnal spikes of the systolic BP of more than 160 mmHg. 24-h urinary normetanephrine level was 5 times the normal amount. The patient underwent PET scanning to detect and localize the suspected pheochromocytoma, but no hypermetabolic activity was found. Further clinical examination found orthostatic tachycardia with concomitant orthostatic hypertension. Noradrenaline was measured after drawing a blood sample in both a supine and a standing position. The patient had a massive elevation of plasma noradrenaline in the standing position. 10-minute tilt and stand tests induced symptoms as well as modifications in heart rate and BP. The patient needed a triple hypotensive therapy (beta-blockers, α1-blockers, central sympatholytics) as well as non-pharmacological treatment (sophrology) in order to control symptoms.
Conclusions: PoTS and OHT are disorders of the autonomic nervous system that can produce substantial disability among otherwise healthy people. The clinical picture of PoTS and OHT can be confused with pheochromocytoma. While the latter requires urgent medical attention, PoTS and OHT are considered to be benign conditions. However, OHT in children might herald sustained arterial hypertension later in life, therefore close follow-up is needed.

PI-4 CARDIOVASCULAR RISK FACTORS IN OBESE CHILDREN. DETERMINATION BY CASE-CONTROL DESIGN OF CAROTID INTIMA MEDIA THICKNESS BY ULTRASOUND AS A PREDICTOR OF CARDIOVASCULAR RISK

Maria Cristina Ontoria Betancort[1], Maria Teresa Rodriguez Bello[1], Carlos Marichal Hernandez[1], Jorge De Luis Yanes[2], Maria Isabel Luis Yanes[1], Victor Manuel Garcia Nieto[1]

[1]Hospital Universitario Ntra Señora De La Candelaria, Santa Cruz De Tenerife, Spain, [2]Hospital Universitario Gregorio Marañon.madrid

Introduction: Obesity is a major childhood health problem. Central or abdominal obesity has been linked to various complications, such as high blood pressure, dyslipidemia, insulin resistance and diabetes mellitus, constituting the so-called "metabolic syndrome". Early ultrasound study of carotid intima media thickness (CIMT) could detect subclinical atherosclerotic lesions. In the pediatric age, studies related to obesity, cardiovascular risk factors and atherosclerosis are limited.To determine the CIMT of both common carotids as a marker of subclinical atherosclerosis by ultrasound measurement in obese children and children with a normal body mass index (BMI) and to determine its relationship with variables related to cardiovascular risk.
Material and methods: Observational case-control study. We studied 69 children of both sexes, between 6 and 14 years old, with a diagnosis of obesity or body mass index (BMI) higher than +2 SD of the mean according to the reference population. A control group of 76 healthy children with similar characteristics was assigned. A history of cardiovascular risk in first-degree relatives was analyzed. In both study groups, somatometry was performed with determination of BMI and waist circumference and complete physical examination with measurement of blood pressure, as well as the determination of analytical parameters related to childhood obesity. Obese children underwent oral glucose overload and abdominal ultrasound and, for all children, carotid ultrasound to determine CIMT
Results: Obese children showed significantly ($p<0.001$) a higher frequency of high blood pressure, hypertriglyceridemia, decreased HDL-c and insulin resistance compared to non-obese children.The mean CIMT was higher in obese children (0.46±0.09 for the right carotid and 0.45 ±0.09 for the left carotid) than in healthy children (0.33±0.04 on the right and 0.36±0.04 on the left) ($p<0.001$). These differences were present since prepubertal times (<0.001). Of these, BMI was the variable that was independently associated with the CIMT of both carotids ($p< 0.001$). The right CIMT was higher in obese patients with hyperuricemia ($p=0.039$) and insulin resistance ($p=0.01$).. Children with hepatic steatosis (50% of obese patients) had a higher CIMT for both carotids ($p=0.003$) compared to obese children with normal liver ultrasounds. Likewise, the presence of hepatic steatosis was independently associated with increased thickness of the right ($p=0.002$) and left ($p=0.001$) intima media.
Conclusions: The obese subjects in the sample presented a higher frequency of parameters associated with cardiovascular risk. The thickness of the intima media of both carotids was significantly higher in both prepubertal and pubescent obese children relative to non-obese children.The thickness of the intima media of both carotids was directly correlated with multiple variables related to cardiovascular risk, with BMI and the presence of hepatic steatosis being the variables that showed an independent association with the CIMT of both carotid arteries.

PI-5 PHARMACOLOGICAL TREATMENT OF ARTERIAL HYPERTENSION IN CHILDREN AND ADOLESCENTS IN LITHUANIA

Dovile Ruzgiene[1], Eleonora Ivanova[2], Augustina Jankauskiene[1]

[1]Pediatric Center, Institute Of Clinical Medicine, Vilnius University, [2]Vilnius University, Faculty Of Medicine

Introduction: The prevalence of arterial hypertension (AH) in pediatric population in Europe ranges from 2.2% to 13% and even more in overweight or obese children. Part of these patients receives pharmacological treatment. The aim of the study was to evaluate the pharmacological treatment of children and adolescents in Lithuania and to compare the treatment choices in 2015 – before the release of 2016 European Society of Hypertension guidelines for the management of high blood pressure (BP) in children and adolescents and after – in 2019.
Material and methods: Big data was extracted from Lithuanian National Health Insurance Fund database for prescriptions covered by insurance for children from 0 to 17 years of age in the year of 2015 and 2019 for primary and secondary AH. Patients were divided in 0-3, 4-7, 8-12, 12-17 age groups. Antihypertensive medications were grouped by their pharmacological class: angiotensin converting enzyme inhibitors (ACEi), angiotensin receptor blockers (ARBs), beta blockers (BB), calcium channel blockers (CCB), diuretics, imidazoline receptor agonists, alpha-blockers, combined ACEi and diuretics.
Results: 1382 prescriptions were done in 2015 and 1233 in 2019 for AH. ACEi were the first choice for primary and secondary AH in all age groups, while BB were in the second place. ARBs were the third or the fourth choice depending on the age group. Enalapril was the most common in ACEi group, while Metoprolol in BB. The pharmacological treatment options did not differ in compared years.
Conclusions: BB are used more often as antihypertensive agents than they should be, while ARBs are underused. There was no difference between primary and secondary AH treatment. No difference in pharmacological AH treatment was made after the release of 2016 BP guidelines.

PI-6 FONTAN ASSOCIATED NEPHROPATHY: DOES DIASTOLIC FUNCTION PLAY A ROLE?

Nicola Bertazza Partigiani[1], Roberta Biffanti[2], Jolanda Sabatino[2], Germana Longo[1], Elisa Benetti[1], Mattia Parolin[1], Marta Rotella[2], Beatrice Binda[4], Massimo Padalino[3], Davide Meneghesso[1]

[1]Pediatric Nephrology And Dialysis Unit, Department Of Women's And Child's Health, University Of Padova Medical School, 35128 Padova, Italy, [2]Paediatric Cardiology, Department Of Women's And Child's Health, University Of Padova Medical School, 35128 Padova, Italy, [3]Pediatric And Congenital Cardiac Surgery Unit, Department Of Cardiac, Thoracic And Vascular Sciences And Public Health, University Of Padova Medical School, 35128 Padova, Italy, [4]University Of Padova Medical School, 35128 Padova, Italy

Introduction: We investigate the long term nephrological outcomes, the physiopathology of kidney involvement and its relationship with cardiac diastolic function in Fontan patients.
Material and methods: This is a prospective study including patients who underwent Fontan completion in our Centre between 1993 and 2016. We excluded patients with major congenital renal anomalies, those who underwent cardiac transplantation and redo-Fontan patients. Patients underwent clinical evaluation, laboratory exams with renal function, kidney US and complete cardiac evaluation.
Results: We enrolled 35 patients, 46% female (N=16), and 54% male (N= 19). Medium age was 17 y.o., (range 10-31 y.o.). Medium time from Fontan completion was 160 months (range 57-340 months). Ten patients had a functional single left ventricle (FSLV, 28,5%) and 21 a functional single right ventricle (FSRV, 60%); 4 patients had an undetermined single ventricle (11,5%). Data from renal function assessment showed 26% of patients with stage 2 CKD (eGFR 60-89 ml/min/1,73mq) and only one

with stage 3 CKD, using cystatin C based equation. Most of the patients with CKD were FSRV (89%). Erased beta-2-microglobulin levels were present in 4 patients. Echocardiographic evaluation of diastolic function showed 2 patients with baseline E/A < 1 (6%, tot N=33) and 11/33 (33%) pts with abnormal E/E' (>12). A statistical relationship between diastolic parameters (E/E') and tubular damage is found (Pearson's R 0,4 and 0,48, respectively, p<0,05). Diastolic function appeared to be associated also with glomerular filtration, with direct correlation between diastolic pulmonary wave deceleration time and creatinine value (Pearson's R 0,49, p<0,05).

Conclusions: Fontan related nephropathy is associated with worsening diastolic function, which was more represented in FSRV patients. Those data suggest renal function should be closely monitored in patients with impaired diastolic function.

PI-7 DEVELOPING A MOBILE APP TO FACILITATE SELF-MANAGEMENT IN PEDIATRIC CHRONIC KIDNEY DISEASE: NEPHROGO

Giedrė Žulpaitė[1], Karolis Vyčius[3], Justė Parnarauskienė[2], Karolis Ažukaitis[2], Augustina Jankauskienė[2]

[1]Faculty Of Medicine, Vilnius University, Vilnius, Lithuania, [2]Clinic Of Pediatrics, Institute Of Clinical Medicine, Faculty Of Medicine, Vilnius University, Vilnius, Lithuania, [3]Ministry Of National Defence Republic Of Lithuania Project 'create Lithuania'

Introduction: Self-management interventions for chronic kidney disease (CKD) have been related to improved control of CKD complications in adults. Mobile apps may serve as simple, time-saving and cheap tool to facilitate self-management in CKD patients and may be an attractive option in the pediatric population. We aimed to create a mobile app for pediatric CKD patients dedicated to monitor patient-reported CKD-specific health status and to improve self-care.

Material and methods: Following the analysis of scientific literature on self-management interventions for CKD patients, a mobile app NephroGo (Android and iOS compatible) was developed using Python-based Django WEB system and Flutter technology. The adult version was updated to address pediatric CKD issues. Nutrition guidance was based on the updated Pediatric Renal Nutrition Taskforce Recommendations and a personalized counter of nutrients with a database of food composition was created.

Results: Health status tracker helps to conveniently store data on blood pressure, urine output, weight, height/length and to monitor the dynamics of well-being. The personalized counter of nutrients and nutrition tracking feature helps patients to find out how much nutrients they have consumed and helps to follow CKD-stage appropriate diet. In addition, the app allows to monitor peritoneal dialysis parameters and provides patient-relevant CKD-specific information.

Conclusions: NephroGo may be an attractive mobile health application to aid and improve self-care and CKD-specific knowledge in children with CKD and their families. Following patient and families feedback a multilingual version will be created.

PI-8 EVALUATION OF AMBULATORY BLOOD PRESSURE MONITORING IN CHILDHOOD PRIMARY HEADACHES

Asiye Bolca[1], SeÇil Arslansoyu Çamlar[2], Cemaliye BaŞaran[3], GÖkÇen Erfidan[3], Demet Alaygut[2], Fatma MutlubaŞ[2], Belde Kasap Demir[4]

[1]University Of Health Scienes Izmir Tepecik Training And Research Hospital Department Of Pediatrics, [2]University Of Health Sciences Izmir Faculty Of Medicine Department Of Pediatric Nephrology, [3]University Of Health Sciences Izmir Tepecik Training And Research Hospital Department Of Pediatric Nephrology, [4]Izmir Katip Celebi University Faculty Of Medicine Department Of Pediatric Nephrology And Rheumatology

Introduction: Primary headache, one of the most common complaints in children may have elevated blood pressure (BP) load even if the office blood pressure measurements are normal and this may have a role in the etiology of primary headache. Ambulatory Blood Pressure Monitoring (ABPM) is an effective and non-invasive tool to diagnose hypertension and identifying risk groups for cardiovascular disease. Herein, we aimed to evaluate ABPM in children followed up with Primary Headache.

Material and methods: Children aged 8-17 years, who diagnosed with primary headache and had office blood pressure below the 95th percentile were included. Age and gender matched healthy control group is also involved. The 24-hour ABPM was performed to the children in the patient and control groups Demographic data, office blood pressures and ABPM results were compared between the patient and control groups.

Results: Our study included 37 patients diagnosed with Primary Headache (30 migraine, 7 tension-type headache) and 37 healthy children in the control group. the patient and control groups were similar in case of age, gender and SDSs of body weight, height, body mass index and office blood pressure.

The ABPM data showed significantly elevated total systolic blood pressure (SBP), daytime SBP, nocturnal SBP, nocturnal systolic load, and nocturnal diastolic load in the study group compare to the control group. There was no difference between groups in terms of total diastolic blood pressure (DBP), daytime DBP, nocturnal DBP, total mean arterial pressure (MAP), daytime MAP, nocturnal MAP, daytime systolic load, daytime diastolic load, systolic dipping and diastolic dipping.

Conclusions: Children who apply to the hospital with a complaint of headache may have abnormal ABPM parameters although they have normal office blood pressure. These children may need to be evaluated to identify risk groups for hypertension and cardiovascular disease.

PI-9 HEPATIC PHENOTYPE AND COMPLICATIONS IN PATIENTS WITH AUTOSOMAL RECESSIVE POLYCYSTIC KIDNEY DISEASE (ARPKD)

Kathrin Burgmaier[1], Ilse J. Broekaert[1], Samuel Kilian[2], Benjamin Leidig[1], Anja BÜscher[3], Ismail Dursun[4], Marc Fila[5], Ibrahim Gokce[6], Nakysa Hooman[7], Matko Marlais[8], Laura Massella[9], Antonio Mastrangelo[10], Djalila Mekahli[11], Monika Miklaszewska[12], Lukasz Obrycki[13], Larisa Prikhodina[14], Bruno Ranchin[15], Lutz T. Weber[1], Elke WÜhl[16], JÖrg DÖtsch[1], Franz Schaefer[16], Max C. Liebau[17]

[1]Department Of Pediatrics, University Hospital Cologne And University Of Cologne, Faculty Of Medicine, Cologne, Germany, [2]Institute Of Medical Biometry, University Of Heidelberg, Heidelberg, Germany, [3]Department Of Pediatrics Ii, University Hospital Essen, Essen, Germany, [4]Department Of Pediatric Nephrology, Erciyes University, Faculty Of Medicine, Kayseri, Turkey, [5]Pediatric Nephrology Unit, Chu Arnaud De Villeneuve-université De Montpellier, Montpellier, France, [6]Research And Training Hospital, Division Of Pediatric Nephrology, Marmara University, Istanbul, Turkey, [7]Department Of Pediatric Nephrology, Ali-asghar Children Hospital, Ali-asghar Clinical Research Development Center (aacrdc), Iran University Of Medical Sciences, Tehran, Iran, [8]Ucl Great Ormond Street Hospital For Children Institute Of Child Health, Ucl, London, Uk, [9]Division Of Nephrology, Department Of Pediatric Subspecialties, Bambino Gesù Children's Hospital, Irccs, Rome, Italy, [10]Pediatric Nephrology, Dialysis And Transplant Unit, Fondazione Irccs Cà Granda, Ospedale Maggiore Policlinico, Milan, Italy, [11]Department Of Pediatric Nephrology, University

Hospitals Leuven And Department Of Development And Regeneration, Pkd Research Group, Ku Leuven, Leuven, Belgium, [12]*Department Of Pediatric Nephrology And Hypertension, Faculty Of Medicine, Jagiellonian University Medical College, Krakow, Poland*, [13]*The Childrens Memorial Health Institute, Warsaw, Poland*, [14]*Department Of Inherited And Acquired Kidney Diseases, Research Clinical Institute For Pediatrics N.a. Acad. Y. E. Veltishev, Pirogov Russian National Research Medical University, Moscow, Russia*, [15]*Pediatric Nephrology Unit, Hôpital Femme Mère Enfant, Hospices Civils De Lyon, Centre De Référence Maladies Rénales Rares, Bron, France*, [16]*Division Of Pediatric Nephrology, Center For Pediatrics And Adolescent Medicine, University Of Heidelberg, Heidelberg, Germany*, [17]*Department Of Pediatrics, University Hospital Cologne; Center For Molecular Medicine, University Hospital Cologne And University Of Cologne, Faculty Of Medicine, Cologne, Germany*

Introduction: Autosomal recessive polycystic kidney disease (ARPKD) is a rare but severe early-onset hepatorenal fibrocystic disease and mainly caused by variants in the PKHD1 gene. Ductal plate malformation as mandatory aspect of the disease is the pathophysiologic basis for development of congenital hepatic fibrosis, which leads to clinical signs of portal hypertension. Here, we aimed to describe the hepatic phenotype of ARPKD patients.
Material and methods: We analysed clinical datasets of 605 ARPKD patients from the ARegPKD registry study for their hepatic characteristics and complications. Portal hypertension was defined as splenomegaly on ultrasound, thrombocytopenia (<150 000/ul), proof of collateral blood flow or a substantial hepatic complication, substantial hepatic complication was defined as occurrence of variceal bleeding, presence of TIPS/surgical portosystemic shunt or isolated liver or combined liver-kidney transplantation (LTx, CLKTx).
Results: Of the 605 analyzed patients with ≥1 follow-up visit, median (Q1-Q3) age at initial diagnosis was 0.2 (0.1-1.6) years, median (Q1-Q3) age at first visit was 1.3 (0.2-7.2) years. Most patients initially presented with kidney-related symptoms, but 14/537 patients (3%) presented with gastrointestinal bleeding and 4/557 (1%) with cholangitis. The 5 (10; 20) year survival without signs of portal hypertension was 77% (55%; 25%), without substantial hepatic complication 94% (79%; 58%). In total, 42 children underwent at least one LTx or CLKTx. Median (Q1-Q3) age at first LTx was 8.7 (5.3-22.4) years and at first CLKTx 8.1 (5.4-14.6) years.
Conclusions: Only a small fraction of patients initially presented with gastrointestinal symptoms. However, 20-year-survival without signs of portal hypertension was only 25%, without substantial hepatic complication 58%, indicating that a relevant fraction of patients suffers from hepatic symptoms in childhood and adolescence. Further analyses aim to delineate early clinical or biochemical risk markers for a substantial hepatic phenotype to counsel wisely this at-risk group and to improve clinical decision-making.

PI-10 GENERATION OF ARPKD KIDNEY ORGANOIDS USING PATIENT-SPECIFIC INDUCED PLURIPOTENT STEM CELLS

Theresa Leonie Fluhr, Mansoureh Tabatabaeifar, Franz Schaefer

University Clinic Heidelberg

Introduction: Autosomal-recessive polycystic kidney disease (ARPKD) is a severe paediatric kidney disorder characterised by the early development of multi-cystic kidneys and leading to a gradual decline in kidney function. Currently, the only treatment options for the resulting end stage renal disease remain dialysis or kidney transplantation.
Patient-derived organoids grown from our two new patient-specific iPSC lines can be used as a new approach for modelling ARPKD, understanding underlying pathomechanisms and identifying cyst-modifying compounds.
Material and methods: Peripheral blood mononuclear cells (PBMCs) form two patients with different PKHD1 mutations were used to generate patient-specific induced pluripotent stem cells according to the Yamanaka protocol. The iPSCs were subsequently grown into patient-derived kidney organoids using the StemDiff Kidney Organoid Kit (Stemcell Technologies). For cyst formation, PKA signalling inside the cells was amplified by treating the organoids with forskolin according to a protocol of Low et al. Cyst development was monitored by brightfield microscopy and compared to organoids derived from wildtype iPSCs functioning as a healthy control. The development of organ-specific cell types such as podocytes, tubular epithelial cells or endothelial cells was observed via immunofluorescence staining.
Results: We were able to prove the development of glomeruli, proximal and distal tubules as well as vascular networks via immunofluorescence staining. While undergoing treatment with forskolin, cyst formation was visible in the patient-derived ARPKD organoids, whereas control organoids seemed to be nearly unaffected by the PKA induction.
Conclusions: Our patient-derived organoids are able to depict kidney organogenesis in vitro developing organ-specific structures and cell-types. Furthermore, they can be used as disease-in-a-dish models in which an ARPKD-like phenotype was inducible. This could be the basis of screening cyst-modifying compounds and developing new treatment strategies. The experimental setup is currently being adapted to enable a high throughput screening process.

PI-11 CARDIOVASCULAR ASSESSMENT IN A COHORT OF ADPKD CHILDREN

Laura Lucchetti[1], Marcello Chinali[2], Alessio Franceschini[2], Carolina Danna[2], Francesco Emma[1], Laura Massella[1]

[1]*Department Of Pediatric Nephrology, Bambino Gesù Childrens Hospital, Rome, Italy*, [2]*Department Of Pediatric Cardiology, Bambino Gesù Childrens Hospital, Rome, Italy*

Introduction: Cardiovascular events represent the first cause of morbidity and mortality in adults with autosomal dominant polycystic kidney disease (ADPKD). Hypertension occurs also in childhood, with a reported incidence ranging 15% to 44%. In 2008, M. Cadnapaphornchai et al reported increased, although not hypertrophic, left ventricular mass indexed to body surface (LVMI) in hypertensive ADPKD children. Our aim was to evaluate cardiac geometry and function in a cohort of ADPKD children.
Material and methods: Patients with a confirmed diagnosis of ADPKD and a first echocardiogram performed before the age of 18 years were enrolled in the study. Renal function, blood pressure, height and body weight were collected. Left ventricular hypertrophy (LVI) was defined as LV mass > $45g/(m^{2.16} + 0.09)$.
Results: Ninety-one patients (46M) were included in the study. The median age at the echocardiography was 11.5 years (1-23 years). Twenty-six (12M) patients started antihypertensive treatment at a median age of 10 years. All had well-controlled blood pressure at the time when echocardiography was performed. Two patients were hypertensive (treatment not yet started). The median LVMI was 33 g ± 7.6 g. No difference was observed between patients treated or not treated for antihypertension ($p= 0.771$). Two patients had LVI; both were obese. No patient had systolic or diastolic dysfunction. No correlation was observed between LVMI and blood pressure. LVMI and GFR were inversely correlated ($r=-0.54$, $p<0.01$).
Conclusions: All our children with ADPKD had normal LVMI and cardiac function, except two obese children. These data indicate that early blood pressure control can prevent median-term cardiovascular damage.

PI-12 EGFR EQUATIONS IN PEDIATRIC ADPKD PATIENTS: ONE OR THE OTHER?

Schellekens Pieter[1], Marcelien Verjans[1], Dachy Angélique[2], Peter Janssens[3], De Rechter Stéphanie[1], Breysem Luc[1], Allegaert Karel[1], Bammens Bert[1], Vennekens Rudi[4], Vermeersch Pieter[1], Pottel Hans[4], Mekahli Djalila[1]

[1]Ku Leuven/ Uz Leuven, [2]Ku Leuven / Chu Liège, [3]Vub, [4]Ku Leuven

Introduction: Pediatric ADPKD could benefit from novel disease altering therapies. However, the lack of sensitive and validated endpoints in this population renders clinical trials very challenging. We aimed to evaluate methods for the estimated glomerular filtration rate (eGFR) in order to identify the most accurate method for this population.
Material and methods: Serum creatinine (SCr) and serum Cystatin C (SCysC) were measured in a large cohort of genotyped ADPKD children with long-term follow-up. Commonly used equations for eGFR were compared for their relative performance, using the reference intervals for healthy children.
Results: We included 68 genetically confirmed ADPKD children (sex ratio 1:1) with a mean age of 10.2 years (min-max: 0-23 years) and with a mean time of follow-up of 3.6 years (1-8 years). SCr was mostly within the reference interval, regardless age and sex. The revised Schwartz formula (CKiD) showed a highly significant and clinically important decline in eGFR with aging (-3.31ml/min/1.73m^2/year, p<0.0001). The recently updated equation by the Schwartz group (CKiDU25) showed a smaller (-0.90 mL/min/1.73m²/year) but significant (p=0.001) decline in eGFR with aging and also showed a significant unexplainable sex difference (p<0.0001). SCr normalized for Q and the related FAS-SCr did not show a clear age or sex dependency (table 1). Finally, CysC based and combined equations were independent of age and sex in this paediatric ADPKD cohort.
Conclusions: The CKiD equation, the most widely used method to calculate eGFR in children, and the CKiDU25 were associated with unexpected age and sex differences in the young ADPKD population. In contrast, FAS-SCr, FAS-CysC and the combined FAS equation which are all normalized by Q, showed an age and sex independency and might therefore be more reliable to monitor kidney function in this population. This finding could help future design of the upcoming clinical trials.

Table 1. Comparison of the different eGFR equations and SCr & SCysC normalized by Q. A p-value of < 0.0023 was considered significant to account for multiple testing

Equation	Gender effect mean value F/M (p-value)	Age effect (mL/min/1.73m²/ year)	133.9 < Fr < 160.1 (mL/min/ 1.73m²)	Fr > 160.1 (mL/min/ 1.73m²)
CKiD	124.9 / 127.4 (NS)	-3.31 (p < 0.0001)	23.6%	12.0%
CKiDU25	110.0 / 124.7 (< 0.0001)	-0.90 (p = 0.0011)	14.6%	5.5%
FAS-Age	119.7 / 125.5 (p = 0.0275)	-0.61 (p = 0.0266)	17.5%	6.6%
FAS-Height	122.2 / 131.7 (p = 0.0020)	-0.98 (p = 0.0005)	20.4%	9.1%
EKFC	107.2 / 111.6 (p = 0.0008)	-0.27 (p = 0.0004)	1.1%	0.0%
LMR18	101.9 / 107.4 (p = 0.0002)	-0.45 (p = 0.0004)	0.4%	0.0%
CKDEPI40	105.3 / 112.0 (p = 0.0001)	-0.08 (NS)	0.4%	0.0%
FAS-CysC	104.7 / 101.7 (NS)	0.096 (NS)	1.8%	0.4%
Fas-Combined	112.3 / 114.0 (NS)	-0.2872 (NS)	6.18%	1.09%
Equation	**Gender effect mean F/M (p-value)**	**Age effect (mL/min/1.73m²/ year)**	**Fr < 0.80 (mg/dL/year)**	**Fr < 0.67 (mg/dL/year)**
SCr/Q	0.93 / 0.88 (p = 0.0068)	+0.0044 (p = 0.0296)	17.1%	6.9%
CysC/Q'	1.06 / 1.07 (NS)	-0.001 (NS)	1.8%	1.5%

F: female, M: male, Fr: fraction, NS: not statistically significant.

PI-13 CYSTIC KIDNEY DISEASE IN ALPORT SYNDROME: ONE NEPHROLOGY CENTER DATA

Marina Aksenova, Natalia Konkova, *Marina Dobrynina*, *Olga Chumak*

Y.veltischev Research And Clinical Institute For Pediatrics, N.pirogov Russian National Research Medical University, Moscow, Russia

Introduction: It is suggested that renal cysts may be an additional incompletely penetrant consequence of a COL4A3/A4/A5 genes mutations.The aim of the study was to determine the incidence of renal in our cohort of children with Alport syndrome (AS).
Material and methods: Demographic, genetic, laboratory (proteinuria (Pr, mg/m^2/day), eGFR (ml/min/1.73m^2), total kidney volume (TKV, cm^3/m^2)) data from 177 children (109M) with AS were analyzed. AS was diagnosed only on the basis of a family history and clinical presentation in 8 (q=0.05), confirmed morphologically in 32 (q=0.18) and by NGS-based genetic tests in 137 pts (q=0.77) including COL4A3/A4/A5 panel in 77 (56%), panel of hereditary renal diseases (HRDP) in 45 (33%) and WES in 15 (11%) pts.
Results: Renal cysts were revealed in 24 (q=0.14) children (XLAS in 23, 15M): 17 pts (q=0.1) had several (≤3) small and **7** pts (q=0.04) had bilateral small (except 1 case) multiple cysts. In 12 pts WES and HRDP did not reveal genetic variants associated with cystic kidney diseases (7 pts had COL4A3/A4/A5 genetic panel). One male with large polycystic kidneys and positive family history in absence of PKD-related genetic variants was excluded from subsequent analysis. In contrast to children without and with several renal cysts, patients with multiple bilateral cysts had lower eGFR (98±20 vs 89±29 vs 45±23 ml/min/1.73m^2, $p_{1,3}$=0.02, $p_{2,3}$=0.03) and higher Pr (73[20;405] vs 250[88;867] vs 1800[460;2400] mg/m^2/day, $p_{1,2}$=0.04, $p_{1,3}$=0.003, $p_{2,3}$=0.005). There was no significant difference in age (11±4.8 vs 14.3±2.5 vs 12.8±3.6) and TKV (196±37 vs 230±57 vs 183±92 cm^3/m^2) between the groups.
Conclusions: We found renal cysts in 14% including polycystic kidneys in 4% of pts with AS. Polycystic kidney disease in AS is associated with high Pr and GFR decreasing and may reflect the AS-nephropathy progression.

PI-14 RENAL FUNCTION IN CHILDREN WITH TUBEROUS SCLEROSIS COMPLEX

Piotr Skrzypczyk[1], Anna Maria Wabik[1], Anna Deja[1], Michal Szyszka[2], Przemyslaw Bombinski[4], Aleksandra Jakimow-kostrzewa[4], Michal Brzewski[4], Sergiusz Jozwiak[3]

[1]Department Of Pediatrics And Nephrology, Medical University Of Warsaw, [2]Department Of Pediatrics And Nephrology, Doctoral School, Medical University Of Warsaw, [3]Department Of Pediatric Neurology, Medical University Of Warsaw, [4]Department Of Pediatric Radiology, Medical University Of Warsaw

Introduction: Renal lesions are observed in many pediatric patients with tuberous sclerosis complex (TSC). Single studies indicate that hyperfiltration is common in this group of patients. Our study aimed to

evaluate renal function and frequency of hyperfiltration in children with TSC based on creatinine and cystatin C levels.
Material and methods: In 44 children with TSC (age ≥2 years, mean age 8.26±5.52 years), we evaluated estimated GFR using simplified creatinine-based Schwartz formula ($eGFR_{Cr}$) and creatinine-, urea-, and cystatin C-based Schwartz formula ($eGFR_{CrUrCys}$) [mL/min/1.73m^2]. Hyperfiltration was defined as eGFR >140 mL/min/1.73m^2. In all patients, we also analyzed clinical and biochemical data and renal lesions by measuring the largest lesion diameter (angiomyolipoma – AML and cysts).
Results: 34 (77.3%) patients had renal AMLs, including 6 with large AMLs (>30 mm) and 36 (81.8%) had renal cysts. $eGFR_{Cr}$ was from 96 to 230, mean 140.1±31.8, $eGFR_{CrUrCys}$ was from 82 to 153, mean 111.0 ±15.6 mL/min/1.73m^2. The difference between $eGFR_{Cr}$ and $eGFR_{CrUrCys}$ was from 0.18 to 88.8, mean 30.0±20.8 mL/min/1.73m^2. Hyperfiltration was found in 17 (38.6%) using $eGFR_{Cr}$ and in 2 (4.5%) using $eGFR_{CrUrCys}$. Serum cystatin C varied from 0.53 to 1.45, mean 0.84±0.20 [mg/L], and was normal in all but three (6.8%) children. There was no difference between patients with hyperfiltration (defined using $eGFR_{Cr}$) and patients with normal $eGFR_{Cr}$ in age, sex, biochemical parameters, and renal lesions. $eGFR_{Cr}$ and the difference between $eGFR_{Cr}$ and $eGFR_{CrUrCys}$ correlated with age (r=-0.387, p=0.009; r=-0.479, p<0.001) whereas there was no such correlation for $eGFR_{CrUrCys}$.
Conclusions: 1. Children with TSC have normal kidney function, even with very large kidney lesions.
2. The simplified Schwartz formula does not accurately assess renal function in children with TSC and may lead to overdiagnosis of hyperfiltration, especially in the youngest patients.
3. Cystatin C -based evaluation of eGFR should be routinely used in pediatric TSC patients.

PI-15 GALECTIN-3 AS A POTENTIAL BIOMARKER OF RENAL AND HEART INVOLVEMENT IN CHILDREN WITH TUBEROUS SCLEROSIS COMPLEX

Piotr Skrzypczyk[1], Beata Kucinska[2], Anna Maria Wabik[1], Michal Szyszka[3], Sergiusz Jozwiak[5], Przemyslaw Bombinski[4], Aleksandra Jakimow-kostrzewa[4], Michal Brzewski[4], Bozena Werner[2], Malgorzata Panczyk-tomaszewska[1]

[1]Department Of Pediatrics And Nephrology, Medical University Of Warsaw, [2]Department Of Paediatric Cardiology, Medical University Of Warsaw, [3]Department Of Pediatrics And Nephrology, Doctoral School, Medical University Of Warsaw, [4]Department Of Pediatric Radiology, Medical University Of Warsaw, [5]Department Of Pediatric Neurology, Medical University Of Warsaw

Introduction: Elevated serum galectin-3 (Gal-3) is associated with numerous kidney and cardiovascular pathologies. Overexpression of Gal-3 was found in TSC (tuberous sclerosis complex)-related tumors and in serum of TSC adult patients with lymphangioleiomyomatosis. The study aimed to analyze serum Gal-3 in pediatric TSC patients and its relation with renal and cardiovascular manifestations of the disease.
Material and methods: In 38 children with TSC (age: 8.26±5.52 years, 19 boys, 19 girls), we evaluated serum Gal-3 levels [ng/mL], anthropometric parameters, renal lesion (by ultrasound and magnetic resonance imaging), heart lesions (by echocardiography), blood pressure, and biochemical parameters. The control group (CG) consisted of 20 healthy children (age: 8.83±4.68 years, 8 boys, 12 girls).
Results: Arterial hypertension was found in 4 (10.5%), renal angiomyolipomas (AML) in 24 (63.2%), and renal cysts in 30 (79.0%) TSC children; 24 (63.2%) had heart rhabdomyomas (not hemodynamically significant), only one child had GFR <90 mL/min./1.73m^2 (77.1). TSC patients did not differ in Gal-3 compared to CG (2.80±1.59 vs. 2.72 ±1.54 [ng/mL], p=0.862). In children with TSC, Gal-3 correlated positively with systolic blood pressure Z-score (r=0.372, p=0.047) and negatively with left ventricular ejection fraction (r=-0.369, p=0.029). There was no difference in Gal-3 between patients with and without renal cysts, AMLs, and heart rhabdomyomas and no relation of Gal-3 with AML and cyst size. Gal-3 did not correlate with rhabdomyoma size, left ventricular mass index, or mitral and tricuspid annular plane systolic excursion. In CG, age was the strongest determinant of Gal-3 (r=-0.670, p=0.001) without relation of Gal-3 and blood pressure; in TSC patients, there was only trend towards negative association between Gal-3 and age (r=-0.294, p=0.073).
Conclusions: 1. In TSC patients, increase in Gal-3 might reflect cardiovascular burden, but this requires further prospective studies.
2. Serum Gal-3 is not associated with presence or size of renal and heart tumors in pediatric patients with TSC.

PI-16 PHENOTYPE ANALYSIS OF A CILIOPATHY COHORT WITH DISEASE-CAUSING VARIANTS AND RENAL INVOLVEMENT: A RETROSPECTIVE SINGLE CENTER STUDY

Lisa-marie Brislinger[1], Korbinian Maria Riedhammer[2], Aruna Marchetto[3], Moneef Shoukier[3], Peter Strotmann[4], Bärbel Lange-sperandio[5], Velibor Tasic[6], Christian W. Schaaf[1], Roman Günthner[2], Jasima Comic[1], Julia Hoefele[1]

[1]Institute Of Human Genetics, Klinikum Rechts Der Isar, Technical University Of Munich, School Of Medicine, Munich, Germany, [2]Department Of Nephrology, Klinikum Rechts Der Isar, Technical University Of Munich, School Of Medicine, Munich, Germany, [3]Pränatal-medizin München, Munich, Germany, [4]Pediatric Nephrology, Childrens Hospital, München-klinik Schwabing, Klinikum Rechts Der Isar, Technical University Of Munich, Munich, Germany, [5]Pediatric Nephrology, Dr. V. Hauner Childrens Hospital, Ludwig Maximilians University, Munich, Germany, [6]University Children's Hospital, Medical Faculty Of Skopje, Skopje, Macedonia

Introduction: Ciliopathies comprise a geno- and phenotypically complex and heterogeneous spectrum of syndromic or isolated disorders. The aim of this study was the retrospective analysis of the phenotype of individuals with disease-causing variants in ciliopathy-associated genes and renal involvement.
Material and methods: The study included 69 individuals from 55 different families with a reported disease-causing variant in a ciliopathy-associated gene and an index individual with a renal phenotype. Data was collected using a standardized questionnaire. The reported variants were reviewed using the standards of the American College of Medical Genetics and current amendments. The cohort was divided into the following groups. 1.) postnatal disease onset vs. prenatal disease onset, and 2.) < 18 years at disease onset vs. ≥ 18 years at disease onset.
Results: Individuals with disease onset < 18 years had a significantly lower median age at end-stage kidney disease than individuals ≥ 18 years (7 [IQR 3 - 11.5] years vs. 47 [31 - 64] years; p < 0.001). Furthermore, individuals with a prenatal onset of disease (24/69 individuals) vs. individuals with a postnatal onset of disease (45/69 individuals) showed a significantly higher risk for the development of extrarenal manifestations (OR = 6.9 [95% confidence interval 1.42 - 33.13]), a significantly higher risk of more than 2 extrarenal organ systems being involved (OR = 12.3 [3.46 - 43.59]), a central nervous system involvement (OR = 8.7 [2.36 - 31.93]) and skeletal anomalies (OR = 4.0 [1.14 - 14.09]).
Conclusions: The results of this study help to improve human genetic counseling by emphasizing the increased probability of a syndromic dis-

ease and a more severe progression in early-onset ciliopathy disease. Especially in prenatal counseling, this should be taken into consideration.

PI-17 AMBULATORY BLOOD PRESSURE CORRELATES WITH MRI-BASED RENAL VOLUME IN CHILDREN AND ADOLESCENTS WITH AUTOSOMAL DOMINANT POLYCYSTIC KIDNEY DISEASE

Kubra Yilmaz[1], Seha Saygili[2], Ozlem Akgun-dogan[3], Zeynep Nagehan Yuruk Yildirim[4], Rumeysa Yasemin Cicek[5], Huseyin Adil Oner[4], Bagdagul Aksu[6], Nazli Gulsum Akyel[7], Ozge Oguzhan[1], Hasan Dursun[8], Sevgi Yavuz[9], Neslihan Cicek[10], Nurver Akinci[11], Ayse Agbas[2], Ahmet Nevzat Nayir[4], Dildar Konukoglu[12], Sebuh Kurugoglu[7], Lale Sever[2], Nur Canpolat[2], Salim Caliskan[2]

[1]Istanbul University-cerrahpasa, Cerrahpasa Faculty Of Medicine, Department Of Pediatrics, Istanbul, Turkey, [2]Istanbul University-cerrahpasa, Cerrahpasa Faculty Of Medicine, Department Of Pediatric Nephrology, Istanbul, Turkey, [3]Acıbadem University School Of Medicine, Department Of Pediatrics, Division Of Pediatric Genetics, Istanbul, Turkey, [4]Istanbul University, Istanbul Faculty Of Medicine, Department Of Pediatric Nephrology, Istanbul, Turkey, [5]Istanbul Basaksehir Cam And Sakura City Hospital, Department Of Pediatric Nephrology, Istanbul, Turkey, [6]Istanbul University, Institute Of Child Health, Department Of Pediatric Basic Sciences, Istanbul, Turkey, [7]Istanbul University-cerrahpasa, Cerrahpasa Faculty Of Medicine, Department Of Pediatric Radiology, Istanbul, Turkey, [8]Istanbul Prof. Dr. Cemil Tascioglu City Hospital, Department Of Pediatric Nephrology, Istanbul, Turkey, [9]University Of Health Sciences, Istanbul Basaksehir Cam And Sakura City Hospital, Department Of Pediatric Nephrology, Istanbul, Turkey, [10]Marmara University School Of Medicine, Department Of Pediatric Nephrology, Istanbul, Turkey, [11]Koc University Hospital, Department Of Pediatric Nephrology, Istanbul, Turkey, [12]Istanbul University-cerrahpasa, Cerrahpasa Faculty Of Medicine, Department Of Biochemistry, Istanbul, Turkey

Introduction: Hypertension (HT) is a major risk factor for cardiovascular disease in patients with Autosomal Dominant Polycystic Kidney Disease (ADPKD). The aim of this study is to determine the frequency of HT in children with ADPKD and reveal the relationship between MRI-based kidney volumes, blood pressures (BP) and kidney functions.

Material and methods: This cross-sectional, single-center study included 89 patients (39 girls, 50 boys) diagnosed with ADPKD, and age- and sex-matched 27 healthy children (13 girls, 14 boys) as controls. BP was determined by both office measurements and 24-hour ambulatory BP measurements. Estimated glomerular filtration rate (eGFR) was calculated from serum creatinine and cystatin C levels using the CKiD equation. The MRI-based total kidney volumes (TKV) of the patients were measured with the manual planimetry method, and corrected by the patient's height.

Results: The median age of patients was 6.9 years, with a median follow-up of 5.2 years. 79 children had a genetically proven diagnosis, whereas 10 children were diagnosed with clinical characteristic. Mean eGFR of the children with ADPKD (112 ml/min/1.73m2) was significantly lower than the control group. None of the patients had an eGFR <90 ml/min/1.73m2. The corrected TKV of the patient group was significantly bigger than the control group. ABPM revealed ambulatory HT in 23%, masked HT in 2%, and white coat HT in 13%. 17% of the patients were using antihypertensive treatment during the evaluation. Totally, 41% of the patients were diagnosed with HT. A high 24-h MAP-SDS was independently associated only with higher levels of TKV [95%CI: 0.002-0.006; p<0.001].

Conclusions: This study revealed a significant relationship between MRI-based kidney volume and blood pressure values measured with ABPM. 24-h ABPM should be used for the evaluation of HT especially in patients with increased renal size.

PI-18 IS A CONCENTRATION OF EXTRACELLULAR DNA DETERMINED BY ETIOLOGY OF AKI?

Alexandra Gaál Kovalčíková Ľubomíra Tóthová[2], Ľubica Janovičová[2], Peter Celec[2], Ľudmila Podracká[1]

[1]Department Of Pediatrics, National Institute Of Children's Diseases And Faculty Of Medicine, Comenius University, Bratislava, Slovakia, [2]Institute Of Molecular Biomedicine, Faculty Of Medicine, Comenius University, Bratislava, Slovakia

Introduction: Despite progress in the management and treatment of acute kidney disease (AKI), it is still a life-threatening entity with increasing prevalence. Regardless the etiology of AKI, a change in conventionally used marker creatinine occurs within several hours or days after damaging insult. This might limit early diagnostics of AKI and increase the risk of irreversible kidney scarring and progression to end-stage renal disease. Recently, extracellular DNA (ecDNA) is gaining popularity as a non-specific marker of tissue damage. ecDNA is released from disintegrated cell (apoptosis, necrosis, NETosis) into various body fluids including plasma and urine. Although several studies described its increased concentrations in AKI, no study rigorously investigated ecDNA in different AKI causes. Thus, we aimed to describe ecDNA concentrations and activity of DNAse in children with different etiologies of AKI as well as in animal models.

Material and methods: In the study were included 28 children with AKI (9.67 ± 6.29) and 27 sex- and age-matched healthy controls (aged 8.57 ± 4.30 years). Patients were diagnosed with glomerulonephritis (GN – 11%), tubulo-interstitial nephritis (TIN – 40%), atypical hemolytic-uremic syndrome (aHUS – 30%) and other etiologies (19%). In the experimental part, the models of adenine nephropathy (n = 30), HUS (n = 30), ischemia-reperfusion injury (n = 14), and 5/6 nephrectomy (n = 24) were studied. Total ecDNA concentrations were determined using fluorescent method (Qubit dsDNA HS Assay Kit, Invitrogen, Carlsbad, CA, USA). Nuclear (ncDNA) and mitochondrial DNA (mtDNA) were quantified by real-time PCR.

Results: Plasma ecDNA and ncDNA were significantly higher in AKI patients compared to controls (ecDNA: 5-fold higher, p<0.001; ncDNA: 10-fold higher, p<0.05). When comparing various etiologies of AKI, plasma ecDNA was significantly increased in TIN and HUS. Interestingly, ncDNA and mtDNA did not differ between AKI causes. Urinary ncDNA was higher in AKI patients compared to controls (800-fold, p<0.05). However, there was no difference in urinary ecDNA, ncDNA and mtDNA between different causes of AKI. Further, no difference in DNase activity between patients and controls was found. In animal models, increased plasma ecDNA was observed in adenine nephropathy (5-fold), HUS (4-fold) and IRI (1.5-fold) but not in 5/6 nephrectomy. Accordingly, similar results were observed in urinary ecDNA together with decreased DNase activity in AKI animals.

Conclusions: Results of the clinical part and the animal experiment indicate that increase in ecDNA in AKI is cause dependent. The higher concentration of ecDNA observed only in tubulo-interstitial nephritis and aHUS is probably related to the development of inflammation. Limitation of this study is low number of participants in subgroups of different AKI etiologies. Further studies involving more patients should elucidate the role of inflammation in ecDNA releasing in different causes of AKI.

The work was support by the Grant Agency of Ministry of Education, Science, Research and Sport of the Slovak Republic VEGA 1/0234/18 and APVV-18-0287.

PI-19 IMPACT OF THE BODY MASS INDEX ON AMBULATORY BLOOD PRESSURE PARAMETERS IN OBESE CHILDREN AND ADOLESCENTS WITH NORMAL OFFICE BLOOD PRESSURE

Ana Kovačević, Ines Vidatić, Iva Škorić, Bernardica Valent Morić

Sestre Milosrdnice University Hospital Center

Introduction: Our goal was to investigate the influence of the degree of obesity on ambulatory blood pressure (BP) parameters in selected group of office normotensive obese children and adolescents. This study is unique because it is the first one which included only obese office normotensive patients in order to establish if the deviation in ambulatory BP parameters is proportional to the increasing body mass index (BMI).
Material and methods: In total, 119 obese patients (55 males, 46.2%) aged 7-18 years (mean 14.22 years), divided into 3 groups based on their BMI Z-score were included in our study. They all underwent ambulatory blood pressure monitoring (ABPM).
Results: This study shows that obese patients, even if office normotensive, have alterations in BP values obtained by ABPM. We found a positive correlation between systolic and diastolic BP and BMI in our patients (p 0.001), however there was no correlation between this parameters on the level of different BMI subgroups. Daytime blood pressure load correlated with rising BMI and was higher in groups II and III compared to group I ($p<0.001$). BMI category did not influence the dipping pattern in our subjects although most of our subjects (66.4%) showed non-dipping pattern for systolic BP. The difference in BPV was confirmed only for daytime systolic and diastolic values between groups I and II ($p=0.019$ and $p=0.002$, respectively).
Conclusions: Systolic and diastolic BP values obtained by ABPM in office normotensive obese children and adolescents are higher in subjects with higher BMI. Patients with increased BMI have increased daytime blood pressure variability and higher percentage of BP readings above 95th percentile. Obese patients show non-dipping pattern, independently on the rising BMI category. This study shows the importance of performing ABPM in obese, office normotensive patients, in order to promptly recognize individuals with higher cardiovascular risk and assure early intervention.

PI-20 THE SEXUAL DIMORPHISM OF KIDNEY GROWTH IN MICE AND HUMANS

Paul Vergnaud, Denise Laouari, Takuo Hirose, Marion Rabant, Mohamad Zaidan, Clement Nguyen, Martine Burtin, Christophe Legendre, Patrice Codogno, Gerard Friedlander, Dany Anglicheau, Fabiola Terzi

Institut Necker Enfants Malades

Introduction: Kidney mass and function are sexually determined but the cellular events and the molecular mechanisms involved in this dimorphism are poorly characterized. The objective of this study was to elucidate them.
Material and methods: We combined in vivo approaches with female and male mice and castration/replacement experiments with in vitro-approaches with renal tubular cells treated with dihydrotestosterone or vehicle.
Results: We showed that males exhibited kidney overgrowth from five weeks of age. This effect was organ-specific, since liver and heart weight were comparable between males and females, regardless of age. Consistently, androgen receptor was expressed in male kidney, but not in liver. In growing mice, androgens led to kidney overgrowth by first inducing a burst of cell proliferation and then an increase of cell size. Remarkably, androgens were also required to maintain cell size in adults. In fact, orchiectomy resulted in smaller kidneys in a matter of few weeks. These changes paralleled the changes of the expression of ornithine decarboxylase and cyclin D1, two known mediators of kidney growth, whereas, unexpectedly, mTORC1 and Hippo pathways did not seem to be involved. Androgens also enhanced in vivo and in vitro renal autophagy, very likely by increasing TFEB nuclear translocation. Functionally, the increase of tubular mass resulted in increased sodium/phosphate transport. These findings were relevant to humans. Indeed, by studying living gender-paired kidney donors-recipients, we showed that tubular cell size increased three months after transplantation in men as compared to women, regardless of the donor gender.
Conclusions: Collectively, these results identify novel signaling pathways that may be involved in androgens-induced kidney growth and homeostasis and suggest that androgens determine kidney size during physiological growth and after transplantation. Thus, targeting these pathways might lead to the development of novel therapeutic strategies for kidney transplantation.

PI-21 PROFOUND CONFORMATIONAL CHANGES OF SERUM ALBUMIN IN CHILDREN WITH NEPHROTIC SYNDROME DETECTED BY LIGHT SCATTERING AND ELECTRON PARAMAGNETIC RESONANCE (EPR) SPECTROSCOPY

Peter Hoyer[1], Haleh Haeri[2], Jana Eisermann[3], Heike Schimm[2], Anja BÜscher[1], Dariush Hinderberger[2]

[1]*University Duisburg - Essen, Kinderheilkunde Ii, Essen, Germany*, [2]*Institute Of Chemistry, Physical Chemistry - Complex Self-organizing Systems, Martin Luther University (mlu) Halle-wittenberg, Germany*, [3]*Department Of Chemistry - Molecular Sciences Research Hub, Imperial College London, London, United Kingdom*

Introduction: Molecular characteristics of the most abundant serum protein, albumin, in patients with nephrotic syndrome (NS) has not be in the focus of recent research. The observation of a patient with NS, refractory to all therapeutic interventions, before and after kidney transplantation clearly points toward malfunction of human serum albumin (HAS) as a remission could repeatedly be achieved by extremely high albumin infusion.
Material and methods: After excluding a mutation in the albumin encoding gene, we set out to characterize the molecular physicochemical properties of serum albumin in patients with MCNS and FSGS by employing dynamic light scattering (DLS), electrophoretic light scattering (ELS), and electron paramagnetic resonance (EPR) spectroscopy. An EPR-active stearic acid (16-DSA) was added to serum samples in in HSA:16-DSA ratios of 1:2, 1:4, and 1:6, probing mainly the high affinity (1:2) or globally up to almost all fatty acid binding sites of albumin.
Results: EPR spectroscopy has shown that fatty acids binding behavior to HSA is significantly different in diseased and healthy HSA [1, 2]. We found changes in the local environment and binding capacity of HSA in NS patients, especially pronounced in the patient with steroid resistant type of NS. These changes are correlated with variations of HSA surface potential/charge as investigated by DLS/ELS.
Conclusions: Our results show profound functional changes in serum albumin from patients with NS, like an increased hydrodynamic radius, a less negative surface potential and lower binding affinity to 16-DSA even at 1:2 ratio. Whether these changes are caused by other serum factors, loss of the binding capacity of podocyte toxic factors, or if albumin with less negative surface potential is prone to filtering through the glomerular basement membrane are subject to further research.
[1] Haeri,H.H. et al. (2019) Fatty Acid Binding to Human Serum Albumin in Blood Serum Characterized by EPR Spectroscopy. ChemistryOpen, 8, 650 - 656.

[2] Haeri H.H. et al. (2020) Identification of Patients with Pancreatic Cancer by Electron Paramagnetic Resonance Spectroscopy of Fatty Acid Binding to Human Serum Albumin. ACS Pharmacol Transl Sci. 3(6):1188-1198.

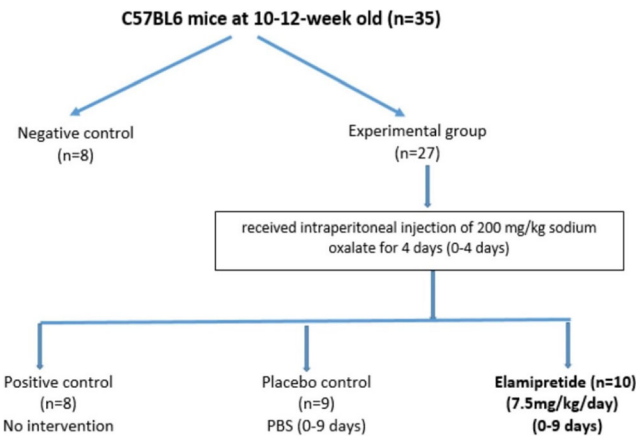
PI-22 EFFECT OF ELAMIPRETIDE ON MITOCHONDRIAL FUNCTIONS IN MICE WITH OXALATE NEPHROPATHY

Ismail Dursun[1], Eylem Taşkın Güven[2], Celal Güven[3], Züleyha Doganyigit[4], Ecma Güvenilir[1], Mehmet Memiş[1], Serpil Taheri[1], Zuhal Hamurcu[1], Meryem Şentürk[5], Hamiyet Altuntaş[6]

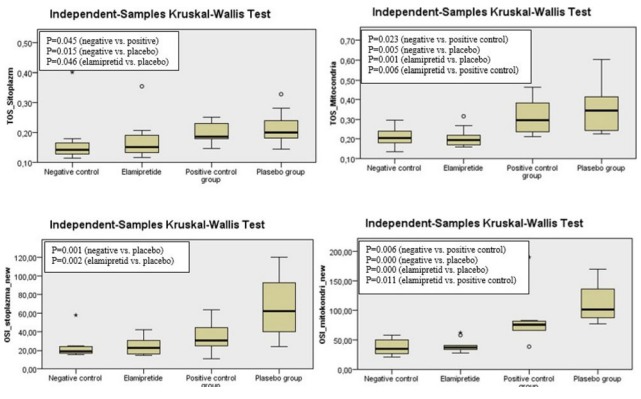
[1]Erciyes University Genome And Stem Cell Center, [2]Niğde Ömer Halis Demir University, Faculty Of Medicine Department Of Physiology, [3]Niğde Ömer Halis Demir University, Faculty Of Medicine Department Of Biophysics, [4]Bozok University, Faculty Of Medicine, Department Of Physiology, [5]Erciyes University, Faculty Of Veterinary, Department Of Biochemistry, [6]Erciyes University Faculty Of Medicine, Department Of Medical Biology, [7]Erciyes University Faculty Of Medicine, Department Of Pediatrics, Division Of Nephrology

Introduction: It has been shown that mitochondrial damage may play an important role in the emerge of the oxalate nephropathy (ON), and prevention of mitochondrial damage is thought to be one of the most important targeted therapies. The aims of this study are 1) To investigate the possibile role of mitochondrial damage in the development of ON in mice 2) To investigate whether mitochondrial targeting tetrapeptide (Elamipretide) treatment has a therapeutic effect on mitochondrial function in a mouse model of ON.

Material and methods: As Figure 1, 35 C57BL6 mice at 10-12-weeks of age were used (8 controls, 27 experimental groups). The study group was administered 200 mg / kg sodium oxalate intraperitoneally (IP) for 4 days. No application was made to the positive control group after the 5th day. Phosphate buffer saline was administered to the placebo group on days 0-9, and elamipretide at a dose of 7.5 mg / kg / day was administered via IP to the drug groups. The experiment was terminated on the 10th day and the mice were bled and their kidneys removed. Histopathological evaluation was made in the kidney tissue. The kidneys were homogenized and biochemical measurements (TAS, TOS, SOD) were made in mitochondria, cytosol. Mitochondria membrane potential (MMP) and ATP levels were determined. Oxidaitve strest index (OSI) was calculated. This study supported by the Erciyes University Scientific Research Center (TDK-2019-9184).



Results: Histologically, oxalate caused apparent tubulointerstitial inflammation which was alleviated by elamipretide. Both mitocondrial and cytosolic oxidant capasity (TOS), OSI were increased and anti-oxidant capasity (TAS) were decreased in mice with ON and all were recovered with elamipretide (Figure 2a-d). We found oxalate resulted in decreased MMP and it was alleviated by elamipretide.



Conclusions: In this study, it has been shown that ***mitochondrial damage occurs in mice with ON and is alleviated by elamipretide***. We think mitocondrial targeting treatment may be used to decrease oxidative stress related injury in kidney tissue in mice with ON.

PI-23 MITOCHONDRIA-TARGETED COQ10 FORMULATION FOR THE TREATMENT OF COQ10 NEPHROPATHIES

Hamide Sena Ozbay[1], Samiye Yabanoglu Ciftci[1], Ipek Baysal[2], Merve Gultekinoglu[3], Cemil Can Eylem[4], Kezban Ulubayram[3], Emirhan Nemutlu[4], Rezan Topaloglu[5], Fatih Ozaltin[5]

[1]Hacettepe University, Faculty Of Pharmacy, Department Of Biochemistry, Sihhiye, Ankara 06100, Turkey, [2]Hacettepe University, Hacettepe University, Faculty Of Health Sciences, Ankara, Turkey, [3]Hacettepe University, Faculty Of Pharmacy, Department Of Basic Pharmaceutical Sciences, Sihhiye, Ankara 06100, Turkey, [4]Hacettepe University, Faculty Of Pharmacy, Department Of Analytical Chemistry, Sihhiye, Ankara 06100, Turkey, [5]Hacettepe University, Faculty Of Medicine, Department Of Pediatrics, Division Of Pediatric Nephrology, Sihhiye, Ankara 06100, Turkey

Introduction: Coenzyme Q_{10} (CoQ_{10}) deficiency causes many organ dysfunctions, with isolated kidney involvement leading to chronic kidney disease in some subtypes. Early administration of oral CoQ_{10} has been observed to reduce proteinuria and has been postponed the onset of chronic kidney disease in these patients, suggesting that it may have renoprotective properties. On the other hand, the limited bioavailability of CoQ_{10} in mitochondria is an obstacle to its effectiveness.

Material and methods We aimed to formulate mitochondria targetted CoQ_{10}-loaded poly(lactic-co-glycolic acid)-poly(ethylene glycol)-triphenylphosphonium nanoparticles (CoQ_{10}-TPP-NPs) that may be used to treat CoQ_{10} nephropathies more effectively. Their effects were evaluated on an in vitro disease model, which was created in HK-2 cell line by siRNA based silencing of the COQ8B. Mitochondrial functions were determined by metabolomic analyses.

Results: The size of these nanoparticles was determined to be around 150 nm, with a zeta potential of +20 mV. The entrapment efficiency of the nanoparticles was found to be 40%. It was found that these nanoparticles did not show cytotoxic effects on HK-2 cells. Metabolomic analyses showed increased rate of tricarboxylic acid cycle in COQ8B$^{-/-}$ cells treated with CoQ_{10}-TPP-NPs as compared to those treated with free-CoQ_{10}.

Conclusions: Our mitochondria-targetted formulation was found to be more effective than free-CoQ_{10} in CoQ_{10} deficient HK-2 cells. If its in vivo effects confirm our in vitro observations, this

formulation would be a promising therapeutic option in the treatment of CoQ_{10} nephropathies.

PI-24 LEVAMISOLE MODULATES PODOCYTES' ACTIN CYTOSKELETON IN VITRO IN IDIOPATHIC NEPHROTIC SYNDROME

This abstract has been withdrawn.

PI-25 CANDIDATE BIOMARKER(S) FOR CYSTINOSIS WITH OMIC-BASED TECHNOLOGY: FROM LABORATORY TO BED-SIDE

Emirhan Nemutlu[1], Cemil Can Eylem[1], Bora GÜlhan[2], Ipek Baysal[3], Samİye Yabanoglu-cİftcİ[4], Sedef Kir[1], Kezban Ulubayram[5], GÜlberk UÇar[4], Osman UĞur Sezerman[6], Fatİh Ozaltİn[2], Rezan Topaloglu[2]

[1]Hacettepe University Faculty Of Pharmacy, Department Of Analytical Chemistry, Ankara, Turkey, [2]Hacettepe University Faculty Of Medicine, Department Of Pediatrics, Division Of Pediatric Nephrology, Ankara, Turkey, [3]Hacettepe University Vocational School Of Health Services, Ankara, Turkey, [4]Hacettepe University Faculty Of Pharmacy, Department Of Biochemistry, Ankara, Turkey, [5]Hacettepe University Faculty Of Pharmacy, Department Of Basic Pharmaceutical Sciences, Ankara, Turkey, [6]Acibadem Mehmet Ali Aydinlar University Faculty Of Medicine, Department Of Biostatistics And Medical Informatics, Istanbul, Turkey

Introduction: Cystinosis is a rare autosomal recessive lysosomal cystine storage disease that stems from defective cystinosin protein caused by mutations in CTNS. The most frequently applied diagnostic method is the confirmation of the increased leukocyte cystine levels. However, this test is expensive, unreproducible and difficult to have an access in everywhere. This study aimed to investigate new biomarkers for the diagnosis and follow-up of this rare disease with multiomics (genomic, proteomic and metabolomic) methods.
Material and methods: Multiomic studies have been performed in plasma and urine samples from three pediatric groups; a) newly-diagnosed cystinosis patients (n=14), b) cystinosis patients under treatment (n=31), c) healthy controls (n=30) and in d) siRNA based cystinosis model in vitro.
Results: Multivariate analysis revealed different plasma and urinary profile between groups. The candidate biomarkers were determined using receiver operating curve (ROC) analysis. Among these metabolites; L-serine, taurine, 4-Trimethylammoniobutanoic acid, glutathione, PE(O-18:1(9Z)/0:0), 2-hydroxyphenyl acetic acid, and 3-indoxyl sulphate in plasma and allo-inositol, oleanonic acid,4-hydroxyphenylpyruvic acid, 2-hydroxybutiric acid, cystine, pyruvic acid, valine, phenylalanine, N-acetyl-L-glutamic acid, 3-aminopropionitrile, ribitol, hydroquinone, glucuronic acid, 3-phosphoglycerate, xanthine, creatinine and 5-aminovaleric acid in urine samples were found as candidate biomarkers in for the differentiation of patients with cystinosis from healthy individuals. Besides, for the first time in the literature, siRNA based CTNS silenced HK2 cells were generated as an in vitro disease model to determine omics differences compared to wild type HK-2 cells to understand metabolic alterations caused by cystinosin deficiency at cellular level.
Conclusions: Our study led to the description of candidate biomarkers and important results, which would pave the way for a better understanding of the pathophysiology of cystinosis.

PI-27 ASSESSMENT OF HLA INCOMPATIBILITY AT THE MOLECULAR COMPARED TO ANTIGENIC HLA LEVEL ENABLES BETTER PREDICTION OF GRAFT FUNCTION DETERIORATION IN PAEDIATRIC KIDNEY TRANSPLANTATION.

Jon Jin Kim[1], Alexander Fichtner[2], Hannah Copley[1], Caner Susal[2], Lars Pape[3], Jun Oh[4], Kai Krupka[2], Burkhard Toenshoff[2], Vasilis Kosmoliaptsis[1]

[1]University Of Cambridge, [2]University Of Heidelberg, [3]University Of Hannover, [4]University Of Hamburg

Introduction: Immune recognition of donor HLA mismatches and subsequent graft rejection remain a major cause of graft deterioration in paediatric kidney transplantation. We aimed to evaluate the potential of computational methods of assessing HLA immunogenicity (molecular mismatching, molMM) and of classical antigen mismatching (antMM) to predict primary alloimmunity risk.
Methods We performed a retrospective analysis of 177 paediatric patients (median age 10.8 [IQR] 5-15 years) from the antibody-mediated

rejection (ABMR) study from the CERTAIN registry, all of whom had prospective monitoring of donor-specific HLA antibody (DSA) post-transplant. We compared four molMM methods: amino acid mismatch scores (AAMS, assessing T- and B-cell alloimmunity), electrostatic mismatch score-3-dimensional (EMS3D, assessing B-cell alloimmunity), and NetMHCIIpan (T-cell alloimmunity) at HLA-peptide affinity binding thresholds of 500 nM (netMHC) and 1000 nM (netMHC1k as implemented in the PIRCHE algorithm).
Results: *De novo* DSA incidence was highest against HLA-DQ (30/177, 17%, Table 1). Higher antMM and molMM were associated with DSA formation. DSA preferentially targeted the highest scoring molMM allele in each individual patient. MolMM methods were more predictive of DSA compared to antMM (Table 1, AUROC results), and EMS3D was the most consistent predictor across all HLA loci (AUROC 0.72-0.75). Biopsy-proven ABMR (11/177, 6%) was associated with increasing recipient age and class II molMM (defined by AAMS or EMS3D or netMHC). Late TCMR (>6 months postrtansplant) was diagnosed on for-cause biopsies in 23 (13%) patients. Accounting for the total HLA burden, neither molMM nor antMM scores were predictive of TCMR outcome. Stratifying HLA mismatches into low or high risk according to molMM score enabled patient classification into strata. TCMR was significantly associated with HLA-DQ molMM using T-cell algorithms - AAMS (HR 1.8, 95%CI 1.2-3.0 per risk stratum) and netMHC (HR 1.5, 95%CI 1.0-2.2 per risk statum).
Summary: Molecular HLA mismatching enabled better prediction of primary alloimmunity risk than conventional antigen mismatching. Incompatibility at the HLA-DQAB loci was the main driver for TCMR and ABMR.

HLA loci (number of DSA, %)	A (22, 12%)	B (15, 8%)	DQ (30, 17%)	DR (25, 14%)
Antigen	0.63	0.46	0.56	0.57
AAMS	0.71	0.67	0.72	0.74
EMS3D	0.74	0.72	0.75	0.73
netMHC	0.71	0.57	0.64	0.69
netMHC1k	0.73	0.52	0.76	0.70

Table1: Area under the receiver operating characteristic curve (AUROC) results for each molMM method compared with antMM. Analysis was performed using the highest score of both alleles, using logistic regression for each HLA allele separately.

PI-28 KIDNEY TRANSPLANT OUTCOMES IN SMALLER CHILDREN COMPARED TO LARGER CHILDREN – REPORT FROM THE UK TRANSPLANT REGISTRY

Jan Dudley[1], Charlie Pickles[2], Chloe Brown[3], Ben Reynolds[4], Nicos Kessaris[1], Stephen Marks[5]

[1]Bristol Royal Hospital For Children, [2]Great North Childrens Hospital, Newcastle, [3]Nhs Blood And Transplant, [4]Royal Hospital For Children, Glasgow, [5]Great Ormond St Hospital For Children

Introduction: Kidney transplantation in young children is often delayed due to technical feasibility and concerns about poorer outcomes. We compared 5 and 10 year graft outcomes in children weighing < 12Kg and < 15Kg, compared with larger children.
Material and methods: Data on all first paediatric (aged <18 years) kidney only transplants performed in the UK between 1 January 2006 and 31 December 2016 were extracted from the UK Transplant Registry (n = 1,352). Donor, recipient and transplant characteristics were compared. Thirty day, one-year and five-year patient and allograft survival were compared using the Kaplan-Meier method.
Results: There was no evidence of a difference in 30-day or one-year kidney allograft survival (p = 0.57, p = 0.73 respectively). Five-year allograft survival for children < 15Kg was better than that for children ≥15kg (92.1% (95% CI: 87.2% - 95.2%) vs 87.0% (95% CI: 84.6% - 89.0%)) but this did not reach statistical significance (p = 0.1). For children < 12Kg, a greater proportion of kidney transplants were from living donors compared with children ≥12kg (p = 0.005). There was no difference overall in renal allograft function between the groups (p = 0.78). Immediate graft function was seen in 77.9% of children <12Kg, compared with 84.5% in children ≥12kg, however, risk-adjusted analysis could not be performed due to the very small number of graft failures in this group.
Conclusions: Patient and renal allograft survival outcomes for kidney transplantation undertaken in small children are at least as good as for larger children and this should be reflected in earlier preparation for transplantation for these children. 10-year graft survival data are currently being analysed.

PI-29 ASSESSMENT OF HLA INCOMPATIBILITY AT THE MOLECULAR COMPARED TO ANTIGENIC HLA LEVEL ENABLES BETTER PREDICTION OF GRAFT FUNCTION DETERIORATION IN PAEDIATRIC KIDNEY TRANSPLANTATION.

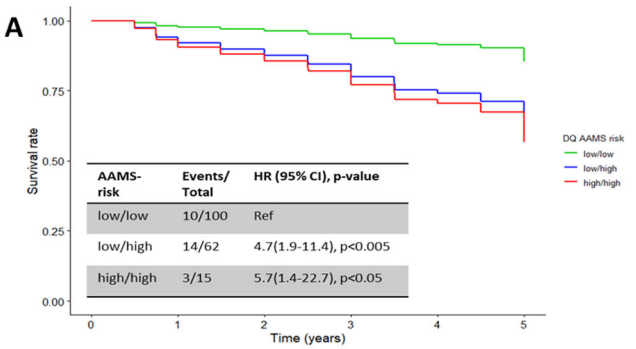
Jon Jin Kim[1], Alexander Fichtner[2], Hannah Copley[1], Caner Susal[2], Lars Pape[3], Jun Oh[4], Kai Krupka[2], Burkhard Toenshoff[2], Vasilis Kosmoliaptsis[1]

[1]University Of Cambridge, [2]University Of Heidelberg, [3]University Of Hannover, [4]University Of Hamburg

Introduction: HLA mismatching has a detrimental effect on graft survival after paediatric kidney transplantation. Assessment of HLA incompatibility at the molecular level (molecular HLA mismatch; molMM) has emerged as a promising method for predicting primary alloimmunity risk. In this study, we aimed to assess whether molMM compared to conventional antigenic mismatching (antMM) enables better prediction of graft function deterioration in paediatric kidney transplantation.
Methods: We performed a retrospective analysis of 177 paediatric patients from the ABMR study of the CERTAIN registry (median follow-up 4.5 (IQR 3-5) years). Only five patients experienced graft loss. Therefore, we used the time to 50% reduction in eGFR, from month-3 post-transplant baseline, as a surrogate endpoint for long-term graft loss (eGFR-50). HLA molMM was assessed using the Cambridge amino acid mismatch score (AAMS) which on a separate study was predictive of primary alloimmunity risk in this cohort. Survival analysis was performed using Cox models, adjusted for donor and recipient baseline characteristics.
Results: 27 (15%) patients met the primary outcome. In multivariable analysis, recipient and donor age, baseline eGFR, and re-transplant status had a significant association with eGFR-50. Importantly, only mismatches at HLA-DQα1β1, and not at other loci, were associated with the primary outcome (adjusted HR (aHR) 10.2; 95% CI, 10.1-10.4 per 10 AAMS increase, and aHR 1.8; 95% CI, 1.02-3.4 per antigen increase). There was a wide range of AAMS values (0-49) within each HLA-DQ antMM (0-2). We used a predetermined molMM score (AAMS=16 derived from analyses of donor-specific antibody risk) to classify patients according to HLA-DQα1β1 mismatch risk into "low/low", "low/high" and "high/high" risk groups. Patient risk for eGFR-50 was associated

with molMM stratification, regardless of their antMM. eGFR-50 risk was equivalent in patients with "low/low" risk mismatches (aHR 2.2, 0.2-23, p=0.5 for 2 v 0 antMM). Patients with "low/high" and "high/high" HLA-DQα1β1 mismatches had worse allograft outcomes ("low/high": aHR 4.7, 1.9-11.4, p<0.05; "high/high" aHR 5.7, 1.4-22.7, p<0.05 versus "low/low", Figure 1). Compared to antMM, molMM showed better stratification of outcomes whilst increasing the number of patients in the low risk group ("low/low" n=100, v 0 antMM n=65).

Conclusion: Assessment of HLA incompatibility at the molecular level enables better stratification of graft deterioration risk compared to conventional serology-based HLA mismatching. Further validation of molMM in independent cohorts is required before clinical implementation.



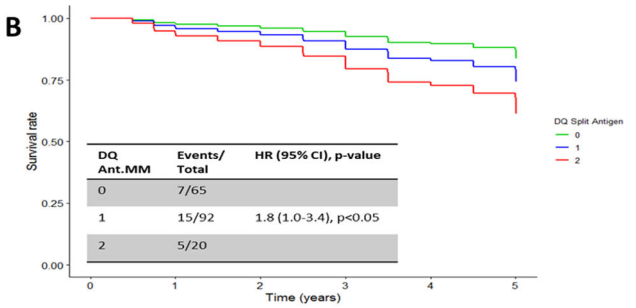


Figure1: Survival analysis for GFR50 outcome based on molMM (A) and antMM (B). Survival curves were adjusted for baseline eGFR, recipient age, donor age and transplant number. Hazard ratios were calculated using multi-variable Cox analysis.

PI-30 RESPONSE TO MRNA VACCINES FOR SARS-COV-2 IN YOUNG KIDNEY TRANSPLANT RECIPIENTS.

Luigi Cirillo[1], Elisa Buti[1], Francesco Citera[2], Vito Terlizzi[3], Francesca Becherucci[1], Carmela Errichiello[1], Giovanni Taccetti[3], Chiara Azzari[2], Paola Romagnani[1]

[1]Nephrology And Dialysis Departmet, Meyer Childrens Hospital, Florence, It, [2]Immunology Department, Meyer Childrens Hospital, Florence, It, [3]Cystic Fibrosis Department, Meyer Childrens Hospital, Florence, It

Introduction: Seroconversion after mRNA SARS-CoV-2 vaccination might be unsatisfactory in Kidney Transplant Recipients (KTRs), ranging in literature between 14-58% after the second dose; however most of the data are based on adult populations given the scarce evidence in the young population.

Material and methods:
Patients aged 16-24 years, renal transplant carriers and age-matched control group without immunosuppression, vaccinated with RNA vaccines; we performed quantitative ELISA assay to quantify IgG anti-spike, anti-capsid and neutralizing (RBD) IgG antibodies for SARS-Cov2; samples were drawn on the day of the second dose of vaccine, one month after and one month after the booster dose.

Results: 18 KTRs and 15 controls with a mean age of 19 years were enrolled. 16 KTRs in therapy with steroids, MMF and CNI; 1 with steroids, CNI; 1 with steroids, MPA and CNI. One patient with a previous diagnosis of COVID19 tested positive for anti-capsid. At the first sampling 7/18 (39%) patients had seroconversion; at the second sampling 13/18 (72%); 11/12 (92%) at the third sample. KTRs neutralizing antibodies were reduced compared to controls after the second dose (p<0.05); antibody levels became equal only after the booster (p<0.05). This in fact produced a statistically significant increase of neutralizing antibodies compared to the second dose in KTRs (p<0.05). The higher quantitative response to the vaccine was observed in those who were in therapy with MPA or did not take antimetabolite, and have lower dose of MMF; a high response was also found in the patient who had had a previous infection. All patients who went adjunctive immunosuppressive treatments other than maintenance in the previous six months did not show seroconversion.

Conclusions: These findings suggest that a third dose of SARS-CoV-2 mRNA vaccine improves the RBD-specific responses of transplant patients treated with immunosuppressive drugs.

PI-31 BELATACEPT OUTCOMES IN PEDIATRIC KIDNEY TRANSPLANTATION: THE ROBERT DEBRE EXPERIENCE

Charlotte Duneton, Elodie Cheyssac, Houaida Dahdouh, Veronique Baudouin, Hogan Julien

Pediatric Nephrology Department, Robert Debré Hospital, Aphp, Paris France

Introduction: Belatacept is associated with reduced dnDSA, improved renal function, and prolonged allograft survival in adult transplant recipients. Its use in older children and young adults is limited. We report outcomes for 13 pediatric patients converted to belatacept.

Material and methods: 13 patients were converted to belatacept between 2018 and 2021 in Robert Debré Hospital. Patients received an induction with basiliximab (n=8) or ATG (n=5). Maintenance immunosuppression included CNI, antimetabolite and steroids. Patients' viral status were monitored monthly and allograft biopsy was performed prior to conversion and 6 months after conversion. The first 5 belatacept injections were administered at 5mg/kg/dose every 2 weeks, then monthly. CNI doses were decreased by 25% at each infusion and stopped after 2 months. Antimetabolite doses were also increased at CNI withdrawal. 6/13 patients were steroid-free at the time of conversion.

Results: Median age at conversion was 17,6 years (range 10,3-19,4) and 3,9 years (IQR 1,3 – 6) post-transplant. Conversion indication was based on medical need for long term CNI avoidance (n=11) or to improve adherence (n=2). CNI was withdrawn in all patients after a median of 42 days (IQR 42-75). GFR was stable or improved over a median follow-up time of 12,1 months (IQR 9,6-20,3). Rejection episodes were observed in 4/13 patients (median 10,3 months, IQR 7,3-19,1): 2 chronic active TCMR IA, 1 mixed acute rejection (TCMR IB and ABMR) and 1 ABMR (both DSA-negative). 2 had prior history of rejection (with normal pre-belatacept biopsies), 1 showed minimal interstitial inflammation without tubulitis (and was off steroids) prior to starting belatacept and 1 had been

converted for adherence problems, which subsequently persisted. Rejection episodes showed good evolution after treatment, but CNI were reintroduced for 2/4. No severe viral complications or development of dnDSA were observed.
Conclusions: Selected pediatric kidney recipients may benefit from long-term CNI toxicity avoidance, but selection criteria need to be refined.

PI-32 EVALUATION OF THE SIGNIFICANCE OF ISOLATED C4D STAINING WITHOUT HISTOLOGICAL EVIDENCE OF REJECTION ON KIDNEY BIOPSIES AFTER ABO-COMPATIBLE TRANSPLANTATION IN PEDIATRIC RECIPIENTS

Charlotte Duneton[1], Marion Rabant[3], Jean Paul Duong-van-huyen[3], Veronique Baudouin[1], Elodie Cheyssac[1], Olivia Gillion Boyer[2], Julien Hogan[1]

[1]Pediatric Nephrology Department, Robert Debré Hospital, Aphp, Paris France, [2]Pediatric Nephrology Department, Necker Enfants-malades Hospital, Aphp, Paris France, [3]Department Of Pathology, Necker Enfants-malades Hospital, Aphp, Paris France

Introduction: C4d staining in peritubular capillaries has been part of ABMR Banff definition since 2003. Despite relatively high specificity, c4d staining shows limited sensitivity, and the clinical significance of C4d+ biopsies without other histological evidence of rejection (WER) is unknown. We aimed to assess the clinical significance of c4d+ biopsies WER on kidney biopsies after ABO-compatible transplantation in pediatric recipients
Material and methods: We retrospectively analyzed patients under 18 years with c4d+WER biopsies performed between 2011-2020, in 2 pediatric transplant centers in Paris, France. All biopsies were reviewed by a single expert pathologist to confirm the immunohistochemical C4d+ staining and to ensure the absence of ABMR or TCMR. C4d+ WER patients were compared with a cohort of C4d- patients matched on recipients' demographics, year and center of transplantation, biopsy indication and time to transplantation.
Results: 6% of biopsies were C4d+WER. 40 C4d+WER children were compared with 40 matched C4d- controls (median age 12 and 11,6 years). 65% were protocol biopsies performed within the 1st year after transplant (median 6,9 months, IQR 2,5–15,2). There was no significant difference regarding recipient, donor, or transplant characteristics. However, C4d+ patients showed significantly more DSA on the day of biopsy (40% vs 17,5%, p=0,047). After a 4-years median follow-up, 4/40 C4d+ patients developed an ABMR within year 1 (10,4 months, IQR 6,2–14,0). All these patients were DSA+ at the time of biopsy. However, among the patients who had a follow-up biopsy (n=32), we noted a negativation of C4d staining in 78%, including 20/32 patients (62%) in the absence of specific treatment. Survival rate without ABMR did not differ between C4d+ and C4d- patients (Fig A). However, C4d+ DSA+ patients developed significantly more ABMR within the 1st-year post biopsy compared with C4d+DSA- and C4d- patients (Fig 2). Interestingly, patient characteristics and outcomes of C4d+DSA- patients did not differ from C4d- patients.
Conclusions: C4d+WER biopsies DSA- do not represent an increased risk of ABMR compared to C4d- patients. Molecular analysis of the biopsies may help to assess the significance and clinical implication of C4d+WER in pediatric kidney transplant recipients.

PI-33 IMMMUNOLOGIC RESPONSE TO BNT162B2 COVID-19 MRNA VACCINE IN ADOLESCENT KIDNEY TRANSPLANT RECIPIENTS. A SINGLE CENTER EXPERIENCE

Varvara Askiti[1], Evangelia Gole[1], Maria Papadimitriou[2], Maria Eirini Gourtzelidou[3], Minos Matsas[2], Andromachi Mitsioni[1]

[1]Pediatric Nephrology Department, Children's Hospital "p&a Kyriakou", Athens, Greece, [2]Department Of Microbiology, Children's Hospital "p&a Kyriakou", Athens, Greece, [3]National And Kapodistrian University Of Athens, Medical School, Athens, Greece

Introduction: Early reports suggest low immunogenicity of SARS-CoV-2 vaccines in adult kidney transplant recipients (KTR). We describe the immunogenicity and safety profile of BNT162b2 mRNA Covid-19 vaccine in adolescents KTR and we compare it with the serologic response of natural infection.
Material and methods: 11 KTR (group A) received two doses of the vaccine between July 2021 and November 2021, while 4 KTR had a PCR-confirmed Covid-19 infection (group B). Serum samples were tested at 20 days and 3 months post the second dose for detection of IgG antibodies against spike protein of SARS-CoV-2, using chemiluminescent microparticle immunoassay (Architect/Alinity,Abbott). IgG results ≥50 AU/ml were considered positive.
Results: Median age was 14,5 years old (IQR 13-17), median time from transplant 16,26 months (IQR 10-58,6). Positive serologic responses were observed in 7/11 (64%) of the vaccinated KTR and 4/4 (100%) of the naturally infected KTR. Three of the four seronegative patients had previously received rituximab (19, 18 and 8 months before vaccination respectively) compared to none of the responders, p=0.024. Antibody titers were 10 times higher in the naturally infected group than vaccinated group [median 122 AU/ml (IQR 20-678) versus 1339 AU/ml (IQR 1234-1384)], p=0.001 Man-Whitney test]. In vaccinated KTR antibody levels increased from a median level of 11.6 AU/ml at 20 days to 122 AU/ml at 3 months post second dose. The vaccine was well tolerated with no rejection episodes. No patient developed COVID 19 infection post vaccination, including the seronegatives, during the follow up period.
Conclusions: SARS-CoV-2 vaccination is safe in adolescents KTRs. Our cohort seem to have better immunologic response than the previously reported in adults but longer time is required to mount an adequate antibody response compared to the general population. Vaccination results in lower antibody titers than natural infection. Rituximab has a negative effect on serologic response.

PI-34 COVID-19 INFECTION AND IMMUNE RESPONSE IN PEDIATRIC KIDNEY TRANSPLANT PATIENTS

Yolanda Calzada Baños, Marta Jiménez Moreno, Ana Cristina Aguilar Rodríguez, Pedro Arango Sancho, Claudia Fortuny Guasch, Raquel Jiménez García, Elena Codina Sampera, Álvaro Madrid Aris

Hospital Sant Joan De Déu

Introduction: COVID-19 has had high morbidity and mortality in kidney transplant patients (RT) and other solid organs.However, its clinical course in pediatric recipients remains poorly defined,with few reported cases.The objective of the study is to characterize the evolution of SARS-Cov-2 infection in pediatric RT as well as to study the humoral response developed after it.
Material and methods: Descriptive prospective single-center study started in October 2020.RT patients aged 18 years or younger under follow-up at our center with a diagnosis of SARS-CoV-2 infection by PCR technique,indicated by the presence of symptoms and/or signs,are included.Suggestive of infection or by study of contacts in confirmed cases. Demographic,clinical and analytical variables,seroconversion study (IgM and IgG) and biobank samples for subsequent study of cellular response and interleukins are analyzed.
Results: Six patients are included,predominantly female (83.3%);median age of 13.5 years (7-16) and median time post-RT of 31.5 months (6-

120).2 patients (33.3%) had high blood pressure,1 had diabetes mellitus,1 was overweight (BMI 25.8) and 1 was obese (BMI 30.9),none had underlying heart disease or lung disease.The most frequent maintenance immunosuppression was tacrolimus,mycophenolate and prednisone (50%).Half of the patients were diagnosed during a school contact study,and the other 50% as a result of family cases,none of which was an index case.2 patients (33.3%) were asymptomatic.The most frequent symptom was cough (4,66.7%) followed by asthenia (2),fever (1) and diarrhea (1).One patient presented mild bronchospasm without hypoxemia,normal chest X-ray.The most frequent analytical finding was lymphopenia or its worsening (5,83.3%) and ferritin elevation (66.7%),with erythrocyte sedimentation rate (ESR),procalcitonin and C-reactive protein being negative in all cases.Mild elevation of D-dimer in 2 patients.No patient presented graft dysfunction.No patient required hospitalization or adjustment of immunosuppression.4 patients (66.7%) developed IgM and IgG that are maintained at 6 months post-infection.2 patients (33.3%) did not present seroconversion,of which 1 presented positivization of IgM and IgG after 3 months,being oriented as asymptomatic reinfection.It remains to analyze cellular response at the time of analysis.

Conclusions: In our pediatric RT cohort,COVID-19 appears to have a benign course similar to studies in non-immunosuppressed children.The study of cellular immunity remains pending at the time of the analysis.

PI-35 SARS-COV-2 ANTIGEN-SPECIFIC CELLULAR AND HUMORAL IMMUNE RESPONSE AFTER TWO OR THREE DOSES MRNA VACCINE BNT162B2 IN ADOLESCENT KIDNEY TRANSPLANT RECIPIENTS

Cyrielle Parmentier[1], Isabelle Nel[2], Laurene Dehoux[3], Marina Charbit[3], Ferielle Louillet[4], Elodie Cheyssac[2], Jean-daniel Delbet[1], Veronique Baudouin[2], Tim Ulinski[1], Guislaine Carcelain[2], Julien Hogan[2]

[1]Trousseau Hospital, Aphp, Paris France, [2]Robert Debre Hospital, Aphp, Paris France, [3]Necker Hospital, Aphp, Paris, France, [4]Rouen Normandie, Chu Hospital, Rouen, France

Introduction: Adolescent kidney transplant recipients (KTRs) are immunocompromised and therefore prioritized for SARS-CoV-2 mRNA vaccination. Data are lacking regarding their humoral and cellular response to COVID-19 vaccination.

Material and methods: We conducted a retrospective study to analyze the early (between 21 and 90 days) humoral immune (ELISA) or/and cellular (interferon-g release assay and flow cytometry) response in 48 KTRs aged 12 to 21 years, using two or three doses of mRNA vaccine BNT162b2.

Results: SARS-CoV-2-vaccination induced seroconversion with a humoral response in 86% patients after 2 doses and 88% after 3 doses. The third dose induced seroconversion in the 3 seronegative patients after two doses. Median antibody levels were 1500 BAU/mL IQ (414; 2860) and 955 (163; 3737) after 2 and 3 doses, respectively. Only 12/28 (43%) patients showed a specific T cell response after the second injection and 11/23 (48%) after a third. Patients with a history of COVID-19 infection received only 2 injections were all responders. KTRs treated with an immunosuppression including mycophenolate were more likely to be non-responders than in those with azathioprine (76% vs. 100%). Likewise, after two doses in KTRs with lymphopenia, 5/7 (71%) patients had no specific T cell response vs. 13/24 (54%) in patients with normal lymphocyte count, and a median specific IgG directed against the spike protein of 261 BAU/mL vs. 1790 BAU/mL (p= 0.02) respectively.

Conclusions: Adolescent KTRs exhibit a high seroconversion rate of 86% after only two doses. Immunosuppressive drug type, as well as lymphopenia are determinants of seroconversion failure suggesting the need for immune monitoring and adaptation of vaccination protocols for this specific population.

PI-36 THE VALUE OF NON-INVASIVE HEMODYNAMIC MONITORING DURING PEDIATRIC HEMODIALYSIS

Anna Végh[1], Lóránt Sagát[2], Ágnes Liebhardt[2], GyÖrgy S. Reusz[1]

[1]First Department Of Pediatrics Semmelweis University, Budapest, Hungary, [2]Faculty Of Medicine Semmelweis University, Budapest, Hungary

Introduction: Both hypo- and hypertension during hemodialysis (HD) has been associated with an increased cardiovascular risk. Currently, measuring blood pressure (BP) is the primary method for estimating hemodynamic changes caused by fluid removal (UF). The aim of this study is to assess hemodynamic changes by non-invasive monitoring with electrical velocimetry ™ (EV) during hemodialysis (HD) on pediatric patients.

Material and methods: Maintenance HD patients of a single pediatric center took part in this study. Anthropometric data, laboratory results and history were collected. Multiple HD sessions were recorded for each patient. Hemodynamic data was measured during HD with an ICON® monitor (Osypka Medical, Germany), BP was recorded every 25 minutes. A BCM device (Fresenius, USA) was used to assess fluid status before and after dialysis.

Results: Thirty-eight dialysis sessions of thirteen pediatric patients were included in this study, with 10 (71,42%) males. Median [IQR] age was 15,8 [13,3-16,2] years. Median [IQR] predialytic overhydration was 2,8 [0,4-5,2] percent of dry weight.

Significant decrease of stroke volume (SV) (p=0,015) and thoracic fluid content (TFC) (p<0,0001), as well as increase of heart rate (HR) could be observed, with a correlation to UF: SV (p=0,004; **Figure 1**.), TFC (p<0,0001; **Figure 2.**), HR (p=0,05; **Figure 3.**). Although cardiac output (CO) and mean arterial pressure (MAP) was stable in the whole cohort, subgroup analysis revealed different patterns. Pre- and postdyalitic hypertension was present in 18-18 (47,4-47,4%) patients. No hypotensive event occurred. A 10% change in CO was considered significant. CO increased at 15 (39,5%), decreased at 12 (31,6%), and stayed stable at 11 (28,9%) sessions.

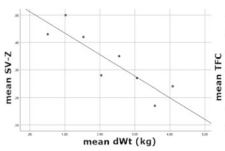


Figure 1.

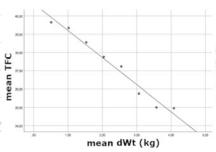


Figure 2.

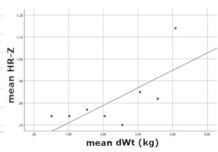


Figure 3.

Conclusions: While MAP stayed stable, EV provided more detailed insight into the compensatory mechanisms during HD. EV could potentially optimize HD management in pediatric patients by revealing hemodynamic changes that go unnoticed by BP monitoring.

PI-37 BENEFITS OF BNP/NT-PRO-BNP SERUM LEVEL EVALUATION FOR DRY WEIGHT ADJUSTMENT IN PAEDIATRIC HAEMODIALYSIS PATIENTS

Antoine Mouche[1], Cyrielle Parmentier[1], Fatma Fendri[1], Claire Herbez-rea[1], Theresa Kwon[2], Laurene Dehoux[3], Jean-daniel Delbet[1], Tim Ulinski[1]

[1]Hopital Trousseau, Paris (aphp), [2]Hopital Debre, Paris (aphp), [3]Hopital Necker, Paris (aphp)

Introduction: Dry weight (DW) adjustment in children on haemodialysis (HD) can be challenging. It relies on clinical evaluation and additional supports. Our aim was to study the benefits of cardiac

biomarkers assessment, in addition to the more commonly used technique bioimpedance spectroscopy (BIS) and clinical signs for DW prescription in paediatric HD patients.
Material and methods: Prospective observational study including 41 children on HD in three paediatric HD centres in the Paris region. During one session, BIS was performed before the session and serum levels of BNP and NT-proBNP were analysed before and after the session.
Results: Median pre-dialysis level of BNP was 87 ng/L [24–192] and NT-proBNP 968 ng/L [442–4828]. Cardiac biomarkers levels showed positive correlation with the BIS hydration status evaluation (p=0.004). The most appropriate cut-off for pre-dialysis BNP to detect significant overhydration (OH) was 165 ng/L (sensibility 0.67, specificity 0.84). Based on the BIS evaluation, only 32% of patients with high blood pressure (BP) had OH, whereas in the normal BP group, 33% had significant OH.
Conclusions: DW prescription for children on HD should not only rely on clinical evaluation, particularly BP, and should include additional helpful parameters. BIS is well validated in children, but it has limitations in non-cooperative patients. Cardiac biomarkers, especially BNP, have proven in this study to be well correlated to hydration status evaluated by BIS, and thus could add precious informations for individual patient management and dry weight assessment.

PI-38 HEALTH-RELATED QUALITY OF LIFE IN PATIENTS RECEIVING IN-CENTRE DIALYSIS

Claire Dempster, Francesca De Zan, Jo Wray, Rukshana Shroff

Nephrology Unit, Great Ormond Street Hospital For Children Nhs Foundation Trust, Great Ormond Street, London, United Kingdom

Introduction: End-stage kidney disease is associated with impaired quality of life (QoL). Few studies have investigated the mental health and well-being of children on dialysis. Through validated questionnaires we explored children's and parents' QoL perception and identified risk factors for a concerning score.
Material and methods: Participants from the "3H - HDF-Hearts-Height" multicentre study in children on hemodialysis (HD) and hemodiafiltration (HDF) completed Paediatric QoL questionnaire (PedsQL) and the Strengths and Difficulties (SDQ) questionnaires.
Results: 100 children and parents answered at least one questionnaire and were enrolled in the study. Children on dialysis scored significantly lower for health related QoL on the PedsQL than healthy children ($p < 0.001$ in all the domains). Differences between children and parents' answers in both questionnaires were not significant in all the domains ($p > 0.15$). On univariable analysis school attendance significantly reduced the probability of a concerning score, both in children's reported outcomes for emotional (95%CI 0.083 – 0.61; p = 0.003), school (95%IC 0.029 – 0.722; p = 0.018) and total domain (95%CI 0.023 – 0.49; p = 0.004) and in parents' reports on school (95%CI 0.023 – 0.49; p = 0.035) and total (95%CI 0.04 – 0.85; p = 0.03) domain. Other significant risk factors for having a concerning score were age above 10 years in social and school domains for children (95%CI 1.11 – 12.0; p= 0.032 and 95%CI 1.18 – 15.57; p = 0.027) and in school (95%CI 1.25 – 14.7; p = 0.021) domain for parents; overnight wakes in children's emotional and school domains (95%CI 1.59 – 10.92; p = 0.004 and 95%CI 1.15 – 8.84, p = 0.026). Severe pruritus significantly affected children's social domain (95%CI 1.52 – 40.4; p= 0.014).
Conclusions: Children on in-centre dialysis and their parents report poorer QoL on PedsQL and SDQ questionnaires compared to their healthy peers. School attendance may be a protective factor for having a better score and consequently a better QoL perception.

PI-39 LOW DIALYSATE SODIUM CONCENTRATION IN PEDIATRIC AND YOUNG ADULT PATIENTS ON MAINTENANCE HEMODIALYSIS: A PROSPECTIVE, RANDOMIZED, CROSSOVER STUDY

Olga Caporale[1], Silvia Consolo[1], Francesca Sofia Grassi[1], Giuseppe Puccio[2], Giovanni Montini[1], Fabio Paglialonga[1]

[1]Pediatric Nephrology, Dialysis And Transplant Unit, Fondazione Irccs Ca Granda Ospedale Maggiore Policlinico, Milan, Italy, [2]Department Of Sciences For Health Promotion, University Of Palermo, Italy

Introduction: The optimal dialysate sodium (dNa) concentration in patients on maintenance hemodialysis (HD) is still debated, especially in pediatrics. Aim of our study was to compare the effect on blood pressure (BP) and interdialytic weight gain (IDWG) of a dNa reduction to 135 mEq/l vs the standard concentration (138 mEq/l) in pediatric and young adult patients on chronic HD.
Material and methods: This single-center, prospective, randomized, crossover study consisted in a randomized sequence of two study phases: "standard dNa" of 138 mmol/L and "low dNa" of 135 mmol/L. Each study phase lasted 4 weeks and was preceded by a 2-week washout-period. Inclusion criteria were age <25 years, pre-HD serum Na (sNa) ≥ 130 mmol/L, hypertension, low incidence of intradialytic events. Study endpoints were: pre- and post-HD systolic and diastolic BP (SBP and DBP), IDWG as percentage of body weight, first-hour refill index (a BVM-based index of pre-HD fluid overload), incidence of symptomatic sessions (hypotension, cramps, vomiting), pre- and postHD sNa.
Results: Fifteen patients were recruited, mean age 17.8 ± 4.4 years (8 patients <18years). Pre- and post-HD SBP and DBP were not significantly different between the two treatments. Mean IDWG was significantly lower in the dNa 135 than in the dNa 138 phase: 2.12±1.39% vs 2.77±1.53% (p=0.008) with a mean reduction of 22.7%. The Refill Index was significantly lower with dNa of 135 mmol/L (p= 0.018). The mean sodium gradient (dNa-sNa) with dNa 138 and 135 mmol/L was 0.17±2.8 and 2.53±2.4 mmol/L respectively (p=0.0001). The incidence of symptomatic sessions was similar (1.0% vs 1.0%).
Conclusions: In a selected population of non-hypotension prone pediatric and young adult HD patients, lowering dNa to 135 mmol/L is associated with a significant reduction in IDWG over a 1-month period. Further longer-term studies are needed to investigate the effect of lowering dNa on BP in this population.

PI-40 HIGHER RELATIVE OVERHYDRATION BLUNTS AUGMENTATION INDEX IN CHILDREN ON DIALYSIS

Stella Stabouli[1], Varvara Askiti[2], Athanasia Chainoglou[1], Georgia Malakasioti[2], Vasiliki Karava[1], Maria Mila[2], Katerina Chrysaidou[1], Smaragdi Marinaki[3], Andromachi Mitsioni[2]

[1]First Pediatric Department, Medical School, Faculty Of Health Sciences, Aristotle University Thessaloniki, Hippokratio Hospital, Thessaloniki, Greece, [2]Nephrology Department, "p&a Kyriakou" Children's Hospital, Athens, Greece, [3]Clinic Of Nephrology And Renal Transplantation, National And Kapodistrian University Of Athens, Medical School, Athens, Greece

Introduction: Cardiovascular disease is highly prevalent in the chronic kidney disease population and associates with higher morbidity and mortality even in young ages. The aim of the present study was to assess the effect of hydration status on ambulatory blood pressure monitoring (ABPM) and pulse wave analysis (PWA) profiles in children on dialysis.

Material and methods: Sixteen patients on peritoneal dialysis (PD) and 12 patients on hemodialysis (HD) with similar age, underwent ABPM, PWA, echocardiography and assessment of hydration status calculating relative overhydration (rel-OH, %) by bioimpedance spectroscopy.
Results: Mean age of the cohort was 12.07±3.242 years. Rel-OH was higher in PD patients compared to HD (6.56±6.43 vs -2.57±6.67 %, p<0.005). Rel-OH did not differ between anuric patients and those with residual kidney function. Na intake was not associated with BP and PWA parameters, neither with rel-OH. We found no significant associations of rel-OH with MAP, central SBP, neither with pulse wave velocity (PWV), cardiac index (CI), and left ventricular mass index (LVMI). However, there was a negative association between heart rate-adjusted augmentation index (AIx75) and rel-OH levels (r=-0.36, p<0.05). The differences in AIx75 between those with normal rel-OH (-7 to 7%) and rel-OH > 7% persisted after adjustment for age, sex, MAP, central SBP, PWV, and dialysis modality (29.15, 95%CI 26.9-31.3 vs 22.25, 95%CI 17.9-26.59 %, respectively, p<0.05). In HD patients we also found no association between interdialytic weight gain (IDWG) % and MAP, central SBP, AIx75, CI, PWV and LVMI. CI was numerically higher in those with IDWG > 4% but the difference was not significant.
Conclusions: In children on dialysis volume overload may blunt the impact of reflecting waves in the pulse wave contour masking functional vascular abnormalities and resulting in lower AIx75 independent of BP and PWV levels. The significance of IDWG versus chronic volume overload needs further research.

PI-41 PREDICTORS OF POOR CARDIOVASCULAR STATUS IN PEDIATRIC HEMODIALYSIS PATIENTS – RESULTS FROM THE INTERNATIONAL PEDIATRIC HEMODIALYSIS NETWORK IPHN

Dagmara Borzych-duzalka[1], Rukshana Shroff[2], Sara Testa[3], Marc Fila[4], Aysun Aysun Bayazit[5], Gema Ariceta[6], Attila Szabo[7], Amrit Kaur[8], Stéphanie Tellier[9], Isabelle Vrillon[10], Loai Eid[11], Yam-ngo Lim[12], Jameela Kari[13], Marsha Lee[14], Bradley A Warady[15], Franz Schaefer[16], Claus Peter Schmitt[16]

[1]Medical University Of Gdansk, Gdansk, Poland, [2]Great Ormond Street Hospital For Children, London, Uk, [3]Fondazione Ospedale Maggiore Policlinico, Milano, Italy, [4]Chu De Montpellier, Montpellier, France, [5]Cukurova University, Faculty Of Medicine, Adana, Turkey, [6]Hospital Universitario Materno-infantil Vall D Hebron, Barcelona, Spain, [7]Semmelweis University, Budapest, Hungary, [8]Royal Manchester Children Hospital, Manchester, Uk, [9]Dialyse Pédiatrique Chu Toulouse, Toulouse, France, [10]Chru Nancy, Nancy, France, [11]Dubai Hospital, Dubai, United Arab Emirates, [12]Kuala Lumpur Hospital, Kuala Lumpur, Malaysia, [13]King Abdulaziz University Hospital, Jeddah, Saudi Arabia, [14]The University Of California, San Francisco (ucsf), San Francisco, USA, [15]Childrens Mercy Hospital, Kansas City, USA, [16]Center For Pediatrics And Adolescent Medicine, Heidelberg, Germany

Introduction: Fluid and salt overload in dialysis patients result in high blood pressure (BP), left ventricular hypertrophy (LVH) and are associated with poor outcome.
Material and methods: 954 pediatric hemodialysis (HD) patients (542M/412F), aged 0 to 21 (median 12) years on chronic HD(F), treated at 65 pediatric dialysis units in 30 countries were prospectively followed by the IPHN.
Results: In 2838 6-monthly observations 28% of patients were normotensive without antihypertensives, while 17% were normotensive on 2.1 ±1.0 antihypertensives and 55% patients were hypertensive. 24% of HD and 33% of HDF patients were normotensive without treatment (p<0.001). Systolic BP-SDS was independently predicted (PE±SEM) by intradialytic weight gain (IDWG; 0.2±0.02, p=0.0006) and younger age (-0.08±0.01, p<0.0001). Diastolic BP-SDS was predicted by younger age (-0.08±0.007, p<0.0001), dialysate sodium (0.05 ±0.01; p=0.006) and dialysis modality (HD versus HDF; PE 0.2 ±0.08; p=0.02). 4% of systolic and 12% of diastolic BP variability was explained by center effects.
Median LV mass index (LVMI) was 41.3 (31.6; 54.4); 51% patients exhibited LVH. In multivariable analysis LVMI was predicted by higher systolic BP-SDS (2.8±0.55, p<0.0001), younger age (-1.15±0.18; p<0.0001) and dialysis modality (HD versus HDF; 5.8±2.03; p=0.004), but not by UF/h, urine output/m^2 or dialysate sodium.
Intradialytic hypotension was reported in 23% of dialysis sessions and independently predicted by HD mode (0.54±0.2, p=0.007), lower urine output/m^2 (-0.44±0.16, p=0.005), higher IDWG (0.14±0.04, p=0.0001) and younger age (-0.05±0.002, p=0.008), but not by dialysate sodium, BP, UF/h and weekly dialysis time. 13% of the variability was explained by center effect.
Conclusions: High blood pressure is still prevalent in the majority of hemodialysis patients despite elaborated antihypertensive therapy. Predictive and modifiable factors of BP and LVH include, dialysis modality, dialysate sodium and IDWG. HDF is superior to HD in terms of BP control, prevention of LVH and intradialytic hypotension, independent of center effects.

PI-42 AMBULATORY BLOOD PRESSURE MONITORING AND PULSE WAVE ANALYSIS PROFILES IN PERITONEAL DIALYSIS AND HEMODIALYSIS PATIENTS

Stella Stabouli[1], Varvara Askiti[2], Athanasia Chainoglou[1], Georgia Malakasioti[2], Vasiliki Karava[1], Maria Mila[2], Katerina Chrysaidou[1], Smarargi Marinaki[3], Andromachi Mitsioni[2]

[1]First Pediatric Department, Medical School, Faculty Of Health Sciences, Aristotle University Thessaloniki, Hippokratio Hospital, Thessaloniki, Greece, [2]Nephrology Department, "p&a Kyriakou" Children's Hospital, Athens, Greece, [3]Clinic Of Nephrology And Renal Transplantation, National And Kapodistrian University Of Athens, Medical School, Athens, Greece

Introduction: Hypertension is highly prevalent in the chronic kidney disease population and associates with higher cardiovascular morbidity. The aim of the study was to compare ambulatory blood pressure monitoring (ABPM) and pulse wave analysis (PWA) profiles in children on peritoneal dialysis (PD) and hemodialysis (HD).
Material and methods: A cohort of 16 patients on PD and 12 on HD, aged 6 to 18 years, underwent ABPM and PWA using the oscillometric Mobil-O-Graph device during the 48h interdialytic period in HD or for 24h in PD.
Results: The prevalence of hypertension was 50% in the PD and 66.7% in the HD group. PD and HD patients did not differ in age (12.9±3.4 vs 10.9±2.5 years, p=0.1), mean arterial pressure (MAP) (93.1±17.6 vs 91.7 ±7.6 mmHg, p=0.7) central SBP (93.9±19.9 vs 96.6±10.1 mmHg, p=0.6) and pulse wave velocity (PWV) levels (4.1±0.6 vs 4.2±0.4 m/sec, p=0.6). The differences remained non-significant when age- or height-adjusted z-scores were used. However, heart rate-adjusted augmentation index (AIx75) (22.1±9.9 vs 30.5±6.1 %, p<0.05), cardiac index (CI) (3.3±0.5 vs 4.3±0.9 l/m^2, p<0.005), and HR (84.9±12.6 vs 94.2 ±9.3 beats/min, p<0.5) were significantly higher in the HD group. In HD patients, MAP and central SBP showed significant increases from the first to the second monitoring day (91.5±5.3 vs 96.3±6.1, p<0.001, and 103.9 ±5.2 vs 108.4±5.6, mmHg, p<0.005, respectively), while AIx75, Cl and PWV presented similar values in both days. The differences in MAP

central SBP and PWV between PD and HD patients remained non-significant both in the first and second monitoring day.
Conclusions: Despite gradual BP increases during the interdialytic period in HD, PD patients had comparable BP and PWV levels suggesting similar cardiovascular risk between the different dialysis modalities. However, the increased levels of AIx75 and CI in HD patients could imply different patterns of vascular and cardiac functional changes.

PI-43 RELATIONSHIP OF CIRCULATING IRISIN WITH BODY COMPOSITION IN CHILDREN WITH CHRONIC KIDNEY DISEASE

Emre CEYHUN[1], Seha SAYGILI[2], Sergen DEVRAN[3], Rüveyda GÜLMEZ[2], Kaan Can DEMİRTAŞ[4], Şevval ÇELEN[5], Ayşe AĞBAŞ[2], Salim ÇALIŞKAN[2], Bülent BAYRAKTAR[3], Nur CANPOLAT[2]

[1]Istanbul University – Cerrahpasa, Cerrahpasa Medical Faculty, Department of Pediatrics, *[2]Istanbul University – Cerrahpasa, Cerrahpasa Medical Faculty, Department of Pediatric Nephrology*, *[3]Istanbul University, Faculty of Medicine, Sports Medicine Department*, *[4]Istanbul University – Cerrahpasa, Cerrahpasa Medical Faculty*, *[5]Istanbul University, Faculty of Sports Science*

Objective: Children with chronic kidney disease (CKD) suffer from decreased exercise capacity due to the uremic environment, protein-energy-wasting, oxidative stress, and inflammation. Low levels of irisin, a recently discovered exercise-induced myokine, have been linked to adverse metabolic outcomes. The aim of this study was to evaluate the serum irisin levels in children with CKD and to analyze its potential associations with body composition, estimated glomerular filtration rate (eGFR), markers of oxidative stress, and inflammation.
Design and methods: This cross-sectional, single-center study enrolled 39 children with CKD (22 male, aged 7.7 to 20.7 years, 26 children CKD stage 3 and 4 and 13 CKD 5D) and age and gender compatible 29 healthy children. Standard deviation scores (SDS) of height and body mass index (BMI) for height age were calculated according to national percentiles. Body composition parameters were measured with the multiple-frequency bioimpedance device. Serum concentrations of irisin, CRP, TNF-alpha, total antioxidant capacity (TAS), and total oxidant capacity (TOS) were analyzed by ELISA assays.
Results: The number of underweight, overweight, and obese children in the CKD group was 4 (10.3%), 5 (12.8%), and 7 (17.9%), respectively. Although there were no differences in BMI-SDS or body composition parameters between the patient and control groups, median serum irisin level was lower in the CKD group than in the control group (3.97 vs 4.54 ng/ml, p= 0.03). However, there was no difference in irisin levels between patients with CKD 3-4 and CKD 5D. Serum irisin levels showed no association with BMI-SDS, body composition parameters, eGFR, CRP, TNF-alpha, TAS, TOS, or serum lipid levels.
Conclusions: This study demonstrates decreased serum irisin concentrations in pediatric CKD patients. However, there is no association of irisin with any body composition parameters, inflammatory or oxidative stress markers. Further studies are needed to investigate the factors affecting serum irisin levels in children with CKD.

PI-44 THE RELATIONSHIP BETWEEN INSULIN RESISTANCE AND SERUM ADIPOKINE LEVELS IN CHILDREN WITH CHRONIC RENAL DISEASE RECEIVING PERITONEAL DIALYSIS

Ceren BilgÜn[1], Nurdan Yildiz[2], Ali Yaman[3], Goncagul Ustunel Haklar[3], Harika Alpay[2]

[1]Marmara University, Medical School, Division Of Pediatrics, *[2]Marmara University, Medical School, Division Of Pediatric Nephrology*, *[3]Marmara University, Medical School, Division Of Biochemistry*

Introduction: Adipose tissue secretes adipokines which have important metabolic and endocrine functions.In chronic kidney disease(CKD), insulin resistance (IR) and malnutrition increase morbidity and mortality by increasing the risk of cardiovascular diseases.
We aimed to evaluate IR,its relationship with adiponectin and resistin in peritoneal dialysis(PD) and predialysis stage 2-4 CKD patients, and to investigate the relationship of IR with peritoneal transport properties and body composition.
Material and methods: Twenty PD, 20 CKD patients and 40 healthy children were included in this prospective cohort study.Demographic, clinical and laboratory findings were recorded from the medical files. Anthropometric measurements and bioimpedance analysis were performed.Serum insulin, adiponectin and resistin levels were measured. HOMA-IR and HOMA-AD were calculated for all groups.
Results: The mean adiponectin levels were 281.6±74.9 ng/mL in PD, 172.3±81.2 in CKD patients and 112.2±57.8 ng/mL in controls. The mean resistin levels in PD, CKD patients and controls were 5.5±2.1, 3.8±1.5 and 1.3±0.5,respectively. Resistin and adiponectin levels were higher in PD patients compared to CKD(p=0.006 and <0.001) and controls(p=<0.001 and < 0.001). There was negative correlation between resistin and e-GFR whereas no significant relation was observed between adiponectin and e-GFR.
Insulin resistance was found in 5(%25) PD,13(%65) CKD patients and 19(%47.5) controls whose HOMA-IR>2.5. HOMA-IR were higher in CKD patients than PD(p=0.018).However, it was not different in PD and CKD patients compared to controls(p>0.05).Resistin was not associated with IR in PD and CKD patients(p>0.05). There was no significant correlation between Kt/v and anthropometric measurements, insulin, HOMA-IR, HOMA-AD, resistin, adiponectin in PD patients(p>0.05).
Conclusions: Insulin resistance may develop in PD patients and in the early stages of CKD, and should be closely monitored to reduce cardiovascular disease in adult life.Resistin is not a good marker to determine IR in children with PD and CKD.Studies with larger series are needed to evaluate the relationship between inflammation, IR and adipokines.

PI-45 ANTI-FGF23 TREATMENT IN A CASE OF AUTOSOMICAL RECESSIVE HYPOPHOSPHATEMIC RICKETS TYPE 2

Nicola Bertazza Partigiani, Davide Meneghesso, Germana Longo, Elisa Benetti, Mattia Parolin

Pediatric Nephrology And Dialysis Unit, Department Of Women's And Child's Health, University Of Padova Medical School, 35128 Padova, Italy

Introduction: Hypophosphatemic rickets is a disorder of phosphate metabolism secondary,to alterations in FGF23 metabolism.Inactivating mutations of the ENPP1 gene(ARHR2)are responsible for the development of rickets, without necessarily developing an increase in FGF23,which is however inappropriate high for the phosphatemia.Burosumab,a recombinant anti-FGF23 monoclonal antibody, is effective in the treatment of PHEX in pediatric patients,but its use in other forms of hypophosphatemic rickets is not proven.
Material and methods: A 9-year-old girl,followed for ARHR2 due to homozygous ENPP1 mutation (c.1709A>Gp.Tyr570Cys),stopped supplementation therapy due to adverse events and subsequent difficult in compliance.For this reason,,treatment with Burosumab was started,initially at a dose of 0.4mg/kg every two weeks,and then increased

to 0.8mg/kg 6 months after for partial response.Patient underwent regular clinical evaluation,laboratory exams, and radiological evaluation, to assess clinical improvement and to recognize signs of Generalized Arterial Calcification of Infancy(GACI), due to ENPP1 mutation.
Results: At the onset she presents valgus knee,mild nephrocalcinosis,marked hypophosphatemia(0.43mmol/L),increase in ALP(732 U/L),hyperphosphaturia and a slight increase in FGF23: 1.57pmol/L (0-0.8).The initial dose allowed a partial improvement,while increasing the dose to 0.8mg/kg,ALP reached 499U/L with serum phosphate of 0.75mmol/L,after 18 months.Growth was good(75-90th percentile).Wrist and knee radiographs demonstrate an improvement in the RSS score from 4 to 2.No signs of vascular calcification were detected.
Conclusions: This case represents the first patient with ARHR2 effectively treated with Burosumab. Initially we scheduled a low dose considering the risk of vascular calcification in the ENPP1 mutation.However we increased the dose obtaining good results on phosphate and bone metabolism,demonstrating the efficacy and safety of Burosumab in this kind of mutation.

PI-47 VITAMIN-D DEPENDENT RICKETS TYPE 1A: PHENOTYPE GENOTYPE CHARACTERIZATION OF 24 PATIENTS WITH CYP27B1 MUTATION

Meaux Marie-noëlle[12], Harambat Jérome[3], Rothenbuhler Anya[4], Léger Juliane[4], Kamenicky Peter[4], Sylvie Soskin[5], Boyer Olivia[4], Emese Boros[6], Danella Pascal[7], Mignot Brigitte[8], Gebhart Maite[8], Vic Philippe[9], Richard Nicolas[10], Thivichon-prince Beatrice[11], Francou Bruno[4], Linglart Agnes[4], Bacchetta Justine[11], Molin Arnaud[2]

[1]Chu De Lyon, Service De Néphrologie, Rhumatologie Et Dermatologie Pédiatriques, Lyon, France, [2]Chu De Caen, Service De Génétique, Caen, France, [3]Bordeaux, [4]Paris, [5]Strasbourg, [6]Bruxelles, [7]Avignon, [8]BesanÇon, [9]Quimper, [10]Caen, [11]Lyon, [12]Chu De Bordeaux, Service De Néphrologie Pédiatrique, Bordeaux, France

Introduction: Vitamin D dependent rickets type 1A (VDDR1A) is an autosomal recessive disease due to biallelic loss of function variants in the CYP27B1 gene, encoding the 1α-hydroxylase enzyme that activates vitamin D. The objectives of this study were to describe clinical data, genetic features and outcomes in a European population of VDDR1A, and to analyze genotype – phenotype correlations.
Material and methods: We performed a multicentric retrospective study. Data from 24 genetically confirmed cases of VDDR1A from 10 centers were retrospectively reviewed. Data are presented as median [min - max].
Results: Clinical symptoms at diagnosis were mainly bone and neurological abnormalities. Age at diagnosis was 1.5 [0.5 - 8.7] years. Laboratory data at diagnosis showed mild hypocalcemia (1.97 [1.40 - 2.40] mmol/L) and hypophosphatemia (- 3.4 [- 13.4 - (-) 0.2] SDS), low 25OHD (23 [7 - 81] ng/mL) and low $1,25(OH)_2D_3$ (14 [7 - 82] pg/mL), secondary hyperparathyroidism with PTH at 6.6 [1.3 - 13.7] times ULN and increased alkaline phosphatases (2041 [521 - 7000] UI/L). Bone X-rays were abnormal for 80% patients. Outcome under substitutive treatment by alfacalcidol (median dose 1 μg/day) was considered satisfactory in case of good adherence. Median adult height was 164 centimeters (160 in women, 167 in men). Median blood pressure at last follow-up was at the upper limit normal. Five cases of nephrocalcinosis were described, which normalized. Dental abnormalities were frequent. There were 17 different mutations and the recurrent p.(Ala129Thr) substitution (1 compound heterozygous and 4 homozygous cases) was associated with a milder phenotype with older age at diagnosis and often normocalcemia.
Conclusions: VDDR1A is a rare, genetically heterogenous disease. Our findings are consistent with previous studies except for younger age at diagnosis, inconstant hypocalcemia and lower 25OHD levels. They highlight the need of closer follow-up of eyes, teeth, kidneys and blood pressure in these patients.

PI-48 FGF23, IRON STATUS AND ANEMIA IN CHILDREN WITH CHRONIC KIDNEY DISEASE

Vasiliki Karava[1], John Dotis[1], Antonia Kondou[1], Athanasios Christoforidis[2], Anna Taparkou[3], Evangelia Farmaki[3], Marina Economou[4], Nikoleta Printza[1]

[1]Pediatric Nephrology Unit, 1st Department Of Pediatrics, Hippokratio General Hospital, Aristotle University Of Thessaloniki, Greece, [2]Pediatric Endocrinology Unit, 1st Department Of Pediatrics, Hippokratio General Hospital, Aristotle University Of Thessaloniki, Greece, [3]Pediatric Immunology And Rheumatology Referral Center, 1st Department Of Pediatrics, Hippokratio General Hospital, Aristotle University Of Thessaloniki, Greece, [4]Pediatric Hematology Unit, 1st Department Of Pediatrics, Hippokratio General Hospital, Aristotle University Of Thessaloniki, Greece

Introduction: This cross-sectional study investigates the association of fibroblast growth-factor 23 (FGF23) with iron status and anemia in children with moderate and advanced chronic kidney disease (CKD).
Material and methods: Serum calcium, phosphorus, 25(OH)D, intact parathormone, c-terminal FGF23 and a-Klotho were measured in 53 patients from 5 to 19 years old with GFR<60 ml/min/1,73m^2. Serum iron (Fe), ferritin and unsaturated iron-binding capacity, blood Hb and red blood cell indices were measured the same day. Transferrin saturation (TS) was calculated for each patient.
Results: LnFGF23 was correlated to lnKlotho (rs=-0.321, p=0.020) after adjustment for CKD stage. In 36 patients with CKD stage 3-4, lnFGF23 was correlated to Fe (rs=-0.415, p=0.013) and TS (rs=-0.356, p=0.036) but not to ferritin (rs=0.062, p=0.725) after adjustment for CKD stage. In CKD stage 5 patients, no correlation was observed between lnFGF23 and iron parameters. No correlation was observed between lnKlotho and iron parameters in both patient groups. In CKD stage 3-4 patients, TS was correlated to Hb (rs=0.465, p=0.004), red blood cell mean corpuscular volume (rs=0.360, p=0.031) and distribution width (rs=-0.392, p=0.018). In this patient group, 10 patients presented low Hb, 8 of which also presented low TS (<16%). In multivariate backward logistic regression analysis, lnFGF23 was associated with low TS and low Hb, after adjustment for bone mineral parameters and CKD stage (OR 4.402, 95% CI 1.118-17.336, p=0.034 and OR 5.145, 95% CI 1.039-25.487, p=0.045 respectively).
Conclusions: In pediatric CKD stages 3-4, FGF23 is possibly implicated in the disturbed iron metabolism and consequent anemia, independently of Klotho.

PI-50 IMPRECISION IN GFR ESTIMATES - IMPACT ON CKD STAGING

Janusz Feber[1], Ivan Blasutig[2], Robert L. Myette[3], Robert Gow[1]

[1]Childrens Hospital Of Eastern Ontario, University Of Ottawa, [2]Cheo, University Of Ottawa, Eastern Ontario Regional Laboratory Association, [3]Cheo, University Of Ottawa, Kidney Research Center, The Ottawa Hospital Research Center

Introduction: Schwartz GFR (SchwGFR) is estimated from height (cm) and serum creatinine (SCr) and is used for staging of chronic kidney disease (CKD). The SchwGFR formula may introduce additional propagation error in GFR estimation, in addition to intrinsic variability/error in measuring SCr and cm.

The aim of the study was to analyze the propagation error (PE) of SchwGFR and its impact on CKD classification.
Material and methods: All available SCr results obtained from June 2021 to December 2021 were retrieved from the lab (924 samples from 648 patients). SchwGFR (point estimates, ml/min/1.73 m2) were calculated using the formula published by Schwartz et al (2009). Lab specific standard deviations (SD) of SCr assays (mean SD = 5 umol/l) and variability of height measurements (mean SD = 0.5 cm) were used to calculate PE in SchwGFR (composite error/SD propagated from SCr and height measurements), which resulted in interval-based GFR (SchwGFRi) with individual 95% confidence intervals. The agreement/disagreement between SchwGFRi and traditional point estimated GFR (SchwGFRp) was then analyzed (Cohen's kappa, test of proportions) to inform classification of CKD stages. Samples were classified as CKD if the lower limit of the SchwGFRi was below the CKD classification threshold of 90 ml/min/1.73 m2.
Results: Mean SchwGFR PE (%) [89% prediction intervals] were: 1.49 [1.04-1.95], 1.83 [1.37-2.31], 2.17 [1.67-2.64] and 2.51 [2.01-2.95] at GFR levels of 30, 60, 90 and 120 ml/min/1.73 m2, respectively. SchwGFRp identified CKD (GFR < 90) in 411/924 samples, whereas SchwGFRi detected CKD in an additional 49/924 (5.3%) samples ($p<0.02$), mostly CKD2.
Conclusions: The propagation error of SchwGFR estimates was relatively small (up to 3%) but increased with increasing GFR and allowed for calculation of individual GFR "trusted" intervals. This led to a significantly higher number (additional 5%) of GFR samples classified as CKD by interval estimates compared to traditional point estimates.

PI-51 ILLNESS-RELATED PARENTAL STRESS AND QUALITY OF LIFE IN CHILDREN WITH CHRONIC KIDNEY DISEASE: A MULTI-CENTRIC STUDY

Elke De Bruyne[1], Lore Willem[2], Koen Van Hoeck[3], Sarah Reynaert[3], Sylvie Vankerckhove[4], Brigitte Adams[4], Stephanie Leroi[5], Laure Collard[5], Aline Michaux[6], Nathalie Godefroid[6], Djalila Mekahli[2], Noël Knops[2], Eline Van Hoecke[1], Sunny Eloot[7], Ann Raes[8], Evelien Snauwaert[8], Johan Vande Walle[8], Elena Levtchenko[2]

[1]Pediatric Psychology, Department Of Pediatrics, Ghent University Hospital, Belgium, [2]Department Of Child Nephrology And Organ Transplantation, Leuven University Hospital, Belgium, [3]Department Of Pediatric Nephrology, Antwerp University Hospital, Belgium, [4]Department Of Pediatric Nephrology, Queen Fabiola Childrens University Hospital Brussels, Belgium, [5]Department Of Pediatric Nephrology, Chc Health Group Montlégia Clinic,liege, Belgium, [6]Department Of Pediatric Nephrology, Saint-luc Brussels University Hospital, Belgium, [7]Department Of Nephrology, Ghent University Hospital, Belgium, [8]Department Of Pediatric Nephrology & Rheumatology, Ghent University Hospital, Belgium

Introduction: Monitoring the psychological well-being of children with chronic kidney disease (CKD) is seen as standard care in pediatric nephrology, as many studies have shown that CKD has a great psychological impact. This multi-centric cross-sectional study investigated quality of life (QoL) and illness-related parental stress in this population by 1/ comparing mean levels of these two variables between several CKD categories, and 2/ exploring their correlation.
Material and methods: We recruited children with CKD and their parents, followed at the 6 Belgian revalidation reference centers for child nephrology. Childrens QoL was assessed by the PedsQL^{TM} 4.0 Generic Core Scales, parental stress was measured by the Pediatric Inventory for Parents (PIP). All patients were divided in categories based on their CKD diagnosis: 1/congenital diseases 2/ tubulopathies and metabolic diseases, 3/ nephrotic syndromes, 4/ acquired diseases with proteinuria and hypertension, and 5/ kidney transplantations.
Results: In total we included 295 children (176 boys; M age= 11.8, SD = 3.7) and 285 parents. Fifty-seven children (19%) had transplant in the past. There were no significant differences in QoL between CKD categories as reported by the children ($p>.05$). In contrast, there were significant differences between CKD categories in QoL ($F(4, 220) = 3.46$, $p<.01$)) and stress ($F(4,269) = 2.92$, $p<.05$), reported by parents, with transplant patients having lower QoL ($t(220) = -3.31$; $p = .001$) and higher parental stress ($t(269) = 2.30$; $p = .02$). Finally, there were significant negative correlations ($p < .001$) between QoL and parental stress.
Conclusions: This multi-centric study showed lower levels of QoL and higher levels of parental stress in transplanted children with CKD, compared to children without transplant, when based on parent reports. More parental stress is associated with worse QoL in the child. These results highlight the importance of a multidisciplinary team with special attention for the parents.

PI-52 KIDNEY DYSFUNCTION IN MEDICALLY STABLE ADOLESCENTS WITH EATING DISORDERS AND THE VALUE OF CYSTATIN-C

Ayse Bilge Baklaci[1], Nuray Kanbur[2], Berna Oguz[3], Filiz Akbiyik[4], Melis Pehlivanturk Kizilkan[2], Sinem Akgul[2], Orhan Derman[2], Ercan Ayaz[3], Mithat Haliloglu[3], Rezan Topaloglu[5], Ali Duzova[5]

[1]Department Of Pediatrics, Hacettepe University Faculty Of Medicine, Ankara, Turkey, [2]Division Of Adolescent Medicine, Hacettepe University Faculty Of Medicine, Ankara, Turkey, [3]Department Of Radiology, Hacettepe University Faculty Of Medicine, Ankara, Turkey, [4]Ankara City Hospital Siemens Healthineers Laboratory, Ankara, Turkey, [5]Division Of Pediatric Nephrology, Hacettepe University Faculty Of Medicine, Ankara, Turkey

Introduction: Eating disorders may cause renal complications. Creatinine measurement may be misleading due to loss of muscle mass in this group of patients. We aimed to determine the frequency of kidney dysfunction, and to evaluate the value of cystatin-C in adolescents with eating disorders.
Material and methods: Medically stable but not weight restored 41 patients (36 female, 5 male; mean age 15.93 ± 1.47 years; mean follow-up time 12.1±12.3 months) with anorexia nervosa or bulimia nervosa according to DSM-5 at our tertiary referral center, between January 2020 and August 2020, were included. Serum biochemistry markers, serum cystatin-C, complete blood count, urinalysis, urinary protein and electrolytes, estimated glomerular filtration rate (eGFR), 24-hour ambulatory blood pressure (BP) monitorization (ABPM), and renal ultrasonography were evaluated in all patients. An age and sex matched control group was composed for serum creatinine and cystatin-C measurements.
Results: Microalbuminuria, macroalbuminuria, hypostenuria, leukocyturia (sterile), and hypercalciuria was seen in 19.4%, 2.8%, 32.5%, 7.5%, and 15.8% of cases, respectively. Although serum creatinine levels were comparable to control, serum cystatin-C levels were found to be significantly lower; eGFR values, calculated with different creatinine and/or cystatin-C based methods, showed that 0-9.8% of patients had an eGFR <90 ml/min/1.73m^2. Ultrasonographic examination did not reveal nephrolithiasis or nephrocalcinosis. 24-hour ABPM showed that BP levels were significantly low; more remarkable for systolic BP (SBP) and during daytime (31.3% had daytime SBP <5th percentile).
Conclusions: Cystatin-C may also have limitations for the evaluation of kidney functions due to decreased adipose tissue in patients with eating disorders. There were significant inconsistencies between different

creatinine and cystatin-C based eGFR methods. ABPM may be useful in addition to routine laboratory tests. Longitudinal studies are needed to determine the values of these methods in the management of adolescents with eating disorders.

PI-54 EFFICACY AND SAFETY OF CINACALCET THERAPY IN YOUNGER CHILDREN WITH SECONDARY HYPERPARATHYROIDISM RECEIVING MAINTENANCE DIALYSIS.

Ilona Zagożdżon, Irena Bałasz- Chmielewska, Aleksandra Skibiak, Aleksandra Zurowska

Department Of Pediatrics, Nephrology And Hypertension. Medical University Of Gdansk, Poland

Introduction: Secondary hyperparathyroidism (SHPT) commonly occurs in children receiving dialysis causing numerous complications due to CKD- MBD. When conventional therapy of SHPT is ineffective calcimimetics may control symptoms but have been approved for children over 3 years of age. Scarce information is available for their use in the youngest age group.
The aim of the study was to assess the efficacy and safety of cincacalcet therapy in children below 3 years of age.
Material and methods: Case records of 31 chidren dialysed under 3 yrs of age were screened for the use of calcimimetics. Clinical symptoms and iPTH, Ca and P levels and response to treatment were analysed. Clinical manifestation of SHPT were bone deformities in one case and growth retardation in two cases.
Results: Three patients were identified all of whom had started peritoneal dialysis in infancy (1-8 months age). At a mean age of 27 months cinacalcet was introduced at an initial dose of 0.23mg/kg. Initial mean iPTH was 1968 pg/ml and calcium 11,7mg/dl. Therapy was continued over 9-36 months. The cinacalcet dose was titrated every 4 weeks based on iPTH and calcium threshold to a maximum dose 2,2mg/kg. The patients remained on stable doses of vitamin D analogues and phosphate binders. A spectacular 70% reduction of iPTH was observed while corrected calcium decreased insignificanty to 10,8mg/dl. Improvement of bone deformities was noted after 24 months of treatment. A single episode of mild asymptomatic hypocalcaemia occurred in the first month of treatment in one child and precoccius puberty in another.
Conclusions: 1. Cinacalcet therapy was effective in children with SHPT below 3 years age.
2. With careful monitoring Cinacalcet was safe to use with insignificant decrease in calcium levels during treatment.
3. The association of cinacalcet treatment and precocius puberty requires further evaluation as two previous reports have been published concerning this finding.

PI-55 IDENTIFYING AREAS FOR IMPROVEMENT IN PATIENT EXPERIENCES OF CHILDREN WITH KIDNEY AND ONCOHEMATOLOGICAL DISEASES AND THEIR PARENTS: A FOCUS GROUP STUDY

Karolis Azukaitis[1], Goda Vaitkeviciene[1], Birute Mockeviciene[2], Augustina Jankauskiene[1], Danguole Jankauskiene[2]

[1]Clinic Of Pediatrics, Institute Of Clinical Medicine, Faculty Of Medicine, Vilnius University, Vilnius, Lithuania, *[2]Mykolas Romeris University, Vilnius, Lithuania*

Introduction: Understanding patient experiences within healthcare system is an integral part of patient-centered care and essential for healthcare quality improvement. We aimed to evaluate problematic experiences of children with conditions requiring long-term treatment and their families throughout whole patient journey by the example of pediatric nephrology and oncohematology.
Material and methods: We conducted 2 focus groups with purposively sampled children (n=11) and parents (n=12) from Lithuania. Groups consisted of patients and parents of children with long-standing history of glomerular diseases, chronic kidney disease, solid tumors, leukemia and after hematopoietic stem cell or kidney transplantation. Unstructured interviews aiming to assess their experiences requiring improvement throughout different stages of patient journey (diagnosis, treatment, ongoing care and rehabilitation) were conducted. All data were transcribed and then thematically analyzed using content analysis and NVivo package for qualitative studies.
Results: Insufficient disease-specific information and psychological support, as well as complicated access to specialized healthcare services were identified as relatively consistent problems throughout all stages of patient journey. Lengthy initial diagnosis process within primary healthcare and perceived poor quality of their services were highlighted during the diagnosis stage. Non-immediate access or procedural complexity to access certain therapies and financial burden on family were additional problems identified during the treatment and ongoing care stages. Inefficient and non-flexible reintegration to educational system and lack of disease-specific knowledge by staff were most important during rehabilitation stage. Lack of empathy from healthcare staff was commonly mentioned across variety of contexts.
Conclusions: Improving patient information on their health and healthcare plan, psychological support and accessibility to specialized care are top priorities for healthcare improvement in children and their families with kidney and oncohematological diseases. Empathic communication, as well as improvements in diagnostic pathways within primary healthcare and reintegration to education system are important. Results, however, should be interpreted considering country specificity.

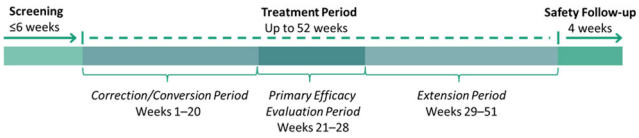
PI-56 SAFETY AND EFFICACY OF VADADUSTAT FOR THE TREATMENT OF PEDIATRIC PATIENTS WITH CHRONIC KIDNEY DISEASE (CKD)–RELATED ANEMIA

Franz Schaefer[1], Alicia Neu[2], Rosa Real[3], Andrew Blair[4], Christine Solinsky[4], Zhiqun Zhang[4]

[1]Heidelberg University, *[2]Johns Hopkins University*, *[3]Otsuka America Pharmaceutical, Inc.*, *[4]Akebia Therapeutics, Inc.*

Introduction: Vadadustat (VADA) is an oral hypoxia-inducible factor (HIF) prolyl hydroxylase inhibitor. We aimed to assess the safety, efficacy, and pharmacokinetic (PK)/pharmacodynamic (PD) properties of once-daily VADA in pediatric patients with CKD-related anemia.
Material and methods: Two phase 3, multicenter, single-arm, open-label studies are evaluating once-daily oral VADA for treatment of pediatric patients (≥4 months to <17 years) with dialysis-dependent (DD) or non–dialysis-dependent (NDD) CKD-related anemia either naïve to erythropoiesis-stimulating agents (ESA-naïve trial; NCT05082584) or after conversion from an ESA (ESA-treated trial; NCT05082571). Key inclusion criteria are diagnosis of anemia of CKD, estimated glomerular filtration rate of >10 and <60 mL/min/1.73 m^2 or a diagnosis of DD-CKD, transferrin saturation ≥20%, and mean hemoglobin (Hb) ≥9.0 and ≤12.0 g/dL (ESA-treated trial) or <10.0 g/dL (ESA-naïve trial). Patients are being enrolled in staggered cohorts stratified by age (12-17 y, 6-11 y, 2-5 y, 4 mo-<2 y); target enrollment for each study is up to 71 patients. The treatment period will consist of a correction/conversion period (weeks 1–20), primary evaluation period (PEP) (weeks 21–28),

and extension period (weeks 29–51), in which all patients will receive once-daily VADA from baseline through week 52. A 4-week safety follow-up period is planned after end of treatment (**Figure 1**). Primary efficacy endpoint is mean change in Hb between baseline and PEP. Additional endpoints include time to achieve Hb ≥10.0 g/dL and proportion of patients with mean Hb values of 10.0–12.0 g/dL during PEP and extension period; safety and tolerability, including adverse events; and PK/PD endpoints.



Results: These studies will be the first evaluation of VADA in pediatric patients with anemia of CKD.
Conclusions: Two studies are assessing safety, efficacy, and PK/PD of VADA in pediatric patients with CKD-related anemia (DD or NDD) either naïve to or after conversion from an ESA.

PI-57 USING THE URINE PROTEOMIC SPECTRUM TO OBSERVE CHILDREN HAVING CHRONIC KIDNEY DISEASE

Ekaterina P. Krivonosova, Gadgy M. Letifov

Federal State Budgetary Educational Institution Of Higher Education "rostov State Medical University" Of The Ministry Of Health Of The Russian Federation

Introduction: The study of the proteomic spectrum of urine allows us to assess the likelihood of progression of various nephropathies, which is relevant, given the increasing incidence of diseases of the urinary system in children, including those with asymptomatic course.
The aim of the study was to search for informative non-invasive markers of renal parenchyma damage in children with chronic kidney disease.
Material and methods: Proteomic study of urine was performed using proteomics methods (MALDI-TOF-MS/MS, Ultraflex II, Bruker, USA). Information about molecular interactions was obtained using the STRING 10.0 database. The study included 30 children aged 1 to 18 years with chronic kidney disease, the leading laboratory symptoms were microalbuminuria (MAU) and microhematuria.
Results: The level of MAU in the examined children was: A0 (up to 10 mg / day) - in 17% (5 patients), A1 (from 10 to 30 mg / day) - in 13% (4 children), A2 (from 30 to 299 mg / day) - in 60% (18 children), A3 (from 300 mg / day and more) - in 10% (3 people). 45 different proteins have been isolated. In isolated erythrocyturia, tubulointerstitial nephritis antigen (100%), aquaporin-1 (75%), platelet growth factor β (65%), vasorin (50%), and antiepithelial membrane antigen (50%) were most frequently detected. With the addition of MAU, the frequency of detecting a molecule of damage to the kidney tissue (100%), an apoptosis-inducing factor (100%), aquaporin-1 (100%), and neutrophilic gelatinase-bound lipocalin (75%) increased. As the process progressed, in stage III chronic kidney disease, the proteomic spectrum of urine was determined by the factor stimulating prostacyclin, interleukin 16, matrix metalloproteinase, tolloid-like protein 2.
Conclusions: Thus, the assessment of the proteomic spectrum of urine makes it possible to identify non-invasive markers of the progression of damage to the tubulointerstitial tissue of the kidneys associated with various pathologies accompanied by MAU and microhematuria.

PI-58 FOLLOW UP OF EXTREMELY PRETERM NEONATES WITH NEPHROCALCINOSIS – A SINGLE CENTER EXPERIENCE

Austeja Ivaskeviciene, Violeta Sevcenko, Rasa Garunkstiene, Arunas Liubsys, Andrius Cekuolis, Rimante Cerkauskiene

Vilnius University Faculty Of Medicine, Vilnius, Lithuania

Introduction: Preterm neonates are at risk of nephrocalcinosis (NC). The etiology of NC is multifactorial. There is insufficient data whether prematurity increases the risk of NC and renal failure later in life, or if there are any other risk factors such as hypervitaminosis D. The aim of our study was to investigate whether extreme prematurity, vitamin D concentration, hypercalcemia and hypercalciuria affect kidney function and nephrocalcinosis development.
Material and methods: A prospective study of extremely preterm infants ≤ 31 weeks of gestational age with NC born in Vilnius University Hospital Santaros Klinikos between 2018 - 2022 was performed. The following data was collected at birth with a follow up at 1, 6, 12 and 24 months: weight, height, vitamin D, serum calcium, urine calcium/creatinine ratio and renal ultrasound.
Results: Out of 160 extremely preterm infants 45 (28%) had NC, 66.7% of them were boys. Mean age was 27.4 ± 2.2 weeks. Mean birth weight 1063.1 ± 326.0 g, and height 34.9 ± 4,3 cm. At age of 6 months calcium/creatinine ratio was 1,28 [0.45-1.99] with NC vs. 0.45 [0.22-1.04] without NC. There was no correlation between vitamin D concentration and NC in all age groups. NC persisted in 69.8%, 62.5% and 46.2% after 6, 12, 24 months respectively. After 6 and 12 months NC was more frequent 6.4 [1.53-26.78] (p=0,01) and 5.6 [1.146-27.37] (p=0,015) after more than 66 days of hospitalization in neonatal period respectively. A positive family history of kidney stones was 26.7%.
Conclusions: Longer hospitalization and hypercalciuria, but not vitamin D and serum calcium concentration were risk factors for NC. Long term follow up of extremely premature infants with NC is recommended.

PI-59 EVALUATION OF TRACE ELEMENT LEVELS IN CHRONIC KIDNEY PATIENTS IN CHILDHOOD

Ali MuratoĞlu[1], Beltİnge DemircioĞlu KiliÇ[2], Mehtap Akbalik Kara[2], Seyithan Taysi[3], Mithat BÜyÜkÇelik[2], AyŞe Balat[2]

[1]Hatay Education And Research Hospital, Department Of Pediatrics, Hatay/turkey, [2]Gaziantep University, Department Of Pediatric Nephrology, Gaziantep/turkey, [3]Gaziantep University, Faculty Of Medicine, Department Of Medical Biochemistry, Gaziantep, Turkey

Introduction: Trace elements are found in very low concentrations in biological fluids or tissues, which can sometimes cause deficiency and toxicity in chronic kidney disease (CKD) patients. In our study, we aimed to evaluate trace element levels in childhood CKD.
Material and methods: Children who were diagnosed as CKD (n:64) with GFR below 60 ml/min/1.73 m^2 and healthy children (n:25) were included in the study. Patients with CKD were divided into stage 3-4 CKD (n:27), haemodialysis (HD) (n:12) and peritoneal dialysis (PD) (n:25) groups. Zinc (Zn) and copper (Cu) were measured in serum with a fully automatic biochemistry autoanalizer. Selenium (Se), manganese (Mn), lead (Pb), cadmium (Cd), chromium (Cr) and nickel (Ni) were measured by inductively coupled plasma optical emission spectrometry (ICP-OES) method.
Results: Serum Cu median value; In the control group, 178 μg/dl (135.2-270.3); In stage 3-4 CKD patients, 129.4 μg/dl (108.6-139.1); In HD patients, 106.35 μg/dl (96.25-130.5); In PD patients, 135.4 μg/dl (122.5-144.4) was detected. Copper level was found higher in the control

group compared to the patient groups, and this difference was significant (p=0.001). Serum Cr median value; In the control group, 101.7 μg/L (91.3-170.2); In stage 3-4 CKD patients, 102.1 μg/L (82.3-140.2); In HD patients, 147.2 μg/L (101.95-191.65); In PD patients, 112.1 μg/L (92.3-132.1) was detected. Crom level was found higher in HD group, but this difference wasn't significant (p=0.458). There was no significant difference between the groups in serum Zn, Ni, Se, Mn, Pb and Cd levels.
Conclusions: In the evaluation of eser elements, it was determined that only Cu level was low in the patients. Although Cr level, which is one of the toxic trace elements, is higher in HD patients compared to other groups, a statistically significant level could not be determined. We believe that, multicenter studies with more patients and control groups are needed on this subject.

PI-60 IMPAIRED T CELL RECEPTOR REPERTOIRE IN CHILDREN WITH CHRONIC KIDNEY DISEASE

Emine Ulgen[1], AyÇa Kiykim[2], Seha Saygili[3], Nihan Burtecene[2], Haluk ÇokuĞraŞ[2], Salim ÇaliŞkan[3], Nur Canpolat[3]

[1]Department Of Pediatrics, CerrahpaŞa Faculty Of Medicine, Istanbul University-cerrahpaŞa, Istanbul, Turkey, [2]Department Of Pediatric Allergy And Immunology, CerrahpaŞa Faculty Of Medicine, Istanbul University-cerrahpaŞa, Istanbul, Turkey, [3]Department Of Pediatric Nephrology, CerrahpaŞa Faculty Of Medicine, Istanbul University-cerrahpaŞa, Istanbul, Turkey

Introduction: Chronic kidney disease (CKD) is associated with immune dysregulation, including impaired cellular immune function. T lymphocytes are actively involved in the cellular immune response by recognizing and presenting antigens and cause the target cell to be lysed with a cytotoxic effect. The T cell receptor (TCR) on the surface of T lymphocytes provides clonal expansion and response against the antigen. In this study, we evaluated the TCR Vβ repertoire to investigate possible disturbances in the cellular response due to uremia.
Material and methods: This single center cross-sectional study included 35 patients with CKD younger than 20 years (21 CKD stage 3-4 and 14 CKD stage 5-dialysis) and 15 age- and sex-matched healthy controls. The clinical data of the patients were obtained from their medical records. Lymphocyte subgroups and TCR Vβ repertoire were assessed with a flow cytometry device simultaneously with complete blood count.
Results: The median age of the patients 12.3 (8.6-19) years and the median lymphocyte count was 2300 (1800-2900)/μl. There were no significant differences between patients and controls in terms of age, lymphocyte count, or lymphocyte subgroups (CD3, CD4, CD8 T cells, CD4/CD8 ratio and CD16/56 NK). However, the expressions of TCR Vβ 9, TCR Vβ 11, and TCR Vβ 16 were significantly lower and the expression of TCR Vβ 17 was significantly higher in patients than in controls ($p<0.05$ for all). Subgroup analysis showed that there were significant differences in CD4 percentages, CD4/CD8 ratio, Vβ11 and Vβ12 between the CKD and dialysis groups ($p<0.05$ for all).
Conclusions: Despite lymphocyte count and lymphocyte subsets comparable to those of healthy controls, children with CKD have an impaired TCR Vβ repertoire. This deterioration is more pronounced in dialysis patients.

PI-61 EFFECTS OF TRANSITIONING FROM IMMEDIATE RELEASE TO EXTENDED RELEASE CYSTEAMINE THERAPY IN NORWEGIAN PATIENTS WITH NEPHROPATHIC CYSTINOSIS

Sonja Amdal Aase[1], Maria Radtke[2], Christian Siva[3], Bjørn Egil Vikse[4], Damien Brackman[5], Helga Gudmundsdottir[6], Brita Forsberg[7], Anna Bjerre[8]

[1]Stavanger University Hospital, [2]St:olavs University Hospital, [3]Vestfold Hospital, [4]Haugesund Hospital, [5]Haukeland University Hospital, [6]Ullevål University Hospital, [7]Chiesi Global Rare Diseases, Nordics, [8]Oslo University Hospital

Introduction: Nephropathic cystinosis is a rare lysosomal storage disorder where accumulation of cystine, and formation of crystals, progressively damages various organs, including kidneys, retina, muscles and central nervous system.
Cysteamine has been available since 1997 as immediate release (IR) formulation, Cystagon®, and since 2013 as extended release (ER) formulation, Procysbi®. The 10 Norwegian patients were all switched from IR- to ER-cysteamine in 2015-2016.
The aim of this long-term, retrospective study was to evaluate implementation of ER-cysteamine in patients already treated with IR-cysteamine. Primary efficacy endpoints were white blood cell (WBC) cystine levels and estimated glomerular filtration rate (eGFR) change per year.
Material and methods: All 10 Norwegian paediatric- and adult patients participated in the study and data were obtained retrospectively from up to 6 years prior to the switch, until time of inclusion.
Results: Mean level of WBC cystine remained stable between the IR- and ER treatment periods (1.19 versus 1.38 nmol hemicystine/mg protein) despite a dose reduction in most patients on ER-treatment. Mean eGFR change per year was more pronounced during ER-treatment compared to during IR-treament (-4.41 versus -0.17 ml/min/1.73m^2/year). However, single events, such as tubulointerstitial nephritis and kidney transplantation, could have influenced the results. Z-height score developed positively from -1.062 before switch to -0.330 after switch. Four of seven patients with halitosis, reported improvement of symptoms after switch, one reported unchanged symptoms and two reported worsened symptoms. Most adverse drug reactions (ADRs) were of mild to moderate severity during both periods. However, one patient developed two serious ADRs during ER-treatment and was eventually switched back to IR-formulation.
Conclusions: The results from this long-term retrospective study indicates that a switch from IR- to ER-cysteamine was feasible and safe under routine clinical practice. However, due to the low number of patients, and the retrospective collection of data, it is hard to draw any definitive conclusions.

PI-62 KIDNEY AND BONE MARROW INVOLVEMENT IN IPEX SYNDROME WITH ATYPICAL PRESENTATION: THE "FIL ROUGE" OF TREG BETWEEN IPEX FEATURES AND OTHER CLINICAL ENTITIES?

Edoardo La Porta[1], Micaela Gentile[1], Andrea Angeletti[1], Gianmarco Ghiggeri[1], Gianluca Caridi[1], Francesca Lugani[1], Lorenzo Nescis[1], Enrico Fiaccadori[2], Alberto Magnasco[1], Antonella Trivelli[1], Enrico Verrina[1]

[1]Nephrology, Dialysis And Transplantation, Irccs Gaslini Hospital, Genoa, Italy, [2]Dipartimento Di Medicina E Chirurgia, Università Di Parma, Parma, Italy

Introduction: The transcription factor Forkhead box protein P3 (FOXP3) is central to the function of regulatory T cells (Treg). Mutations in the FOXP3 gene lead to a systemic disease called immune dysregulation, polyendocrinopathy and enteropathy, X-linked syndrome (IPEX). Some FOXP3 mutations were associated with atypical presentation including rare disease
Material and methods: We reported two cases of IPEX characterized by kidney and hematologic involvement
Results: *Patient 1.* A 16-year male with a clinical diagnosis of ALPS treated with sirolimus. Due to the onset of proteinuria and decreased

kidney function, a kidney biopsy was performed, with diagnosis of membranous glomerulopathy (MGP). PLA2R on serum and tissues were negative. A mutation of the FOXP3 gene c.779T>A (p.L260Q), never reported before and predicted to be Likely Pathogenic was found. The patient does not present the classical triad of IPEX, and Treg resulted normal. He was treated with steroids and continues sirolimus with good control of proteinuria and stable kidney function.

Patient 2. A 2-year child was diagnosed with a bone marrow failure, genetic investigations were negative. He presented elevated serum IgG4 and kidney failure. A kidney biopsy showed MGP associated with TIN. IF and IHC for PLA2R resulted positive. IHC for IgG4 resulted positive. After the diagnosis of IgG4 RD, steroid therapy was started, without clinical response. Thereafter the patient underwent bone marrow transplant from his brother (HLA-identical). We performed a new genetic exam. A hemizygous mutation of the FOXP3 gene c.1087A>G (p.I363V), already described in the literature, was found, also in the mother and in the proband's brother, thus a diagnosis of IPEX was done. Therefore, Treg resulted normal for brother and mother but not in our patient. The patient underwent kidney transplantation, and after one year he presents normal kidney function.

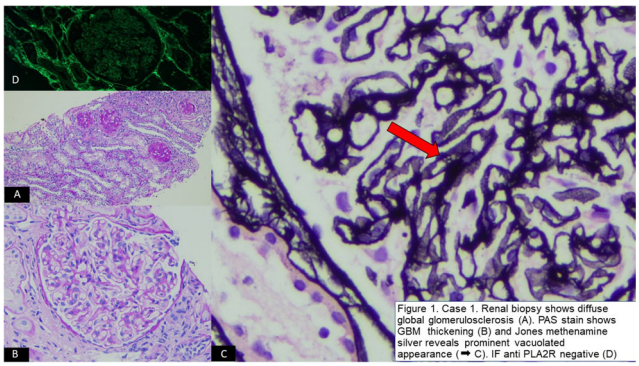


Figure 1. Case 1. Renal biopsy shows diffuse global glomerulosclerosis (A). PAS stain shows GBM thickening (B) and Jones methenamine silver reveals prominent vacuolated appearance (➡ C). IF anti PLA2R negative (D)

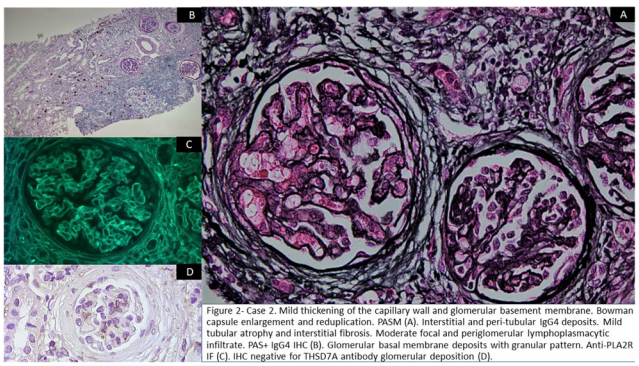


Figure 2- Case 2. Mild thickening of the capillary wall and glomerular basement membrane. Bowman capsule enlargement and reduplication. PASM (A). Interstitial and peri-tubular IgG4 deposits. Mild tubular atrophy and interstitial fibrosis. Moderate focal and periglomerular lymphoplasmacytic infiltrate. PAS+ IgG4 IHC (B). Glomerular basal membrane deposits with granular pattern. Anti-PLA2R IF (C). IHC negative for THSD7A antibody glomerular deposition (D).

Conclusions: MGP pathogenesis in IPEX is consistent with recent evidence of unbalance of Th17/Treg in idiopathic MGP, with significant reduction of Treg cell and FOXP3 expression. IPEX poses a diagnostic challenge considering the spectrum of different phenotypes, but to recognize kidney involvement, together with the growing use of wide genomic analysis, could play a central role.

PI-63 PAEDIATRIC PATIENTS UNDERGOING RENOVASCULAR SURGERY HAVE EXCELLENT CLINICAL OUTCOMES AND IMPROVED QUALITY OF LIFE

Momitha Chaudhury, Nicos Kessaris, Colin Forman, George Hamilton, Meryl Davis, Premal Patel, Kishore Minhas, Nadine Dobby, Kjell Tullus Samiran Ray, Jelena Stojanovic

Great Ormond Street Hospital For Children Nhs Foundation Trust

Introduction: Renovascular hypertension is the 3rd most common cause of hypertension in children accounting for 5-10% of childhood hypertension. Hypertension can initially be medically managed however, endovascular interventions or vascular surgery may be necessary in patients who do not achieve good blood pressure(BP) control on medication only. To our knowledge, this is the first study looking into quality of life(QoL) and clinical outcomes in children who underwent renovascular surgery.
Material and methods: Retrospective study on all 22 patients who underwent a renal auto-transplant and/or a renovascular surgery between 2000 and 2021 at Great Ormond Street Hospital for Children NHS Foundation Trust. Data collection was obtained through electronic patient records. The Wilcoxon signed rank test was used. The median and interquartile ranges were reported when data was not normally distributed. Quality of life was assessed using the validated PedsQL™ Transplant Module questionnaires.
Results: A minority of children (12.3%) with renovascular hypertension required vascular surgery following which, there was a significant improvement in BP control in those who had an auto transplant and an aortic bypass as well as needing to use fewer anti-hypertensive drugs and reduction in number of angioplasties required post-surgery. Patient survival was 100% and kidney function was preserved in all patients.
QoL improved with patients needing to take fewer medications and having fewer concerns regarding medicine side effects and body image. Median score was 70 (100 being highest). Patients reported better QoL than their parents. This study identified the importance of holistic care and need for good communication between healthcare professionals and patients in reducing anxiety associated with treatment as well as an enhanced psychological support particularly for adolescents.
Conclusions: The study reports excellent patient survival, improved BP control, preserved kidney function and improved QoL in patients undergoing renovascular surgery.

PI-64 CLINICAL AND METABOLIC PROFILE OF CHILDREN WITH NEPHROLITHIASIS IN WESTERN MAHARASHTRA

Madhura Fadnis, Jyoti Singhal, Jyoti Sharma

Kem Hospital,pune

Objective: To study the clinical profile and metabolic abnormalities in children with nephrolithiasis.
Material and methods: A chart review of children aged less than 18 years, diagnosed to have nephrolithiasis on ultrasonography.Details recorded were demography, history, serum biochemistry and 24 hour/ spot urine tests for metabolic workup, genetic tests and stone analysis by Fourier transform infrared spectroscopy (FTIRs) when available.
Results: Of 151 records retrieved, 111 children had followed up for evaluation. Boys were predominantly affected (M: F = 2.6:1).The mean age at onset of symptoms was 57±40 months. The most common presenting complaint was gross hematuria in 61(36%); Others were abdominal pain 41(24%),urinary tract infection 26(15%) and urinary retention causing anuria 25(15%).Family history of was present in 60(40%).Anatomical defects were identified in 6 children;pelviureteric junction obstruction in 4(3%), duplex collecting system and posterior urethral valves in 1 boy each. Five children presented with acute kidney injury requiring renal replacement therapy. Obstructive calculi were present in 42(28%); most common site was pelviureteric junction in 21(50%). Metabolic evaluation available for 111 patients revealed hypercalciuria in 14(13%), hyperuricosuria in 9(8%),hyperoxaluria in 5(5%), cystinuria in 3(3%),hypocitraturia in 2(2%) and no cause was found in 76 (68%).Four patients were diagnosed as distal renal tubular acidosis.FTIRs performed in 34 children showed calcium oxalate in11(32%), uric acid in9 (26%).Genetic tests possible in

5 patients showed a mutation in GRHPR gene (primary hyperoxaluria type 2) and SLC3A43 (hypercalciuria) in one each. Three children had hyperoxaluria on 24 hour urine collections but no mutation on NGS. Recurrence of calculi was seen in 19/111patients (12%), 8 of whom had metabolic derangement on presentation. Forty-five (30%) patients required surgical intervention.
Conclusions: Despite thorough evaluation, majority of the children might not have an underlying metabolic abnormality. Anatomical defects should be suspected when evaluating children with calculi.

PI-65 SWITCHING FROM IMMEDIATE TO EXTENDED RELEASE CYSTEAMINE IN PATIENTS WITH NEPHROPATHIC CYSTINOSIS IN SPAIN: FROM CLINICAL TRIALS TO CLINICAL PRACTICE

Gema Ariceta[1], Fernando Santos[2], Andrés Lopez Muñiz[3], Alvaro Hermida[4], Maria Luisa Matoses[5], Ana Ventura[6], Paloma Leticia Martín-moreno[7], Esther González[8], Julia Vara[9]

[1]Pediatric Nephrology. Hospital Universitari Vall D Hebron. Barcelona, [2]Paediatric Nephrology. Hospital Universitario Central De Asturias. University Of Oviedo, [3]Nephrology Department. Complejo Hospitalario Universitario A Coruña, [4]Internal Medicine Service. Hospital Clínico Universitario De Santiago De Compostela, [5]Paediatric Nephrology Department. Hospital Universitario La Fe (valencia), [6]Nephrology Department. Hospital Universitario La Fe (valencia), [7]Department Of Nephrology, Clinica Universidad De Navarra, Navarra Institute For Health Research (idisna) (pamplona), [8]Nephrology Department. Hospital 12 De Octubre (madrid), [9]Paediatric Nephrology Department. Hospital 12 De Octubre (madrid)

Objectives: The objective of RELUCIR study was to evaluate, under clinical practice conditions, the effectiveness and safety of switching from immediate-release (IR) to extended-release (ER) cysteamine in patients with nephropathic cystinosis (NC) in Spain.
Material and methods: Observational, retrospective multicentre study in patients with NC of any age, that had received IR cysteamine for at least 12 months, had switched to ER cysteamine, and had been receiving ER cysteamine for at least 6 months prior to inclusion into the study.
Results: A total of 9 patients, 4 children and 5 adults, 10.1-34.4 years old, were included. All 5 adult patients and one adolescent had received a kidney transplant (3 in one patient). Despite individual variations, no significant differences in grouped WBC (white blood cells) cystine levels were observed after the switch. In patients with preserved kidney function, eGFR remained stable after the switch. There was no significant difference in the cysteamine dose received before and after the switch. However, we observed that some patients were receiving <50% of recommended doses of cysteamine and showed elevated levels of WBC cystine. A significant improvement in height, weight and corresponding Z scores after the switch was observed, particularly in paediatric patients ($p<0.05$). There was a trend towards reduction in the number of hospitalizations and a significant reduction in the hospitalizations stays (days of hospitalization; $p<0,0001$) after the switch to ER-cysteamine, especially in those patients who were admitted due to medication or to the disease itself. A reduction in the appearance of halitosis, body odour and gastrointestinal effects, as well as PPI use were observed after the change to ER-cysteamine in some of the patients.
Conclusions: Switching from IR to ER cysteamine in clinical practice might help to improve tolerability and growth in children with NC and to reduce hospitalization stays.

PI-66 PROXIMAL TUBULOPATHY AND SEVERE MULTI-ORGAN DYSFUNCTION IN AN INFANT WITH NEK-8 MUTATION

Alejandro Zarauza SantoveÑa[1], Laura Garcia Espinosa[1], Gema Muñoz Bartolo[2], César Abelleiras Pardeiro[3], Julían Nevado Blanco[4], Laura Espinosa Román[1]

[1]Pediatric Nephrology. Hospital Universitario La Paz. Madrid., [2]Pediatric Hepatology. Hospital Universitario La Paz. Madrid., [3]Pediatric Cardiology. Hospital Universitario La Paz. Madrid., [4]Ingemm (instituo De Genética Médica Y Molecular), Hulp-idipaz And Ciberer. Madrid Spain

Introduction: NEK-8 gene encodes a ciliary protein involved in organ development. Mutations in NEK-8 have been described as causing nephronophtisis, although renal phenotype can be diverse. Multiorgan involvement has been reported, such as hepatopathy or cardiomyopathy, usually with poor evolution.
Material and methods: We present a case of a child with severe hepatic dysfunction, cardiac involvement and a rare renal presentation including CAKUT and proximal Fanconi tubulopathy.
Results: Male infant born full-term from healthy non-consanguineous parents. Prenatal ultrasound showed severe left hydronephrosis with thickened parenchyma and cysts. Postnatal evaluation confirmed functional annulment of left kidney, with normal right kidney. Cystography showed mild left vesicoureteral reflux. After birth he developed severe and progressive cholestasis with hypoacolia. Etiological study of cholestasis, including exploratory laparoscopy, hepatic biopsy and genetic study, was negative. After finding of hyperchloremic metabolic acidosis, a tubular function study revealed signs of proximal tubulopathy (hyperuricosuria, glycosuria, mild renal phosphate loss, low molecular weight proteinuria, generalized aminoaciduria). Sodium bicarbonate was initiated at 3 months, and common causes of tubulopathy with hepatopathy were ruled out.
Liver disease progressed to cirrhosis and was complicated by an arterioportal shunt leading to congestive heart failure and requiring embolization. Finally, at 10 months of age he received a liver transplant, without complications and good further liver function. After liver transplantation, he had hypertension and progressive declining in glomerular filtration rate. He currently has stage 3 CKD with an estimated GFR of 33 mL/min/1.73m2. Cardiologically, he maintains complex mitral valve disease with good myocardial function.
At 20 months, a NGS renal disease panel confirmed two non-described missense mutations (compound heterozygosis) in NEK-8, pending parental study.
Conclusions: Renal manifestations of NEK8 mutations may include unusual manifestations. Accurate genetic diagnosis provides prognostic information and allows anticipation of potential complications.

PI-67 CLİNİCAL SPECTRUM OF CUBULIN MUTATIONS

Neslihan Cicek[1], Harika Alpay[1], Sercin Guven[1], Ozde Nisa Turkkan[1], Serim Polat[1], Ece Demirci Bodur[1], Ceren Alavanda[2], Nurdan Yildiz[1], Pinar Ata[2], Ibrahim Gokce[1]

[1]Marmara University School Of Medicine, Department Of Pediatric Nephrology, [2]Marmara University School Of Medicine, Department Of Genetics

Introduction: Cubulin is one of the receptor proteins responsible for the reabsorption of albumin in proximal tubule and is encoded by CUBN gene. We aimed to evaluate clinical and genetic characterization of five patients with proteinuria who had mutations in CUBN gene.
Material and methods: Patients' demographic charecteristics, serum creatinine, albumin, vitamin-B12 level, urine analysis, 24-hour urine protein, microalbumin, beta2 microglobulin, estimated glomerular filtration rates(eGFR), treatments, kidney biopsies and genetic analysis were evaluated.

Results: Five patients(1 female, 4 male) were evaluated with a mean admission age of 7.8±2.9 years and a follow up time of 7.7±5.6 years. All patients were referred to Pediatric Nephrology Department with an incidental finding of persistent protenuria. Serum albumine, creatinine, eGFR were in normal ranges, urine analysis revealed no hematuria at admission and at last visit and C3, C4, ANA, anti-DNA were negative for all patients. 24-hour urine protein at admission was 18.1±4 mg/m^2/hour and microalbumin was high in all patients. The maximum proteinuria during follow-up was 27.8±1.5 and 15.9±3.8 mg/m^2/hour at last visit. Serum vitamin B12 was low in two patients and was normal in three patients. Renal ultrasonography was normal in all patients. Renal biopsy was performed in three patients, two demonstrated normal light microscopy, one focal segmental glomerulosclerosis(FSGS) and immunofluorescence examination was negative in three patients. Genetic tests revealed four homozygous and one compound-heterozygous mutation in C-terminal part of CUBN gene. All patients had normal eGFR and still non-nephrotic range proteinuria at last visit.
Conclusions: CUBN gene mutations should be considered in patients with isolated non-nephrotic range proteinuria and normal kidney function. Diagnosing these patients, who are thought to have a better prognosis, is important in terms of avoiding unnecessary treatment and predicting the prognosis. Besides CUBN gene mutations may also be presented as FSGS which extends the spectrum of renal manifestation of these patients.

PI-68 EFFICACY OF ANGIOTENSIN-CONVERTING ENZYME INHIBITORS IN CHILDREN WITH DENT DISEASE

Larisa Prikhodina, Svetlana Papizh, Zilya Bashirova, Varvara Obukhova, Tatyana Lepaeva, Tatyana Nikishina

Research & Clinical Institute For Pediatrics Pirogov Russian National Research Medical University

Introduction: Dent disease (DD) is an X-linked proximal tubulopathy characterized by low molecular weight proteinuria, hypercalciuria and nephrocalcinosis with progression to end-stage kidney disease in males. Data on efficacy of angiotensin-converting enzyme (ACE) inhibitors in children with DD are sparse. The aim of the study was to investigate the efficacy of ACE inhibitors in boys with DD.
Material and methods: We conducted retrospective longitudinal study of 15 boys aged 12.0 (IQR: 9.0; 15.5) years at the last follow up including 12 patients with DD1 and 3 with DD2 types confirmed by direct Sanger sequencing. The median proteinuria before treatment with ACE inhibitors was 0.92 (0.67; 1.42) g/m^2/day. The median eGFR before ACE inhibitors was 82.1 (70.2; 89.5) ml/min/1.73m^2. The median age of starting ACE inhibitors was 7.5 (6.0; 9.5) years. The initial ACE inhibitors dosage was 0.13 (0.11; 0.2) mg/kg/day. The median time of treatment with ACE inhibitors was 35.0 (23.0; 85.0) months.
Results: Treatment with ACE inhibitors led to reduction (>50%) in proteinuria in 9/15 (60%) children with DD including all 3 boys with DD2 type. 2/15 (13.3%) and 4/15 (26.7%) boys with DD treated with ACE inhibitors had the same or increased proteinuria, respectively. Increased eGFR at the last follow-up was found in 8/15 (53.3%) children with DD including all 3 boys with DD2 type. The median rate of increasing eGFR from baseline level on the treatment with ACE inhibitors was 11% (3.4%; 26.4%). 7/15 (46.7%) boys with DD had declined eGFR at the last follow up by 10.6% (-30.1%; -6.2%). All of them had DD 1 type.
Conclusions: We conclude that treatment with ACE inhibitors lead to decreasing of proteinuria in 60% of patients and increasing of eGFR in 53.3% of boys with DD 1 and 2 types.

PI-69 SUCCESSFUL TREATMENT OF IGA NEPHROPATHY (IGAN) WITH TARGETED-RELEASE BUDESONIDE IN A 13-YEAR-OLD BOY

Śladowska-kozłowska Joanna, Galata Barbara, Tönshoff Burkhard

Children's Hospital, University Of Heidelberg, Germany

Introduction: The NEFIGAN and the NefIgArd trials demonstrated that the oral, targeted-release-formulation (TRF) of the glucocorticoid budesonide, delivering the drug to the distal ileum, suppressed the dysfunctional mucosal immune system and, in conjunction with optimal renin-angiotensin-aldosterone system blockade (RAASB), safely reduced proteinuria and improved renal function in IgA-Nephropathy (IgAN) in adults. Also, the safety profile of TRF-budesonide is proposed to be superior to systemic glucocorticoid therapy given its extensive first pass metabolism with <10% entering the systemic circulation. To our knowledge, only one case report detailing the first successful trial of this therapy in a paediatric patient was published. Here, we want to share our experience with TRF-budesonide therapy.
Material and methods: A 13-year-old boy who underwent a routine laboratory blood examination for a scheduled jaw cysts extraction was unexpectedly diagnosed with IgAN. At the time of the diagnosis his serum creatinine was 2.4 mg/dl, and urinary protein-to-creatinine ratio (uPCR) was >700 g/mol. He had hypertension and left ventricular hypertrophy (LVH); the family history was positive for celiac disease of the father. A kidney biopsy demonstrated features of IgA mesangioproliferative glomerulonephritis with focal segmental sclerosis and crescents (14%). Celiac disease was excluded. He received pulse methylprednisolone therapy on 3 consecutive days, followed by oral prednisone over 2 months (for 1 month 75 mg daily, then 50 mg every other day) as well as maximally tolerated RAASB. Because of persisting proteinuria in the nephrotic range therapy with mycophenolate mofetil (MMF) was initiated. After 3 months only small reductions of proteinuria and serum creatinine were achieved. A second course of systemic glucocorticoids was not considered, because he had suffered from the steroid-associated side effect of depressive mood during the first course of steroids. We therefore initiated treatment with 15 mg TRF-budesonide daily continuing MMF and RAASB.
Results: The blood pressure normalised reaching values <50^{th}percentile, the LVH decreased. 3 and 6 months after initiation of TRF-budesonide the patient responded well with a 86% decrease of uPCR down to 100 g/mol and a 34% decrease of serum creatinine down to 1.6 mg/dL. Budesonide was well tolerated, and no side effects were observed.
Conclusions: This case report of successful and well tolerated treatment of IgAN with TRF-budesonide in a paediatric patient underlines the potential of this drug for this disease, for which no approved therapy is currently available. Controlled trails on TRF-budesonide in children with IgAN are required.

PI-70 IGAN IN CHILDREN: A RETROSPECTIVE SINGLE CENTER STUDY

Luca Antonucci, Laura Fuiano, Barbara Ruggiero, Alessandra Gianviti, Marina Vivarelli, Francesca Diomedi-camassei, Francesco Emma

Bambino Gesù Children Hospital

Introduction: IgA nephropathy (IgAN) is the most common glomerulonephritis in childhood. Long-term outcome is generally good, but the risk of progression is about 20-30% at 20 years from onset. Disease course is variable and difficult to predict, ranging from spontaneous remission to rapid progression. Most pediatric studies have focused on identifying risk factors of progression. Conversely, few data exist on IgAN remission. Our study aimed to identify the probability of a complete clinical remission of pediatric IgAN and its predictive factors.
Material and methods: In our monocentric retrospective study, we selected all IgAN histological confirmed cases from 1986 to 2018. The

biopsy were reclassified according to MEST-C score of Oxford classification. Complete clinical remission was defined as absence of proteinuria, hematuria, hypertension, renal failure, in patients off therapy for more than 1 year. The Kaplan-Meier method and Cox proportional hazard model were used for the analysis.

Results: A total of 153 patients were enrolled, with an age at onset of 10.6 ±4 years. At biopsy, prevalence of proliferative lesions (M 41%, E 18%, C 19%) was higher compared to chronic ones (S 32%, T 4%). The estimated probability of complete remission was about 40% at 10 years of follow-up. The probability of recurrence at 6 years from remission was about 20%. Uni-multivariate analyses showed that age at onset and presence of S1 lesions at biopsy were significantly associated with lower chances of complete remission (HR 0.899; p0.041, and HR 0.160; p0.013, respectively). No specific treatment predicted outcome.

Conclusions: Our study has documented that pediatric IgAN has a significant probability of complete remission at 10 years from onset, and a limited probability of recurrence within 6 years from remission. The greatest chances of complete remission were observed in younger children and in the absence of glomerulosclerosis on biopsy.

PI-71 CLINICAL FACTORS AT PRESENTATION INFLUENCING OUTCOME OF BIOPSY PROVEN IGA VASCULITIS NEPHRITIS - A MULTICENTRE STUDY OF 1175 CHILDREN

Katharina Rohner[1], Matko Marlais[2], Yo Han Ahn[3], Alaa Ali[4], Abrar Alsharief[5], Biswanath Basu[6], Anja Blejc[7], Evrim Kargin Cakici[8], Nur Canpolat[9], Eugen Yu-hin Chan[10], Stephane Decramer[11], Filipa Durao[12], Anne M Durkan[13], Ali Duzova[14], Thomas Forbes[15], Junior Gahona Villegas[16], Nilufer Goknar[17], Valentina Gracchi[18], Tulin Gungor[8], Tomoko Horinouchi[19], Belde Kasap Demir[20], Yasuko Kobayashi[21], Mikael Koskela[22], Eda Didem Kurt-sukur[14], Claudio La Scola[23], Dean Langan[24], Xiaozhong Li[25], Gabriele Malgieri[26], Antonio Mastrangelo[27], Jeesu Min[3], Malgorzata Mizerska-wasiak[28], Nabila Mussaoui[11], Aytul Noyan[29], Matti Nuutinen[30], Takayuki Okamoto[32], Louise Oni[33], Michiel Oosterveld[34], Malgorzata Pańczyk-tomaszewska[28], Gonul Parmaksiz[29], Andrea Pasini[23], Pornpimol Rianthavorn[35], Yunyan Shen[25], Rajiv Sinha[36], Rezan Topaloglu[14], Diletta Domenica Torres[37], Tomohiro Udagawa[38], Yok Chin Yap[39], Kjell Tullus[2]

[1]University Childrens Hospital Zurich, [2]Great Ormond Street Hospital London, [3]Department Of Pediatrics, Seoul National University Childrens Hospital & Seoul National University College Of Medicine, Seoul, Republic Of Korea, [4]Great North Childrens Hospital Newcastle, Uk, [5]Queen Silvia Childrens Hospital, GÖteborg, Sweden, [6]Div. Of Pediatric Nephrology, Department Of Pediatrics, Nilratan Sircar Medical College & Hospital, Kolkata, India, [7]Department Of Nephrology, University Childrens Hospital, University Medical Centre Ljubljana, Slovenia, [8]Dr Sami Ulus Maternity And Child Health And Diseases Training And Research Hospital, Department Of Pediatric Nephrology, Ankara, Turkey, [9]Cerrahpasa Medical School, Istanbul University-cerrahpasa, Turkey, [10]Paediatric Nephrology Centre, Hong Kong Childrens Hospital, Hong Kong, [11]Hôpital Des Enfants, Toulouse, France, [12]Pediatric Nephrology And Kidney Transplantation Unit, Department Of Pediatrics, Hospital Santa Maria, Centro Hospitalar Universitário Lisboa Norte, Lisbon, Portugal., [13]Dept Of Nephrology, The Children's Hospital At Westmead, Sydney, Australia, [14]Hacettepe University School Of Medicine, Department Of Pediatrics, Division Of Nephrology, Ankara, Turkey, [15]Department Of Nephrology, Royal Children's Hospital, Victoria, Australia, [16]Baca Ortiz Pediatric Hospital (quito-ecuador), Pontifical Catholic University Of Ecuador, [17]Istanbul Medeniyet University, Turkey, [18]Beatrix Children's Hospital, University Medical Center Groningen, University Of Groningen, Groningen, The Netherlands, [19]Department Of Pediatrics, Kobe University Graduate School Of Medicine, Kobe, Japan, [2] İzmir Katip Çelebi University, Department Of Pediatrics, Division Of Pediatric Nephrology And Rheumatology, Turkey, *[21]Gunma University Graduate School Of Medicine, Department Of Pediatric, Gunma, Japan, [22]Department Of Pediatric Nephrology And Transplantation, New Childrens Hospital, University Of Helsinki And Helsinki University Hospital, Helsinki, Finland., [23]Irccs Azienda Ospedaliero-universiatria Di Bologna, Italy, [24]University College London, London, Uk, [25]The Childrens Hospital Of Soochow, China, [26]Division Of Nephrology,dialysis And Transplant, Aorn Santobono Pausilipon, Napoli, Italy, [27]Nefrologia, Dialisi E Trapianto Pediatrico (ndtp), Fondazione Irccs Ca Granda Ospedale Maggiore Policlinico, Milano, Italy, [28]Department Of Pediatrics And Nephrology, Medical University Of Warsaw, Poland, [29]Baskent University Adana Dr. Turgut Noyan Training And Research Center, Andana, Turkey, [30]Clinic For Children And Adolescents, Oulu University Hospital, Oulu, Finland, [31]Royal Belfast Hospital For Sick Children, Belfast, Ireland, [32]Department Of Pediatrics, Hokkaido University Graduate School Of Medicine, Sapporo, Japan, [33]Department Of Womens And Childrens Health, Alder Hey Childrens Nhs Foundation Trust Hospital And University Of Liverpool, Uk, [34]Emma Children's Hospital, Amsterdam University Medical Center, Amsterdam, Netherlands, [35]Department Of Pediatrics, Chulalongkorn University, Bangkok, Thailand, [36]Division Of Pediatric Nephrology, Institute Of Child Health, Kolkata, India., [37]Pediatric Nephrology And Dialysis Unit, Pediatric Hospital Giovanni Xxiii, Bari, Italy, [38]Tokyo Medical And Dental University, Tokyo, Japan, [39]Department Of Paediatric, Hospital Tunku Azizah, Women And Children Hospital Kuala Lumpur, Malaysia*

Introduction: IgA Vasculitis Nephritis (IgAVN) is in a large majority of cases self-limiting but a small proportion of affected children develop marked proteinuria, worsening kidney function and even kidney failure. The aim of our study was to define the clinical parameters at onset that have an effect on outcome in a large cohort of children with kidney biopsy-proven IgAVN.

Material and methods: Data were collected through a retrospective international survey from December 2020 to August 2021. Anonymised demographic and clinical data (including renal outcome data) were collected for children (0-18 years of age) with typical symptoms of IgA-Vasculitis, biopsy-proven IgAVN and a follow up of at least 12 months.

Results: Data from 1175 patients were collected from 42 international paediatric nephrology centres. Median age at kidney biopsy was 8.3 years, 43 % female, median duration of follow up 3.7 years. At time of biopsy 37 patients (3.5 %) had an estimated glomerular filtration rate (eGFR) below 30 ml/min/1.73m^2 and 2.6 % needed kidney replacement therapy. 92.5% of the patients had proteinuria, 50% within nephrotic range. With increasing age children had lower eGFR but also lower proteinuria at presentation (p=0.000 and p=0.008).

54.1 % had normalised their kidney function, 40.4 % had CKD Stage 1 and 2, 5.5 % CKD 3-5 at last follow up. 76.6 % had normalised urinary protein excretion, 1.5% continued to have nephrotic range proteinuria. Lower eGFR (p=0.000), hypoalbuminemia (p=0.027) and nephrotic syndrome (p=0.002) at onset were predictors of impaired eGFR at last follow up.

Conclusions: This cohort is the largest study to date of children with biopsy proven IgAVN and confirms the overall favourable outcome in the medium term. Nephrotic syndrome and impaired kidney function at onset increased the risk of developing CKD in the medium term.

PI-72 IGA NEPHROPATHY IN CHILDREN WITH MINIMAL PROTEINURIA: TO BIOPSY OR NOT TO BIOPSY?

Cambier Alexandra[1], Jean-phillipe Roy[1], Claire Dossier[2], Natacha Patey[1], Olivia Boyer[3], Marion Rabant[3], Jean Daniel Delbet[4], Anne-laure Lapeyraque[1], Julien Hogan[2]

[1]Sainte Justine Hospital, [2]Robert Debré Hospital, [3]Necker Hospital, [4]Trousseau Hospital

Introduction: Glomerular inflammation severity and tubulointerstitial lesions have been shown to correlate with the amount of proteinuria in childhood IgA nephropathy (cIgAN). However, data are lacking regarding the severity of histopathologic findings in cIgAN with minimal to absent proteinuria since kidney biopsy indications are not well defined in such cases.
Material and methods: Data on 140 consecutive cIgAN patients with a kidney biopsy from 4 different centers in Paris (France) and Montreal (Canada) were reviewed in order to select patients with urine protein/creatinine ratio (UP/Cr) < 0.03 g/mmol and normal renal function (estimated glomerular filtration rate (eGFR) > 90ml/min/1.73m^2) at biopsy and before any treatment.
Results: A total of 28 cIgAN (mean age 11.02 years ± 3.740); median follow-up 3 years [0.84-11.55] were included. History of macroscopic hematuria was present in 92.8%. At first evaluation, mean eGFR was 115.5 ± 18.2 ml/min/1,73m^2, median UP/Cr was 0.02 [0.011-0.03] g/mmol. Microscopic or macroscopic hematuria was present in 35.7 and 64% respectively. Kidney biopsy optic microscopy showed: mesangial (M1), endocapillary (E1) or extracapillary (C1) proliferation in 53.5, 32.1 and 7% of patients, respectively. Chronic lesions were also present: glomerulosclerosis (S1) and tubular atrophy/interstitial fibrosis in 42% and 7%. Podocytopathic features were also present in 21%.
ACE inhibitors or an immunosuppressive therapy were prescribed in respectively 42.8% and 21.5 % of cases and no treatment in 35.7%. At last follow-up, median eGFR was 102 [91.63-124.4] ml/mn/1.73m^2 and median UP/Cr was 0.028 [0.01-0.03] g/mmol. eGFR was < 90 in 6 patients (21.4%) and 2 (7%) had increased proteinuria (UP/Cr >0.03 g/mmol).
Conclusions: cIgAN with minimal to absent proteinuria at biopsy can be associated with worrisome glomerular acute and chronic lesions.

PI-73 EVALUATION OF BK VIRUS FREQUENCY AND ITS RELATIONSHIP WITH CLINICAL PARAMETERS IN CHILDREN RECEIVING IMMUNOSUPPRESSIVE TREATMENT FOR KIDNEY DISEASE EXCLUDING TRANSPLANTATION

Buket Ugurtay[4], Zeynep Nagehan Yuruk Yildirim[1], Sevim Mese[5], Cemile Pehlivanoglu[2], Betul Sozeri[6], Nurver Akinci[3], Mustafa Onel[1], Bagdagul Aksu[1], Ali Agacfidan[5], Alev Yilmaz[1], Ahmet Nayir[1]

[1]Istanbul University, Istanbul Faculty Of Medicine, Division Of Pediatric Nephrology, [2]Istanbul Umraniye Training And Research Hospital, Division Of Pediatric Nephrology, [3]Bezmialem Vakif University Hospital, Division Of Pediatric Nephrology, [4]Istanbul University, Istanbul Faculty Of Medicine, Department Of Pediatrics, [5]Istanbul University, Istanbul Faculty Of Medicine, Department Of Microbiology, [6]Istanbul Umraniye Research And Training Hospital, Division Of Pediatric Rheumatology

Introduction: The full spectrum of BKV-related kidney diseases in immunocompromised patients remains unclear. The aim of our study was to evaluate the frequency of BK viruria and viremia in patients who received immunosuppressive treatment due to kidney diseases other than transplantation and to evaluate its relationship with clinical parameters.
Material and methods: A total of 46 children, who were using immunosuppressive treatment for kidney disease except renal transplantation and 28 healthy children were included in the study. BKV quantitation was performed in urine and serum samples by real time PCR.
Results: Twenty-four (52.2%) patients were receiving only methylprednisolone, rest of the patients were receiving cyclosporine, mycophenolate mofetil, canakinumab, eculizumab, cyclophosphamide (alone or in combination) with or without methylprednisolone at the time of sampling. BKV-DNA was detected in the urine samples of 2 (4.35%) patients while there were no BKV-DNA positivity in plasma samples. One of these patients was being followed up with the diagnosis of systemic lupus erythematosus (SLE) and has been receiving cyclosporine and steroid at the time of sampling. The other with the diagnosis of steroid-dependent nephrotic syndrome and receiving steroid at the time of sampling. Both patients had used cyclophosphamide in the past. Also rituximab was used for the patient with SLE. There were no positivity in the plasma and urine samples of the healthy control group.
Conclusions: The use of steroids alone as immunosuppressive therapy does not appear to be an important risk factor for BKV reactivation. It does not seem necessary to perform BKV-DNA screening in pediatric patients receiving mild immunosuppressive therapy. It has been thought that BKV reactivation may be more frequent, especially if the underlying disease is SLE or if intense immunosuppression is used, and BKV screening may be performed in these patients. Our results suggest that frequency of BK viruria and viremia are very low in healthy children.

PI-74 SARS-COV-2 ASSOCIATED ACUTE INTERSTITIAL NEPHRITIS IN AN ADOLESCENT

Karolis Azukaitis[1], Justinas Besusparis[2], Arvydas Laurinavicius[2], Augustina Jankauskiene[1]

[1]Clinic Of Pediatrics, Institute Of Clinical Medicine, Faculty Of Medicine, Vilnius University, Vilnius, Lithuania, [2]Institute Of Biomedical Sciences, Faculty Of Medicine, Vilnius University, Vilnius, Lithuania

Introduction: Acute interstitial nephritis (AIN) has been recently recognized as one of the infrequent kidney involvement phenotypes among adult patients with severe acute respiratory syndrome coronavirus 2 (SARS-CoV-2) infected patients. Although SARS-CoV-2 associated intrinsic kidney disease has been still scarcely reported in children, at the time of this case report only one case of AIN has been published.
Material and methods: Case report of a 12 year old boy with AIN. Immunohistochemistry (IHC) for Anti-SARS-CoV-2 spike glycoprotein antibody (abcam: ab272504) and electron microscopy (EM) was performed.
Results: The patient presented with fatigue, anorexia and polydipsia following a real-time polymerase chain reaction confirmed SARS-CoV-2 infection 6 weeks ago. Initial work-up revealed estimated glomerular filtration rate (eGFR) of 23 ml/min/1.73 m^2, high erythrocyte-sedimentation rate (120 mm/h), anemia (hemoglobin 9.8 g/dL), thrombocytosis (517 x 10^9) and increased C-reactive protein (49.6 mg/L). Urine tests showed glucosuria, low-molecular weight proteinuria and microhematuria with hyaline and granular casts on microscopy. Antibodies against SARS-CoV-2 S protein receptor binding domain confirmed prior infection with high titres (453 BAU/ml). Kidney biopsy showed diffuse active interstitial nephritis with negative immunofluorescence and positive IHC for SARS-CoV-2 in interstitial inflammatory infiltrate and EM revealed SARS-CoV-2-like viral particles. Kidney function continued to deteriorate despite several days of watch and wait period (eGFR nadir 19.8 ml/min/1.73 m^2) and thus treatment with methylprednisolone pulse-dose therapy (1000 mg thrice) was initiated. This was followed by oral methylprednisolone 48 mg once daily which led to complete normalization of kidney function within 3 weeks.
Conclusions: Our case adds to the emerging evidence of SARS-CoV-2 as a potential etiological agent of AIN in children. In the light of evolving virus spread among children and potential asymptomatic course, epidemiological history, serologic testing and SARS-CoV-2 detection in biopsy should be added to the work-up of children with AIN of unknown etiology.

PI-75 TWO PHASE III TRIALS EVALUATING CROVALIMAB IN PATIENTS WITH ATYPICAL HAEMOLYTIC UREMIC SYNDROME (AHUS): COMMUTE-A AND COMMUTE-P

Neil Sheerin[1], Larry A. Greenbaum[2], Shuichi Ito[3], Chantal Loirat[4], Shoichi Maruyama[5], Ming-hui Zhao[6], Khaled Benkali[7], Christelle Pieterse[8], Mona D. Shah[9], Alexandre Sostelly[8], Sasha Sreckovic[8], Fadi Fakhouri[10]

[1]Translational And Clinical Research Institute, Newcastle University, Newcastle Upon Tyne, United Kingdom, [2]Emory University And Children's Healthcare Of Atlanta, Atlanta, Georgia, USA, [3]Department Of Paediatrics, Graduate School Of Medicine, Yokohama City University, Yokohama, Japan, [4]University Hospital Robert DebrÉ, Paris, France, [5]Nagoya University Graduate School Of Medicine, Nagoya, Japan, [6]Peking University First Hospital, Beijing, China, [7]Certara, Inc., Paris, France, [8]F. Hoffmann-la Roche Ltd, Basel, Switzerland, [9]Genentech, Inc., South San Francisco, California, USA, [10]Vaudois University Hospital Center (chuv), Lausanne, Switzerland

Introduction: aHUS is a life-threatening disease of complement dysregulation, characterised by thrombotic microangiopathy (TMA). While treatment with C5 inhibition is effective, currently approved therapies require regular intravenous infusions. Crovalimab, a novel anti-C5 monoclonal antibody, allows for small-volume, subcutaneous self-injections. Crovalimab is being tested for treatment of aHUS in two global, Phase III single-arm trials: COMMUTE-a and COMMUTE-p.

Material and methods: COMMUTE-a (NCT04861259) will enrol 3 cohorts of patients with aHUS aged ≥ 12 years and weighing ≥ 40 kg (N ≈ 90): **1) Naive (n ≈ 60)**: complement inhibitor-naive patients; **2) Switch (n ≈ 30)**: patients switching from eculizumab/ravulizumab; and **3) C5 SNP (n < 5)**: patients with a known single-nucleotide polymorphism (SNP).

COMMUTE-p (NCT04958265) will enrol 3 cohorts of patients with aHUS aged ≥ 28 days to < 18 years and weighing ≥ 5 kg (N ≈ 35): **1) Naive (n ≈ 20)**: complement inhibitor-naive patients; **2) Switch (n ≈ 10)**: patients switching from eculizumab/ravulizumab; and **3) Pretreated (n < 10)**: patients who received and discontinued prior eculizumab/ravulizumab treatment.

In both COMMUTE-a and COMMUTE-p trials, patients will receive weight-based crovalimab as a weekly loading series (Weeks 1-4; intravenous dose on Day 1, followed by subcutaneous dosing for subsequent loading doses), followed by self-administered, subcutaneous maintenance doses once every 4 weeks (or once every 2 weeks if < 20 kg; Week 5 onward). The primary objective for both studies is to evaluate the efficacy of crovalimab in Naive patients, based on the proportion of patients with complete TMA response any time from baseline to Week 25.

Results: COMMUTE-a and COMMUTE-p are currently enrolling.

Conclusions: Both COMMUTE-a and COMMUTE-p will assess the efficacy and safety of crovalimab in patients with aHUS.

PI-76 ARE THE COMPLEMENT GENE MUTATIONS AFFECTING CLINICAL OUTCOMES IN CHILDREN WITH C3 GLOMERULOPATHY ?

Neslihan Günay[1], Ismail Dursun[2], Ibrahim Gokce[3], Mehtap Akbalık Kara[4], Demet Tekcan[5], Neslihan Çiçek[3], Meral Torun Bayram[6], Mustafa Koyun[7], Nida Dinçel[8], Hasan Dursun[9], Seha Saygılı[10], Zeynep Nagehan Yürük Yıldırım[11], Selçuk Yüksel[12], Osman Dönmez[13], Sibel Yel[2], Beltinge Demircioğlu[4], Özlem Aydoğ[5], Bahriye Atmış[14], Aysun Çaltık Yılmaz[15], Sevcan A Bakkaloğlu[16], Mehmet Baha Aytaç[17], Mehmet taşdemir[18], Belde Kasap[19], Alper Soylu[6], Elif Çomak[7], Aslı Kantar Özşahin[8], Alper Kaçar[9], Nur Canpolat[10], Alev Yılmaz[11], İlknur Girişgen[12], Kadirye Betül Akkoyunlu[13], Harika Alpay[3], Hakan M Poyrazoğlu[2]

[1]Kayseri City Training and Research Hospital, Pediatric Nephrology Clinic, Kayseri/Turkey, [2]Erciyes University Medical Faculty, Department of Pediatric Nephrology, Kayseri /Turkey, [3]Marmara University Medical Faculty, Department of Pediatric Nephrology, İstanbul/Turkey, [4]Gaziantep University Medical Faculty, Department of Pediatric Nephrology, Gaziantep/Turkey, [5]Ondokuz Mayıs University Medical Faculty, Department of Pediatric Nephrology, Samsun/Turkey, [6]Dokuz Eylül University Medical Faculty, Department of Pediatric Nephrology, İzmir/Turkey, [7]Akdeniz University Medical Faculty, Department of Pediatric Nephrology, Antalya/Turkey, [8]Behçet Uz Pediatric Diseases Training And Research Hospital, Pediatric Nephrology Clinic, İzmir/Turkey, [9]Dr. Cemil Taşcıoğlu Hospital Pediatric Nephrology Clinic, İstanbul/Turkey, [10]Cerrahpaşa University Medical Faculty, Department of Pediatric Nephrology, İstanbul/Turkey, [11]İstanbul University Medical Faculty, Department of Pediatric Nephrology, İstanbul/Turkey, [12]Pamukkale University Medical Faculty, Department of Pediatric Nephrology, Denizli/Turkey, [13]Uludağ University Medical Faculty, Department of Pediatric Nephrology, Bursa/Turkey, [14]Gazi University Medical Faculty, Department of Pediatric Nephrology, Ankara/Turkey, [15]Çukurova University Medical Faculty, Department of Pediatric Nephrology, Adana/Turkey, [16]Keçiören Training and Research Hospital, Pediatric Nephrology Clinic, Ankara/Turkey, [17]Kocaeli University Medical Faculty, Department of Pediatric Nephrology, Kocaeli/Turkey, [18]Koç University Medical Faculty, Department of Pediatric Nephrology, İstanbul/Turkey, [19]Tepecik Training and Research Hospital, Pediatric Nephrology Clinic, İzmir/Turkey

Objectives: C3 glomerulopathy (C3G) is a complement-mediated disease. Abnormalities in complement genes are implicated in the development of C3G. The aim of this study is to determine the factors affecting the clinical course of pediatric C3G patients with and without mutations in genes regulating alternative complement pathway.

Methods: Sixty pediatric patients with C3G from 18 referral centers in Turkey were included in study. Patients were classified according to whether they had any complement genetic mutations or not. Demographic data, clinic-pathologic findings, treatment, and outcome data were compared and survey analysis of groups with and without mutations was performed by Kaplan-Meier analysis.

Results: We found mutation in complement system regulating genes in 17 of 60 patients. The most common mutation was in the CFH gene (53.3%). Mean age at diagnosis was 11.2±4.1 years and 56% (n:28) of all patients were male. The age at diagnosis was statistically significantly higher in the group with mutation (12.9±3.6). Median follow-up time was 45 months. The follow-up time of the group with mutation was significantly longer (59/35). While the patients without mutation were most frequently presented with the nephrotic syndrome(%39.5(n:17)), the group with mutations presented with asymptomatic urinary findings(%47.1(n:8)), and the difference was significant(p:0.043). No significant difference was found in terms of serum BUN, creatinine, C3, C4 and urine protein to creatinine at the time of admission. While low albumin(<3gr/dl) was detected in 69.8%(n:30) of patients without mutation, this rate was 29.4%(n:5) in the group with mutations and it was statistically significant(p:0.008). When the histopathological features were compared, no difference was found. During the follow-up, 10 of all patients developed CKD, 4 of which were from the mutation group. In Kaplan-Meier analysis, the mean time to develop CKD was 110.4±9.7 months in the group without mutation, while it was 139.1±30.5 months in the group with mutation. The difference was not found statistically significant. In survival analysis in patients in all the groups and with mutation group, MMF treatment did not change the CKD process.

Conclusions: Complement mutations detected in pediatric C3G patients have no effect on clinical characteristics and survival.

Table: Comparison of clinical and laboratory parameters according to mutation status

Variables	Genetic analysis Mutation Ø (n:43) Gender*	Mutation+ (n:17)	*p*
Girl, n(%)	23(%53.5)	9 (%52.9)	1.000
Boy,n(%)	20(%46.5)	8(%47.1)	
Age at diagnosis, year (mean±SD)**	10.5±4.1	12.9±3.6	**0.039**
Follow-up time, months (median(min-max))***	35(5-140)	59 (9-204)	**0.034**
Laboratory parameters**			
BUN mg/dL	24.4±19.8(n:43)	15.8±8.7 (n:17)	0.094
Cr, mg/dl	0.98±1.36(n:43)	0.62±0.28 (n:16)	0.302
First eGFR	103.4±50.9(n:42)	124.2±43.3(n:17)	0.145
<60 n(%)	8(19)	1(5.9)	
≥60 n(%)	34(81)	16(94.1)	0.382
Last eGFR (mean±SD)	116.6±65.7(n:40)	92.5±45.6(n:16)	0.187
Albumin,gr/dl	2.57±0.95 (n:43)	3.39±0.95(n:30)	**0.004**
<3 n(%)	30(69.8)	5(29.4)	
≥3 n(%)	13(30.2)	12(70.6)	**0.008**
C3, mg/dl	44.6±35.5(n:43)	50.5±45.9(n:17)	0.595
C4, mg/dl	18.2±6.7(n:43)	24.5±14.9(n:17)	0.111
UP/Cr	7.40±6.7(n:31)	2.47±2.7(n:4)	0.154
Clinical presentation n(%)*			
Nephrotic Syndrome	17(39.5)	5(29.4)	
Nephritic Syndeome	19(44.2)	4(23.5)	
Asymptomatic urinary abnormality	7(16.3)	8(47.1)	**0.043**
Dialysis need at the time of admission, n(%)*			
No	39(90.7)	15(88.2)	0.551
Yes	4(9.3)	2(11.8)	
Histopathological features light microscopy,n(%)			
Membranoproliferative GN	28(%65.1) 17(%39.5)	12(%70.6) 8(%47.1)	0.769 0.809
Mesangial proliferative GN	12(%28.6) 7 (%16.3)	3(%17.6) 3(%17.6)	0.516 0.260
Cresentric GN	11 (%25.6)	4(%23.5)	1.000
Arteriolar sclerosis	15(%34.9)	8(%47.1)	0.562
Interstitial fibrosis			
Global sclerosis			
Electron microscopy, n(%)*			
C3	36(%83.7)	13(%76.5)	0.712
DDD	7(%16.3)	5(%23.5)	

* chi-square test **independent sample t test ***Mann-Whitney U test

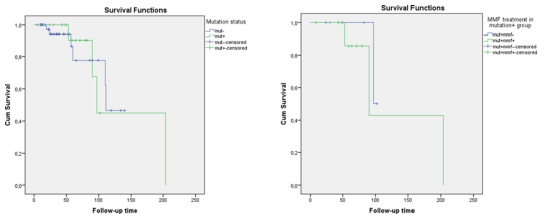


Figure 1(left):Kaplan-Meier analysis for CKD in the group with and without mutation. Mean CKD development time in the group with and without mutation, respectively 139.1±30.5 and 110.4±9.7 months (p:0.98 Figure2(right)Kaplan-Meier analysis for CKD in the group with mutations by MMF use status. Mean CKD development time in the group with MMF use and don't, respectively 133.5±44.9 and 99.5±1.76 months (p:0.572)

PI-77 RENAL INVOLVEMENT IN CHRONIC INFLAMMATORY BOWEL DISEASES: A SERIES OF PEDIATRIC CASES IN FRANCE

Rym Khellaf, Christine Pietrement, Aurelie Pons

Chu De Reims

Introduction: Inflammatory bowel diseases (IBD), including Crohns disease, ulcerative colitis and unclassified colitis, are multisystem diseases. Renal manifestations are not uncommon, and heterogeneous, inaugural or not, caused by IBD itself, their complications, or their treatments. Despite the frequency of IBD in children, there are few data on IBD-related renal damage in this population. The aim of this study is to describe these renal disorders in a series of pediatric IBD cases, in order to highlight the diagnostic and therapeutic difficulties.
Material and methods: This study is a retrospective observational study of a series of pediatric cases collected after a call for observations to pediatric nephrologists of the Society of Pediatric Nephrology and the French Group of Pediatric Gastro-Enterology and Hepatology.
Results: We present the preliminary results of the ongoing study of 6 patients in 5 hospitals in France. 5/6 patients had Crohns disease. 4/6 were treated with PENTASA(1 associateed with Infliximab), 1 with Vedolizumab, and 1 was not treated. Renal involvement manifested as acute kidney injury in all patients and occurred at the same time as digestive manifestations in two cases. Proteinuria and hematuria were not always present. When performed (3 cases), renal biopsy revealed serious tubulointerstitial nephritis. The renal lesions were treated with corticosteroids or simply monitored. 3 cases were attributed to a iatrogenic cause, and IBD treatment was suspended (PENTASA 2/6, VEDOLIZUMAB 1/6). During follow-up, renal function normalized in 2/6 patients, and 4/6 kept chronic kidney disease of varying severity.
Conclusions: The etiological diagnosis of IBD-related kidney damage remains very difficult and often unclear in clinical practice, which makes therapeutic decisions difficult and may delay diagnosis and management, increasing the risk of renal sequels.

PI-78 NIKI-TAG (NEPHROTOXIC INJURY IN KIDS-TAG) A COMPUTER ALERT SYSTEM TO PREVENT AKI (ACUTE KIDNEY INJURY) LINKED TO NEPHROTOXIC DRUG PRESCRIPTION IN HOSPITALISED CHILDREN

This abstract has been withdrawn.

PI-79 IDENTIFYING ACUTE KIDNEY INJURY – A PILOT ELECTRONIC-ALERT SYSTEM TO IDENTIFY AKI IN AT RISK GROUPS IN A TERTIARY PAEDIATRIC HOSPITAL

Colin Higgins, Kathryn Mullan, Emma Ohagan, Mairead Convery, Grace Mccall

Royal Belfast Hospital For Sick Children, Belfast Health And Social Care Trust, Northern Ireland

Introduction: Acute Kidney Injury (AKI) is an often under-recognised problem in the paediatric population. It is associated with increased morbidity and mortality along with an increased incidence of chronic kidney disease. Paediatric patients at high risk of AKI have been identified within the following clinical areas: Paediatric Intensive Care Unit (PICU), Haematology/Oncology and Cardiology. An AKI E-Alert system was subsequently piloted across the three aforementioned three areas to identify incidence of AKI in a tertiary paediatric hospital.
Material and methods: All patients of the three clinical areas between 1st August and 31st October 2020 with an AKI stage 1 alert were included. The serum creatinine level (SCr) at the time of the alert was manually evaluated to assess true incidence of AKI using KDIGO classification. Baseline SCr was identified as the lowest SCr result on the Electronic Care Record system in the preceding 12 months or when this was unavailable, using the upper limit of the reference range for age.
Results: Over a 3 month period there were 159 AKI stage 1 alerts affecting a total of 86 patients. Based on KDIGO classification, we identified 39 patients with AKI stage 1 (45%), 36 patients with AKI stage 2 (42%) and 7 patients with AKI stage 3 (8%). 4 patients were excluded as they were incorrectly highlighted as having had an AKI (5%). Haematology/Oncology patients most frequently developed true AKIs (51%) and had the highest number of AKI stage 2-3s.
Conclusions: The implementation of this pilot AKI E-Alert system provides crucial information regarding the incidence of inpatient AKI in high risk paediatric patient groups. This will ultimately allow for early recognition and timely treatment of AKI, thus reducing the long term burden on paediatric renal health. This pilot also highlights the need for funding of an AKI service throughout the hospital.

PI-80 STUDY FOR THE EVALUATION OF TOXICITY AND RENAL FUNCTION IN PEDIATRIC PATIENTS TREATED WITH BEVACIZUMAB (BERETOX)

Pedro Arango Sancho, Ana Cristina Aguilar Rodríguez, Marta Jiménez Moreno, Elena Codina Sampera, Raquel Jiménez García, Yolanda Calzada Baños, Ofelia Cruz Martínez, Álvaro Madrid Aris

Hospital Sant Joan De Déu

Introduction: Anti-vascular endothelial growth factor (VEGF) antibodies,such as Bevacizumab (BVZ),act on tumor angiogenesis trying to stabilize/remit the tumor mass.This is widely expressed in the glomerulus,playing a key role in its maintenance and proliferation,involving endothelium and podocytes.The objective of our study was to describe the nephrotoxicity associated with treatment with BVZ in pediatrics,as well as the study of kidney damage and function after its withdrawal
Material and methods: Retrospective study including 66 patients affected by solid tumors of the CNS treated between August 2006 and November 2020 with BVZ,without previous nephropathy or history of nephrotoxicity during their treatment.The mean age of the patients was 6.65 years and their total survival from diagnosis of about 5.04 years,with up to 57.6% of the cohort dying during the study. All patients received prior and/or adjuvant therapies including nephrotoxic drugs (cyclosporine, cisplatin or cyclophosphamide) or radiotherapy (50%)
Results: The indication for BVZ in the majority was tumor progression (n=48; 80%),with complete/partial remission in 17%.The mean treatnebt duration was 13.66 months (1-62),18.3 months in nephrotoxicity patients (2.8-34.2).Survival after BVZ was>90 months in 9% of cases,most frequent being 12-36 months (22.5%).Proteinuria and hypertension was observed in 15.10%. When comparing the cumulative dose and duration of treatment with the finding of proteinuria and hypertension,a correlation was observed(100%/50% if >30g and 6.3% /3.2% if >2 years).Long-term analysis of renal function was not possible in the entire cohort due to high mortality (57%),with only 28 patients in long-term follow-up among them,if we exclude the patients who received a nephrotoxic agent after BVZ,only two patients who received BVZ without previous nephrotoxicity (7.14%) presented a glomerular filtration deficit (83 and 88ml/min/1.73 m2).
Conclusions: Long-term renal evaluation in these patients is highly hampered by poor survival,probable previous nephrotoxic damage,and the difficulty of reliably determining renal function in historical cohorts.All patients with proteinuria and hypertension due to BVZ normalized these parameters after its withdrawal.The alterations observed in long-term renal function are minimal and do not allow conclusions to be drawn.Knowledge of the mechanisms of nephrotoxicity,as well as its long-term effects,is essential for the development of new guidelines and preventive strategies that minimize the risk and impact on survival of these patients.

PI-81 RENAL FUNCTION IN SURVIVORS OF HYPOTHERMIA-TREATED HYPOXIC-ISCHAEMIC ENCEPHALOPATHY AFTER 10-12 YEARS

Katarina Robertsson Grossmann[3], Liya Vishnevskaya[2], Mats Blennow[3], Peter Barany[1], Milan Chromek[3]

[1]*Karolinska University Hospital, Department Of Pediatric Nephrology*, [2]*Karolinska University Hospital, Department Of Radiology, Intervention Unit*, [3]*Karolinska Institutet, Department Of Clinical Science, Intervention And Technology, Division Of Pediatrics*

Introduction: Therapeutic hypothermia (TH) is standard of care for infants with moderate-severe hypoxic-ischaemic encephalopathy (HIE) in most high-income countries as it reduces the risk of death or severe neurologic disability. Perinatal asphyxia is a common cause of neonatal acute kidney injury (AKI), which remains a frequent complication also in infants treated with TH. Studies on long term renal outcome after TH are scarce. We recently reported that 21% of survivors in our cohort of children with TH-treated HIE had decreased estimated glomerular filtration rate (eGFR) according to the Schwartz-Lyon equation at age 10-12 years. We now sought to investigate renal functions in greater detail in our cohort.
Material and methods: Our cohort consisted of children born in Stockholm 2007-2009 with a history of TH-treated HIE. At age 10-12, we calculated cystatin C-estimated GFR (cyst C eGFR). Children with decreased cyst C eGFR were examined with iohexol clearance as well. Furthermore, we measured urine-albumin/creatinine ratio in a morning sample, blood pressure (BP) and renal volume on magnetic resonance tomography.
Results: Forty-eight children participated in the assessment. Five per cent (2/42) had decreased cyst C eGFR, and one child (2% 1/42) had decreased GFR according to iohexol clearance. Microalbuminuria was seen in one child (2%, 1/43), and an elevated office BP in three children (7%, 3/45). Subsequent ambulatory 24-hour BP-measurement revealed high normal BP in one case only (2%, 1/45). No child had hypertension. Mean renal volume was 232.2 cc (SD 40.01, 95% CI 217.7-246.6 cc). There was no difference in any of the parameters between children who had suffered neonatal AKI and those who had not.
Conclusions: In our cohort, the Schwartz-Lyon equation appears to overestimate the incidence of decreased GFR. Cyst C eGFR and iohexol clearance confirmed a decreased GFR in only 5% and 2% of children, respectively. Albuminuria and elevated BP were rare.

PI-83 RENAL COMPLICATIONS AT THE HEMATOPOIETIC STEM CELL TRANSPLANTATION EARLY PHASE

SeÇil Kezer, Mehmet Sait DoĞan, Nihan Bayram, Serra Elibol, YÖntem Yaman, Murat Elli, Sema Anak, Cihangir AkgÜn, Önder YavaŞcan

Introduction: Hematopoietic stem cell transplantation (HSCT) is a life-saving therapy for many patients with malign disease, as well as patients with some nonmalignant disorders. Renal complications directly associated with HSCT include a wide range of structural and functional abnormalities. The aim of the study is to investigate the renal complications at the HSCT early phase.
Material and methods: The first 3-month follow-up of 117 patients with HSCT between January 2019-December 2021 was retrospectively evaluated. Renal complications were evaluated as acute kidney injury (AKI) and non-AKI problems (hypertension, proteinuria, increased echogenicity or kidney enlargement on ultrasonography, electrolyte imbalance). Pediatric RIFLE criteria were used for the diagnosis of AKI. The patients were analyzed according to age, gender, donor type, primary diagnosis and complications of HSCT.
Results: Renal complications (60% male) were detected in 83 patients (AKI in 36 patients, Non-AKI in 47 patients). There was no significant relationship between age, donor type, primary diagnosis, and the presence of HSCT complications and of renal complications (Table 1). Continuous dialysis treatment was applied to only 1 patient who died due to multiple organ failure and AKI.

Table 1. Renal complications and patient characteristics

	Renal Complications +			-	*p*
	Non AKI[a]	*AKI*	Total (AKI+Non AKI)		
Patient number n (%)	*47*	*36*	83 (71)	34 (29)	
Hypertension	*25*	*25*			
Proteinuria	*18*	*14*			
Hyperechogenicity on US	*0*	*8*			
Kidney enlargement on US	*24*	*19*			
Male/Female n (%)	*26/21*	*24/12*	50(60.2)/ 33(30.8)	23(67.6)/ 11(32.4)	*0.4*
Age (year) n (%)					
0-5	*26*	*18*	44 (72.2)	17 (27.8)	*0.2*
5-10	*12*	*4*	16 (72.8)	6 (27.2)	
>10	*9*	*14*	23 (67.7)	11 (32.3)	
Donor type n (%)					
Autologous	*15*	*7*	22 (61.2)	14 (38.8)	*0.2*
Allogeneic	*26*	*21*	47 (77.1)	14 (22.9)	
Haplo-identical	*6*	*8*	14 (70)	6 (30)	
Malign/Nonmalign n (%)	*21/26*	*15/21*	36(43.3)/ 47(54.7)	11(32.3)/ 23(77.7)	*0.5*
Complication n (%)					
No	*26*	*16*	42 (72.7)	21 (33.3)	*0.3*
GVHD	*15*	*9*	24 (77.5)	7 (22.5)	
VOD	*4*	*5*	9 (64.3)	5 (35.7)	
GVHD + VOD	*2*	*5*	7 (87.5)	1 (12.5)	

US: Ultrasonography, GVHD: Graft versus host disease, VOD: Veno-occlusive disease
[a]*: More than one non-AKI problem was detected in the patients.*

Conclusions: Acute renal complications are common complication of HSCT. Multiple factors may contribute to renal complications. It is difficult to determine the cause of complications in each patient. Early diagnosis, treatment and identification of nephrotoxic agents are the most important elements of the management of the disease.
HSCT associated kıdney injury was important due to long-term complications (CKD, hypertension, proteinuria). CKD patients require adequate diagnosis and specific follow up. Liver and kıdney enlargement and hyphosfatemıa are early prognostıc factors for AKI.

PI-84 ACUTE KIDNEY INJURY IN EXTREMELY AND VERY LOW BIRTH WEIGHT INFANTS

Sandra Habbig[1], Kathrin Burgmaier[1], Melanie Zeiher[1], Anna Weber[1], Katrin Mehler[2], Angela Kribs[2]

[1]*University Of Cologne, Faculty Of Medicine And University Hospital Cologne, Department Of Pediatrics, Pediatric Nephrology, Cologne, Germany*, [2]*University Of Cologne, Faculty Of Medicine And University Hospital Cologne, Department Of Pediatrics, Neonatology, Cologne, Germany*

Introduction: Acute kidney injury (AKI) in preterm infants has gained enormous attention in recent years as multi-center studies revealed a prevalence of AKI of up to 50 % in these infants and identified AKI as an independent risk factor for mortality.
Material and methods: We here present a retrospective analysis of extremely-low-birth-weight (ELBW, birth weight < 1000 g) and very-low-birth-weight (VLBW, birthweight 1000-1499 g) infants born in our

tertiary center in 2020. We applied the most recent KDIGO definition for neonatal AKI to identify AKI in this cohort.
Results: This study includes 128 preterm infants (68 ELBW and 60 VLBW) with a median birthweight of 962 (range 210-1490) g. Of the entire cohort, 25 patients (20%) suffered from at least one episode of neonatal AKI, in eight patients (6% of the whole cohort) AKI was classified as severe (stage 2 or 3). The incidence of AKI differed significantly between ELBW and VLBW infants (28 versus 10%, p=0.011). Both, length of intensive care as well as length of hospitalisation, was significantly longer in the infants with AKI as compared to those without AKI. Infants with AKI had significantly lower Apgar Scores at 10 minutes compared to infants without AKI. Mechanical ventilation and therapy with resuscitation drugs was performed more frequently in the cohort with AKI (68% vs. 21%; p<0.001; 40% vs. 11%; p< 0.001). In addition, more infants with AKI received indomethacin due to persisting ductus arteriosus Botalli.
Conclusions: As already shown in other studies, neonatal AKI occurred more frequently in ELBW as compared to VLBW infants. Infants with AKI had a lower gestational age and birth weight, lower APGAR values and needed a less intensive therapy including nephrotoxic medication. Interestingly, the overall incidence of AKI was substantially lower in our cohort as compared to recent studies which might be related to the very restrictive use of intubation and mechanical ventilation in our center.

PI-85 FIRST EXPERIENCE OF CONTINUOUS FLOW PERITONEAL DIALYSIS IN NEONATES WITH HYPERAMMONEMIA DUE TO INBORN ERRORS OF METABOLISM.

Hai Liang Tan[1], Ming Jie Chuah[1], Jia Yi Tham[1], Elaine Ee Lane Wong[1], Chee Lee Chan[1], Lilian Ping Ling Ngo[2], Azie Jumaatul Adawiyah Nabir[2], Zuraidah Hj Abd Latif[2], Yok Chin Yap[1]

[1]Department Of Paediatric, Hospital Tunku Azizah, Women And Children Hospital Kuala Lumpur, Malaysia, *[2]Department Of Paediatric, Hospital Ampang, Malaysia*

Introduction: The prognosis of neurological outcome in hyperammonemia caused by inborn errors of metabolism (IEM) depends on plasma ammonia levels and duration of hyperammonaemic coma. Current guidelines suggest the use of continuous renal replacement therapy (CRRT) in the treatment of refractory hyperammonemia. Due to limited vascular access available in a newborn, conventional peritoneal dialysis (PD) has been studied but showed limited efficacy in reducing ammonia levels. This study aims to evaluate if continuous flow peritoneal dialysis (CFPD) is an effective treatment in the management of hyperammonemia due to IEM in newborns.
Material and methods: Three neonates were managed with CFPD in addition to standard medical therapy. CFPD was performed with two bedside-placed catheters sized 10F for inflow and 12F for outflow. The peritoneum was initially filled with 20ml/kg of dialysate during catheter insertion followed by continuous dialysate inflow with the use of an infusion pump at initial rate of 10ml/kg/hour then titrated up as tolerated. The dialysate outflow was gravity dependent with the drainage bag placed 80cm below the outflow catheter. Dialysate was bicarbonate buffered with 2% dextrose concentration. Infusion of dialysate was stopped every 4th hour to allow passive drainage.
Results: The mean age of the patients were 9.3 ± 6.7 days old. The mean plasma ammonia level before dialysis was 1256 ± 466 μmol/L. The mean inflow rate of dialysate was 54 ± 19.9 ml/kg/hour. The mean outflow rate was 51.5 ± 22.9 ml/kg/hour. Plasma ammonia levels below the critical level of 200 μmol/L were achieved within 18 ± 2.6 hours. There were no peritonitis episodes or increased ventilatory support requirements. Minor complications were hypokalaemia that was corrected intravenously and one episode of blocked catheter, solved by switching the inflow and outflow catheter. All patients survived and were discharged home.
Conclusions: This first report of using CFPD for the management of hyperammonemia due to IEM shows that it is effective in removing ammonia rapidly and safely.

PI-86 NOVEL CELLULAR MODEL OF INHIBITION OF GLYCOLATE OXIDASE DEPENDENT OXALATE PRODUCTION BY BBP-711

Thulashitha Rajasingham[1], Andy Whitney[2], Justin Lafountaine[1], Uma Sinha[1]

[1]Cantero Therapeutics, A Bridgebio Company, *[2]Applied Molecular Transport*

Introduction: Primary hyperoxaluria type 1 (PH1) is an autosomal recessive disorder characterized by overproduction of oxalate. Glycolate oxidase (GO) is a logical target for inhibition as substrate reduction therapy for PH1. BBP-711 is a small molecule inhibitor of GO in development for the treatment of PH1. The purpose of this study was to evaluate the in vitro activity of BBP-711 to inhibit GO-dependent production of oxalate in liver cells.
Material and methods: Western blot analysis was used to confirm expression of GO in HepaRG cells and immunohistochemistry was used to confirm localization of GO in peroxisomes. Inhibitory potency of BBP-711 was measured in cultured confluent HepaRG cells over a 7-day period while 5 mM of glycolate was used to enhance oxalate production. Given the structural similarity of the substrate binding sites of GO and lactate dehydrogenase A (LDHA), BBP-711 activity was evaluated with purified LDHA to exclude off-target effects.
Results: GO expression in HepaRG cells was confirmed by western blot analysis. Colocalization of GO and the peroxisome marker catalase was confirmed by immunohistochemistry of the cells. BBP-711 demonstrated dose-dependent inhibition of oxalate production by HepaRG cells over the duration of the experiment (mean IC_{50} value ± SD of 414 ± 200 nM, 521 ± 47, 481 ± 69 nM at 24 hours, 48 hours and 72 hours, respectively). In vitro assays demonstrated that relative to its inhibitory potency against GO (IC_{50}= 15.4 nM), BBP-711 demonstrated only marginal inhibition (>10%) of LDHA at a much higher concentration (10 μM).
Conclusions: This novel cellular assay demonstrates the localization of GO in the peroxisome and the ability of BBP-711 to inhibit peroxisomal GO-mediated oxalate production at therapeutic target concentrations.

PI-87 CORRELATION BETWEEN THE METABOLIC PROFILE OF UROLITHIASIS IN CHILDREN WITH IDIOPATHIC HYPERCALCIURIA AND THE COMPOSITION OF THE DEPOSIT ASSESSED BY INFRARED SPECTROSCOPY

Malgorzata Placzynska, Joanna Milart, Katarzyna Jobs, Arkadiusz Lubas

Military Institute Of Medicine

Introduction: Urolithiasis is an increasingly common condition. Each patient after stone passage should have stone analysis performed. Every child with a urinary stone should be given a complete metabolic evaluation and the stone analysis is its essential component.
Material and methods: The aim of the study was to establish the relationship between the metabolic profile of urolithiasis in children and the composition of the excreted stone.The study involved 26 children with urolithiasis associated with idiopathic hypercalciuria (aged 1-17 years) from whom stones were obtained for the analysis. The urine pH and the

24-hour urine excretion of phosphorus and oxalate as well as spot urinalysis including ratio of crystaloids to creatinine from the second voided urine sample of the day were assessed. Urinary stones were analyzed by infrared spectroscopy. The relationship between the metabolic data and the stone type was then analyzed, taking into account two types of minerals: stones with a predominance of calcium oxalate dihydrate (weddellite) and calcium oxalate monohydrate (whewellite).

Results: The ROC analyzes showed that lower values of each of 4 variables: phosphate and oxalate excretion, magnesium-creatinine ratio, urine pH are characteristic for patients with stones containing > 60% of weddellite. The most useful in identifying these stones were the product of the excreted phosphate and oxalate, and then the excreted phosphate and oxalate, and the urine pH. In the regression analysis finding a reduced value of each of these 4 variables increased more than sixfold the chance of diagnosing urolithiasis with stone composed of over 60% of weddellite (p=0,019).

Conclusions: The reduced excretion of phosphates and oxalates in the 24-hour urine sample and the reduced magnesium-creatinine ratio, as well as the reduced urine pH in patients with idiopathic hypercalciuria, more than six times increases the probability of diagnosis of calcium oxalate stones with a predominance of > 60% of weddellite.

PI-88 RESPONSE TO LUMASIRAN IN PRIMARY HYPEROXALURIA TYPE 1 (PH1): FIRST CLINICAL EXPERIENCES

Sina Saffe[1], Markus J Kemper[1], Matthias Hansen[2], Nele Kanzelmeyer[3], Anja BÜscher[4], Jun Oh[5], Katja Doerry[5]

[1]Department Of Paediatrics, Asklepios Klinik Nord, Hamburg, Germany, [2]Paediatric Nephrology, Clementine Kinderhospital, Frankfurt, Germany, [3]Paediatric Nephrology, Medizinische Hochschule Hannover, Germany, [4]Paediatric Nephrology, University Hospital Essen, Germany, [5]Paediatric Nephrology, University Hospital Hamburg, Germany

Introduction: Primary hyperoxaluria type 1 (PH1) is characterised by hepatic overproduction of oxalate, leading to hyperoxaluria which can result in nephrocalcinosis, urolithiasis and renal failure. Conservative management of PH1 aims to reduce the urinary oxalate concentration (high fluid intake, citrate, pyridoxine). The only curative treatment to date is a liver transplant. Lumasiran is a novel drug that has been approved for use in PH1 patients in November 2020. It reduces the oxalate synthesis in the liver by RNA interference. Data on its effect in routine clinical setting are scarce.

Material and methods: We present data on 6 patients (3 girls) with genetically confirmed PH1, including 2 on dialysis.

Results: Median age at start of treatment was 10.8 (1.8-15.8) years. In those with preserved renal function median reduction of hyperoxaluria was 66% (range 50.4-82%) after 2 months and 58% (4.4-71%) after 6 months. One of these patients had rapid normalization initially but urinary oxalate levels started rising again after the third dose and reached values similar to before the treatment was started. In 2 patients urinary oxalate decreased to healthy control values. In the 2 patients on dialysis, frequency of sessions could be decreased in one patient because plasma oxalate levels decreased but had to be increased in the other due to increasing systemic oxalosis. Treatment was well tolerated, the only noticeable side effects were injection site reactions.

Conclusions: Even though there is promising data regarding the benefits of Lumasiran in PH1, this could not be replicated in all of our patients. Our data highlights the need to regularly re-evaluate hyperoxaluria during Lumasiran treatment also in the long-term; registry data seem mandatory for all PH1 patients treated with Lumasiran to monitor effectiveness and side effects.

PI-89 GENOTYPE-PHENOTYPE CORRELATION IN A COHORT OF INDIVIDUALS WITH DISEASE-CAUSING VARIANTS IN COL4A3/COL4A4 ASSOCIATED WITH TYPE-IV-COLLAGEN-RELATED NEPHROPATHY (ALPORT SYNDROME AND THIN BASEMENT MEMBRANE NEPHROPATHY)

Simmendinger Hannes[1], Riedhammer Korbinian Maria[1], Tasic Velibor[3], Putnik Jovana[4], Abazi-emini Nora[3], Stajic Natasa[4], Weidenbusch Marc[5], Patzer Ludwig[6], Lungu Adrian[7], Milosevski-lomic Gordana[4], Braunisch Matthias[2], GÜnthner Roman[2], Comic Jasmina[1], Hoefele Julia[1]

[1]Institute Of Human Genetics, Klinikum Rechts Der Isar, Technical University Of Munich, School Of Medicine, Munich, Germany, [2]Department Of Nephrology, Klinikum Rechts Der Isar, Technical University Of Munich, School Of Medicine, Munich, Germany, [3]University Children's Hospital, Medical Faculty Of Skopje, Macedonia, [4]Institute For Mother And Child Health Care Of Serbia "dr Vukan Čupić", Department Of Nephrology, University Of Belgrade, Faculty Of Medicine, Belgrade, Serbia, [5]Nephrologisches Zentrum, Medizinische Klinik Und Poliklinik Iv, Klinikum Der Universität MÜnchen, Ludwig-maximilians University, Munich, Germany, [6]Childrens Hospital St. Elisabeth And St. Barbara, Halle/saale, Germany, [7]Fundeni Clinical Institute, Pediatric Nephrology Department, Bucharest, Romania

Introduction: Type-IV-collagen-related nephropathies comprising Alport syndrome (AS) and thin basement membrane nephropathy (TBMN) show a broad phenotypic spectrum ranging from isolated microscopic hematuria (MH) to end-stage kidney disease (ESKD). Monoallelic disease-causing variants in COL4A3/*COL4A4* are associated with TBMN (also described as autosomal dominant AS) and biallelic variants with autosomal recessive AS (ARAS). The aim of this retrospective cross-sectional study was to analyze clinical and genetic data regarding a possible genotype-phenotype correlation in individuals with disease-causing variants in COL4A3/*COL4A4*.

Material and methods: 89 individuals carrying at least one COL4A3/*COL4A4* variant classified as (likely) pathogenic according to the American College of Medical Genetics guidelines and current amendments were recruited. Clinical data concerning the prevalence and age of first manifestation of MH, proteinuria, ESKD, eye anomalies and hearing impairment were raised.

Results: Individuals with monoallelic non-truncating COL4A3/*COL4A4* variants reported a significantly higher prevalence of MH (46/48 (96%) vs. 14/24 (63%), $p = 0.001$) and proteinuria (21/48 (44%) vs. 4/24 (17%), $p = 0.035$) than individuals with truncating COL4A3/*COL4A4* variants. Individuals with biallelic truncating COL4A3/*COL4A4* variants had a significantly lower median age at first manifestation of MH (3 years vs. 6 years, $p = 0.030$) and proteinuria (3 years vs. 9 years, $p = 0.017$) than individuals with biallelic non-truncating COL4A3/*COL4A4* variants. In addition, individuals with biallelic truncating variants in COL4A3/*COL4A4* were significantly younger at first manifestation of proteinuria than individuals with biallelic non-truncating/truncating COL4A3/*COL4A4* variants (median: 3 years vs. 9 years, $p = 0.015$).

Conclusions: In this study, for the first time, an association of heterozygous non-truncating COL4A3/*COL4A4* variants in TBMN/ADAS individuals with a more severe phenotype could be shown indicating a potential dominant-negative effect as an explanation for this observation. The results for individuals with ARAS support the literature data that biallelic truncating variants in COL4A3/*COL4A4* lead to more severe clinical manifestations of AS.

PI-90 A RARE CAUSE OF COMBINED HEPATIC AND RENAL FAILURE: NPHP19 DUE TO A NOVEL DCDC2 VARIANT IN TWO SIBLINGS

Gizem Yildiz[1], Meral Torun Bayram[1], Ahmet Okay ÇaĞlayan[2], Ayfer Ülgenalp[2], Alper Soylu[1], Salih Kavukcu[1]

[1]Dokuz EylÜl University Medical Faculty, Department Of Pediatric Nephrology, Izmir, Turkey, [2]Dokuz EylÜl University Medical Faculty, Department Of Pediatric Genetics, Izmir, Turkey

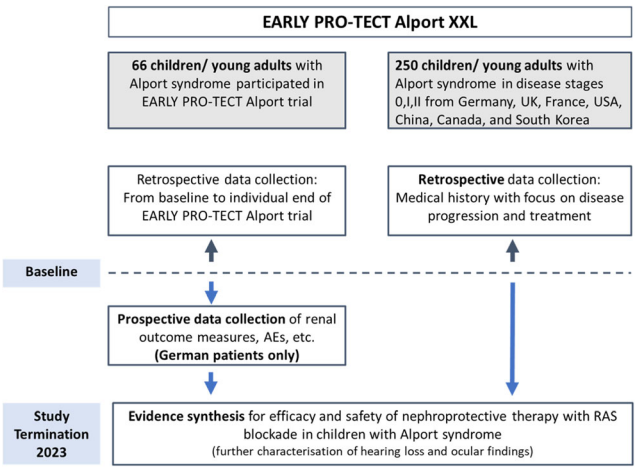
Introduction: Nephronophitisis-19 (NPHP19) is a rare autosomal recessive renal-hepatic ciliopathy due to variants in DCDC2 gene. This syndrome has only been reported in three patients previously. Hepatic involvement precedes renal involvement and is characterized by early onset hepatosplenomegaly, ductal plate malformation, hepatic fibrosis and cholestasis. Renal findings which may not be present at diagnosis include increased echogenicity, severe interstitial fibrosis, tubular dilatation with prominent epithelial luminal budding. We describe here two Turkish siblings with NPHP19 having previously undefined cardiac involvement.
Material and methods: Case Report
Results: Two siblings presented due to chronic renal disease (CKD) and proteinuria at the ages of 7.5 (female) and 10 (male) years. Their parents were first degree relatives. Both had a history of liver transplantation at the ages of 2.5 and 1.5 years, respectively. They had neonatal cholestasis and liver pathology was characterized by biliary cirrhosis and hepatic fibrosis. Both had small echogenic kidneys and nephrotic range proteinuria. CKD stage was 3 and 2 in the girl and the boy, respectively. Kidney biopsy in the boy disclosed focal segmental glomerulosclerosis. Valvular aortic stenosis was detected in both siblings. End stage renal disease developed in the girl at 9 years of age and she died at 10-year of age due to sudden cardiac arrest. The boy is 13-year-old at present and still had stage 2 CKD and nephrotic range proteinuria. Genetic analysis for nephronophitisis panel revealed a novel likely pathogenic homozygous frameshift mutation in exon 7 of DCDC2 gene (chr6:24278358, c.840dup, p.Glu281ArgfdTer27) in both siblings.
Conclusions: Renal involvement has been reported in two of three NPHP19 patients in the literature. We described two siblings with novel homozygous DCDC2 mutation who developed renal involvement during the first decade. ESRD developed at 9 years of age in one patient. In addition, both had cardiac involvement characterized by valvular aortic stenosis which has net been described previously.

PI-91 EARLY PRO-TECT ALPORT XXL: A WORLDWIDE OBSERVATIONAL STUDY TO IMPROVE EVIDENCE FOR PREEMPTIVE START OF ACE INHIBITOR THERAPY IN CHILDREN WITH ALPORT SYNDROME

Oliver Gross, Jan Boeckhaus

University Medicine Goettingen, Nephrology And Rheumatology

Introduction: Children with the type IV collagen disease Alport syndrome (AS) develop renal failure early in life. ACE-inhibition is the first-line off-label therapy and can delay renal failure in a time dependent manner. The EARLY PRO-TECT Alport trial was the first randomized and placebo-controlled trial to evaluate safety and efficacy of ACE-inhibition in children with AS. The trial indicated safety and efficacy of nephroprotective therapy without statistical significance.The aim of this observational study is to analyse renal outcome and adverse effects in the longterm observation of children from the EARLY PRO-TECT Alport trial as well as other children in disease stages 0,I,II from all over the world.
Material and methods: Supported by the EARLY PRO-TECT Alport XXL team international and the study group of the German Society of Pediatric Nephrology, we developed a predefined study protocol:
Alport Syndrome disease stages are defined as:
0 Microhaematuria without microalbuminuria
I Microalbuminuria (30-300 mg albumin/gCrea)
II Proteinuria >300 mg albumin/gCrea
Primary Endpoint: time to progress of AS to the next disease stage under treatment
Secondary Endpoint: Time to progress of AS to the next disease stage under treatment compared to children without treatment.
Results: In addition to the 14 German trial sites of EARLY PRO-TECT Alport, we contacted our international cooperation partners with leading pediatric nephrology centers in the US, UK, South Korea, and China, using University based individual cooperation contracts. We cordially invite all pediatric nephrologists to contribute to this international effort. At present, we have data of long-term outcome of more than 150 children in our system. Data collection will end in 2023.



Conclusions: PLEASE JOIN OUR INTERNATIONAL EFFORT! Using a Bayesian evidence synthesis approach, different levels of evidence will contribute to enrich the EARLY PRO-TECT Alport data in order to reach the significant levels for efficacy of pre-emptive start of ACE-inhibitor therapy in children with AS. Our international approach also aims to set up a political statement for the worldwide need for early diagnosis of children with AS.

PI-92 AUTOSOMAL DOMINANT HYPOCALCEMIA. WHEN BARTTER SYNDROME "PUSHES BOUNDARIES"

Marta Jiménez Moreno, Pedro Arango Sancho, Ana Cristina Aguilar Rodríguez, Raquel Jiménez García, Elena Codina Sampera, Yolanda Calzada Baños, Álvaro Madrid Aris

Hospital Sant Joan De Déu

Introduction: Bartter syndrome (BS) is a heterogeneous disorder related to multiple genetic mutations in transporters located in the thick loop of Henle.In 2002 it was described that autosomal dominant hypocalcemia (ADH) can be associated with BS.The calcium-sensitive receptor (CaSR) is a G protein-coupled receptor that is responsible for regulating calcium hemostasis,controlling the secretion of parathormone (PTH) and renal calcium excretion.Activating mutations of CaSR lead to inhibition of the ROMK channel and inhibit PTH production because of hypocalcemia.The clinical presentations and time of onset of the Bartter phenotype differ according to the type of mutation
Material and methods: A 6-year-old patient who consulted for colic abdominal pain and 24-hour paresthesia in hands and feet.Born at term, blood tests with ions and normal PTH at birth.Under follow-up for non-compaction cardiomyopathy.Asymptomatic to date.Mother,aunt (twins) and maternal cousin in follow-up with endocrinology and

cardiology with current diagnoses of pseudohypoparathyroidism and non-compaction cardiomyopathy
Results: The initial study revealed hypokalemic metabolic alkalosis (potassium 2.5 mmol/l),hypomagnesemia and hypocalcemia,abnormally normal PTH for serum calcium.In the urine,hypercalciuria (calcium/creatinine ratio 1.02 mg/mg) is noted,with elevated fractional excretions of sodium,chloride,potassium and transtubular potassium gradient (TTKG).Renal ultrasound showing severe bilateral nephrocalcinosis.Given the suspicion of Bartter syndrome associated with an alteration in phosphocalcic metabolism, a genetic study was carried out detecting an activating mutation of the CaSR both in the patient and her relatives,being diagnosed with autosomal dominant hypocalcemia associated with Bartter syndrome
Conclusions: When faced with patients with characteristics of renal salt-wasting syndrome together with alterations in phosphocalcic metabolism (hypocalcaemia and hypomagnesaemia),we must consider the differential diagnosis with this rare entity.At the present time,genetic results are pending for the patients heart disease,since no relationship between the two pathologies has been described to date.The genetic study avoids erroneous diagnoses allowing an adequate optimization of the treatment from the early stages of the disease,as well as the planning of a genetic advice before possible future pregnancies

PI-93 EPIDEMIOLOGY OF INS DURING COVID PANDEMIC COMPARED TO THE LAST DECADE: A REGIONWIDE RETROSPECTIVE STUDY IN THE PARIS AREA

Victoire Thenot[1], Cyrielle Parmentier[2], Olivia Boyer[3], Fouad Madhi[4], Sylvie Nathanson[5], Ferielle Zenkhri[6], Philippe Blanc[1], Nasser Mendli[8], Aurelien Galerne[9], Alexis Mandelcwajg[10], Julien Hogan[1], Claire Dossier[1]

[1]Pediatric Nephrology, Robert-debre Hospital, Paris, Aphp, *[2]Pediatric Nephrology, Armand-trousseau Hospital, Paris, Aphp*, *[3]Pediatric Nephrology, Necker Hospital, Paris, Aphp*, *[4]Department Of Pediatrics, Chi Creteil*, *[5]Department Of Pediatrics, Ch De Versailles, Le Chesnay*, *[6]Department Of Pediatrics, Bicetre Hospital, Kremlin-bicetre, Aphp*, *[7]Department Of Pediatrics, Ch Poissy*, *[8]Department Of Pediatrics, Ch Gonesse*, *[9]Department Of Pediatrics, Jean-verdier Hospital, Bondy, Aphp*, *[10]Department Of Pediatrics, Ch De Saint-denis*

Introduction: The etiology of idiopathic nephrotic syndrome (INS) remains partially unknown. Viral infections have been reported associated to INS onset. Thus, both SARS-CoV-2 infection and lockdown measures may have influenced INS incidence. The aim of this study was to describe the incidence of childhood INS during the COVID19 pandemic and compare it to the pre-COVID period.
Material and methods: A written questionnaire was sent to the 37 pediatric departments in Paris area. Children aged 1 to 15 years, living in the Paris area, with INS onset between January 2017 and December 2020 were included. To estimate incidence, population-based denominators were obtained from the National Institute for Statistics and Economic Studies (INSEE). Data were compared to the NEPHROVIR cohort over the 2008-2013 period.
Results: Between 2007 and 2020, 248 cases of INS were reported. Median age was 5.6 years, M/F ratio 2.1/1 and 94 % were steroid sensitive. Annual incidences were 2.66, 2.49, 2.91 and 2.40 per 100 000 children aged $\leq$ 15 years for 2017, 2018, 2019, 2020 respectively, with no significant difference between years and also no difference with the 2008-2013 period. During the lockdown measures (2months), incidence was 0.76 versus 2.71 for the rest of the study period (46 months) ($p < 0.05$). During week 15 (6-12 April 2020), with the peak of hospital admissions for COVID-19, no case of INS was reported.
Conclusions: Over the last decade, incidence of childhood INS was stable in the Paris area. In 2020, no peak of incidence occurred concomitantly to COVID pandemic. In addition, during lockdown measures, incidence of INS was significantly lower. Interestingly, incidence of other respiratory viral infections were reported reduced during lockdown measures. Together, these results argue again for a link between INS onset and viral infections, while COVID-19 does not appear to be a significative trigger for INS onset in children.

PI-94 INTERACTION BETWEEN RENIN AND KALLIKREIN IS IMPORTANT DURING HUMAN KIDNEY DEVELOPMENT AND IN THE CONTROL OF BLOOD PRESSURE IN HEALTHY AND CNF KIDNEYS

Ivona Kosovic[1], Natalija Filipovic[1], Marijan Saraga[2], Merica Glavina Durdov[3], Katarina Vukojevic[1], Mirna Saraga-babic[1]

[1]Department Of Anatomy, Histology And Embryology, University Of Split, School Of Medicine, Split, Croatia, *[2]Department Of Paediatrics, University Hospital In Split, Split, Croatia*, *[3]Department Of Pathology, University Hospital In Split, Split, Croatia*

Introduction: Blood pressure is controlled by kallilkrein-kinin system (KKS) and renin-angiotenzin-aldosteron system (RAAS). In developing kidneys, renin progenitor cells appear in the kidney arterial tree and juxtaglomerular apparatus (JGA), while kallikrein expression characterizes distal nephron. Both renin and kallikrein seem to influence kidney development and maturation. Data on kallikrein expression are conflicting, especially during human kidney development. In this study, we analyze the spatiotemporal and functional relationship between kidney renin and kallikrein during human development, in the postnatal period, and in the nephrotic syndrome of the Finnish type (CNF).
Material and methods: The spatiotemporal expression of renin and kallikrein was analyzed in human kidneys by double immunofluorescence techniques in paraffin sections of 10 human conceptuses 8 to 38 weeks old, in 1,5-year-old healthy kidney tissues and in the CNF.
Results: In the embryonic period, renin - kallikrein expression characterized metanephric cup cells, S-form nephrons, immature glomeruli, and collecting ducts (CD). In fetal and postnatal kidneys, kallikrein was partly expressed in distal tubules (DT) and CD, and strongly along connecting tubules (CT), while renin expression characterized DT and CD, and partly CT. Their co-expression was observed in small areas of DT and CT. In CNF kidneys, renin and kallikrein co-expression was stronger and with longer overlapping areas than in the healthy kidneys.
Conclusions: Our data suggested the interaction of renin and kallikrein during kidney maturation and in the blood pressure control. The two proteins were predominantly expressed in neighboring structures, while their co-expression characterized CT region. In CNF, their changed expression indicated increased efforts of affected kidneys in the blood pressure control.

PI-95 EXPRESSION OF NOTCH2, WNT4 AND SNAIL IN DEVELOPING AND HEALTHY POSTNATAL HUMAN KIDNEYS AND IN PATHOLOGICALLY CHANGED KIDNEYS AFFECTED BY CNF AND FSGS

Marin Ogorevc[1], Ivona Kosovic[1], Marija Juric[1], Natalija Filipovic[1], Ivana Bocina[2], Katarina Vukojevic[1], Snjezana Mardesic[1], Benjamin Benzon[1], Marijan Saraga[3], Mirna Saraga-babic[1]

[1]Department Of Anatomy, Histology And Embryology, University Of Split, School Of Medicine, Split, Croatia, *[2]Department Of Biology, Faculty Of Science, University Of Split*, *[3]Department Of Paediatrics, University Hospital In Split, Split, Croatia*

Introduction: During early human kidney development, metanephric mesenchyme cells undergo mesenchymal–to–epithelial transition (MET). In the S-shape nephron, cells of the proximal nephron differentiate into podocytes and parietal epithelial cells (PECs). Near the vascular glomerular pole, PECs share podocyte markers and ultrastructure. While podocytes are unique terminally differentiated cells, PECs form a simple squamous epithelium. Pathologically changed kidneys are characterized by the proliferation and detachment of podocytes and PECs, and the appearance of cellular bridges between them. We analyzed the expression of Notch2, WNT4 and Snail in developing and postnatal human kidneys, in focal segmental glomerulosclerosis (FSGS) and nephrotic syndrome of the Finnish type (CNF).
Material and methods: Expression of Notch2, WNT4 and Snail was analyzed in sections of 10 human conceptuses 8 to 38 weeks old, in 3 healthy postnatal kidneys and in 7 CNF and FSGS samples, using immunohistochemistry and electron microscopy (EM).
Results: In developing kidneys, WNT4 expression increased moderately in podocytes and strongly in PECs, while in CNF and FSGS kidneys decreased in both cell populations. Notch2 was widely expressed in the developing kidneys, moderately in podocytes and strongly in PECs. In CNF, expression of Notch2 increased, while it decreased in FSGS. Snail was expressed moderately in early nephrogenesis, while in later development Snail increased only in PECs. In CNF and FSGS kidneys Snail increased both in the podocytes and PECs. EM showed cytoplasmic processes between podocytes and PECs in developing and healthy kidneys, in CNF and FSGS.
Conclusions: Developmental expression of WNT4, Notch2 and Snail indicate their role in MET and nephrogenesis of human kidneys, while changed expression pattern characterizes kidneys affected by CNF and FSGS. Cellular bridges characterize both developing podocytes and PECs, while they multiply in CNF and FSGS kidneys.

PI-96 MYCOPHENOLATE MOFETIL (MMF) VERSUS CYCLOPHOSPHAMIDE (CYC) TO PREVENT RELAPSE IN CHILDREN WITH STEROID-DEPENDENT NEPHROTIC SYNDROME (SDNS): A MULTICENTRER, RANDOMIZED, CONTROLLED TRIAL

Veronique Baudouin[1], Annie Lahoche[2], Isabelle Vrillon[3], Stephane Decramer[4], Sylvie Cloarec[5], Gwenaelle Roussey[6], Denis Morin[7], Philippe Eckart[8], Jerôme Harambat[9], Tim Ulinski[13], Christine Pietrement[10], FranÇois Nobili[11], Djamal-dine Djeddi[12], Claire Dossier[1], Corinne Alberti[1], Julien Hogan[1]

[1]*Robert Debre University Hospital- Aphp*, [2]*Chu Lille*, [3]*Chru Nancy*, [4]*Chu Toulouse*, [5]*Chu Tours*, [6]*Chu Nantes*, [7]*Chu Montpellier*, [8]*Chu Caen*, [9]*Chu Bordeaux*, [10]*Chu Reims*, [11]*Chu BesanÇon*, [12]*Chu Amiens*, [13]*Trousseau Hospital-aphp*

Introduction: Previous studies demonstrated the efficacy of CYC and MMF in preventing relapses in children with SDNS but no study to date provided a clear comparison between these two treatments. This study aim at demonstrating that MMF is superior to CYC in preventing relapses in children with SDNS.
Material and methods: We included 70 children (2-16 years old) with SDNS in this open-labeled, randomized, controlled trial. Patients were included during a relapse and received a standardized steroid regimen. Oral CYC was administered at 2mg/kg/d for 12 weeks (cumulative dose 168mg/kg) and MMF at 1200mg/m²/d for 18 months.
Results: 70 children were include in 15 centers: 34 patients were randomized to receive CYC and 35 to MM. Patients' characteristics did not differ between treatment groups. There was no significant difference in relapse rates at 24 months between the CYC group (58%) and the MMF group (57%), p=0,97. There were no differences between relapsers and non-relapsers in terms of sex, disease duration and cumulative dose of steroid in the year prior to inclusion. Younger age was associated with a higher rate of relapse (75% in children <6 vs. 45% in children>6, p=0,02). Among younger children, CYC tended to be associated with a higher rate of relapse compared to MMF (86% vs. 62%, p=0,15), while no difference was found in older children. No significant differences in digestive, infectious or hematological complications were found and 4 patients (12%) in the CYC reported alopecia.
Conclusions: Overall, MMF was not superior to CYC in preventing relapse in children with SDNS. Children under 6 have the highest risk of relapse and MMF may be superior to CYC in this subpopulation.

PI-97 PLASMA EXCHANGE OR IMMUNOADSORPTION FOR RECURRENT FOCAL SEGMENTAL GLOMERULOSCLEROSIS: CLEAR DIFFERENCES IN VITRO

This abstract has been withdrawn.

PI-98 10-YEAR EXPERIENCE OF RENAL BIOPSY MONITORING FOR TACROLIMUS TOXICITY IN NEPHROTIC SYNDROME

Rebecca Calthorpe, Ai May Lee, Tom Mcculloch, Andrew Maxted

Nottingham University Hospitals Nhs Trust

Introduction: Calcineurin inhibitors, including tacrolimus, are considered 2nd or 3rd line treatment for the management of complex nephrotic syndrome. Current practice in our centre is to perform a routine renal biopsy for assessment of nephrotoxicity after 2 years; defined as excessive glomerulosclerosis with or without arteriolar hyalinosis. Significant nephrotoxicity would necessitate the need to consider a switch to non-nephrotoxic immunosuppression. The aim of this audit was to review the management of patients with nephrotic syndrome following evidence of tacrolimus toxicity on renal biopsy.
Material and methods: This was a retrospective audit conducted at Nottingham Children's Hospital tertiary paediatric nephrology centre. Inclusion criteria were patients with a diagnosis of nephrotic syndrome, on tacrolimus therapy undergoing a routine renal biopsy to assess for evidence of tacrolimus toxicity over a 10-year period (1/1/2011 to 31/12/2020). Patients were identified using a histopathology biopsy database with clinical information collected from electronic medical notes.
Results: 44 patients were included (male 66%). The mean age of starting tacrolimus was 4 years, with the interval between commencing treatment and biopsy 34 months. On the patient's first renal biopsy, 41% (18/44) had histological evidence of nephrotoxicity. Consequently 44% (n=8) had treatment discontinued, however 25% (n=2) were recommenced on tacrolimus due to multiple relapses on alternative agents. For patients who were continued on tacrolimus despite evidence of nephrotoxicity (56%, n =10), 3 had evidence of nephrotoxicity on subsequent biopsies.
Conclusions: These results demonstrate that a significant proportion of patients had histological evidence of nephrotoxicity on renal biopsy secondary to tacrolimus. However, decisions to discontinue tacrolimus were multifactorial and tacrolimus was not stopped based on biopsy results alone. Intriguingly, repeat biopsies on some patients with initial toxicity showed apparent histological improvement on subsequent biopsy. Future work will compare our practices with that of other UK centres and answer whether biopsy monitoring on tacrolimus should be mandated.

PI-99 LOW POST VACCINE ANTIBODY TITERS IN CHILDREN WITH IDIOPATHIC NEPHROTIC SYNDROME TREATED WITH RITUXIMAB

Claire Herbez Rea[1], Julie Alsuguren[1], Cyrielle Parmentier[1], Claire Dossier[2], Olivia Boyer[3], Julien Hogan[2], Jean-daniel Delbet[1], Tim Ulinski[1]

[1]Trousseau Hospital, [2]Robert Debre Hospital, [3]Necker Hospital

Introduction: Infections remain a major complication in children with idiopathic nephrotic syndrome (INS). We report post-vaccination antibody (Ab) levels for hepatitis B, measles, diphtheria, tetanus and varicella in patients with steroid dependent INS treated with Rituximab (RTX).
Material and methods: We carried out a retrospective study in the pediatric nephrology units of Trousseau, Necker and Robert Debre hospitals. All children with a diagnosis of INS treated with RTX were included. Post vaccine Ab titers and the following clinical and biological parameters were analyzed: age at diagnosis of INS, number of relapses before and after RTX, duration of INS, other immunosuppressive treatment, immunoglobulin levels, duration of B cell depletion, time between the last RTX dose and time interval between the last relapse and post-vaccination serologies. The primary endpoint was a positive antibody titer.
Results: 37 patients were included. The median age at INS diagnosis was 3.5 (2.5-6.3) years, the median duration of the INS was 9 (5.0-12.5) years, the median time between the last relapse and post-vaccination serology sampling was 17 (6.5-31) months, the median IgG level was 8.3 (6.6-10.4) g/L. 23/37 (62%) patients received calcineurin inhibitors during disease course, 25/37 (67%) received MMF, 15/37 (40%) received both and 24/37 (65%) received Levamisole. The median duration of B cell depletion was 7.5 (3.3-11.5) years and the time between the last RTX dose and vaccine serologies was 14 (6-29) months. The percentage of positive vaccine titers for hepatitis B, measles, diphtheria / tetanus and chickenpox were 35%, 62%, 48% and 32% respectively, all lower than in healthy children. When compared to INS patients on oral drugs without RTX, the percentages of positive titers were the same for hepatitis B and measles, but lower for diphteria/tetanus and varicella (85%-83%).
Conclusions: INS patients treated with RTX had inadequate post-vaccination antibody titers, which did not correlate with any specific treatment modality.

PI-100 DARATUMUMAB ENABLES SUSTAINED REMISSION AFTER IMMUNOADSORPTION IN REFRACTORY MULTIDRUG RESISTANT NEPHROTIC SYNDROME

Claire Dossier[1], Anne-laure Leclerc[2], Marc Fila[3], Robert Novo[4], Lise Allard[5], Theresa Kwon[1], Julien Hogan[1]

[1]Pediatric Nephrology, Robert-debre Hospital, Aphp, Paris, [2]Pediatric Nephrology, Hospices Civiles De Lyon, [3]Pediatric Nephrology, Chu Montpellier, [4]Pediatric Nephrology, Chu Lille, [5]Pediatric Nephrology, Chu Bordeaux

Introduction: Multidrug Resistant Nephrotic Syndrome (MRNS) is a dramatically challenging condition that may lead to end-stage renal disease and post-transplant recurrence. Immunoadsorption of Immunoglobulins (IA) has been reported safe and efficient to induce remission, however most patients relapse after discontinuation, and some happen to be IA-dependent. Long-lived plasma-cells may be responsible for refractory NS. We report the use of Daratumumab(DARA), an antiCD38 monoclonal antibody that targets plasma cells, in IA-dependent MRNS.
Material and methods: In this retrospective multicenter study, we included children with MRNS that reached complete remission after IA, but relapsed when lowering frequency of IA sessions despite B-cell depletion. We report on a further attempt of IA withdrawal adding 4 weekly infusions of daratumumab of 1000mg/1.73m^2.
Results: Four boys and 2 girls were included. Median age at diagnosis was 6.1 years (range 5.5-7.9). Renal biopsy showed FSGS in 3 patients and MCD in 3. All had negative genetic testing. All were resistant to ciclosporine and/or tacrolimus, 4 also received MMF and 3 rituximab(RTX). Median time between INS diagnosis and IA initiation was 1 year (0.5-5.6). All patients achieved complete remission after IA but relapsed after a first discontinuation attempt, despite B-cell depletion with RTX(n=3) or Obinutuzumab(OBI) (n=3). Complete remission was again obtained with intensive IA in all but one with partial remission. All patients received a new infusion of anti-CD20 (obinutuzumab) followed by 4 injections of daratumumab. Complete remission was sustained in all patients enabling IA withdrawal. Proteinuria relapsed in 4 patients (RPC 0.05-0.10g/mmol) and was successfully treated with either a single reinjection of daratumumab (n=2) or combined to OBI and/or IA. All patients were in complete remission at 7 months (range 1.5-17.5) following IA discontinuation.
Conclusions: The association of plasma-cell depletion with daratumumab to B-cell depletion allowed IA discontinuation in all patients with Ig-IA dependent MRNS. Further studies are needed to confirm the efficacy of daratumumab in children with MRNS and better define its place in the treatment strategy.

PI-101 HEALTH-RELATED QUALITY OF LIFE AND PSYCHOSOCIAL ADJUSTMENT OF PATIENTS WITH RECENTLY DIAGNOSED IDIOPATHIC NEPHROTIC SYNDROME

Floor Veltkamp[1], Lorynn Teela[2], Hedy A. Van Oers[2], Elske M. Maknienhuis[1], Lotte Haverman[2], Antonia H.m. Bouts[1]

[1]Amsterdam University Medical Centers, University Of Amsterdam, Emma Childrens Hospital, Department Of Pediatric Nephrology, *[2]Amsterdam University Medical Centers, University Of Amsterdam, Emma Childrens Hospital, Department Of Child And Adolescent Psychiatry And Psychosocial Care*

Introduction: Idiopathic nephrotic syndrome (INS) in children with relapsing disease is associated with lower health-related quality of life (HRQoL) and psychosocial functioning. However, little is known about HRQoL and psychosocial functioning in children with recently diagnosed steroid-sensitive nephrotic syndrome (SSNS). The aim of this study was to assess these outcomes and related variables in children with new onset SSNS.
Material and methods: Dutch and Belgian children (n=46) aged 2-16 years with first onset SSNS participated in a randomised placebo-controlled trial (LEARNS) between April 2018 and December 2020. To measure HRQoL and psychosocial functioning, children (age ≥8 years, self-report) and/or their parents (proxy-report) completed two online generic patient-reported outcome measures (PROMs) at 4 weeks after first onset: the Pediatric Quality of Life Inventory 4.0 and the Strengths and Difficulties Questionnaire. Total and subscale scores and the proportion of children with impaired HRQoL (<1 SD) or clinical scores for strengths and difficulties ($<10^{th}$ and $>90^{th}$ percentile, respectively) were compared to the Dutch norm population. Multivariate regression analyses were used to asses related variables of HRQoL.
Results: Forty patients (87%) completed the PROMs. Older children (8-18 years) reported significantly lower HRQoL on the total scale and the physical and emotional functioning subscales compared to the reference group. A high proportion (>45%) of these children reported impaired HRQoL scores. No differences in HRQoL between children aged 2-4 or 5-7 years and the reference group was found, except for higher scores on social functioning (5-7 years). A small proportion of children scored within the clinical range on psychosocial functioning. No variables related to HRQoL or psychosocial functioning could be identified.
Conclusions: In children with new onset SSNS receiving high doses of steroids, HRQoL and psychosocial functioning was moderately affected, which was more profound in older children. Longitudinal data could provide important insight in the evolution and predictors of HRQoL in SSNS.

PI-102 PATIENTS WITH CONGENITAL ANOMALIES OF KIDNEY AND URINARY TRACT (CAKUT) SHARE COMMON HOTSPOT LOCI WITH LOW FREQUENCY VARIANTS

Jakob Zapušek[1], Mario Gorenjak[2], Danijela Krgović[3], Nataša Marčun Varda[4]

[1]Department Of Paediatrics, General Hospital Ptuj, Potrčeva Cesta 23, 2250 Ptuj, *[2]University Of Maribor, Faculty Of Medicine, Centre For Human Molecular Genetics And Pharmacogenomics, Taborska Ulica 8, 2000 Maribor, Slovenia*, *[3]Laboratory Of Medical Genetics, University Medical Centre Maribor, Ljubljanska 5, 2000, Maribor, Slovenia*, *[4]Department Of Paediatrics, Nephrology Unit, University Medical Centre Maribor, Ljubljanska 5, 2000, Maribor, Slovenia*

Introduction: Congenital anomalies of kidney and urinary tract (CAKUT) are common pathology and important cause of chronic kidney disease in children. The aim of our study was to find genetic correlations between our cohort of children with severe CAKUT phenotypes and relevant genetic findings, using next-generation sequencing (NGS).
Material and methods: Cinical study was performed including 324 CAKUT patients, who had a blood sample for DNA isolation withdrawn. Afterwards, 113 patients with most severe CAKUT were selected for Whole Exome Next-generation sequencing using NovaSeq 6000 Sequencing System and Agilents' SureSelect Human All Exon V6 workflow. All enrolled children had more than one CAKUT and almost all of them (93.8%) had vesicoureteral reflux (VUR).
Results: We found 11 very low population frequency variants (mutations/insertions/deletions) that were present in all 113 patients. 7 of them were splice region mutations, 3 frameshift mutations and 1 missense mutation. Every locus with identified variants in all 113 patients presents many insertions/deletions, thus each locus can be interpreted as a mutation hotspot. Two MUC16 variants on chromosome 19 have been also discovered and may contribute to the phenotype as compound heterozygous mutations if present in different alleles. Further research in terms of genetic testing of parents is needed to elucidate the role of MUC16 in CAKUT. We hypothesise that compound heterozygous variants in MUC16 may present a monogenic VUR aetiology.
Conclusions: In every single included patient with severe CAKUT 11 mutual mutations were found, which is rather uncommon in such studies, promising important findings, warranting further elucidation and confirmation. Both mutations on chromosome 19 are especially interesting as they may present a monogenic VUR aetiology in terms of compound heterozygote.

PI-103 CHANGES IN ANTIBIOTIC RESISTANCE IN CHILDREN WITH THE FIRST FEBRILE URINARY TRACT INFECTION IN LAST TWO DECADES

Ana Petrovic, Srdjan Nikolovski, Dusan Paripovic, Gordana Milosevski Lomic, Milica Vukanovic, Brankica Spasojevic, Mirjana Kostic

University Childrens Hospital – Tirsova

Introduction: Urinary tract infection (UTI) is one of the most common childhood diseases and causes of antibiotic prescriptions. Antibiotic treatment is usually initiated empirically, based on symptomatology, blood and urine analysis, knowing the potential pathogen distribution and sensitivity patterns in population. However, resistance patterns should be monitored regularly in order to keep guidelines up to date. The aim of this study was to assess and compare antibiotic sensitivity and resistance patterns during 2005-2011 and 2017-2021.
Material and methods: We retrospectively reviewed hospital records for patients aged 1 months to 3 years, who were discharged from Nephrology department, with a principal diagnosis of the first febrile UTI, during the two study periods: January 1^{st}, 2005 – December 31^{st}, 2011 (Group 1) and January 1^{st}, 2017 – June 30^{th}, 2021 (Group 2). Urine samples from each patient were obtained at admission before initiating antimicrobial therapy. Urine culture and antibiogram results were analysed. Antibiotic resistance rates were checked for: ampicillin, ceftriaxone, ciprofloxacin, nitrofurantoin, trimethoprim-sulfamethoxazole (TMP-SMX), gentamicin, amikacin, ceftazidime and imipenem.
Results: A total of 690 patients were included in the study, 502 (72.8%) in Group 1 and 188 (27.2%) in Group 2. The median age of all patients was 0.4 (IQR 0.3-0.8) years. E. coli was the most common isolated pathogen taking place in 591/690 cases. Significantly higher antibiotic resistance rates of E. coli isolates were observed with ciprofloxacin, gentamicin and amikacin in Group 2, compared to Group 1. Decrease of resistance rates to TMP-SMX was recorded in Group 2, compared to Group 1. No significant changes in resistance rates to ampicillin, ceftriaxone, nitrofurantoin, ceftazidime and imipenem were noticed.

Conclusions: Comparing two study periods, resistance to ciprofloxacin, gentamicin and amikacin have increased during time, while resistance to TMP-SMX has decreased. Our findings support the need for periodical assessment of antibiotic resistance.

PI-104 GENOME-WIDE ASSOCIATION STUDY IN PATIENTS WITH ANATOMICAL OBSTRUCTIONS OF THE LOWER URINARY TRACT

Loes F. M. Van Der Zanden[1], Carlo Maj[2], Oleg Borisov[2], Iris A. L. M. Van Rooij[1], Josine S.l.t. Quaedackers[3], Martijn Steffens[4], Luca Schierbaum[5], Sophia Schneider[5], Lea Waffenschmidt[5], Lambert A.l.m Kiemeney[1], Liesbeth De Wall[6], Stefanie Heilmann[5], Wolfgang RÖsch[7], Jan Gehlen[8], Johannes Schumacher[8], Maria Szczepanska[15], Katarzyna Taranta-janusz[9], Grazyna Krzemien[10], Agnieszka Szmigielska[10], Michiel Schreuder[11], Stefanie Weber[12], Marcin Zaniew[13], Nel Roeleveld[1], Heiko Reutter[14], Wout Feitz[6], Alina C. Hilger[16]

[1]Radboud Institute For Health Sciences, Department For Health Evidence, Radboud University Medical Center, Nijmegen, The Netherlands, [2]Institute For Genomic Statistics And Bioinformatics, Medical Faculty, University Of Bonn, Germany, [3]Department Of Urology, University Medical Center Groningen, Groningen, The Netherlands, [4]Department Of Urology, Isala, Zwolle, The Netherlands, [5]Institute Of Human Genetics, University Hospital Of Bonn, Bonn, Germany, [6]Radboud Institute For Molecular Life Sciences, Division Of Pediatric Urology, Department Of Urology, Radboudumc Amalia Children's Hospital, Nijmegen, The Netherlands, [7]Department Of Pediatric Urology, Clinic St. Hedwig, University Medical Center Regensburg, Regensburg, Germany, [8]Center For Human Genetics, University Hospital Of Marburg, Marburg, Germany, [9]Department Of Pediatrics And Nephrology, Medical University Of Białystok, Białystok, Poland, [10]Department Of Pediatrics And Nephrology, Medical University Of Warsaw, Warsaw, Poland., [11]Radboud Institute For Molecular Life Sciences, Department Of Pediatric Nephrology, Radboudumc Amalia Children's Hospital, Nijmegen, The Netherlands, [12]University Children Hospital Marburg, Philipps University Marburg, Marburg, Germany., [13]Department Of Pediatrics, University Of Zielona Góra, Zielona Góra, Poland, [14]Division Of Neonatology And Pediatric Intensive Care, Department Of Pediatrics And Adolescent Medicine, Friedrich-alexander University Of Erlangen-nÜrnberg, Erlangen, Germany, [15]Department Of Pediatrics, Faculty Of Medical Sciences In Zabrze, Medical University Of Silesia In Katowice, Zabrze, Poland, [16]Department Of Pediatrics And Adolescent Medicine, Friedrich-alexander University Of Erlangen-nÜrnberg, Erlangen, Germany

Introduction: Congenital lower urinary tract obstruction (LUTO) is most often caused by an anatomical blockage of the urethra most commonly posterior urethral valves (PUV), a male limited phenotype affecting 1 in 4,000 male live births. Little is known about the genetic background of anatomical LUTO. Here, we report the first study using genome-wide association methods to analyze anatomical LUTO in 4 cohorts of patients and controls.
Material and methods: The final meta-analysis comprised 758 patients and 4,823 ethnically matched controls and comprised 5,754,208 variants that were genotyped or imputed and passed quality control in all 4 cohorts.
Results: No genome-wide significant locus was identified, but 33 variants showed suggestive significance ($P<1\times10^{-5}$). When considering only loci with multiple variants residing within <10kB of each other showing suggestive significance and with same effect direction in all four cohorts, three loci comprising together nine variants remained. Loci resided on chromosome 13, 16, and 20.
Conclusions: The present GWAS and meta-analysis is the largest genetic study on anatomical LUTO performed to date. The fact that no genome-wide significant locus could be identified, could be explained by the lack of power or may indicate that common variants do not play a major role in the aetiology of anatomical LUTO. Nevertheless, three loci yielded suggestive association. Future studies are warranted to support these loci.

PI-105 THE DIAGNOSIS OF INTRARENAL REFLUX USING CONTRAST ENHANCED VOIDING UROSONOGRAPHY

Andrea Cvitkovic Roic[1], Iva Palcic[1], Danko Milosevic[2], Daniel Turudic[3], Goran Roic[4]

[1]Clinic For Pediatric Medicine Helena, [2]University Of Zagreb Medical School, [3]University Hospital Center Zagreb, [4]Children´s Hospital Zagreb

Introduction: Contrast-enhanced urosonography (ceVUS) has shown capable diagnostic accuracy for the diagnosis of vesicoureteral refux (VUR) in children but the ability of ceVUS to detect intrarenal refux (IRR) is not yet sufficiently researched. The purpose of our study is to assess the ability of ceVUS to detect IRR as well as its association with age, gender, and the grade of VUR.
Material and methods: The study included 5153 children who were referred to our clinic for ceVUS. All children underwent sonographic examinations, which were performed on a LOGIQ S8 machine equipped with dedicated software for contrast-enhanced studies with harmonic imaging. Standard ultrasound of the urinary tract was followed by bladder catheterisation and instillation of physiological normal saline and the US contrast medium (SonoVue®, Bracco).
Results:
VUR was diagnosed by ceVUS in 1959 out of 5153 children (38%), of whom IRR was found in 233 of 1959 children (11.9%). A total of 285 ureteral units showing IRR were found. High grades of VUR (IV+V) with IRR were found in a total of 235 of 285 (82.81%) renal units. Bilateral IRR was found in 53 patients, usually with a high-grade VUR on both sides. Most children had VUR grade IV, predominantly those younger than 1 year. The younger the child, the higher the likehood of higher grade of VUR (p=0,02).
Conclusions: CeVUS, combined with harmonic imaging and second-generation ultrasound contrast media, enabled IRR detection in almost 12% of our patients with VUR. IRR is most commonly found in children under 1 year of age with VUR grades IV and V.

PI-106 URINE DKK3 AND CHRONIC KIDNEY DISEASE PROGRESSION IN CHILDREN WITH CAKUT, IS IT A RELIABLE BIOMARKER?

AyŞe Seda PinarbaŞi[1], NeslİhanGÜnay[2], İnayet GÜntÜrk[3], DİdemBarlak Ketİ[4], SİbelYel[6], Sekure RabİaUlueren[5], Cevat Yazici[4], Muammer Hakan PoyrazoĞlu[6], İsmaİl Dursun[6]

[1]Diyarbakir Children Hospital, Pediatric Nephrology, [2]Kayseri State Hospital, Pediatric Nephrology, [3]NiĞde Ömer Halisdemir University, Faculty Of Medicine, Department Of Biochemistry, [4]Erciyes University, Faculty Of Medicine, Department Of Biochemistry, [5]Erciyes University, Faculty Of Medicine, [6]Erciyes University, Faculty Of Medicine, Department Of Pediatric Nephrology

Introduction: Chronic kidney disease (CKD) is an important public health issue. Although serum creatinine is most commonly used marker to estimate glomerular filtration rate (GFR), it is affected by many variables such as muscle mass, the search continues for different biomarkers. Recently, DKK3 levels secreted into the urine has been emerged to use as

a short-term, non-invasive biomarker of evaluation of CKD progression. This study was conducted to determine the role of urine DKK3 levels in predicting CKD stage and progression in children with congenital abnormalities of the kidney and urinary tract (CAKUT).
Material and methods: CKD Stage 1-4 patients followed up with CAKUT were included into the study. Urine DKK3 levels, eGFR and urine albumin to creatinine ratio were measured at baseline and at least 6 months of follow-up in children with CAKUT. These values were compared between the CKD groups and with the control group.
Results: A total of 113 patients with CKD stage 1-4 and 28 healthy controls were included into the study. Baseline urine DKK3/creatinine ratio was negatively correlated with eGFR and positively correlated with urine albumin creatinine ratio. When the patients were re-evaluated 6 months later, no correlation was found between urine DKK3 and eGFR changes. Only baseline eGFR was found to be independent predictor for increased urine DKK3 to creatinine ratio in multivariate regression. At the cutoff of 1675,48 pg/mg with 71.1% sensitivity and 71.4% specify.
Conclusions: Urine DKK3 is thought to be an important marker for the progression of CKD as an indicator of renal fibrosis. In this study, we showed high level of urinary DKK3 especially in patients with stage 3-4 CKD in the CAKUT group, which is one of the most important causes of CKD in children, but it is insufficient to predict worsening of eGFR in short-term follow-up of children with CAKUT.

PI-107 A GENETIC INVESTIGATION OF MONOZYGOTIC TWINS DISCORDANT FOR SOLITARY FUNCTIONING KIDNEY

Sander Groen In T Woud[1], Alexander Hoischen[3], Richarda De Voer[3], Marcel Nelen[3], Wout Feitz[4], Nel Roeleveld[1], Loes Van Der Zanden[1], Michiel Schreuder[2]

[1]Radboud University Medical Center, Department For Health Evidence, Radboud Institute For Health Sciences, Nijmegen, The Netherlands, *[2]Radboudumc Amalia Children's Hospital, Department Of Pediatric Nephrology, Radboud Institute For Molecular Life Sciences, Nijmegen, The Netherlands*, *[3]Radboud University Medical Center, Department Of Human Genetics, Radboud Institute For Molecular Life Sciences, Nijmegen, The Netherlands*, *[4]Radboudumc Amalia Children's Hospital, Department Of Urology, Radboud Institute For Molecular Life Sciences, Nijmegen, The Netherlands*

Introduction: The aetiology of solitary functioning kidney (SFK) is likely multifactorial, and some genetic and environmental causes have been identified. With our current understanding, however, a majority of cases cannot yet be explained. The objective of this study was to determine whether postzygotic mutations could play a role in the aetiology of SFK by investigating monozygotic twins discordant for SFK.
Material and methods: Two monozygotic twin pairs, each consisting of one child with congenital SFK and a healthy sibling, were identified in the SOFIA study. DNA was isolated from saliva samples and exomes were captured using Twist Biosciences enrichment kits. Next, whole exome sequencing was performed with a targeted sequencing depth of 300 (NovaSeg6000, Illumina). For both twin pairs, variants present in at least 2 of the reads of the affected but not the unaffected child, as identified with GATK's MuTect2 tool, were classified as postzygotic variants. Coding postzygotic variants were ranked for biological and technical plausibility based on literature and sequencing results, respectively.
Results: Exome sequencing was successful in all four samples, with mean sequencing depths between 120 and 281. In twin pair one, 17 postzygotic variants were called, of which one variant was likely to be true positive. This constitutes a synonymous variant in the VGLL4 gene, which will be confirmed using targeted sequencing. In twin pair two, 108 postzygotic variants were called. However, all variants with high biological plausibility were deemed as technical artefacts after visualization using the Integrative Genomics Viewer.
Conclusions: A postzygotic mutation was identified in one of two monozygotic twin pairs discordant for SFK. This variant warrants further investigation before its clinical relevance can be determined. Although no obvious causal variant was found in the current study, this novel method of studying postzygotic genetic variants could enhance our understanding of the genetic aetiology of SFK.

PI-108 PREDICTABILITY FACTORS OF PATIENT SURVIVAL IN CASE OF RENAL OLIGOHYDRAMNIOS

Mathilde Baudoin, Claire Herbez-rea, Ferdinand Dhombres, Isabelle Guellec, Jean Marie Jouannic, Tim Ulinski

Trousseau Hospital - Aphp.sorbonne University

Introduction: Renal oligohydramnios (ROH) is a poor prognostic factor of neonatal survival in CAKUT patients, lung hypoplasia being the main cause of mortality. We aimed to describe the foetal morbidity and mortality in case of ROH and to find predictive risk factors for morbidity which may help in antenatal counceling and post natal care.
Material and methods: All data were retrospectively collected at the obstretrics, neonatology and pediatric nephrology units of Trousseau hospital, from 2008 to 2020. All fetuses with renal oligohydramnios were included.
Results: We included 66 foetuses with renal parenchymal pathologies or posterior urethral valves (PUV) (N=25), bilateral kidney agenesis (N=10), hypodysplasia (N=16), polycystic kidney disease (N=10) causing oligamnios or anamnios, identified on antenatal ultrasound. Total mortality was 75% (50/66) including 35% antenatal deaths (22 terminations of pregnancy and 1 intrauteral death), 10 died immediately after birth, 17 died in neonatal intensive care unit and 16 survived. The presence of pneumomediastinum and pneumothorax was not different in survivors and non-survivors. The mortality in case of PUV was 52% (13/25) including 5/13 antenatal, 6/13 in ICU and 2/13 in neonatology, 3 immediately after birth. For patients with hypodysplasia 88% (14/16) have died including 3 before birth, 3 immediately after birth, and 8 in ICU. Foetuses with kidneys having features of hypodysplasia on ultrasound at T2 and those with anamnios or oligohydramnios before 32 weeks GA had a higher risk to die. There was a significant difference in plasma creatinine of the surviving patients compared to the deceased patients, from postnatal day 3 onwards (183 μmol/L [88; 255] vs. 295 μmol/L [247; 326]; p=0.038).
Conclusions: Pulmonary hypoplasia was not associated with an increase of neonatal mortality in this specific patient setting. However, the increase of serum creatinine (from day 3 onwards) and oligohydramnios detected before 32 weeks GA were different in survivors vs. non-survivors.

PI-109 PRETERM BIRTH: IS IT A RISK FACTOR FOR HYPERTENSION AND REDUCED KIDNEY VOLUME?

Ozge Oguzhan[1], Ayse Agbas[2], Seha Saygili[2], Nazli Gulsum Akyel[3], Kubra Yilmaz[1], Sebuh Kurugoglu[3], Dildar Konukoglu[4], Nur Canpolat[2], Salim Caliskan[2], Lale Sever[2]

[1]Istanbul University-cerrahpasa, Cerrahpasa Faculty Of Medicine, Department Of Pediatrics, Istanbul, Turkey, *[2]Istanbul University-cerrahpasa, Cerrahpasa Faculty Of Medicine, Department Of Pediatric Nephrology, Istanbul, Turkey*, *[3]Istanbul University-cerrahpasa, Cerrahpasa Faculty Of Medicine, Department*

Of Pediatric Radiology, Istanbul, Turkey, [4]*Istanbul University-cerrahpasa, Cerrahpasa Faculty Of Medicine, Department Of Medical Biochemistry, Istanbul, Turkey*

Introduction: The aim of this study is to evaluate the kidney volume and its relationship with kidney function and blood pressure (BP) in children born prematurely with low birth weight.
Material and methods: This cross-sectional observational case-control study included 50 preterm-born children (mean age 11.5±1.9 years, 58 % female) with low birth weight (preterm group) and 27 term-born children (control group). The preterm group was divided into subgroups according to birth weight and gestational age, as small and appropriate for gestational age, SGA and AGA groups, respectively. Estimated glomerular filtration rate (eGFR, by Zappitelli formula using creatinine and cystatin C) and microalbuminuria (urine albumin-to-creatinine ratio) were assessed. Blood pressure (office and 24-hour ambulatory blood pressure monitoring) and kidney volume measurements (ultrasound, USG and magnetic resonance imaging, MRI) were performed. Kidney volumes were adjusted by body surface area and expressed as total kidney volume index; USG-TKVi and MRI-TKVi, respectively.
Results: The median USG-TKVi of the preterm group was smaller than the control group [131 (115;143) cm3/m2 vs 143 (127;159) cm3/m2, respectively, p=0.036]; however, the MR-TKVi was not significantly different between the two groups. There was no difference in eGFR, BP-SDSs between preterm and control groups. In univariate analysis, both USG- and MRI-TKVi were positively correlated with birth weight-SDS (p=0.027, r=0.312 and p=0.040, r=0.292, respectively) and MRI-TKVi was also correlated with eGFR (p=0.008, r=0.373), but there was no correlation with gestational week or BP-SDSs. In the subgroup analysis, the SGA group (n=16) had significantly lower median USG-TKVi [119 (110;130) cm3/m2 vs 136 (124;146) cm3/m2, respectively, p=0.017] and MRI-TKVi [109 (103;123) cm3/m2 vs 123 (115;139) cm3/m2, respectively, p=0.022)] compared to the AGA group (n=34), but the BP-SDs were not different.
Conclusions: Preterm-born children, especially who were SGA, have lower kidney volume compared to healthy counterparts. Therefore, long-term follow up of these children is important.

PI-111 NOVEL RENAL PHENOTYPES IN KBG SYNDROME: A CASE SERIES

Natalie Wyatt, Faidra Veligratli, Aoife Waters

Great Ormond Street Hospital

Introduction: KBG syndrome is a rare autosomal dominant syndrome associated with mutations or deletions in ANKRD11. Named for the three families first identified, features of KBG syndrome include macrodontia of upper central incisors, distinctive facies, skeletal abnormalities, seizures, and developmental delay. To date renal manifestations have not been described in the literature. ANKRD11 gene encodes for ankyrin repeat domain-containing protein which inhibits ligand-dependent activation of transcription. It is expressed in the kidney. We present two cases which, for the first time, describe kidney anomalies in children with KBG syndrome.
Material and methods: We retrospectively reviewed paediatric cases of KBG syndrome associated with renal anomalies presenting to Great Ormond Street Hospital in 2021. Genetic diagnosis was made with the GOSHome gene panel test (DDTOP).
Results: Two cases have been identified. A 4-year-old female presenting with seizures, developmental delay, and bilateral conductive hearing loss. Genetic testing identified a heterozygous frameshift mutation in ANKRD11 c.1356_1359del p.(Asn452Lysfs*2). Ultrasound revealed a left kidney duplex malformation and a normal right kidney in the context of normal kidney function, blood pressure and negative urine dipstick test. A second unrelated, female with developmental delay and abnormal feet posturing with a heterozygous mutation in ANKRD11 c.6538dup, p.(SER2180fs) was incidentally found to have chronic kidney disease (eGFR is 45ml/min/1.73m^2) during investigation of suboptimal growth. Ultrasound revealed bilateral pelvicalyceal and ureteric dilatation, loss of corticomedullary differentiation but appropriately grown kidneys. The long-term prognosis is unknown and thus both are undergoing follow up.
Conclusions: To date more than 200 cases of KBG syndrome have been reported in the literature. This is the first description of an association with kidney anomalies. Imaging and assessment of kidney function should be performed in those diagnosed with KBG syndrome. Furthermore, these cases highlight the importance of investigating the genetic basis of CAKUT in the context of extra-renal manifestations.

PI-112 NON-INVASIVE URINARY BIOMARKERS OF KIDNEY INJURY IN CHILDREN WITH SICKLE CELL DISEASE – THE BIODREPA STUDY

Rute Baeta Baptista[1], Carolina Santos[2], Joao Ferreira Simoes[3], Ana Araujo Carvalho[3], Marisa Oliveira[4], Sara Batalha[4], Raquel Maia[4], Paula Kjollerstrom[4], Marta Verissimo[5], Teresa Ferreira[5], Marta Contreiras[6], Andreia Matos[2], Margarida Abranches[1], Manuel Bicho[2], Edgar Almeida[7]

[1]*Paediatric Nephrology Unit, Department Of Paediatrics, Hospital Dona Estefania, Centro Hospitalar Universitario De Lisboa Central, Lisbon, Portugal*, [2]*Laboratorio De Genetica E Instituto De Saude Ambiental, Faculdade De Medicina, Universidade De Lisboa, Lisbon, Portugal*, [3]*Department Of Paediatrics, Hospital Dona Estefania, Centro Hospitalar Universitario De Lisboa Central, Lisbon, Portugal*, [4]*Paediatric Haematology Unit, Department Of Paediatrics, Hospital Dona Estefania, Centro Hospitalar Universitario De Lisboa Central, Lisbon, Portugal*, [5]*Department Of Paediatrics, Hospital Prof. Doutor Fernando Fonseca, Amadora, Portugal*, [6]*Department Of Paediatrics, Hospital Beatriz Angelo, Loures, Portugal*, [7]*Centro Cardiovascular Da Universidade De Lisboa (ccul), Centro Academico Medico De Lisboa (caml), Faculdade De Medicina, Universidade De Lisboa, Lisbon, Portugal*

Introduction: Sickle cell nephropathy (SCN) affects 30-50% of SCD patients and may lead to premature death. Glomerular filtration ratio (GFR) and urine albumin-to-creatinine ratio (uACR) become altered only in advanced SCN. The main goal of the BIODREPA study is to assess non-invasive biomarkers of kidney injury as a tool to detect SCN in children.
Material and methods: Multicentre prospective cohort study of paediatric patients with SCD. The primary outcome was a composite of decreased GFR (<90 mL/min/1,73m2), albuminuria (uACR >30 mg/g), or abnormal findings in kidney ultrasound. Urinary levels of alpha-glutathione S-transferase (a-GST), neutrophil gelatinase-associated lipocalin (NGAL), and kidney injury molecule-1 (KIM-1) were compared between groups defined according to the primary outcome.
Results: We report data from the 65 patients included in the initial cross-sectional analysis of the BIODREPA study. The median age was 8.4 years (IQR 5.5-12.5) and 57% were male. Median GFR was 149 mL/min/1,73 m2 (IQR 124-164). The primary outcome was met by 59.7% of the cohort (low GFR in 0%; albuminuria in 6.2%; and abnormal kidney ultrasound in 57.1%). The median (IQR) urinary levels of a-GST, NGAL, and KIM-1 normalized to urinary creatinine in ng/mg were: 20.8 (11.1-53.6), 3.2 (1.4-7.9), and 1.7 (0.7-3.3), respectively. In a comparison between groups according to the primary outcome, a-GST urinary levels were significantly higher in patients with albuminuria (52.1 versus 19.1 ng/dL, p-value 0.014). Despite that, none of the biomarkers was a

significant predictor of the primary renal outcome in the logistic regression model adjusted for age and sex.
Conclusions: In the preliminary results of the BIODREPA study, the prevalence of renal involvement among children with SCD was about 60%. The urinary a-GST may be the most promising of the biomarkers studied for early detection of SCN. Future research should prospectively assess consecutive patients with sickle cell disease to identify early predictors of kidney injury.

PI-113 THE IMPORTANCE OF REANALYSIS OF NGS DATA AND FURTHER FUNCTIONAL ANALYSIS ON THE EXAMPLE OF 5 PATIENTS WITH A CLINICALLY DIAGNOSED HYPOPHOSPHATEMIC RICKET.

Margarita Sharova[1], Svetlana Papizh[2], Olga Levchenko[1], Alexandra Filatova[1], Andrey Marakhonov[1], Anatoliy Tulpakov[1], Mikhail Skoblov[1]

[1]Research Centre For Medical Genetics, Moscow, Russia, *[2]Veltishev Research & Clinical Institute Of Pediatrics, Pirogov Russian National Research Medical University, Moscow, Russia*

Introduction: Pathogenic variants in SLC34A1 and SLC34A3 genes encoding Na/Pi transporters are identified in patients with Hereditary Hypophosphatemic Rickets with Hypercalciuria (HHRH; MIM#241530) and Idiopathic Infantile Hypercalcemia, type 2 (IIH2; MIM#616963).
Material and methods: Five patients with hypophosphatemic ricket were referred for molecular genetic testing. Four patients underwent WGS followed by reanalysis of genomic data; NGS panel data were reanalyzed for one. Experimental investigation of the effect of two variants on splicing was performed using minigene assay and RNA analysis.
Results: Primary NGS data analysis didn't reveal causative variants for patients or there was only one variant in SLC34A1 and SLC34A3. Secondary investigation of NGS data revealed previously missed in-frame deletion p.91_97del in SLC34A1with global frequency 1,7% in two patients in compound heterozygous state. Intronic deletion c.925+20_926-48del101 in SLC34A3 was found in two brothers. Both variants were previously described as pathogenic. Missed synonymous VUS c.1449G>A in SLC34A1 was detected in one. Minigene assay showed that the variant leads to truncation of exon 13 by 34 nucleotides with PTC formation that allows us to reclassify the variant as likely pathogenic. Another synonymous c.846G>A variant in SLC34A3 was also missed during primary investigation. Functional study was performed by RT-PCR and identified the variant as splicing one, leading to skipping of exon 8 without frameshift.
Conclusions: A molecular genetic diagnosis of HHRH and IIH2 is extremely significant for early diagnosis, correct treatment optionsand genetic counseling. Reanalysis of genomic data is important not only over time, but also by different clinical bioinformaticians. Some pathogenic variants with high global frequency could be filtered out during early stages of NGS analysis. Functional analysis allows to reclassify VUS and investigate the pathogenicity previously described variants.

PI-114 END-STAGE KIDNEY DISEASE IN PRIMARY HYPEROXALURIA TYPE 1: LOOKING FOR DETERMINANTS

This abstract has been withdrawn.

PI-115 A STROMME SYNDROME PATIENT WITH END STAGE RENAL DISEASE AND A NOVEL PATHOGENIC VARIANT IN CENPF GENE

Demet Alaygut[1], Berk Özyilmaz[2], ÖzgÜr Özdemİr ŞİmŞek[1], GÖkÇen ErfİdAn[1], SeÇİl Arslansoyu Çamlar[1], Belde Kasap DemİR[3], Fatma Mutlubas[1]

[1]University Of Health Sciences Tepecik Training And Research Hospital Department Of Pediatric Nephrology, [2]University Of Health Sciences Tepecik Training And Research Hospital Department Of Genetic Diseases Evaluation Center, [3]Katip Celebi University Faculty Of Medicine Department Of Pediatric Nephrology And Rheumatology

Introduction: Stromme syndrome is an autosomal recessive congenital disorder involving multiple systems that shares clinical features with ciliopathies. Here, we report a girl with a novel homozygous pathogenic variant in CENPF gene, presented with end stage renal diseases (ESRD) with Stromme syndrome clinical features.
Material and methods: An 8-year-old girl was admitted with anemia, growth retardation and metabolic acidosis. (Urea: 173 mg/dl, Cre: 10 mg/dl, PTH: 947, Ca 6.6 mg/dl, P 8.3 mg/dl, K 5.95, Hb: 7.5 g/dL HCO_3: 12 9 . She was hypertensive (170/100 mmHg). Weight was 26 kg (25-50 p, -0.43 SDS), height was 114 cm (< 3p, -2.93 SDS), head circumference was 43 cm (< 3p, -6.27 SDS), dysmorphic facial appearance was present. Emergency hemodialysis was performed. She was investigated for the etiology of chronic kidney disease. C3 and C4 levels were found to be normal. Tubular tests were defective (Pro/ Cre: 3.2 mg/mg, FeNa 8.2, FeK 59.8, TPR: 51%). Microcephaly, mega cisterna magna, hypoplasia in the right cerebellar hemisphere were detected due to cranial MRI. Posterior embryotoxon was detected in the left eye. For the molecular genetic evaluation of the patient, SNP microarray analysis was performed at fist-step and no clinically significant variant was detected.
Results: For further evaluation, WES was performed. The patient was found to carry a homozygous c.117del (p.Gln40SerfsTer26) variant in (ENST00000366955.3) CENPF gene. This variant was not previously reported in databases or literature and according to the ACMG criteria (PVS1, PM2, PP3) it was interpreted as a novel "Pathogenic" variant.
Conclusions: The centromeric protein F (CENPF) human ciliopathy gene was included in the ciliopathy-related diseases group in 2016. By writing this report we have contributed to the literature by presenting a Stromme Syndrome patient with end stage renal disease and a novel pathogenic variant in CENPF gene.

PI-116 CURRENT CLINICAL PRACTICE OF GENETIC SCREENING IN PATIENTS WITH HEREDITARY DISEASES: FINDINGS FROM THE EUROPEAN RARE KIDNEY DISEASE REGISTRY (ERKREG)

Giulia Bassanese[1], Tanja Wlodkowski[1], Aude Servais[2], Dario Roccatello[3], Laurence Heidet[4], Francesco Emma[5], Gema Ariceta[6], Stéphane Decramer[7], Elena Levtchenko[8], Augustina Jankauskiene[9], Giovanni Montini[10], Jun Oh[12], Jaap Groothoff[11], Franz Schaefer[1]

[1]Division Of Pediatric Nephrology, Center For Pediatrics And Adolescent Medicine, University Of Heidelberg, Heidelberg, Germany, [2]Nephrology And Transplantation Department, Centre De Référence Des Maladies Rénales Héréditaires De L'enfant Et De L'adulte, Necker University Hospital, Aphp, Université De Paris, Paris, France, [3]Nephrology And Dialysis Unit, San Giovanni Hub Hospital And Department Of Clinical And Biological Sciences, University Of Turin, Turin, Italy, [4]Aphp, Pediatric Nephrology Unit, Centre De Référence Des Maladies Rénales Héréditaires De L'enfant Et De L'adulte (marhea), Hôpital Universitaire Necker-enfants Malades, 75015, Paris, France, [5]Division Of Nephrology, Bambino Gesù Children's Hospital Irccs, Rome, Italy, [6]Department Of Paediatric Nephrology, Hospital Universitario Vall D'hebron, Barcelona, Spain, [7]Pediatric Nephrology, Internal Medicine And Rheumatology, Southwest Renal Rares Diseases Centre (sorare), University Children's Hospital, Toulouse, [8]Department Of Pediatric Nephrology And Development And Regeneration, University Hospitals Leuven, University Of Leuven, Leuven, Belgium, [9]Pediatiic Center, Institute Of Clınıcal Medicine, Vilnius Universıty, Vilnius, Lithuania, [10]Pediatric Nephrology, Dialysis And Transplant Unit, Fondazione Ca' Granda Irccs, Policlinico Di Milano, Milan, Italy, [11]Department Of Pediatric Nephrology, Amsterdam University Medical Center, Amsterdam, The Netherlands., [12]Pediatric Nephrology, University Medical Center Hamburg-eppendorf, Hamburg, Germany

Introduction: Genetic screening is rapidly moving from a research tool to the first-line diagnostic procedure for hereditary kidney diseases. We examined the current state of genetic screening for hereditary nephropathies in Europe.
Material and methods: Since 2019, the European Rare Kidney Disease Registry (ERKReg) has monitored diagnostic procedures in >13.000 patients at 75 centers in 24 European countries. Here we evaluated the use of genetic screening in 4.242 registry patients diagnosed with a rare kidney disease from 01/16-12/21.
Results: Genetic screening was performed in 1.259 patients, including 53% of all patients with tubulopathies (TP) and metabolic nephropathies (MNP), 39% of ciliopathy patients, 38% of patients presenting with thrombotic microangiopathies (TMA), 27% of glomerulopathy and 11% of CAKUT patients. Genetic screening for hereditary diseases was ordered twice as common in pediatric as in adult patients (36% v. 18%). Among the patients screened, a genetic diagnosis was established in 96% of MNP, 92% of ciliopathies, 91% of TP, 68% of TMA and 56% of CAKUT cases.
The screening turnaround time shortened over time to a current median of 3.4 months. Notable differences were noticed between countries, with a range of 1.3 months in Germany to 10.5 months in Italy. Sanger sequencing of individual genes was used in 22%, NGS gene panels in 63% and whole exome sequencing in 15% of patients. Median turnaround time was 2.5 months for Sanger, 3.3 months for NGS panel screening, and 5.8 months for whole exome sequencing. In those patients in whom a genetic diagnosis was established, median time to genetic diagnosis was 10 months from disease onset, and 8.5 months from referral to the specialist center.
Conclusions: Genetic screening yields remarkably high diagnostic confirmation rates. While decreasing, laboratory turnaround times still require improvement to maximize the clinical usefulness of genetic screening.

PI-117 EVOLUTION OF HEMATURIA OVER TIME AND CORRELATION WITH DIFFERENT GENOTYPES IN A COHORT OF ALPORT CHILDREN AND YOUNG ADULTS

Laura Lucchetti[1], Alessandra Terracciano[2], Anna Ewa Kaminska[2], Alma Iafisco[1], Antonio Novelli[2], Francesco Emma[1], Laura Massella[1]

[1]Division Of Nephrology, Bambino Gesù Childrens Hospital Irccs, Rome, Italy, [2]Laboratory Of Medical Genetics, Translational Cytogenomics Research Unit, Bambino Gesù Childrens Hospital, Irccs, Rome, Italy

Introduction: Hematuria is usually the earliest finding in patients with Alport Syndrome (AS). However, adults with preserved kidney function and very mild or no hematuria have also been reported. In these patients, renal biopsy usually shows focal segmental glomerulosclerosis and the diagnosis can be missed if electron microscopy is not performed.
Aim: To evaluate the evolution of hematuria over time and its correlation with genotype in patients with AS.
Material and methods: We collected patients with X-linked, autosomal recessive or autosomal dominant AS (XL-AS, AR-AS, AD-AS) followed at our center, diagnosed before age 18 years, with eGFR ≥70 ml/min/1.73m^2 at diagnosis and at least 5 years of follow-up.

Hematuria was stratified in four classes according to the number of red blood cells in the urinary sediment (1=0-15; 2=16-50; 3=51-150; 4=>150). Data were grouped according to patient age (0-11, 12-18, >18 years). Genotypes were grouped based on the theoretical impact of pathogenic variants as follows: severe genotype = AR-AS or XL-AS males with large deletions, splice-site substitutions, or frameshift variants; intermediate genotype = XL-AS males with missense variants; mild genotype = XL-AS females or AD-AS.
Results: Eighty-nine patients (54M) were included in the study (mean age at diagnosis of 9.9 ± 5.3 years). A statistically significant decrease in the magnitude of hematuria over time was observed in all patient categories. The percentage of urine samples with highest score reduced from 33% in 0-11 years group to 11% in >18 years group (D = 22%, $p<0.05$). This effect was more pronounced in severe genotype group with a reduction of 34% overtime.
Conclusions: The natural evolution of hematuria in patients with AS is to decrease overtime. In some cases, hematuria may become very mild or even absent. This may explain the unexpected findings of pathogenic variants in COLIV genes in adult patients diagnosed with FSGS.

PI-118 HYPONATREMIA, A LOW-COST DIAGNOSTIC TOOL, THAT PREDICTS SEVERE CARDIAC INVOLVEMENT IN CHILDREN WITH MIS-C

Kyriaki Papadopoulou-legbelou, Maria Kavga, Evangelia Desli, Panagiota Karananou, Olga Vambertzi, Sofia Markidou, Elissavet Vakouftsi, Paraskevi Panagopoulou, Efimia Papadopoulou-alataki, Michail Portokalas, Athanasia Nikolakaki, Sofia Chantavaridou, Maria Fotoulaki, Despoina Tramma

Aristotele University Of Thessaloniki

Introduction: Objective: To highlight the role of low sodium in early recognition of severe cardiac involvement in children presenting with multisystem inflammatory syndrome (MIS-C).
Material and methods: We describe a case series of nine patients, aged from 3,5 to 15,5 years that presented to our hospital during last winter and fulfilled the criteria of MIS-C with fever, elevated inflammatory markers and involvement of at least two organ systems, following a recent Covid-19 infection.
Results: Eight out of 9 patients experienced gastrointestinal symptoms, 2/9 had lymphopenia, 3/9 thrombocytopenia and 2/9 children had severe hypoalbuminemia. Only one child developed decreased renal function, which was confirmed by high plasma urea and creatinine levels. However, hyponatremia (sodium:125-135mmol/L), was detected in 8/9 children (88.8%), which was severe in 3 children (33.3%). Severe hyponatremia was associated with severe cardiac involvement, as well as with remarkable high brain natriuretic peptide (BNP) values, combined with only a slight increase in troponin levels. Echocardiogram revealed mild left ventricular dysfunction (EF:50-55%) in 6/9 children, pericardial effusion (mild in 5/9, moderate in 2/9), and coronary artery dilation in 2/9 children. All patients were treated with intravenous human immunoglobin (IVIG), corticosteroids and low-dose of aspirin and only one child needed cardiac support with milrinone.
Conclusions: Children with MIS-C often develop hyponatremia, the severity of which is related to the severity of the disease. Hyponatremia in MIS-C has been associated with inappropriate secretion of antidiuretic hormone, probably caused by the cytokine storm. Furthermore, high BNP levels in MIS-C patients, are mainly related to myocardial edema and not to myocardial cell damage, as observed in myocarditis, and to natriuresis, probably another underlying mechanism associated with hyponatremia.

PI-119 EVALUATION OF BLOOD PRESSURE AND RENAL FUNCTIONS IN CHILDREN WITH COVID-19 INFECTION

Özge ÖzÇelik[1], Selma Oktay Ergin[2], Medine Helin Tanfer[1], Nazlı Umman[1], Ahmet Irdem[2], Hasan Dursun[3]

[1]Health Sciences University, Prof. Dr. Cemil Taşçıoğlu City Hospital, Department Of Pediatrics, [2]Health Sciences University, Prof. Dr. Cemil Taşçıoğlu City Hospital, Department Of Pediatric Cardiology, [3]Health Sciences University, Prof. Dr. Cemil Taşçıoğlu City Hospital, Department Of Pediatric Nephrology

Introduction: COVID-19 may result in pathologies such as high blood pressure and renal dysfunction. The aim of this study is to investigate the rate of hypertension and the renal damage in children who had COVID-19 infection.
Material and methods: A total of 97 children, 61 girls and 36 boys, aged between 4-18 years, who were admitted to the Pediatric Nephrology and Pediatric Cardiology outpatient clinics with a history of COVID-19 at least 3 months ago are included in this study. A total of 22 healthy children, 13 girls and 9 boys, similar to the patient group in terms of age and gender, were included in the control group. A 24-hour ambulatory blood pressure measurement was performed in all of the cases included in the study. Blood and urinalysis results were recorded retrospectively from patient files and hospital electronic medical records.
Results: When the patient and control groups were compared in terms of ambulatory blood pressure monitoring findings, hypertension was found in 35 (36%) of 97 patients included in the study, whereas hypertension was found in only 1 (4.5%) of 22 patients in the control group. The mean 24-hour, day and nighttime mean arterial pressure, nighttime systolic blood pressure and blood pressure load are statistically significantly higher in the group that had COVID-19 compared to the controls. In addition, 24-hour, day and nighttime heart rate, systolic and diastolic dipping are found to be statistically significantly lower in the group who had COVID-19. Statistically significant elevations were found in the group with COVID-19 infection in terms of hemoglobin, hematocrit, creatinine, and potassium.
Conclusions: In this study, the hypertension rate of patients with COVID-19 infection is found to be higher than the control group, and it is an important finding that they are non-dipper in addition to being hypertensive. Moreover, the blood creatinine level, which is one of the biochemical parameters retrieved from the patients, is found to be higher than the controls group, suggesting that COVID-19 infection affects renal glomerular functions but not renal tubular functions.

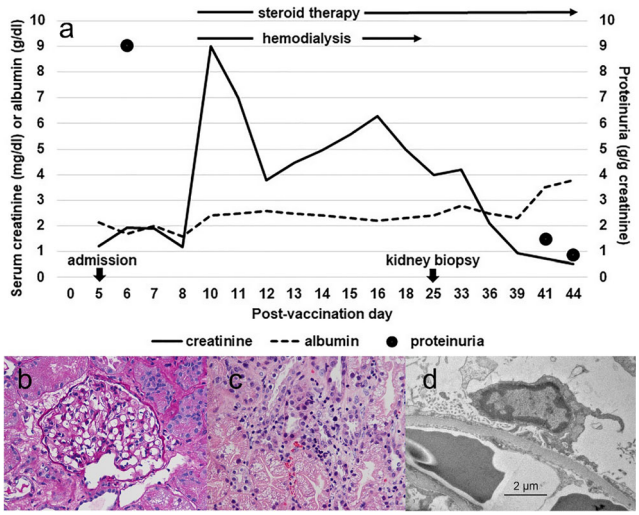
PI-120 CHILDHOOD NEPHROTIC SYNDROME AND ACUTE KIDNEY INJURY FOLLOWING THE SARS-COV-2 BNT162B2 VACCINE

Pondtip Jongvilaikasem[1], Nuanpan Siripen[2], Yong Poovorawan[3], Pornpimol Rianthavorn[2]

[1]Pediatric Division, Hat Yai Medical Education Center, Hat Yai Hospital, Songkhla, Thailand, [2]Division Of Nephrology, Department Of Pediatrics, Faculty Of Medicine, Chulalongkorn University, [3]Center Of Excellence In Clinical Virology, Faculty Of Medicine, Chulalongkorn University

Introduction: Widespread vaccination is a critical tool to protect everyone from coronavirus disease 2019. The SARS-CoV-2 BNT162b2 vaccine (COMIRNATY, Pfizer-BioNTech) has received emergency authorization for children aged five years and above. Although results from the vaccine safety monitoring are reassuring, it is important to continue to monitor the safety of the vaccine.

Material and methods: The clinical course of a male adolescent who developed minimal change disease (MCD) and acute kidney injury (AKI) after first injection of COMIRNATY was retrospectively reviewed.
Results: A previously healthy 14-year-old boy developed anasarca five days after the first vaccination. Clinical evaluation revealed hypertension, nephrotic syndrome (proteinuria 4+ and 9 g/g creatinine of spot urine, albumin 2 g/dl and cholesterol 257 mg/dl), and AKI (creatinine 2 mg/dl). Diagnostic workup, including C3/C4, ANA, ANCA, hepatitis B surface antigen, and antibodies to hepatitis C virus, were negative. Ten days after vaccination, he became anuric with peak creatinine of 9 mg/dl. Three doses of daily pulse methylprednisolone followed by 60 mg of daily oral prednisolone were administered. He received acute hemodialysis for three weeks (Figure 1a). Kidney biopsy showed unremarkable glomeruli, diffuse tubular epithelial injury, interstitial inflammatory cell infiltration, and negative immunofluorescence staining. Electron microscopy showed diffuse foot process effacement (Figure 1b-d). After the 5-week treatment with corticosteroids, the patient was in partial remission (creatinine 0.53 mg/dl and proteinuria 0.9 g/g creatinine). Total immunoglobulins specific to the receptor-binding domain of the SARS-CoV-2 spike protein using Elecsys® (Roche Diagnostics, Basel, Switzerland) was 5.6 U/ml (positive ≥0.8 U/ml).



Conclusions: The temporal association with vaccination in the patient suggests that a T-cell mediated immune response to viral mRNA could induce podocytopathy. The incidence of MCD following COMIRNATY injection needs to be determined. Including nephrotic syndrome into pediatric safety concerns should be considered to raise clinician and parental awareness of this potential adverse effect.

PI-121 "ANTIBODY RESPONSE TO SARS-COV2 MRNA VACCINES IN IMMUNOSUPPRESSED CHILDREN: VARIAN-COV STUDY"

Jessica Serafinelli[1], Antonio Mastrangelo[1], William Morello[1], Chiara Tamburello[1], Martina Rossano[2], Giovanni Filocamo[2], Antonella Petaccia[2], Francesca Minoia[2], Massimo Oggioni[3], Ferruccio Ceriotti[3], Giovanni Montini[1]

[1]Pediatric Nephrology, Dialysis And Transplant Unit, Irccs Ca Granda Ospedale Maggiore Policlinico, Milan, Italy, [2]Pediatric Intermediate Care Unit, Irccs Ca Granda Ospedale Maggiore Policlinico, Milan, Italy, [3]Clinical Laboratory, Irccs Ca Granda Ospedale Maggiore Policlinico, Milan, Italy

Introduction: SARS-CoV-2 vaccination provides protection in immunocompetent persons, while antibody response in immunosuppressed children is unclear. We evaluate the humoral immunogenicity of mRNA SARS-CoV-2 vaccines in immunosuppressed nephropathic children.
Material and methods: Single centre prospective observational study (April 2021-January 2022). Three groups of patients >5 years of age, receiving 2 doses of SARS-CoV-2 mRNA-vaccines, were enrolled: Kidney group (K), children undergoing immunosuppression because of glomerulonephritis, nephrotic syndrome, SLE/vasculitis; Rheumatologic group (R), children immunosuppressed because of juvanile idiopathic arthtitis/uveitis, connectivitis; and Control group (C), non-immunosuppressed children. IgG against receptor-binding domani of the spike protein and nucleocapsid were measured with the Elecsys® assay (Roche), before the vaccination (T0) and 2 weeks after the second dose (T1). Patients testing positive for nucleocapsid-IgG were excluded, because of the previous infection. Clinical, demographic and lab data were collected. The study was approved by the Ethics Committee.
Results: Excluding patients with a previous infection (presence of nuclecapside-IgG at T0) and those lost to follow-up, we report data for 41 (median age 17,3y, 37% male), 19 (16,5y, 53% male) and 40 (16,7y, 32% male) subjects from K, R and C group respectively. Ongoing immunosuppression was different for the 2 disease groups: in the K group mainly MMF, steroids, calcineurin inhibitors and anti-CD20, while in the R group MTX±anti-TNF/IL6. 73% of patients of the K group and 100% of both the R and C group developed RBD-IgG. Furthermore, lower titres (median 568 U/ml, IQ 93-2147) were present in the K group, compared to R patients (3675, 1237-10928) and the C group (6427, 3478-9752) (p <0,0001)., while no statistically differences were present between the R and C groups.
Conclusions: Immunosuppressed children showed a good response to SARS-CoV-2 mRNA vaccines, although underlying disease and type of immunosuppression affect the grade of response.

PI-122 COVID-19 IN CHILDREN WITH CHRONIC KIDNEY DISEASE; DOES IT DIFFER MUCH?

Demet Baltu, Eda Didem Kurt Sukur, Tugba Tastemel Ozturk, Bora Gulhan, Fatih Ozaltin, Ali Duzova, Rezan Topaloglu

Department Of Pediatric Nephrology, Faculty Of Medicine, Hacettepe University, Ankara, Turkey

Introduction: Although the clinical course of coronavirus disease-2019 (COVID-19) is milder in children, more data on pediatric chronic kidney disease (CKD) is needed. This study aimed to assess the incidence and severity of COVID-19 in pediatric CKD patients followed up at a tertiary center.
Material and methods: A questionnaire including demographics, COVID-19 history, symptoms, vaccination status was applied to patients with glomerular disease treated with immunosuppression, CKD stage 2–5, dialysis patients, and kidney transplant recipients followed up between March 2020-January 2022. Medical records were retrospectively reviewed. Acute kidney injury (AKI) was staged according to KDIGO criteria. COVID-19 was diagnosed by polymerase chain reaction in nasal swab samples and severity was categorized according to the National Institute of Health criteria.
Results: 220 patients were included, 48 were found (21.8%) to have experienced COVID-19. There was no significant difference regarding age, gender, underlying kidney disease, CKD stage, dialysis status, type or number of immunosuppressive medications, and glomerular filtration rate between patients with and without COVID-19 history. Mean age of COVID-19 positive patients was 14.04 ± 4.12 years. Most were infected

by a household member (43.8%) and during outpatient or inpatient care (18.8%). Four (8.3%) were asymptomatic, 43 (89.6%) had mild infection. Eleven (22.9%) patients were hospitalized, severe COVID-19 was observed in only one patient with kidney transplant who needed non-invasive mechanical ventilation. Eleven (22.9%) patients with COVID-19 were previously vaccinated (mostly two doses of BNT162b2 messenger RNA). Laboratory tests were available in 19 patients, AKI was detected in 4 (8.3%); as stage 1 in all. Median follow-up after COVID-19 was 4.6 (IQR;7.5) months. During follow-up one patient (with lupus nephritis) had re-infection, no patient developed multisystem inflammatory syndrome. All patients fully recovered, no renal flare or death was observed.

Conclusions: Although the vaccination rate was low in our cohort, majority of the children with COVID-19 showed a mild course. Along with the vaccination, general precautions seemed to be successful for this population.

PI-123 THE EFFECTS OF COVID-19 RESTRICTIONS IN CHILDREN WITH PRIMARY HYPERTENSION

Emre LeventoĞlu[1], Pelin PekÇetin ŞiŞik[2], İsmail Eray Çelik[2], Bahar BÜyÜkkaragÖz[1]

[1]Gazi University, Faculty Of Medicine, Department Of Pediatric Nephrology, [2]Gazi University, Faculty Of Medicine, Department Of Pediatrics

Introduction: Primary hypertension has been increasingly reported in parallel to the increase in the prevalence of obesity in children, both of which are important components of metabolic syndrome. The aim of this study is to investigate the effects of COVID-19 restrictions, which are believed to induce lifestyle changes and physical inactivity, on the parameters of metabolic syndrome in children with primary hypertension.

Material and methods: This is an observational, pre-post study conducted on pediatric patients with primary HT. The first phase of the study was the period prior to the state of alarm being put in place in Turkey, and the second phase was up to the date when the restrictions were cancelled. Anthropometric and blood pressure measurements, laboratory tests and hypertensive-mediated organ damage at the both phases of the study were compared.

Results: Severe restrictions due to COVID-19 pandemic were associated with an increase in body mass index (BMI) (26.4±7.3 vs. 27.2±7.1, p=0.002), antihypertensive drug use (n=53 (57.6%) vs. n=59 (64.1%), p=0.000), fasting blood glucose level (89.4±12.6 vs. 94.1±14.2, p=0.013) and a borderline elevation in total cholesterol (21 (22.8%) vs. 28 (30.4%), p=0.000). It negatively affected end organs; with increased frequency of interventricular septum hypertrophy (n=12 (13%) vs. n=17 (18.5%), p=0.031).

Conclusions: COVID-19 restrictions were associated with an increased risk for parameters associated with metabolic syndrome in patients with primary hypertension. Physicians should carefully monitor the weight, blood pressure, fasting plasma glucose level and total cholesterol levels in patients during restricted periods such as COVID-19 pandemic.

PI-124 ONE-YEAR FOLLOW-UP DATA OF ARTERIAL ABNORMALITIES IDENTIFIED IN KIDNEYS TRANSPLANTED INTO CHILDREN DURING THE FIRST COVID-19 PANDEMIC WAVE

Mathilde Grapin[1], Laureline Berteloot[2], Romain Berthaud[1], Sarah Temmam[3], Cecile Lozach[2], Marina Avramescu[1], Elisa Zanelli[2], Thomas Blanc[4], Carmen Capito[4], Christophe Chardot[4], Sabine Sarnacki[4], Nicolas Garcelon[1], Florence Lacaille[4], Marina Charbit[1], Myriam Pastural[1], Marion Rabant[1], Nathalie Boddaert[2], Marianne Leruez-ville[3], Marc Eloit[3], Isabelle Sermet-gaudelus[3], Laurene Dehoux[1], Olivia Boyer[1]

[1]Néphrologie Pédiatrique, Centre De Référence Marhea, Hôpital Necker Enfants Malades, Aphp, Institut Imagine, Université De Paris, Paris, France, [2]Imagerie Pédiatrique, Hôpital Necker Enfants Malades, Aphp, Institut Imagine, Université De Paris, Paris, France, [3]Laboratoire Découverte De Pathogènes, Institut Pasteur, Paris, France, [4]Chirurgie Pédiatrique, Hôpital Necker Enfants Malades, Aphp, Institut Imagine, Université De Paris, Paris, France

Introduction: Graft artery stenosis can have a significant short- and long-term negative impact on kidney graft function. We previously reported an unusual number of graft arterial anomalies following kidney transplantation (KTx) in children during the first COVID-19 pandemic wave (Berteloot et al. Am J Transplant21). We report herein the one-year follow-up of these patients.

Material and methods: In this retrospective study, we included all children who received a KTx at our center from February-July 2020.

Results: Among the 9 children who received a KTx at our center between February and July 2020 (8 boys, median age 10 years (3-17)), 8 presented Doppler features suggesting arterial stenosis, with an unusual extensive pattern (Fig.1) after a median delay of 13 days (8-113). For comparison, persistent spectral Doppler arterial anomalies were observed in only 5% of children following KTx at our center over the previous 5-year period, and were all focal anastomotic stenoses. In addition, five children had lymphoceles, which required surgical management as compared to only one patient in the 5 previous years (1%). We retrospectively diagnosed SARS-CoV-2 infection in 6/8 children with arterial stenosis on serologies performed at D0, including one boy with a history of positive RT-PCR 120 days before KTx. None of the patients had reported any symptom suggestive of COVID-19. The remaining 2 patients had received a graft from an asymptomatic deceased adolescent donor with a positive serology at D0. These data led us to suspect immune postviral graft vasculitis, triggered by SARS-CoV-2.

At one year post-transplantation, the outcome was favorable in the 8 isolated KTx recipients. 4/8 children had normal blood pressure and 4 had controlled high blood pressure on mono or bitherapy. Doppler anomalies had resolved in 5/8 and persisted in 3/8 with a trend for improvement of peak systolic velocities and no severe consequences on kidney function and histology. Indeed, the median GFR was 91ml/min/1.73m² (65-129), with unspecific and mild lesions on 4/8 protocol kidney biopsies (IFTA 1 or Cpt 1). One liver-kidney graft recipient had persistent hypertension and diffuse irregular inflammatory parietal thickening of the whole vascular graft associated with a parietal thrombus upstream of the birth of the 2 hepatic arteries

Conclusions: Our case series suggests a risk of postviral kidney graft vasculitis in children with a recent SARS-CoV-2 infection in the recipient or donor. Pre-transplant vaccination against COVID-19 is mandatory in children>5 years and their kidney donor candidates at our center. We also strongly recommend vaccination of all people aged >5 years in the household.

PI-125 COVİD-19 VACCINE-RELATED SIDE EFFECTS AMONG ADOLESCENTS WITH CHRONIC KIDNEY CONDITIONS: A SINGLE-CENTER EXPERIENCE

Demet Baltu, Eda Didem Kurt Sukur, Tugba Tastemel ÖztÜrk, Bora GÜlhan, Fatih Ozaltin, Ali Duzova, Rezan Topaloglu

Department Of Pediatric Nephrology, Faculty Of Medicine, Hacettepe University

Introduction: Considering the uncontrolled pandemic there is an urgent need for studies on the safety profile of the coronavirus disease-2019 (COVID-19) vaccination in children with chronic kidney conditions. Currently in Turkey COVID-19 vaccines are in use for the adolescent population. We aimed to investigate the side-effect and safety profile of COVİD-19 vaccines available for adolescents with chronic kidney disease (CKD) at our center.
Material and methods: The study population included patients with CKD stage 2–5, glomerular disease treated with immunosuppression, patients on dialysis, and kidney transplant recipients followed-up during the pandemic. A questionnaire including demographic and medical information, history of COVID-19 infection, vaccination status, and vaccine-related side effects was administered to the patients.
Results: Ninety eight patients (55 girls, 43 boys) were vaccinated by CoronaVac-inactivated SARS-CoV-2 (n=16) or BNT162b2 messenger RNA (mRNA) COVİD-19 (n= 82) vaccine. The mean age was 16.90 ± 2,36 years and median follow-up 4,9 (0,5-11,03) months. There were 36 stage 2-5 CKD, 8 dialysis, and 24 transplant patients in the cohort. The most common side effects were local pain (46,9 %), fatigue (17,3 %) and fever (11,2 %). No serious side effects were observed. Median duration of the symptoms was 2 (1-30) days. The longest symptom took 30 days; as dizziness in one patient with the BNT162b2 mRNA vaccine. No renal disease flare was observed post-vaccination and 11 (11,2 %) patients experienced mild COVID-19 infection (according to NIH criteria). Although side effects with mRNA seemed more frequent than the inactivated vaccine, it was statistically insignificant (p=0,10). No significant relationship was found between frequency of side effects and age, glomerular filtration rate, immunosuppressive treatments, CKD stage, and the underlying disease.
Conclusions: Although studies with longer follow-up are needed to evaluate the efficacy and side effects of COVID-19 vaccines, our early experience showed that vaccination is safe in the young population with CKD.

ELECTRONIC POSTERS

EP-1 SERUM CATHELICIDINE AND 25(OH) D VITAMIN LEVELS IN CHILDHOOD URINARY TRACT INFECTIONS

Alper ÇiÇek[1], Pelin Elibol[1], Banu İŞbilen BaŞok[2], Dilek Orbatu[3], Emel Berksoy[1], Demet Alaygut[4], Oya Baltali Hidir[3]

[1]University Of Health Sciences Tepecik Training And Research Hospital Department Of Emergency Medicine, [2]University Of Health Sciences Tepecik Training And Research Hospital Department Of Biochemistry, [3]University Of Health Sciences Tepecik Training And Research Hospital Department Of Pediatrics, [4]University Of Health Sciences Tepecik Training And Research Hospital Department Of Pediatric Nephrology

Introduction: Vitamin D stimulates the formation of cathelicidin, an essential antibacterial peptide found mostly in the urinary system. In this study, cathelicidine and vitamin D levels and the relationship between them were investigated in the differentiation of lower/upper urinary tract infection.
Material and methods: Pre-treatment the complete blood count, serum biochemistry, C-reactive protein, procalcitonin, 25-hydroxy vitamin D (ng/mL), and serum cathelicidin (ng/mL) of children aged 0 to 18 years who presented to the Pediatric Emergency Clinic with a urinary tract infection (UTI) were all measured. Demographic data (age, gender), urinary tract infection type (lower/upper), urine analysis results, and blood laboratory results were recorded. The control group consisted of healthy children whose blood samples were taken during routine control.
Results: The study included 72 patients, 36 patients, and 36 control groups in total. The mean age was 83.8±66.22 months, with forty patients (%55,6) female and 32 (%44.4) male. In terms of gender, there was no significant difference between the patient and control groups (p=0.343). The patient groups white blood cell count, neutrophil count, and C-reactive protein (CRP) levels were substantially greater than the control groups (p=0.05). In patients with lower and upper urinary tract infections, there was no significant difference in cathelicidin (5.7±3.7, 9.6±10.9; p=0.810) or vitamin D (23.3±9.5, 25.9±12.5; p=0.795) levels. In the control group, there was a positive correlation between vitamin D and cathelicidine levels (r:346, p=0.03). There was no significant difference in cathelicidin values in patients with upper urinary tract infection compared to the control group (p=0.054).
Conclusions: Although the difference was not statistically significant, serum cathelicidin levels were higher in patients with upper urinary tract infection than in the control group. Larger-scale studies may provide insight into whether cathelicidin can be used as a biomarker.

EP-2 COMPARISON OF CHILDREN WITH URINARY TRACT INFECTIONS HAVING NORMAL ULTRASONOGRAPHY/ NORMAL DMSA SCAN VERSUS NORMAL ULTRASONOGRAPHY/PATHOLOGICAL DMSA SCAN

Mehmet PektanÇ, Gizem Yildiz, Meral Torun Bayram, Alper Soylu, Salih Kavukcu

Dokuz EylÜl University Medical Faculty, Department Of Pediatric Nephrology, Izmir, Turkey

Introduction: Children with urinary tract infections (UTI) and abnormal ultrasonography (USG) are recommended to undergo voiding cystourethrography (VCUG) and DMSA scintigraphy. Yet, children with normal USG might still have abnormal scintigraphy. We aimed to compare the children with normal USG/normal scintigraphy with those having normal USG/abnormal scintigraphy.
Material and methods: Children with UTI and normal USG were grouped as normal vs pathologic scintigraphy. Two groups were compared for demographic, clinical, laboratory and imaging data. Patients with resolving vs persistent pathologic scintigraphy findings were also compared for the same parameters.
Results: 147 patients [122 (83%) female; mean age 62±47 months (1-203)] were enrolled. Serum creatinine was normal in all patients. Patients with pathologic scintigraphy (n=58) were older, had higher creatinine and erythrocyte sedimentation rate compared to those with normal DMSA (n=89). They also had higher frequency of increased C-reactive protein (CRP) level, vesicoureteral reflux (VUR), antibiotic prophylaxis and antireflux surgery. VUR was the most significant parameter predicting pathological scintigraphy. Patients with persistent pathological scintigraphy (n=30) had lower BMI SDS, lower urine specific gravity and higher CRP compared to those with resolving scintigraphy findings (n=9). VCUG was performed in 30 out of 46 children under 2 years of age and 19 of them (63%) had VUR. While DMSA scan was pathologic in 6 (32%) of those with VUR, only 1 of 11 (9%) children without VUR had pathological DMSA scan.
Conclusions: VUR and high CRP increase the probability of pathologic DMSA in children presenting with UTI and normal USG. Thus, ordering DMSA in children with UTI despite normal USG is feasible if CRP is increased at presentation. Presence of VUR and pathologic DMSA in 2/3 and 1/3 of children <2 years of age, respectively, contradicts the American Academy of Pediatricss suggestion that "if urinary system ultrasonography is normal in children under 2 years of age with UTI, no additional examination is required."

EP-3 ROLE OF CELL-FREE HEMOGLOBIN IN APOLIPOPROTEIN L1-MEDIATED SICKLE CELL NEPHROPATHY

Oyindamola C. Adebayo[1], Cheng Cheng[2], Agathe B. Nkoy[2], Pepe M. Ekulu[3], Lambertus P. Van Den Heuvel[2], Elena Levtchenko[2], Veerle Labarque[1]

[1]Centre For Molecular And Vascular Biology, Department Of Cardiovascular Sciences, Katholieke Universiteit Leuven, Leuven, Belgium, [2]Department Of Development And Regeneration, Katholieke Universiteit Leuven, Leuven, Belgium, [3]Division Of Nephrology, Department Of Paediatrics, University Hospital Of Kinshasa, Faculty Of Medicine, University Of Kinshasa, Democratic Republic Of Congo.

Introduction: Apolipoprotein L1 (*APOL1*) risk variants (G1 and G2) have been reported to be associated with increased risk of sickle cell nephropathy (SCN) in African population. APOL1 risk variants are associated with hemoglobinuria, albuminuria and hyperfiltration in sickle cell disease (SCD), but the underlying mechanism remains unknown. We hypothesized that cell-free hemoglobin (Hb) released during hemolysis in patients with SCD is a second-hit factor for APOL1-mediated SCN.
Material and methods: Conditionally immortalized podocytes (ciPodocytes) expressing different genotypes of APOL1 were stimulated with sickle hemoglobin (Hb S) (0–500 μg/ml, corresponding to 0–30 μM of heme molar equivalents) or heme (0–30 μM) for different time periods. Subsequently, the effect of HbS or heme on ciPodocytes were assessed via Resazurin cell viability assay and real-time quantitative PCR (RT-qPCR).
Results: Exposure of ciPodocytes to at least 100 μg/ml of cell-free HbS or 12 μM of heme for 24 hours causes a significant decreased in cell viability. This decrease in cell viability was independent on the APOL1 genotype of the podocytes. APOL1 G0/G0 ciPodocyte cell line also showed a time and concentration dependent upregulation of APOL1 mRNA.
Conclusions: This study provides proof that cell-free Hb or heme at a concentration seen in vivo in patients with SCD can be toxic to the podocytes and can lead to higher expression of APOL1.

EP-4 INTERLEUKIN-10 IN THE NEONATAL MOUSE MODEL OF OBSTRUCTIVE UROPATHY

Maja Wyczanska[1], Ursula Keller[1], Barbara Schraml[2], Bärbel Lange-sperandio[1]

[1]Dr. V. Hauner Children's Hospital, Lmu Munich, [2]Institute Of Cardiovascular Physiology And Pathophysiology, Biomedical Center (bmc), Lmu Munich

Introduction: Urinary tract obstruction during renal development leads to inflammation, leukocyte infiltration, tubular cell death and interstitial fibrosis. Interleukin-10 (IL-10) is an anti-inflammatory cytokine, produced mainly by monocytes/macrophages and regulatory T-cells. IL-10 inhibits the innate and adaptive immune response. A protective, anti-inflammatory and antifibrotic effect of IL-10 after unilateral ureteral obstruction (UUO) was shown in the adult mouse kidney. We studied the role of IL-10 in the neonatal mouse kidney with UUO.
Material and methods: Newborn transgenic mice (IL-$10^{-/-}$) and C57BL/6 wildtype-mice (WT) were subjected to either UUO or sham operation at day 2 of life. Whole kidneys were harvested at day 3, 7, and 14 of life. The kidneys were analyzed by immunohistochemistry (inflammation (F4/80, CD3), cell death (TUNEL, PAS), fibrosis (MT, collagen I, α-SMA)), by Westernblot (PARP, caspase-8, RIPK3 (necroptosis), GSDME (pyroptosis), α-SMA, vimentin, TGF-ß, E-cadherin, β-catenin) and by FACS-analysis (leukocyte subpopulations: B-cells, $CD11b^{hi}$, cDC1, cDC2, $Ly6C^{+}$, $MHCII^{+}F4/80^{hi}$, $MHCII^{-}F4/80^{hi}$, neutrophils, monocytes, T-cells).
Results: UUO induced a continuous increase in leukocyte infiltration with $Ly6C^{+}$ inflammatory monocytes, $CD11b^{hi}$ macrophages (d14), cDC1 (dendritic cells) (d14), neutrophils and T-cells (d14) in the neonatal kidney. IL-$10^{-/-}$ mice showed reduced infiltration of $CD11b^{hi}$, $Ly6C^{+}$, cDC1 and T-cells, and increased infiltration of neutrophils after UUO compared to WT. As an indicator for apoptosis, PARP expression and caspase-8 cleavage increased after UUO. IL-$10^{-/-}$ mice with UUO exhibited a decrease in necroptosis (RIPK3/RIP3) but no difference in apoptosis (PARP and caspase-8), pyroptosis (GSDME) and fibrosis (α-SMA, vimentin, E-cadherin, β-catenin) compared to WT. Expression of TGF-β increased in WT-mice with UUO at d14 and was less in pronounced in IL-$10^{-/-}$ mice.
Conclusions: Contrary to our expectations, IL-$10^{-/-}$ mice demonstrated no increase in cell death, inflammation, or profibrotic cytokines in comparison to WT following UUO. These results suggest that IL-10 signaling plays a minor role in tuning down the inflammatory response following UUO in the neonatal kidney than in the adult kidney.

EP-5 SAFETY AND EFFICAY OF AVACOPAN (CCX168) IN A PEDIATRIC PATIENT WITH C3 GLOMERULOPATHY

Federica Zotta, Marco Busutti, Andrea Cappoli, Ines Lerario, Antonio Gargiulo, Francesco Emma, Marina Vivarelli

Irccs, Division Of Nephrology And Dialysis Bambin Gesu Pediatric Hospital, Rome Italy

Introduction: C3 glomerulonephritis (C3GN) is a subtype of C3 glomerulopathy, characterized by the alternative complement activation and by dominant C3 immunofluorescence .
Avacopan, called CCX168, is an orally administered small-molecule C5aR antagonist that blocks the effects of C5a, which is one of the most potent pro-inflammatory mediators of the complement system.
Results: CASE REPORT
An 11-yr old girl with a biopsy-proven C3GN, was initially treated with three IV boli of methyllprednisolone then tapered to oral prednisone (PDN) associated with mycophenolate mofetil (MMF) and ACE inhibitor. In the following months, cyclosporine (CyA) was added to the therapy due to the relapse of proteinuria (UPCR 1.19 mg/mg).
The patient never achieved complete remission and therefore was enrolled in the ChemoCentryx CL011_168 study.
In the first 26 weeks, during the double-blind period the girl developed pneumoniae with a significant increase in proteinuria.
At the beginning of the open-label phase of the trial UPCR was 2.09 mg/mg. Following start of avacopan, a progressive reduction of proteinuria was observed reaching values around 0.5 mg/mg. In the last 4 weeks of the study, avacopan was discontinued, with subsequent proteinuria increase to > 1 mg/mg. The patient also reported increased fatigue.
After about 3 months, authorization for a compassionate use of avacopan was obtained with improvement in physical well-being and reduction of proteinuria to around 0.5 mg/mg. CyA was discontinued but it was rapidly reintroduced due to a transient increase of proteinuria. In the following months, proteinuria remained low despite the interruption of MMF. At the last follow up, UPCR was 0.29 mg/mg and renal function was normal. The drug was well tolerated.
Conclusions: To the best of our knowledge, this is the first report on the safety and efficacy of avacopan in a pediatric case of C3GN.

EP-6 UNDERDIAGNOSIS OF ACUTE KIDNEY INJURY IN CRITICALLY ILL CHILDREN

Patrícia Costa Reis[1], Beatriz Nicolau[3], Paulo Jorge Nicola[4], Cristina Camilo[2], Rosário Stone[1], Marisa Vieira[2]

[1]Pediatric Nephrology And Kidney Transplantation Unit, Pediatrics Department, Hospital De Santa Maria, Lisbon, Portugal, [2]Pediatric Intensive Care Unit, Pediatrics Department, Hospital De Santa Maria, Lisbon, Portugal, [3]Clínica Universitária De Pediatria, Faculdade De Medicina, Universidade De Lisboa, Lisbon, Portugal, [4]Unidade De Epidemiologia, Instituto De Medicina Preventiva E Saúde Pública, Faculdade De Medicina, Lisbon, Portugal

Introduction: Acute kidney injury is common in critically ill children and it is frequently associated with important long-term sequelae. Recognition of AKI and referral to a pediatric nephrology clinic are important to improve the outcome of these patients. Our goals were to study the incidence of AKI in a pediatric intensive care unit (PICU), whether the team was recognizing AKI and if the patients were being referred for follow-up.
Material and methods: Retrospective study of all the patients admitted to a PICU in an academic hospital during 6 months (Jan-Jun 2021). Newborns, children with previous chronic kidney disease and readmissions were excluded. AKI was defined according to Kidney Disease: Improving Global Outcomes (KDIGO).
Results: We studied 135 patients (median age 8 years old; 50% males; 75% with chronic diseases; 61% post-surgery). AKI was identified in 61 patients (45%): 44% mild; 49% moderate and 7% severe. Five patients (8%) had creatinine elevation, 33 (54%) had oliguria and 23 (38%) had both criteria. The admission diagnosis was paediatric inflammatory multisystem syndrome temporally associated with SARS-CoV-2 (PIMS-TS) in 13 patients and 10 (77%) developed AKI. PIMS-TS was a risk factor for AKI ($p<0.05$). Mechanical ventilation was more frequent in patients with AKI (34% vs 5%; $p<0.0001$). Patients with AKI had higher average hospital stay ($p<0.0001$) and higher mortality (7% vs 0%; $p<0.05$). Only 15 patients (25%) had AKI diagnosis at the PICU discharge note. There was only one referral to follow-up at the Pediatric Nephrology clinic of a patient with severe AKI.
Conclusions: In this cohort of critically ill children, AKI was frequent and it was associated with a higher hospital stay and death. Notably, AKI was underdiagnosed and pediatric nephrologists rarely followed these patients. New strategies will be put into practice to improve the care of these children.

EP-7 DO CYP POLYMORPHISMS AFFECT PHARMACOKINETICS OF TACROLIMUS IN CHILDREN WITH NEPHROTIC SYNDROME?

Zainab Arslan[1], Dalvir Kular[2], Carmen Bugarin_diz[2], Ania Koziell[2]

[1]Great Ormond Street Hospital For Children, London, Uk, [2]Immunology And Microbial Sciences And Genetics And Molecular Medicine, Kings College London, Uk

Introduction: Tacrolimus is a well-established immunosuppressive agent used to achieve disease control in nephrotic syndrome (NS) in children. CYP3A genes are primarily responsible for tacrolimus metabolism, with the differences in CYP3A5 genotype reported between different ethnic groups considered a key reason for the observed variation in its effects. The aim of this study was to correlate CYP3A5 genotypes with reported ethnicity, tacrolimus dose requirements with side effects experienced and degree of nephrotoxicity seen on renal biopsy.
Material and methods: Analysis of all children with NS, at a single tertiary paediatric nephrology centre, who had whole genome sequencing (WGS) performed, was undertaken. CYP3A5 variants were then annotated and divided into "slow", "intermediate" and "fast" metabolising groups based on genotype. Patient demographics and clinical data were collected from electronic patient records.
Results: Of the 51 children included in the study, 35 (69%) were slow metabolisers, 10 (20%) intermediate and 6 (11%) fast metabolisers of tacrolimus. Slow metabolisers were primarily from a European (EUR) background (69%) and required low doses of tacrolimus with 25% experiencing tacrolimus associated side effects and 17% experiencing tacrolimus associated nephrotoxicity on renal biopsy. Intermediate metabolisers included 30% EUR and 20% of African ethnicity (AFR) and required intermediate doses of tacrolimus. 30% of these had compliance associated raised creatinine but none experienced tacrolimus induced nephrotoxicity on renal biopsy. The fast metaboliser group included patients mainly from an African (67%) background (33% from South-Asian ethnicity) and all required high tacrolimus doses. Transient neutropenia was the only side effect reported. No tacrolimus induced nephrotoxicity was seen on renal biopsy in this cohort.
Conclusions: This study is the first to correlate CYP3A5 genotype with ethnicity, tacrolimus dose requirement and side effects including nephrotoxicity based on histological findings in childhood onset NS and provides useful insights into dosing according to metabolising status. We recommend routine use of CYP3A5 genotyping to allow individualising dosages for children receiving tacrolimus.

EP-8 GENETIC CAUSES OF STEROID-RESISTANT NEPHROTIC SYNDROME IN RUSSIAN CHILDREN: RESULTS OF THE MONOCENTER COHORT STUDY

Anastasiia Milovanova, Petr Ananin, Alexander Pushkov, Kirill Savostyanov, Tatiana Vashurina, Olga Zrobok, Olga Komarova, Alla Ryaposova, Svetlana Dmitrienko, Andrey Fisenko, Alexey Tsygin

National Medical Research Center Of Childrens Health

Introduction: Genetic causes of nephrotic syndrome are not thoroughly studied, and nowadays, including more populations in these researches can complete a picture of the mutation spectrum characterizing certain regions.
Material and methods: Up to date, we examined 220 children with primary nephrotic syndrome (NS), including congenital and infantile ones. All children underwent a molecular genetic examination by the new generation sequencing (NGS) of target regions of 200 genes associated with hereditary kidney diseases.
Results: As a result of our study, 142 nucleotide variants were identified in 31 genes, causative mutations were found in 61.3% of cases.Nucleotide variants in the NPHS2 gene were detected in 27 children (12.2%), COL4A5 - in 18 children (8.1%), WT1 - in 17 children (7.7%), mutations in other genes were found less than 5% each. We verified the major mutations prevailing in Russian children: c.259G>T, p.E87*, c.868G>A, p.V290M, and c.686G>A, p.R229Q in NPHS2 gene. The first variant occurred in 14 (51.9%) children; 10 were in the homozygous state. Two others we identified in 6 children each. The most common nucleotide variant in the NPHS1 gene was the nonsense mutation c.3478C>T, p.R1160*, which occurred in the genomes of 3 children. During the follow-up period, 56 children (25.5%) reached CKD stage 3, and 37 children (16.8%) - CKD stage 5. We didnt reveal any gene with a greater likelihood of decreased kidney function.
Conclusions: Based on a voluminous monocenter study, a genetic characteristic of Russian children is given for the first time, reflecting the spectrum and frequencies of nucleotide variants that cause NS.

EP-9 HEMOLYTIC UREMIC SYNDROME (SP-HUS) ASSOCIATED WITH STREPTOCOCCUS PNEUMONIAE. THE

ROLE OF COMPLEMENT IN A RARE PRESENTATION OF THE DISEASE

Ana Cristina Aguilar Rodríguez, Pedro Arango Sancho, Elena Codina Sampera, Yolanda Calzada Baños, Marta Jiménez Moreno, Raquel Jiménez García, Álvaro Madrid Aris

Hospital Sant Joan De Déu

Introduction: Hemolytic uremic syndrome (HUS) is an acute clinical picture characterized by endothelial damage secondary to various causes that leads to thrombotic microangiopathy (TMA) and progressive decline in renal function.There are several underlying causes,including a very rare but serious one caused by invasive Streptococcus pneumoniae (SN) infection that is associated with a worse clinical course and higher mortality.This presentation is on average in younger patients compared to typical HUS.Our objective is to present a case of Sp-HUS with an atypical evolution and resolution, associated with alterations in complement regulatory proteins

Material and methods: We present the case of a 5-year-old boy with no pathological history of interest who at 20 months of age presented a picture of acute otitis media treated with amoxicillin-clavulanic acid without improvement and an unfortunate clinical course leading to a worsening of the general condition and a hemodynamic instability.In the emergency department,a chest x-ray was performed, showing complicated pneumonia that required pleural drainage,along with blood tests showing anemia,thrombocytopenia,coombs+ test, and positive PCR for SN.These results being compatible with a diagnosis of Sp-HUS

Results: From that moment,treatment was started with ceftriaxone,transfusion therapy,bicarbonate supplementation and antihypertensive therapy. During hospitalization,he developed anuric acute renal failure that required 12 sessions of renal replacement therapy,beginning with spontaneous diuresis on day 15 of his evolution,with subsequent recovery of renal function until reaching an estimated GFR by creatinine after 2 years of follow up of 87 ml/min/1.73 m2, as well as complete normalization of proteinuria.The renal biopsy performed at the time of the symptoms showed no signs compatible with TMA or signs of chronicity.Genetic study carried out subsequently detected an alteration in the sialization of factor H (FH) probably due to the pneumococcus and responsible for the clinical picture of our patient

Conclusions: Sp-HUS has a more severe initial behavior with a longer duration of oliguria and thrombocytopenia or extrarenal manifestations (hearing loss),requiring renal replacement therapy in around 70-80%.aHUS risk variants in the CFH-CFHR3-CFHR1 region could contribute to disease predisposition to Sp-HUS.Transient desalination of complement FH by pneumococcal neuraminidase may play a role in disease pathogenesis

EP-10 TREATMENT AND OUTCOME OF ANTI-FACTOR H AUTO-ANTIBODY-ASSOCIATED AHUS IN CHILDREN

Marion Ferri[1], Federica Zotta[2], Claire Dossier[1], Andrea Pasini[3], Veronique Fremeaux-bacchi[4], Julien Hogan[1], Marina Vivarelli[2]

[1]Pediatric Nephrology, Robert Debre Hospital, Aphp, Paris, France, [2]Division Of Nephrology And Dialysis, Irccs Ospedale Pediatrico Bambino Gesu, Rome, Italy, [3]Pediatric Nephrology, Azienda Ospedaliero Universitaria Santorsola Malpighi, Bologna, Italy, [4]Laboratory Of Immunology, Inserm Umrs 1138, Hopital Europeen Georges Pompidou, Aphp, Paris, France

Introduction: Guidelines suggest combining plasma exchange (PEX) and immunosuppressive drugs to treat anti-CFH HUS. Several questions remain including the use of eculizumab (ECZ), the interest of associating rituximab and the timing of treatment withdrawal.

Material and methods: We reviewed 9 cases of pediatric anti-CFH HUS treated in Italy and France. Treatment consisted in PEX or ECZ followed by mycophenolate mofetyl (MMF) as maintenance treatment in severe cases in Italy and ECZ (+/- Ig-Immunoadsorption (Ig-IA)) and Rituximab (RTX) followed by steroids and MMF as maintenance treatment in France.

Results: Median age at onset was 4,3[3,0;5,3] and median GFR was 17[15;22]ml/min/1,73m². All patients presented manifestations of TMA and 3 extrarenal manifestations. Median anti-CFH antibodies levels was 8129[17654;23188]UA/mL. Treatment consisted in ECZ in 7 patients combined with PEX/Ig-IA in 2 patients and 1 patient received PEX only. RTX was added in 3 patients Median duration of ECZ treatment was 24 months and MMF was added in 5 patients. Anti-CFH ab levels decreased in all but one patient with no clear difference between treatment groups. Ab levels decreased spontaneously in patients without maintenance therapy.

All patients rapidly improved with all but one patient recovering a normal kidney function (median time to normalization 1,4[0,5;2,2]months) and all reaching hematological remission in a median time of 2,0[1,0;2,3] months.

At last follow-up, 4 patients were under treatment (2ECZ, 2 MMF) and 5 were off treatment without experiencing relapse. Anti-CFH levels at treatment withdrawal ranged from 0 to 1645UA/mL.

Conclusions: ECZ is effective to achieve remission in anti-CFH HUS. MMF and/or RTX seem effective in decreasing anti-CFH ab levels but spontaneous decrease is seen in some patients. All treatment may be safely withdrawn in most patients, though the timing and the risk of relapse are unknown.

EP-11 ROLE OF NON-DONOR SPECIFIC ANTIBODIES IN PEDIATRIC KIDNEY TRANSPLANTATION

Maria Sangermano[1], Susanna Negrisolo[2], Andrea Carraro[2], Germana Longo[1], Davide Meneghesso[1], Mattia Parolin[1], Elisa Benetti[1]

[1]Pediatric Nephrology Dialysis And Transplant Unit, Department Of Women's And Children's Health, Padua University Hospital, Padua, Italy,, [2]Laboratory Of Immunopathology And Molecular Biology Of The Kidney, Department Of Women's And Children's Health, Padua University Hospital, Padua, Italy,

Introduction: Late allograft failure remains a considerable problem in renal transplantation. While the role of donor specific anti-HLA antibodies (DSA) in the pathogenesis of allograft damage has been largely demonstrated, the role of non-donor specific antibodies (NDSA) is still controversial. This study was aimed to evaluate the occurrence of NDSA in pediatric renal transplant recipients and their correlation with clinical outcomes.

Material and methods: We retrospectively analyzed 52 pediatric renal transplant recipients undergone to anti-HLA antibodies monitoring between 2015 and 2018. Antibodies were measured out 6, 12 and 24 months after transplantation. Protocol biopsies were performed at the same timeline. Collected data included creatinine, eGFR, proteinuria, immunosuppressive therapy, viral infections, rejections. Patients were divided into 4 groups: without antibodies (NA), with NDSA only (NDSA), with both DSA and NDSA (DSA+NDSA), and with DSA only (DSA).

Results: All groups had similar demographic and clinical characteristics. Occurrence of DSA and NDSA was similar (15% of patients) 6 months after-transplantation (PO), while 8% had both DSA and NDSA. 12 months PO, 19% of patients had DSA, 21% NDSA and 12% both. 24 months PO, 15% had DSA, 11% NDSA and 10% both. Protocol biopsies

showed subclinical rejection (acute or chronic) in 23% of patients at 6 months PO (5% antibody mediated rejection (AMR) and 75% T-cell mediated rejection (TCMR), in 25% at 12 months (23% AMR, 77% TMCR) and in 30% at 24 months (25% AMR, 75% TCMR). Statistical analysis showed no significant correlation between NDSA only and rejection, but NDSA seemed to play synergistic action with DSA in AMR. Compared to others, NDSA children had worse eGFR and higher proteinuria (p=0.02), as well as DSA+NDSA group (p=0.026).
Conclusions: NDSA do not seem to cause rejection per se, however they play a synergistic action with DSA in AMR. Proteinuria is significantly higher in patients with NDSA compared to other groups, suggesting a contribution in allograft damage. Our results suggest that NDSA should be considered as a wake-up call for graft outcome and their regular monitoring may be a useful tool in clinical practice.

EP-12 COEXISTENCE OF POST-COVID-19 MULTISYSTEM INFLAMMATORY SYNDROME AND THROMBOTIC MICROANGIOPATHY IN THREE CHILDREN

H.gozde Onal[1], Hulya Nalcacioglu[1], Burcu Bozkaya Yucel[2], Demet Tekcan Karali[1], Emine Hafize Erdeniz[3], Ozlem Aydog[1]

[1]Ondokuz Mayis University Faculty Of Medicine, Pediatric Nephrology Department, Samsun, Turkey, [2]Ondokuz Mayis University Faculty Of Medicine Pediatric Rheumatology Department, Samsun, Turkey, [3]Ondokuz Mayis University Faculty Of Medicine, Pediatric Infectious Disease Department, Samsun, Turkey

Introduction: Severe acute respiratory syndrome coronavirus 2 (SARS-CoV2) infection affects every organ, including the kidney, ranging from an asymptomatic state to critical illness as well as a post-inflammatory response. Multisystem Inflammatory Syndrome in Children (MIS-C) characterized by fever and organ dysfunction in the setting of recent COVID-19 infection. The cause of kidney involvement in COVID 19 is multifactorial, including acute tubular injury, direct renal cell invasion, immune dysregulation with systemic cytokine activation, and thrombotic microangiopathy (TMA).
Material and methods: We presented three patients with MIS-C and Thrombotic Microangiopathy (TMA) related to COVID-19.
Results: Three of our patients' presented with persistent fever, diarrhea, nausea, and vomiting. A high level of clinical suspicion for MIS-C was supported by laboratory findings (elevated ESR, CRP, D-dimers, and fibrinogen) along with positive IgG SARS-CoV-2 antibodies. In the third patient, both PCR PCR test for SARS-CoV-2 and IgG SARS-CoV-2 antibodies was positive. Three patients received intravenous immunoglobulin (IVIG) (2 g/kg), intravenous pulse methylprednisolone (10-30 mg/kg/day) for 3-6 days, followed by oral prednisolone treatment (2 mg/kg/day). Two patients had signs of TMA (non-immune hemolytic anemia, thrombocytopenia, and acute kidney injury) with hyper inflammation at the time of admission and received renal replacement therapy. On the fourth day of admission, the third patient developed oliguria, anemia, and thrombocytopenia compatible with the hemolytic uremic syndrome. Their coagulation tests, ADAMTS 13 activity, was normal and serum complement level 3 was low in two patients and normal range in one patient. Eculizumab was administered to the first and second cases who had no response to plasmapheresis, a persistent anuric condition, after the second dose of Eculizumab, clinical improvement, including down-trend of the hemolytic episode and increase in urine output. Despite improved kidney function, persistently high levels of ferritin, LDH, and ongoing decline of platelet count, anakinra was added to the treatment in one patient. At the follow-up, three patients had normal kidney functions.
Conclusions: Herein, we presented three patients with MIS-C and hemolytic uremic syndrome. We must be aware of the unusual implications of COVID-19-related illness to early diagnosis and treatment.

EP-13 LIPOSOMAL AMPHOTERICIN B NEPHROTOXICITY IN CHILDREN: A CROSS-SECTIONAL ANALYSIS

Alexandra Andrade[1], Sara GonÇalves Dias[2], Ana Araújo Carvalho[3], Rute Baeta Baptista[4], Telma Francisco[4], Margarida Abranches[4]

[1]Department Of Paediatrics, Hospital Central Do Funchal, Funchal, Portugal, [2]Department Of Paediatrics, Hospital Divino Espírito Santo, São Miguel, Portugal, [3]Department Of Paediatrics, Chulc, Lisbon, Portugal, [4]Paediatric Nephrology Unit, Department Of Paediatrics, Chulc, Lisbon, Portugal

Introduction: Nephrotoxicity is a common and potentially severe adverse effect associated with liposomal amphotericin B treatment.
Material and methods: We performed a cross-sectional analysis of data from all paediatric patients treated with liposomal amphotericin B in our centre from january/2017 to december/2021. We aimed to analyse the incidence of nephrotoxicity. The composite renal outcome was defined by the occurrence of AKI (creatinine rise >1.5x baseline) or tubulopathy (need for electrolyte replacement, glycosuria or polyuria).
Results: A total of 76 children were included, with a median age of 3.8 years (P25-P75: 1.2-10.3) and 50% were male. Median treatment duration was 10 days (P25-P75: 6-16). During the course of treatment, 17 (22%) patients died and 6 (8%) had to withdrawal treatment due to adverse effects. The composite renal outcome was achieved in 68 (89%) patients, with AKI occurring in 36 (47%) and tubulopathy in 58 (83%). Serum creatinine increased in 54 (71%) patients, rising 0.14 mg/dL (P25-P75: 0.05-0.5; range 0.01-2.28) or 44% (P25-P75: 16-168) above baseline. Peak creatinine was reached by day 7 (P25-P75 3-10; range 0-21) and returned to baseline by day 13 (P25-75: 6-16; range 0-32). Among the 56 (75%) who needed electrolyte replacement, maximum doses were 3.5 mEq of KCl/Kg/day (P25-P75: 2.0-5.4) and 5.5 mEq of NaCl/Kg/day (P25-P75: 3.0-13.0). Minimum serum potassium levels were significantly higher in patients with AKI (3.0 [P25-75: 2.9-3.1] versus 2.7 [2.5-3.0], p-value 0.04). No other significant differences were found. The presence of comorbidities was a significant predictor of tubulopathy in a logistic regression model adjusted for age, sex, and the severity of AKI (OR 5.9, CI95% 1.4-24.7, p-value 0.015).
Conclusions: Amphotericin B adverse effects led to treatment withdrawal in 8% of our cohort. Nephrotoxicity occurred in 89% of our patients and 75% needed electrolyte replacement. These data emphasize the need for serial clinical and laboratorial monitoring during treatment.

EP-14 SYSTOLIC BLOOD PRESSURE DIPPING AS A PREDICTOR OF CKD PROGRESSION IN CHILDREN

Anna Deja[2], Piotr Skrzypczyk[1], Beata Leszczynska[1], Maria Daniel[1], Malgorzata Panczyk-tomaszewska[1]

[1]Department Of Pediatrics And Nephrology, Medical University Of Warsaw, [2]Department Of Pediatrics And Nephrology, Doctoral School, Medical University Of Warsaw

Introduction: Elevated MAP 24h in ABPM and proteinuria are well-established risk factors for progression of CKD in children with scarce data on significance of other ABPM parameters. The study aimed to analyze risk factors for CKD progression with emphasis on detailed ABPM data.
Material and methods: In 55 children (38 boys, 17 girls) with CKD II–V observed for at least one year or until initiation of renal replacement therapy, we analyzed ABPM (blood pressure during 24h, activity, resting period, blood pressure loads and dipping), clinical and biochemical parameters.

Results: Causes of CKD were: CAKUT in 22, hereditary disorders in 12, glomerular kidney diseases in 7, other in 14. 38 (69.1%) patients had arterial hypertension, and 18 (32.7%) had proteinuria. At the beginning of observation eGFR was 66 (IQR: 42.8–75.3)mL/min/1.73m^2, the observation period was 27 (16–36) months. Mean annual eGFR decline was 2.9±5.7mL/min/1.73m^2/year. eGFR decline correlated with age (r=0.30, p=0.026), diuretic use (r=0.27, p=0.047), initial proteinuria (r=0.34, p=0.013), nighttime systolic and mean blood pressure (r=0.27, p=0.045 and r=0.29, p=0.032) and systolic and diastolic blood pressure dipping (r=-0.37, p=0.006, r=-0.29, p=0.034, respectively). In Kaplan-Meier analysis risk factors for 1 grade CKD progression were presence of proteinuria (p=0.031) and arterial hypertension (p=0.018). There was no relation between MAP 24h Z-score and rate of GFR decline (p=0.739). There was no difference in GFR decline between those with MAP 24h below and above 50th percentile (p=0.992). In multivariate analysis (general regression model), systolic blood pressure dipping (beta=-0.43, p<0.001), presence of proteinuria (beta=-0.35, p=0.004), and age (beta=0.25, p=0.038) were the only predictors of GFR decline.
Conclusions: 1. Targeting blood pressure to MAP 24h below 50th percentile might not be the best suited way to slow progression of CKD in all populations
2. Systolic blood pressure dipping may be valuable indicator of CKD progression in children.

EP-15 THE ROLE OF CELLULAR METABOLISM ON CELL ADHESION IN CYSTINOSIS PODOCYTES: A POSSIBLE THERAPEUTIC STRATEGY?

Sante Princiero Berlingerio[1], Sarah Tassinari[2], Tjessa Bondue[1], Marc Fransen[1], Benedetta Bussolati[2], Lambertus Van Den Heuvel[1], Elena Levtchenko[1]

[1]Ku Leuven, [2]University Of Torino

Introduction: Cystinosis is a rare, incurable autosomal recessive storage kidney disease caused by mutations in CTNS gene, which encodes the cystine transporter cystinosin and leads to lysosomal cystine accumulation in all the body. In addition to proximal tubular cells, cystinosis also affects the glomerulus since podocytes are lost into urine leading to proteinuria and kidney failure. Cysteamine, the current treatment, decrease cystine accumulation but does not reverse proximal tubular injury (renal Fanconi Syndrome) neither glomerular injury. These evidences suggest that different mechanisms are involved and further studies are necessary to understand the disease in order to develop new therapeutic options.
Material and methods: Immortalized patient-derived cystinosis and healthy podocytes were used and the results were validated in our in-house developed cystinosis zebrafish model. To study the altered metabolic pathways, metabolomic analysis (LC-MS), flow cytometry, RT-qPCR, western blot, chemical and redox-sensing fluorescent probes were used.
Results: Cystinotic podocytes present a peculiar cellular metabolism, characterized by impaired glycolytic and TCA cycle, increased ROS levels and cell detachment. Interestingly, the latter can be rescued in vitro and in vivo with targeted treatment.
Conclusions: An impaired metabolism is a critical feature in podocytes and it elucidates the importance to investigate more targeted therapies in combination with the standard of care cysteamine.

EP-16 BARTTER SYNDROME- STILL A DIAGNOSTIC CHALLENGE

Maša Davidović, Ivanka Kos, Ivan Jakopčić, Hana Matković, Maja Ban, Lovro Lamot, Kristina Vrljičak

Division Of Nephrology, Dialysis And Transplantation, Department Of Pediatrics, University Hospital Center Zagreb, University Of Zagreb School Of Medicine, Zagreb, Croatia

Introduction: Bartter syndrome is an inherited tubulopathy that presents in infancy or childhood with metabolic alkalosis, hypokalemia and normal blood pressure. There are 4 types of Bartter syndrome, with types I, II and IV being more severe, and type III (classic form) being a milder form with a variable presentation, sometimes mimicking other tubulopathies. Therefore, Bartter syndrome should be a permanent differential diagnosis in children with various tublopathies.
Material and methods: Case report.
Results: We report a case of a 10-year-old boy who was followed by a pediatric nephrologyst from infancy because of incomplete diabetes insipidus. During that time his electrolytes and blood pressure were normal. At the age of 7 hypercalciuria and hyperreninemia were noted for the first time, as well as hyperparathyroidism and osteopenia. Serial renal ultrasounds were normal. Later on, hypokalemia was also noted. His diagnose was then subjected to reevaluation. Because of severe hypercalciuria, CT urography was done and revealed multiple urolites. Genetic analysis was then finally preformed (Blueprint Nephrolitiasis panel with 35 genes) identifying a novel hemizygous frameshift variant c.38_59delinsAGTCAC, p. (Gly13Glufs*22) and a heterozygous deletion chr1:g.16351202_16372237del which encompasses exons of both CLCNKB and CLCKNA genes. The diagnosis of Bartter syndrome was thereby confirmed.
Conclusions: Type III Bartter syndrome (classic Bartter syndrome) has a very variable presentation, sometimes overlapping with other tubulopathies. It is caused by loss-of-function mutations in CLCNKB gene encoding the voltage-gated chloride channel CICKb, which is expressed in the thick ascending limb of the loop of Henli, but also in the distal convulated tubule, possibly explaining the variable features. While recognizing atypical presentations is still the first step towards the correct diagnosis, modern targeted clinical panels ease the way. Discovering novel mutations is important because of better understanding of molecular mechanisms as well as targeted treatment for the individual patient.

EP-17 RELATION BETWEEN OBESITY-RELATED COMORBIDITIES AND KIDNEY FUNCTION IN CHILDREN

Mark J.c.m. Van Dam[1], Hans Pottel[2], Anita C.e. Vreugdenhil[1]

[1]Centre For Overweight Adolescent And Children's Healthcare (coach), Department Of Pediatrics, School Of Nutrition And Translational Research In Metabolism (nutrim), Maastricht University Medical Centre +, Maastricht, The Netherlands, [2]Department Of Public Health And Primary Care, Ku Leuven Campus Kulak Kortrijk, Kortrijk, Belgium

Introduction: While the association between obesity in adults and kidney disease is well-established, studies in children with obesity have yielded inconsistent results. Discrepancies in estimating glomerular filtration rate (eGFR) equations might play a major role, which was therefore evaluated in this study. Another aim of this study was to examine whether normalized serum creatinine (SCr) for age and sex can be used as a kidney biomarker.
Material and methods: In this cross-sectional study, 600 children with overweight and obesity were included (53.5% girls, mean age 12.2 years, mean BMI z-score 3.31). Children underwent clinical and laboratory examination, air displacement plethysmography, blood-pressure measurement and polysomnography. SCr was normalized using Q-age and Q-height polynomials and creatinine-based eGFR-equations were compared.
Results: Normalized SCr (SCr/Q) and nearly all GFR estimations significantly correlated with fat mass, waist to hip ratio, homeostasis model

assessment for insulin resistance, HDL cholesterol, triacylglyceride, serum uric acid and alanine transaminase concentrations. The presence and strength of these correlations was not confirmed by all formulas, suggesting dependency on the mathematical form of the eGFR-equation.
Conclusions: In conclusion, correlations between SCr/Q or creatinine-based eGFR and obesity-related comorbidities can be found in children with overweight and obesity, but depend very much on the eGFR-equation of choice. SCr/Q might be an appropriate kidney biomarker for assessing correlations with cardiovascular and metabolic risk factors in children with overweight and obesity.

EP-18 NEW FAMILY WITH KCNJ16 GENE TUBULOPATHY, CHARACTERIZED BY HYPOKALEMIA, IMPAIRED ACID-BASE HOMEOSTASIS AND SENSORINEURAL HEARING LOSS

Alejandro Garcia-castaño[1], Sara Gómez-conde[2], Leire Madariaga[1], Gema Ariceta[1]

[1]Biocruces Bizkaia Health Research Institute, Barakaldo, Spain. Renaltube Group, *[2]Biocruces Bizkaia Health Research Institute, Barakaldo, Spain Pediatric Nephrology Department, Cruces University Hospital, University Of The Basque Country Upv/ehu Barakaldo, Spain.*, *[3]Biocruces Bizkaia Health Research Institute, Barakaldo, Spain Renaltube Group. Pediatric Nephrology Department, Cruces University Hospital, University Of The Basque Country Upv/ehu Barakaldo, Spain.*, *[4]Pediatric Nephrology Department. University Hospital Vall D´hebrón. Universitat Autònoma Barcelona, Barcelona. Spain. Renaltube Group*

Background: A new tubular entity caused by biallelic variants in KCNJ16 gene and mixed proximal and distal tubulopathy manifestations such as hypokalemia, acid-base disorder and salt-wasting associated with sensorineural hearing loss (SNHL), has been recently identified. In the kidney, the potassium channel subunit KCNJ16 forms functional heteromers with KCNJ15 in the proximal tubule, and with KCNJ10 in the distal nephron, playing a key role on tubular transport processes and potassium and pH sensing.
Aim: To describe a new family affected with KCNJ16 gene tubulopathy, phenotype variation in two siblings, and patient follow-up, to expand the knowledge about this entity.
Material and methods: Medical records review, molecular diagnosis by Next-Generation Sequencing (NGS), using the Ion GENESTUDIO S5 SYSTEM sequencer with ION CHEF for exome analysis and later sequencing by Sanger.
Results: Two infant brothers from a consanguineous Pakistani family, apparently healthy at birth presented with failure to thrive, hypokalemia of renal origin, and mild metabolic acidosis. **Genetic diagnosis:** c.409C>T p.Arg137Cys homozygous mutation in the KCNJ16 gene (ENST00000392671.6) were identified in both brothers, each one segregated for patients' parents who are asymptomatic carriers.

	Sibling 1		Sibling 2	
	At diagnosis	Last follow up	At diagnosis	Last follow up
age	17 months	10.3 (years)	7 months	3 years
symptoms	-failure to thrive & growth retardation since 4 months of age, -vomits, feeding difficulties -polydipsia-polyuria -developmental delay	-mild fatigue - moderate salt avidity -learning difficulties	failure to thrive, & growth retardation vomits	mild polyuria
Weight Kg (perc)	7.5 (<3)	39.5 (50-75)	6.650 (<3)	13.750 (3-10)
Height cm (perc)	83 (<3)	137 (25-50)	65 cm (3)	92.5 (3-10)
Serum Na, K, Cl,	126/1.5/87	138/3.2/ 107	133/2.7/ 95	140/ 3.3/ 107
(mEq/L) Mg (mg/dL)	2.2	1.9	Not available	1.9
acid-base (venous)	pH 7.39, bicarbonate 22	pH 7.23, bicarbonate 21.4	pH 7.45, bicarbonate 17	pH 7.32, bicarbonate 25.2
Serum Cr mg/dL	0.28	0.64	0.35	0.44
FENa%,	0.15%,	0.32%,	0.8%	1.19%
FEK%,	106.5%	44.9%	29.7%	58.4%
FECl%	1.89%	1.5%	1.4%	1.48%
UCa/Cr (mg/mg)	0.1	0.01	0.34	0.01
eGFR	122	88	77	87
hearing	Not available	mild neurosensorial hypoacusia	Not available	normal
Kidney ultrasound	normal	normal	normal	normal
Treatment		KCl + K citrate + indometacin		KCl + K citrate + indometacin

Conclusions: This new family with KCNJ16 gene tubulopathy was characterized by early failure to thrive, variable acid-base balance anormalities, moderate polyuria-polydipsia and salt wasting, hypokalemia of renal origin and hypocalciuria. Over time one patient developed bilateral sensorineural hearing loss but both siblings preserved kidney function.

EP-19 A SINGLE-CENTER ANALYSIS ON GENETICS OF KIDNEY STONE FORMERS: HAVE WE LEARNED ANYTHING?

George Claudiu Costea, Anca Elena Marin, Mihai Gurgu, Mona Irina Matei, Florina Badea, Adrian Catalin Lungu, Ovidiu Limoncu, Cristina Stoica

Fundeni Clinical Institute

Introduction: Urolithiasis is defined as the pathological entity in which calculi are formed within the urinary tract, opposed to nephrocalcinosis, which occurs secondary to calcium deposition within the renal parenchyma. We aimed to analyze the cases of pediatric urolithiasis and nephrocalcinosis in which genetic testing has been performed within the last year in our department.
Material and methods: Next generation sequence technology in an accredited clinical laboratory was performed in selected cases of pediatric urolithiasis. The criteria for patient selections were at least one of the following: young age at diagnosis, bilateral calculi, family history or recurrence of the disease. A number of 28 cases were analyzed after obtaining the informed consent for participating in this study.
Results: We had a number of 28 subjects enrolled in this study; of these, 69.7% (17) were males. The mean age of our patients was 6.51 years (range from 6 months to 17 years). Positive analysis was identified in 57.1% (16) of cases. No statistically significant correlation was found between age at diagnosis and positive genetic finding. However, a statistically significant correlation was made between positive family history and positive genetic finding (p-0.03, Pearson correlation).No statistically significant correlation has been found between the site of nephrolithiasis (unilateral or bilateral) and the rate of positive findings.
Conclusions: Our study included more male patients, consistent with the literature supporting male gender to be a risk factor for nephrolithiasis and nephrocalcinosis. Children with positive family history of urolithiasis or nephrocalcinosis should undergo genetic testing for identifying the cause. Although our study did not statistically correlate young age and bilateral nephrolithiasis with gene pathogenic variants, we still recommend genetic testing in these cases.

EP-20 CHALLENGES IN FOLLOW-UP OF CHILDREN SURVIVING EPISODE OF ACUTE KIDNEY INJURY IN A RESOURCE LIMITED-SETTING

Agathe Nkoy[1], Ndiyo Yoly[1], Matoka Therance[1], Odio Matondo[1], Betukumesu Dieumerci[1], Kazadi Orly[1], Veerle Labarque[2], Lambertus P. Van Den Heuvel[3], Levtchenko Elena[3], Ekulu Pepe[1]

[1]Division Of Nephrology, Department Of Pediatrics, University Hospital Of Kinshasa, Faculty Of Medicine, University Of Kinshasa, Kinshasa, Democratic Republic Of Congo., [2]Department Of Pediatric Haematology, University Hospital Leuven, Leuven, Belgium, [3]Department Of Development And Regeneration, Ku Leuven, Leuven, Belgium.

Introduction: While acute kidney injury (AKI) has been reported as an important risk factor of chronic kidney disease (CKD), the long-term follow-up of children who survive to AKI remains a big challenge in resource limited-settings. Our study aimed to assess the kidney outcome of children who experienced AKI and survived to hospital discharge in a resource-limited setting.
Material and methods: This is a retrospective review on the kidney outcome of children admitted for AKI and survived after hospital discharge from January 2018 to December 2021 at the University Hospital of Kinshasa. The main outcome parameters were proteinuria by dipstick ($\geq +1$), albuminuria (uACR ≥ 30 mg/g), decreased estimated glomerular filtration rate (eGFRcr < 60ml/min/1.73m²). CKD was defined as the presence of albuminuria and/or decreased eGFRcr for more than three months.
Results: From January 2018 to December 2021, 218 children (126 boys and 92 girls) were admitted for AKI. The median age was 7 years (1.5 months-16 years). The leading causes of AKI were severe malaria (95/218; 43.6%), sepsis (41/218; 18.8%), and HUS (38/218; 17.4%). Dialysis was indicated and performed in 148 of 218 (67.9%). At the discharge, 180/218 (82.6%) recovered efficient diuresis and 40 (18.3%) died probably due to a late transfer. Out of 180 children, only 71 (39.4%) returned for follow-up. There were 26, 9, 11 and 12 children who returned for follow-up at 3, 6, 9, ≥12 months after the discharge, respectively. CKD was found in 2/71 children (2.8%).
Conclusions: These results emphasize the pressing need to implement a strategic policy for the retention of patients surviving from an episode of AKI for an effective periodic follow-up.

EP-21 GROSS HEMATURIA AFTER COVID 19 IMMUNIZATION IN PEDIATRIC PATIENTS AND LITERARY REVIEW

Andrea Puma, Michela Gritti, Chiara Tosolini, Sara Picassi, Laura Venditto, Giorgio Piacentini, Milena Brugnara

Department Of Surgical Sciences, Dentistry, Gynecology And Pediatrics, Pediatric Division, University Of Verona, Verona, Italy

Introduction: From the beginning of massive vaccination programme several immune-mediated reactions, including cases of myocarditis and newly diagnosed or relapsed glomerulonephritis (GN), have been reported. Recent reviews report of IgA nephropathy (IgAN) flare-up presenting as gross hematuria, following COVID-19 vaccination in adult patients. However, evidence of de novo or flare up GN in pediatric patients is lacking, and, to date, there are only few case reports that includes children and adolescents.
Material and methods: We evaluated characteristics and clinical outcomes of 3 patients who had a new onset gross hematuria following mRNA COVID-19 vaccination and provide a brief literature review of pediatric cases of GN after vaccine.
Results: In this case series we report 3 cases of new onset macroscopic hematuria post–COVID-19 mRNA vaccines. In particular one patient showed a severe IgAN with gross hematuria, proteinuria and decreased renal function within few days after third dose of mRNA COVID-19 vaccination.
Conclusions: mRNA COVID-19 vaccine appeared to relate to de novo or relapsing GN in some individuals but the mechanism by which COVID-19 vaccination is associated with GN flares is still unclear. Pediatricians lack data to advise patients about the chance of relapse of GN after COVID19 vaccination and the current recommendation is based on case reports and clinical experience. The risk and severity of de novo or relapsing GN after COVID-19 vaccine is not fully understood and this can be challenging during vaccination counselling. Further research is required to understand the relation between COVID-19 vaccination and GN which can provide a more scientific basis for patient counselling, guidance in risk stratification and monitoring for relapse.

EP-22 EXTENSIVE REVIEW OF PHARMACOKINETIC AND PHARMACODYNAMIC PROPERTIES OF ACE-INHIBITORS AND ANGIOTENSIN RECEPTOR BLOCKERS IN CHILDREN WITH RENAL IMPAIRMENT

Eva Degraeuwe[1], Louis Sandra[2], Paulien Devos[2], Elke Gasthuys[2], Evelien Snauwaert[1], Ann Raes[1], Johan Vande Walle[3]

[1]Department Of Internal Diseases And Paediatrics, Faculty Of Health And Medical Sciences, Ghent University, [2]Laboratory Of Medical Biochemistry And Clinical Analysis, Faculty Of Pharmaceutical Sciences, Ghent University, [3]Ghent University Hospital (uz Ghent)

Introduction: Childhood hypertension is affecting around 2 to 4% of the paediatric population, where 90% of the Western cases is caused by renal impairment. ACE-inhibitors (ACE-I) and Angiotensin Receptor Blockers (ARB's) are most frequently prescribed for blood pressure reduction. Both therapeutics are currently being used off-label, despite being available for over three decades. The aim of this study is to retrospectively summarize and to compare findings of conducted pharmacokinetic (PK) and pharmacodynamic (PD) studies investigating all drugs of the ACE-I and ARB's classes, including a potential recommendation for improved study design.
Material and methods: This review focused on the clinical trials investigating PK and PD properties of ACE-I and ARB's. 60 studies were selected, including 19 randomized controlled trials. Analysis was conducted with a focus on trial design and endpoints, i.e. safety and efficacy.
Results: Between ACE-I and ARB's, geographical location, drug intake and formulations were comparable. Study population differed, for example studies on ARB's focused on both primary and secondary hypertension, where studies on ACE-I focused on secondary hypertension. Sampling regimens varied, where studies investigating the PK of ARB's were frequently based on single dosing at non-steady state. For both classes, low reporting of estimated glomerular filtration rate (eGFR) (23.3%) and the exclusion of participants with an eGFR under 30 was apparent. Around 90% of the individual reportings using ACE-I achieved an antihypertensive effects of $\geq$ 6 mmHg. ACE-I were generally well tolerated when considering safety parameters and serious adverse events. Limited studies investigated the long-term effects of ACE-I and ARB's on cardiovascular morbidity and mortality.
Conclusions: Standardization of methodology and reporting of results is imperative for PKPD studies, to allow a better comparison of results and to aim towards appropriate labelling. Inclusion across and stratification for age categories and eGFR ranges is recommended.

EP-23 ARTERIAL STIFFNESS AND AMBULATORY BLOOD PRESSURE MEASUREMENTS IN CHILDREN WITH FAMILIAL MEDITERRANEAN FEVER

Emre LeventoĞlu[1], Bahar BÜyÜkkaragÖz[1], Emine Nur Sunar Yayla[2], Pelin Esmeray Şenol[2], Sevcan A. BakkaloĞlu[2], Sevcan A. BakkaloĞlu[1]

[1]Gazi University, Faculty Of Medicine, Department Of Pediatric Nephrology, [2]Gazi University, Faculty Of Medicine, Department Of Pediatric Rheumatology

Introduction: Familial Mediterranean Fever is an autoinflammatory disease which may cause the endothelial dysfunction and arterial stiffness. In this study, arterial stiffness indicators like PWV, AIx and ABPM were performed in the patients with FMF and it was aimed to evaluate whether there are any differences between the patients in terms of endothelial damage.
Material and methods: This is a single center, prospective, case-control study conducted on pediatric patients with FMF. Patients were divided into groups according to their treatment modalities: colchicine-only therapy (group 1) vs. colchicine and an IL-1 antagonist together (group 2).
Results: The study group comprised 63 pediatric FMF patients (28 boys and 35 girls). The frequency of normotensive state was lower and stage I HT was higher in group 2; ABPM revealed that the nighttime systolic BP load and the frequency of nocturnal hypertension were significantly higher in group 2; measures of arterial stiffness including mean aortic pressure, pulse pressure and PWV were significantly higher in group 2.
Conclusions: Endothelial damage develops due to inflammation in FMF patients and arterial stiffness increases due to this damage. The arterial stiffness is more pronounced in group 2, due to more severe clinical symptoms and an augmented inflammatory milieu.

EP-24 URINE PROTEIN ARRAY ANALYSIS TO IDENTIFY KEY INFLAMMATORY MARKERS IN CHILDREN WITH IGA VASCULITIS NEPHRITIS

Julien Marro[1], Andrew Chetwynd[1], Rachael Wright[1], Louise Oni[1]

[1]Department Of Women's And Children's Health, Institute Of Life Course And Medical Sciences, University Of Liverpool, Liverpool, United Kingdom, [2]Department Of Paediatric Nephrology, Alder Hey Children's Nhs Foundation Trust Hospital, Liverpool, United Kingdom

Introduction: Chronic kidney disease is a recognised complication of Immunoglobulin A (IgA) Vasculitis (previously Henoch Schonlein Purpura, HSP). The exact pathophysiology of this disease remains largely unknown and identifying urinary inflammatory markers could aid identification of targets for earlier diagnosis and/or treatment.
The aim of this pilot study was to conduct a large protein panel evaluation of urine samples in children with IgA Vasculitis to discover proteins associated with nephritis.
Material and methods: Paediatric patients with IgAV and healthy controls (HC) were recruited as part of the IgA Vasculitis Study (Alder Hey Children's NHS Foundation Trust, Liverpool, REC 17/NE/0390). Patients with a diagnosis of IgAV were grouped into those with nephritis (IgAVN) and those without (IgAVwoN). Nephritis was defined as a urinary albumin to creatinine ratio (UACR) >30 mg/mmol. Determination of relative levels of 126 proteins (encompassing inflammatory cytokines and known markers of kidney inflammation) was performed using commercially available proteome profiler array kits (Human Kidney Biomarker and Human XL Cytokine kits, R&D System Ltd). ImageJ software was used for the pixel density analysis and data was normalised to control points, volume of urine and urinary creatinine. Statistical analysis was performed using MetaboAnalyst 5.0 software.
Results: 11 children were included in this study (HC n=3, IgAVN n=4, IgAVwoN n=4). Median age was 7.6 years [4.0-13.44] and male:female ratio was 1.2:1. For IgAVN, median UACR was 542.2mg/mmol [110.4-2,357.37]. Multivariate analysis identified 28 proteins which were significantly different when comparing IgAVN to IgAwoN and HC; 13 proteins were also significantly different between IgAwoN and HC. There was no difference between all IgAV and HC.
Conclusions: This data has identified key urinary proteins that provide insight into the pathophysiology of IgAVN. The identified proteins include components of the complement pathway, CXCR pathway and RAAS system. Further analysis is required to validate these findings.

EP-26 40TH ANNIVERSARY OF FIRST PEDIATRIC KIDNEY TRANSPLANTATION IN UNİVERSİTY HOSPİTAL CENTER ZAGREB

Ivanka Kos, Kristina Vrljičak, Maja Ban, Hana Matković, Maša Davidović, Lovro Lamot, Jasna Slaviček

Division Of Nephrology, Dialysis And Transplantation, Department Of Pediatrics, University Hospital Centre Zagreb, University Of Zagreb School Of Medicine, Zagreb, Croatia

Introduction: Kidney transplantation (KT) is the optimal treatment for children with end-stage renal disease (ESRD). The first pediatric KT was introduced in 1964. in Pittsburgh, and the first pediatric KT in our Center was performed in 1982. to a 12 -year old boy with nephrotic syndrome.
Methods: This is a retrospective, descriptive study.
Results: In 40 years (1982-2022), 148 children have been followed at UHC Zagreb after KT. Out of this number, 82,4% were performed in Croatia. There were more kidney transplants from deceased than from living donors (67.5% versus 32.4%), with 13.5% being preemptive. The majority of transplanted children (42.5%) had congenital anomalies of the kidneys and urinary tract as their primary disease, while other common causes of ESRD included glomerulonephritis (14,8%), FSGS (12.2%), nephronophthisis (5.4%), congenital nephrotic syndrome (3.3%), polycystic kidney disease (3.3%), IgA nephropathy (2,7%), and Alport syndrome (2.7%). Most of the patients (65.5%) were 6 - 15 years old, and 58% were male.
Results: It was observed that one- year graft survival rates were 69% (1982 to 1990), 95% (1990-2000), 92.3% (2000-2010), 91.4% (2010-2020), while five- year graft survival rates were 53.8% (1982 to 1990), 76,2% (1990-2000), 81.5% (2000-2010), 85.7% (2010-2020). Ten- year patient survival rates were 85% (1982 to 2000), and 96.1% (2000-2020).
Conclusion Outcomes after pediatric KT in Croatia have been markedly improved after the first decade, mainly due to greater surgical experience, improved immunosuppression and donor selection criteria. Patient survival and 5- year graft survival rates have dramatically improved after the first decade and continued to improve thereafter. One- year graft survival has remained the same since the 1990s. Despite the better outcomes following initial transplantation, many challenges such as newer immunosuppressive agents with optimal balance, improved surgical techniques and better antiviral prophylaxis, remain to be implemented in the years to come.

EP-27 HYPERTENSION SUBTYPES EFFECTING LEFT VENTRICULAR HYPERTROPHY IN OBESE HYPERTENSIVE CHILDREN

Belde Kasap Demir[1], Cemaliye Başaran[2], Tülay Demircan[3], Eren Soyaltın[4], Gökçen Erfidan[2], Özgür Özdemir Şimşek[2], Seçil Arslansoyu Çamlar[5], Demet Alaygut[5], Fatma Mutlubaş[5]

[1]Department Of Pediatrics, Division Of Pediatric Nephrology And Rheumatology, Izmir Katip Çelebi University, Izmir, Turkey, [2]Department Of Pediatrics, Division Of Nephrology, University Of Health Sciences Izmir Tepecik Training And Research Hospital, Izmir, Turkey, [3]Department Of Pediatrics, Division Of Cardiology, University Of Health Sciences Izmir Tepecik Training And Research Hospital, Izmir, Turkey, [4]University Of Health Sciences, Başakşehir Çam And Sakura City Hospital, Department Of Pediatrics, Division Of Nephrologyistanbul, Turkey, [5]Izmır Faculty Of Medicine, Department Of Pediatrics, Division Of Nephrology, University Of Health Sciences Turkey, Izmir, Turkey

Introduction: We aimed to evaluate the BP indices effecting LVH in obese children. Since BMI is a strong and independent predictor of LVH, we evaluated obese children with HT determined with office BP measurements and/or ABPM.
Material and methods: The obese cases (BMI>95p) who were detected with ABPM for the first time between December 2018 and December 2021 were included in the study. Office systolic and diastolic HT is defined as BP measures >95p according to AAP 2017. Systolodiastolic HT was defines as both systolic and diastolic BPs >95p. Ambulatory systolic HT is defined as daytime and/or nighttime systolic HT, and ambulatory diastolic HT is defined as daytime and/or nighttime diastolic HT. Ambulatory systolodiastolic HT is defined as both sytolic and diastolic HT in the same monitorization. “Isolated systolic HT” is defined as mean office and ambulatory systolic BPs ≥95p and office and ambulatory diastolic BPs<95p. “Isolated diastolic HT” is defined as office and ambulatory diastolic BPs ≥95p and office and ambulatory systolic BPs <95p. “Isolated nocturnal hypertension” is defined as mean nighttime systolic and/or diastolic BP ≥95p and mean daytime BP<95p. “Isolated daytime hypertension” is defined as mean daytime systolic and/or diastolic BP ≥95p and mean nighttime BP<95p. “Combined daytime and nighttime hypertension” is defined as mean nighttime and daytime systolic and/or diastolic BPs≥95p regardless of the BP load.
Results: There were 128 obese cases who were detected with ABPM and echocardiographic examination in the defined period. Those who had both normal office and ambulatory HT were 47 cases. The 81 cases who had either office or ambulatory HT were assessed. Nineteen (23%) of the cases had LVHT. The age, gender, body weight, height and BMI SDSs were similar between the cases with and without LVH. Office and ABPM systolic and diastolic BP parameters and HT rates, systolic and diastolic dips and daytime and nighttime loads were similar between the groups. Isolated nocturnal and daytime HT, combined daytime and nighttime HT and isolated diastolic HT rates were similar between the groups. However, the isolated systolic HT was significantly more common in obese cases with LVH (p=0.025).
Conclusions: Isolated systolic HT is the hypertension subtype contributing to LVH in obese hypertensive patients

EP-28 IMMUNOGENICITY AND ANTIBODY PERSISTENCE OF A BOOSTER 13-VALENT PNEUMOCOCCAL CONJUGATE VACCINE DOSE IN CHILDREN WITH IDIOPATHIC NEPHROTIC SYNDROME (INS)

Varvara Askiti[1], Konstantina Kitsou[2], Argyroula Zampetoglou[1], Andromachi Mitsioni[1], Vana Spoulou[2]

[1]Pediatric Nephrology Department Athens Childrens Hospital "p&a Kyriakou", [2]Immunobiology And Vaccinology Research Laboratory, First Department Of Paediatrics, “aghia Sophia” Children’s Hospital, School Of Medicine, National And Kapodistrian University Of Athens

Introduction: Children with INS under immunosuppression often demonstrate lower vaccine-derived protection. We evaluated the immunogenicity of a 13-valent Pneumococcal Conjugate Vaccine (PCV13) dose and the persistence of antibodies induced by PCV13 against Pneumococcal Serotypes, PS1, PS3, PS7F, PS9V and PS19A, in children with INS under corticosteroids and immunomodulatory treatments (IMT)-Cyclosporine-A and mycophenolate mofetil-compared to healthy children.
Material and methods: Blood was collected from INS patients, aged 3.3-16.4 years under corticosteroids (n=17)-GroupA, IMT (n=10)-GroupB and from age-matched healthy controls (n=17)-GroupC, prior, one-month (1M) and 12 months (12M) after a PCV13 booster. All children have received PCV immunization in infancy. Serum was separated, frozen, and stored at −20ºC until tested. The World Health Organization ELISA protocol was used for the detection of PS-specific immunoglobulin G (IgG) antibodies in serum samples. Mann-Whitney U-test and Wilcoxon-rank test were performed.

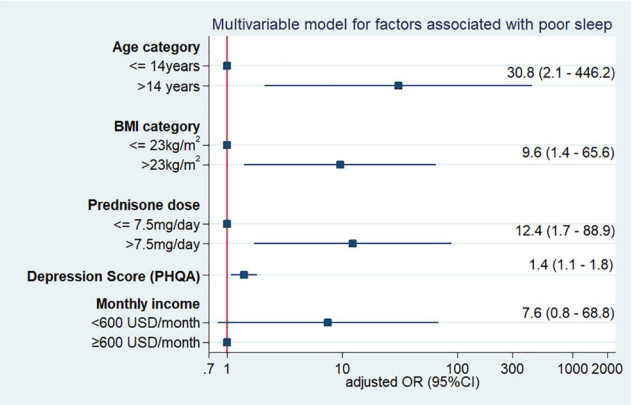
Results: Compared to baseline, significantly increased (protective) antibody titers were achieved at both timepoints in all groups for all Serotypes tested ($p<0.05$). No statistically significant differences in the titers against the Serotypes evaluated were observed at baseline among groups. No statistically significant differences were observed between GroupA and GroupC in the anti-PS antibody titers for all serotypes tested at any timepoint. Significantly lower antibody titers were observed in GroupB compared to GroupC for PS1, PS3 and PS9V at 1M and 12M (PS1: 1M: p=0.013 and 12M: p=0.02, PS3: 1M: p=0.015 and 12M: p=0.04, PS9V: 1M: p=0.023, 12M: p=0.04). No statistically significant differences were observed between GroupB and GroupC in the antibody titers against PS7F and PS19A at any timepoint.
Conclusions: These results indicate a negative effect of IMT on PS-specific antibody responses to PCV13.

EP-29 SLEEP DISTURBANCES IN ADOLESCENTS WITH SYSTEMIC LUPUS ERYTHEMATOSUS

Pattareeya Yotasan[1], Stephen J Kerr[2], Montida Veeravigrom[3], Nuanpan Siripen[1], Pornpimol Rianthavorn[1]

[1]Department Of Pediatrics, Faculty Of Medicine, Chulalongkorn University, *[2]Research Affairs, Faculty Of Medicine, Chulalongkorn University.*, *[3]Section Of Child Neurology, Department Of Pediatrics, The University Of Chicago Biological Sciences*

Introduction: Sleep disturbances are understudied in adolescents with systemic lupus erythematosus (SLE). The aims of this study were to examine the prevalence of sleep disturbances and determine factors related to poor sleep in adolescents with SLE.
Material and methods: The Pittsburgh Sleep Quality Index (PSQI) and the Patient Questionnaire of Adolescents (PHQA) were administered to children aged 13-18 years with SLE to evaluate sleep quality and depression, respectively. Participants with PSQI≥5 were considered to be poor sleepers. Participants with PHQA≥5 were considered to be at risk of depression. Variables which were significantly different between good sleepers and poor sleepers were used to run univariable and multivariable logistic regression models.
Results: Fifty-seven patients (46 females, 81%) with a median age (interquartile range) of 15 years (13.5-17) and a median disease duration of 44 months (17.8-69) were enrolled. The median body mass index (BMI) was 22.6 kg/m2 (18.6-26.2), and 22 participants (38.6%) were overweight (BMI>23 kg/m2). All participants received daily prednisolone with a median dose of 10 mg (5-30). The median PSQI was 4 (2-5). Eighteen participants (31.6%) were poor sleepers. The median PHQA was 3 (1-7). Eighteen participants (31.6%) were at risk of depression. Five factors significantly different between poor sleepers and good sleepers, namely age >14 years (middle vs. early adolescent), being overweight, daily prednisolone >7.5 mg (high vs. low daily prednisolone dose), PHQA score, and monthly household income <600 US dollars (lower vs. higher income), and were included in a multivariate model. Figure outlines the adjusted odds ratio and confidence interval (CI) of factors associated with poor sleep quality. The adjusted odds of having poor sleep in middle adolescents, overweight participants, or taking high daily prednisolone dose were significantly increased by 30.8, 9.6 and 12.4 fold, respectively. There was a 1.4-fold increase in the odds of having poor sleep for every unit increase in PHQA score. The odds of having poor sleep in participants with lower income were increased 7.6 fold. Although not statistically significant, the 95%CI were consistent with an elevated odds of poor sleep in those with lower incomes.



Conclusions: A substantial number of adolescents with SLE had sleep disturbances and depressive mood. Routine assessment and management of sleep problems and mood disorders should be part of the comprehensive care of adolescents with SLE.

EP-30 PERSISTENT MULTIPLE POST-COVID COMPLICATIONS WITH AUTONOMIC DYSFUNCTION IN A 16- YEAR OLD GIRL WITH END-STAGE RENAL DISEASE

Ivanka Kos[1], Katja Dumić Kubat[2], Dorotea Bartoniček[3], Ivan Lehman[4], Maja Ban[1], Hana Matković[1], Lovro Lamot[1], Kristina Vrljičak[1]

[1]Division Of Nephrology, Dialysis And Transplantation, Department Of Pediatrics, University Hospital Centre Zagreb, University Of Zagreb School Of Medicine, Zagreb, Croatia, *[2]Division Of Endocrinology And Diabetes, Department Of Pediatrics, Uhc Zagreb, University Of Zagreb School Of Medicine, Zagreb, Croatia*, *[3]Division Of Cardiology, Department Of Pediatrics, Uhc Zagreb, University Of Zagreb School Of Medicine, Zagreb, Croatia*, *[4]Division Of Neurology, Department Of Pediatrics, Uhc Zagreb, University Of Zagreb School Of Medicine, Zagreb, Croatia*

Introduction: The world is facing COVID-19 pandemic with a complex presentation. Moreover, many people are suffering from a variety of post-COVID symptoms. Although it appears that children experience less severe disease, those with underlying conditions such as chronic kidney disease (CKD) are more susceptible to infection and post-COVID syndrome
Material and methods: We report a 16-year old girl who has been on peritoneal dialysis (PD) for the past 10 years. A kidney transplant was performed at the age of eight, complicated by early graft loss. The further course was characterized by many CKD related comorbidities including hypertension
Results: In December 2020 she had a COVID-19 infection with pneumonia and encephalopathy. In the following months she developed suppurative hidradenitis, partially improved after adalimumab treatment. Pericarditis was diagnosed five months post-COVID and successfully treated with NSAIDs and intravenous immunoglobulins. More than 6 months after COVID-19 infection she suffered from numerous complications, including paroxysmal involuntary leg movements, memory and mood problems. The most severe of post-COVID disorders was refractory hypertension with blood pressure (BP) exceeding 200/100 mmHg. The Schellong test showed BP drops of >30%. Quantitative autonomic nervous system testing confirmed adrenergic score4, and cardiovagal score1 leading to diagnosis of orthostatic dysregulation with pronounced supine hypertension. Different combinations of antihypertensives have been tried, but without anticipated effect. Despite the conversion to hemodialysis and achieved fluid loss, BP

remained high for weeks. Recently, one year after COVID-19 infection, gradual improvement in BP has occurred
Conclusions: There is a growing body of literature describing post-COVID complications in children with severe chronic diseases. In order to understand the complex underlying pathophysiological mechanism, further investigations and follow-up are required.

EP-32 EVALUATION OF UNCLASSIFIED AMBULATORY BLOOD PRESSURE MONITORING PHENOTYPES IN TERMS OF LEFT VENTRICULAR HYPERTROPHY

Belde Kasap Demir[1], Cemaliye Başaran[2], Tülay Demircan[3], Gökçen Erfidan[2], Özgür Özdemir Şimşek[2], Seçil Arslansoyu Çamlar[4], Demet Alaygut[4], Fatma Mutlubaş[4], Cem Karadeniz[5]

[1]Department Of Pediatrics, Division Of Pediatric Nephrology And Rheumatology, Izmir Katip Çelebi University, Izmir, Turkey, [2]Department Of Pediatrics, Division Of Nephrology, University Of Health Sciences Izmir Tepecik Training And Research Hospital, Izmir, Turkey, [3]Department Of Pediatrics, Division Of Cardiology, University Of Health Sciences Izmir Tepecik Training And Research Hospital, Izmir, Turkey, [4]Izmr Faculty Of Medicine, Department Of Pediatrics, Division Of Nephrology, University Of Health Sciences Turkey, Izmir, Turkey, [5]Department Of Pediatrics, Division Of Pediatric Cardiology, Izmir Katip Çelebi University, Izmir, Turkey

Introduction: We aimed to evaluate the clinical significance of unclassified groups as an ambulatory blood pressure (ABP) phenotype with regard to left ventricular hypertrophy(LVH).
Material and methods: Office and ABPM measures of cases who were also evaluated for LVH were included in the study. According to office BPs(OBPs), mean ambulatory SBP or DBPs(MABPs), and SBP or DBP loads(BPLs), ambulatory BPs were classified as normal (NT;N OBPs, MABPs<95p, BPLs<25%), white coat hypertension (WCHT;OBPs≥95p, MABPs<95p, BPLs<25%), prehypertension (Pre-HT;OBP≥90p, MABPs<95p, BPLs≥25%), masked hypertension (MHT;OBP<95p, MABPs>95p, BPLs≥25%), and ambulatory hypertension (AHT;OBP>95p, MABPs>95p, BPLs≥25%). Unclassified cases (UC1) with N OBPs, MABPs<95p, but BPLs≥25% and those (UC2) with OBPs≥95p, MABPs<95p, and BPLs≥25% were defined. LVMI, LVMI/95p values and LVH ratios were compared between NT, UC1 and MHT groups and between WCHT, UC2 and AHT groups.
Results: A total of 533 cases were included. There were 52 cases in the NT, 47 cases in UC1, 50 cases in MHT, 79 cases in WCHT, 104 cases in UC2, and 165 cases in AHT group. Cases in MHT group were older than those of the NT and UC1 groups. Gender, BMI, BMI SDS, LVMI, LVMI/95p values were similar between the NT, UC1 and MHT cases and between WCHT, UC2 and AHT cases. LVH in MHT was significantly higher than UC1 and NT groups. LVH was significantly higher in the AHT group compared to WCHT, and similar between WCHT and UC2 groups.
Conclusions: This is the first study assessing the importance of unclassified groups in terms of LVH. Being in the UC1 or UC2 groups do not change the status of NT or WCHT. As the only difference between NT and UC1 groups and WCHT and UC2 groups is the BPLs, we may conclude that having BPLs>25% does not contribute to LVH in children.

EP-33 ACUTE HEMODIALYSIS EXPERIENCE IN PEDIATRIC PATIENTS WEIGHING LESS THAN 15 KG

Ömer Nazim GÜlÇek[1], Bora Gulhan[2], Nesrin TaŞ[2], GÜlŞah Özdemir[2], Demet Baltu[2], Tuba Tastemel ÖztÜrk[2], Eda Didem Kurt ŞÜkÜr[2], Fatih Ozaltin[2], Ali Duzova[2], Rezan Topaloglu[2]

[1]Hacettepe University School Of Medicine, Department Of Pediatrics, Ankara, Turkey, [2]Hacettepe University School Of Medicine, Department Of Pediatrics, Division Of Pediatric Nephrology, Ankara, Turkey

Introduction: There are few studies on the prognosis of pediatric patients who have received renal replacement therapy (RRT). This study aims to examine the clinical features of patients who have undergone acute RRT while weighing less than 15 kg.

Material and methods: Patients who have undergone acute RRT while weighing less than 15kg with a minimum follow-up period of 6 months were included. Survived patients were examined at the last visit.
Results: A total of 80 patients (42 female, 38 male) were included in the study. Among them, 43 patients (53.8%) received hemodialysis (HD) and 37 patients (46.2%) received peritoneal dialysis (PD). The age, height, weight, and body surface area of HD patients were found to be significantly higher than those of PD patients ($p<0.001$). Most of the patients less than 5 kg (96%) received PD and most of the patients between 10.1-15 kg (88.6%) received HD. At the end of the dialysis process, 41 patients (51.3%) survived. It was determined that the height, weight, and body surface area values of the surviving patients at the beginning of dialysis were higher than those of the deceased patients ($p<0.001$). In multiple regression analysis, only low weight and vasopressor treatment were found to be negatively associated with survival. Dialysis modality did not affect survival. At the last visit 31 patients could be evaluated. Non-nephrotic proteinuria was found in six (19.3%) of 31 patients (38.8%). Stage 2 chronic kidney disease (CKD) was diagnosed in three patients (9.6%). Office blood pressure measurement revealed hypertension in one patient. Masked hypertension was detected in one patient with 24-hr ABPM.
Conclusions: PD and HD can be used safely, and effectively in patients weighing less than 15 kg. Findings such as CKD, proteinuria, and hypertension may be observed in these group of patients and they should be followed up regularly.

EP-34 IS IT JUST SARS-COV-2 RELATED SEPTIC SHOCK?

GÖkÇen Erfidan[1], ÖzgÜr Özdemir ŞimŞek[1], Demet Alaygut[2], SeÇil Arslansoyu Çamlar[2], Fatma MutlubaŞ[2], Belde Kasap Demir[3]

[1]University Of Health Sciences Izmir Tepecik Training And Research Hospital, Department Of Pediatric Nephrology., [2]University Of Health Sciences Izmir Faculty Of Medicine, Department Of Pediatric Nephrology., [3]Izmir Katip Celebi University, Faculty Of Medicine, Department Of Pediatric Nephrology And Rheumatology.

Introduction: Pediatric COVID-19 disease has been defined as a mild illness compared to that in adults, as presenting mostly with fever and cough. However, some severe cases and multisystemic inflammatory syndrome in children (MISC) have been described worldwide.
Material and methods: Since the criteria of MISC overlaping with sepsis and septic shock, the differential diagnosis could be challenging. Here, we present a case with active COVID-19 and septic shock, misdiagnosed with MISC.
Results: A 15-year-old female patient applied with fever for two days, stomachache and vomiting. She had cadaveric renal transplantation seven years ago. She had history of recurrent urinary infections, vesicoureteral reflux (VUR) to graft kidney, two subureteric transurethral injections, and one ureteroneocystostomy procedure. There had been no graft dysfunction or rejection so far (basal serum creatinine: 0.9 mg/dL). She had not yet been vaccinated for COVID-19.
On physical examination, she had fever (39.5°C), tachycardia, tachpnea, hypotension, diffuse abdominal tenderness with prominancy over graft. Laboratory results showed leukocytosis, neutrophilia, high levels of acute

phase reactants (APR), pyuria and acute graft dysfunction with significantly elevated serum creatinine (4.5 mg/dL).
After sodium chloride %0.9 boluses, adrenaline infusion was started due to hypotension. Her PCR test revealed delta-variant SARS-CoV-2 with negative ELISA for both IgG and IgM. She diagnosed MISC based on the CDC criteria. However due to tenderness over graft, pyuria and very high levels of acute phase reactants, a contrast enhanced abdominal computed tomograpghy was performed, showing multiple hypodense wedge-like lesions and microabscess on graft kidney, heterogeneity of perirenal fat tissue. She was diagnosed with acute focal bacterial nephritis, an upper urinary tract infection in a spectrum between acute pyelonephritis and renal abscess.
Conclusions: When making the diagnosis of MIS-C, clinicians should focus on the criteria of "no alternative plausible diagnosis" as children mostly have mild form of the disease.

EP-35 SUCCESSFUL OBINUTUZUMAB AND DARATUMUMAB TREATMENT IN A PATIENT WITH STEROID RESISTANT NEPHROTIC SYNDROME RELAPSE POST-TRANSPLANT UNRESPONSIVE TO IMMUNOADSORPTIO

Cyrielle Parmentier[1], Jean Daniel Delbet[1], Antoine Mouche[1], Claire Herbez-rea[1], Veronique Baudoin[2], Claire Dossier[2], Julien Hogan[2], Tim Ulinski[1]

[1]Trousseau Hospital-Sorbonne University, [2]Robert Debré Hospital -aphp

Introduction: Idiopathic nephrotic syndrome relapse post-transplant unresponsive to immunoadsorption (IA) is a dilemma and no reliable treatment strategy has been identified to induce remission so far.
Material and methods: A 4- year-old girl presented first with pure nephrotic syndrome (NS). Initial biological analysis were not in favor for other renal pathologies. She did not reach remission after 30 days of oral steroids at 60 mg/m2 and she remained resistant to steroid pulses and combined tacrolimus/steroid treatment, to IV cyclosporine, rituximab and to 30 sessions of plasma exchange. Her renal function declined and hemodialysis was started. Bi-nephrectomy was performed as edema were still massive despite CKD stage 5. Ten months later she received an allograft from a deceased donor and idiopathic NS relapsed immediately post transplant. She did not respond to a combination of immunoadsorption (50 sessions), high dose tacrolimus treatment combined with oral high dose steroids and steroid pulses and mycophenolate mofetil. She received obinutuzumab 1g/1.73m2 injections weekly for one month and then daratumumab 1g/1.73m2 weekly for one month while IA sessions were continued.
Results: CD20 and CD38 were negative and proteinuria started to decrease 7 days after the 1st daratumumab injection and was negative 2 weeks later. We decreased IA sessions over 2 months without proteinuria relapse. 4 months later CD38 positive cells recovered in peripheral blood and nephrotic proteinuria reappeared. We performed a daratumumab and obinutuzumab reinjections leading to INS remission without considerable side effects. Graft function and all standard parameters are normal.
Conclusions: Anti CD20 and anti CD38 treatment seems to be a promising strategy in post TPL INS relapse without any response to standard treatment options.

EP-36 A SINGLE-CENTER EXPERIENCE: COVID-19 INFECTION IN PATIENTS FOLLOWED FOR CHRONIC KIDNEY DISEASE

Özgür Özdemir Şimşek[1], Gökçen Erfidan[1], Belde Kasap Demir[2], Demet Alaygut[3], Seçil Arslansoyu Çamlar[3], Fatma Mutlubaş[3]

[1]Department Of Pediatrics, Division Of Nephrology, University Of Health Sciences Izmir Tepecik Training And Research Hospital, Izmir, Turkey, [2]Department Of Pediatrics, Division Of Pediatric Nephrology And Rheumatology, Izmir Katip Çelebi University, Izmir, Turkey, [3]Izmir Faculty Of Medicine, Department Of Pediatrics, Division Of Nephrology, University Of Health Sciences Turkey, Izmir, Turkey

Introduction: Although renal involvement is observed in Covid-19 infection, the role of existing renal problems in morbidity and mortality has been demonstrated by studies. Here, we present our Covid-19 experience in patients followed for chronic kidney disease (CKD).
Material and methods: Medical records of 16 patients, who were followed up for CKD and found to be positive for covid-19 PCR in our hospital were reviewed retrospectively.
Results: This study included 16 patients (F/M:9/7). Sociodemographic, clinical and laboratory findings of the patients, treatment changes and treatments applied are shown in Table 1. An increase in creatinine was observed in all 5(%83) patients, except for one asymptomatic transplant patient, and hemodialysis was applied to one patient during this period. There was an increase in the need for dialysis in 2 patients who received dialysis, and hemodialysis was applied to the peritoneal dialysis patient during the infection period. An increase in creatinine was observed in 1 patient who was followed up only because of CAKUT among the other patients. Two patients who received renal replacement therapy and six patients with an increase in serum creatinine were hospitalized. No patient died. Patients whose signs of infection regressed and were found to be PCR negative were reverted to their previous treatments.

Table 1. The demographic, clinical and laboratory findings of the patients, treatment changes and treatments

COVID-19 PCR(+)	Renal Replacment Group (n=8) (Renal tx n=6, Periton Dx n=1, Hemodialysis n=1)	Other Group (n=8) (TINU n=2, cAKUT n=2, nephrotic syndrome n=4)
Presenting symptoms		
Fever	7	5
Cough	7	0
Weakness or myalgia	4	0
Headache	4	0
Gastrointestinal symptoms	1	0
Upper respiratory tract symptoms	3	0
Shortness of breath or hypoxia	1	0
Hypotension	1	3
Asymptomatic		
Severity of the disease		
Asymptomatic	1	3
Mild disease	4	5
Moderate disease	0	0
Severe disease	1	0
Critical illness	2	0
Abnormal radiologic findings	3	0
Lymphopenia (<1500 cells)	7	0
Respiratory support		
No respiratory support	7	0
Oxygen treatment	0	0
Mechanical ventilation	1	0
Drug treatment		
Favipiravir	7	1

Antibiotics	3	0
Oseltamivir	2	0
IVIG	2	0
Immunosuppressive modification		
Mycophenolate withdrawal	5	
	2	
Mycophenolate dose reduction	0	
	7	
TAC withdrawal or dose reduction		
Increase in steroid dose		
Hospitalization	6	1
ICU admission	2	0
Outcome		
Recovery	8	8
Death	0	0

Conclusions: 87% of transplant patients and all dialysis patients had immunosuppressive use. While the other group was hospitalized only because of high creatinine, there was a significant difference in the rate of hospitalization in transplant patients and patients who received dialysis treatment. Contrary to the literature, think that the use of immunosuppressive therapy and the use of renal replacement therapy may lead to an increase in morbidity from Covid-19 infection in patients.

EP-37 PREVALENCE OF HYPERTENSION USING AMBULATORY BLOOD PRESSURE MONITORING IN PAEDIATRIC KIDNEY TRANSPLANT RECIPIENTS TREATED WITH EARLY STEROID WITHDRAWAL IMMUNOSUPPRESSIVE REGIMES.

Thomas Dowsett, Mohan Shenoy

Royal Manchester Childrens Hospital

Introduction: To assess prevalence of hypertension and to assess BP control following kidney transplantation (KT) using ambulatory blood pressure monitoring (ABPM) in children initiated on an early steroid withdrawal immunosuppression regimen.
Material and methods: Cross sectional study of children followed up in a single centre transplant clinic from April 2021 -March 2022 age >5 years who have been commenced on an early steroid withdrawal immunosuppression regimen.
Results: A total of 50 patients completed successful 24-hour ABPM monitoring. The mean age was 13.7 years (range 6-18) and 65% of patients were male. The average time since KT was 3.6 years and the mean eGFR was 61ml/min/1.73m^2 (range 32-96). In the cohort 12% were overweight and 6% were obese. A total of 5% of patients were on antihypertensive medication (AHM) prior to KT. At the time of the study 18% were treated with AHM and 33% were on corticosteroids. A mean arterial pressure (MAP) of <50th centile was recorded in 62% of patients. Average systolic blood pressure (SBP) recordings were <50th centile for 82% (n=41) of patients and none had average SBP >90th centile. The average diastolic BP (DBP) was <50th centile in 53% patients with 30% and 9% having an average DBP in the 50-90th and >90th centile, respectively. A nocturnal dip in the MAP of >10% was observed in 59% of patients.
Conclusions: There appears to be a low prevalence of hypertension as determined by ABPM in children initiated on early steroid withdrawal immunosuppression. BP control in this population appears to be excellent with majority having MAP and systolic values <50th centile. Lack of nocturnal dip in BP is the most common abnormality.

EP-38 HEMOLYTIC UREMIC SYNDROME (HUS) IN A PEDIATRIC PATIENT WITH MULTISYSTEM INFLAMMATORY SYNDROME IN CHILDREN (MIS-C) ASSOCIATED WITH COVID-19

Ester Conversano, Federica Zotta, Francesco Emma, Marina Vivarelli

Division Of Nephrology And Dialysis, Bambino Gesù Hospital, Irccs, Rome, Italy

Introduction: There have been few reports of HUS triggered by Sars-CoV2 infection. We report a child who developed HUS concomitant to MIS-c associated with COVID-19.
Material and methods: A 1-year-old female presented in a Ukrainian hospital with fever, bloody diarrhea and lethargy. Laboratory investigation revealed anemia (8,7 g/dl), thrombocytopenia (63 10^3 /uL), acute kidney injury (creatinine 0,7 mg/dl) and signs of hemolysis. Urine analysis showed microscopic hematuria (153/pf) and proteinuria (6 g/l). C3 and C4 levels and ADAMTS13 activity were normal. The microbiological VTEC analysis tested negative. After a few days, the patient developed acute myocardial dysfunction and bilateral pneumonia. Laboratory findings showed elevated inflammatory markers, Pro-BNP and D-Dimers, along with positive IgG SARS-COV2 antibodies, consistent with MIS-c. Afterwards, neurological impairment and myocardial function worsened, leading to mechanical ventilation and extracorporeal membrane oxygenation. The patient was treated with intravenous immunoglobulins at dosage of 2 g/kg and heparin. On day 30, the patient was discharged with normal renal function, no significant proteinuria, mild hypertension and left ventricular ejection fraction of 50%. The patient was referred to Bambino Gesù Pediatric Hospital; genetic and functional testing for complement dysregulation was performed and are ongoing.
Results: There is a growing interest in the role of complement activation in Sars-CoV2-related endothelial dysfunction and pro-coagulative state. In a series of paediatric renal thrombotic microangiopathy (TMA) associated with MIS-c or acute SarsCoV-2 infection, higher serum C5b9 was associated with more severe renal damage. Moreover, two HUS patients concomitant to SarsCoV-2 infection responded to eculizumab. Genetic susceptibility for complement dysregulation was reported in an adult series of COVID-19-related renal TMA.
Conclusions: These findings postulate the role of COVID-19 as a trigger for complement-driven HUS and suggest the importance of genetic testing in patients with HUS secondary to COVID-19.

EP-39 INTERACTION BETWEEN COAGULATION AND COMPLEMENT IN MULTIORGAN SYSTEM FAILURE

Danko Milosevic[1], Zoltán Prohászka[2]

[1]University Of Zagreb, School Of Medicine, [2]Research Laboratory, 3rd Department Of Internal Medicine And Mta-se Research Group Of Immunology And Hematology, Hungarian Academy Of Sciences And Semmelweis University, Budapest, Hungary,

Introduction: Clostridium difficile colitis is a rare cause of life-threatening multi-organic failure in children. Several septic anticoagulant proteins (protein C, antithrombin, thrombomodulin, TFPI - tissue factor inhibitor) may be reduced due to reduced synthesis, consumption, or degradation of proteases (plasmin) in patients with disseminated intravascular coagulation (DIC).

Material and methods: In this example, we want to show the causal relationship between coagulation and the complement in multiorgan system failure.
Results: A fourteen-month-old boy with a severe, life-threatening illness was admitted to the intensive care unit due to a sudden onset of fever, hematemesis, hematuria, and bloody diarrhea with a rapid spread of hematoma and generalized edema. The diagnosis of DIC is supported by clinical presentation, elevated aPTT, and decreased antithrombin III with low platelet counts. Plasma fibrinogen was elevated alongside LDH, D-dimers, and hemoglobin, with a decreased haptoglobin and C3, indicating complement consumption. Significantly reduced C1q with negative anti-C1q reflects the presence of global complement activation. Massive complement consumption is indicated by decreased factor H, factor I, and factor B with alternative complement activation pathway activity (30%). ADAMTS13 metalloprotease (46%) excludes thrombotic thrombocytopenic purpura. Following administration of Eculizumab at the recommended dose on day 10 of illness, the childs general condition soon improved with the cessation of bloody diarrhea. The second occurrence of bloody diarrhea after day 5 was stopped with a subsequent dose of Eculizumab. The most compelling solution to the success of therapy in our patient is DIC with massive complement consumption that can result in sepsis-induced thrombotic microangiopathy. A rapid clinical and laboratory response (increase in platelets and C3) after administration of Eculizumab is consistent with this outcome.
Conclusions: Recent animal studies demonstrate the interaction between coagulation disorders, the complement system, and innate immunity during septic progression. Eculizumab therapy has never been used in children in patients with DIC to the best of our knowledge. We cannot recommend the use of Eculizumab in all patients with MOF and DIC. Instead, a personalized approach in patients with massive consumption of complement with short-term use of Eculizumab may be considered a temporary solution to eliminate the life-threatening effect of complement at that time.

EP-40 EVALUATION OF EPICARDIAL ADIPOSE TISSUE IN PEDIATRIC PATIENTS WITH CHRONIC KIDNEY DISEASES

ÖzgÜr Özdemir ŞimŞek[1], Tülay Demircan[2], Gökçen Erfidan[1], Seçil Arslansoyu Çamlar[3], Fatma Mutlubaş[3], Belde Kasap Demir[4], Demet Alaygut[3]

[1]Department Of Pediatrics, Division Of Nephrology, University Of Health Sciences Izmir Tepecik Training And Research Hospital, Izmir, Turkey, [2]Department Of Pediatrics, Division Of Cardiology, University Of Health Sciences Izmir Tepecik Training And Research Hospital, Izmir, Turkey, [3]Izmr Faculty Of Medicine, Department Of Pediatrics, Division Of Nephrology, University Of Health Sciences Turkey, Izmir, Turkey, [4]Department Of Pediatrics, Division Of Pediatric Nephrology And Rheumatology, Izmir Katip Çelebi University, Izmir, Turkey

Introduction: Cardiovascular diseases (CVD) are the most important causes of morbidity and mortality in chronic kidney disease (CKD). In addition to many reasons, factors such as inflammation and endothelial dysfunction are thought to be responsible for the increased CVD risk.
The aim of this study is to evaluate epicardial adipose tissue (EAT), which plays an active role in the development of CVD and atherosclerosis by secreting inflammatory substances in patients with CKD.
Material and methods: Retrospectively, the age at diagnosis, current disease status (eGFR), treatments used, renal replacement therapy and type, weight, height, BMI, mean blood pressure, presence of edema, laboratory parameters, and ECO findings were evaluated from the file data of patients with CKD (Group 1). EAT 1 (parasternal long axis) and EAT 2 (parasternal short axis) measurements were evaluated. Healthy screening ECHO cases were included as the control group (Group 2).
Results: The study and control groups consisted of 27 patients (14 males) and 15 patients (7 males), respectively. There was no difference between the groups in terms of gender, age, BMI. It was observed that 19 patients were followed up with Stage 3 and below CKD, and 8 patients with Stage 4-5 CKD. Peritoneal dialysis was applied to 6 patients. When both groups were evaluated in terms of ECO findings, a significant difference was found between the groups in terms of IVSD, LVPWD, Mitral a, LV mass, EAT 1 and some ECG parameters (Table 1).
Conclusions: Cardiovascular morbidity and mortality are high globally in renal diseases and early diagnosis of cardiovascular diseases is important in this group of patients. Measurement and tracking of EAT can give us information in this sense as a non-invasive parameter.

EP-41 CASE OF IGG-4 RELATED DISEASE WITH KIDNEY INVOLVEMENT IN CHILD. EFFICACY OF LONG TERM IMMUNOSUPPRESSIVE TREATMENT.

Alexey Tsygin[1], Svetlana Dmitrienko[1], Petr Ananin[1], Olga Vorobyova[2], Kirill Savostyanov[1], Tatiana Vashurina[1], Andrei Fisenko[1]

[1]National Medical And Research Centre For Childrens Health, Moscow, [2]National Centre For Clinical Morphology Diagnostics

Introduction: IgG4-related disease (IgG4-RD) is a rare immune mediated disease with multiple organ involvement with non-predominant kidney damage described mostly in adult population.
Material and methods: We report a case of 7 year old male with unremarkable family history, pregnancy and delivery.
Results: At first year of life recurrent episodes of diarrhea, once with bloody stool. Since 2 y.o. episodes of fever with elevated WBC, ESR, low grade anemia, microhematuria, pyuria and sterile urine culture. Normal GFR, slightly elevated bilirubin. Till 4 y.o. gradual decrease of GFR to 50 ml/min/1,73 m^2 with concomitant changes oh renal scan was observed. VUR was excluded, clinical exome sequencing didn't show any pathogenic variant. FEGDS – gastritis, jejunitis, US-scan – enlarged mesenterial lymph nodes.
Kidney biopsy at 4,6 years revealed: Diffuse severe chronic tubulointerstitial nephritis with high concentration of IgG4+ plasma cells (CD138+/IgG4+ >30 cells/hpf) and tubular basement membrane electron dense deposits; marked tubular atrophy and interstitial fibrosis (50%); global (56%) and secondary "perihilar" segmental (9%) glomerulosclerosis; without glomerular hypercellularity, crescents and arteriolosclerosis. Subsequently, 5-fold increase of serum IgG4 was found confirming IgG4-RD. This disease affects predominantly GI system, but kidney involvement previously was described in adults only.
Prednisolone 1 mg/kg was initiated and within 7 months the child become symptom-free with completely normal blood tests and GFR 88 ml/min/1,73 m^2 and normal serum IgG4. The dose was slowly tapered and after 1,5 years after initiation discontinued due to hypertension and growth problem. Control of serum IgG4 brought us to mofetyl mycophenolate initiation and this treatment lasts one year with increase of GFR to 102 ml/min/1,73 m^2.
Conclusions: Our observation suggests that IgG4-RD may be the cause of tubulointerstitial kidney damage in children responsive to immunosuppressive treatment. To elaborate optimal strategy multicenter studies are needed.

EP-42 EIGHT CASES OF SECONDARY PSEUDOHYPOALDOSTERONISM, A RARE CASE OF SALT WASTING SYNDROME IN INFANTS

Kenan DoĞan[1], Fatih Kilci[2], Merve AktaŞ ÖzgÜr[1], Mehmet Baha AytaÇ[1], Kenan Bek[1]

[1]*Kocaeli University School Of Medicine, Department Of Pediatrics, Division Of Pediatric Nephrology, Kocaeli, Turkey*, [2]*Kocaeli University School Of Medicine, Department Of Pediatrics, Division Of Pediatric Endocrinology, Kocaeli, Turkey*

Introduction: Secondary pseudohypoaldosteronism (PHA) is a rare salt-wasting syndrome that develops due to transient peripheral resistance to aldosterone. It usually presents with hyponatremia, hyperkalemia, and metabolic acidosis in infants. Here, we present eight infant cases who presented with severe hyponatremia and hyperkalemia and were diagnosed with secondary PHA.
Material and methods: Data were analyzed retrospectively from patient files. Adrenal precursor, renin and aldosterone values of the cases were standardized according to age and gender.
Results: Four of the all cases were boys. Median age was 45 days (9-90). Severe hyponatremia and hyperkalemia were the common laboratory features of the cases. Median serum sodium and potassium levels were 120.5 meq/L (108-126) and 6 meq/L (5.9-11.7), respectively. Although all cases had normal genital appearance, all of them were evaluated with the presumptive diagnosis of congenital adrenal hyperplasia. While serum adrenal precursors were normal, renin and aldosterone levels were high, suggesting PHA. All patients underwent urinary ultrasonography. All cases had urinary tract infection and/or urinary tract malformation (Six of them had urinary tract malformation and five of them had urinary tract infection), so they were diagnosed with secondary PHA. After the treatment (hydration and/or antibiotic) serum renin and aldosterone levels of patients were found to be within normal limits.
Conclusions: Secondary PHA should be kept in mind in cases of hyponatremia and hyperkalemia in infancy. Although congenital adrenal hyperplasia should be the first diagnosis to be excluded in the presence of concurrent hyponatremia and hyperkalemia in the infantile period, evaluation should also include urinalysis and renal imaging.

EP-43 ACUTE KIDNEY INJURY SECONDARY TO URINARY TRACT OBSTRUCTION, A RARE COMPLICATION OF CIRCUMCISION

Efrat Ben-shalom, Vardit Peles, Shimrit Tzvi-behr, Rachel Becker-cohen, Choni Rinat, Jenny Weinbrend-goichberg, Sapir Choshen, Yaacov Frishberg

Shaare Zedek Medical Center, Jerusalem, Israel

Introduction: Circumcision is prevalent worldwide. In Israel, over 90% of men are circumcised. Muslim boys are generally circumcised shortly after birth, and Jewish circmcision is performed on the eighth day of life. Complications are uncommon and are mainly surgical and infectious. Urinary tract obstruction is a rare complication of circumcision. The objective of this retrospective study was to define the incidence of urinary tract obstruction following circumcision in a single tertiary medical center, describe the characteristics of affected babies and review the outcome.
Material and methods: Study participants were identified from medical records of all male babies aged 0-30 days presenting at the pediatric emergency department during the years 2000-2020, with serum creatinine levels ≥ 1 mg/dl. Clinical and laboratory data were collected from the electronic records.]
Results: 8091 male post-circumcision newborn babies were admitted to the pediatric emergency room during the study period, 46 (0.57%) of them had elevated serum creatinine. We identified 10 babies (0.12%) with acute kidney injury attributed to urinary tract obstruction after circumcision. Average age at presentation was 10.1 days (8-13). Eight babies (80%) had a complicated postnatal course. The main findings on physical examination were distended abdomen, abdominal wall discoloration and leg edema. Average creatinine on admission was 1.76 mg/dl (1.0-3.28). Additional laboratory abnormalities were hyperkalemia 6.2 mEq/L (4.5-7.6) and hyponatremia 125 mEq/L (118-134). All hospitalized patients developed post-obstructive diuresis. All patients had complete resolution of kidney function and laboratory abnormalities. Six children (60%) had long term follow up, none had evidence of kidney damage.
Conclusions: Urinary tract obstruction with acute kidney injury is a rare severe complication of circumcision. Timely diagnosis and appropriate treatment can result in complete resolution of kidney function.

EP-44 CLINICAL-RADIOLOGICAL CORRELATION IN THE DIAGNOSIS OF ACUTE FOCAL BACTERIAL NEPHRITIS IN CHILDREN AND A NEW RADIOLOGICAL DIAGNOSTIC CRITERIA

Demet Alaygut[1], Ceren Sarioglu[2], GÖkÇen Erfİdan[1], ÖzgÜr Özdemİr ŞİmŞek[1], SeÇİl Arslansoyu Çamlar[1], Fatma MutlubaŞ[1], Belde Kasap Demİr[1]

[1]*University Of Health Sciences Tepecik Training And Research Hospital Department Of Pediatric Nephrology*, [2]*University Of Health Sciences Tepecik Training And Research Hospital Department Of Pediatric Radiology*

Introduction: Acute lobar nephronia (ALN), also known as acute focal bacterial nephritis, is a suppurative focal form of acute bacterial infection. Computed tomography (CT) is currently recognised as the most sensitive and specific imaging modality to diagnose ALN. Although it has a typical appearance, an objective, measurement-based diagnostic criteria has never been developed so far.
Material and methods: Patients followed up with the diagnosis of upper urinary tract infection and clinically indicated for CT were included in the study. The renal parenchymal involvement was defined as low-density areas in the kidney parenchyma on the contrast-enhanced CT scan. According to the morphological characteristics, the radiological diagnosis was categorized as acute pyelonephritis (APN) and ALN. The faint distinction of the hypodense areas from the normal renal parenchyma was accepted as the APN. The sharp distinction of the hypodense areas from the normal renal parenchyma was accepted as the ALN. The other characteristics of the hypodense areas were evaluated, including the site (right kidney/left kidney/bilaterally), the number (solitary/multiple), and the shape (wedge-like/non-wedge-like). For the quantitative analysis to differentiate ALN from the APN, the mean Hounsfield unit (HU) of the hypodense areas and those of adjacent normal kidney parenchyma were calculated on the contrast-enhanced CT scan. To eliminate variations in CT acquisition parameters and contrast material amount among patients, two formulas were used: Normal parenchyma HU – Hypodense area HU (Formul 1 (F1)) and Normal parenchyma HU / Hypodense area HU (Formul 2 F2))
Results: A total of 33 kidney units (15 APN (Group 1), 18 ALN (Group 2)) of 23 patients (18 ALN, 5 APN) were evaluated. Demographic and clinical results of the cases are shown in Table 1. When APN and ALN lesion HUs were evaluated, there was no difference between lesions and normal parenchyma, whereas F1 and F2 were different between both groups ($p=0.001$ and $p<0.001$, respectively) (Table 2) In the ROC analysis performed to predict that the lesion is ALN on CT, F1 had 83% sensitivity and 80% specificity for ≥ 49.5, while F2 ≥ 1.57 had 89% sensitivity and 80% specificity. There was no significant correlation between laboratory parameters and F1 and F2.

Table 1. Demographic, clinical and radiological characteristics of the patients

Parameters	
Age (month)	95,9 ± 58,5 (10-192)
Gender	6 (26,1%) M, 17 (73.9 %) F
Nationality	17 (73.9 %) Turkish 6 (26.1 %) Syrian
Highest fever	39,2 ± 0,42 (38,5-40,0)
Duration of fever (day)	3,17 ± 1,6 (1-7)
Laboratory data	
CRP (n=23)	181,7 ±81,7 (39-369)
Procalcitonin (n=21)	7,33 ± 8,8 (0,04-27,9)
WBC (n=23)	16.573 ± 5626 (8800-31900)
Neutrophil / Lymphocyte (n=23)	12,7 ±15,7 (0,94-69,5)
Albumin (n=23)	3,9± 0,4 (3,0-4,6)
Sodium (n=23)	133,17 ± 2,9 (130-140)
Urine kultur positivity	13 (56,5 %) 11 E.coli 2 Klebsiella pneumoniae
MERS	3
Lesion location on CT	Right kidney 11 (47,8%) Left kidney 11 (47,8 %) 1 Tx kidney
Number of lesions on CT	Multipl lesions 20 (87%) 3 single lesions (13%)

Table 2. Lesion HU values of APN and ALN and the difference between formulas

	APN (Group 1) N=15	ALN (Group 2) N=18	p
Lesion HU values	108,47 ± 51,16 (56-237)	84,11 ±32,5 (52-183)	0,063
Normal parenchymal HU values	155,2 ± 70,2 (93-364)	158,33 ±60,6 (104-356)	0,772
F1	46,73 ±24,1 (20-127)	74,2 ± 33,8 (37-173)	**0,001**
F2	1,46 ±0,15 (1,18-1,69)	1,91 ± 0,35 (1,41-3,05)	**< 0,001**

Conclusions: It is not easy to distinguish between ALN and APN clinically and radiologically. This study introduces an objective diagnostic criterion for the diagnosis of ALN.

EP-45 COMPARISON OF CASES WITH MODERATE AND SEVERE PRIMARY VESICOURETERAL REFLUX ASSOCIATED RENAL PARENCHYMAL INJURY

GÜlcan ErbaŞ[1], GÖkÇen Erfidan[2], ÖzgÜr Özdemir ŞimŞek[2], Cemaliye BaŞaran[2], Caner Alparslan[2], SeÇil Arslansoyu Çamlar[3], Demet Alaygut[3], Belde Kasap Demir[4], Fatma MutlubaŞ[3]

[1]University Of Health Sciences Izmir Tepecik Training And Research Hospital Department Of Pediatrics, [2]University Of Health Sciences Izmir Tepecik Training And Research Hospital Department Of Pediatric Nephrology, [3]University Of Health Sciences İzmir Faculty Of Medicine Department Of Pediatric Nephrology, [4]Izmir Katip Çelebi University Faculty Of Medicine Department Of Pediatric Nephrology And Rheumatology

Introduction: Primer Vesicoureteral Reflux (VUR) related renal parenchymal damage can be presented as congenital (CPD) or acquired parenchymal damage (APD). We aimed to compare the patients with VUR staged 3 or more according to type of parenchymal damage.
Material and methods: We analysed the records of the children who were followed-up in our center between 2010-2020 with primary VUR staged-3 (moderate) or 4-5 (severe). They were grouped as CPD, APD and normal parenchyma (NP).
Results: The study included 149 patients and 218 renal reflux units (RRU). The APD, CPD, NP groups included 42, 40, 67 patients and 48, 40, 130 RRUs, respectively. The APD had female predominancy (73.8%), while the CPD had male predominancy (72.5%) (p<0.001). The age of diagnosis was significantly higher in the APD (p<0.001). The diagnosis was coincidental in CPD, while mostly after urinary tract infection (UTI) in APD and NP groups (p<0.001). Severe VUR mostly diagnosed by antenatal hydronephrosis in CPD and NP groups, and by UTI in APD group (p=0.002). Getting UTI ≥2 times before diagnosis was most common in APD group, while no UTI was found before diagnosis at a rate of 67.5% in CPD (p<0.001). The main treatment modality for moderate VUR was continuous antibiotics prophylaxis (CAP) in NP, endoscopic subureteric transurethral injection in APD, and ureteroneocystostomy in CPD groups (p<0.001). Severe VUR patients were mostly followed up with CAP in the NP group and with surgical treatment in the APD and CPD groups (p<0.001). Moderate VUR patients did not have UTI under CAP in the NP, APD and CPD groups by the rate of 95.7%, 62.5% and 66.7%, respectively (p=0.017).
Conclusions: Moderate and severe VUR will present with normal or damaged renal parenchyma. Patients with CPD may emerge with different mechanisms and may show different clinical courses compare to APD.

EP-46 URINARY STONES IN CHILDREN: A SINGLE CENTER EXPERIENCE

Mesut Saygin[1], Secil Kezer[2], Cihangir Akgun[2], Onder Yavascan[2]

[1]Department Of Pediatrics, İstanbul Medipol University School Of Medicine, İstanbul, Turkey, [2]Division Of Pediatric Nephrology, Department Of Pediatrics, İstanbul Medipol University School Of Medicine, İstanbul, Turkey

Introduction: The incidence of urolithiasis is increasing related to environmental and nutritional factors nowadays. We aimed to evaluate the demographic and clinical features, etiologies, treatment approaches and follow-up of urolithiasis.
Material and methods: Medical records of children with urolithiasis were evaluated retrospectively in İstanbul Medipol Unıversity Hospital between 2018-2021. We performed preventive measures of stones (encouraged to drink a lot of extra fluids, eat less sodium, preventing excessive clothing that increases sweating) and medical treatment (alkalinization of urine) and in small stones (<5-7 mm and/or no causing symptoms or blocking the urinary tract). In patients with larger stones or block the urine flow underwent surgery treatments (lithotripsy, ureteroscopy or percutaneous nephrolithotomy).
Results: 176 patients with urolithiasis were evaluated retrospectively. 77 patients (43%) were girls and 99 patients (57%) were boys. Median follow time is 8.9 and median age of admission is 10 months. 48.9% of patients had a family history, 27.8% patient had urinary tract infection (UTI). We determined to stone associated with metabolic disorder in 27 (15.3%), urinary tract infection in 4 (2.3%) and urinary tract abnormality in 7 (4%) patients. In 138 (78.3%) patients, no reason was found in the

etiological assessment. Only preventive measures of stones were suggested in 63 (35.7%) patients. Medical treatment, shock wave lithotripsy, ureteroscopy and percutaneous nephrolithotomy were performed in 90 (50.8%), 10 (5.6%), 9 (5.1%) and 4 (2.2%) patients, respectively. Stone free rate was found to be 40.9% (72 patients). However recurrence rate was 2.3% (4 patients) after stone free period. We showed that decrease of stone size/number in 35 (19.9%), increase of stone size/number in 8 (4.5%) and no change in the size and number of stones in 13 (7.4%) patients.
Conclusions: The frequency of urinary system stone disease is increasing in children and it is one of the preventable causes of kidney damage. Kidney stones usually regress with adequate fluid consumption, proper nutrition advice and medication in childhood.

EP-47 MENDELIAN STEROID RESISTANT NEPHROTIC SYNDROME IN CHILDHOOD: IS IT AS COMMON AS REPORTED?

Zainab Arslan[1], Hazel Webb[1], Emma Ashton[2], Becky Foxler[1], Kjell Tullus[1], Aoife Waters[1], Detlef Bockenhauer[1]

[1]Great Ormond Street Hospital For Children, London, Uk, [2]Ne Thames Regional Genetics Laboratory, Gosh Nhs Foundation Trust, London, United Kingdom

Introduction: Primary steroid resistant nephrotic syndrome (SRNS) is thought to have either genetic or immune-mediated aetiology. Children with a genetic cause of SRNS tend to not respond to immunosuppression and usually progress to chronic kidney failure in childhood. Some forms of genetic SRNS allow for targeted therapies. Knowing which children to screen for genetic causes can be difficult. Numerous studies have described the prevalence of genetic causes of primary SRNS to be between 30-40%, but their cohorts include children with congenital and infantile NS and familial cases. We aimed to look at the prevalence of a Mendelian cause for children 1-year of age or older with sporadic SRNS.
Material and methods: Retrospective electronic patient record analysis of all children with sporadic SRNS who presented at our centre over the last 15 years was undertaken. All statistical analysis were performed using SPSS Version 27.
Results: Of the 49 children who met the inclusion criteria, 5 (10%) had causative variants identified, predominantly in NPHS2. None responded to immunosuppression. Of the 44 (90%) who had no genetic cause identified, 33 (75%) had complete or partial remission after commencing second-line immunosuppression and 67% of these had eGFR >90 ml/min/1.73m^2 at last clinical follow-up. Of the children who did not respond to immunosuppression, 64% progressed to kidney failure.
Conclusions: Those with identified genetic cause were significantly (p= 0.003) less likely to respond to immunosuppression and more likely (p=0.026) to progress to chronic kidney disease. Understanding the genetics along with response to immunosuppression informs management in this cohort of patients.

EP-48 HEMODIALYSIS IN CHILDREN WITH BODY WEIGHT < 12KG: A SINGLE CENTRE EXPERIENCE

Varvara Askiti, Georgia Malakasioti, Argyroula Zampetoglou, Andromachi Mitsioni

Pediatric Nephrology Department, Childrens'hospital P&a Kyriakou", Athens, Greece

Introduction: Despite recent technological advances in pediatric nephrology, haemodialysis in infants is associated with significant morbidity and mortality. Due to the limited number of infants on haemodialysis worldwide, there are only few studies on this field. We aimed to describe our experience in children with End Stage Renal Failure (ESRF) and body weight (BW) <12 kg treated with haemodialysis in the last 8 years in regards to outcomes, morbidity and complications.
Material and methods: 11 patients were included in this study (54% boys), with BW 5.8 – 12 kg and age 0.71 – 4.9 years (Median: 2.1 years). 37% had a diagnosis of congenital nephrotic syndrome, 36% had corticosteroid-resistant nephrotic syndrome, 18% had renal hypoplasia/dysplasia and 9% hyperoxaluria. 40% of the patients had residual renal function. All patients were treated with at least 3 sessions of haemodialysis per week lasting 4-5 hours.
Results: The main problem was vascular access. All patients had a permanent double lumen jugular vein catheter. The mean catheter survival time was 31.5 months (8.8-53.1). Each patient had approximately 3 different catheter insertions (0.46/patient/year). The main indications for catheter removal were mechanical problems (90%) and infections (10%) with most common bacteria isolated Staph. Aureus and Staph. Epidermidis. The mean number of catheter related infections was 0.35/patient/year (0-1.21). Other complications included hemodynamic instability during hemodialysis, hyperkalemia, seizures, allergic reactions, extracorporeal circuit thrombosis. In all children weight and height z scores improved (50% of the patients were on growth hormone treatment). During the follow up period, 2 patients (18%) received a renal transplant, 7 patients (64%) continue on hemodialysis and 2 patients (18%) changed dialysis mode due to vascular access problems.
Conclusions: Haemodialysis in experienced centres is safe for children with BW<12 kg. Vascular access problem is the main complication but longevity can be prolonged with good line care.

EP-49 A CASE OF COMPOUND HETEROZYGOUS MUTATIONS IN SLC34A3 CAUSING HYPOPHOSPHATEMIC RICKETS WITH HYPERCALCIURIA

Michela Mariapia Gritti, Andrea Puma, Sara Picassi, Chiara Tosolini, Laura Venditto, Milena Brugnara

Pediatria C, Odb Verona

Introduction: We report a case of hypophosphatemic rickets with hypercalciuria caused by compound heterozygous mutations in SLC34A3 gene in an extremely preterm girl with horseshoe kidney.
Material and methods: Our patient is a girl born extremely preterm at 23 gestational weeks with a birth weight of 440 g. She developed severe bronchopulmonary dysplasia needing steroids and diuretics for the first months of life. Our patient has horseshoe kidney and since the first months of life her renal ultrasounds have shown bilateral nephrocalcinosis with consequent pyelectasia. Bone densitometry detected osteopenia, worsening at the following checks. Laboratory tests revealed hypercalcemia, hypercalciuria, in the presence of low values of PTH and of 25-hydroxyvitamin D. To the contrary, calcitriol was highly increased and the patient showed hypersensitivity to the treatment with vitamin D. Genetic testing for CYP24A1 mutations resulted negative. Over time laboratory tests have shown a trend to low levels of phosphataemia and incostant phosphate reabsorption. Nephrocalcinosis has been significant and the patient underwent lithotripsy at the age of ten. The genetic testing was then widened with clinical exome sequencing.
Results: Clinical exome sequencing revealed compound heterozygous mutations c.1304delG and novel c.986_1000del in SLC34A3 gene which encodes the sodium-phosphate cotransporter NaPi2a. These mutations cause FGF23 suppression and consequent calcitriol increase. The clinical features described for homozygous mutations are similar to our patient's. Therapy consists in phosphorus supplementation.

Conclusions: Nephrocalcinosis and rickets with hypercalciuric hypercalcemia and low levels of PTH and 25-hydroxyvitamin D may be consistent with mutations of tubular phosphate transporter, even without clear alterations of phosphataemia and phosphaturia. The extreme prematurity of our patient acted as a confounding factor but the genetic testing has been diagnostic and suggested a targeted therapy with good prognosis.

EP-50 ATYPICAL POST-INFECTIOUS GLOMERULONEPHRITIS AND C3 GLOMERULOPATHY: EXPANDING THE SPECTRUM

Luca Antonucci, Antonio Gargiulo, Alessandra Gianviti, Francesca Diomedi-camassei, Francesco Emma, Marina Vivarelli

Bambino Gesù Children Hospital

Introduction: Post-infectious glomerulonephritis (PIGN) is the most common and benign hypocomplementemic nephritis in children. However, cases of atypical PIGN (aPIGN) with prolonged C3 consumption and/or atypical membranoproliferative (MPGN) pattern have been reported (Sethi, 2013). In this setting, C3 glomerulopathy (C3G) is the main differential diagnosis. Dysregulation of alternative complement pathway is a well-known pathogenetic mechanism of C3G. However, similar alterations have been reported even in aPIGN, suggesting a disease spectrum. Our case report is paradigmatic of this concept.
Material and methods: A 5-year-old child presented persistent macrohematuria since 4 weeks earlier, during an upper airways infection. Blood pressure was normal, blood tests showed normal renal function, C3 hypocomplementemia, and urinary protein creatinine ratio (UPCR) of 1.7 mg/mg. C4, immunoglobulin and autoantibodies were normal: PIGN was suspected. At 6 weeks, macrohematuria persisted, C3 was still low and UPCR was 2 mg/mg. Kidney biopsy was performed.
Results: Histological picture showed a light microscopy MPGN pattern and isolated C3 immunofluorescence compatible with C3G. By electron microscopy, electron dense deposits were mainly mesangial and subendothelial, but also intramembranous, as seen in dense deposit disease. Moreover, numerous humps were present. C3 glomerulonephritis was diagnosed. Therapy with oral prednisone and ramipril was started and led to rapid normalization both of UPCR (in 2 weeks) and of C3 levels (in 4 weeks).
Conclusions: In our case, acute clinical course (in total 10-12 weeks from onset to remission) with rapid response to prednisone and numerous humps suggest a PIGN diagnosis, while intrainfectious macrohematuria, prolonged C3 reduction, MPGN pattern with heterogeneous distribution of electron dense deposits, especially intramembranous, suggest C3G. This clinical and histological picture of true atypical PIGN show that, in some cases, a distinction between PIGN and C3G is difficult and, probably, a spectrum of forms, between the purely acute self-limiting and the chronic forms, exists.

EP-51 THE PREVALENCE AND RISK FACTORS FOR ARTERIAL HYPERTENSION IN CHILDREN WITH IDIOPATHIC NEPHROTIC SYNDROME

Piotr Skrzypczyk[1], Anna Maria Wabik[1], Anna Deja[2], Michal Szyszka[2], Anna Ofiara[1], Malgorzata Panczyk-tomaszewska[1]

[1]*Department Of Pediatrics And Nephrology, Medical University Of Warsaw*, [2]*Department Of Pediatrics And Nephrology, Doctoral School, Medical University Of Warsaw*

Introduction: Arterial hypertension (AH) is supposed to be uncommon in children with idiopathic nephrotic syndrome (INS). The study aimed to analyze prevalence and risk factors of AH in children with INS.
Material and methods: In 153 children with INS (9.32±3.84 years, 100 boys), we evaluated systolic and diastolic blood pressure (mm Hg, Z-scores), age at onset of the disease, anthropometric data, response to steroids, results of kidney biopsy, number of INS relapses, and medications.
Results: Mean age at disease onset was 4.39±2.38 years, 67 patients had SSNS, 12 FRNS, 58 SDNS, and 16 SRNS, kidney biopsy was performed in 26: 5 MCD, 11 mesangial proliferation, 10 FSGS. AH at disease onset was found in 10 (6.5%) patients, after prednisone initiation in following 11 (7.2%). After observation period of 4.93±3.80 years, AH was found in 31 (20.3%) children: in 3 (4.5%) patients with SSNS, 2 (16.7%) with FRNS, 20 (34.5%) with SDNS, 6 (37.5%) with SRNS. Patients with AH had lower height Z-score (-0.46±1.20 vs. 0.18±1.15, p=0.006), higher BMI Z-score (1.15±1.10 vs. 0.41±1.09, p<0.001), number of INS relapses (8.32±6.78 vs. 4.41±3.82, p<0.001), and current prednisone dose (0.49±0.67 vs. 0.18±0.29 [mg/kg/24h], p<0.001). Systolic blood pressure Z-score at the end of observation correlated with age (r=-0.192, p=0.017), BMI Z-score (r=0.368, p<0.001), and present prednisone dose [mg/kg/24h] (r=0.186, p=0.021), diastolic blood pressure Z-score with BMI Z-score (r=0.231, p=0.004), and number of INS relapses (r=0.163, p=0.044). In Cox Proportional Hazard, presence of SDNS and SRNS were the only predictors of AH (SDNS: HR=4.80, 95CI(1.30-17.75), SRNS: HR=4.77, 95CI(1.06-21.53)).
Conclusions: 1. Approximately one-third of patients with SDNS and SRNS develop arterial hypertension in the course of the disease.
2. Steroid dependence and steroid resistance are the strongest predictors of AH in children with INS. Other determinants of blood pressure elevation in this group of patients are: high BMI and high number of relapses.

EP-52 MOLECULAR ANALYSIS IN CHILDREN WITH SEVERE HYPERTENSION SECONDARY TO MID AORTIC SYNDROME

Ester Conversano[1], Laura Lucchetti[1], Federica Zotta[1], Laura Massella[1], Ugo Giordano[2], Massimo Rollo[3], Mario Giordano[4], Silvia Morlino[5], Marina Vivarelli[1], Francesco Emma[1], Marco Spada[6]

[1]*Division Of Nephrology And Dialysis, Bambino Gesù Hospital, Irccs, Rome, Italy*, [2]*Arterial Hypertension Unit, Bambino Gesù Hospital, Rome, Italy*, [3] Department Of Interventional Imaging And Radiology, Bambino Gesù Hospital, Irccs, Rome, Italy, [4]*Nephrology Unit, Giovanni Xxiii Pediatric Hospital, Bari, Italy*, [5]*Medical Genetics, House For Relief Of Suffering, S. Giovanni Rotondo, Foggia, Italy*, [6]*Division Of Hepatobiliopancreatic Surgery And Liver And Kidney Transplantation, Bambino Gesù Hospital, Irccs, Rome, Italy*

Introduction: Mid Aortic Syndrome (MAS) is a rare vascular abnormality characterized by segmental coarctation of the abdominal aorta associated with variable involvement of aortic branches. MAS presents with severe hypertension (HNT), requiring multiple treatments. About 60% of cases are idiopathic, whereas 40% are secondary to great vessels vacuities or associated with genetic disease.
Material and methods: We describe five patients with MAS referred to Bambino Gesù Childrens Hospital, one of them followed up together with Giovanni XXIII Pediatric Hospital (table).
Results: All the patients had severe early-onset HNT. At least two drugs were needed in all but one case. Patients 1, 2 and 5 required renal artery angioplasty, not allowing any anti-HTN treatment reduction. Patient 5 developed uncontrolled HNT, left ventricular hypertrophy and hypertensive retinopathy. Renal auto-transplantation and resection of the stenotic renal artery tract was performed, leading to a significant improvement in HTN control. In all our patients, MAS was associated with a genetic disorder.

Patient (N°)	1	2	3	4	5
Age at onset (yrs)	6	5	6	7	6
Genetic findings	FBN2 (VUS)	KRAS mosaic	NF1	NF1	FBN2
Vascular abnormalities (stenosis)	Supra-renal aorta, bilateral renal artery (left post-stenotic aneurysm), inferior mesenteric artery	Sub-renal aorta, bilateral renal artery; inferior mesenteric artery	Renal aorta, bilateral renal artery, celiac tripod, superior mesenteric artery	Renal aorta, bilateral renal artery (right post stenotic aneurysm)	Renal aorta, bilateral renal artery; ectasia of ascending aorta; bicuspid aortic valve
Drugs (N°)	2	3	3	1	4
Follow up (yrs)	3	9	15	8	1
Endovascular/ surgical treatment	Percutaneous angioplasty	Percutaneous angioplasty	-	-	Percutaneous angioplasty Renal auto-transplant
Reduction of anti-HTN treatment	-	-	-	-	1 drug stopped, 1 reduced dosage

Conclusions: MAS secondary to a genetic disease should be suspected in children with very early onset HNT. Endovascular treatment may fail, and renal auto-transplantation can be proposed successfully.

EP-53 COMPARISON OF GRAFT SURVIVAL AFTER PEDIATRIC KIDNEY TRANSPLANTATION FROM LIVING AND DECEASED DONORS: 2007-2018 FRENCH COHORT

Manon Aurelle[1], Emilie Savoye[2], Myriam Pastural[2], Julie Bernardor[15], Olivia Boyer[4], Sylvie Cloarec[9], Stéphane Decramer[7], Olivier Dunand[12], Marc Fila[14], Florentin Garaix[13], Jérôme Harambat[6], Julien Hogan[3], Annie Lahoche[10], Gwenaelle Roussey[5], Isabelle Vrillon[11], Ariane Zaloszyc[8], Justine Bacchetta[1], Anne-laure Leclerc[1], Bruno Ranchin[1]

[1]Department Of Pediatric Nephrology, Hospices Civils De Lyon, Centre De Référence Maladies Rénales Rares-néphrogones, Filière Orkid, Lyon, France, [2]Agence De La Biomédecine, Direction Prélèvement Greffe Organes-tissus, Saint-denis La Plaine, France, [3]Department Of Pediatric Nephrology, Hôpital Robert-debré, Aphp, Paris, France, [4]Department Of Pediatric Nephrology, Reference Center For Idiopathic Nephrotic Syndrome In Children And Adults, Imagine Institute, Paris University, Necker Hospital, Aphp, Paris, France, [5]Pediatric Department, Nantes University Hospital, Nantes, France, [6]Department Of Pediatrics, Pediatric Nephrology Unit, Centre De Référence Des Maladies Rénales Rares Du Sud-ouest, Sorare, Bordeaux University Hospital, Bordeaux, France, [7]Department Of Pediatric Internal Medicine, Rheumatology And Nephrology, Centre De Référence Des Maladies Rénales Rares Du Sud-ouest, Sorare, Toulouse University Hospital, Toulouse, France, [8]Department Of Pediatric Nephrology, Hôpitaux Universitaires De Strasbourg, Strasbourg, France, [9]Department Of Pediatric Nephrology, Hôpital Bretonneau Et Hôpital Clôcheville, Chu Tours, 2 Bd Tonnellé, 37044, Tours Cedex, France, [10]Department Of Pediatrics, Nephrology Unit, Centre Hospitalier Régional Universitaire Lille, Lille, France, [11]Pediatric Department, Centre Hospitalier Universitaire Nancy, Nancy, France, [12]Department Of Pediatric Nephrology, Centre Hospitalier Universitaire La Réunion, Réunion, France, [13]Pediatric Department, Hôpital De La Timone, Marseille, France, [14]Pediatric Nephrology Department – Chu Arnaud De Villeneuve - Montpellier, [15]Department Of Pediatric Nephrology, Chu De Nice, Hôpital Archet,151 Route Saint-antoine De Ginestière, 06200 Nice, France

Introduction: It is usually recognized that living donor (LD) kidney transplantation (KT) has better long-term results than deceased donor (DD) KT. We compared graft survival according to the type of donor (DD or LD) in the most recent French pediatric cohort.

Material and methods: We included all pediatric patients who received a first isolated KT in France between 2007 and 2018. The primary endpoint was graft survival at 5 years. We also described and compared the duration of dialysis, causes of kidney failure, HLA, EBV and CMV matching, donor characteristics and ischemia times in the 2 groups (Mean±SD).

Results: 852 and 191 patients were included in the DD and the LD groups, respectively. The age at transplantation was slightly lower in DD group: 11.1±5.0 vs 12.0±4.4 years (p=0.01). The 5-year renal graft survival was 89% (CI95%: 87-91) and 89% (CI95%: 83-93%) in the DD and LD groups (p=0.678), respectively. Similar results with death censoring were observed: 90% (CI95%: 88-92%) versus 91% (CI95%: 85-94%). In the LD group, there were more preemptive transplants (52.2 vs 23.6%, p<0.01), but the same duration in dialysis before KT among dialyzed patients (18.9±25.1 vs 19.2±19.6 months, p=0.94), a shorter waiting time (6.5±6.8 vs 10.5±10.5 months) (p<0.01), shorter cold (3.3 ±2.9 vs 15.9±5.1 hours) and warm ischemia times (48±24 vs 54±36 minutes) (p<0.01 and p=0.01), older donor (42.4±6.9 vs 15.2±7.7 years) (p<0.01) and better HLA matching (mismatches A: 0.8 vs 1.2, B: 0.9 vs 1.4, DR: 0.7 vs 0.8 and DQ: 0.6 vs 0.7 (p<0.01).

Conclusions: The 5-year graft survival is currently the same between DD and LD in the most recent French pediatric cohort. The next step is to compare these results with the 1995-2006 pediatric cohort to analyze factors associated with graft loss in both groups and in both cohorts.

EP-54 A CASE OF POLYCYSTIC KIDNEY DISEASE WITH ADDITIONAL ABNORMALITIES: HOW DEEP SHOULD WE DIVE INTO GENES?

Ivan Jakopčić, Hana Matković, Maja Ban, Maša Davidović, Lovro Lamot, Ivanka Kos, Kristina Vrljičak

Division Of Pediatric Nephrology, Dyalisis And Transplantation, Department Of Pediatrics, University Hospital Center Zagreb, University Of Zagreb School Of Medicine, Zagreb, Croatia

Introduction: Polycystic kidney disease (PKD) is a genetic disorder with a variable clinical presentation dominantly characterized by cystic expansion of the kidney in addition to various extrarenal manifestations, most commonly cysts and aneurysms. Therefore, when other abonrmalities are present, expanded genetic testing might indicate an additional underlying cause.

Material and methods: Case report
Results: We present the case of a 16 months old girl with arterial hypertension and polycystic kidney disease. She was born premature with unremarkable gestation. Shortly after birth she developed lower extremities edemas, oligury and hyponatremia. From the second day of her life increased blood pressure was measured. Renal ultrasound revealed large kidneys with many small cysts in parenchyma while echocardiogram indicated hypertrophy of interventricular heart septum. Due to involvement of various organ systems at the age of 11 months comprehensive genetic testing was performed. Sequence analysis using the Blueprint Genetics (BpG) Whole Exome Plus identified a heterozygous nonsense variant PKHD1 c.9319C>T, p.(Arg3107*), a heterozygous missense variant PKHD1 c.4882C>G, p.(Pro1628Ala), and a heterozygous missense variant PTPN11 c.178G>A, p.(Gly60Ser). The two former variants are classified as pathogenic and the latter as likely pathogenic.
Conclusions: While mutations in PKHD1 gene are related to defects in cilia-mediated signaling activity resulting in cyst formation and recessive form of PKD, PTPN11 muatations have been associated with dysregulation of the RAS-MAPK signaling pathway recognized as the molecular cause underlying a group of clinically related developmental disorders with features including reduced growth, facial dysmorphism, cardiac defects, ectodermal anomalies, variable cognitive deficits, and susceptibility to certain malignancies. Consequently, there is a wide range of possible genotypic-phenotypic correlations arising from those mutations, previously undescribed in the same patient.

EP-55 A RARE CAUSE OF TROMBOTIC MICROANGIOPATHY IN A BOY OTHER THAN HEMOLYTIC UREMIC SYNDROME

GÖkÇen Erfidan[1], ÖzgÜr Özdemir ŞimŞek[1], SeÇil Arslansoyu Çamlar[2], BurÇak Tatli GÜneŞ[3], Demet Alaygut[2], Fatma MutlubaŞ[2], Belde Kasap Demir[4]

[1]University Of Health Sciences, Izmir Tepecik Training And Research Hospital, Department Of Pediatric Nephrology, [2]University Of Health Sciences, Izmir Faculty Of Medicine, Department Of Pediatric Nephrology, [3]University Of Health Sciences, Izmir Tepecik Training And Research Hospital, Department Of Pediatric Hematology And Oncology, [4]Izmir Katip Celebi University, Faculty Of Medicine, Department Of Pediatric Nephrology And Rheumatology

Introduction: Hemolytic uremic syndrome (HUS) is one of the common causes of acute kidney injury characterized by trombotic microangiopathy (TMA) secondary to endothelial damage.
Material and methods: Classical triad is non-immune hemolytic anemia, thrombocytopenia, and renal impairment. The presentation may rarely overlap with the other forms of hemolytic anemia.
Results: An African 15-month-old boy applied with fever, vomiting, and watery diarrhea. He had no history of a chronical illness, drug USAge or hospitalization. On physical examination, he had tachycardia (154 beat/min), fever (38.5ºC), hypertension (systolic-99p, diastolic-96p) and signs of dehydration. Sodium chloride %0.9 bolus (20 cc/kg) was started. Meanwhile, the laboratory examination showed anemia (Hemoglobin [Hb]:3.8 gr/dL), thrombocytopenia (Plt:17x10^3/uL), high lactate dehydrogenase (LDH:919 U/L) with impaired renal function (Urea:43 mg/dL, Creatinine:0.7 mg/dL). Peripheral blood smear showed schistocytes (3%). C3 was found decreased (0.47 g/L), while C4, ADAMTS13, haptoglobulin and homocysteine were normal. Direct coombs was negative. He had been diagnosed with HUS. Shigatoxin was found negative. Symptomatic treatment was started. During the follow-up, urine output did not decrease at all. After eight days of hospitalization, he discharged healthily (Hb:10.9 gr/dL, Plt:218x10^3/uL, LDH:376 U/L, Urea: 17 mg/dL, Creatinine: 0.4 mg/dL). After seven months, he presented with fever and decreased appetite. Physical examination revealed signs of dehydration and significant hepatosplenomegaly. He had severe anemia (Hb:3.4 gr/dL), thrombocytopenia (Plt:63x10^3/uL), high LDH (569 U/L) and normal kidney functions (urea:14 mg/dl, creatinine:0.4 mg/dL). Peripheral blood smear revealed anisocytosis, poikilocytosis, polychromasia, schistocytes (3-4%) and sickle cells. Direct coombs test was positive. Hemoglobin electrophoresis showed HBF:13.1%, HBA2:3.3%, HBS:83.1%, HBA:0.5%. He diagnosed with sickle cell disease.
Conclusions: Sickle cell disease is a common inherited hemoglobinopathy that may complicate with hyperhemolytic and vasooclusive crisis. Although TMA associated with sickle cell disease is a rare entity, there have been reported cases in literature. It should be considered in the differential diagnosis of HUS.

EP-56 EFFICACY OF TREATMENT OPTIONS OF IGA NEPHROPATHY IN CHILDREN – NATIONAL STUDY

Malgorzata Mizerska-wasiak[1], Agnieszka Such-gruchot[1], Karolina Cichoń-kawa[1], Jadwiga Małdyk[2], Monika Miklaszewska[3], Dorota Drożdż[3], Agnieszka Firszt-adamczyk[4], Roman Stankiewicz[4], Beata Bieniaś[5], Przemysław Sikora[5], Agnieszka Rybi-szumińska[6], Anna Wasilewska[6], Maria Szczepańska[7], Magdalena Drożyńska-duklas[8], Aleksandra Żurowska[8], Agnieszka Pukajło-marczyk[9], Danuta Zwolińska[9], Łukasz Obrycki[10], Sylwia Szymańska[11], Jacek Zachwieja[12], Monika Pawlak-bratkowska[13], Marcin Tkaczyk[13], Malgorzata Pańczyk-tomaszewska[1]

[1]Department Of Pediatrics And Nephrology, Medical University Of Warsaw, Poland, [2]Department Of Pathology, Medical University Of Warsaw, Poland, [3]Department Of Pediatric Nephrology, Jagiellonian University, Cracow, Poland, [4]Department Of Pediatrics And Nephrology, Ludwik Rydygier Hospital, Toruń, Poland, [5]Department Of Pediatric Nephrology, Medical University Of Lublin, Poland, [6]Department Of Pediatrics And Nephrology, Medical University Of Bialystok, Poland, [7]Department Of Pediatrics, Smdz In Zabrze, Silesian Medical University, Katowice, Polan, [8]Department Of Pediatrics, Nephrology And Hypertension, Medical University Of Gdańsk, Poland, [9]Department Of Pediatric Nephrology, Wroclaw Medical University, Poland, [10]Department Of Nephrology, Kidney Transplantation And Hypertension, Children's Memorial Health Institute, Warsaw, Poland, [11]Department Of Pathology Children's Memorial Health Institute, Warsaw, Poland, [12]Department Of Pediatric Nephrology And Dialysis, Medical University Of Poznań, Poland, [13]Department Of Pediatrics, Immunology And Nephrology, Polish Mothers Memorial Hospital Research Institute, Łódź, Poland

Introduction: IgA nephropathy (IgAN) is the most common glomerulonephritis worldwide. The treatment of IgAN in children is still under debate.
Material and methods: We studied 199 patients with IgAN diagnosed by renal biopsy with predominantly IgA deposits from the Polish Pediatric IgAN Regisrty, which includes IgAN diagnosed since 2000.
In all patients, the following were analyzed: clinical symptoms of disease onset and the end of follow-up (proteinuria mg/kg/day, hematuria, GFR, IgA, C3), renal biopsy result with Oxford classification (MEST-C) and treatment used as: R- renoprotection alone (ACEI/ARB), S- steroid therapy combined with renoprotection, AZA- Azathioprine with steroids and renoprotection, IS- other immunosuppressive treatment (Cyclophosphamide, Cyclosporine A, Mycophenolate mofetil), combined with steroid therapy and renoprotection.
Results: Mean age of onset was 11.74 ± 4.11 years, proteinuria 14.05 (5-967) mg/kg/day, GFR 97.25 ± 31.34ml/min.
Time to kidney biopsy 0.4 (0-13) years; follow up period was 3.03 (0.1-15.48) years.

R was used in 50%, S in 22.3%, AZA 31.5%, IS in 22.3%.
Analysis of treatment effect showed significantly higher GFR follow (p=0.03), proteinuria follow NS in children from AZA group than R. Children treated with AZA compared to IS showed significantly lower proteinuria follow (p=0.037) and higher GFR (NS); comparison of AZA to S showed GFR higher (NS), proteinuria NS. In AZA group MESTC>3 was found significantly more often than in R group (45.45%vs1.96%) and significantly more often than in P group (44.12%vs14.29%).
Kaplan-Meyer analysis showed significantly better renal survival with normal GFR in children in gr AZA than in gr R (10-year survival 63% vs 31%), despite worse Oxford classification scores.
Conclusions: Analysis of long-term outcomes in children with IgAN demonstrates significant benefits for maintaining normal renal function from treatment with combination therapy with steroids and AZA and renoprotection compared to R alone. Analysis of treatment with other immunosuppressive drugs, requires studies on larger groups of patients.

EP-57 EXOME SEQUENCING IN INDIVIDUALS WITH CONGENITAL ANOMALIES OF THE KIDNEY AND URINARY TRACT (CAKUT): A SINGLE-CENTER EXPERIENCE

Jasmina Ćomić[1], Korbinian M. Riedhammer[1], Velibor Tasic[2], Jovana Putnik[3], Nora Abazi-emini[2], Aleksandra Paripovic[3], Natasa Stajic[3], Valbona Nushi-stavileci[4], Matthias C. Braunisch[5], Julia Hoefele[1]

[1]Institute Of Human Genetics, Klinikum Rechts Der Isar, Technical University Of Munich, School Of Medicine, Munich, Germany, [2]University Children's Hospital, Medical Faculty Of Skopje, Macedonia, [3]Institute For Mother And Child Health Care Of Serbia "dr Vukan Čupić", Department Of Nephrology, University Of Belgrade, Faculty Of Medicine, Belgrade, Serbia, [4]Pediatric Clinic, University Clinical Center Of Kosovo, Prishtina, Kosovo, [5]Department Of Nephrology, Klinikum Rechts Der Isar, Technical University Of Munich, School Of Medicine, Munich, Germany

Introduction: Individuals with congenital anomalies of the kidney and urinary tract (CAKUT) show a broad spectrum of malformations. CAKUT can occur in an isolated fashion or as part of a syndromic disorder and can lead to end-stage kidney disease (ESKD). A monogenic cause can be found in approximately 12% of affected individuals.
Material and methods: 86 unrelated individuals with CAKUT were analyzed by exome sequencing (ES). Prioritized rare variants were rated according to the recommendations of the American College of Medical Genetics and the Association for Clinical Genomic Science. Clinical data were collected using a standardized questionnaire.
Results: In the study cohort, 7/86 individuals had a (likely) pathogenic variant in PAX2, PBX1, EYA1 or SALL1 gene. Additionally, in one individual, a chromosome 17q12 deletion syndrome (including the HNF1B) was detected. 62 individuals had a kidney affection, 36 of them bilateral. All solved cases (8/86, 9%) had bilateral kidney affection.
Conclusions: Although the diagnostic yield in CAKUT cohorts is low, our single-center experience argues, that, in individuals with bilateral kidney affection, genetic diagnostics should be considered.

EP-58 EFFECTS OF LONG-TERM CYCLOSPORINE A TREATMENT IN CHILDREN WITH NEPHROTIC SYNDROME

Gutting Miriam Lerch Christian Nele Kanzelmeyer

[1]Hannover Medical School, Pediatric Nephrology, Germany, [2]Pediatric Practice, Garbsen, Germany

Introduction: Idiopathic nephrotic syndrome (NS) affects 1–3 per 100.000 children per year. Steroids are used as first-line therapy and remission is achieved in up to 85 %. 80% of the children experience at least one relapse. For patients with frequently relapsing NS cyclosporine A (CsA) treatment is recommended aiming for complete remission. CsA therapy is effective but the benefical effects of CsA are often accompanied by acute and chronic CsA nephrotoxicity, hypertension, hypertrichosis and gingival overgrowth. Aim of this study is to analyze the effects of long-term CsA treatment in children with NS.
Material and methods: We performed a retrospective study in children (<18 years) with NS treated with CsA to control frequent relapsing nephrotic syndrome, steroid resistant nephrotic syndrome and steroid dependent nephrotic syndrome in our clinic from 2007 to 2020. Possible patients were identified by searching our database of children with nephrotic syndrome who fulfilled the above criteria. For statistical analysis we used a Bayesian mixed model approach.
Results: A total of 75 patients (34% female) were enrolled in the study. At the beginning of treatment with CsA the median age was 6 years [first quartile 3.4; third quartile 10.3], serum creatinine was 35 μmol/l [27; 44] and estimated glomerular filtration rate (eGFR, bedside formula by Schwartz) was 130 ml/min/1,73m2 [113; 146]. Duration of treatment with CsA was 4.5 years [2.7; 8.0]. Preliminary data analysis showed that either treatment duration or cumulated CsA dose or cumulated trough levels are able to predict course of eGFR.
Conclusions: Children with NS that are treated with CsA for a long period of time a decrease in eGFR can be observed. Other factors like gender, age and clinical course of NS have no influence. Whether the CsA induced decrease on kidney function is reversible has to be analyzed.

EP-59 A RARE SALT WASTING SYNDROME IN INFANTS: SECONDARY PSEUDOHYPOALDOSTERONISM; 8 DIFFERENT CASES

Kenan DoĞan[1], Fatih Kilci[2], Merve AktaŞ ÖzgÜr[1], Mehmet Baha AytaÇ[1], Kenan Bek[1]

[1]Kocaeli University School Of Medicine, Department Of Pediatrics, Division Of Pediatric Nephrology, Kocaeli, Turkey, [2]Kocaeli University School Of Medicine, Department Of Pediatrics, Division Of Pediatric Endocrinology, Kocaeli, Turkey

Introduction: Secondary pseudohypoaldosteronism (PHA) is a rare salt-wasting syndrome that develops due to transient peripheral resistance to aldosterone. It usually presents with hyponatremia, hyperkalemia, and metabolic acidosis in infants. Here, we present eight infant cases who presented with severe hyponatremia and hyperkalemia and were diagnosed with secondary PHA.
Material and methods: Data were analyzed retrospectively from patient files. Adrenal precursor, renin and aldosterone values of the cases were standardized according to age and gender.
Results: Severe hyponatremia and hyperkalemia were the common laboratory features of the cases. Although all cases had normal genital appearance, all of them were evaluated with the presumptive diagnosis of congenital adrenal hyperplasia. While serum adrenal precursors were normal, renin and aldosterone levels were high, suggesting PHA. Urinary ultrasonography was performed in all cases. Six of them had urinary tract malformations and urinary tract infections, so they were diagnosed with secondary PHA. After the treatment (hydration and/or antibiotic) serum renin and aldosterone levels of patients were found to be within normal limits.
Conclusions: Secondary PHA, known as the resistance of renal tubule cells to aldosterone, is a salt wasting syndrome seen in infants and should be kept in mind in cases of hyponatremia and hyperkalemia in infancy.

Although congenital adrenal hyperplasia should be the first diagnosis to be excluded in the presence of hyponatremia and hypokalemia in the infantile period, evaluation should also include urinalysis and renal imaging.

EP-60 XANTHOGRANULOMATOUS PYELONEPHRITIS IN A 6 MONTH OLD INFANT PRESENTED WITH NEPHROCALCINOSIS AND AFUNCTIONAL LEFT KIDNEY

Hana Matković[1], Lovro Lamot[1], Ivanka Kos[1], Maja Ban[1], Maša Davidović[1], Ivan Jakopčić[1], Stela Bulimbašić[2], Tomislav Luetić[3], Kristina Vrljičak[1]

[1]Division Of Pediatric Nephrology, Dialysis And Transplantation, Department Of Pediatrics, Uhc Zagreb, University Of Zagreb School Of Medicine, Zagreb, Croatia, *[2]Department Of Pathology And Cytology, Uhc Zagreb, University Of Zagreb School Of Medicine, Zagreb, Croatia*, *[3]Department Of Surgery, Uhc Zagreb, University Of Zagreb School Of Medicine, Zagreb, Croatia*

Introduction: Xanthogranulomatous pyelonephritis (XPN) is a rare presentation of severe chronic pyelonephritis characterised by renal parenchymal destruction by granulomatous tissue, inflammatory infiltration and fibrosis. The possible underlying mechanism involves urinary tract obstruction due to congenital anomalies, nephrolithiasis and recurrent urinary infection, resulting in unspecific symptoms such as abdominal pain, fever, abdominal mass, growth and weight retardation with persistent anemia, leucocytosis, elevated erythrocyte sedimentation rate (ESR), bacteriuria and pyuria. Therefore, the range of possible differential diagnosis is wide.
Material and methods: Case report
Results: The presented patient was born from an uneventful pregnancy and delivery. He first came to our attention at the age of 6 months due to acute cystopyelonefritis. Subsequently, left-sided hydroureteronephrosis (without vesicoureteral reflux), lymphadenopathy, severe anemia, elevated ESR and liver enzymes, as well as hypergammaglobulinemia and hypertriglyceridemia were noted. Other immune mediated, infectious, metabolic and malignant conditions were excluded by thorough diagnostic workup, including bone marrow analysis and whole exome sequencing. Abdominal ultrasound and computed tomography revealed unilateral nephrocalcinosis, and dynamic renal scintigraphy afunction of the left enlarged kidney. Despite antibiotic therapy, recurrent sterile pyuria with ongoing inflammation was present, necessitating a short course of glucocorticoids which led to gradual improvement of laboratory findings. Finally, left nephrectomy was performed with pathohistological finding suggestive of XPN.
Conclusions: Regardless of diligent diagnostic workup, the precise mechanisms leading to XPN and intense immune reaction in our patient remains inconclusive. The literature data suggest that it might be a result of a macrophage defect in microbial processing, although no other signs of this process were present in the presented patient. Since this condition is very rarely present in children, especially infants, increased reporting of such challenging cases might increase the awareness, alleviate diagnostic dilemmas and steer the most appropriate treatment options.

EP-61 RECURRENT URINARY TRACT INFECTION MAY BE A WARNING FACTOR FOR PATIENTS WITH VOIDING DISORDERS

ÖzgÜr Özdemir ŞimŞek[1], Dilnur Sevinç[2], Gökçen Erfidan[1], Seçil Arslansoyu Çamlar[3], Fatma Mutlubaş[3], Belde Kasap Demir[4], Demet Alaygut[3], Sibel Tiryaki Birol[5]

[1]Department Of Pediatrics, Division Of Nephrology, University Of Health Sciences Izmir Tepecik Training And Research Hospital, Izmir, Turkey, *[2]Department Of Pediatric Surgery, University Of Health Sciences Izmir Tepecik Training And Research Hospital, Izmir, Turkey*, *[3]Izmır Faculty Of Medicine, Department Of Pediatrics, Division Of Nephrology, University Of Health Sciences Turkey, Izmir, Turkey*, *[4]Department Of Pediatrics, Division Of Pediatric Nephrology And Rheumatology, Izmir Katip Çelebi University, Izmir, Turkey*, *[5]Department Of Pediatric Surgery, Division Of Pediatric Urology, University Of Health Sciences Izmir Tepecik Training And Research Hospital, Izmir, Turkey*

Introduction: Dysfunctional voiding may lead to chronic kidney disease in some patients. Prognostic criteria to predict kidney injury are not defined. The aim of this study is to assess the clinical course of patients with dysfunctional voiding to determine possible differences in patients with kidney injury.
Material and methods: Medical records of 19 patients with dysfunctional voiding followed in our hospital were reviewed retrospectively. Patients with anatomic and neuropathic disorders were excluded.
Results: This study included 19 patients (F/M:13/6) with dysfunctional voiding. The mean age at diagnosis was 72±52 months, and the follow-up period was 51±41 months. 16 of 19 patients presented with both daytime and night-time urinary incontinence, 1 patient only had enuresis. Two patients were continent, presented only with urinary tract infection (UTI). At the time of diagnosis, 15 patients had UTI and 13 patients had recurrent UTI in the follow-up. Thirteen (68.4%) of 18 patients to who DMSA was performed had scars in the first scan. 5 of the patients had vesicoureteral reflux (VUR) and one of them had no scars on DMSA. There were two patients with chronic kidney disease (CKD) (Stage IIIA- and IIIB), and these 2 patients did not have VUR. Asignificant amount of residual urine was found in 13 of the patients in repetitive evaluations according to ICCS criteria. When the admission symptoms and uroflow examinations are evaluated; it was observed that recurrent UTI at the time of admission was the only independent risk factor for the development of CKD (p=0.016).
Conclusions: The first complaint in patients with bladder and bowel dysfunction is usually urinary incontinence. Different parameters are used to predict upper tract injury in different uropathies. Patients with VUR and scars in DMSA are alerting findings in dysfunctional voiding. Recurrent UTI was observed as the only independent risk factor for CKD in our study.

EP-63 A 14-YEAR-OLD GIRL WITH IGA-NEPHROPATHY AND TINU: AN UNUSUAL ASSOCIATION

Rachele Spagnol, Elisa Benetti, Mattia Parolin, Germana Longo, Susanna Negrisolo, Andrea Carraro, Davide Meneghesso

Pediatric Nephrology - University Of Padua

Introduction: Immune-mediated kidney diseases in children often follow an infectious event. IgA nephropathy (IgAN) is the most common primary chronic glomerulonephritis. Among acute immune-mediated renal diseases is tubulo-interstitial nephritis (TIN), which can be associated to uveitis in TINU syndrome. Some authors have reported the concurrence of TINU syndrome and IgAN in adults. Only one case is described in children, however the diagnosis of IgAN preceded that of TINU by one year.
Material and methods: In this work we describe the case of a girl who presented histological features of both TINU and IgAN at the time of the diagnosis.
Results: Two weeks after a Mycoplasma lung infection, a 14-year-old girl presented inappetence, fatigue and weight loss with anemia and

increased creatinine levels; tubular proteinuria and glycosuria were found. Ultrasound showed hyperechogenic and swollen renal cortexes, suggestive of acute kidney injury. The clinical presentation was therefore indicative of acute TIN. Ophthalmological evaluation was performed, with finding of bilateral anterior uveitis, configuring a diagnosis of TINU. Renal histology confirmed the clinical hypothesis of TINU, however optical microscopy and immunofluorescence also showed IgA deposit in the mesangium and the presence of a fibro-cellular crescent, typical features of IgAN. A two months therapy with oral prednisone 1 mg/kg/daily was started with normalization of renal function and urine tests at 1 month's follow-up.
Conclusions: To our knowledge this is the first pediatric case with a simultaneous diagnosis of two immune-mediated renal pathologies, both triggered by an infectious event. Our case presented with clinical and laboratory characteristics compatible with TINU syndrome, with an excellent response to the oral corticosteroid; therefore it is conceivable that the IgAN a pre-existing subclinical condition.

EP-64 PRETERM BIRTH AND ITS EFFECTS ON GLOMERULAR FILTRATION RATE AND POTENTIAL QTC PROLONGATION

Anke Raaijmakers[2], Thomas Salaets[1], Zhenyu Zhang[3], Dongmei Wei[3], Jan A Staessen[3], Yuling Yu Yu[3], Karel Allegaert[4]

[1]Division Of Pediatric Cardiology, Department Of Pediatrics, University Hospitals Leuven, Leuven, Belgium, [2]Department Of Pediatrics, Hospitals Zna Jan Palfijn, Merksem, Belgium, [3]Research Unit Hypertension And Cardiovascular Epidemiology, Ku Leuven Department Of Cardiovascular Sciences, Leuven, Belgium, [4]Department Of Development And Regeneration, Ku Leuven, Leuven, Belgium

Introduction: Whether preterm birth is associated with cardiac conduction or repolarization abnormalities in later life is still poorly explored, with conflicting data on QTc prolongation in former extreme low birth weight (ELBW, <1000 g) infants. Former preterms are known to have a poorer cardiovascular outcome compared to their term born peers and QTc is often used to decide whether children are fit to receive certain drugs.
Material and methods: Twelve lead electrocardiograms (ECG) at rest, collected in the PREMATurity as predictor of children's Cardiovascular and renal Health (PREMATCH) study in former ELBW cases and term controls during pre-adolescence (8–14 years) were analyzed on eGFR (cystatin C) and corrected QT time (QTc, Bazett) and QT dispersion (QTd). ECG findings were compared between groups (Mann −Whitney), and associations with clinical and biochemical findings were explored (Spearman). In ELBW cases, associations between QTc and perinatal characteristics (at birth, neonatal stay) were explored (Mann −Whitney, Spearman).
Results: As expected, former preterms scored lower on eGFR compared to the control group (94 vs. 107 ml/min/1.73m^2), but QTc and QTd were similar between 93 ELBW cases and 87 controls [409 (range 360–465) versus 409 (337–460); 40 (0–100) versus 39 (0–110)] ms. Age, height, weight, or body mass index were not associated with the QTc interval, while female sex (median difference 11.4 ms) and lower potassium ($r = -0.26$) were associated with longer QTc interval. We could not observe any significant association between QTc interval and perinatal characteristics.
Conclusions: Although former preterms had a lower eGFR, there were no differences in QTc or QTd between ELBW and term controls in ECGs at rest in pre-adolescents. As such, QTc risk for medicines should not be handled differently in former preterms.

EP-65 ATYPICAL HEMOLYTIC-UREMIC SYNDROME IN A CHILD WITH PRE-B ACUTE LYMPHOBLASTIC LEUKEMIA ON MAINTENANCE THERAPY: CASE REPORT, THE ROLE OF RETICULOCYTES AND ASPARTATE-AMINOTRANSFERASE TO PLATELET RATIO INDEX IN THROMBOTIC MICROANGIOPATHIES

Danko Milosevic[1], Daniel Turudic[3], Ernest Bilic[3]

[1]University Of Zagreb, School Of Medicine, [2]Department Of Pediatrics, General Hospital Zabok And Hospital Of Croatian Veterans, [3]Department Of Pediatric Hematology And Oncology, University Hospital Centre Zagreb

Introduction: Atypical hemolytic uremic syndrome (aHUS) is a rare complement-mediated disease, rarely combined with acute lymphoblastic leukemia (ALL). Some authors believe that leukemia in children is a rare but possible risk for the occurrence of thrombotic microangiopathies.
Material and methods: We present a girl with aHUS who responded well to maintenance therapy for pre-B ALL (BFM ALL IC-2009 protocol). In this child, Eculizumab was administered at a very early stage of aHUS and achieved a rapid response to therapy.
Results: Common and additional hemolysis parameters followed up decreases in hemolysis: Reticulocyte Production Index (RPI) and RPI adjusted for age (RPI/A) to platelet ratio, as well as aspartate-aminotransferase-platelet ratio index (APRI). RPI and RPI/A to platelet ratio are markers of bone marrow response to anemia and hemolysis, primarily serving as a marker of red blood cell vs. platelet recovery. APRI/d-dimer with platelet relationship could be helpful to mark the exact recovery point of aHUS patients and a prognostic marker of Eculizumab treatment success.
Conclusions: To our knowledge, this is the seventh case of HUS / aHUS on maintenance therapy and the first with clearly documented use of Eculizumab. A systematic review of the literature found 14 out of 312 similar articles; five children had aHUS before the onset of ALL and two simultaneously. No clinical recurrences of ALL or aHUS during Eculizumab treatment or residual renal impairment were observed during the one-year follow-up. Such a small number of published cases poses a limitation to the conclusion that both diseases have a mutual genetic background trigger. Possible undiagnosed/unpublished cases may be assumed.

EP-66 PERITONITIS IN CHILDREN ON PERITONEAL DIALYSIS IN UHC ZAGREB BETWEEN 2011.-2021: A SINGLE CENTER EXPERIENCE

Maja Ban, Jasna Slaviček, Ivanka Kos, Hana Matković, Maša Davidović, Lovro Lamot, Kristina Vrljičak

Division Of Nephrology, Dialysis Nd Transplantation, Department Of Pediatrics, Uhc Zagreb, Department Of Pediatrics, University Of Zagreb School Of Medicine, Zagreb, Croatia

Introduction: The objective was to determine the average rate and most frequent causes of peritonitis in children on peritoneal dialysis (PD) treated in University Hospital Center Zagreb, Croatia.
Material and methods: We included 38 patients (20 male, 18 female) followed in our Center who started PD in the period between 1.1.2011. and 31.12.2021. We analysed the duration and all the complications of PD, particulary the incidence and causes of peritonitis.
Results: 13 patients started PD before the age of 6 and 6 patients after the age of 15. Most frequent cause of kidney failure were congenital anomalies (CAKUT) followed by various types of nephrotic syndrome. 19 patients received a kidney transplant, 2 patients are currently on haemodialysis and 17 patients are still on PD.

26 patients had 0-1 peritonitis (68,4%), 7 patients had 2-3 episodes (18,4%) and 5 patients had 4 or more peritonitis episodes (13,1%). 31 patient (81,5%) kad low (<0.5 episodes) and 7 patients (18,4%) had high (>0.5) episodes per patient years. The overall peritonits per patient year was 0,31. Most frequently isolated microbial agent was S. Aureus, followed by P. aeruginosa and sterile peritonitis. Peritonitis was the cause of peritoenal catheter replacement in 14 cases.
Conclusions: Peritonitis remains one of the most severe complications of PD. Adequate training of caretakers and medical staff performing PD is the best way to decrease the incidence of bacterial infections. Having a national registry of PD patients could help analyse weak spots and improve the overall care for patients on PD.

EP-67 MULTICYSTIC DYSPLASTIC KIDNEY AND A RARE GENETIC ABNORMALITY IN A GIRL INFANT

GÖkÇen Erfidan[1], ÖzgÜr Özdemir ŞimŞek[1], Berk Özyilmaz[2], SeÇil Arslansoyu Çamlar[3], Belde Kasap Demir[4], Fatma MutlubaŞ[3], Demet Alaygut[3]

[1]University Of Health Sciences Izmir Tepecik Training And Research Hospital Department Of Pediatric Nephrology, [2]University Of Health Sciences Izmir Tepecik Training And Research Hospital Medical Genetic Diseases Center, [3]University Of Health Sciences İzmir Faculty Of Medicine Department Of Pediatric Nephrology, [4]Izmir Katip Çelebi University, Faculty Of Medicine, Department Of Pediatric Nephrology And Rheumatology

Introduction: Multicystic dysplastic kidney (MCDK) is a common developmental anomaly of kidney characterized by multiple non-communicating cysts and non-functioning dysplastic parenchyma. It is usually asymptomatic, incidental diagnosis is common if not detected antenatally.
Material and methods: The exact mechanism of disrupted nephrogenesis is unknown. However, some of the mutations and chromosomal abnormalities have been identified in this regard.
Results: A full term 3.4 kg female baby was born to a 31 year-old mother via caesarean section. There was no history of teratogen exposure, maternal disease, or medication. The parents had non-consanguineous marriage. On prenatal follow-up, multiple cystic lesions in the right kidney were reported, the largest of which was 80*127 mm. Baby cried immediately at birth and Apgar score was 5/8. On physical examination, she had abdominal distension with palpable mass. Also, she had sacral dimple and hairy patch. Abdominal ultrasonography showed right kidney with massive cystic lesions filling the whole abdomen, and hyperechogenic left kidney with grade 2 hydronephrosis. Sacral ultrasonography was normal.
Urine output was normal (3 cc/kg/hour). While postnatal serum creatinine (SCr) level was 1,2 mg/dL in the first 24 hours, it increased to 1,6 mg/dl after 48 hours. Voiding cystoureterogram showed no vesicoureteral-reflux. On clinical follow-up, she couldn't tolerate feeding per oral or nasogastric tube due to abdominal distension. Therefore, nephrectomy was performed. After the procedure, she well-tolerated feeding and discharged. After one month, SCr level decreased to 0.8 mg/dl. Anteroposterior diameter of left renal pelvis regressed from 18 mm to 13 mm. Pathologic examination of the right kidney supported the diagnosis of MCDK. Genetic analysis showed 17q12 deletion.
Conclusions: "17q12 microdeletion syndrome" is an autosomal dominant inherited chromosomal anomaly including HNF1B gene region. It presented with developmental kidney anomalies, diabetes and neurodevelopmental disorders. This case is reported due to atypical presentation of MCDK and underlying rare genetic abnormality.

EP-68 HYPEROXALURIA AND LOW CALCIUM INTAKE IN PEDIATRICS: BEWARE OF DIET!

Estelle Wagner, Cécile Acquaviva-bourdain, Laurence Dubourg, Garnier Charlotte, Bacchetta Justine

Hcl

Introduction: Secondary hyperoxaluria may be due to increased dietary intake of oxalate/oxalate precursors, decreased calcium intake or intestinal malabsorption. In pediatrics, primary genetic forms (PH) are the most frequent, but in adults, almost half cases of hyperoxaluria have a nutritional origin.
Material and methods: We report on a 5-year-old girl, referred to our tertiary care for hyperoxaluria. She was a single child born from non-consanguineous parents without familial renal history. She displayed cloudy urines in infancy; renal ultrasounds found multiple bilateral lithiasis. A first evaluation was performed, finding high levels of urinary oxalate/creatinine ratio (Ox/creatU), between 120-304 μmol/mmol (N 17-100). Genetic analysis ruled out PH 1, 2 and 3.
A thorough evaluation was then performed because of the discrepancy between overt hyperoxaluria and negative genetic testing. The patient also had multiple food allergies (including cow's milk protein). A detailed nutritional assessment found a low-calcium diet, with only 340 mg of nutritional calcium per day (as opposed to the recommended 800 mg for age), only of vegetal origin (thus of lower efficiency for calcium intestinal absorption). She did not receive calcium supplementation. Biochemical evaluation found normal renal function, and confirmed hyperoxaluria (Ox/creatU 132) with normal glycolate, as well as normal calcium, phosphate, PTH and ALP levels but increased 1.25-dihydroxyvitamin D (350 pmol/L, N <200) and hypocalciuria (<0,11 mmol/L).
Results: Dietician advice were given, aiming at normal calcium intake. Six months later, both 1.25-dihydroxyvitamin D and Ox/creatU normalized (86 μmol/mmol and 157 pmol/L, respectively), as well as renal ultrasounds.
Conclusions: Enteric hyperoxaluria is common in adults, but less frequent in children. Low calcium intake in children may be seen more frequently, because of increased prevalence of cow's milk allergy, avoidance of dairy products and vegan diets. A thorough dietetic assessment is crucial in the evaluation of pediatric nephrolithiasis.

EP-69 CLINICAL PROFILE AND SHORT-TERM OUTCOME OF PRIMARY VESICOURETERAL REFLUX IN CHILDREN

Madhura Fadnis, Jyoti Singhal, Shashank Shrotriya, Jyoti Sharma

Kem Hospital,pune

Introduction: To study the clinical characteristics and short-term outcome of children diagnosed with primary vesicoureteral reflux (VUR).
Material and methods: It was a retrospective chart review of children less than 18 years of age, attending the Pediatric Nephrology Service of a tertiary care center in India. They had been diagnosed with VUR on micturating cystourethrography (MCU). We excluded children with VUR associated with other disorders like posterior urethral valves, urethral stenosis or neurogenic bladder. We noted the demographic profile, presenting complaints, grade of VUR and renal scarring/ hypodysplasia and treatment details.As per the earlier guidelines, all children with dilating reflux were initiated on UTI prophylaxis and underwent ureteric reimplantation for standard indications. Short term outcome with respect to development of hypertension, proteinuria, stage of CKD and resolution of reflux were recorded.
Results: Of the 69 patients with VUR, boys outnumbered girls (M: F ratio: 1.4:1). The median age at diagnosis was 12 months and urinary tract infection (UTI) was the presenting complaint in 59 (85%) children. Of the total renal units of 136(2 patients had a solitary kidney), total refluxing

units were 103, 78 (75%)were dilating (grade I and II) and 25(25%) were non-dilating(grades III, IV and V).Total scarred/dysplastic units ondimercaptosuccinic acid scintigraphy (DMSA) scan were 69(51%).Children with dilating reflux had significantly higher proportion of renal scarring/dysplasia($p<0.05$). Those with an antenatal diagnosis of hydronephrosis were predominantly boys and had a higher prevalence of dilating reflux. Spontaneous resolution of reflux was seen in 7/36(19%) of the patients at a median age of 52 months while 22(32%) children needed surgical intervention.
Conclusions: Primary VUR, irrespective of grade of reflux, is brought to attention most often following a UTI. Dilating reflux (III to V) is most likely to be identified on antenatal ultrasonography, is more common in boys and more likely to be associated with scarring/dysplasia.

EP-70 A PALE GIRL WITH DARK URINE: AN UNCOMMON CASE OF MACROHEMATURIA WITH A FAVOURABLE OUTCOME

Maja Ban, Ivanka Kos, Lovro Lamot, Hana Matković, Maša Davidović, Ivan Jakopčić, Kristina Vrljičak

Division Of Nephrology, Dialysis And Transplantation, Department Of Pediatrics, Uhc Zagreb, Department Of Pediatrics, University Of Zagreb School Of Medicine, Zagreb, Croatia

Introduction: Autoimmune hemolytic anemia is a rare cause of hemolytic anemia in children, most commonly mediated by „warm" IgG antibodies. As opposite, „cold" IgG antibody mediated anemia occurs in only 1% of autoimmune hemolytic anemias in children.
Material and methods: We present a 2.5-year-old, otherwise healthy girl who initially came to our attention because of pallor and dark urine. Two weeks earlier she had symptoms suggestive of both upper respiratory and urinary tract infection which was treated with wide-spectrum antibiotics and led to the complete resolution of symptoms. Two weeks later she developed fever with joint pain, headache and macrohaematuria. A wide differential diagnosis was considered, including hemolytic-uremic syndrome.
Results: Her laboratory findings showed hemolytic anemia (Hgb 92), mild thrombocytopenia (Trc 145), normal renal function and mild hyperlipidemia with only a few erythrocytes in the urine sediment. As a part of workup, the screening for antibodies against red blood cells was performed, revealing positive direct (DAT) and negative indirect (IAT) antiglobulin test. Finally, more specific test revealed positive „cold" biphasic IgG anti P1 antibodies which are patognomonic for paroxysmal cold hemoglobinuria.
Conclusions: Paroxysmal cold hemoglobinuria (Donath-Landsteiner anemia) is a form of autoimmune hemolytic anemia characterised with abrupt onset of hemolytic anemia and hemoglobinuria after exposure to cold weather. Although rare in children, it should not be disregarded in the differential diagnosis of macrohematuria. Treatment is symptomatic with warm electrolyte infusions and erythrocte transfusions. The outcome is favourable with self-limiting course and has no repercussions on patients future life, although recurrent episodes of hemoglobinuria have been described.

EP-71 A CASE WITH VESICOURETERAL REFLUX AND MORNING GLORY ANOMALY: WHAT IS YOUR DIAGNOSIS?

GÖkÇen Erfidan[1], ÖzgÜr Özdemir ŞimŞek[1], SeÇil Arslansoyu Çamlar[2], Demet Alaygut[2], Fatma MutlubaŞ[2], Berk Özyilmaz[3], Gamze TÜre[4], Belde Kasap Demir[5]

[1]University Of Health Sciences Izmir Tepecik Training And Research Hospital Department Of Pediatric Nephrology, [2]University Of Health Sciences Izmir Faculty Of Medicine Department Of Pediatric Nephrology, [3]University Of Health Sciences Izmir Tepecik Training And Research Hospital Medical Genetic Diseases Center, [4]University Of Health Sciences Izmir Tepecik Training And Research Hospital Department Of Ophthalmology, [5]Izmir Katip Celebi University Faculty Of Medicine Department Of Pediatric Nephrology And Rheumatology

Introduction: Morning glory anomaly (MGA) is a rare congenital abnormality of the optic disc. It may be related with neurological, neurovascular and renal abnormalities.
Material and methods: We report a case with coexistence of MGA and vesicoureteral reflux (VUR) with family history of renal failure and blindness.
Results: A 9-year-old female patient applied with the history four pyelonephritis attacks, the last of which occurred two months ago requiring hospitalization. Physical examination revealed no growth retardation or urogenital malformation. Laboratory results showed high serum creatinine (0.9 mg/dL) with estimated glomerular filtration rate of 85 ml/min/1.73m^2, and non-nephrotic proteinuria (8 mg/m^2/hour). Bilateral kidney sizes were normal based on age and sex reference values on ultrasonography, no hydronephrosis was detected. Voiding cystourethrography showed Grade 3 VUR in the right kidney. DMSA performed three months after the last pyelonephritis revealed hypoactive lesions on the left kidney with a split function of 45%. The detailed family history revealed consanguineous parents, two siblings with severe visual impairment, end-stage renal disease in father, aunt and paternal grandmother. Her eye examination revealed MGS. Genetic analysis performed to explain eye and renal anomalies detected a pathogenic heterozygous mutation [c.430C>T(p.Gln144Ter)] on PAX-2 gene. She was diagnosed as papillorenal syndrome. The genetic analysis of the other family members are currently ongoing.
Conclusions: PAX2 gene encodes a transcription factor which is essential for embryonic development of the kidney, eye and central nervous system. Pathogenic variants of PAX2 cause autosomal dominantly inherited disorders with wide phenotypic variability. 92% of the affected individuals have renal abnormalities while 77% have ophthalmologic abnormalities. Both abnormalities together are named papillorenal syndrome. Reported renal abnormalities with PAX2 mutations are renal hypodysplasia, chronic kidney disease, hereditary hyperuricemia, focal segmental glomerulosclerosis and VUR. PAX-2 mutations should be detected in cases with renal abnormalities and eye abnormalities like MGA and suggestive genetic background.

EP-72 CLINICAL OUTCOMES OF PRA POSITIVITY IN THE POST-TRANSPLANT PERIOD

Özgür Özdemir Şimşek[1], Gökçen Erfidan[1], Neslihan Güney[2], Tülay Kılınçarslan[3], Seçil Arslansoyu Çamlar[4], Demet Alaygut[4], Fatma Mutlubaş[4], Belde Kasap Demir[5]

[1]Department Of Pediatrics, Division Of Nephrology, University Of Health Sciences Izmir Tepecik Training And Research Hospital, Izmir, Turkey, [2]Department Of Pathology,university Of Health Sciences Izmir Tepecik Training And Research Hospital, Izmir, Turkey, [3]Department Of Medical Biology And Genetics, Izmir Katip Çelebi University, Izmir, Turkey, [4]Izmır Faculty Of Medicine, Department Of Pediatrics, Division Of Nephrology, University Of Health Sciences Turkey, Izmir, Turkey, [5]Department Of Pediatrics, Division Of Pediatric Nephrology And Rheumatology, Izmir Katip Çelebi University, Izmir, Turkey

Introduction: Despite advances in kidney transplantation, late graft failure still poses a problem. With Panel Reactive Antigen(PRA), both antibody screening and antibody identification can be done. In particular, identification of donor-specific antigens(DSA) that are well-known risk factors for shortened graft survival is important. In this study, we evaluated whether PRA positivity could be a stimulant for rejection in patients with stable graft functions.
Material and methods: We used to monitor PRA and/or DSA twice a year in the last 5 years in addition to annual protocol biopsies in the first five years. We included the patients with positive PRAs and concurrent renal biopsies, and recorded the clinical outcome of the cases in the long term follow up. Patients with elevated serum creatinine were excluded.
Results: This study included 12 patients (F/M:2/10) with kidney transplantation and increased MFI levels for PRA, and only 3 of those patients had DSA. While de novo DSA appeared in 2 patients in the 3rd year, it was seen in the other patient at the 5th year. 6 of the patients were transplanted from cadaveric donors. Only 1 patient had class I positivity. Eight patients had non-nephrotic proteinuria, and one of the patients with de novo DSA did not have proteinuria. The 3 patients with de novo DSA positivity and 5 of the remaining 9 patients with increasing titers of PRA had simultaneous rejection in their biopsies, which were all compatible with humoral rejection. All 8 patients with rejection had class II positivity. None of the 4 patients with PRA positivity but non-specific findings in the simultaneous biopsies had rejection in the follow-up.
Conclusions: Recognition of rejection at an early stage before creatinine elevation is important for graft survival. The use of increased titers of PRA, even not determined as DSA, may be a warning marker for asymptomatic rejection and this data may be evaluated in larger series.

EP-73 THREE SIBLINGS WITH STEROID-RESISTANT NEPHROTIC SYNDROME (SRNS)

Merve Aktas Ozgur, Kenan Dogan, Mehmet Baha Aytac, Kenan Bek

University Of Kocaeli Pediatric Nephrology Department

Introduction: Alport syndrome (AS) is characterized by hematuria and progressive renal disease with ocular and cochlear involvement. Here we present three siblings with hematuria and SRNS whose parents are consanguineous and with no family history of hematuria, kidney disease, deafness.
Material and methods: Data were analyzed retrospectively from the patient file.
Results: Sibling 1: A4-year-old girl presented with gross hematuria and bufissur edema. Laboratory findings revealed hypoalbuminemia, hematuria, and proteinuria. She received an initial steroid treatment however she failed to achieve remission after 4 weeks of prednisone (60 mg/m2). Therefore renal biopsy was performed. Light microscopy showed mesangial hypercellularity. No immune deposit was observed on immunofluorescence staining. Electron microscope examination could not be performed. Pulsed methylprednisolone and then cyclosporine-A was administered however her severe proteinuria persisted. Hearing loss was detected in the outpatient clinic controls. Genetic analysis showed the heterozygote mutation in NPHS1, PLCE1, LAMB2, and homozygote mutation in COL4A4(c.4523-1G>A). Angiotensin-converting enzyme inhibitors were given for treatment. ESKD developed 10 years after disease onset.
Sibling 2: 4-year-old-old girl presented with edema. Laboratory findings revealed proteinuria, hematuria, and hypercholesterolemia. Genetic analysis showed the heterozygote mutation in NPHS1, PLCE1, LAMB2, and homozygote mutation in COL4A4.
Sibling 3: A 3,5-year-old boy presented with weakness and fever. Laboratory findings revealed hypoalbuminemia, hematuria, severe proteinuria. Genetic analysis showed the heterozygote mutation in INF2, LAMB2. Alport gene analysis result awaited.
Conclusions: This article aims to draw attention to the diagnosis of autosomal recessive AS in three siblings with SRNS.

EP-74 VALUE OF NEUTROPHIL-LYMPHOCYTE AND PLATELET-LYMPHOCYTE RATIOS IN THE EVALUATION OF ACUTE REJECTION AND CHRONIC ALLOGRAFT NEPHROPATHY IN CHILDREN WITH KIDNEY TRANSPLANTATION

Hülya Ercan Emreol[1], Bahar Büyükkaragöz[2], İpek Işık Gönül, Sevcan A Bakkaloğlu[2], Kibriya Fidan[2], Oğuz Söylemezoğlu[2], Aydın Dalgıç, Necla Buyan[2]

[1]Gazi University, Department Of Pediatrics, Ankara, Turkey, *[2]Gazi University, Department Of Pediatric Nephrology, Ankara, Turkey*

Introduction: Neutrophil-lymphocyte ratio (NLR) and platelet-lymphocyte ratio (TLR) have become accepted markers of inflammation in recent years and are used to assess disease activity in some diseases. In this study, we investigated the relationship between NLR and TLR values and acute rejection (AR) attacks as well as their role in determining chronic allograft nephropathy (CAN) in the follow-up of kidney transplant (KTx) patients.
Material and methods: 58 KTx patients aged 5-18 years with at least 5-year follow-up at our center were included. Patients with a history of secondary KTx, concomitant malignancy and a shorter follow-up time were excluded. Physical examination, medical history, laboratory parameters in the post-KTx 1st, 3rd and 6th month, 1st, 2nd, 3rd, 4th and 5th years, and renal biopsy reports were reviewed.
Results: Both NLR and TLR were significantly higher during AR attacks (p=0.003 for NLR, p=0.002 for TLR). Although both NLR and TLR values were higher in patients with CAN at the end of 5-year post-KTx follow-up, the difference was not statistically significant (p=0.69 for NLR and p=0.55 for TLR). When the patients with and without CAN within 5 years were compared, the ones with CAN development had significantly higher NLR and TLR values in all periods in the post-KTx first 2 and 4 years, respectively. From the patients with AR attacks, those who subsequently developed CAN had higher NLR in the post-KTx first 3 years, and TLR was higher in post-KTx all time periods, although without a statistically significant level.
Conclusions: This is the first study on evaluation of NLR and TLR in children with KTx. Our results indicate that both values can be used as useful and easily accessible markers in AR diagnosis and CAN prediction, the two major causes of post-KTx graft loss. Pediatric studies with larger populations are needed to support our findings.

EP-75 A RARE CAUSE OF POSTRENAL KIDNEY INJURY IN A BOY

ÖzgÜr Özdemir ŞimŞek[1], SeÇil Arslansoyu Çamlar[1], Sibel Tiryaki[2], Onur Oztan[3], Doga Luleyap[4], Fatih Durak[4], Demet Alaygut[1]

[1]University Of Health Sciences Tepecik Training And Research Hospital Department Of Pediatric Nephrology, *[2]University Of Health Sciences Tepecik Training And Research Hospital Department Of Pediatric Surgery*, *[3]Katip Celebi University Faculty Of Medicine Department Of Pediatric Surgery*, *[4]Katip Celebi University Faculty Of Medicine Department Of Pediatric Intensive Care Unit*

Introduction: Acute kidney injury (AKI) occurs due to prerenal, renal, and postrenal causes. Here, a case that presented with postrenal acute

kidney injury after appendectomy and was diagnosed with Burkitts lymphoma as a result of pathological examination is presented.
Material and methods: An 8-year-old boy, who underwent appendectomy due to abdominal pain and vomiting and was discharged 2 days later, was admitted to the emergency service on the 3rd postoperative day with complaints of nausea, vomiting, and decreased urine output. The growth of the patient admitted to the emergency department was normal. The blood pressure was 145/90mmHg, consistent with stage-II hypertension. The patient, who was seen with tachycardia, tachypnea, and orthopnea in physical examination and the abdomen was distended. He had (+1) pretibial pitting edema. Laboratory tests revealed WBC:14700/mm^3, Hb:8.5gr/dL, plt 556000/mm^3, urea 58mg/dL (N:10-38), serum creatinine 3.7 mg/dL (N:0.5-1.2), uric acid 12 mg/dL (N:2-5.5), albumin 3.1g/dL (N:3.5-5.5), LDH 443U/L (N:110-295), sodium 135 mmol/L (N:134-150), potassium 4.6 mmol/L(N:3.5-5.5), calcium 8.8mg/dL (N:8.8-10.8), phosphorus 9.9mg/dL (N:4-7), and CRP 87mg/L (N:0-5). His urine output was 1.1 cc/kg/h.
Results: Contrast-enhanced computed tomography was performed on the patient. Bilateral ureteral traces could not be evaluated. Diagnostic laparotomy was performed in the case with diffuse abdominal edema. It was observed that the appendix epiploic was diffuse and severely edematous. A double JJ catheter was placed in the bilateral ureters and the operation was terminated by taking the lymph node for the pathological sample. In 72 hours, creatinine decreased to 0.79 mg/dL and urine output returned to normal. The patient with CD10 and CD 20 positivity was diagnosed with Burkitt lymphoma.
Conclusions: In fast-growing hematological malignancies such as lymphoma, kidney functions may be impaired due to tumor involvement, or it may occur due to compression in intra-abdominal masses. Intra-abdominal masses should be kept in mind, especially in patients with elevated uric acid and LDH, and postrenal kidney injury.

EP-76 CLINICAL FEATURES AND ASSOCIATED ANOMALIES IN CHILDREN WITH MULTICYSTIC DYSPLASTIC KIDNEY

Matjaž Kopač, Robert Kordič

University Medical Centre Ljubljana

Introduction: To evaluate clinical features and associated congenital genitourinary anomalies in children with multicystic dysplastic kidneys (MCDK).
Material and methods: In this retrospective study, 80 children with unilateral MCDK were analyzed, evaluated between 2012 and 2020. Data were obtained from electronic and paper health care records.
Results: There were 62,5 % boys and 37,5 % girls. Follow-up time was 8,0 +/- 5,2 y (mean +/-standard deviation). MCDK were detected with prenatal ultrasound in 82,3 % of them. None of them had hypertension. 43,8 % of these children had associated congenital genitourinary anomalies, most commonly vesicoureteral reflux (16.3 %), followed by cryptorchidism (in 15 % of them, but in 24.0 % of boys) and urinary tract dilatation (10 %). 6,3 % of them had chromosomopathy and two among them had the 22q11.2 deletion – DiGeorge syndrome. 10 % of them had urinary tract infection. All of them had normal kidney function except one child with dysplasia of the contralateral kidney. Urinalysis was normal in 90 % of them. Spontaneous involution of MCDK occured in 38,8 % of children in observed period, with average age of involution at 4,1 +/- 3,6 y. Nephrectomy was done in 12,5 % of them, at average age of 2,0 y.
All of the boys with cryptorchidism had undergone surgery except one. In one boy with bilateral cryptorchidism and right-sided MCDK there was an absent vas deferens on the right side. Inguinal hernia was found and repaired during cryptorchidism surgery in eight boys.
Conclusions: Children with a unilateral MCDK have a very good prognosis if the contralateral kidney is normal. It affects boys more commonly than girls. Most of them are detected prenatally. Associated congenital genitourinary anomalies are common, especially vesicoureteral reflux and cryptorchidism.

EP-77 ANTI-COMPLEMENT FACTOR H ATYPICAL HAEMOLYTIC UREMIC SYNDROME OCCURRING AFTER MENINGOCOCCAL B VACCINATION

Francesca Becherucci[1], Ester Conversano[2], Paola Romagnani[3], Francesco Emma[4], Marina Vivarelli[4]

[1]Nephrology And Dialysis Unit, Meyer Childrens University Hospital, Florence, Italy, [2]Paediatric Department, Institute For Maternal And Child Health - Irccs "burlo Garofolo", Trieste, Italy, [3]Department Of Experimental And Clinical Biomedical Sciences "mario Serio", University Of Florence, Florence, Italy, [4]Division Of Nephrology And Dialysis, Bambino Gesù Hospital, Irccs, Rome, Italy

Introduction: Anti-complement factor H (FH) atypical Haemolytic Uremic Syndrome (aHUS) is due to the formation of anti-FH antibodies predisposed by deletions in complement FH-related protein (*CFHR*) genes[1]. The interrelation of the genetic predisposition to the antibody formation is still unclear and probably due to a "second hit" mechanism. We report a case of anti-FH aHUS occurring after meningococcal B (MenB) vaccination.
Material and methods: We report a case of anti-FH aHUS occurring after meningococcal B (MenB) vaccination.
Results: A 5-year-old female patient was referred to a Romanian hospital for HUS, presenting with severe haemolytic anaemia, thrombocytopenia and acute kidney injury (serum creatinine 0.97 mg/dl). Fever, prodromic gastrointestinal symptoms and other accompanying features were not reported. The patient had undergone MenB vaccination booster two weeks before the onset of the clinical picture. C3 was reduced and microbiological analysis tested negative for VTEC. The patient was discharged without sequelae.
Subsequent follow-up showed progressive hypertension that was successfully treated with RAS blockers. Haptoglobin was borderline, with normal LDH levels and without other signs of haemolysis; kidney function kept normal, urinary sediment was negative and sC5-b9 levels were normal. Functional testing for complement deregulation revealed high titre of anti-FH antibodies (1723 U/l, normal values <100 U/l). Genetic analysis with whole-exome sequencing showed a homozygous deletion of CFHR1 gene, but no rare pathogenic variants in complement genes.
Severe hypertension despite mild non-relapsing aHUS suggested chronic post-AKI damage or latent disease activation. In this view, mycophenolate mofetil plus eculizumab were initiated to reduce anti-FH antibodies and allow the continuation of the vaccination schedule. At four months follow-up, anti-FH titles slightly decreased (1340 U/l).
Conclusions: The MenB vaccines contain a polysaccharides epitope with structural similarity to human FH, potentially indicuing molecular mimicry and exposing to the risk of eliciting anti-FH antibodies in genetically predisposed subjects. However, aHUS after MenB exposure is not reported to date[2]. Interestingly, complete deletion of CFHR1 or CFHR3, observed in 5-6% of the general population, is not considered a risk factor for aHUS, but is associated with an increased risk of developing anti-FH antibodies.[1]
This case raises the concern about MenB vaccine as "second hit" in aHUS first episode or relapses among patients with CFHR protein mutations and increased risk of producing anti-FH antibodies.

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EP-78 ARTERIAL STIFFNESS AND CARDIAC FUNCTIONS IN CHILDREN WITH NEPHROTIC SYNDROME

Emre LeventoĞlu[1], Akif Kavgaci[2], Fatma İncedere[2], Semiha TokgÖz[2], Bahar BÜyÜkkaragÖz[1]

[1]Gazi University, Faculty Of Medicine, Department Of Pediatric Nephrology, [2]Gazi University, Faculty Of Medicine, Department Of Pediatric Cardiology

Introduction: Nephrotic syndrome (NS) is one of the most common glomerular disorders of children. It may predispose to accelerated cardiovascular disease, expecially atherosclerosis and cause arterial stiffness due to hypoalbuminemia, hyperlipidemia or useage of steroids. In this study, arterial stiffness indicators like PWV, AIx and ABPM were performed; also cardiac functions were evaluated by echocardiography in the patients with NS. It was aimed to evaluate whether there are any differences between the patients in terms of endothelial damage or cardiac functions.
Material and methods: This is a single center, prospective, case-control study conducted on pediatric patients with NS. Patients were divided into groups according to their treatment modalities: steroid-only therapy (group 1) vs. steroid and another immunsupressive together (group 2). In addition, patients were evaluated in terms of endothelial damage and cardiac functions according to cumulative steroid doses.
Results: Eighteen patients in group 1 and 14 patients in group 2 were included in the study. There was no difference between the groups in terms of sex.
It was observed that age, BMI, cumulative steroid dose per kilogram, and hyperlipidemia were higher; mean arterial pressure, pulse pressure, stroke volume, central blood pressure, PWV were higher; daytime systolic and diastolic blood pressure loads and nocturnal diastolic blood pressure loads were higher; the left ventricular and interventricular septum wall thicknesses were higher, the left ventricular myocardial tissue performance index was lower, and the aortic diameter was larger in group 2.
In addition, as the cumulative steroid dose per kilogram increased; BMI, hyperlipidemia, stroke volume, left ventricular and interventricular septum wall thicknesses, left ventricular mass index increased; left ventricular myocardial tissue perfusion index decreased.
Conclusions: Cardiovascular dysfunction is more pronounced in group 2 due to more severe clinical symptoms. Even if patients with a diagnosis of NS are in remission, they should be evaluated regularly from a cardiovascular point of view.

EP-79 RECURRENCE OF SEMAPHORIN 3B ASSOCIATED MEMBRANOUS NEPHROPATHY AFTER KIDNEY TRANSPLANTATION IN A CHILD

Marc Fila[1], Hanna Debiec[1], Helene Perrochia[3], Nabila Djouadi[1], Marie Christine Verpont[2], David Buob[4], Pierre Ronco[5]

[1]Pediatric Nephrology Department Chu Arnaud De Villeneuve - montpellier University, [2]Sorbonne University Inserm S1155 Paris, [3]Department Of Pathology Chu Gui De Chauliac - Montpellier University, [4]Department Of Pathology - Chu Tenon Aphp - Paris, [5]Department Of Nephrology Centre Hospitalier Du Mans

Introduction: Membranous nephropathy is rare in pediatric patients although its diagnostic may be underestimated in children who are responsive to corticosteroid therapy prescribed for a suspicion of minimal change disease. It is most often associated with an autoimmune disease, mostly lupus. The occurrence of early onset membranous nephropathy (MN) associated with semaphorin 3B has been reported to date in 9 children and 2 adults.
Material and methods: Biopsies were performed on native kidney and one and 5 months after transplantation. semaphorin 3B antigen was detected in immune deposits by immunohistochemistry and confocal microscopy on paraffin-embedded biopsies. Anti-semaphorin antibodies were detected by Western blot and analyzed sequentially
Results: We report the first case of early recurrence after transplantation in a 7-year old boy who presented with severe nephrotic syndrome and advanced kidney failure. There was no evidence of hereditary or associated autoimmune disease. Abundant, almost deposits were seen by electron microscopy and bright granular, subepithelial staining was observed for semaphorin 3B antigen. Western blot analysis of serum revealed the presence of anti-semaphorin antibodies. Recurrence of MN occurred 25 days after transplantation and manifested as nephrotic range proteinuria despite conventional immunosuppressive therapy. Kidney biopsies confirmed histological MN recurrence with colocalization of semaphorin 3B antigen and IgG as early as one months. The patient was treated with rituximab. Proteinuria dramatically decreased within 2 months and complete remission was obtained within 4 months Anti-semaphorin 3B antibodies were then not detected 40 days after rituximab. At one year after kidney transplantation, the patient has a sustained complete immunologic and clinical remission
Conclusions: This case provides evidence that anti-semaphorin 3B antibodies are pathogenic and should be monitored in these patients.

EP-80 TSEN2 GENE MUTATION ASSOCIATED ATYPICAL HUS AND ECULIZUMAB: A CASE REPORT

Faidra Veligratli[1], Elizabeth Forsythe[1], Michal Malina[2], Aoife Waters[1]

[1]Great Ormond Street Hospital, London, Uk, [2]The Newcastle Upon Tyne Hospitals, Uk

Introduction: Most cases of atypical Haemolytic Uraemic Syndrome (aHUS) are associated with mutations of genes encoding proteins involved in the alternate complement pathway. Recently, aHUS has been described in a syndrome involving congenital malformations associated with mutations in the TSEN2 gene. Here we report the case of 2 siblings with a similar phenotype and a mutation within the TSEN2 gene region.
Material and methods: This is a case report.
Results: A 4-year-old girl presented with non-specific symptoms of weakness and lethargy and was diagnosed with atypical HUS. She had a past medical history of faltering growth since the age of 2 years, with microcephaly and reported swallowing difficulties. Family history of childhood death in sibling with similar presentation at the same age, including faltering growth and swallowing difficulties, led to extensive investigations. Our patient was initially treated in intensive care due to severe hypertension. She received eculizumab infusions with subsequent response of thrombocytopenia and anaemia but no improvement in renal impairment. DNA analysed by direct sequencing of the entire coding regions for the CFH, CFI, CD46, C3, CFB, DGKE and MMACHC genes did not reveal any known pathogenic variants of atypical HUS. A kidney biopsy confirmed the picture of chronic thrombotic microangiopathy. Whole exome sequencing revealed a homozygous variant of uncertain significance in the TSEN2 gene, coding for the tRNA-splicing endonuclease subunit SEN2 (TSEN2 NM_025265.4:c.-17-2A>C p.).

Conclusions: This case report adds to the evidence suggesting a role for TSEN2 in immune responses and aHUS development. Experimental work is underway to elucidate the possible mechanisms associated with TSEN2 related kidney disease.

EP-81 ALBUMIN-BILIRUBIN (ALBI) GRADE AMONG CHILDREN WITH STEROID SENSITIVE AND RESISTANT NEPHROTIC SYNDROME: COULD THE INITIAL VALUES FORECAST THE OUTCOME?

Drago Baković[1], Jelena Benčić[1], Ivan Jakopčić[2], Hana Matković[2], Maja Ban[2], Maša Davidović[2], Ivanka Kos[2], Kristina Vrljičak[2], Lovro Lamot[2]

[1]University Of Zagreb School Of Medicine, Zagreb, Croatia;, [2]Division Of Nephrology, Dialysis And Transplantation, Department Of Pediatrics, University Hospital Center Zagreb, University Of Zagreb School Of Medicine, Zagreb, Croatia;

Introduction: Despite the wide use of steroids for idiopathic nephrotic syndrome (INS) treatment during the past 60 years, the reliable marker of response is still lacking. Recently, a parameter known as the albumin-bilirubin (ALBI) grade has been used in several malignant, infectious, and immune-mediated diseases to predict the prognosis and response to treatment.
Material and methods: This was a single centre retrospective pilot study investigating the ALBI grade in children with non-genetic INS. Both albumin and bilirubin concentration were measured before the standard steroid regime was commenced, while patients were further divided according to response during ≤6 weeks of treatment. The ALBI grade was calculated based on the formula previously reported in the literature (log10 bilirubin [μmol/L] × 0.66) + (albumin [g/L] × −0.0852).
Results: Total of 18 responders and 6 non-responders were included, with the mean age of 6.14 and 6.02 years, respectively. The average ALBI score was -1,31 for responders and -1,14 for non-responders.
Conclusions: The crucial role of the immune system in the pathogenesis of non-genetic INS has been widely accepted in recent years, although the precise mechanisms remain inconclusive, and most probably multifactorial. Consequently, a possible systemic subclinical inflammation involving the liver might contribute to the underlying disease processes. In this context, the albumin and bilirubin might be further explored beyond their mechanical role in the maintenance of oncotic pressure, as a negative marker of inflammation and liver damage. Therefore, despite the lack of statistical significance, most probably due to a small number of patients, the result of our study indicates that the ALBI grade, highly representative of the complex albumin and bilirubin interactions, might have a value as a marker of outcome in INS.

EP-82 THE CLINICAL COURSE OF EPHROTIC SYNDROME: A CASE SERIES

Hulya Nalcacioglu, H. Gozde Onal, Demet Tekcan Karali, Ozlem Aydog

Ondokuz Mayis University Faculty Of Medicine, Pediatric Nephrology Department, Samsun, Turkey

Introduction: Nephrotic syndrome is a common renal disease worldwide and an important chronic renal disease in children. This study aimed to retrieve the follow-up clinical data of NS patients and investigate the therapeutic response in children.
Material and methods: We reviewed the medical records of 168 children with NS in our center from 2001 to 2021. We extracted all patients' demographic data, clinical features, laboratory values at the time of diagnosis, and receipt and response to steroids and other immunosuppressants were examined. All used medications and side effects were noted during the examination. Children with secondary or congenital causes of nephrotic syndrome (age, <1 year) were excluded.
Results: 162 out of 168 patients were evaluated. At the onset of idiopathic NS, the median age was four years, ranging from 1 year to 17.0 years. Fourteen patients (8.6%) were older than ten years at onset. The median follow-up duration was 61 months (IQR, 90 months). The median age at the last examination was 11.7 years, ranging from 2.4 years to 18.2 years. Among patients 111 patients (68.5%) steroid-sensitive, 38 (23.5%) steroid-dependent and 13 (8 %) were steroid-resistant NS. 62 (38.3%) patients under other maintenance therapy (CNI, MMF, CYC, or RTX).45 patients underwent renal biopsy. Cyclosporine treatment is the most frequent therapy (53 patients, 31 steroid dependent), followed by MMF with 20 patients. Twelve patients received rituximab therapy (9 were steroid-dependent, three patients were in the steroid-resistant group). Ninety-five patients (58.6%) achieved remission, defined as the absence of relapses being off therapy at the end of the last examination. During the follow-up, side effects were observed in 40 patients (23.8%), mostly due to steroid USAge. Four patients reached CKD stage 5, and 2 of them underwent renal transplants; one patient was on peritoneal dialysis, the other was on hemodialysis.
Conclusions: Most NS patients were steroid-sensitive and reached remission in our series, but many had relapses. Cyclosporine treatment is the most frequent steroid-sparing therapy. Progression to end-stage renal disease occurred in a few patients due to focal segmental glomerulosclerosis

EP-83 COMPARISON OF THE EFFECTIVENESS OF CITRATE AND HEPARIN IN PATIENTS ADMITTED TO THE PEDIATRIC INTENSIVE CARE UNIT WITH RENAL DYSFUNCTION AND RECEIVING CONTINUOUS RENAL REPLACEMENT TREATMENT

Seyma Koksal Atis[1], Muhterem Duyu[2], Alev Yilmaz[1]

[1]Istanbul University Institute Of Health Sciences, [2]Istanbul Medeniyet University Hospital

Introduction: Continuous renal replacement therapies (CRRT) are the most common treatment modalities for critically ill patients in pediatric intensive care units. Blood in the CRRT circuit coagulates when it encounters a foreign surface. Anticoagulation methods are used to prevent coagulation. Although citrate is the most used anticoagulation method in adults, data are limited in the pediatric population. In this study, it was aimed to compare these coagulation methods in terms of the effectiveness of citrate and heparin, filter life and side effects on the patient in patients who applied to the Pediatric Intensive Care Unit with renal dysfunction and underwent CRRT.
Material and methods: This study had 131 patients between January 1, 2015, and January 1, 2021. Patients who underwent CRRT with heparin and citrate were grouped. Cycle life, clotting cycle life, complications (bleeding, metabolic) occurring in patients, and transfusion rates were noted.
Results: We were observed that 43 (32,8%) of 131 patients included in the study had kidney disease. It was seen that 55 patients received treatment with citrate and 76 patients with heparin. The mean filter life was 51 hours (IQR 24-67) in the citrate group and 29.5 hours (IQR 17-48) in the heparin group ($p = 0.002$). The mean lifespan of the coagulated hemofilters was 38 hours (24-50) in the citrate group and 15 hours (10-23) in the heparin group ($p < 0{,}0001$). There was no statistically significant difference between the 2 groups in terms of metabolic complications and bleeding ($p > 0{,}05$).

Conclusions: Citrate is a safe and effective anticoagulation method for continuous renal replacement therapy in children because it appears to prolong circuit survival with a lower incidence of coagulation.

EP-84 MEMBRANOUS NEPHROPATHY IN PAEDIATRIC PATIENTS

Inês Martins, Cláudia Silva, Jéssica Sousa, Joana Suarez, Telma Francisco, Rute Baeta Baptista, Margarida Abranches

Hospital Dona Estefânia

Introduction: Membranous Nephropathy (MN) is a morphological pattern characterized by thickening of the glomerular basement membrane. Although it is one of the most frequent causes of nephrotic syndrome in adults, it accounts for < 5% of cases among children.
Material and methods: Case 1: A 16-year-old girl treated with penicillamine for Wilson's disease presented with nephrotic syndrome. Penicillamine was withhold. Maintenance therapy with zinc acetate was initiated and, transiently, enalapril and furosemide were added. Due to worsening oedema, prednisolone was started. Kidney histopathology was consistent with membranous nephropathy type I. Serum anti-PLA2R and tissue staining for PLA2R were both negative. Nephrotic syndrome remitted in one month, and the proteinuria resolved in six months.
Case 2: A 16-year-old girl with inflammatory bowel disease, pyoderma gangrenosum and a previous history of nephritic-nephrotic syndrome presented with worsening systemic inflammation and increasing proteinuria. Amyloidosis was excluded. The kidney histopathology revealed membranous nephropathy type I. Serum anti-PLA2R antibodies and tissue staining for PLA2R were negative. Systemic inflammation was controlled under infliximab with progressive proteinuria resolution.
Case 3: A 15-year-old previously healthy girl presented with nephrotic proteinuria without hypoalbuminemia. The immunological and infectious studies were unremarkable. Kidney histopathology showed membranous type II with parietal granular deposits of IgG (mostly IgG1 +++ and IgG3 +/-), IgM (++), C1q (++) and IgA (+/-). Serum anti-PLA2R and tissue staining for PLA2R were negative.
Results: MN in paediatric patients is often secondary to drug exposure or systemic illness, as seen in cases 1 and 2, respectively. In case 3, despite the absence of circulating anti-PLA2R antibodies and negative tissue staining for PLA2R, the immunohistochemical findings suggest secondary MN. However, no other criteria of systemic illness have been found.
Conclusions: These three cases illustrate that proteinuria is the clinical hallmark of MN and demonstrate the diversity of the underlying aetiology of this histological pattern, whose identification is paramount to guide treatment.

EP-85 LMX1B-ASSOCIATED STEROID-RESISTANT NEPHROTIC SYNDROME IN CHILDREN

Larisa Prikhodina[1], Svetlana Papizh[1], Tatyana Nikishina[1], Tatyana Lepaeva[1], Patricia Povilaitite[2], Ekaterina Stolyarevich[3]

[1]*Research & Clinical Institute For Pediatrics Pirogov Russian National Research Medical University*, [2]*State Budgetary Institution Of The Rostov Region, Pathologoanatomic Bureau*, [3]*Moscow City Hospital #52*

Introduction: Pathogenic variants in the LIM homeobox transcription factor 1 beta (*LMX1B*) lead to nail-patella syndrome (NPS) (MIM #161200), characterized by nails, patellae's and elbows abnormalities and steroid-resistant nephrotic syndrome (SRNS) as well as to isolated SRNS with FSGS10 (MIM # 256020). The aim of the study was to investigate clinical and molecular characteristics in children with LMX1B-associated SRNS.
Material and methods: Retrospective analysis of phenotype and genotype of 3 girls with LMX1B-associated SRNS was conducted. The median follow-up period was 6.0 (2.0; 14.0) years. LMX1B variants were identified by NGS (n=3).
Results: Among 3 patients with LMX1B-associated SRNS, NPS had 1 girl and isolated SRNS had 2 children. The onset of SRNS with hematuria was at the age of 6 years in the case with NPS and at 4 and 2.5 years in patients with isolated SRNS. Kidney biopsy revealed FSGS in all patients. Electron microscopy (EM) showed moderate effacement of podocyte foot processes and irregular thickening of GBM in all 3 patients. Specific ultrastructural lesions were found in all children: type 3 collagen fibrils in GBM in 2 girls with NPS and isolated SRNS, myelin bodies within podocytes in 1 patient with isolated SRNS. Pathogenic heterozygous previously described variants in the LMX1B gene were identified: c.788T>G (p.Val263Gly) in the girl with NPS, and the same c.737G>A (p.Arg246Gln) in 2 girls with isolated SRNS. At the last follow-up of patients aged 18.0 (8.0; 18.0) years progression to CKD2 was found in 2/3 children with NPS and with isolated SRNS.
Conclusions: We found that children with LMX1B-associated SRNS as involved in NPS and as isolated SRNS had FSGS with specific ultrastructural lesions on EM. NGS in all children with SRNS can improve early diagnosis of isolated LMX1B-associated SRNS.

EP-86 CONGENITAL ANOMALIES OF THE KIDNEY AND URINARY TRACT – A CASE REPORT

Armanda Rebelo[1], Lourdes Mota[1], Catarina Neves[1], Armando Reis[2]

[1]*Hospital Distrital Da Figueira Da Foz*, [2]*Centro Materno-infantil Do Norte*

Anamnesis/Background: An eleven-year-old boy was admitted to our hospital emergency department due to recurrent haematuria. He reported three transient episodes of terminal macroscopic haematuria in the last five months (the first one a few days after a bicycle accident and another immediately after exercise), without other associated symptoms (fever or other genitourinary signs). He was previously healthy, was under no medication, had no known allergies and his vaccines were in schedule. There was no history of renal diseases in the family.
Physical Examination: At our observation he was apyretic and had no abnormalities at physical examination, namely on the cardiovascular and abdominopelvic exams. Height was at percentile (P)50-85, body mass index at P15-50 and blood pressure inferior to P90.
Complementary Diagnostic Tests: Urinalysis evidenced haematuria and a normal protein/creatinine ratio (0,14mg/mg). There were no abnormal findings on blood analyses (blood count, C-reactive protein, creatinine, blood urea nitrogen, ionogram, hepatobiliary parameters and autoimmune study). The renal-bladder ultrasonography described a diffusely thickened bladder with regular contours and a post-voiding residue >10% of the maximum bladder volume, no endoluminal abnormalities and preserved parenchymal renal differentiation. A CT-urogram showed a bilateral piellic dilation (right pelvis: 25mm; left pelvis: 13mm); the right ureter was dilatated (maximum gauge: 11mm), with a slight delay in elimination; the bladder had diffused parietal thickening that was prominent at the level of the vesical trigone (maximum thickness: 14mm). Voiding cystourethrography confirmed the diagnosis of posterior urethral valves.
Learning Points of the Case: Posterior urethral valves are one of the most common congenital anomalies of the kidney and urinary tract, and can occur in varying degrees of severity, depending on the time of diagnosis. Infants and older boys with undiagnosed posterior urethral valves can present in a diversity of ways, including macroscopic haematuria. An early diagnosis is essential to preserve renal function.

EP-87 A RARE CAUSE OF CHILDHOOD NEPHROTIC SYNDROME: RENAL AMYLOIDOSIS

GÖkÇen Erfidan[1], ÖzgÜr Özdemir ŞimŞek[1], Demet Alaygut[2], SeÇil Arslansoyu Çamlar[2], Fatma MutlubaŞ[2], Belde Kasap Demir[3]

[1]University Of Health Sciences Izmir Tepecik Training And Research Hospital Department Of Pediatric Nephrology, [2]University Of Health Sciences Izmir Faculty Of Medicine Department Of Pediatric Nephrology, [3]Izmir Katip Celebi University Faculty Of Medicine Department Of Pediatric Nephrology And Rheumatology

Introduction: Familial Mediterranean Fever (FMF) is an autosomal recessively inherited autoinflammatory disorder which has a classical presentation with self-limited periodic attacks of fever and polyserositis in childhood. However, it may rarely present with unexplained proteinuria/nephrotic syndrome due to amyloid nephropathy.
Material and methods: Herein, we describe a case with nephrotic syndrome diagnosed with amyloidosis on renal biopsy.
Results: A 9-year-old female patient admitted with edema of ankles and eyes for two months. She had a history of recurrent fever, stomachache lasting for one day. The parents described occasionaly artralgia unrelated with fever periods. The family history was irrelevant, no consanguineous marriage was noted. On physical examination, she had normal growth parameters (height of 1.36 m, SDS:-0.12 and weight of 32 kg, SDS:0.02). The blood pressure was normal. She had abdominal distention with palpable hepatosplenomegaly. She had bilateral pretibial edema. Laboratory tests showed hypoalbuminemia (1.7 mg/dL), nephrotic proteinuria (355 mg/m^2/day). Viral serology was negative. The levels of C3 and C4 were normal. Anti-nuclear antibody, anti-double strand DNA, anti-neutrophil cytoplasmic antibodies were negative. Erythrocyte sedimentation rate was 125 mm/hour. Tuberculin skin test was negative. During her hospitalization, we observed periodic fever attacks at three-day intervals, one of them was accompanied with arthritis. Due to atypical presentation, the renal biopsy was performed resulting amyloid-A deposition. Based on the anamnesis and clinical observations, we preliminary diagnosed FMF. Serum amyloid A level was found 746 mg/L (N: 0-6.5). Colchicine (1 mg/day) was started. Anti-IL1, anakinra (100 mg/day) was started after genetic analysis showing homozygous M694V mutation. Now, she is on third month of treatment. She still needs regular albumin infusions with resistant nephrotic syndrome.
Conclusions: Renal amyloidosis is the most severe complication of FMF, mainly affected the patients with homozygous M694V mutations. The patients presenting with nephrotic syndrome may diagnosed after renal biopsy.

EP-88 KIDNEY OUTCOMES IN CHILDREN WITH POSTERIOR URETHRAL VALVES

Rosário Stilwell[1], Cláudia Rodrigues[1], António Bento Guerra[1], Ema Santos[2], Pedro Morais[2], Rute Baeta Baptista[1], Telma Francisco[1], Fátima Alves[2], Margarida Abranches[1]

[1]Paediatric Nephrology Unit, Department Of Paediatrics, Hospital Dona Estefânia, Centro Hospitalar Universitário De Lisboa Central, Lisbon, Portugal, [2]Paediatric Urology Unit, Department Of Paediatric Surgery, Hospital Dona Estefânia, Centro Hospitalar Universitário De Lisboa Central, Lisbon, Portugal

Introduction: Posterior urethral valves (PUV) affect approximately 1:5000 live male births, with about 50% progressing to end-stage renal disease within ten years. We aimed to identify predictors of kidney outcomes for children diagnosed with PUV.
Material and methods: This retrospective single-centre cohort study included children who underwent a PUV ablation between January 1, 2015 and December 31, 2020. Patients with less than three months of follow-up were excluded. The primary outcome was a composite of glomerular filtration rate (GFR) below -2 SD of expected for age or renal replacement therapy (RRT).
Results: From a total of 34 patients in the cohort, 18 (53%) were diagnosed prenatally and 10 (29%) were referred to us from Portuguese-speaking African countries (PALOP). Prevalence of kidney hypoplasia was higher among patients born in a PALOP (100% versus 74%, p=0.04). Patients with a prenatal diagnosis were more frequently born preterm (56% versus 7%, p=0.003), were younger at first urethral catheter placement (6.0±14.4 days versus 14.2±32.4 months, p=0.03), and had higher baseline serum creatinine (1.64±1.61 versus 0.55±0.21 mg/dL; p=0.029) than patients diagnosed postnatally. Mean follow-up time was 3.3±1.2 years. At last follow-up, median age was 2.7 years [interquartile range (IQR) 0.9-3.9] and median GFR was 92.6 mL/min/1.73m2 [IQR 70.5-114.4]. Twelve patients (43%) met the primary outcome. Two patients needed (6%) peritoneal dialysis and another 10 (29%) had a low GFR for age. In the logistic regression analyses adjusted for age at last follow-up prenatal diagnosis (OR 8.4, 95% CI 1.3-53.0, p=0.023), prematurity (OR 49.8, 95% CI 3.5-70.5, p=0.04), and higher serum creatinine at baseline (OR 4.9, 95% CI 1.2-20.6, p=0.031) were significant predictors of the primary outcome.
Conclusions: Prenatal diagnosis of PUV, prematurity and higher serum creatinine before PUV ablation may predict adverse kidney outcomes (low GFR and need for RRT). Our sample size and time of follow-up may have limited our conclusions.

EP-89 A CASE OF A GIRL WITH HEMATURIA AS AN EARLY SIGN OF MULTISYSTEM INFLAMMATORY SYNDROME (MIS-C)

Maria Sangermano[1], Giulia Rubin[1], Annamaria Bonutti[1], Germana Longo[2], Paola Ferrarese[1], Massimo Bellettato[1]

[1]Pediatric Department, Neonatal Intensive Unit And Pediatric Intensive Unit - San Bortolo Hospital, Vicenza, [2]Pediatric Nephrology Dialysis And Transplant Unit, Department Of Women's And Children's Health, Padua University Hospital, Padua, Italy,

Introduction: Multisystem Inflammatory Syndrome (MIS-C) is a rare complication in children with temporal association with COVID 19. MIS-C criteria include: persistent fever, multisystem involvement and elevated markers of inflammation. Renal involvement is a rare reported manifestation of MIS-C, most commonly presented as acute kidney injury (AKI) and seldom as hematuria, proteinuria and pyuria.
The aim of this case is is to evaluate the diversity of renal clinical manifestations of MISC.
Material and methods: We consulted the databases of PubMed and Google Scholar using the keywords: kideny, MIS-C, AKI. The following parameters were noted from the studies including MIS-C patients: renal impairment, treatment, and outcome.
Results: We present a case of 11 years old girl without comorbidities, who presented to our pediatric emergency for persistent fever, abdominal pain, hematuria. Her recent medical history included exposure to SARS-COV2 infection in December 2021. At the physical examination, she was febrile and well appearing. Blood tests showed neutrophilic leukocytosis, elevated markers of inflammation, troponin, proBNP, D-dimers, fibrinogen, creatinine and urea. Urine test showed hematuria, associated with proteinuria and pyuria. Positive IgG SARS-COV2 confirmed clinical suspicion of MIS-C. Within the next 2 days, her condition worsened with AKI, oligoanuria and left ventricular systolic dysfunction (EFLV 40%). The patient was admitted to pediatric intensive care to initiate

inotropic support and she was treated with intravenous immunoglobulin (IVIG) and high-dose cortisone, with instantaneous clinical and laboratory improvement.
Conclusions: Despite numerous reports of MIS-C cases in children, there are still many uncertainties regarding clinical presentation. AKI is the most commonly clinical manifestation of renal involvement in patients with MIS-C, specially among patients with cardiac dysfunction. Pathogenesis of AKI appears to be predominantly pre-renal, but abnormal urinalysis with hematuria and proteinuria is an indicator of renal parenchymal injury. We believe that reporting various manifestations in MIS-C patients will lead to improve diagnosis, treatment and outcome of this novel condition.

EP-90 KIDNEY OUTCOMES IN CHILDREN WITH POSTERIOR URETHRAL VALVES

Rosário Stilwell[1], Claúdia Rodrigues[2], António Bento Guerra[3], Ema Santos[4], Joana Patena Forte[4], Rute Baeta Baptista[1], Telma Carvalho Francisco[1], Fátima Alves[4], Margarida Abranches[1]

[1]Paediatric Nephrology Unit, Department Of Paediatrics, Hospital Dona Estefânia, Centro Hospitalar Universitário De Lisboa Central, Lisbon, Portugal, [2]Department Of Paediatrics, Centro Hospitalar Médio Tejo, Portugal, [3]Department Of Paediatrics, Hospital Do Espírito Santo, Évora, Portugal, [4]Paediatric Urology Unit, Department Of Paediatric Surgery, Hospital Dona Estefânia, Centro Hospitalar Universitário De Lisboa Central, Lisbon, Portugal

Introduction: Posterior urethral valves (PUV) affect approximately 1:5000 live male births, with about 50% progressing to end-stage renal disease within ten years. We aimed to identify predictors of kidney outcomes for children diagnosed with PUV.
Material and methods: This retrospective single-centre cohort study included children who underwent a PUV ablation between January 1, 2015 and December 31, 2020. Patients with less than three months of follow-up were excluded. The primary outcome was a composite of glomerular filtration rate (GFR) below -2 SD of expected for age or renal replacement therapy (RRT).
Results: From a total of 34 patients in the cohort, 18 (53%) were diagnosed prenatally and 10 (29%) were referred to us from Portuguese-speaking African countries (PALOP). Prevalence of kidney hypoplasia was higher among patients born in a PALOP (100% versus 74%, p-value=0.04). Patients with a prenatal diagnosis were more frequently born preterm (56% versus 7%, p-value=0.003), were younger at first urethral catheter placement (6.0±14.4 days versus 14.2±32.4 months, p-value=0.03), and had higher baseline serum creatinine (1.64±1.61 versus 0.55±0.21 mg/dL; p=0.029) than patients diagnosed postnatally. Mean follow-up time was 3.3±1.2 years. At last follow-up, median age was 2.7 years [interquartile range (IQR) 0.9-3.9] and median GFR was 92.6 mL/min/1.73m^2 [IQR 70.5-114.4]. Twelve patients (43%) met the primary outcome. Two patients needed (6%) peritoneal dialysis and another 10 (29%) had a low GFR for age. In the logistic regression analyses adjusted for age at last follow-up prenatal diagnosis (OR 8.4, 95% CI 1.3-53.0, p=0.023), prematurity (OR 49.8, 95% CI 3.5-70.5, p=0.04), and higher serum creatinine at baseline (OR 4.9, 95% CI 1.2-20.6, p=0.031) were significant predictors of the primary outcome.
Conclusions: Prenatal diagnosis of PUV, prematurity and higher serum creatinine before PUV ablation may predict adverse kidney outcomes (low GFR and need for RRT). Our sample size and time of follow-up may have limited our conclusions.

EP-91 USE OF THE NEW EQUATION TO ESTIMATE GLOMERULAR FILTRATION RATE IN ADOLESCENTS DURING TRANSITION TO ADULT HEALTHCARE

Elena Kulakova[1], Tatjana Nastausheva[1], Tatjana Zvyagina[2], Inna Kondratjeva[1], Mariya Skrylnikova[1]

[1]N.n. Burdenko Voronezh State Medical University, [2]Voronezh Regional Children's Clinical Hospital №1

Introduction: Several new GFR-estimating equations were developed to solve the problem of a discrepancy in the estimated GFR (eGFR) in patients during the transition from adolescence to adulthood. The purpose of our study was to investigate use of the CKiD under 25 (CKiDU25) equation in comparison with the Schwartz bedside equation in adolescents during transition to adult healthcare.
Material and methods: We conducted a retrospective analysis of the medical records of 287 seventeen-year old patients (47.4% females) with CKD or/and AKD without AKI who were hospitalized at the Voronezh Regional Children's Clinical Hospital. We calculated and compared the eGFR using the Schwartz bedside, age-dependent CKiDU25, and constant CKiDU25 based on creatinine. The absolute difference was calculated by subtracting Schwartz bedside values from age-dependent CKiDU25 values. The results were presented as a median and interquartile range [IQR].
Results: The median eGFR values using the Schwartz bedside, age-dependent CKiDU25, and constant CKiDU25 were 88 [78–103], 99 [86–109], and 86 [76–96] mL/min/1.73m2, respectively. The median eGFR values for males using the same equations were 82 [72–91], 100 [87–111], and 83 [73–93] mL/min/1.73m2, and those for females were 98 [85–110], 98 [85–109], and 89 [77–100] mL/min/1.73m2, respectively. The median absolute difference between the age-dependent CKiDU25 and Schwartz bedside in males was 16,7 [14,6–19,5], and that for females was -0,8 [-1,4– -0,1] mL/min/1.73m2. We found that 71 (24,7%) adolescents (100% males) needed reclassification upward of their KDIGO GFR category using age-dependent CKiDU25 instead of the Schwartz bedside equation, particularly between the G2–G1 and G3A–G2 categories. There was no need to change the GFR categories in females and patients of both genders with eGFR <30 mL/min/1.73m2.
Conclusions: The biggest absolute difference between the age-dependent CKiDU25 and Schwartz bedside equations was observed in male adolescents with well-preserved kidney function.

EP-92 UTILIZING AMBULATORY BLOOD PRESSURE MONITORING FOR THE DIAGNOSIS OF MASKED HYPERTENSION

Priyanka Chati

L.j. Murphy Childrens Hospital

Introduction: The incidence of hypertension (HTN) in patients with chronic kidney disease (CKD) is increasing. Masked HTN is defined as a normal blood pressure in the clinic, but an elevated blood pressure outside of the clinic. Studies have shown that masked HTN is associated with increased risk for target organ damage including left ventricular hypertrophy, proteinuria, and decreased glomerular filtration rate resulting in end stage renal disease and mortality in the CKD population. If masked HTN is identified early, it can serve as a modifiable risk factor. ABPM is a 24 hour BP monitoring procedure used for a more accurate diagnosis of HTN. In comparison to blood pressures measured at clinic visits, ABPM serves as a more accurate representation to identify a patient

with an underlying history of HTN and is more predictable of target organ damage.

Material and methods: The aim in this quality improvement study was to increase the percentage of ABPM procedures performed in children ages 6-21 with a diagnosis of CKD from 0 to 10% in 1 year. A list of CKD patients ages 6-21 years were identified in the nephrology clinic. Chart review was performed to confirm which patients had ABPMs performed and what percentage of these were diagnosed with masked HTN. Exclusion criteria included patients out of this age range and those with known diagnosis of HTN. The measures included the percentage of patients using ABPM (process) and the percentage of patients identified as having masked HTN with ABPM. There were a total of two interventions performed including placing sticky note reminders in patient charts to screen for masked HTN using ABPM and a poster placed in the clinic room targeted towards patients with CKD.

Results: 92 nephrology clinic patients met inclusion criteria. The percentage of patients using ABPM increased from 0% to 18% (n=17) by the end of December 2021. 53% (n=9) of these patients were identified as having masked HTN.

Conclusions: This quality improvement initiative was successful in achieving its aim to increase ABPM screening in pediatric patients with CKD. Doing so provides an opportunity to identify, treat masked HTN, and ultimately address a major contributing factor to progressive loss of kidney function in this vulnerable population.

EP-93 VANIN-1 AND PERIOSTIN AS A BIOMARKERS OF ACTIVE AUTOIMMUNE PROCESS OR RENAL FIBROSIS IN CHILDREN WITH IGAN OR IGAVN – PILOT STUDY

Malgorzata Mizerska-wasiak[1], Emilia Platos[2], Karolina Cichoń-kawa[1], Malgorzata Pańczyk-tomaszewska[1]

[1]Department Of Pediatrics And Nephrology, Medical University Of Warsaw, Poland, [2]Science Students' Association At The Department Of Pediatrics And Nephrology, Medical University Of Warsaw, Poland

Introduction: This study aimed to evaluate the usefulness of vanin-1 and periostin in urine as a markers of the autoimmune process in kidneys and renal fibrosis in IgA nephropathy (IgAN) and IgA vasculitis with nephritis (IgAVN).

Material and methods: From group of 194 patients from the Department, who were included to the Polish Pediatric Registry of IgAN, IgAVN we qualified 51 patients to the study (20 with IgAN and 31 with IgAVN) between the ages of 3 and 17, diagnosed based on kidney biopsy. The parameters assessed at the onset of the disease were age, protein in the 24-hour urine collection, eGFR, creatinine, albumin, IgA, IgM, IgG, complement components: C3 and C4. Kidney biopsy was classified according to the Oxford Classification. All of the patients received glucocorticosteroids, immunosuppressive or renoprotective therapy. After treatment administration we implemented follow-up measurement and tested the levels of vanin and periostin in the urine Control group consisted of 18 healthy individuals.

Results: The concentration of vanin-1 was significantly higher in IgAN and IgAVN than in the control group (203.44 (2.49-421.6) vs 190.41 (1.06-533.01) vs 109±135.98 pmol/l respectively). Furthermore, no significant differences were found between the concentration of vanin-1 between IgAN and IgAVN. In the study group, periostin concentration did not differ between IgAN, IgAVN and the control group. The concentration of vanin/creatinine correlates positively with the level of IgA and negatively with the serum level of C3 at the end of the observation.

Conclusions: Urinary vanin -1 concentration may be probably useful as a marker of active autoimmune process in IgAN, IgAVN in children, but the study needs confirmation on a larger group of children, along with evaluation of the dynamics of this marker. Urinary periostin is not a good marker for children with IgAN and IgAVN, especially in stage 1 and 2 CKD.

EP-94 HYPONATREMIA AND IMPAIRED RENAL FUNCTION IN COVID-19 PEDIATRIC PATIENTS. A SINGLE CENTER RETROSPECTIVE STUDY

Despoina Tramma, Panagiota Karananou, Paraskevi Panagopoulou, Olga Vambertzi, Sofia Markidou, Efimia Papadopoulou-alataki, Elissavet Vakouftsi, Maria Kavga, Anna Fragoulidou, Nikolaos Gkiourtzis, Agni Glava, Evangelia Desli, Theodora Delaporta, Maria Moutafi, Maria Ntoumpara, Konstantinos Heirakis, Aristides Christakopoulos, Maria Fotoulaki, Kyriaki Papadopoulou-legbelou

Aristotle University Of Thessaloniki

Introduction: A literature review showed a high prevalence of hyponatremia in children with COVID-19 pneumonia, while some studies have reported that hyponatremia is relatively common in Multisystem Inflammatory Syndrome in Children (MIS-C). The objective of the present study was to investigate the prevalence of hyponatremia and impaired renal function among children (0-16 years) hospitalized with COVID-19 infection

Material and methods: We conducted a retrospective, observational study and screened hospitalized patients with laboratory-confirmed SARS-CoV-2 admitted to 4th Pediatric Department of Aristotle University of Thessaloniki between December 2020 and December 2021

Results: Over a period of 12 months, a total of 103 COVID-19 patients [57 (55%) male; 49 (48%) <12 months-old (6d-12m)] were hospitalized. Among them 37 (36%) were hyponatremic (Na<136 mmol/L) Hyponatremic patients were more likely to be infants (<12 months) (25/37, 68%), without sex predominance, presenting with fever and anorexia, but without severe respiratory or gastrointestinal symptoms. Serum potassium ≥4mmol/L(4-6,1) was observed in all 103 patients. Eleven out of 103 patients had severe pneumonia and among them only 3 (27%) had hyponatremia (aged 2, 13 and 14 years old). Hyponatremic patients over 12 months-old presented with syncope (3/12), seizures (1/12), gastrointestinal symptoms (4/12) and rhinitis (1/12). Slightly impaired renal function was found in 50% of patients with hyponatremia and in 27% of normonatremic patients Hyponatremic patients were hospitalized for a median of 6 days (range: 3-13) whereas normonatremic 4.5 days (range: 1-11). Disease outcome was good for all 103 patients including those with sodium homeostasis disturbance. Intravenous fluids, for at least 24 hours, was given to all hyponatremic patients under 12 months of age.

Conclusions: Clinicians should take into consideration hyponatremia in all children with COVID-19, especially under 12 months old, to investigate the cause and manage it accordingly, with careful fluid and electrolyte administration and monitor o fluid balance.

EP-95 SOLITARY FUNCTIONING KIDNEY: ASSOCIATIONS BETWEEN URIC ACID, KIDNEY LENGTH AND FUNCTION

Vaiva Cenkute[1], Dovile Ruzgiene[1], Andrius Cekuolis[2], Arunas Malikenas[2], Augustina Jankauskiene[1], Karolis Azukaitis[1]

[1]Clinic Of Pediatrics, Institute Of Clinical Medicine, Faculty Of Medicine, Vilnius University, Vilnius, Lithuania, [2]Vilnius University Hospital Santaros Klinikos, Vilnius, Lithuania

Introduction: Children with solitary functioning kidney (SFK) may be at risk of kidney function impairment due to reduced nephron number. We

aimed to analyze the association between SFK length, uric acid and SFK function in children.
Material and methods: A single-center prospective cross-sectional study of children with SFK (congenital or acquired; nephrectomies due to malignancy were excluded) >2 years old. SFK length was standardized by calculating height-specific z-scores (SFKz) according to 2021 reference values. Estimated glomerular filtration rate (eGFR) was calculated using updated Schwartz equation. Compensatory hypertrophy (CH) was defined as SFK length >95th percentile.
Results: 41 children aged 7.6±8.8 years (51% boys) with a mean eGFR of 88.75±22.73 ml/min/1.73 m^2 were enrolled. Mean SFKz was 1.5±1.7 and 22 (54%) had CH, while SFKz was below 50th percentile in 9 (22%). 10 (24%) patients had anomaly of SFK. Primary SFK cause was agenesis (n=26), multicystic dysplastic kidney (n=7), nephrectomy (n=6) or undetermined (n=2). 19 (46%) patients had impaired kidney function, with 15 and 4 patients in chronic kidney disease stages 2 and 3, respectively.
Estimated GFR correlated significantly with SFKz (r=0.56, p<0.001) and uric acid (r=-0.53, p<0.001), and associations persisted after adjustment for SFK anomaly and cause (both not associated with eGFR, p>0.05). A model incorporating uric acid and SFKz could explain 40% of eGFR variability. SFKz (OR 1.63 per z-score; 95% CI 1.09 to 2.68), uric acid (OR 0.98 per mmol/L; 0.96 to 0.99) and SFKz below 50th percentile (OR 5.83; 1.18 to 43.8) but not lack of CH (OR 2.41; 0.70 to 8.81) were associated with decreased eGFR (<90 ml/min/1.73 m^2).
Conclusions: SFK length associates with SFK function and may be a simple routine marker to estimate risk of kidney function impairment. The value of uric acid to predict kidney function decline should be investigated in longitudinal studies.

EP-96 POSTERIOR REVERSIBLE ENCEPHALOPATHY SYNDROME (PRES) AND ASSOCIATION WITH COVID-19

Serim Pul, SerÇin GÜven, Nurdan Yildiz, Ece Demirci Bodur, Neslihan ÇiÇek, Özde Nisa TÜrkkan, İbrahim GÖkÇe, Harika Alpay

Marmara UnİversİTy, PedİatrİC Nephrology

Introduction: Posterior reversible encephalopathy syndrome(PRES) is a disease characterized by headache, seizures,visual disturbances,mental changes and focal neurological findings.Available data suggest that PRES is a possible neurological complication of COVID-19. We presented clinical and radiological characteristics of two cases with PRES and COVID-19.
Results: Case-1 Nine-year old girl with nephrotic syndrome, who was receiving corticosteroid therapy, was admitted with fever, cough and increased generalized edema. Her COVID-19 RT-PCR test was positive and blood pressure(BP) was 140/90mmHg. Her BP was regulated with enalapril and amlodipine treatments and she was discharged from the hospital on the sixth day.Fifteen days after discharge,she was admitted with generalized tonic clonic seizure.Her BP was 150/100mmHg and brain magnetic resonance imaging(MRI) revealed findings compatible with PRES.Blood pressure was regulated initially by esmolol infusion.She was completely recovered within three weeks and discharged with normal BP regulated by oral antihypertensive therapy.
Case-2: Nine-year old girl who was on continuous ambulatory peritoneal dialysis due to end-stage kidney disease secondary to spina bifida and neurogenic bladder was admitted with fever and swelling in the right leg. Antibiotic therapy was started with early diagnosis of septic arthritis. On the first week, The SARS-Cov-2 PCR test, which was performed due to recurrence of fever on the first week, was positive. There was no systemic involvement. She was discharged three days later but readmitted with vomiting and confusion two days after discharge. Her blood pressure was 160/100 mmHg and she did not have hypervolemia.Brain MRI finding was compatible with PRES.Her BP was initially regulated with esmolol infusion and then oral antihypertensive therapy.Her symptoms resolved within 10 days.In her last visit,she was normotensive and had no neurological findings.
Conclusions:: Our cases suggested that PRES may be a potentially neurological complication associated with COVID-19. Acute blood pressure elevation which itself may be the cause of hypertensive encephalopathy, may further facilitate the development of PRES in these patients. Tight blood pressure control should be provided in COVID-19 patients and clinicians should consider the possibility of PRES in case of any change in consciousness for appropriate treatment to prevent sequelae of encephalopathy.

EP-97 THE FREQUENCY OF KIDNEY INVOLVEMENT IN PEDIATRIC COVID-19 CASES; SINGLE CENTER EXPERIENCE FROM A PANDEMIC HOSPITAL

Ayse Agbas[1], Gulsen Akkoc[2], Cevher Kizilirmak[3], Nurcihan Caliskan[4], Elvan Bayramoglu[5], Murat Elevli[3]

[1]Pediatric Nephrology, Haseki Training And Research Hospital, University Of Health Sciences, Istanbul, Turkey, [2]Pediatric Infectious Diseases, Haseki Training And Research Hospital, University Of Health Sciences, Istanbul, Turkey, [3]Pediatrics, Haseki Training And Research Hospital, University Of Health Sciences, Istanbul, Turkey, [4]Department Of Biochemistry, Haseki Training And Research Hospital, University Of Health Sciences, Istanbul, Turkey, [5]Pediatric Endocrinology, Haseki Training And Research HospİtaI, University Of Health Sciences, Istanbul, Turkey

Introduction: SARS-CoV-2 has a tropism to the kidney. Hematuria, proteinuria and acute kidney injury have been evaluated in adult population; however, data are scarce for the pediatric population.
Material and methods: This retrospective study evaluated the prevalence of hematuria, proteinuria and acute kidney injury (AKI) in SARS-CoV-2 positive children and adolescents attending to emergency outpatient clinic, during the first eight months of the pandemic. Patient aged between 1-18 years old, having both urinalysis and serum creatinine measurement were consecutively included, patients with urinary tract infection were excluded. “Elevated serum creatinine” and AKI were defined as serum creatinine is above and 1.5 times higher than age-specific upper limit of reference interval, respectively.
Results: A total of 228 patients were evaluated [median (IQR) age 12.7 years (7.5; 16,1), 51.3 % were male]. The prevalence of hematuria, proteinuria and elevated serum creatinine was 15.8% (36/228), 6% (14/228) and 3% (7/228), respectively. None of the patients met the AKI criteria. Kidney involvement (at least one of hematuria, proteinuria and/or elevated serum creatinine) was observed in 23.2% of all cohort (53/228) and 43.5% (10/23) in hospitalized children. Urine density >1020 was present in 52%, serum urea-to-creatinine ratio >40 was present in 56.4% of the patients.
Conclusions: Children generally present with a milder COVID-19 disease course; AKI is not common. However, about one fourth of children have findings of kidney involvement and about half have findings of dehydration, which emphasizes the importance of hydration and avoidance of nephrotoxic drugs in the management of COVID-19.

EP-98 IMPORTANCE AND CLINICAL RELEVANCE OF SYMPTOMS OF ARTERIAL HYPERTENSION IN CHILDREN AND ADOLESCENTS ACCORDING TO THE FINAL DIAGNOSIS OF HYPERTENSION.

Julia Mirecka[6], Malgorzata Stanczyk[1], Aleksandra Olejniczak[2], Justyna Zamojska[3], Marta Gruca[3], Karolina Kowara-dzik[4], Agnieszka Wosiak[5], Agnieszka Szadkowska[2], Elzbieta Smolewska[3], Marcin Tkaczyk[1]

[1]Department Of Paediatrics, Nephrology And Immunology, Medical University Of Lodz, Poland, [2]Department Of Paediatrics, Endocrinology, Diabetology And Nephrology, University Centre Of Paediatrics Of M. Konopnicka, Lodz, Poland, [3]Department Of Cardiology And Paediatric Rheumatology, Medical University Of Lodz, Poland, [4] Children's Department, Provincial Hospital Complex Of S. Rybicki, Skierniewice, Poland, [5]Institute Of Information Technology, Lodz University Of Technology, Poland, [6]Department Of Paediatrics, Immunology And Nephrology, Polish Mother's Memorial Hospital Research Institute Of Lodz, Poland

Introduction: Among the data collected during history taking in children with suspected hypertension there is an information on the symptoms of arterial hypertension. Symptoms commonly considered to be typical of high blood pressure are headache, epistaxis, palpitations, sleep disturbances, blurry vision, fatigue and decreased exercise tolerance. The literature doesn't provide much data on the frequency of symptoms of arterial hypertension in children. The predictive value of the symptoms in children referred to the diagnosis of hypertension is uncertain.
The aim of the study was to determine whether the symptoms reported by children with suspected hypertension are related to elevated blood pressure and whether analysis of symptoms considered typical for hypertension is of diagnostic importance.
Material and methods: The study was retrospective analysis of data of 471 patients aged 5 to 18 referred for the diagnosis of hypertension. The medical records were analyzed with regard to a kind of reported symptoms and the significance of symptoms in relation to the final diagnosis.
Results: More than half of the patients (55.5%) were asymptomatic. The most common symptom was headache (28%) and less commonly reported were: chest pain (6.6%), syncope (6.5%), epistaxis (6%), dizziness (5.3%), weakness (4, 3%), palpitations (3.4%), flushing (1%). In asymptomatic patients, the diagnosis of elevated blood pressure or hypertension was more frequent than the diagnosis of normal blood pressure. Patients with headaches were mostly normotensive, as well as patients with chest pain. Syncope was most commonly observed in patients with essential hypertension. Epistaxis, weakness and flushing were not characteristic of patients diagnosed with arterial hypertension.
Conclusions: Hypertension in children is mainly asymptomatic. The symptoms so far described as indictive of hypertension are not predictive of this condition. The key to proper and timely diagnosis of hypertension is routine blood pressure measurements during medical check-ups.

EP-99 RENAL INVOLVEMENT IN TYPE I INTERFERONOPATHY

Marina Aksenova[1], Anna Kozlova[2], Yulia Rodina[2], Vasily Burlacov[2], Anna Scherbina[2]

[1]Y.veltischev Research And Clinical Institute For Pediatrics, N.pirogov Russian National Research Medical University, Moscow, Russia, [2]Dmitry Rogachev National Research Center Of Pediatric Hematology, Oncology And Immunology, Russia

Introduction: Interferonopathies are a group of monogenic autoinflammatory disorders characterized by a dysregulation of the interferon pathway and manifested by early onset vasculitis, interstitial lung disease, familiar polyarthritis and SLE-like disease specific features.The aim of study was to present the case of SLE-like disease in children with autosomal dominant type 1 interferonopathy caused by deoxyribonuclease-1 gene (*DNASE1*) mutation.
Material and methods: Clinical, laboratory and anamnestic data of patient were summarized.
Results: The 16 yo girl examined for an acute hemorrhagic rash was diagnosed with cytopenia (WBC $4.9x10^3/\mu l$, Plt $142x10^3/\mu l$, Er $2.6x10^6/\mu l$, Hb 109 g/l) and was treated with dexamethasone. Antibiotic-resistant fever and severe hemolytic anemia (Hb 45 g/l) appeared after 2 months. The girl was admitted to the Immunology Department due to ineffective therapy and a positive family history (ex.letalis in sibs from immune pancytopenia). The patient presented with hemorrhagic rash, anasarca, pericarditis, ascites, blood hypertension (145/90 mm Hg), pancytopenia (WBC $1.76x10^3/\mu l$, Plt $52x10^3/\mu l$, Er $2.1x10^6/\mu l$), Coombs positive (3+) hemolytic anemia (Hb 69 g/l), nephrotic syndrome (serum albumin 20 g/l, proteinuria 2000 mg/m^2/d) with AKI (serum CysC 3,6 mg/l, creatinine 178 μmol/l), low C3/C4 blood activity and persistence of multiple autoantibodies (DNA-Ab, ANA, ANCA, cardiolipin-Ab). Kidney biopsy shown focal proliferative nephritis with mesangial and peripheral positivity of IgG++, IgM+, IgA++, C3++, C1q++, κ+, λ++ (lupus nephritis class III). Induction with Solu Medrol (1g/d, №3) and Rituximab (375 mg/m^2/week №4) and symptomatic treatment were started followed by therapy with Prednisone (1 mg/kg/d) and Mycophenolate mofetil (MMF, 1.2 g/m^2). Later MMF was replaced by tofacitinib due to persistent anemia and genetic results: the variant in DNASE I (c.397C>T, p.Arg133Ter) was revealed. The patient is in treatment-induced immunological remission now.
Conclusions: A positive family history and the presence of multiple autoantibodies in patients requires suspicion of primary immunodeficiency. An accurate diagnosis can lead to specific treatment; the diagnosis is important for the future therapy strategy (duration and type), prognosis of the patients and their families (given the dominant inheritance of the disease).

EP-100 ENDOTHELIAL DYSFUNCTION - AS A PREDICTOR OF SECONDARY NEPHROPATHIES IN ENDOCRINE DISEASES IN CHILDREN

Lyudmila Kutsenko, Albina Vyalkova

Federal State Budgetary Educational Institution Of Higher Education "orenburg State Medical University" Of The Ministry Of Health Of The Russian Federation

Introduction: Endothelial dysfunction correlates with the levels of biomolecular markers of inflammation and is an early sign of renal damage (Kurumova K.O., 2010).
Material and methods: The level of endothelin-1 (ET-1) blood and lipocalin associated with neutrophil gelatinase (NGAL) urine was assessed in 150 children aged 3 to 17 years: with endocrinopathies without renal pathology (45), secondary nephropathies in type 1 diabetes mellitus (DM1, n=25), obesity (CEE, n=20), autoimmune thyroiditis (AIT, n=15), 30 conditionally healthy children of the control group.
Results: Statistically significant differences in the level of ET-1 blood and NGAL urine were found in patients with secondary nephropathy (DM1-109.37±8.73pg/ml, 20.03±5.92ng/ml; ECR 112.78±3,48pg/ml, 3.52±1.20ng/ml; AIT 101.2± 8.34pg/ml,10.44±3.89ng/ml) compared with children without nephropathy (88,83±1.71,100.31±2.58,68.29 ±6.83 pg/ml; 3.84±1.41, 1.0±0.05, 1.87±0.46ng/ml) and the control group (26.8±3.7pg/ml, 1.87±0.46ng/ml). The average rate of systolic and diastolic intrarenal blood flow velocity in children with nephropathies is significantly lower than in patients with endocrinopathies without kidney damage ($p<0.05$). In 100% of children with nephropathies hyperfiltration was revealed ($p<0.05$), in 60%- microalbuminuria (MAU) (DM1–100%, IVF-40%) in the absence of MAU in children without kidney damage.
In children with secondary nephropathies, a direct correlation was established between ET-1 and NGAL levels with hyperfiltration (DM1 r=0.6,

0.64; IVF-0.29, 0.27; AIT-0.24, 0.22), MAU (r=0.45, 0.41; 0.34, 0.27; 0.34, 0.46), inverse-with parameters of intrarenal hemodynamics ($p<0.05$).
The clinical significance of ET-1 and NGAL as biomarkers of secondary nephropathies in endocrinopathies was confirmed by the indicators of relative risks (DM1–3.16, 5.09; CEO–2.27, 4.88; AIT-3.25, 4.12), sensitivity (0.64, 0.8; 0.55, 0.8; 0.86, 0.73), specificity (0.92, 0.92; 0.85, 0.9; 0.53, 0.93).
Conclusions: Elevated levels of ET-1 blood and urine NGAL is a predictor of secondary nephropathies in pediatric endocrinopathies.

EP-101 THE EFFECT OF NUTRITION ON RENAL FUNCTIONS IN VERY LOW BIRTH WEIGHT INFANTS

Nagihan Çiftçi Pınar[1], Umut Selda Bayrakçı[2], Şerife Suna Oğuz[3]

[1]Ankara City Hospital, [2]Ankara City Hospital, Department Of Nephrology, [3]Ankara City Hospital, Department Of Neonatalogy

Introduction: Early enteral feeding (EF) and the timing of full enteral feeding (FEF) in the neonatal period are controversial. Kidney functions during the transition period from total parenteral nutrition (TPN) to enteral nutrition (EN), while various physiological adaptation processes of immature kidney were going on, were evaluated.

Material and methods: 168 VLBW newborns were evaluated. The subjects were divided into two groups according to the transition time to full enteral nutrition (FEN). Group 1 was determined as the early FEN group and Group 2 as FEN after the postnatal 14th day and received TPN. BUN, serum creatinine (SCr), BUN/SCr were checked within the first 48 hours of life, between 48 hours and 7 days, between 7-30 days, and after the first month of life. Renal function tests (RFT) of patients that received TPN or early EF were compared.
Results: Early FEF was given to 79 and delayed complete EF was given to 82 infants. BUN, SCr, and BUN/SCr values of all newborns decreased, as postnatal day after birth increased. This decrease was significantly higher in Group 1 than in Group 2. This gives us the idea that the BUN/creatinine ratio will decrease rapidly with the provision of FEN. eGFR, BUN, creatinine, BUN/SCr values; which show kidney maturation, returned to their normal course earlier in the group that switched to enteral nutrition early.
Conclusions: RFT returned to normal faster in the FEN group than in the TPN group, and this decrease is significant for each value. Maturation of the kidney is earlier in newborns who switch to early EF.

EP-102 CONSCIOUS SEDATION IN PAEDIATRIC RENAL BIOPSIES: AN AUDIT OF COMPLICATION RATES USING KETAMINE AND MIDAZOLAM, AND ENTONOX

Emily Broad, Drew Maxted

Nottingham Childrens Hospital, Queens Medical Centre

Introduction: Our primary aim was to audit a change in kidney biopsy sedation protocol in children at a tertiary paediatric nephrology centre. The updated guideline entailed a switch from Pethidine and Diazepam to Ketamine and Midazolam.
Our secondary aim was to assess the safety and efficacy of Entonox as the primary sedating agent.
Material and methods: All paediatric patients who had a renal biopsy between September 2018 and December 2021 were identified. Digital health records were then analysed, reviewing their biopsy audit form and subsequent medical and nursing notes. Complications of the biopsy procedure itself were not included. We purely analysed for complications of sedation such as respiratory depression, oxygen requirement, hypotension, or the need for an alternative form of sedation.
Results: We undertook 170 renal biopsies in the outlined time period, 30 were excluded due to being under general anaesthetic (usually due to age). Of the remaining 140 biopsies, 43 biopsies used the old protocol, 39 the new protocol, and 58 used Entonox. Of the 39 patients on the new protocol of Ketamine and Midazolam, no patients had any sedation caused complications. This compares equally to no patients on the old protocol. Furthermore, when looking at the 58 patients who received solely Entonox, the only complication that arose was 4 patients needing further intravenous sedation, one of these patients also vomited after Entonox.
Conclusions: We have demonstrated the safety and efficacy of using a Ketamine and Midazolam combination for conscious sedation during elective renal biopsies for children. Furthermore, we demonstrated that Entonox alone is a safe and efficacious method of sedation for many renal biopsy patients. Entonox is frequently used for painful procedures, but not commonly for renal biopsies. It has a reduced side effect profile and quicker recovery time and is a viable alternative to intravenous sedation.

EP-103 USE OF TARGETED RELEASE FORMULATION-BUDESONIDE FOR IGAN: A CASE REPORT

Luca Antonucci, Laura Lucchetti, Maria Cristina Mancuso, Alessandra Gianviti, Francesco Emma, Marina Vivarelli

Bambino Gesù Children Hospital

Introduction: The best treatment in IgA nephropathy (IgAN) is still debated. Based on the evidence that a dysfunctional mucosal immune system can lead to abnormal glomerular IgA deposition, release of steroid directly to ileal Payer's patches appears a promising prospective. The trials NEFIGAN and NEFIGARD have demonstrated that targeted release-formulation (TRF)-budesonide, in conjunction with renin-angiotensin-aldosterone system –inhibitors (RAASi), efficiently and safely reduced proteinuria in adults, leading to FDA approvement of TARPEYO as the first specific drug for adult IgAN. In pediatric IgAN, the main therapies remain RAAS-inhibitors and oral steroids. To our knowledge, this is one of the few reports of TRF-budesonide therapy in a pediatric patient.
Material and methods: A 13-year-old boy underwent a kidney biopsy for recurrent macrohematuria and proteinuria, showing IgAN, with a MEST-C score of M1-E1-S0-T0-C1. At admission, serum creatinine was 0.67 mg/dL and UPCR 0.76 mg/mg with macrohematuria. Three methylprednisolone pulses were performed, followed by a 6-month course of prednisone, along with RAASi therapy. After an initial improvement, 10 months from onset, macrohematuria became constant and UPCR increased again up to 0.48 mg/mg. A second renal biopsy showed an increase in sclerotic lesions (S1). Prednisone was discontinued, and a trial with TRF-budesonide 9 mg/day started, increasing ramipril dose to 7.5 mg/day.
Results: One month later, macrohematuria finally disappeared and UPCR decreased to 0.36 mg/mg, with a stable renal function. After 5 months, due to reduction in morning cortisol levels, we started to wean TRF-budesonide by 3 mg every 3 months, increasing ramipril dose to 10 mg/day. Macrohematuria recurred occasionally during intercurrent infectious episodes and proteinuria newly increased to UPCR 0.62 mg/mg, with a slight worsening of renal function.
Conclusions: Our case suggests that TRF-budesonide may be an effective dose-dependent treatment, even in complex pediatric IgAN cases. However, a pediatric clinical trial to identify correct dosage and tolerability of TRF-formulation, is needed.

EP-104 PROGRESSION OF CHRONIC KIDNEY DISEASE IN CHILDREN WITH RENAL HYPOPLASIA

Florina-raluca Badea, Anca-elena Marin, George-claudiu Costea, Cristina Oprea, Diana Stoica, Mona Irina Matei, Ovidiu Limoncu, Adrian Lungu, Cristina Stoica

Fundeni Clinical Institute

Introduction: Renal hypoplasia is the most common entity of the congenital anomalies of the kidney and urinary tract (CAKUT), which represent the first cause of end-stage renal disease in children. The aim of this study was to analyze the progression of CKD in children with renal hypoplasia and to identify the potential predictive and progression factors.
Material and methods: We performed a retrospective study, collecting data from the patients who were admitted to our clinic in the last 10 years with the diagnosis of unilateral or bilateral renal hypoplasia, excluding any acquired causes (obstructive or reflux nephropathy, chronic pyelonephritis, glomerulosclerosis, vascular anomalies). We divided the patients into 2 groups (unilateral and bilateral renal hypoplasia) and 4 categories, according to their age (0-4years, 5-9years, 10-14years and 15-19years).
Results: Among 53 patients, 19 had bilateral hypoplasia and 34 had unilateral hypoplasia, simple (38.23%) or associated with other malformations of the contralateral kidney (the most common were renal ectopia 38% and renal dysplasia 19%).The percentage of patients with a glomerular filtration rate<30 ml/min/1.73m^2 was 14.7% in the unilateral hypoplasia group, while in the bilateral hypoplasia group was 68.42%. According to the age categories, the group of 10-14 years had the highest number of patients with a GFR<30 ml/min/1.73m^2 (28.57% in the unilateral hypoplasia group and 100% in the bilateral hypoplasia group). 47.36% of the patients with bilateral hypoplasia started renal replacement therapy, while in the unilateral hypoplasia group only 3%, in the cases associated with other malformations. Among the risk factors, we identified low birth weight, prematurity, the association with urinary tract infections and the development of hypertension, albuminuria and proteinuria.
Conclusions: This study provides a characterization of renal hypoplasia patients and their progression to CKD, but further analysis and prospective studies are required in order to clarify the management.

EP-106 TREATMENT OPTIONS FOR X-LINKED HYPOPHOSPATAEMIA

Sylva Skalova, Marie Kopecka, Tomas Filipsky

Department Of Pediatrics, Charles University, Faculty Of Medicine In Hradec Králové; University Hospital Hradec Králové

Introduction: X-linked hypophosphataemia (XLH) is a rare disease caused by inactivating mutations in the gene encoding neutral endopeptidase-regulating phosphate (PHEX). The disease is manifested mostly during the first and second year of life by rickets, dysproportionate short stature, limb pain and some other typical complications
Material and methods: Case report A boy with a positive family history of XLH was diagnosed at 3 years of age. Intolerance to conventional phosphate and calcitriol therapy in this boy lead to discontinuation of treatment after approximately 2 years of administration. Conventional treatment was resumed at 9 years of age, but due to intolerance it was not properly given again and there was no improvement in clinical, laboratory and radiological signs of XLH. Pain and deformities of the boys lower limbs significantly reduced the routine activities with the need to use splints.
Results: After switching the patient at the age of 10.5 years to the treatment with burosumab, there was a partial improvement in pain and a gradual adjustment of laboratory parameters. The boy´s younger brother was diagnosed with XLH at the age of 2,5 years and was also initially started on phosphate solution and calcitriol. Due to the intolerance of phosphate solution with repeated vomiting, the boy was switched fter 18 months to the treatment with burosumab with good effect on improvement of clinical difficulties as well as laboratory parameters. Compared to the older sibling, the effect of burosumab treatment in this younger boy was faster and more pronounced.
Conclusions: Early initiation of XLH treatment significantly improves the prognosis of quality of life aspects. A limitation and disadvantage of conventional therapy is often poor tolerance of oral phosphate preparations leading to non-adherence and thus insufficient therapeutic effect. Burosumab is a more effective alternative to XLH without side effects.

EP-107 SERPINB11 GENE VARIANT-RELATED LIVER INJURY IN STEC-HEMOLYTIC UREMIC SYDNROME: A CASE REPORT

Nazli Umman[1], Mey Talip Petmezci[2], Cansu Altuntas[4], Özge ÖzÇelik[1], Biray ErtÜrk[3], Hasan Dursun[5]

[1]Hsu, Prof. Dr. Cemil TaŞÇioĞlu City Hospital, Department Of Pediatrics, İstanbul, Turkey, [2]Hsu, Prof. Dr. Cemil TaŞÇioĞlu City Hospital, Department Of Pediatric Intensive Care Unit, İstanbul, Turkey, [3]Hsu, Prof. Dr. Cemil TaŞÇioĞlu City Hospital, Department Of Medical Genetics, İstanbul, Turkey, [4]Hsu, Prof. Dr. Cemil TaŞÇioĞlu City Hospital, Department Of Pediatric Gastroenterology, İstanbul, Turkey, [5]Hsu, Prof. Dr. Cemil TaŞÇioĞlu City Hospital, Department Of Pediatric Nephrology, İstanbul, Turkey

Introduction: Liver damage of hemolytic uremic syndrome due to Shiga toxin producing Escherichia coli (STEC-HUS) are uncommon. Here, we present a case who were followed up in our clinic with the diagnosis of STEC-HUS and progressed to liver damage with findings thought to be related to serpin family B member 11 gene c.268G>T (p.Glu90Ter) homozygous variant.
Material and methods: A boy aged 3 years were referred to our clinic with a preliminary diagnosis of STEC-HUS. Hemoglobin, thrombocyte and haptoglobin levels were low in patient, whereas lactic dehydrogenase, urea and creatinine levels were found to be high. E.Coli O157:H7 was detected in stool examination in patient. Many schistocytes were found in peripheral smears.
Results: The patient was diagnosed with STEC-HUS, and we performed hemodialysis, plasma exchange, plasmapheresis and gave supportive treatments to the patient. He was successfully treated in the intensive care unit and significant improvement was noticed after plasmapheresis and continuous veno-venous hemodialysis. Meanwhile, cholestasis developed in patient and the total bilirubin levels of the patient was found to be high. Ursodeoxycholic acid was added to the treatment. In the follow-up period, renal functions recovered completely in patient. However, liver damage findings did not improve, and chronic liver damage developed. Liver enzymes and bilirubin values were still high at 3 months of follow-up. Gene mutations that may cause liver damage were investigated in patient, and a c.268G>T (p.Glu90Ter) homozygous variant was detected in exon 9 of the serpin family B member 11 gene in patient. Patients was included in the liver transplantation program due to chronic liver disease in the 6th month and first years of his follow-up.
Conclusions: The main symptom in our patient was renal involvement and liver malfunction. Hepatic manifestation of STEC-HUS may have been a result of hemolysis in the endothelium of hepatic vessels and

subsequent organ ischemia. The occurrence of liver injury in STEC-HUS cases may be associated with the serpin family B member 11 gene c.268G>T (p.Glu90Ter) homozygous variant.

EP-108 HYPERTENSION: IS IT A COMMON COMPLICATION OF MULTICYSTIC DYSPLASTIC KIDNEY?

Ebru Burcu Demirgan[1], Esra Karabag Yilmaz[1], Seha Kamil Saygili[1], Ayse Kalyoncu Ucar[2], Ayse Agbas[1], Ruveyda Gulmez[1], Salim Caliskan[1], Nur Canpolat[1]

[1]Department Of Pediatric Nephrology, Istanbul University-cerrahpasa, Cerrahpasa Faculty Of Medicine, Istanbul, Turkey, [2]Department Of Radiology, Istanbul University-cerrahpasa,cerrahpasa Faculty Of Medicine, Istanbul,Turkey

Introduction: Hypertension is considered a potential complication of multicystic dysplastic kidney(MCDK). However, there is limited evidence on the prevalence of hypertension based on ambulatory blood pressure monitoring(ABPM). The aim of this study was to determine the prevalence and risk factors for hypertension in children with MCDK.
Material and methods: Fifty-four children with MCDK and 20 healthy children participated in this single-center cross-sectional study. Hypertension was determined by office measurement and ABPM. Compensatory hypertrophy was defined as kidney volume above the 95th percentile on kidney ultrasound. Estimated GFR was calculated using the modified Schwartz formula. Microalbuminuria was defined as urine albumin-to-creatinine ratio (ACR) >30 mg/g.
Results: The median (ıqr) age of patients was 10.4 (7.5-13.5) years, and the male-to-female ratio was 1.1/1.0. The left kidney was affected in 59% of cases (n=32). Total involution was observed in 30 patients (56%), regression of MCDK in 20 patients (37%), and nephrectomy was performed in four patients (7%) due to unchanged or increased size. In the contralateral kidney, compensatory hypertrophy was noted in 50 patients (92.5%) and a concomitant urinary abnormality in ten patients (13.5%). There were no differences between patients and controls in terms of age, sex, BMI, SD scores of ABPM values, eGFR, or urine ACR. A total of three patients (5.6%) had hypertension (one sustained hypertension, one masked hypertension, and one controlled hypertension). In addition, seven patients (12.9%) had white coat hypertension. Four patients had hyperfiltration (GFR>130 mL/min/1.73m^2) and five patients had microalbuminuria. The 24-hour MAP-SDS showed no association with the presence of MCDK, contralateral kidney volume, or eGFR.
Conclusions: Approximately 6% of children with MCDK have hypertension according to ABPM. Our results do not suggest a causal relationship between hypertension and MCDK itself or the contralateral kidney. Identification of risk factors for hypertension in this population remains a challenge.

EP-109 KIDNEY TRANSPLANT FROM A LIVING CROSS-DONOR OF INCOMPATIBLE BLOOD GROUP IN A HYPERSENSITIZED PEDIATRIC PATIENT

Yolanda Calzada Baños[1], Pedro Arango Sancho[1], Ignacio Revuelta[2], Marta Jiménez Moreno[1], Ana Cristina Aguilar Rodríguez[1], Elena Codina Sampera[1], Raquel Jiménez García[1], Álvaro Madrid Aris[1]

[1]Hospital Sant Joan De Déu, [2]Hospital Clinic

Introduction: To present our experience in the first crossover ABO-incompatible kidney transplant in a pediatric recipient in Spain
Material and methods: Patient with genetic nephrotic syndrome (WT1 mutation).First transplant at 4 years from a deceased donor and isogroup (group O).cPRA 0%, negative crossmatch.Induction: thymoglobulin (prolonged ischemia), methylprednisolone, tacrolimus, and mycophenolate. At 24 hours, she presented graft thrombosis and restarted hemodialysis. The patient becomes hypersensitized (cPRA 100%) and the relatives are incompatible
Results: Desensitization was performed with immunoabsorption, rituximab (375mg/m2, 2 doses) and intravenous immunoglobulins.The reduction of cPRA to 96% at the expense of class I was verified,carrying out fortnightly maintenance immunoabsorption sessions.Despite this,potential donors are always ruled out because they present DSA.After 4 years of hemodialysis, it is possible to carry out a living cross-donor transplant from a 62-year-old woman with an incompatible blood group (group A).Negative current crossmatch (LB and LT).It presents historical DSA that remained negative after desensitization (DPB1*04, DRB1*15) with maximum MFI of 10950. ABO desensitization is performed, intensified by HLA memory.Initial titers of IgM anti-A isohemagglutinins 1/16 and IgG anti-A 1/64.He received 3 doses of rituximab at 375 mg/m2/dose (D-14, D-7 and D-1) and performed 5 immunoabsorption sessions (days D-5 to D-1).He received intravenous immunoglobulins at replacement doses and a plasma exchange on the day of the transplant (titers 1/1 of IgM-antiA and IgG-antiA at the end). Induction: thymoglobulin, methylprednisolone, mycophenolate, and tacrolimus. Post-transplant maintains isohemagglutinins up to 1/2 after 3 weeks. Protocol biopsy at 2 weeks and 6 months without rejection,positive CD4 in the context of ABO incompatible.Singel antigen serial without DSA.He is currently 12 months post-transplant,without infectious complications and with a normally functioning graft (GFR 60ml/min/1.73m2)
Conclusions: For hypersensitized patients, access to a compatible donor may require rethinking the limits considered unacceptable in a pediatric transplant

EP-110 URINARY PROTEOLYSIS FACTORS AS A PROGNOSTIC MARKER FOR THE PROGRESSION OF X-LINKED ALPORT SYNDROME IN CHILDREN

Zilya Bashirova, Ismail Osmanov

Pirogov Russian National Research Medical University, Moscow, Russian Federation

Introduction: Alport syndrome is a glomerulopathy with typical pathological changes in the GBM. However, studies have shown that tubular damage and interstitial fibrosis contribute to the progression of Alport syndrome. In this study, we wanted to assess whether urinary protheolis factors are associated with disease progression and to determine their prognostic value in children with X-linked Alport syndrome.
Material and methods: 32 children (15M/17F) with X-linked Alport syndrome and normal renal function (CKD gr.1) were examined. All children received therapy with ACE inhibitors. The median age was 10.5 (IQR: 7.5;15). The median follow-up period was 5.5 (IQR:3.5;6.5) years. The control group consisted of 12 age-matched healthy children with normal renal function. Laboratory tests included serum creatinine, MMP-2, MMP-3 and MMP-9 and their inhibitors TIMP-1 and 2, PAI-I in urine (were corrected for urinary creatinine excretion), determined by ELISA. A decrease in eGFR of ≥30% over 2 years from baseline was chosen to represent the rapidly progressive course. 28.1% of children had a rapidly progressive course of the disease (7M/2F), 71.9%-a slowly progressive course (8M/15F). The association of baseline urinary levels of MMPs and their inhibitors with eGFR and progression of patients with Alport syndrome to a later stage of CKD during the follow-up period was used to assess the prognostic value of the marker.

Results: Decreased MMP-9 levels (100% vs. 47.8% (p=0.012), OR=1.82 (95%CI: 1.23-2.71)) and increased TIMP-1 levels (88.9% vs 30.4% (p=0.005), OR=18.2 (95%CI: 1.96-175)) in the urine is statistically significantly more common in children with a rapidly progressive course of the disease than with a slowly progressive course of the disease. Could not find relationship between MMP-2, TIMP-2, PAI-I and disease progression.
Conclusions: Our study suggests that urinary MMP-9 and TIMP-1 are a promising biomarker for accelerated decline in kidney function in children with X-linked Alport syndrome.This may help identify patients at high risk of progression for targeted clinical management and improve patient stratification in future studies.

EP-111 KIDNEY AND BLADDER FUNCTION IN CHILDREN WITH BLADDER EXSTROPHY.

Michal Maternik[1], Chudzik Ilona[1], Drozynska-duklas Magdalena[1], Andrzej Golebiewski[2], Leszek Komasara[3], Aleksandra Zurowska[1]

[1]Department Of Paediatrics, Nephrology And Hypertension Medical University Of Gdansk Poland, [2]Department Of Paediatric Surgery And Urology Medical University Of Gdansk, Poland, [3]Department Of Paediatric Surgery And Urology, Copernicus Hospital, Gdansk, Poland

Introduction: Bladder exstrophy is a rare urological condition with an incidence of 1 in 10,000 - 50,000 live births. Bladder closure in early childhood creates a risk for upper urinary tract damage due to bladder dysfunction, VUR, hydronephrosis and recurrent UTI's. There is limited data on the long term outcome of kidney function in this cohort.
Material and methods: Bladder exstrophy patients treated at our institution during the last 10 years were identified . Kidney damage was assessed by evaluating their last available eGFR, albuminuria and beta2-mikroglobulin levels. Kidney corticomedullary differentiation and the presence of hydronephrosis were evaluated by ultrasound and kidney differential function by dynamic scintigraphy. Continence status, bladder capacity and performance of CIC were used as markers of bladder function.
Results: Fifteen subjects were identified: eight females and seven males of an average age of 5 years (1-11) . None of the children had a decreased eGFR <90ml/min/1,73m2, and all had normal blood pressure values. Albuminuria was present in a single subject and increased beta2microglobuline in two. Loss of corticomedullary differentiation was present in in 3 children in at least one renal unit. Bilateral hydronephrosis was present in 6 subjects and differential kidney function was abnormal in 5. Bladder capacity was decreased in 13 children when compared expected bladder capacity for age (EBC); 8/13 children above 5yrs old were incontinent and 7/15 required CIC (clean intermittent catheterization).
Conclusions: At a mean age of 5 yrs 33% have developed stage I chronic kidney disease underscoring that regular nephrological evaluation is mandatory in children born with bladder exstrophy. The majority of children with bladder extrophy demonstrate bladder dysfunction and incontinence is frequent.

EP-112 MULTIDRUG THERAPY OF ACUTE COMPLICATIONS AFTER HEMATOPOIETIC STEM CELL TRANSPLANTATION INCLUDING BK-VIRUS-MEDIATED HEMORRHAGIC CYSTITIS

Katarzyna Gąsowska, Jolanta Goździk, Katarzyna Zachwieja, Dorota Drożdż

Jagiellonian University Collegium Medicum

Introduction: 11-year-old girl with acute monocytic leukemia was admitted for allogenic bone marrow transplant. A preparatory therapy for transplantation was carried out, the prophylaxis of transplant rejection included methotrexate, ATG and cyclosporine. Hematopoietic cells were transplanted from an unrelated donor, compatible with 9/10 HLA.
Material and methods: In the posttransplant course from day 1, intensive gastrointestinal mucositis was observed requiring opioid analgesics and parenteral nutrition which then progressed to gastrointestinal bleeding. On day +3 patient's general condition worsened, she developed fever, dyspnea with the necessity of oxygen therapy, and inflammatory markers increased. Staphylococcus hominis was found in blood culture, followed by Klebsiella pneumoniae (day +8). Antibiotic therapy was modified, immunoglobulins and steroid therapy were applied and intensive symptomatic treatment was implemented. Chest X-ray revealed signs interstitial pneumonia with a possible etiology of pneumocystosis, therefore pentamidine was also used. Severe hemorrhagic diarrhea and increasing parameters of renal failure (from day +3) were observed (maximum serum creatinine 110 umol/l, cystatin C 2,26 mg/). The doses of drugs were adjusted to GFR (45 ml/min/1,73m2), cyclosporin (then serum drug concertation 98,07 ng/ml) was changed to tacrolimus (maximum serum concertation 33,8 ng/ml) – that was gradually discontinued. From day +12; massive hemorrhagic cystitis occurred, (increased BKV replication was observed). Ciprofloxacin was administered, followed by cidofovir resulting in decreased hematuria - without complete disappearing of bladder inflammation. On day +49 CMV reactivation was found. Ganciclovir was administered.
Results: Due to the persistence of hemorrhagic cystitis, non-standard therapy with leflunomide was implemented. Gradually, complete resolution of the hemorrhagic cystitis was observed. Methylprednisolone treatment was started and tacrolimus was discontinued. After clinical stabilization, the girl was discharged home. The development of stage 3 chronic kidney disease and arterial hypertension remained as complications in further follow-up.
Conclusions: Intensive multi-drug therapy of acute post-HSCT complications may lead to chronic kidney disease as a result of drug-related toxicity.

EP-113 THE MAJORITY OF MNE AS WELL AS NMNE PATIENTS HAVE A THERAPEUTIC WINDOW FOR DESMOPRESSIN THERAPY

Sevasti Karamaria[1], Lien Dossche[1], Ann Raes[1], Evelien Snauwaert[2], Johan Vande Walle[1]

[1]Ghent University Hospital, Ghent University, [2]Ghent University Hospital

Introduction: Nocturnal enuresis is caused by nocturnal urine production and functional bladder capacity mismatch. ICCS suggests patient classification into MNE and NMNE, where MNE is likely to respond to desmopressin and/or alarm. This led to the misconception that NMNE could not benefit from desmopressin. With the recent ICCS standardization, most patients are now labeled NMNE. Desmopressin's anti-diuretic effect and renal concentrating response have no direct correlation with bladder dysfunction and should be evaluated independently. Many patients have LUTS and nocturnal polyuria and could benefit from desmopressin in combination therapy.
Material and methods: Aim: identify patients in a tertiary center who might benefit from desmopressin (defined as urinary osmolality (Uosmol)<850 mOsm/l) and study the timing overnight.
Methods: retrospective analysis of 398 enuretic children who performed a 24h-urine concentration profile at home (4 daytime (D1-D4), 4 nighttime urine collections (N1-N4)).

Results: 212 children (>50%) had Uosmol<850 mOsm/l at the 1st-night collection (N1), and would benefit from a short-term desmopressin activity; however, in a significant percentage, Uosmol is low later in the night (181 N2, 169 N3, 167 N4), needing a longer action duration. 50 patients didn't reach Uosmol>850mOsm/l over 24h, suggesting lower maximal renal concentration capacity of the normal spectrum or high 24h fluid intake.
Conclusions: Classification into MNE and NMNE is mainly bladder/LUTS driven and is widely accepted to predict the anti-enuretic effect of therapy, thus an indication for desmopressin. However, many patients have a combination of LUTS and abnormal circadian diuresis pattern. Desmopressin's anti-diuretic effect may be expected in most patients with high diuresis and low Uosmol overnight. >50% of patients have a low Uosmol early the night, hence a therapeutic window for desmopressin. In 1/3 patients, Uosmol remains longer low, needing longer-acting V2-stimulation, without risk of too prolonged action. It is evident that desmopressin's PK/PD characteristics do not fulfill these promises.

EP-114 NEPHROLITHIASIS IN AN INFANT AS DIAGNOSTIC KEY OF A GENERALIZED PEROXISOMAL DISORDER

Héctor Ríos Duro[1], Jose Antonio Arranz Amo[3], Silvia Franch Salvadó[2], Alejandro Cruz Gual[1], Mercedes López González[1], Víctor Pérez Beltrán[1], Marina Muñoz López[1], Gema Ariceta Iraola[1]

[1]Department Of Pediatric Nephrology, Hospital Universitari Vall D´hebron, [2]Department Of Pediatrics, Hospital De Tortosa-verge De La Cinta, [3]Metabolic Laboratory, Hospital Universitari Vall D´hebron

Introduction: Peroxisome biogenesis disorders (PBDs) are a heterogeneous group of inherited metabolic diseases caused by mutations in PEX genes. Impaired peroxisomal function in affected individuals can manifest a complex spectrum of clinical phenotypes. Nephrological clinical features are described: kidney cortical microcysts and calcium-oxalate stones, attributed to potential oxalate metabolism dysfunction.
Material and methods: We describe a patient affected by PBDs diagnosed after spontaneous expulsion of a renal stone.
Results: A two-month-old boy without familial or personal history of interest was admitted because of cholestasis detected during patient evaluation for growth retardation, bloody stools and irritability. On physical examination, patient's appearance was characterized by facial asymmetry, marked philtrum and epicanthus inversus. Etiological studies, included evaluation for infection, renal function and metabolic disorders, didn´t reveal any abnormality, except mild elevation of liver enzymes.
At 4 months of age, the patient spontaneously passed a kidney stone. Kidney function was normal but ultrasound showed bilateral nephrocalcinosis grade 1. Mild hypertransaminasemia persisted without cholestasis. Further metabolic assessment demonstrated repeated hyperoxaluria oxalate/creatinine 392 mmol/mol and 551 mmol/mol (normal value for age 370 mmol/mol), increased glycolate and increased of 2-hydroxy-sebacic acid in urine, all together lead to the suspicion of a peroxisomal disorder. Stone composition was mainly calcium oxalate monohydrate. Extended metabolic study confirmed elevated very-long-chain-fatty acids in plasma and genetic test confirmed the diagnosis of peroxisomal disorder. The patient carried a compound heterozygous pathogenic variants (c.2097dupT, c.3077T>C) in PEX1 gen.
Treatment with increased fluid intake and urine alkalinization with citrate salts was performed. Kidney ultrasound normalized 3 months later and oxaluria decreased over time to normalize after two years.
Conclusions: Children affected by PBDs should be monitored for hyperoxaluria, which can lead to kidney stone disease and kidney chronic damage. Kidney ultrasound may be useful to detect renal stones. This case shows the relevance of early recognition and follow-up in this entity.

EP-115 SCHIMKE IMMUNO-OSSEUS DYSPLASIA: EXPERIENCE IN MANAGING A PATIENT AFTER KIDNEY TRANSPLANTATION

Olga Raikevich-liachovskaja[1], Sergej Baiko[2]

[1]The 2nd City Children's Clinical Hospital, Minsk, Belarus,, [2]Belarusian State Medical University, Minsk, Belarus

Introduction: Schimke immuno-osseous dysplasia (SIOD) is a rare disorder, characterized by spondyloepiphyseal dysplasia, defective cellular immunity, nephrotic syndrome with an outcome in most cases in the end-stage renal disease (ESRD).
Material and methods: We describe a clinical case of a patient with SIOD after kidney transplantation.
Results: At the age of 3 years a boy was diagnosed with nephrotic syndrome resistant to steroids and cyclosporine A, which led to the ESRD by the age of 5. At the age of 5,8 years he received a kidney from a deceased donor. On the eve of operation, the patient received an intravenous induction therapy: methylprednisolone (MP) and basiliximab. The maintenance therapy included MP, azathioprine and tacrolimus. By the second month after the transplantation frequent viral infections, leukopenia and lymphopenia appeared, which required the reduction of azathioprine. After the normalization of leukocyte level the boy was swiched to mycophenolate sodium (MS). It was accompanied by a significant decrease in the level of leucocytes and lymphocytes, frequent herpes infections. The reduction of MS didn't lead to a positive result, and antimetabolites were canceled. MP (4 mg) was prescribed every day (before that every other day). Subsequently while taking reducing azathioprine or MS dose leukopenia increased, infectious complications reoccurred. Antimetabolites were finally canceled in 5,5 years after transplantation. Thus, a 2-component immunosuppressive therapy is optimal for the patient: MP and tacrolimus. During the last 3 years kidney function remains stable. The current blood level of creatinine is 63 μmol/l.Donor-specific antibodies were not revealed.
Conclusions: The immunosuppressive therapy for patients with SIOD after kidney transplantation should be individual because they have primary T-cell immunodeficiency.

EP-116 NETRIN-1 AS A PROGNOSTIC MARKER OF SHORT-TERM RENAL OUTCOME IN CHILDREN AFTER CARDIAC SURGERIES – PRELIMINARY STUDY.

Beata Leszczynska[1], Anna Deja[1], Michal Buczynski[2], Michal Zawadzki[2], Maria Daniel[1], Katarzyna Szymanska-beta[3], Malgorzata Panczyk-tomaszewska[1]

[1]Department Of Pediatrics And Nephrology, Medical University Of Warsaw, [2]Department Of Cardiosurgery And Pediatric Surgery, Medical University Of Warsaw, [3]Department Of Anesthesiology And Intensive Therapy For Children, Pediatric Clinical Teaching Hospital, University Clinical Teaching Center Of Medical University Of Warsaw

Introduction: Acute kidney injury (AKI) is a common complication of pediatric cardiosurgical procedures and is associated with elevated mortality. Netrin-1 is an anti-inflammatory protein secreted by proximal

tubule epithelial cells in reaction to hypoxic or toxic injury. The aim of the study was to evaluate the role of netrin-1 as a marker of short-term renal outcome in children after cardiac surgeries.
Material and methods: The study involved 32 children (22 girls, 10 boys) who underwent cardiac surgery with extracorporeal circulation due to congenital heart defects. We evaluated urinary netrin-1 before, 6, 24 and 48 hours after the surgery, as well as biochemical and clinical parameters. eGFR was calculated using Schwartz formula with cystatin C.
Results: Median age was 6.5 months (IQR 4.0-13.5). Median duration of extracorporeal circulation was 111 min (IQR 77-130), median aortic clamp time was 51 min (IQR 34-69). 48 hours post-surgery, AKI (according to KDIGO guidelines) was present in 3 patients and GFR decline – in 19 patients (median ΔGFR -9.6 ml/min/1.73m^2, IQR -18.9–2.71). Initial netrin-1 correlated positively with ΔGFR after 48 hours. The cohort was divided into two groups concerning initial netrin-1: detectable (group A, n=11) and undetectable (group B, n=21). After 48 hours GFR increased in group A and decreased in group B with significant difference between the groups (ΔGFR 7.3 ml/min/1.73m^2 and -4.8 ml/min/1.73m^2 respectively, p=0.046). In multivariate analysis, detectable netrin-1 (beta=0.34, p=0.02) and serum urea (beta=0.55, p<0.01) were predictors of GFR increase. Netrin-1 ≥2.55pg/ml was predictive of GFR increase with 82.4% sensitivity and 53.3% specificity. We found no correlation between netrin-1 after 6, 24 or 48 hours and post-surgical biochemical or clinical parameters.
Conclusions: 1. Urinary netrin-1 might be an indicator of subclinical kidney injury associated with renal hypoxia due to cardiac defect.
2. Children with detectable urinary netrin-1 might benefit from the surgery concerning short-term renal outcome.

EP-117 FULL-HOUSE NEPHROPATHY AFTER SARS-COV-2 INFECTION: ABOUT A CASE

Julie Tenenbaum[1], Agnès Chevalier[1], Marc Fila [1], Floriane Hemery[1], Lydia Ichay[1], Hélène Perrochia[2], Denis Morin[1]

[1]Department Of Pediatric Nephrology, Centre Hospitalier Universitaire De Montpellier, Montpellier, France, [2]Pathology Laboratory, Centre Hospitalier Universitaire De Montpellier, Montpellier, France

Introduction: We report the case of a 12-years old girl, arrived from Sudan a month ago, who present sudden edema revealing a nephrotic syndrome. A macroscopic hematuria appear one week after edema followed by an acute kidney failure with a nadir of serum creatinine of 222 µmol/L after 18 days of evolution.
Material and methods: Kidney biopsy reveal a diffuse mesangial and endocapillary proliferation with many parietal glomerular deposits of C3, IgG, C1q, kappa and lambda light chains evoking a full-house nephropathy (FHN).
No oncological or auto-immune cause was found. Complement factors C3 and C4 were normal, serum ANA and ANCA were negative, with no elements for a systemic lupus erythematosus (SLE). Various viral serologies were negative, a part of SARS-CoV-2 serology with negative anti-nucleocapsid (N) antibodies and positive anti-spike (S) antibodies at a level of 133 UA/mL one month after diagnosis. The patient had no clinical manifestations of SARS-CoV-2 infection.
Results: Therapeutic management consists of 3 methylprednisolone pulses of 1g/1.73m² followed by oral corticotherapy of 60 mg/day, associated to MMF at 1200 mg/m²/day in 2 doses.
After one month of treatment with a good tolerance, we observe a decrease of proteinuria becoming non nephrotic (proteinuria/creatininuria 70 mg/mmol), persistence of a microscopic hematuria and improvement of serum creatinine (87 µmol/L).
The patient currently benefits from a monthly follow-up.
Conclusions: Non-lupus FHN represents a diagnostic and therapeutic challenge because it is a new entity which still needs further studies. We need to follow serologies suggestive of SLE because FHN may be initial manifestation of SLE.
This is the first described case of FHN after a SARS-CoV-2 infection and we need to collect more data to appreciate the potential severity of this pathology.

EP-118 ERICONS – EARLY RITUXIMAB IN CHILDHOOD ONSET NEPHROTIC SYNDROME – STUDY PROTOCOL.

Aleksandra Zurowska, Magdalena Drozynska-duklas, Ilona Zagozdzon, Irena Balasz-chmielewska, Iga Zaluska-lesniewska, Michal Maternik

Department Of Paediatrics, Nephrology And Hypertension Medical University Of Gdansk Poland

Introduction: Initial treatment protocols for idiopathic nephrotic syndrome (INS) are based on high dose steroids which induce remission in the majority of children. Nevertheless >45% of children may relapse and many will demonstrate a protracted steroid dependent(SDNS) or frequently relapsing (FRNS) disease lasting throughout childhood. Subjects with SDNS/FRNS are at risk of frequent hospitalizations and numerous side effects of steroid and immunosuppressive treatment. Rituximab, an anti-CD20 antibody has raised hopes of higher cure rates and decreased treatment related morbidity. Randomized trials on the efficacy of early rituximab treatment of SDNS/FRNS in children are lacking.
Material and methods: A multicentre, randomized double blinded study has been designed to assess the efficacy and safety of early treatment with rituximab for children with SDNS/FRNS prior to any traditional immunosuppressive therapy. Recruitment is planned from - 9 major University Hospitals. 60 children are to be randomised (30 to rituximab, 30 to a placebo arm) following remission of NS achieved with steroids. Rituximab will be given in two infusions of 375mg/m2 and compared to placebo with further cessation of steroids. The primary endpoint of the study is time of survival without relapse in double blinded phase. The secondary endpoints are: time to treatment failure, total dose of steroids, correlation of relapse with lymphocyte B counts, rituximab concentration and presence of anti- rituximab antibodies, immunofenotype and genotype of subjects. Urine and blood tests will be collected according to the study protocol during screening and at monthly control visits for 12 months observation period. Adverse effects of treatment will be monitored over 12 months following infusion of study drug and will be registered according to GCP rules.
Results: The study is funded by Medical Research Agency, Poland, Project number: 2019/ABM/01/00024. The study has been accepted by regulating agencies and bioethical committee in 2020 and registered in EudraCT: 2020-004982-37.
Conclusions: The first patient was included in December 2021. Recruitment is planned till June 2023.

EP-119 IMPACT OF IMPROVED TREATMENT COMPLIANCE WITH DELAYED-RELEASE CYSTEAMINE BITARTRATE CAPSULES ON THE COSTS OF END STAGE RENAL DISEASE FOR PATIENTS WITH NEPHROPATHIC CYSTINOSIS IN THE UNITED KINGDOM

Seun Lashilola[1], Weiwei Xu[1], Giacomo Brandi[2], Khashayar Azimpour[2], Sara Carlot[2]

[1]Iqvia, [2]Chiesi

Introduction: Good treatment compliance in nephropathic cystinosis (NC) may lead to healthcare cost-savings by avoiding/postponing end stage renal disease (ESRD). This study compared the lifetime ESRD cost implications of delayed-release cysteamine-bitartrate (DR-CYS) with immediate-release-CYS (IR-CYS) resulting from a difference in compliance for NC patients in the United Kingdom (UK).
Material and methods: A partitioned-survival model involving three health states- ESRD-free, post-ESRD, and death - was developed in Microsoft Excel® to model lifetime patient progression using 1-year cycles. Health state membership was determined by independently modelled survival-curves. Reference survival curves (time-to-ESRD and time-to-death) were derived from Brodin-Sartorius et al.(2012) study. For each treatment arm, the time-to-ESRD curve was modified by a compliance-dependant hazard-ratio (HR); the time-to-death curve was modified by HR suggested by clinical experts. Compliance was measured using a "composite compliance score" (CCS), a function of mean treatment duration and mean-annual cystine level. The annual cost of ESRD, comprising of dialysis and kidney transplant costs, was applied to the time spent in the post-ESRD state. Costs (2020 prices) were discounted at 3.5%.
Results: The ESRD lifetime-costs were £144,000 and £209,243 for DR-CYS and IR-CYS patients respectively, resulting in cost-savings of £65,243/patient in the DR-CYS arm. The difference in dialysis costs (£211,225 [DR-CYS] vs. £295,146 [IR-CYS]) contributed significantly to the DR-CYS cost-savings of £58,379. Scenario analyses showed results were most sensitive to assumptions regarding treatment duration. Assuming mean treatment duration for DR-CYS patients (22 years in the base case) equalled life expectancy (61 years), DR-CYS costs-savings increased to £209,127.
Conclusions: The study results demonstrated that improved compliance with DR-CYS in NC patients resulted in substantial ESRD lifetime cost-savings in the UK. As this study exclusively focused on a vital complication of NC, the results are likely a significant underestimation of true costs-savings.

EP-120 ACUTE PHOSPHATE NEPHROPATHY IN AN ADOLESCENT WITH KIDNEY TRANSPLANTATION

Emre LeventoĞlu[1], Bahar BÜyÜkkaragÖz[1], İpek IŞik GÖnÜl[2], Kibriya Fidan[1], BetÜl ÖĞÜt[2], OĞuz SÖylemezoĞlu[1], Sevcan A. BakkaloĞlu[1], Necla Buyan[1]

[1]Gazi University, Faculty Of Medicine, Department Of Pediatric Nephrology, [2]Gazi University, Faculty Of Medicine, Department Of Pathology

Introduction: Acute phosphate nephropathy is an unusual and overlooked cause of acute or chronic kidney dysfunction. Patients exposed to high doses of phosphorus who had already impaired kidney functions are at greater risk of developing acute phosphate nephropathy.
Material and methods: We present an adolescent with kidney transplantation who developed acute phosphate nephropathy.
Results: A 15-year-old adolescent with autosomal recessive polycystic kidney disease received a living donor kidney transplantation in October 2017. Her phosphate and parathyroid hormone (PTH) levels prior to transplantation were 6.2 mg/dL and 1194.7 pg/mL, respectively.
After transplantation, serum creatinine rapidly decreased to 0.56 mg/dL, but hypophosphatemia of 1.38 mg/dL was present. Oral phosphate solution at a dose of 50 mg/kg/day was prescribed. On the post-transplant 13th day, elevation of serum creatinine to 1.22 mg/dL was noted without any symptoms. Kidney ultrasound and mercapto-acetyl-triglycine scintigraphy were normal.
In kidney biopsy, blue-violet crystal structures compatible with diffuse luminal calcium phosphate deposition, especially in the distal tubular segments were detected in the tubular system. These crystals were positively stained with von Kossa dye and did not reflect under polarized light. With these findings, the patient was diagnosed as acute phosphate nephropathy.
The oral phosphorus solution was discontinued and serum creatinine declined to 0.73 mg/dL. However, after a very short time, the oral phosphorus solution had to be started again (30 mg/kg/d) because the serum phosphorus decreased to 1.1 mg/dL and the patient had profound muscle weakness. Abundant hydration was provided throughout the whole early post-transplant period when PTH levels remained high.
Control allograft biopsy was performed in the 1st post-transplant year, and it revealed persistence in the findings in terms of acute phosphate nephropathy as well as moderate degrees of tubular atrophy and interstitial fibrosis. Nevertheless, in the 4th post-transplant year, no significant deterioration in the kidney functions; her current serum creatinine level is 1.36 mg/dL.
Conclusions: With this case, we would like to emphasize that it should be kept in mind that even when using a drug that seems innocent, especially in kidney transplant recipients, the possibility of causing allograft dysfunction and the physiologic/pathophysiologic mechanisms should be considered while prescribing each drug.

EP-121 EARLY HIGH-DOSE STEROID TREATMENT IS LIFE-SAVING IN RITUXIMAB-RELATED ACUTE LUNG INJURY

İbrahim GÖkÇe, Serim Pul, Ece Demirci Bodur, Özde Nisa TÜrkkan, SerÇin GÜven, Neslihan ÇiÇek, Nurdan Yildiz, Harika Alpay

Marmara UnİversİTy, PedİatrİC Nephrology

Introduction: Rituximab(Rtx) is a chimeric monoclonal antibody against CD20 antigen on the surface of mature B lymphocytes. In nephrology practice, it is used in the treatment of antibody-mediated rejection(AMR) and steroid-dependent nephrotic syndrome. İts reported side effects are ranging from mild reactions such as local erythema to acute lung injury(ALI) and anaphylaxis in acute phase, and increased risk of infection and malignancy in a longer period. Premedication is recommended especially in atopic individuals. In this report, a case of ALI occured in an atopic patient receiving Rtx for AMR is presented.
Material and methods: Case: Our patient with CKD secondary to bilateral hypodysplasia had kidney transplantation from a living donor when he was 12 years old. Seven years after transplantation, kidney biopsy was performed due to high titer of positive donor-specific antibody. AMR was diagnosed and plasmapheresis was initiated after three doses of 500mg pulse methylprednisolone. Seven sessions of plasmapheresis was completed with premedication. IVIG treatment could not be completed because of development of anaphylaxis. Rituximab treatment was planned. After antihistaminic and 40mg methylprednisolone, 500mg Rtx was infused slowly. Mild nausea was occured during the infusion but resolved spontaneously. Sudden onset of dyspnea was observed following the end of the infusion, didnt respond to adrenaline. There was bilateral crackles on pulmonary examination and capillary oxygen saturation was 85% in room air. Computed tomography(CT) showed bilateral diffuse ground glass opacities. After excluding infectious and cardiac etiologies, 250mg methylprednisolone was given due to findings consistent with ALI approximately 6 hours after the RTX infusion. The steroid was tapered within days, after administration of 160mg/day methylprednisolone for the first three days. He recovered completely on the third day. Currently his pulmonary function is completely normal.
Conclusions: Rituximab is a high-risk agent and should be used with attention. One of the most serious side effects is ALI. It is a rare but potentially fatal pulmonary toxicity and severe/permanent damage may occur. Early high-dose steroid therapy is the most important factor

affecting mortality and morbidity in ALI. In possible cases, rituximab related ALI should be kept in mind and high-dose steroid should be started as soon as possible.

EP-122 GENETIC TESTS IN NON-NEUROGENIC NEUROGENIC BLADDER: TWO SIBLINGS WITH OCHOA SYNDROME

Serim Pul[1], İbrahim GÖkÇe[1], Ceren Alavanda[2], ÇaĞri Akin Şekerci[3], Ece Demirci Bodur[1], Özde Nisa TÜrkkan[1], SerÇin GÜven[1], Neslihan ÇiÇek[1], Nurdan Yildiz[1], SelÇuk YÜcel[3], Pinar Ata[2], Harika Alpay[1]

[1]Marmara UnİversİTy, PedİatrİC Nephrology, [2]Marmara UnİversitY, GenetİCs, [3]Marmara UnİversİTy, Urology

Introduction: Neurogenic bladder(NB) is a clinical entity characterized by dysfunctional contraction of detrusor muscle secondary to damaged spinal cord or lower urinary tract obstruction(LUTO). Besides, in non-neurogenic neurogenic bladder(NNNB), findings are similar with NB but without LUTO or spinal injury. Patients with Ochoa syndrome(urofacial syndrome) have a typical smile like crying and NNNB; which occurs due to Heparanase2(HPSE2) mutation. In this report, two siblings with voiding dysfunction diagnosed as Ochoa syndrome are presented.

Material and methods: Cases A six-year-old male was admitted to nephrology outpatient clinic with all day urinary incontinence. On physical examination, marked growth retardation and dysmorphic face were remarkable. He was diagnosed as chronic kidney disease(CKD) secondary to hydronephrotic scarred kidneys. Vesicoureteral reflux(VUR) was not detected in voiding cystourethrography(VCUG). There was no spinal pathology in magnetic resonance imaging(MRI). Posterior urethral valve was not detected on cystoscopy, but severe trabeculation and multiple diverticules were seen. A Low-capacity, high-pressure and hypocompliant bladder was detected by urodynamics and NNNB was diagnosed. He is still being followed up as stage-3 CKD with clean intermittent catheterization(CIC) and anticholinergic treatment.

A ten-year-old girl, sister of the 1st patient, was also admitted to nephrology outpatient clinic with all day urinary incontinence and recurrent urinary tract infection. She always shows a facial expression that resembles crying. She was defined as CKD secondary to hydronephrotic scarred kidney. Left sided VUR was detected in VCUG. High-pressure NB was diagnosed by urodynamics. There was no spinal pathology. She is still being followed up with CIC and anticholinergic treatments.

Genetic examination was performed due to NNNB and dysmorphic face in both siblings and homozygous c.436_437 deletion was found in the HPSE2 gene consistent with Ochoa syndrome.

Conclusions: Early diagnosis and treatment of NB is important to prevent kidney damage. Genetic examination should be kept in mind especially in familial cases and if there was no identified cause for the etiology of NB.

EP-123 IMMUNOHISTOCHEMICAL EXPRESSION PATTERN OF FGFR1, FGFR2 AND RIP5, IN DEVELOPING AND POSTNATAL KIDNEYS OF DAB1-/- (YOTARI) MICE

Nela Kelam, Anita Racetin, Sandra Kostić, Katarina Vukojević, Snježana Mardešić

University Of Split School Of Medicine

Introduction: *Yotari* (Dab1-/-) mice are the autosomal recessive mutant mouse that emerged spontaneously during the generation of mice with a mutation in the gene encoding the inositol-1,4,5-trisphosphate receptor. Our latest research revealed the congenital anomalies of the kidney and urinary tract (CAKUT) phenotype culminating in renal hypoplasia followed by foot process effacement in the kidney glomeruli and functional decline kidney tissue of yotari. This study aimed to determine how functional silencing of the Dab1 gene affects the spatial and temporal patterns of expression of fibroblast growth factor receptor 1 (FGFR1), fibroblast growth factor receptor 2 (FGFR2), and receptor-interacting protein kinase 5 (RIP5) in the developing and postnatal kidneys of yotari-mice as possible predictors of normal kidney formation and function.

Material and methods: Animals were sacrificed on the gestation days E13.5 and E15.5 and postnatal days P4, P11, and P14. Analyses were performed using immunohistochemistry and fluorescent microscopy, followed by quantification of positive cells in kidney substructures. Data were analyzed by two-way ANOVA.

Results: Dab1$^{-/-}$ animal kidneys exhibit decreased FGFR1/FGFR2 expression, suggesting the involvement of the observed markers in generating the CAKUT phenotype leading to renal hypoplasia. Between all developmental periods tested, there was no alteration in the immunoreactivity of RIP5 cells in the substructures of yotari kidneys.

Conclusions: The abundant presence of observed proteins in kidneys and the dynamics of their expression found in this study suggest that FGFR1, FGFR2 and RIP5 play essential roles not only in early metanephric mesenchymal patterning, ureteric bud branching morphogenesis, nephrogenesis, and nephron progenitor survival but also in the maintenance of overall homeostasis and the maturation of kidney structures in the postnatal phase. Our results emphasize the crucial significance of the examined markers throughout normal kidney development and their potential involvement in renal pathology and diagnostics, where they could potentially serve as biomarkers and therapeutic targets.

EP-124 NEPHRITIS FOLLOWING SARS-COV-2 VACCINE: CAUSAL OR INCIDENTAL CORRELATION ?

Luigi Annicchiarico Petruzzelli[1], Vittorio Serio[1], Giuseppina Marino Marsilia[3], Severo Campione[3], Francesca Diomedi Camassei[2], Daniela Molino[1], Gabriele Malgieri[1], Carmine Pecoraro[1]

[1]Aorn Santobono Pausilipon Childrens Hospital-naples-italy, [2]Irccs Ospedale Pediatrico Bambino Gesu Childrens Hospital - Rome - Italy, [3]Aorn Antonio Cardarelli Hospital-naples-italy

Introduction: The global coronavirus 2019 (COVID-19) pandemic required vaccination even in children to reduce infection.

We describe two patients with nephritis following SARS-CoV-2 vaccine: the first one developed acute kidney injury (AKI) (due to) and minimal change disease (MCD) nephrotic syndrome following injection of the BNT162b2 COVID-19 vaccine (Pfizer-BioNTech) and the second one developed gross hematuria, after second injection of the BNT162b2 COVID-19 vaccine .

Material and methods: In the first case a 12-year-old previously healthy boy was referred to our hospital following the development of peripheral edema and nephrotic range proteinuria. Nine days before he had received the first injection of the vaccine. Seven days after injection, he developed leg edema, which rapidly progressed to anasarca with significant weight gain. On admission, serum creatinine was 1.5 mg/dL and 24-hour urinary protein excretion was 5 grams with fluid overload. Kidney biopsy was performed showed minimal change disease, tubular obstruction with cytoplasmic degeneration; Immunofluorescence (IF) was negative, on EM GBM with aspects of rehash, stretches of capillary wall with tortuous course and aspects of collapse, swelling and extensive fusion of the pedicels. There were no electron-dense deposits As kidney function continued to decline over the next days, empirical prednisone 1g mq/d treatment and renal replacement therapy with ultrafiltration were starte. Seven days

after steroid therapy, kidney function began to improve, gradually returning to normal.
In the second case a 14-year-old boy, with a history of an episode of previous HSP without renal involvement, developed gross hematuria Henoch-Schönlein Purpura nephritis associated, with nephritic proteinuria two day after BNT162b2 COVID-19 vaccine (Pfizer-BioNTech) second dose. Kidney biopsy was performed showing mild mesangial hypercellularity, crescents and immunofluorescence mesangial IgA staining. Pulses and oral steroid therapy plus MMF therapy was started with clinical remission.
Results: The association between nephritic, nephrotic syndrome, AKI has been previously reported in pediatric and adults following covid 19 vaccines.
Conclusions: Pathogenesis is not completely understood but current literature suggest that T cell dysfunction / triggering might be the main underlying immunological mechanism.

EP-125 THE ROLE OF URINARY N-ACETYL-β-D-GLUCOSAMINIDASE IN EARLY DETECTION OF TUBULAR DAMAGE AND ACUTE KIDNEY INJURY AMONG PEDIATRIC PATIENTS WITH NEOPLASTIC DISORDERS

Erika Biró[2], István Szegedi[2], Csongor Kiss[2], Anna V.oláh[1], Mark Dockrell[3], Robert G.price[4], Tamás Szabó[2]

[1]Department Of Laboratory Medicine, Faculty Of General Medicine, University Of Debrecen Hungary, [2]Department Of Pediatrics, Faculty Of General Medicine, University Of Debrecen, Hungary, [3]Epsom And St Helier University Hospital Nhs Trust, Uk, [4]King's College, University Of London, Uk

Introduction: Renal injury is a frequent complication during the treatment of childhood cancers, due to tumor lysis syndrome, nephrotoxic drugs, septic periods. As a consequence of multiple acute AKI episodes with variable and multifactorial origin along the course of oncological treatment, there is a notably higher chance for the occurance of chronic kidney disease at long term.
Material and methods: We investigated the diagnostic value of urinary N-acetyl-β-D-glucosaminidase (uNAG) as an early marker of renal tubular damage and acute kidney injury. This retrospective clinical analysis included 415 uNAG measurements in 35 children with neoplastic disorders, who had serial uNAG tests (min. 5 samples/patient). Renal function was determined by cystatin-C and creatinine based GFR, relative increase of uNAG index, along with patients' general condition and medication were registered. We focused on detecting both clinical and subclinical AKI episodes (according to Biomarker-Guided Risk Assessment with the using pRIFLE criteria and /or elevated uNAG levels) and the incidence of chronic kidney damage.
Results: 63 episodes in 27 patients were identified during the observation period with positivity at least in one parameter of the kidney panel. We detected 20/63 clinical and 12/63 subclinical renal episodes. In 28/63 episodes only uNAG values was elevated without acute therapeutic consequence. Almost complete recovery of tubular damage was observed in 13/27 patients, while chronic tubuloglomerular injury occurred in 5/27 patients.
Conclusions: We found that repeated measurements of uNAG, in combination with other renal parameters, is a sensible, non-invasive and economic marker for monitoring renal function in both acute and follow up periods. The serum creatinine value is not an early marker of renal damage and its level is often measured to be low among cancer patients due to malnutrition and decreased muscle mass. Our observation highlight the importance of using multi-marker AKI signaling approach to assess changes in kidney/tubular function in pediatric hemato-oncology patients.

EP-126 DE NOVO SOX4 VARIATION ASSOCIATED WITH RENAL DEVELOPMENT ABNORMALITY

Laurene Dehoux[1], Marina Avramescu[1], Anne Couderc[1], Mathilde Grapin[1], Alban Lermine[3], Rosa Vargas-poussou[2], Olivia Gillion-boyer[1], Laurence Heidet[1]

[1]Pediatric Nephrology Department,hopital Necker Enfants Malades, Aphp Paris, France, [2]Genetic Department, Hopital Europeen Georges Pompidou, Paris, France, [3]Director Of Bioinformatics And I.t. Department, Sequioa Genomic Plateform, France

Introduction: SOX4 belongs to group C of SRY-related (SOX) transcription factors which play key roles in multiple developmental pathways, including neurogenesis and skeletogenesis. SOX 4 pathogenic variants are associated to developmental delay, intellectual disability, and mild facial and digital morphological abnormalities
Material and methods: We reported for the first time a 3-year-old girl with the association of intrauterine and postnatal growth retardation, oligoamnios, antenatal micro and macrocystic kidneys, neonatal kidney failure rapidly progressing to ESRD from 12 months old, feeding difficulties, microcephaly, retarded psychomotor and intellectual development, especially in language and walk, neonatal hypotonia and spastic diplegia with right reducible equinovarus foot, distal end of spinal cord, thin aspect of chiasma and optic nerves without vision defect, transitory hepatic cytolysis, adenoid and tonsil hypertrophy leading to hypoacousia and sleep apnea, mild facial dysmorphism with wide mouth with a cupid bow, posteriorly rotated ears.
Results: The child was found through trio-based genome sequencing to carry a de novo missense SOX4 (NM_003107.3) variant c.176_177delinsAA (p.Ile59Lys). That variant was never reported, but a de novo variant affecting the same amino-acid (c.176T>G p.Ile59Ser) was reported in a French child presenting with mild learning difficulties, deep-set eyes, infra-orbital grooves, upturned nares, wide mouth with cupid bow and full lips, mild 5th finger clinodactyly, and dysplastic 5th toenails (Zawerton et al. Am. J Hum Genet 104:246-259, 2019).
Conclusions: Although SOX4 was shown to be required for normal renal development in vivo in mice (Huang et al Develop Dynamics 242:7906799 2103), this case is, to our knowledge, the first case of renal phenotype associated with a SOX4 probably pathogenic variant. Although additional cases will be required before asserting causality link between the variant and the renal disease, we believe this case may be the first case of renal development defect due to SOX4 pathogenic variant.

EP-127 A RARE CASE OF CHILDHOOD RENAL MANIFESTATION OF SARCOIDOSIS IN A 10 YEARS OLD GIRL.

Monika Abramczyk, Ilona Zagożdżon, Aleksandra Żurowska

Department Of Paediatrics, Nephrology And Hypertension, medical University Of Gdańsk

Introduction: Sarcoidosis is a multiorgan inflammatory disease characterized by formation of non-necrotizing granulomas. The diagnosis is based on the clinical presentation, granulomatous inflammation on tissue biopsy and exclusion of alternative causes of granulomatous disease. Common manifestations include uveitis/iritis, lymphadenopathy, hepatosplenomegaly, arthritis, parenchymal lung disease and skin rash. Elevated serum ACE levels are characteristic. Kidney involvement is exceptional in children.
Material and methods: The aim of our study was to present a rare case of kidney sarcoidosis in a child. A ten years old girl was admitted to hospital due to acute kidney injury (AKI) after a gastrointestinal infection.

Stool culture was negative and despite parenteral fluids AKI persisted (creatinine level -1,41mg/dl, eGFR 46 ml/min/1,73m2).
Results: Persistent sterile pyuria and albuminuria and a high CRP level were noted. A renal ultrasound revealed bilateral microabscess-like lesions in enlarged kidneys. Following lack of improvement to wide-spectrum antibiotics a kidney biopsy was performed showing a granulomatous interstitial nephritis. Mycobacterium tuberculosis, Mycoplasma pneumoniae and Chlamydia trachomatis infections were excluded. Elevated serum ACE (79,2 U/L) confirmed the diagnosis of kidney sarcoidosis. Extrarenal involvement was excluded by lung HR-tomography and abdominal MRI. The girl was treated with pulses of methyl-prednisolone followed by oral steroids for 2 years. Kidney function improved but eGFR was decreased. 6months following treatment multiple renal scars were visualized by MRI and chronic kidney disease was diagnosed (creatinine 1,33mg/dl, eGFR 51,5 ml/min/1,73m2).
Conclusions: 1. Single kidney organ manifestation of sarcoidosis in children is exceptional but needs to be considered among the differential diagnosis of atypical interstitial nephritis.
2. Despite early treatment and improvement following steroids permanent kidney damage may ensue due to renal scarring.

EP-128 VALUE OF URINARY BIOMARKERS IN DIAGNOSIS OF PEDIATRIC ACUTE KIDNEY INJURY AND PROGNOSIS OF ITS COURSE

Antanas Naujokaitis[1], Zina Doviltyte[1], Diana Dobiliene[2], Jurate Masalskiene[2], Sarunas Rudaitis[2]

[1]Medical Academy, Lithuanian University Of Health Sciences, Kaunas, Lithuania, [2]Department Of Pediatrics, Medical Academy, Lithuanian University Of Health Sciences, Kaunas, Lithuania

Introduction: Objectives: To determine the value of urinary neutrophil gelatinase-associated lipocalin (uNGAL) and interleukin 18(uIL-18) in the diagnosis of pediatric acute kidney injury (AKI) and prognosis of its course.
Material and methods: The study included 138 subjects: 107 critically ill patients who met the inclusion criteria and 31 healthy children as the control group. Serum creatinine; urinary creatinine (uCr), uNGAL, and uIL-18 were assessed in the case group on days 1 and 3. The case group was divided into AKI (n=32) and non-AKI (n=75) subgroups according to pRIFLE criteria.
Results: The uNGAL level on day 1 was 0.17 (0.01–2.68) ng/ml in the control group, 2.5 (5.23-6.78) ng/ml in the non-AKI subgroup and 2.99 (1.44-10.45) ng/ml in the AKI subgroup (P=0.04). On day 3 the levels were 0.17 (0.01–2.68) ng/ml, 1.84 (0.42-6.88) ng/ml and 7.56 (0.79-12.56) ng/ml respectively (P=0.018). The uNGAL/uCr ratio on day 1 was 0.22 (0.03–6.46) ng/ml in the control group, 4.67 (1.1–14.11) ng/mg in the non-AKI subgroup, and 12.10 (2.47–90.27) ng/mg in the AKI subgroup (P=0.007). On day 3 the ratios were 0.22 (0.03–6.46) ng/ml 3.94 (1.79–14.66) ng/ml and 12.48 (2.62–14.48) ng/ml respectively (P=0.015). In the AKI subgroup, the uIL-18 level on day 1 was 60.99 (56.17–68.67) ng/l in children whose AKI resolved within a 5-day period, and 69.78 (62.17–74.22) ng/l in children whose AKI persisted or progressed. The uIL-18 level of > 69.24 pg/mL on day 1 was associated with an 8-fold increased risk of AKI progression (OR = 8.33, 95% CI = 1.39 to 49.87, P = 0.023).
Conclusions: The uNGAL level and the uNGAL/uCr ratio on days 1 and 3 were found to be reliable prognostic biomarkers of AKI. The uIL-18 level was found to be a significant prognostic factor for AKI progression in the AKI subgroup.

EP-129 NEW ONSET NEPHROTIC SYNDROME IN AN ADOLESCENT BOY RELATED TO COVID-19

Seçil Arslansoyu Çamlar[1], Özgür Özdemir Şimşek[2], Gökçen Erfidan[2], Neslihan Güney[3], Fatma Mutlubaş[1], Demet Alaygut[1], Dilek Yılmaz Çiftdoğan[4], Belde Kasap Demir[5]

[1]Izmır Faculty Of Medicine, Department Of Pediatrics, Division Of Nephrology, University Of Health Sciences Turkey, Izmir, Turkey, [2]Department Of Pediatrics, Division Of Nephrology, University Of Health Sciences Izmir Tepecik Training And Research Hospital, Izmir, Turkey, [3]Department Of Pathology,university Of Health Sciences Izmir Tepecik Training And Research Hospital, Izmir, Turkey, [4]Department Of Pediatrics, Division Of Pediatric Infectious Disease, Izmir Katip Çelebi University, Izmir, Turkey, [5]Department Of Pediatrics, Division Of Pediatric Nephrology And Rheumatology, Izmir Katip Çelebi University, Izmir, Turkey

Introduction: Kidney involvement associated with COVID-19 is proteinuria, hematuria, collapsing glomerulonephritis, nephritis and acute kidney injury. Nephrotic syndrome(NS) is a rare renal manifestation.
Material and methods: A 16-year-old boy presented with two-week history of progressive bilateral eyelid and facial swelling of both lower limbs. He had no fever, sore throat, cough, breathing difficulty or anosmia. He displayed a weight gain of 8kg since the onset of swelling. On physical examination he had a bodyweight of 73kg(50-75p), height of 179cm(75-90p), blood pressure of 150/92mmHg(>95p+12). He had periorbital and +4 pretibial pitting edema. Respiratory sounds were poorly heard in bilateral basal areas. He had scrotal edema.
His parents were diagnosed with COVID-19 infection 3weeks ago. The other 2 siblings living together in the same house did not have any symptoms related to COVID.
Laboratory tests revealed urea 33mg/dL, creatinine 0.8mg/dL, albumin 1.8g/dL, serum total cholesterol, triglyceride, and low-density lipoprotein cholesterol were high. Urine analysis revealed protein (+4), and urine microscopy showed dysmorphic erythrocytes. Urine protein was 16g/day. Serology revealed normal serum complements and negative levels for ANA. The covid-19 test with PCR was negative. The COVID-19 IgM and IgG ELISA antibody test was positive.
Results: Renal biopsy was performed. He had tip variant of focal segmental glomerulosclerosis on renal biopsy. Immunofluorescence studies revealed only segmental +1/+2 mesangial granular IgM deposition in the basement membrane. The edema was treated with furosemide. After five doses of pulse methylprednisolone (1gr/day), we continue with oral steroid with 2mg/kg(60mg). On the 14th day of treatment, he was in remission. The urine output had improved, and his weight had trended down to 67kg. After recovery from NS relapse and Covid-19, the patient was discharged from our hospital.
Conclusions: The clinical presentation and well respond to prednisone treatment of new-onset NS associated with COVID-19 infection is similar to NS associated with other viral infections.

EP-130 RISK FACTORS FOR URINARY TRACT INFECTIONS IN INFANTS WITH HIGH GRADE VESICOURETERAL REFLUX (VUR) – A MULTICENTER PROSPECTIVE STUDY.

Anna Kranz, Aleksandra Żurowska

Medical University Of Gdansk

Introduction: High grade VUR (III-V) can be associated with hypertension and/or end stage renal disease due to the frequent coexistence of kidney dysplasia and acquired damage caused by scarring following

urinary tract infections (UTIs). The aim of the study was to assess the potential risk factors for developing UTI in infants with III-V VUR.
Material and methods: A prospective study was performed at 6 university centres in Poland identifying over a 3 year period consecutive children with grade III-V VUR diagnosed in the first 12 months of life. Risk factors for developing UTI in infancy were analyzed.
Results: 218 infants (137 boys/81 girls) were included in the study: 25,7% with grade III, 34,8% grade IV and 39,5% grade V. Primary VUR was predominant (87,6%). 126 infants demonstrated bilateral and 92 unilateral VUR. UTI developed during the first year of life in 120/218 infants (55,0%); 22/120 (20%) demonstrated urosepsis. 29% (63/218) of initial UTI's occurred by 2 months of life. Median time of initial UTI was 8 weeks. UTI's were distributed equally among infants with grade III (64%) vs. IV-V (51.8%); with unilateral (58.7%) vs. bilateral (52,4%) VUR and in those with secondary (63%) vs. primary VUR (54%). UTI was significantly more common in girls (77,8%) vs. boys (42%) [p<0.0001].
Conclusions: The risk of developing UTI in infant with high grade VUR reaches 55% by the age of 12 months. The initial UTI frequently occurs early in the first 2 months of life. Risk factors for UTI are age <2months and female gender.

EP-131 ICAM-1 LEVEL IN CHILDREN WITH GLOMERULAR DISEASES

Aksana Kandratsenka[1], Hanna Bialkevich[2], Ina Kazyra[2], Aleksandr Sukalo[2]

[1]10th City Childrens Clinical Polyclinic, *[2]Belarusian State Medical University*

Introduction: The intercellular adhesion molecule-1 (ICAM-1) plays an important role in numerous cellular immune responses, and can be considered as a possible marker of the progression of glomerular diseases (GD).
Material and methods: We examined 93 patients with CKD, who were observed in Belarusian Center of pediatric nephrology and renal replacement therapy 2nd Childrens hospital Minsk: primary immune GD (n=14), secondary immune GD (n=28), nonimmune GD (n=29), kidney transplant recipients (n=7). The comparison group included patients with non-glomerular nephropathies (n=29). Healthy children without renal pathology (n=5) were examined as a control group. The concentration of ICAM-1 in the blood serum and urine of patients was determined by the ELISA method.
Results: In the majority of patients with immune GD, ICAM-1 was detected in both serum and urine in 29/40 (72.5%). Moreover, there was no significant difference in the concentration of ICAM-1 between the groups of patients with primary and secondary immune GD. In the majority of patients with non-glomerular CKD, it was also possible to determine the concentration of ICAM-1 both in serum in 15/17 (88%), $\chi 2=8,33$, p<0,01vs immune and in urine in 3/7 (43%). It is interesting to note that only in 10/27 (37%) of patients with nonimmune GD had an intercellular adhesion molecule in the blood serum, $\chi 2=11,15$, p<0,001vs immune. In the urine of this group ICAM-1 was not determined. Only two kidney transplant recipients were able to determine the concentration of ICAM-1 in the blood serum (the cause of ESKD in these children were diseases of non-immune etiology).
Conclusions: Thus, it can be concluded that further studies to determine ICAM-1 in blood serum and urine as a marker of glomerulopathy progression may be of interest in patients with immune glomerulopathies.

EP-132 EVALUATION OF CARDIOVASCULAR RISK FACTORS AND THE RELATIONSHIP BETWEEN RENALAZ IN CHRONIC KIDNEY PATIENTS IN CHILDHOOD

Zeynep GÖktÜrk ErdoĞan[1], Beltİnge DemircioĞlu KiliÇ[2], Seyithan Taysi[3], Mehtap Akbalik Kara[2], Mithat BÜyÜkÇelik[2], AyŞe Balat[2]

[1]Gaziantep Abdulkadir Yuksel State Hospital, Department Of Pediatrics, Gaziantep/turkey, *[2]Gaziantep University, Department Of Pediatric Nephrology, Gaziantep/turkey*, *[3]Gaziantep University, Faculty Of Medicine, Department Of Medical Biochemistry, Gaziantep, Turkey*

Introduction: Cardiovascular sistem (CVS) complications are the most important causes of mortality and morbidity in children with chronic kidney disease (CKD). Renalase is a monoamin oxidase which is released from the kidney and degrades circulating catecholamines and regulated by renal function, amount of catecholamine. The aim of this study was to determine the risk factors for CVS complications in children with CKD and evaluate the relationship between risk factors and renalase.
Material and methods: Children who were diagnosed as CKD (n:108) with GFR below 60 ml/min/1.73 m^2 and healthy children (n:50) were included in the study. Patients with CKD were divided into stage 3-4 CKD (n:50), haemodialysis (HD) (n:14) and peritoneal dialysis (PD) (n:44) groups. Renelase levels were measured by ELISA Biotek ELx800 (USA).
Results: The median value of renalase was measured 30.28 ng/ml (17.73-39.6) in healthy volunteers, 27.4 ng/ml (15.65-37.94) in stage 3-4 CKD, 23.45 ng/ml (15.85-34.71) in HD patients, and 21.17 ng/ml (17.11-33.24) in PD patients. Renalase levels were found to be lower in CKD, but there was no significant correlation with CKD (p=0.185). The patients were evaluated in terms of cardiovascular risk factors. 15.7% hypertension (HT), 38.9% hypoalbuminemia, 67.6% anemia, 49.1% hypercholesterolemia, 38% hypertriglyceridemia, 13.9% CRP elevation and 1.9% obesity were detected. When the cardiovascular risk factors were evaluated according to groups; hypoalbuminemia (70.5%), anemia (88.6%), total cholesterol (70.5%) and LDL elevation (68.2%) were most common in the PD group, hyperparathyroidism was seen mostly in the stage 3-4 CKD group (74%) and hypertrigiliseridemia (57.1%) was most frequent in the HD group (p <0.05). There was no correlation between hematological and biochemical parameters and renalase levels.
Conclusions: Cardiovascular risk factors such as anemia, hypoalbuminemia and hyperlipidemia were found to be increased in children with CKD, but there was no significant relationship between blood renalase levels and cardiovascular risk factors.

EP-133 EVALUATION OF CLINICAL COURSE AND RENAL PROGNOSIS IN CHILDREN WITH PRIMARY VESICOURETERAL REFLUX

Emel Saribas, Cagla Cagli, Derya Cevizli, Bahriye Atmis, Aysun K. Bayazit

Cukurova University, Department Of Pediatric Nephrology, Adana, Turkey

Introduction: Vesicoureteral reflux (VUR) is one of the most important causes of urinary tract infection and renal failure in children.VUR may be an isolated abnormality (primary VUR) that is diagnosed mostly after urinary tract infection (UTI).The aim of this study was to evaluate the clinical course and renal prognosis in children with primary VUR.
Material and methods: We retrospectively analyzed of 199 children with primary VUR who were followed-up in our tertiary center. We reviewed medical records of children with VUR and recorded demographic features, clinical course, laboratory and radiological data of the patients.
Results: The median age of 199 children was 48 (0.03–192) months.82 children (41.2%) were male.The median follow-up period was 39 (2-209) months. In a total of 293 reflux units, 36 (12.3%) were grade 1,46

(15.7%) were grade 2, 88 (30%) were grade 3,66 (22.5%) were grade 4 reflux and 57 (19.5%) were grade 5 reflux. Boys were diagnosed at younger age as compared to girls (24 vs 72 months)($p<0.001$).The most common presenting symptoms were UTI, antenatal hydronephrosis and abdominal pain, respectively. While reflux resolved spontaneously in 44 (22.1 %) of the patients, surgery was required in 132 (45.1%) patients. Of 132 patients who underwent surgical treatment, 70 (53%) underwent subureteric injection of dextranomer/hyaluronic acid, 52 (39.4%) patients ureteroneocystostomy, and 10 (7.6%) patients nephroureterectomy. The median age at diagnosis was 16 months for those who recovered spontaneously, and 60 months for those requiring surgical treatment($p<0.001$).The scar rate in DMSA scan was significantly higher in those requiring surgical treatment (62.5% vs 31.8%,$p<0.001$). Patients with scarring in DMSA had a later diagnosis age and lower eGFR at the last visit. As the VUR degree increased in the patients, the rate of kidney scarring increased($p<0.001$). In a total of 132 patients who required surgical treatment, 19 (14.4%) had reflux after surgery. Stage 2 and higher stage CKD developed in 23(11.6%) patients during follow-up.
Conclusions: Later age at diagnosis and high grade of VUR were risk factors for renal scarring in children with primary VUR. Spontaneous resolution of VUR was seen in those diagnosed at an earlier age.

EP-134 ARE FURTHER INVESTIGATIONS NEEDED IN CHILDREN WITH HORSESHOE KIDNEY?

Bahriye Atmis, A. Asena Emiroglu Taskin, Derya Cevizli, Aysun K. Bayazit

Cukurova University, Faculty Of Medicine, Department Of Pediatric Nephrology, Adana, Turkey

Introduction: Horseshoe kidney (HSK) is the most common renal fusion anomaly that is usually asymptomatic, but that increases the risks of kidney stones and urinary tract infection (UTI). We aimed to identify the clinical course, kidney outcomes and prognosis in children with HSK.
Material and methods: We retrospectively analyzed of 46 children with HSK who were followed-up in our center. We reviewed medical records of children with HSK and recorded demographic features, clinical course, laboratory and radiological data of the patients.
Results: The mean age of 46 children was 122±55.6 months. Twenty children (43.5%) were male and 26 children (56.5%) were female. The median age at diagnosis was 40 (1-180) months. The median age at diagnosis was found lower in male patients than female patients (13.5 months vs 54 months) ($p=0.022$). Parental consanguinity was found in 18 (39.1%) patients. While 13 (28.3%) patients were coincidentally diagnosed with ultrasonography, the second most common presenting symptom (21.7%) was UTI. UTI was seen in 26 (56.5%) of the patients during the follow-up period. Of the 25 (54.3%) patients who underwent voiding cystourethrography, four (8.7%) had unilateral vesicoureteral reflux (VUR) and one (2.2%) had bilateral VUR. Renal cortical scarring was found in 14 (30.4%) of the patients. While the mean differential function of the right kidney was 48.3±10.9% in DMSA, the mean differential function of the left kidney was 51.9±10.9%. The mean estimated glomerular filtration rate was 144±9.9 ml/min/1.73m^2 at last visit. Two (4.3%) of patients had stage 2 chronic kidney disease (CKD). Other urological abnormalities including nephrolithiasis, kidney cyst, hypospadias and ureteropelvic junction obstruction were seen in seven (15.2%) of patients. One patient had Cornelia de Lange syndrome and one had Schimke immuno-osseous dysplasia. The mean eGFR was not found significantly different in patients with other urological abnormalities than those without ($p=0.103$).
Conclusions: Although children with HSK have a good prognosis, they should be followed for progressive CKD. In addition, children with HSK should be carefully examined for accompanying other abnormalities and further investigations have to be considered.

EP-135 USE OF CONTINUOUS GLUCOSE MONITORING SYSTEM FOR MANAGEMENT OF PATIENTS WITH FANCONI-BICKEL SYNDROME

Justė Parnarauskienė[1], Giedrė Maželytė[2], Rūta Repečkienė[2], Viktoras Sutkus[1], Rimantė Čerkauskienė[1]

[1]Center For Coordination Of Rare Diseases, Vilnius University Hospital Santaros Klinikos, Vilnius, Lithuania, [2]Vilnius University, Faculty Of Medicine, Vilnius, Lithuania

Introduction: Fanconi-Bickel syndrome (FBS) is a rare autosomal recessive disorder characterized by impaired glucose liver homeostasis and proximal renal tubules dysfunction. It is caused by defects in the glucose transporter 2 (GLUT2) encoded by SLC2A2 gene. Clinical manifestations of FBS include acidosis, fasting hypoglycemia, postprandial hyperglycemia, and severe growth disorder. There are limited data on optimal nutritional therapy and a lack of comprehensive clinical evaluation in FBS-treated patients. We aimed to evaluate the efficacy of a diet with extended release waxy-maize cornstarch (ERWMC) and continuous glucose monitoring (CGM) on metabolic response and growth.
Material and methods: We report three cases with FBS treated in Vilnius University Hospital Santaros Klinikos with uncooked corn starch (UCCS) or ERWMC (Glycosade®), carbohydrate-controlled diet and applied CGM system (FreeStyle Libre 2, Abbott) using a smartphone.
Results: Patient 1 glycemic control with UCCS and carbohydrate-controlled diet showed slow but steady growth (height always about -9 cm below 3rd percentile of height by age), no significant decrease in liver length or lipid changes. Growth retardation (height growth from -2 cm below 3rd percentile of height by age at 3 months to -20 cm below 3rd percentile of height by age at 4 years) and metabolic changes were most prominent for Patient 2 due to lack of parental compliance to the prescribed diet and UCCS assignments regime. Adequate growth, 14 mm decrease in liver length and positive lipid changes (reduction of total cholesterol by 1.5 mmol/l) have been observed in Patient 3 after the introduction of ERWMC and CGM system.
Conclusions: For Patients with FBS a CGM system can be a useful tool to dose ERWMC, to choose food properly and to keep normal glycemia around the clock to ensure good metabolic response and patient growth.

EP-136 DOES COVID-19 FACILITATE NEW-ONSET DIABETES MELLITUS AFTER TRANSPLANTATION

Nurdan Yildiz[1], Ece Demirci Bodur[1], Neslihan ÇiÇek[1], Ahmet Kahveci[2], SerÇin GÜven[1], Özde Nisa TÜrkkan[1], Serim Pul[1], Belma HaliloĞlu[2], İbrahim GÖkÇe[1], Harika Alpay[1]

[1]Marmara University Pediatric Nephrology, [2]Marmara University Pediatric Endocrinology

Introduction: New-onset diabetes mellitus after transplantation(NODAT) is an important complication that may be caused by immunosupressive drugs(steroids and calcineurin inhibitors), existing or new onset risk factors such as obesity, infections, family history and rejection attacks. It usually occurs in the first three months after transplantation. It has been shown that new onset DM is associated with COVID-19. We report a patient who had a renal transplantation three months ago and presented with hyperglycemia during SARS-Cov-2 infection(COVID-19).
Material and methods: An eight-year-old girl had a kidney transplant from her mother three months ago due to end-stage kidney disease of unknown etiology. Her immunosupressive regimen was prednisolone, tacrolimus and mycofenolate mofetil. Her blood tests for routine outpatient control revealed a venous glucose of 1049 mg/dL, blood and urine

ketone were negative and she had no acidosis. A week ago, since she had runny nose and her brother had COVID-19 infection, SARS-Cov-2 RT-PCR test was performed and was negative. At admission, she had no complaints but repeated RT-PCR test was positive, her physical examination and chest X-Ray were normal. On repeated measurement, blood glucose was 1000 mg/dl. Subcutaneous insulin was started, tacrolimus was changed to cyclosporin A. HbA1c was 10.5% whereas it was 6.5% ten days ago. In the first 5 days, her blood sugar was resistant to high-dose insulin and remained >400 mg/dl. Her need for subcutaneous insulin was gradually decreased and stopped within ten days. She is still using prednisolone and cylosporine A and is closely monitored for blood sugar without insulin treatment.
Conclusions: A rapid increase in HbA1c in ten days and concomitant COVID-19 in our patient suggest that COVID-19 may have facilitated the development of acute hyperosmolar hyperglycemia.Serum glucose levels should be monitored closely in kidney transplant recipients during COVID-19,especially in the early post-transplant period.

EP-137 THERAPEUTIC PLASMA EXCHANGE IN A PEDIATRIC NEPHROLOGY UNIT

Adriana Monica Bungardi, Bogdan Bulata, Cornel Aldea, Dan Delean

Emergency Hospital For Children Cluj-napoca, Romania

Introduction: Therapeutic plasma exchange (TPE) is considered to have an immunomodulating effect, not counting the removal of pathogenic substances or components in the blood given certain conditions. The aim of this study is to describe the conditions that required TPE according to The American Society for Apheresis (ASFA) 2019 indications in our tertiary referral center.
Material and methods: We conducted a retrospective study between January 2003 and December 2021 on pediatric patients suffering from a condition requiring TPE at some point during the course of illness.
Results: There were 44 patients (65% females) included, with a median age of 146.5±52 months at the time of therapy. The median length of hospitalization was 14.5±43 days in the Nephrology Department/ICU. 40 (90%) of the disorders were included in categories for which TPE is accepted as first- or second-line therapy, with donor fresh frozen plasma as the only replacement fluid. A total of 169 TPE procedures were performed, with a median of 4, the apheresis schedule being determined by the patients condition. The kidney and the nervous system were the most affected organs (17 and 16, respectively), most of them requiring adjunctive immunosuppressive therapy. The third cause was acute poisoning (Amanita mushroom ingestion), found in 11/44 cases. Renal replacement therapy - hemodialysis was required in 13 (76%) cases. A low-risk allergic reaction and two citrate-induced hypocalcemia were the only complications related to the extracorporeal procedure.
Conclusions: TPE can reduce further damage and possibly reverse the pathologic process. According to the ASFA's indications, most of the disorders were included in categories for which this extracorporeal blood purification technique is accepted as first- or second-line therapy. In the studied group, the majority of patients had a favorable outcome, similar to the data reported in the literature. TPE safe with no significant adverse events.

EP-138 NEW CRITERIA FOR RENAL SCINTIGRAPHY AFTER URINARY TRACT INFECTION – ARE THOSE ADEQUATE?

Marta Carvalho, Teresa Almeida Lopes, Filipa Cunha, Catarina Neves

Hospital Distrital Da Figueira Da Foz

Introduction: The recommendations for imagiological investigation after urinary tract infection (UTI) remain controversial, mainly for renal scintigraphy (RS). In our hospital, RS was performed more than 6 months after all febrile UTI. After 2018, new criteria were applied, and RS was only performed in atypical or recurrent UTI or when abnormalities in renal ultrasonography (RUS) were found. Our aim was to analyse all RS to evaluate the new criteria adequacy.
Material and methods: Retrospective study of patients diagnosed with UTI from 2011 to 2020 that underwent RS. Analysis of the new criteria adequacy and their correlation with the RS results. Altered RS was defined as presence of renal scars or renal differential function above 10%.
Results: In this study, 232 children were included. Their median age at the UTI was 14 months old and 60% were female. Infection with non-*Escherichia coli* pathogens was identified in 17% of the cases and 22% of children had recurrent UTI. Abnormal findings were identified in 7% of the RUS. From the 83 RS performed according to the new criteria, 22 were altered (27%), while in the 149 RS that would not be performed, 18 (12%) were altered.
The relation between the new criteria to perform RS and altered RS was statistically significative ($p<0.05$), with an odds ratio of 2.6 (1.3-5.2). Considering each criterion individually, the relations between recurrent or atypical UTI and altered RS were not statistically significative, while between abnormal findings in RUS and altered RS was statistically significative ($p<0.05$).
Conclusions: We verified that the new criteria increase the probability of altered RS and avoid the realization of a significant number of unjustified RS. However, even in the absence of criteria, a percentage of lesions can be identified. More studies are needed to clarify the impact of these lesions in children's prognosis.

EP-139 DIFFERENT COURSE OF BONE DISEASE IN TWO SIBLINGS WITH NEPHROPATHIC CYSTINOSIS ON CYSTEAMINE TREATMENT

Maria Fourikou[1], Stella Stabouli[2], Efstratios Kasimatis[3], Erasmia Sampani[3], Nikolaos Laliotis[4], Athina Ververi[1], Persephone Augoustides-savvopoulou[2], Konstantinos Kollios[1]

[1]Third Department Of Paediatrics, Hippokration General Hospital, Thessaloniki, Greece, *[2]First Department Of Paediatrics, Hippokration General Hospital, Thessaloniki, Greece*, *[3]Department Of Nephrology, Hippokration General Hospital, Thessaloniki, Greece*, *[4]Department Of Orthopaedics, Inter Balkan Medical Center, Thessaloniki, Greece*

Introduction: Cystinosis is an autosomal recessive inherited disease caused by mutations in the CTNS gene, affecting lysosomal cystine transport, leading to excessive intracellular cystine accumulation. Cysteamine therapy has improved patient survival and brought to light the presence of "cystinosis metabolic bone disease" in patients presenting with severe bone symptoms despite treatment.
Objective: This study aimed to describe the clinical course of two siblings with nephropathic cystinosis.
Material and methods: Clinical presentation, investigations and management were followed from patient health records.
Results: The index case is an 18-year-old male who presented at the age of 12 months with hypotonia, growth delay, rickets and Fanconi syndrome. Cystine levels in leucocytes were elevated and genetic analysis revealed homozygosity for the Q264X loss of-function mutation in the CTNS gene. Cysteamine treatment was started after diagnosis but his compliance to treatment over the years became poor. He developed CKD with severe hyperparathyroidism exacerbating the bone deformities and resulting in walking disability at the age of 16 years. One year later he progressed to end-stage renal disease and underwent renal transplantation at the age of 18. Currently he is on a twice daily dosage regimen with

delayed - release mercaptamine bitartrate. The second case, a 10-year-old male, is the younger sibling of the index case. Despite the known family history, prenatal genetic testing has been declined by the parents. He was diagnosed on the 5th day of life and treatment with cysteamine was initiated at two months of age. He presented with Fanconi syndrome at 12 months of age but there have been no clinical manifestations of bone disease so far.

Conclusions: Early treatment of nephropathic cystinosis is pivotal for prognosis. The direct effects of both CTNS mutations and cysteamine compliance on bone disease require further elucidation.

EP-140 OUTCOMES OF ANTENATAL HYDRONEPHROSIS AND COMPARISON OF HYDRONEPHROSIS GRADING SYSTEMS

Bilge Sandal, NilÜfer GÖknar, Pinar Turhan, Emre KeleŞoĞlu, Diana ÜÇkardeŞ, Cengiz Candan

Istanbul Medeniyet University Pediatric Nephrology Department

Introduction: Antenatal hydronephrosis represents a wide spectrum of urinary anomalies, ranging from transient hydronephrosis to urinary tract obstruction or vesicoureteral reflux (VUR). Several grading systems have been used, of which the diameter of anteroposterior renal pelvis, classification of SFU (Society of Fetal Urology) and UTD (Urinary tract dilatation) are the most common ones. The aim of this study is to evaluate the outcomes of children with antenatal and early postnatal hydronephrosis, to assess the relation of hydronephrosis severity with definitive diagnosis and to compare hydronephrosis grading systems.

Material and methods: This retrospective study included all cases of hydronephrosis diagnosed antenatally from 2013 to 2019. Minimum follow up was six months.

Results: A total of 180 children (141 boys) were included in the study; 21% had ureteropelvic junction obstruction, 14% had vesicoureteral reflux, 5% had posterior urethral valve, and 2% had ureterocele. The incidence of ureteropelvic junction obstruction increased with increasing grades with all hydronephrosis grading systems. However, there were no correlation between the presence of vesicoureteral reflux and grade of hydronephrosis. Parenchymal changes on ultrasonography were also important for the diagnosis of obstructive uropathy independent of the severity of hydronephrosis. The incidence of urinary tract infection was significantly higher in children with urinary anomalies than children with transient hydronephrosis or non-obstructive dilatation ($p < 0.001$). For predicting urinary tract abnormality; classification of anteroposterior diameter had 85.3% sensitivity and 71.3% specificity, classification of SFU had 94.1% sensitivity and 52.8% specificity, classification of UTD had 83.8% sensitivity and 79.6% specificity.

Conclusions: Antenatal and postnatal US are sensitive tools for detecting hydronephrosis. Children with moderate and severe hydronephrosis have a high risk of having severe uropathies. Although there is no consensus on which staging system should be used, hydronephrosis gradings systems accurately predicts severity of kidney and urinary tract abnormalities.

EP-141 THE LEVEL OF TRANSFORMING GROWTH FACTOR BETA-1 (TGF - β1) IN URINE AS A MARKER OF KIDNEY DAMAGE IN CHILDREN WITH JUVENILE IDIOPATHIC ARTHRITIS

Svitlana Samsonenko, Tamara Borysova

Dnipro State Medical University

Introduction: To date, it has been proven that kidney damage in children with JIA is characterized by a subclinical course and often remains undiagnosed. With a duration of JIA for more than three years, the patients were found to have proteinuria, a decrease in the glomerular filtration rate and the concentration function of the kidneys. According to the results of a retrospective cohort study, it was found that 8% of children with JIA at 65 months from the onset of the disease have arterial hypertension or minimal proteinuria. The authors found that the main risk factor for the development of kidney damage in these patients was long-term exposure to non-steroidal anti-inflammatory drugs (NSAIDs) and methotrexate in active forms of the disease.

Material and methods: 80 children from JIA were examined. The age of the subjects was 10.4 ± 4.41 (10.6-15.0) years. Girls - 46 (57.5%), boys - 34 (42.5%). The debut of JIA was noted at the age of 5.8 ± 4.14 (4.9; 2.9) years. Children were distributed according to the clinical course of JIA: systemic arthritis - 9 (11.3%), polyarthritis - 47 (58.8%), oligoarthritis - 24 (30.0%). All patients at the time of examination were receiving a pain-modifying drug (methotrexate), and a history of (Plaquenil and Delagil) 31 (38.8%) patients. During the examination period, 22 (27.5%) patients received NSAIDs, 25 (31.3%) patients received immunobiological therapy. The urinary TGF-β1 is a solid phase enzyme-linked immunosorbent assay (ELISA) based on the sandwich principle.

Results: On average TGF-β 1 was 20.26 ± 16.34 (14.02; 12.5-17.98) pg/ml. The level of TGF-β 1 was significantly higher when comparing the group with polyarthritis and the combined group of patients for oligoarthritis or systemic arthritis: 23.02 ± 18.773 (15.08; 12.72-24.48) versus 16.34 ± 11.203 (13.5; 12.19-14.66) pg / ml, $p<0.01$. In patients with high JIA activity, the level of the urinary marker TGF-β1 was significantly higher than 29.15 ± 34.198 (15.21; 14.02- 37.95) in comparison with patients in remission 18.92 ± 15.041 (13.44; 12.18-15.31), $p<0.05$. The dependence of the level of the urinary marker TGF-β1 on the treated JIA was also noted. Thus, patients receiving NSAIDs have significantly higher levels of TGF-β1 25.97 ± 20.430 (17.69; 14.84-27.88) than children who did not receive NSAIDs at the time of examination 18.10 ± 14.105 (13.4; 12.18-15.01), $p<0.001$. When using immunobiological drugs, a significant difference was noted in the reference values of the renal marker TGF-β1. In the group of children receiving immunobiological therapy at the time of examination, the level of TGF-β1 was significantly lower than 14.76 ± 5.994 (12.92; 12.04-14.05) than in the group not receiving immunobiological drugs 22.76 ± 18.823 (14.4; 13.32-22.15), $p<0.01$.

Conclusions: Urinary excretion of TGF-β1 in children with JIA is associated with high activity of the underlying disease. The use of NSAIDs is accompanied by a significant increase in the excretion of TGF-β1 in the urine. In patients receiving immunobiological drugs, the content of TGF-β1 in urine was significantly lower.

EP-142 THE ROLE OF URATE IN HYPERTENSIVE CHILDREN AND ADOLESCENTS IN CARDIOVASCULAR RISK DETERMINATION

Mirjam Močnik, Sonja Golob Jančič, Martina Filipič, Nataša Marčun Varda

Department Of Paediatrics, University Medical Centre Maribor

Introduction: Urate is increasingly recognised as a cardiovascular risk factor. It has been associated with hypertension, metabolic syndrome, obesity, chronic kidney disease and diabetes. Its prognostic role is less clear. The aim of our study was to evaluate the association between serum urate and pulse wave velocity, a measure of arterial stiffness in hypertensive children and adolescents.

Material and methods: 318 children, adolescents and young adults with hypertension were included in the study. In all, anthropometric, blood pressure, pulse wave velocity and serum urate measurements were made. Participants were further divided in age subgroups. Variables were compared between boys and girls, between participants with or without obesity and with or without elevated urate.
Results: In multiple regression analysis for urate as independent variable gender, height, BMI and diastolic pressure were found to be statistically significant. The difference between urate levels were found between boys and girls ($p<0.001$), obese and non-obese ($p<0.001$); however, pulse wave velocity did not differ between hyper- and eu-uricemic group ($p=0.573$). Similar results were found in age subgroups, except for gender difference in the youngest group.
Conclusions: Associations between urate, gender, diastolic blood pressure and obesity were confirmed, however, no significant associations between pulse wave velocity and urate were detected.

EP-143 PHENOTYPE COMPARATIVE ANALYSIS IN CHILDREN WITH LOSS-OF-FUNCTION SODIUM–PHOSPHATE TRANSPORTERS NAPI-IIA AND NAPI-IIC

Svetlana Papizh[1], Larisa Prikhodina[1], Margarita Sharova[2], *Mikhail Skoblov*[2]

[1]*Veltishev Research & Clinical Institute Of Pediatrics, Pirogov Russian National Research Medical University, Moscow, Russia*, [2]*Research Centre For Medical Genetics, Moscow, Russia*

Introduction: Proximal tubular sodium–phosphate transporters NaPi-IIa (*SLC34A1)* and NaPi-IIc (*SLC34A3)* play an important role in renal tubular phosphate reabsorption and mineral metabolism. Loss-of-function mutations in the SLC34A1 and SLC34A3 genes lead to phosphate-wasting disorders as idiopathic infantile hypercalcemia type 2 (IIH2; MIM #616963) and hereditary hypophosphatemic rickets with hypercalciuria (HHRH; MIM #241530), respectively. The aim of the study was to compare the clinical features of IIH2 and HHRH in Russian children with homozygous/compound heterozygous mutations in SLC34A1 and SLC34A3 genes.
Material and methods: 14 children (9M/5F) aged 5.0 (2.0; 7.0) years with homozygous (n=4) and compound heterozygous (n=10) SLC34A1/SLC34A3 mutations were examined. The median age at the first examination in patients with IIH2 (n=6, 3M/3F) was 4.0 (2.0; 5.0) years; in children with HHRH (n=8, 6M/2F) was 6.5 (2.5; 9.0) years, $p=0.22$. Molecular genetic analysis was performed in all children by NGS.
Results: Among patients with IIH2 medullary nephrocalcinosis (NC) was revealed in 6/6 (100%), decreased TmP/GFR level in 4/6 (66.6%), bone deformation and hypercalciuria in 2/6 (33.3%), hypercalcemia, hypophosphatemia, increased serum ALP, decreased serum PTH, urolithiasis in 1/6 (16.6%) children. Patients with HHRH had hypercalciuria in 8/8 (100%), increased serum ALP, decreased serum PTH and TmP/GFR levels, NC in 7/8 (87.5%), bone deformation in 6/8 (75%), hypophosphatemia in 4/8 (50%), hypercalcemia and urolithiasis in 2/8 (25%) subjects. Serum level of $1.25(OH)D_3$ was within normal range in all children with IIH2 and HHRH. In patients with HHRH median height SDS Z score and serum PTH level were significantly lower compared with IIH2 patients: -1.1 (-1.29; -0.38) vs. -0.48 (-0.87; 0.32), $p=0.04$ and 8.1 (5.45; 12.25) vs. 33.2 (18.7; 49.3), $p=0.007$, respectively. CKD2 had 6/6 (100%) of patients with IIH2 and 7/8 (87.5%) with HHRH.
Conclusions: IIH2 was characterized by medullary NC and decreased eGFR in children aged from 2 to 5 years. The most prevalent features of HHRH in patients at the same age were hypercalciuria, increased serum ALP, decreased serum PTH and TmP/GFR level, medullary NC and declined eGFR. Growth retardation and low serum PTH were the most common signs in children with HHRH.

EP-144 ISOLATED FANCONI SYNDROME IN PATIENT WITH BCS1L-RELATED MITOCHONDRIAL DISEASE

Svetlana Papizh

Veltishev Research & Clinical Institute Of Pediatrics, Pirogov Russian National Research Medical University, Moscow, Russia

Introduction: Defects in the BCS1L gene is the most frequent cause of mitochondrial complex III deficiencies. The two major phenotypes associated with disease-causing variants in BCS1L are GRACILE (MIM #603358) (growth restriction, aminoaciduria, cholestasis with iron overload in the liver, lactic acidosis and early death) and Björnstad (MIM #262000) (brittle hair and sensorineural hearing loss) syndromes.
Material and methods: The aim of the study was to present a child with isolated Fanconi syndrome due to mutation in the BCS1L gene.
Results: The girl was born from non-consanguineous parents with birth weight 2900 g and height 49 cm. Her growth and developmental milestones were appropriate until the age of 1 year. At the age of 1.2 years the child presented with failure to thrive and windswept deformities of the knee. At first admission at the age of 4 years the girl had full-blown Fanconi syndrome including polyuria (3.5 L/m^2 per day), phosphaturia with decreased TmP/eGFR (0.4 mmol/mmol), glycosuria (4+), low molecular weight proteinuria with high urinary β-2 microglobulin level (15.4 mg/L), increased of fractional excretion of uric acid (53%), potassium (42%) and sodium (3%), aminoaciduria, metabolic acidosis (pH 7.3, HCO_3^- 17 mmol/l), short stature (height 87 cm (<3 pc), weight 11.2 kg (<3 pc)) and rickets (windswept deformities of the knee, metaphyseal fraying, widening of growth plates and severe osteomalacia). Her eGFR was 72.2 ml/min/1.73 m^2. Kidney ultrasound revealed medullary nephrocalcinosis grade 2. Neurological and glycaemic status, liver function and serum lactate level were normal. The slit-lamp examination did not show any cystine crystals in the cornea. Audiometrically her hearing was normal. MRI of the brain was normal. NGS revealed compound heterozygous mutations c.439C>T in exon 3 and c.424A>G in exon 3 of BCS1L gene. The girl was treated with L-Carnitine (52 mg/kg/d), phosphate supplements (35.4 mg/kg/d), calcitriol (40 ng/kg/d) and sodium bicarbonate (6.5 mEq/kg/d). 12-month therapy leads to improvements in her growth (by 2.3 kg in weight and 9 cm in height), normalization of serum bicarbonate (HCO_3^- 24 mmol/l) and phosphate (1.37 mmol/l) levels and decreased clinical features of rickets. We did not reveal any multisystemic features of mitochondrial dysfunction.
Conclusions: The mitochondrial dysfunction due to mutation in the BCS1L gene can presented with isolated Fanconi syndrome. BCS1L-related disease needs to be considered in non-classical phenotypes of the GRACILE and Björnstad syndromes, and this may improve early clinical recognition of the disease.

EP-145 10 YEARS OF URINARY TRACT INFECTION - MICROBIOLOGY AND ANTIMICROBIALS RESISTANCE

Marta Carvalho, Mariana Florido, Filipa Cunha, Catarina Neves

Hospital Distrital Da Figueira Da Foz

Introduction: Urinary tract infection (UTI) is one of the most common bacterial infections in paediatric age, being essential to know the local microbiology and antimicrobial resistance patterns. The aim of this

study is to characterize the UTI pathogens and their resistances to the antibiotics most frequently used in treatment (cefuroxime and amoxicillin-clavulanic acid) or prophylaxis (trimethoprim-sulfamethoxazole and nitrofurantoin).

Material and methods: Retrospective study of the paediatric UTI between 2011 and 2020. Collection of demographical data, causative pathogens, antibiotic susceptibility profile and analysis of the entire population and by age group (under 1 year old (yo), from 1 to 5 yo, 6 to 9 yo and over 9 yo).

Results: During the time considered, 1311 UTI were confirmed. The median age of the patients was 5 yo and 79% were female. The most frequently isolated pathogens were Escherichia coli (72%), Proteus spp. (13%), coagulase-negative Staphylococci (9%) and Klebsiella spp. (3%). All of them had a higher incidence in female patients, except for Klebsiella spp.

Escherichia coli was the most frequent pathogen in all age groups. Proteus spp. infection was more common in the 1 to 5 yo group (63% of the infections by Proteus spp.) and 93% of the infections by coagulase-negative Staphylococci occurred in the over 9 yo group. The highest global resistance was to ampicillin (40%), followed by trimethoprim-sulfamethoxazole (16%) and nitrofurantoin (13%). The resistance was 10% to amoxicillin-clavulanic acid and 4% to cefuroxime and, from the main pathogens, Klebsiella spp. presented the highest resistance rates (26% to both).

Conclusions: The most frequent pathogen involved in UTI is Escherichia coli as described in literature. The rate of resistance to cefuroxime remains low, supporting its use as an empirical therapy in cases of febrile UTI. New options for prophylactic antibiotics must be considered due to the high resistance rates to those currently used.

EP-146 USE OF TACROLIMUS IN STEROID RESISTANT NEPHROTIC SYNDROME: A SINGLE CENTER EXPERIENCE

Cemaliye Basaran[1], Gokcen Erfidan[1], Ozgur Ozdemir Simsek[1], Belde Kasap Demir[2], Secil Arslansoyu Camlar[3], Demet Alaygut[3], Fatma Mutlubas[3]

[1]University Of Health Sciences, Izmir Tepecik Training And Research Hospital, Department Of Pediatrics, Division Of Nephrology, Izmir, Turkey, [2]Izmir Katip Çelebi University, Faculty Of Medicine, Department Of Pediatrics, Division Of Pediatric Nephrology & Rheumatology, Izmir, Turkey, [3]University Of Health Sciences, Izmir Faculty Of Medicine, Department Of Pediatrics, Division Of Nephrology, Izmir, Turkey

Introduction: The etiology of steroid resistant nephrotic syndrome (SRNS) in children is still unclear. In some children, mutations in podocyte-related genes and in some children an immune-derived factor in the circulation has been blamed. Patients do not respond adequately to immunosuppressive therapy or have frequent attacks. It is a disease that still challenges pediatric nephrologists due to end-stage renal disease and post-transplant recurrence. Since there is no approved treatment option, many immunosuppressive treatments other than steroids are used in the treatment of these patients.

Material and methods:

We wanted to present our 5 patients with SRNS who were followed up in our center, were treated with tacrolimus, and were in clinical remission. The characteristics of our patients are given in the table.

Results:

case	gender	age-age on admission (month)	mutation	biopsy	previous treatments	time using tacrolimus (months)	serum albümin-proteinuria on admission (*spot protein/Cr or 24h collected urine protein*)	current serum albümin-proteinuria (*spot protein/Cr or 24h collected urine protein*)
M.M	female	53-33	*PLCE1 p.Val1896Ile (c.5686G>A)heterozigot *NPHS1 p.Glu117Lys (c.349G>A)heterozigot	MCD	Steroid Cyclosporine ACE inh.	14	1,3g/dL 5,32 mg/mg	4,2g/dL 0,12 mg/mg
A.T	female	184-147	*NPHS1 p.Arg1092Leu (c.3275G>T)heterozigot *COQ6 p.Val406Met (c.1216G>A)homozigot	FSGS	Steroid Cyclosporine Coenzim Q ACE inh. ARB Statin	9	2g/dL 5,5g/24hour	3,8g/dL 940 mg/24hour
P.C	female	160-106	*NPHS1 p.Met477Leu (c.1429A>T)heterozigot *PTPRO p.Pro804Ser (c.2410C>T)heterozigot	MCD	Steroid Cyclosporine ACE inh. ARB	36	2,1g/dL 1,4 g/24hour	4.7g/dL 99 mg/24hour
N.T	female	256-91	*EMP2 p.Met159Val (c.475A>G)heterozigot *COQ6 p.Val406Met (c.1216G>A)homozigot *APOL1 p.Arg255Lys (c.7646G>A)heterozigot	FSGS	Steroid CyC Cyclosporine Coenzim Q ACE inh. ARB Statin Rituximab Unilateral Nephrectomy	12	1,4g/dL 8,5g/24hour	4,7g/dL 750mg/24hour
I.K	male	74-55	*NPHS2 p.Arg229Gln (c.686G>A)heterozigot *NPHS1 p.Asn1077Ser (c.3230A>G)heterozigot *TRPC6 p.Lys803Arg (c.2408A>G)heterozigot	FSGS	Steroid Cyclosporine ACE inh.	15	2g/dL 6,8mg/mg	3,6g/dL 0,53mg/m^2/hour

CyC:Cyclophosphamide, ACE inh:Angiotensin converting enzyme inhibitors, ARB:Angiotensin receptor blockers, Cr: creatinine, MCD:Minimal change disease, FSGS: Focal segmental glomerulosclerosis
Conclusions: In SRNS, there is no response to immunosuppressive drugs such as steroids. Calcineurin inhibitors are recommended in its treatment. Complete or partial remission can be obtained in some SRNS patients, although rare, with tacrolimus treatment. For this purpose, studies involving larger patient populations are needed.

EP-147 A RARE CASE OF HEMOPERITONEUM IN AN ADOLESCENT ON AUTOMATED PERITONEAL DIALYSIS

Vasiliki Karava[1], Athanasia Chainoglou[1], Katerina Chrysaidou[1], Sofia Goutou[1], Kyriaki Charpantidou[1], Vasiliki Georgopoulou[2], Stella Stabouli[1]

[1]Pediatric Nephrology Unit, 1st Department Of Pediatrics, Hippokratio General Hospital, Aristotle University Of Thessaloniki, Greece, [2]Radiology Department, Hippokratio General Hospital, Thessaloniki, Greece

Introduction: Hemoperitoneum is a usually benign problem of peritoneal dialysis (PD), which predominantly develops shortly after peritoneal catheter insertion and rarely in chronic PD state. Excluding peritonitis, this condition is commonly attributed to vascular damage by the peritoneal catheter with favorable outcome after administration of heparin-mixed dialysate flushing and infusion of cool dialysate. Significant abdominal hematoma has been rarely reported only in adult patients.
Material and methods: A 19-years old adolescent on PD for 6 years presented acute abdominal pain localized on right iliac region and bloody peritoneal dialysate. Dialysate leucocyte cell count was negative, blood platelet count and coagulation test were normal. Due to persistent abdominal pain and acute blood hemoglobin level reduction of 3 mg/dl, abdominal CT scan was performed showing a retroperitoneal hematoma of 8.6 x 4.7 x 3.2 cm at the right paracolic gutter. After intravenous contrast injection, a small leakage of superior mesenteric vein was observed, possibly attributed to trauma by the peritoneal catheter.
Results: The patient was initially treated with intraperitoneal antibiotics until the bacterial dialysate culture came out negative. Low molecular weight heparin was added in the peritoneal dialysate solutions and the patient remained on strict decubitus position. Blood hemoglobin level was stabilized, abdominal pain resolved, and effluent became clear within 7 days. CT scan performed one week later showed reduction of abdominal hematoma. The patient was discharged 10 days later.
Conclusions: Although hemoperitoneum is a usually benign complication of pediatric chronic PD, significant abdominal hematoma should be excluded in case of persistent abdominal pain without laboratory evidence of peritonitis.

EP-148 COVID19 MANIFESTATIONS IN HEMODIALYSIS CHILDREN

Selsabil Nouir, Mariem Bouden, Sameh Mabrouk, Fadwa Majdoub, Houda Ajmi, Miniar Tfifha, Jalel Chemli, Noura Zouari, Saoussen Abroug

Pediatric Department, Sahloul Hospital, Tunisia

Introduction: Since its emergence, COVID19 has impacted several countries. Hemodialysis patients have weakened immune systems; therefore they have an increased risk for getting an infection and are particularly vulnerable to the development of severe forms
Material and methods: We retrospectively studied hemodialysis patients diagnosed with COVID -19 from 2020 to 2022 in Sahloul-Sousse dialysis center. The demographic, clinical, biological and radiological descriptions are carried out from medical data. This study aim to identify the different features of COVID 19 in hemodialysis patients.
Results: Seven patients were included(4 boys and 3 girls) with a mean age of 18 years. Two patients presented inbreeding in their history, one patient had epilepsy. All patients had end-stage renal failure 71.5% of patients were on hemodialysis, the rest were on peritoneal dialysis. All patients had high blood pressure, 48.8% had osteodedystrophy and 71.5% had a dilated left ventricle without heart failure. COVID-19 infection was confirmed by PCR in all cases. Revealing symptoms were dominated by fever(100%), cough(85.7%) and headaches(42.8%). On physcial examination, most found signs were crackling(42.8%) and tachycardia(42.8%), Chest-Xray revealed alveolar-interstitial syndrome in most patients(85.7%). CT scan was performed in 85% of cases showed COVID-19 pneumonia(50%), and average pericardial effusion(16.7%). Biological abnormalities were dominated by coagulopathy (high D dimer levels in 71.4%) and inflammatory syndrome(71.4%) 3 patients were transferred to ICU and received ventilator support. Two patients had a vascular refill. All patients received IV antibiotics. Corticosteroids were given to 3 patients. One patient received an antiviral infusion and 1 patient was given a fresh-frozen-plasma transfusion. The course was marked by bacterial superinfection(42.8%)with onset of sepsis in 14.2% of cases. 42.8% of patients experienced fluid overload to whom dialysis rate was increased. All patients have recovered without any special damage
Conclusions: COVID-19 infection is a global threat; its course is impredictable and undergoing dialysis is among the leading factors associated with a higher risk of severe presentation

EP-149 DIFFUSE LARGE B-CELL LYMPHOMA AS A MANIFESTATION OF POST-TRANSPLANT LYMPHOPROLIFERATIVE DISORDER IN A CHILD AFTER KIDNEY TRANSPLANTATION.

Aleksandra Skibiak, Ilona Zagozdzon, Aleksandra Zurowska

Department Of Paediatrics, Nephrology And Hypertension, Medical University Of Gdansk, Poland

Introduction: PTLD is a rare but life-threatening condition seen after solid organ or haematopoetic stem cell transplantation. It represents a group of B-lymphocyte proliferations mostly induced by Epstein-Barr virus in the context of immunosuppression.
Material and methods: The aim of our study was to describe diffuse B-cell lymphoma as an oncological complication of prolonged immunosupression after kidney transplantation.
Results: A twelve- years old boy with Jeune's syndrome and bilateral kidney dysplasia with end stage renal disease was hospitalised 9 months after his second kidney transplantation due to continuous pain in the right lower extremity. He had lost his first transplant due to polyoma BK virus nephropathy. 10 months later he was re-transplanted from a deceased donor. On admission right lower extremity paresis was observed. Graft function was normal but elevated D-dimers and LDH were noted. PCR test for EBV infection showed 10,500 copies/ml. On abdominal ultrasound a focal polycyclic solid mass with heterogeneous echogenicity was visualised. The tumor covered the aorta with preserved blood flow. The MRI revealed a pathological mass in the supra and mesogastrium which penetrated the spinal canal and infiltrated the bones. Histopathological examination of the pathological mass revealed a B-cell non-Hodgkins lymphoma (DLBCL EBV+). There were no abnormalities in the bone marrow biopsy. The boy was treated with Rituximab. His immunosuppression was tapered and finally ceased. He achieved remission of the disease and complete resolution of neurological symptoms with preservation of graft function after 12 month of treatment.
Conclusions: 1. Post-transplant lymphoproliferative disorder can manifest as diffuse large B-cell lymphoma in children following kidney Tx.

2. Despite extensive spinal cord lesions and infiltration of aorta and complete resolution of lymphoma can be achieved and normal kidney function can be maintained.

EP-150 ACUTE KIDNEY INJURY IN PREMATURE INFANTS WITH NECROTIZING ENTEROCOLITIS

Iuliia Kyslova, Nataliia Chornopyshchuk, Olga Yablon, Oleksandr Mazulov, Anastasiia Konoplitska

National Pirogov Memorial Medical University, Vinnytsya

Introduction: Neonatal acute kidney injury (AKI) is an important contributing factor to morbidity and mortality of critically ill neonates. Prematurity itself is an independent risk factor for AKI as a result of incomplete nephrogenesis and low nephron number. Neonatal disease caused by perinatal hypoxia is also strongly associated with AKI.
Material and methods: We analyze the frequency of acute kidney injury in premature infants with necrotizing enterocolitis (NEC) and gestational age less than 32 weeks who died. The study involved 21 of premature infants with necrotizing enterocolitis and gestational age less than 32 weeks who died. Comparison group - 25 infants who survived with similar stages of NEC. Statistical processing of the data obtained was carried out on a personal computer using STATISTICA 6.1 and IBM SPSS.
Results: The average body weight of children was 1371.2 ± 70.5 g, gestational age 29.0 ± 0.5 weeks of gestation, predominantly boys (66.7%). 12 children were diagnosed with stage III of necrotizing enterocolitis, 9 children - stage II of necrotizing enterocolitis. Acute kidney injury developed in 15 (71.4%) premature infants with NEC who died. The levels of creatinine (114.3 to 426 μmol/l) and urea (15.4 to 46.7 mmol/l) in serum of premature infants with NEC who died was significantly increased ($p<0.01$). The level of cystatin C in the blood serum of children who died was significantly increased (Me 3.45 [2.86; 4.52] ng/ml, $p <0.05$). Acute kidney damage manifested itself with anuria - in 10 (66.7%) infants, and oliguria developed in 5 infants. In addition, 13 premature infants (61.9%) were confirmed to have an intrauterine infection, 6 (28.6%) premature infants had sepsis, and 18 (85.7%) premature infants had been diagnosed with hypoxic damage of the brain. The odds ratio (OR) of lethal outcome in premature infants with gestational age less than 32 weeks with NEC and development of acute kidney injury (OR = 12,364; 95% CI: 3,415-44,768) and $\chi 2$ Pearson ($\chi 2$ = 17,578, $p <0,001$).
Conclusions: Acute kidney injury significantly associated with lethal outcome in preterm infants with gestational age less than 32 weeks and necrotizing enterocolitis.

EP-151 NUTRITIONAL MANAGEMENT OF PAEDIATRIC NEPHROTIC SYNDROME

Niki Koutsikou

Pentelis General Childrens Hospital, Greece

Introduction: Nephrotic syndrome is a relatively common glomerular disease during childhood, characterized by the clinical triad of proteinuria, hypoalbuminaemia and oedema. Nutritional management is an important part of nephrotic syndromes overall management and its aims are managing signs and symptoms, replacing the nutrients losses through the urine, improving nutritional status and reducing the risk of greater renal damage.
Material and methods: A Pubmed search was conducted up to September 2021. The literature search was performed using the terms "nephrotic syndrome", "children", "paediatric" and "nutritional management". 23 studies were eligible for the review.
Results: Although high-protein diets might cause glomerular hypertrophy and hyperfiltration and worsen proteinuria, adequate dietary energy and protein intake is of great importance, in order to reduce the risk of protein-energy malnutrition. 100-130 kcal/kg/day and 0.8g of protein/kg/day seem to be adequate. Increase of protein intake by 1 gram for every gram of protein lost through the urine is suggested. Dietary saturated fat must be limited, while emphasis should be given on mono-unsaturated and poly-unsaturated fatty acids. Supplemental administration of cholecalciferol and calcium is suggested in cases of low levels of 25-OH-D3 and/or ionized calcium and/or elevated levels of PTH. In case of anemia, supplemental administration of vitamin B12 and iron is suggested. Supplemental administration of 10 mg/day zinc may reduce nephrotic syndromes relapse rate, whereas supplemental administration of magnesium reduces thrombosis risk. Dietary sodium intake should not exceed 35mg/kg/day, while water consumption should not exceed urine output plus insensible losses. Many clinical studies have found a relationship between food hypersensitivity and nephrotic syndromes clinical manifestations severity, while various mechanisms explaining this relationship have been proposed.
Conclusions: Dietary management of nephrotic syndrome in children should be individualized, due to the great heterogeneity of its clinical manifestations. A dietitian is an important member of the multidisciplinary team that manages children with nephrotic syndrome.

EP-152 MEGACYSTIS IN TWO FEMALE FOETUSES

Sina Saffe[1], Susanne Schmidtke[2], Christiane Goedecke[3], Markus J Kemper[1]

[1]Department Of Paediatrics, Asklepios Klink Nord, Hamburg, Germany, [2]Neonatology And Paediatric Intensive Care, Asklepios Klink Nord, Hamburg, Germany, [3]Department Of Paediatric Surgery, Asklepios Klinik Nord, Hamburg, Germany

Introduction: We are presenting two case histories of female newborns, diagnosed prenatally with a megacystis and hydronephrosis. Foetal megacystis usually indicates a lower urinary tract obstruction (LUTO); in males most commonly due to posterior urethral valves. A LUTO in females is rare, with a urethral stenosis being the leading differential.
Material and methods: Patient A was diagnosed in week 12 of pregnancy and underwent further investigations including a detailed ultrasound scan and genetic analysis. Patient B was found to have a megacystis, hydronephrosis and dilated bowel loops prenatally, no additional investigations were performed. Both patients had a vesico-amniotic shunt placed, despite normal amniotic fluid volumes.
Results: Patient A was born by caesarean section at 37+4 weeks. She was noted to have a sinus urogenitalis, a ventrally displaced anus and a uterus duplex. Renal function was normal, the hydronephrosis resolved following suprapubic catheter insertion. A micturating cysto-urethrogram showed grade II-III vesicoureteral reflux. The infant was diagnosed with an anorectal/urogenital malformation syndrome; a correction operation was performed at the age of 7 months.
Patient B was born by vaginal delivery at 39+4 weeks. On day three, she developed an ileus requiring surgery. An enlarged bladder and bowel malrotation were seen intraoperatively and a stoma was formed. Two further operations were required due to repeated episodes of ileus. Enteral feeding was not tolerated because of severe hypoperistalsis of the gastrointestinal tract. In week five she developed peritonitis, leading to SIRS and ultimately death. Genetic analysis confirmed a diagnosis of megacystis microcolon intestinal hypoperistalsis syndrome (MMIHS).

Conclusions: Anorectal/urogenital malformations and MMIHS are both rare causes of a megacystis. The differential diagnosis of a megacystis in female foetuses is more complex than in males, which has implications for prenatal counselling. The vesico-amniotic shunt insertions in our patients can be considered controversial.

EP-153 CLINICAL SIGNIFICANS OF NR3C1 GENE VARIANTS IN CHILDREN WITH NEPHROTIC SYNDROME

Nur Umit[1], Asli Toylu[2], Elif Comak[3], Ipek Demir[1], Gulsah Kaya Aksoy[3], Mustafa Koyun[3], Sema Akman[3]

[1]Akdeniz University Medical Faculty, Pediatrics, Antalya, Turkey, [2]Akdeniz University Medical Faculty, Medical Genetics, Antalya, Turkey, [3]Akdeniz University Medical Faculty, Pediatric Nephrology, Antalya, Turkey

Introduction: The NR3C1 gene, which encodes the intracellular glucocorticoid receptor, has been shown to affect glucocorticoid receptor expression, function, and sensitivity to steroid therapy. The findings of studies evaluating the clinical significance of NR3C1 gene variants and their association with steroid responsiveness in children with nephrotic syndrome (NS) are different from each other. The aim of this study was to evaluate the clinical significance and the relationship between steroid responsiveness and NR3C1 gene variants' distrubitions in children with NS.
Material and methods: Demographic characteristics, clinical and laboratory data, and NR3C1 c.-13-6284C>T (rs10052957) and c.2298T>C (rs6196) variants of paitents were evaluated. The patients were classified as SSNS (steroid-sensitive NS), SDNS (steroid-dependent NS) and FRNS (frequently relapsing NS) and SRNS (steroid-resistant NS).
Results: A total of and 45 healthy children and 123 patients, 49 (39.8%) of these were girls, with a mean age at diagnosis of 12,4 (3,08-19,8) years and a median follow-up period of 6,08 (1-18,83) years were included in the study. Sixty-three patients were diagnosed with SRNS, 35 with SSNS, 17 with SDNS, and 8 with FRNS; Sixty of patients were steroid-responsive. The rs10052957 and rs6196 variants were distributed similarly in both the patient and control groups, steroid-resistant and steroid-responsive patients. Furthermore, the rs10052957 and rs6196 variant distributions were not different between clinical subgroups (SSNS, SDNS, FRNS and SRNS). The frequency of rs10052957-CT variants was significantly lower in SSNS patients than in non-SSNS patients. In the patients wtih hypertension at diagnosis, the frequency of the rs10052957-T pathogenic allele was higher than in the group without hypertension.
Conclusions: These findings suggest that the NR3C1 gene rs10052957 and rs6196 variants were not associated with the steroid responsiveness in children with NS, although the rs10052957-T allele was significantly more frequent in patients with hypertension at the time of diagnosis.

EP-154 MYELIN REGULATORY FACTOR (MYRF) RELATED UROGENITAL SYNDROME: A CASE PRESENTATION

NilÜfer GÖknar, Diana ÜÇkardeŞ, Hande Nur HasanoĞlu, Emre Kelesoglu, Kerem Özel, Cengiz Candan

Istanbul Medeniyet University

Introduction:
Myelin Regulatory Factor (MYRF), is a transcription factor which is expressed in nervous system, stomach, lung, kidney and small intestine. The phenotypes identified in these subjects included a variety of congenital heart defects, genitourinary anomalies, congenital diaphragmatic hernia, and pulmonary hypoplasia.
Material and methods:
We presented a case with MYRF mutation who presented with chronic kidney disease and ambiguous genitalia.
Results: She was born at 28-week gestation and 1160g weight. She was prenatally diagnosed bilateral cystic dysplastic kidneys and ambiguous genitalia. Her physical exam was notable for ambiguous genitalia, hemangioma on labium majus, and anal atresia with perineal fistula. Laboratory results showed elevated serum urea (46mg/dl) and creatinine levels (1.18mg/dl). Increased cortical echogenicity of left kidney and multiple cysts on right kidney was noted in urinary ultrasonography. Tc-DMSA showed that right kidney had no radiotracer accumulation. Brain MRI showed cavum septum pellucidum and Echocardiographic examination patent foramen ovale. Chromosomal analysis revealed 46,XX karyotype. Congenital pouch colon was demonstrated during rectal contrast enema graphy. Whole exom sequence reported a heterozygous variant, c.191C>T(p.Pro64Leu), in MYRF. On follow up, she experienced recurrent urinary tract infections. Voiding cystourethrography couldn't be done due to urethral anatomy. Cystoscopy showed that bladder was rudimentary. High left ureterocutaneostomy was done and multiple segmental ureteral stenosis in the left ureter was detected. On last control she had stage 4 chronic kidney disease.
Conclusions: Herein we reported a girl with severe urogenital anomalies and chronic kidney disease. MYRF related urogenital anomalies are present in 75% of individuals being affected but chronic kidney disease was not reported. Children with MYRF mutation must be checked for renal anomalies.

EP-155 EVALUATION OF PULSE WAVE ANALYSIS IN OBESE PATIENTS WITH AND WITHOUT METABOLIC SYNDROME

Cemaliye Basaran[1], Gokcen Erfidan[1], Ozgur Ozdemir Simsek[1], Secil Arslansoyu Camlar[2], Demet Alaygut[2], Fatma Mutlubas[2], Cem Karadeniz[3], Bumin Nuri Dundar[4], Belde Kasap Demir[5]

[1]University Of Health Sciences, Izmir Tepecik Training And Research Hospital, Department Of Pediatrics, Division Of Nephrology, Izmir, Turkey, [2]University Of Health Sciences, Izmir Faculty Of Medicine, Department Of Pediatrics, Division Of Nephrology, Izmir, Turkey, [3]Izmir Katip Çelebi University, Faculty Of Medicine, Department Of Pediatrics, Division Of Pediatric Cardiology, Izmir, Turkey, [4]Izmir Katip Çelebi University, Faculty Of Medicine, Department Of Pediatrics, Division Of Pediatric Endocrinology, Izmir,turkey, [5]Izmir Katip Çelebi University, Faculty Of Medicine, Department Of Pediatrics, Division Of Pediatric Nephrology & Rheumatology, Izmir, Turkey

Introduction: Our primary aim in the study was to compare the pulse wave analysis (PWA) of children with metabolic syndrome (MS) and non-MS obese children to those with non-obese children. Our second aim was to examine the reflections of additional risk factors that children with MS have in addition to obesity on PWA.
Material and methods: All the obese cases evaluated between June 2019 and June 2021 were evaluated retrospectively**.** 41 patients with MS, 36 non-MS obese patients, and 34 healthy children of similar age and gender were included. Anthropometric measurements, biochemical evaluations, 24-hour ambulatory blood pressure measurement (ABPM) and PWA measurements were evaluated. The presence of left ventricular hypertrophy (LVH) was investigated by echocardiography. Then, the differences or similarities of all groups with respect to each other were evaluated.
Results: Weight SDS, height SDS, BMI SDS values were highest in the MS and non-MS obese groups ($p<0.05$). In ABPM measurements,

systolic and MAP BP SDSs, loads; in PWA, nighttime central SBP, 24-hour, daytime, nighttime pulse pressure values and 24-hour, daytime and nighttime pulse wave velocity (PWV) rates; left ventricular mass index (LVMI), and relative wall thickness measurements in the cardiac evaluations were significantly higher in MS and non-MS obese cases when compared to the control group ($p<0.05$). 24-hour and daytime systolic and diastolic central BPs are significantly different in 3 groups with the highest in cases with MS ($p<0.05$).
Conclusions: Our study is the first study in which the PWA of children with MS and non-MS was evaluated using the oscillometric technique. Obesity causes higher office, ambulatory and central BP, PWV and LVMI, however additional risk factors leading to MS do not contribute to these parameters except 24 hour and daytime systolic and diastolic central BP values.

EP-156 HYPOCITRATURIA IN CHILDREN: ETIOLOGY AND RELATIONSHIP TO RENAL PROGNOSIS

Bahriye Atmis[1], Aliye Kidi[2], Derya Cevizli[1], Emel Saribas[1], Cagla Cagli[1], Aysun K. Bayazit[1]

[1]Cukurova University, Faculty Of Medicine, Department Of Pediatric Nephrology, Adana, Turkey, [2]Cukurova University, Faculty Of Medicine, Department Of Pediatrics, Adana, Turkey

Introduction: Hypocitraturia is one of the most common factors causing urinary system stone diseases. Hypocitraturia may be idiopathic, or it may be due to some kidney diseases, high protein-low alkaline diet and intestinal diseases. We aimed to determine the etiological factors, to examine the clinical features and renal prognosis of children with hypocitraturia in our center.
Material and methods: We retrospectively analyzed of 59 children with hypocitraturia who were diagnosed and followed-up in our center. We reviewed medical records of patients with hypocitraturia and recorded demographic features, clinical course, laboratory and radiological data of the patients.
Results: Thirty-six (61%) of patients were male. The median age at diagnosis was 4 (0-11) months. The median follow-up period was 30 (1-171) months. The median age of patients was 9 (1.5-18.5) years. Primary diagnosis of 49 (83.1%) patients were nephrolithiasis, of 4 (6.8%) patients were distal renal tubular acidosis, of two (3.4%) patients were primary hyperoxaluria type 1, of one (1.7%) patient was Lesch-Nyhan syndrome and of one patient (1.7%) was cystinuria. The consanguineous marriage rate was 49.2%. Prematurity and history of hospitalization in Neonatal Intensive Care Unit was found in nine (15.3%) of patients. There was no significant difference in mean estimated glomerular filtration rate at diagnosis and at the last visit. The mean urine citrate level was 156±52 mg/g cr at diagnosis and 535±502 mg/g cr at the last visit($p=0.004$). The median oral citrate treatment dose was 10 (6-75) meq daily. There was no significant difference between weight and height SDS of the patients at the time of diagnosis and at the last visit. The mean serum calcium level was 9.85 ±0.73mg/dl at diagnosis. The mean 25(OH)vitamin D level was 23.1±2.34 ng/ml at diagnosis. Nephrolithiasis was found in 49 (83.1%) of patients and nephrocalcinosis was found in 11 (18.6%) of patients. There was no significant difference between gender, prematurity and vitamin D levels in patients with nephrolithiasis and nephrocalcinosis.
Conclusions: Although hypocitraturia is frequently seen in patients with nephrolithiasis, it may accompany some other tubular and metabolic diseases. In spite of hypocitraturia improves with oral citrate therapy, persistence of nephrolithiasis in patients may be related to the underlying disease.

EP-157 FSGS RECURRENCE TO THE GRAFT IN HEREDITARY STEROID-RESISTANT NEPHROTIC SYNDROME: A RUSSIAN EXPERIENCE

Anastasiia Milovanova, Petr Ananin, Alexander Pushkov, Kirill Savostyanov, Tatiana Vashurina, Olga Zrobok, Olga Komarova, Alexey Tsygin

National Medical Research Center Of Childrens Health

Introduction: Kidney transplantation in hereditary nephrotic syndrome (NS) seems to be rather perspective due to the low risk of recurrent FSGS. However, despite this, there are reports of recurrence even in undeniably genetically determined cases (NPHS2 compound heterozygous, for example). There is no explanation of that phenomenon because its considered that FSGS recurrence is mainly circulating plasma factor associated.
Material and methods: We analyzed histories of 25 of 135 children with hereditary NS who underwent kidney transplantation.
Results: Among transplanted patients, there were children with causative mutations in *NPHS2* gene (7 children - 28%), NPHS1, WT1 and SMARCAL1 (3 children - 12% each), NUP93 and ARHGAP24 (2 children - 8% each) and INF2, PLCE1, MYH9 and children with co-regulation and co-expression of two genes - NPHS2 + *LMX1B* and NUP93 + *TRPC6* (1 child - 4% each). The mean age was 9 yr 10 m, SD 44.6 m. The mean onset age was 21 m, SD 30.8 m. In the onset, only four children had isolated NS or proteinuria, 17 - high blood pressure, and 13 - hematuria. Twenty children had a history of steroid therapy, all of them demonstrated resistance. In addition, 17 patients received therapy with calcineurin inhibitors, and only one child achieved partial remission after two months of the treatment. The average age of ESRD was 79.3 m (SD 50.3 m). Seven children initially started peritoneal dialysis, 6 - hemodialysis, 6 - had an experience of both of them, and 6 underwent preemptive kidney transplantation without efferent kidney replacement therapy methods. Only seven children got a deceased donor kidney transplantation. The FSGS recurrence in the graft was verified in 3 children (NS with causative mutations in NPHS2, MYH9, ARHGAP24). Kidney biopsy excluded acute rejection signs. All of them had living-related donor transplantation.
Conclusions: Disease recurrence to the graft in genetic NS occurs in a low percentage of cases (12%). Still, nowadays, mechanisms of it are unclear and require further study.

EP-158 EARLY CYSTEAMINE TREATMENT FOR OF NEPHROPATHIC CYSTINOSIS: RENAL OUTCOME OF RUSSIAN CHILDREN

Valentina Maltseva, Petr Ananin, Tatyana Vashurina, Kirill Savostyanov, Alexander Pushkov, Olga Zrobok, Andrey Fisenko, Alexey Tsygin

Federal State Autonomous Institution "national Medical Research Center For Childrens Health" Of The Ministry Of Health Of The Russian Federation

Introduction: Nephropathic cystinosis is an inherited autosomal recessive disease that leads to early-onset chronic renal failure in consequence to accumulation a lysosomal cystine in cells caused by mutations in the CTNS gene. Early initiation of cysteamine delays progression to end stage kidney disease (ESKD).
Material and methods: Retrospective analysis of renal function of 32 children with nephropathic cystinosis (17 male, 53%) diagnosed in our Center in the period 2008-2021. We analyzed the progression to ESRD in initiated cysteamine treatment groups A (1.0-2.5 years; n=13, 40.6%), B (2.6-5.0 years; n=5, 15.6%), C (after 6.0 years; n=4, 12.5%) and D

(without cure; n=10, 31.3%). Renal survival probability rates were calculated according to Kaplan-Meier, log-rank test to compare survival curves.
Results: Median age at initiating of cysteamine therapy was 1.7 years in A group (IQR: 1.1 – 2.1; range: 0.8 – 2.4); median was 3.0 years in group B (IQR: 2.8 – 3.1), median was 5.9 years in group C (IQR: 5.7 – 6.3), median was 8.7 years (mean 11.1 years; IQR: 7.8 – 12.5, range 6.0 - 26.5) in group D. Twenty one (66%) children reached ESRD at mean 10.5 years (median: 9.6; IQR: 8.2 – 13.3; range: 6.5 – 15.4), of which 16 (50%) patients had a kidney transplantation, not including 3 deaths. Log-rank analysis showed that early starting cysteamine therapy significantly delayed the ESRD onset (p = 0.032), median survival time in A group was 11.8 years (95%Cl 8.0 – 15.6) vs. 8.9 years (95%Cl 2.5 – 15.3) in B group vs 7.7 years (95%Cl 5.4 – 10.1) in C group vs. 7.8 years in group D (95%Cl 7.2 – 8.3), respectively.
Conclusions: Orally initiation of cysteamine significant the delays ESKD in children with cystinosis.

EP-159 THE VALUE OF CONTRAST ENHANCED ULTRASOUND IN YOUNG CHILDREN WITH URINARY TRACT INFECTION

Stroescu Ramona[1], Gafencu Mihai[1], David Vlad[1], ChiŞavu Flavia[2], Isac Raluca[1], Ştefl ea Ruxandra[1], DoroŞ Gabriela[1]

[1]University Of Medicine And Pharmacy "victor BabeŞ" TimiŞoara, România, [2]"louis Ţurcanu" Emergency Hospital For Children, TimiŞoara, România

Introduction: Urinary tract infections (UTIs) have been considered to be the principal cause of permanent renal parenchymal damage and scarring in children. Vesicoureteral reflux (VUR) is found in 30% to 40% of children with UTI; reflux, especially of higher grades, increases the risk of recurrent UTIs and renal scarring, with associated sequelae in later life (proteinuria, hypertension, eclampsia and end-stage renal disease). Aim: Assessing the need to perform voiding urosonography as a screening method for VUR in young children with UTIs.
Material and methods: Renal ultrasounds were performed on 179 patients with UTIs hospitalized during April 2019 –December 2020.The patients were aged between 0.4 months – 10 years, with an average of 3 years ±2.8 months. Of these, 109 patients (60.9%) had a normal renal ultrasound report.
Results: Patients with history of more than 2 infections (58 patients - 32.5%) underwent voiding urosonography. Secondary VUR due to posterior urethral valve was found in 6 patients. 32 patients had grade 3, 4 or 5 VUR, and were transferred to the surgery department. 15 (25.8%) patients detected with reflux had no pelvic or distal ureteral dilatation on renal ultrasound. In 5 patients VUR could not be detected.
Conclusions: Renal ultrasound is important in order to establish a complete diagnosis and subsequent monitoring of UTI in children. Voiding urosonography is a reliable, sensitive, safe and radiation-free method of investigation of vesicoureteral reflux in children.

EP-160 DIAGNOSTIC ROLE OF TYPE 1 PLASMINOGEN ACTIVATION INHIBITOR FOR CHRONIC KIDNEY DISEASE IN CHILDREN

Svetlana Chesnokova, Albina Vyalkova

Orenburg State Medical University

Introduction: Early diagnosis of chronic kidney disease (CKD) in children is an urgent problem in pediatrics and nephrology. In the development of nephrosclerosis, an important role is played by mechanisms due to impaired synthesis and degradation of the main elements of the extracellular matrix and a deficiency of fibrinolysis, regulated by type I plasminogen activator inhibitor (PAI-1).
Objective of the study: To evaluate the diagnostic role of PAI-1 in chronic kidney disease in children.
Material and methods: In patients with various stages of CKD (n=90), children with chronic kidney disease (CKD) without signs of CKD (n=30) and 30 apparently healthy children of the control group, the quantitative level of PAI-1 in the blood was determined by ELISA.
Results: Statistically significant differences in the level of the main inhibitor of fibrinolysis, PAI-I, were revealed in patients with initial stages of CKD (39.5±0.52ng/ml) compared with children with CKD without signs of CKD (29.3±4.32), p<0.05. With the progression of CKD (stage III–IV), a significantly higher content of PAI-I in the blood was found (up to 73.4±4.96ng/ml,p<0.001).
Significant differences in the parameters of intrarenal hemodynamics (Vs and Vd) were proved in patients with CKD without signs of CKD and with CKD at stage I (Vs 21.9±0.4mm/sec and 20.3±0.4mm/sec p<0.001; Vd 7.4±0.08mm/s and 9.33±0.28mm/s,p<0.05). With the progression of CKD, there is a decrease in Vs (with CKD II-17.3±0.55mm/sec; with CKD III-IV 12.9±0.4mm/sec,p<0.05) and Vd (with CKD II-5.6 ±0.05mm/sec, with CKD III-IV 5.2±0.05mm/sec,p<0.05).
The diagnostic significance of PAI-1 as a marker of early diagnosis of CKD was confirmed in terms of relative risk (RR=2.00), sensitivity (Se=0.33), specificity (Sp=0.93).
Conclusions: The diagnostic informativeness of PAI-1 as a biomarker of early renal damage in children with CKD was proved based on significant differences in the PAI-1 level in patients with subclinical stage of CKD and children with CKD without signs of CKD (p<0.001); indicators of relative risk, sensitivity, specificity.

EP-161 ACUTE PYELONEPHRITIS (AP) IN CHILDREN WHO PREVIOUSLY HAD A CORONAVIRUS DISEASE IN 2019 (COVID-19)

Alina Eremeeva[1], Vladimir Dlin[2], Dmitry Kudlay[1]

[1]I.m. Sechenov First Moscow State Medical University (sechenov University), [2]Veltischev Research And Clinical Institute For Pediatrics

Introduction: The SARS-CoV-2 virus is characterized by a specific three-dimensional protein structure that has a strong affinity for angiotensin converting enzyme 2 (ACE-2) receptors. X. Zhou et al. (2020) showed that in the urothelium of the bladder, ACE-2-positive cells make up 2.4%, and in the proximal convoluted tubules 4%.
The purpose of the study is to determine the clinical and laboratory features of the course of AP in children who have been infected with COVID-19.
Material and methods: The main group consists of 36 patients (4M, 36F) with AP who had COVID-19, in the asymptomatic or mild form and had elevated IgG to SARS-CoV-2. The median age was 7.5 years. The comparison group includes 47 patients (6M, 41F) with AP who have not been infected with COVID-19 (IgM and IgG to SARS-CoV-2 (IgM and IgG to SARS-CoV-2 were normal in all children). The median age was 7.0 years.
Results: In children with AP infected with COVID-19, unlike the comparison group, the duration of fever against the background of antibacterial therapy was longer (p <0.001). Six children of the main group (16.7%) were diagnosed with apostematous pyelonephritis, whereas in the comparison group only one child (2.1%). From laboratory parameters in the main group, the levels of inflammatory markers such as C reactive protein, procalcitonin and fibrinogen, as well as blood creatinine, erythrocyturia and proteinuria (p <0.05) were significantly higher than in the comparison group.

Conclusions: Thus, children with the onset of AP who have suffered from COVID-19, in contrast to the comparison group, are characterized by more longer fever, more frequent development of severe forms of pyelonephritis, higher levels and frequency of inflammatory markers.

EP-162 UPDATE OF PEDIATRIC KIDNEY TRANSPLANT PROGRAM IN UKRAINE

Svitlana Fomina[2], Oleg Godic[1], Daria Diehtiarova[1], Vasyl Nedbala[1], Olga Babicheva[1], Viktoria Apalkova[1]

[1]National Specializad Children's Hospital "okhmatdyt", [2]Si "institute Of Nephrology Of Nanm Of Ukraine"

Introduction: Kidney transplant program was launched in Ukraine in March 2021 after the Law on Transplantation was changed. This study aimed to summarize data of pediatric kidney transplantation (KT) performed in Ukraine and last year update.
Material and methods: The data from The National Registry of Pediatric Kidney Replacement Therapy and local registry of National Specialized Children's Hospital "Okhmatdyt" were analyzed.
Results: There were 106 KTs in children performed in Ukraine from 2010 to 2020, none of them from the deceased donor. The change in legislation contributed to update of the transplant program with the activation of the new leading center "Okhmatdyt" and resulted 18 pediatric KTs from March 2021. The main causes of ESKD were congenital anomalies and dysplasia (72.2%). The age of first manifestation was 7.9±1.0 years, the age of KT was ranged from 6 to 17 years. All patients received pretransplant dialysis (range: 1-52 months). Advanced HLA-typing in 10 genes and pre-transplant cross-match were examined. Living donor kidney transplants (LDKT) from related donors were performed in 8/44.4%, deceased donor kidney transplants (DDKT) were carried out in others 10/55.6% patients from 8 donors. There was one urgent LDKT in patient with dialysate diffusion into pleural cavities. One child underwent the second transplantation after graft failure). During follow-up (4.6±0.6 months) graft survival was 94.4%, (n=17): two persons had delayed renal graft function and rejection episodes and one child was removed for acute rejection and graft rupture. Survival of recipients at the time of analysis was 94.4% (n=17). The girl of 12 years old died due sepsis and COVID-19 complications in 2.5 months after LDKT. Furthermore 5 recipients (age 11.2±2.6 years) transplanted elsewhere were transferred to follow up in "Okhmatdyt" from other clinics. Nephrectomy of non-functioning kidney grafts were done in 3 of them and 2 people are known to have been transplanted under 10 kg, and graft survival period was ranged from 1 to 4 years. One child is suffering from recurrent urinary infection and anemia, and other one is being treated from rejection episode.
Conclusions: The accumulation of clinical experience with the involvement of a multidisciplinary team and best evidence-based practice is necessary to continue the successful improvement of pediatric transplantation program.

EP-163 PROFILES OF SERUM TNF-α AND TGF-β IN CHILDREN AFTER ACUTE KIDNEY INJURY

Svitlana Fomina, Olga Lavrenchuk, Victoria Driyanska, Ingretta Bagdasarova

Si "institute Of Nephrology Of Nanm Of Ukraine"

Introduction: Acute Kidney Injury (AKI) in childhood leads to Chronic Kidney Disease (CKD) with asymptomatic deterioration of kidney function. The pro- and anti-inflammatory cytokines are involved in the progression of the pathological process.
The aim of our study was to investigate the serum levels of Tumor Necrosis Factor-α (TNF-α) and Transforming Growth Factor-β (TGF-β) in children after AKI.
Material and methods: The cross-sectional study included 63 children aged from 6 months to 17 years who were observed in different period post AKI: 1-3 months (early recovery, n=21), 3-12 months (n=9), 1-3 years (n=22) and 3-5 years (n=11). All of them showed recovery kidney function to the level of CKD 1-3. We determined cytokines by ELISE (in pg/ml) and the results were analyzed according to AKI outcome: complete clinical recovery (n=14), CKD 1 (n=22), CKD 2-3 (n=27). Eight healthy children were included as controls.
Results: Differences in TNF-α after AKI were not found depends of outcomes. However, individual analysis showed association of high level (≥8.0) and increasing in the proportion of patients with CKD 2-3 ($p<0.001$) at 12 months of follow up.
Serum TGF-β increased from 25.5 (17.4;43.7) in the first year of follow-up to 38.9 (29.4;64.5) thereafter ($p=0.022$) in all patients who underwent AKI. It was found that in those who had TGF-β level ≥40.5 in the first 3 months of follow-up, the progression of CKD was determined in 86%.
Conclusions: Changes in serum levels of proinflammatory TNF-α and profibrotic TGF-β in the first months after the clinical recovery can be used to predict the progression of CKD. These data can be used for refining the mechanisms of disease and optimization the post-AKI follow-up management.

EP-164 THROMBOTIC COMPLICATIONS IN CHILDREN WITH FIRST-EPISODE STEROID-SENSITIVE NEPHROTIC SYNDROME: A SINGLE-CENTER EXPERIENCE

Agnieszka Such-gruchot[1], Hanna Szymanik-grzelak[1], Małgorzata Pańczyk-tomaszewska[1], Agata Poźniak[2], Michał Brzewski[3]

[1]Department Of Pediatrics And Nephrology, Medical University Of Warsaw, Warsaw, Poland, [2]Student Scientific Group At The Department Of Pediatrics And Nephrology, Medical University Of Warsaw, Warsaw, Poland, [3]Department Of Pediatric Radiology, Medical University Of Warsaw, Warsaw, Poland

Introduction: The aim of the study was to evaluate the clinical course and risk factors of vein thromboembolic complications (VTEC) in children with first-episode of steroid-sensitive nephrotic syndrome (SSNS).
Material and methods: We retrospectively analysed the medical records of children hospitalized due to SSNS in one pediatric nephrology unit between 2012-2019. Demographic data, clinical symptoms at the onset of NS and laboratory parameters were compared between patients with and without VTEC.
Results: Among 106 children with first episode of SSNS, in five VTEC were diagnosed during 2-60 days after onset of NS, on the basis of clinical symptoms and/or results of imaging studies. These were thromboses of femoral vein, central part of the kidney, dorsal veins of the hand, venous sinuses of the brain, and superficial vein in the popliteal fossa region. We found significant higher serum fibrinogen level ($P= 0.022$) and D-dimers ($P=0.0001$) in children with VTEC versus without VTEC, but AUC analysis showed that only D-dimers significantly differentiate thrombosis. The clinical risks factors of VTEC were vascular cannulation(100%), infections(80%), diuretics(80%). In children with VTEC, low molecular weight heparin was used. The outcome was a full recovery in all patients.
Conclusions: VTEC occurs in 4.72% of children with first episode of SSNS. The course of VTEC in children with SSNS may be asymptomatic. The clinical risk factors of VTEC in children with SSNS are vascular cannulation, infections, and diuretics. High D-dimers level is sensitive indicator of thrombosis.

EP-165 STUDY OF PROXIMAL GLOMERULAR AND TUBULAR FUNCTION IN A GROUP OF CHILDREN ADMITTED TO THE INTENSIVE CARE UNIT OF OUR HOSPITAL

Maria Isabel Luis Yanes, Eva Rodriguez Carrasco, Carlos Solis Reyes, Jose Sebastian Leon Gonzalez, Pedro Carballo Martin, Sandra Teresa Moraleda Mesa, Patricia Tejera Carreño, Victor Manuel Garcia Nieto

Hospital Universitario Ntra Señora De La Candelaria, Santa Cruz De Tenerife, Spain

Introduction: "Acute Kidney Injury" or AKI (AKI) is a common condition in intensive care units (ICUs). Patients with AKI have a very high mortality and survivors after suffering from it, are at risk of chronic kidney damage. Proximal tubular function has been poorly studied in patients admitted to pediatric ICU
Material and methods: We studied 38 patients (21V,17M) admitted successively to the Pediatric ICU of our hospital. None of them were diagnosed with AKI. Their age was 49.7±46.8 months (range: 0-159) and the days of stay, 6.9±5.3 days (range: 2-26). The most frequent etiology was respiratory failure (n=18), infectious (n=6) and neurological (n=4) causes and control after surgery (n=4). All patients except on had a PRIMS III score of less than 20. 81.6% (31/38) showed a "renal angina index" (AARI) score of less than 8.
Results: At discharge, 26/38 patients (68.4%) had elevated tubular proteinuria (both ratios (n=14), β2-microglobulin/Cr (n=7) or NAG/Cr (n=5)). The calcium/creatinine ratio was increased in 56.1% of cases (23/37) and the albumin/creatinine ratio was elevated in 25% (9/36). Children with tubular proteinuria showed significantly lower Na+ levels at admission (p=0.03) and higher EfNa at discharge (p=0.002), compared to those without tubular proteinuria. There was no relationship between the latter and the calcium/creatinine ratio, but between the NAG/creatinine ratio and the use of nephrotoxicants (p=0.03). The IAR values were not correlated with the parameters of glomerular function (at admission and discharge) but with those of the NAG/creatinine ratios (r=0.72, p<0.001) and β2-microglobulin/creatinine (r=0.75, p<0.001). In two patients, the value of the GFRe was less than 90 ml/min/1.73 m2 with the two formulas used and, in four others, they did not coincide; none received nephrotoxic drugs.
Conclusions: In patients admitted to our pediatric ICU, we observed at discharge, two apparently independent renal anomalies, namely proximal tubular proteinuria (56.1%) and high calcium/creatinine ratio (59.5%) (immobilization?). Tubular proteinuria was accompanied by saline loss. Urinary elimination of NAG was related to the use of nephrotoxicants. Hyponatremia on admission is likely to favor the appearance of tubular proteinuria. The number of children with reduced GFRe at discharge was very low.

EP-167 DIFFERENCES IN SPECTRUM OF BACTERIAL SPECIES AND THEIR SUSCEPTIBILITY TO ANTIBIOTICS IN CHILDREN HOSPITALISED WITH URINARY TRACT INFECTION IN UROLOGY, NEPHROLOGY, PEDIATRIC AND PICU DEPARTMENTS – THE ROLE OF CAKUT AND GENDER

Agnieszka Seraficka[2], Marcin Tkaczyk[1]

[1]Medical University Of Lodz, [2]Polish Mother;s Memorial Hospital Research Institute

Introduction: Urinary tract infections (UTI) constitute a common problem in paediatric clinical practice. The bacterial etiology differs between community acquired and nosocomial infections. In children who require inpatient treatment a different approach for the empiric therapy should be proposed depending on the risk of multi-drug resistant species. **The aim**-of the study was to compare the etiology and antibiotic resistance of bacteria causing UTI in children hospitalised in a specialistic tertiary reference centre with pediatric, nephrology, urology and ICU departments.
Material and methods: We analysed all positive urine specimens (>=100000 CFU/ml) reported by the hospital microbiology unit in a period of 24 months collected in tertiatry pediatric reference centre with 2 pediatric, 1 nephrology, 1 urology and 2 ICU departments. For the further analysis only those children with clinically confirmed UTI were qualified (515 specimens). The medical files were retrieved with regard to specific clinical data (f.e. gender, presence of CAKUT, clinical symptoms, length of hospitalisation). Urine culture was analysed for bacterial strain identification and standard method of antibiotic resistance according to EUCAST recommendations.
Results: E coli was detected in 44% of cases and was predominant in all of the departments, followed by Klebsiella(14%) and Enterococcus (13%) and Pseudomona aeruginosa (11%). When all children with/no CAKUT were analysed only incidence of Candida sp (0.5-4.4%, p=0.011) and Pseudomonas aeruginosa (13.7% vs 5.6%, p=0.002) was higher in the former. However, when the distribution of species between departments differed significantly showing that in non-CAKUT children ICU stay was connected with lowest E.coli and highest P.aeruginosa, Enterococcus and Klebsiella sp. incidence (p<0.025). In CAKUT patients, urology and ICU showed significantly lower incidence of E.coli (25-37 vs 45-66%; p<0.001).
When the resistance to standard antibiotic was analysed (detailed data will be available on poster), we detected that susceptibility of Klebsiella, P.aeruginosa, E.coli differed between analysed units (p<0.001) with pICU specimens mostly responsible for the difference. Boys and girls had different species distribution with lower incidence of E.coli and higher of Klebsiella sp., P.aeruginosa and Staphylococcus. (p<0.035).
Conclusions: Hospitalisation in pICU and urology department, male gender and presence of CAKUT suggested different distribution of etiology (increase in incidence of other G(-)) bacteria and higher resistance to standard set of antibiotics. Thus, we postulate that empriric therapy for UTI in pediatric tertiary reference centre should differ between urology/ICU and other unit patients and CAKUT/non-CAKUT children.

EP-168 POST-TRANSPLANT LYMPHOPROLIFERATIVE DISEASE IN PEDIATRIC KIDNEY TRANSPLANT RECIPIENTS IN TURKEY

Ismail Dursun[1], Mustafa Koyun[2], Nur Canpolat[3], Hakan Poyrazoğlu[1], Sevcan Bakkaloğlu[4], Elif Çomak[2], Rüveyda Gülmez[3], Meral Torun Bayram[8], Ilmay Bilge[5], Yılmaz Table[6], Osman Dönmez[7]

[1]Erciyes University Faculty Of Medicine, Department Of Pediatrics, Division Of Nephrology, [2]AkdenİZ University, Faculty Of Medicine, Department Of Pediatrics, [3]İstanbul University-cerrahpaşa, Cerrahpasa Faculty Of Medicine, Department Of Pediatric Nephrology, [4]Gazi University, Faculty Of Medicine Department Of Pediatric Nephrology, [5]KoÇ University, Faculty Of Medicine Department Of Pediatric Nephrology, [6]İnÖnÜ University, Faculty Of Medicine Department Of Pediatric Nephrology, [7]UludaĞ University, Faculty Of Medicine Department Of Pediatric Nephrology, [8]Dokuz Eylul University, Faculty Of Medicine Department Of Pediatric Nephrology

Introduction: Post-transplant lymphoproliferative disease (PTLD) is a life-threatening complication of kidney transplantation (KTx), but the impact of PTLD on long-term patient and graft outcomes is not well known in the pediatric population.

Material and methods: Fifteen pediatric patients (8 girls and 7 boys) with PTLD from 8 referral centers in Turkey were included in this retrospective study. Demographic data, clinic and pathologic findings, treatment, and outcome data were evaluated.
Results: The mean age and follow-up duration was 8.4±4.6 and 6.2±4.2 years, respectively. The median time from transplantation to diagnosis of PTLD was 1.1 (0.48-9.6) years. Five patients (33.3 %) were seronegative for EBV VCA IgG at the time of KTx. The most common symptom was loss of appetite. The lymph nodes and central nervous system were involved in seven and five patients, respectively. Polymorphic PTLD was the most common type of PTLD. Reduction or withdrawal of immunosuppression (73%) was the most common treatment modality for PTLD. Switching from MMF to mTOR inhibitors was performed in six and rituximab (RTX) was administered to 13 patients. Chemotherapy (*R-CHOP)* was given to six patients, radiotherapy to three patients, and surgery was performed in two patients. Seven patients (47%) achieved complete remission and five patients (33%) died from PTLD. Nine patients were alive with functioning grafts. There was no statistical significance between surviving and deceased patients in terms of EBV viral load before PTLD or at diagnosis, recipient age, or eGFR at the last visit. Mortality was not associated with any organ involvement, clinical presentation, or laboratory findings at the time of PTLD diagnosis.
Conclusions: Although survival rates in kidney transplantation have increased due to immunosuppressive treatments, PTLD still remains as a serious complication that causes death in a significant rate of patients. Therefore, large-scale pediatric studies for predicting KTx patients who are at increased risk of developing PTLD and determining those with poor prognosis are needed.

EP-169 LATE-ONSET HYPERTENSION IN A CHILD WITH SYNDROME OF AME AND COFFIN-SIRIS

Emre LeventoĞlu[1], Esra DÖĞer[2], Bahar BÜyÜkkaragÖz[1], Sinem NalÇaci[2], Ganimet Öner[2], Bedriye Nuray Alpman[1], Kibriya Fidan[1], OĞuz SÖylemezoĞlu[1], Sevcan A. BakkaloĞlu[1]

[1]Gazi University, Faculty Of Medicine, Department Of Pediatric Nephrology, *[2]Gazi University, Faculty Of Medicine, Department Of Pediatric Endocrinology*

Introduction: Prevalence of hypertension in childhood has increased over the past decade. This is most likely related to increase in primary hypertension, but secondary hypertension is still prevalent.
Material and methods: Here we present a patient who was diagnosed with AME syndrome, an endocrinological cause of secondary hypertension, and was also diagnosed with Coffin-Siris syndrome with dysmorphic features.
Results: A 6-month old girl was admitted to a hospital with the complaints of failure to thrive, intermittent vomiting and polyuria. She was born to first-degree consanguineous parents. The patient had body weight and height SDS's were -1.6 and -1.31, respectively. She was normotensive. She had hypokalemic metabolic alkalosis with normal kidney functions and increased urine output and hypercalciuria. Urinary ultrasound showed medullary nephrocalcinosis. With the diagnosis of Bartter syndrome, oral potassium and indomethacin were commenced.
She was noticed to be asymptomatically hypertensive [BP: 128/78 mmHg (>99th and 98th percentile)] for the first time in her control at the age of 7.4 years. The BP percentiles from 2.5 to 7.4 years of age are demonstrated in Table 1. Despite the use of indomethacin (2 mg/kg/d) and oral potassium support (8.4 mEq/kg/d), she had hypokalemia (3.09 mmol/L) and metabolic alkalosis (pH: 7.50, HCO_3: 31.1 mmol/L) with normal serum and urine electrolites. Plasma renin activity (0.07 ng/mL.h) and aldosterone concentrations (0.96 ng/dL) were low. She had dysmorphic facial features with a wide nose and a flat nasal bridge, a wide mouth with thick lips, and thick eyebrows. The 5th distal phalanges of the hands were shorter (Figure 1). Permission for use the pictures was obtained from parent. Ambulatory BP monitoring demonstrated severe hypertension; although fundoscopic was normal, echocardiography revealed concentric hypertrophy. WES showed c.554T>C (p.Phe185Ser) homozygous mutation in HSD11B2 gene and c.3827delC (p.Pro1276LeufsTer15) heterozygous mutation in SMARCA4 gene. The patient was diagnosed as syndrome of AME and Coffin-Siris.
Conclusions: Syndrome of AME should be considered in children with low serum renin and aldosterone levels, hypokalemia and metabolic alkalosis even without early-onset hypertension.

EP-170 SINGLE CENTER EXPERIENCE OF DIARRHEA ASSOCIATED HEMOLYTIC UREMIC SYNDROME IN PEDIATRIC INTENSIVE CARE UNIT.

Caner Alparslan[1], Mehmet Nur Talay[2], Aysel Taktak[3], Murat KanĞin[4]

[1]İzmir Tepecik Training And Research Hospital, Pediatric Nephrology, *[2]Diyarbakir Gazi YaŞargil Training And Research Hospital, Pediatric Intensive Care Unit*, *[3]İstanbul Medicalpark, Pediatric Nephrology*, *[4]İstanbul Medipol University, Pediatric Intensive Care Unit*

Introduction: Hemolytic uremic syndrome (HUS) is characterised by acute kidney injury, hemolytic anemia and thrombocytopenia. In children, it is mostly related with diarrhea (D+). In this paper, we aimed to determine clinical parameters and prognostic factors in D+HUS.
Material and methods: This retrospective study was conducted with D+ HUS 15 pediatric patients in a pediatric intensive care unit between March 2019 and August 2020. Patients demographics, initial vital signs, laboratory parameters (hemoglobine, hematocrit, white blood cell, platellets, creatinine, urea, uric acid, lactat dehydrogenase, aspartat aminotransferase, alanine aminotransferase, amilase, albumine, C3 and C4), plasma therapy, plasma exchange, RRT type and duration, and the need for blood products were evaluated. Therefore, extra-renal involvement, eculizumab treatment and last follow-up were recorded.
Results: The study group consisted of 9 males (60%) and the median age was calculated as 18 months. In 60% of the patients, RRT was implemented. Peritoneal dialysis (in 5) was the most preferded dialysis method. Five patients (33%) had extra-renal involvement. Nine patients (60%) had completely recovered, therefore proteinuria, chronic kidney disease, endstage kidney disease and neurologic sequel developed in 3 (20%), 1 (6.6%), 1 (6.6%) and 1 (6.6%), respectively. Hospitalization and oligoanuria duration had a significant impact on sequel development [Hospitalization duration: OR:1.28 (%95 CI:0.77–0.98) (p=0.04), Oligoanuria duration: OR:1.46 (%95 CI:0.94–1) (p=0.04)].
Conclusions: In this study, we showed that hospitalization and oligoanuria duration had a significant impact on sequel development. We believe that there is a need for more clinical studies to delinate more precise mechanisms of the disease and eliminate worse outcomes of D+HUS.

EP-171 ACUTE RENAL FAILURE IN 8-YEAR OLD GIRL WITH PAEDIATRIC INFLAMMATORY MULTISYSTEM SYNDROME TEMPORALLY ASSOCIATED WITH SARS-COV-2

Maria Daniel, Anna Deja, Beata Leszczynska

Medical University Of Warsaw

Introduction: Paediatric inflammatory multisystem syndrome temporally associated with SARS-CoV-2 (PIMS-TS) is a rare complication of

SARS-CoV-2 associated with single or multiorgan dysfunction. AKI occurred in 10-46% of children and young people admitted with PIMS-TS.
Material and methods: 8-year-old female patient with no significant prior medical history presented to tertiary pediatric center emergency department with fever, abdominal pain, vomiting and rash. Her mother had tested positive for COVID-19 three weeks prior, the girl had then diarrhea, PCR test was negative.
Results: Inflammatory markers were elevated, procalcitonin up to 9,51 ng/ml, CRP up 6 mg/dl and ferritin up to 379,6 ng/ml; white blood cell count was normal. She had anemia (Hb 8,5 g/dl), thrombocytopenia (95 $x10^3/\mu l$) and hypertriglyceridaemia. Liver enzymes were also elevated. Patient on admission had acute renal failure (ARF) with serum creatinine up to 4,15 mg/dl and urea up to 172 mg/dl; eGFR Schwartz formula was 13,5 ml/min/1,73 m^2, oliguria was observed. NT-pro-BNP was elevated, echocardiogram was normal. Sars-Cov-2 serology was positive.
After admission she was receiving intravenous immunoglobulins and corticosteroids. She required renal replacement therapy – two hemodialysis were performed. A kidney biopsy was performed: light microscopy revealed acute tubular interstitial nephritis.
She improved rapidly (in 2 days) on a clinical, laboratory and imaging basis. 3 months after PIMS-TS renal function was normal.
Conclusions: ARF is a severe manifestation of PIMS-TS, but prompt recognition and timely implementation of appropriate treatment gives the possibility of recovery and restoration of kidney function.

EP-172 MYCOPHENOLATE MOFETIL IN PEDIATRIC IGA NEPHROPATHY, A SINGLE-CENTER EXPERIENCE

Petr Ananin[1], Anastasiia Milovanova[1], Tatiana Vashurina[1], Ekaterina Stolyarevich[2], Olga Zrobok[1], Tatiana Voznesenskaya[1], Alexey Tsygin[1]

[1]National Medical And Research Centre For Childrens Health, [2]City Clinical Hospital №52, Moscow, Russia

Introduction: Pediatric IgA-nephropathy is predominantly presented with hematuria and gross hematuria episodes and has a mostly benign course. Mycophenolate mofetil (MMF) recently became the most promising immunosuppressive agent in severe forms of IgAN, but there is still no evidence of efficiency in pediatric IgAN.
Material and methods:
We analyzed biopsy results made in the nephrology department of our hospital for the last 4 years (from January 2018 to December 2021) to find patients who received MMF treatment for IgAN.
Results: We found 97 (17,6%) specimens with predominant IgA deposits and mesangial proliferation and/or crescents; 12 (12,4%) of them were excluded due to IgA-vasculitis. We found 25 (29,4%) patients with IgAN and proteinuria >1 g/l before a biopsy. 21 of them had a history of oral steroid treatment, 10 of them did not respond or had a relapse of proteinuria. These ten patients started MMF therapy, mean age 11,8± 3,2 yr (5 boys), eGFR was 104,6±16,5 ml/min. Morphology: 4 had mesangial proliferation, 3 – focal proliferative and sclerosing glomerulonephritis, 2 – focal proliferative glomerulonephritis with less than 50% crescents, 1 – FSGS. After 6 months, 3 had complete remission and 2 – partial. Mean proteinuria level before MMF was 2,2±1,31 g/l, at 6 months – 0,539 ± 0,32 g/l. 8 patients had 12 months follow-up, eGFR was stable. Patients with partial and complete remission had less severe morphologic changes and did not have FSGS or crescents.
Conclusions: MMF may have limited efficiency in cases of pediatric IgAN with proteinuria; multicenter controlled studies are necessary.

EP-173 LONG-TERM FOLLOW-UP OF CHİLDREN WİTH HYDRONEPHROSİS

SongÜl Yilmaz, Z. Birsin ÖzÇakar, Burcu Biral CoŞkun, NilgÜn Çakar, Fatma FatoŞ YalÇinkaya

Ankara University School Of Medicine, Department Of Pediatric Nephrology

Introduction: The aim of this study is to examine the clinical findins, radiological features and prognosis of children with hydronephrosis (HN).
Material and methods: We retrospectively analyzed the hospital records of patients with hydronephrosis that were followed in our department betweeen 2015-2020. Patients diagnosed with posterior urethral valve, vesicoureteral reflux, and ureterovesical junction obstruction were excluded from the study. Hydronephrosis was graded according to the Society for Fetal Urology classification. Patients with stage 1-2 hydronephrosis were grouped as mild hydronephrosis, and those with stage 3-4 hydronephrosis were grouped as severe hydronephrosis.
Results: A total of 219 (142 boys and 77 girls) childen were enrolled. The median age at diagnosis was 1.5 (1-204) months. Twenty eight patients (12.8%) had right sided HN, 110 (50.2%) had left sided HN and 81 (37%) had bilateral HN. Mild hydronephrosis was detected in 147 patients (67.1%) and severe HN in 72 (32.9%) patients. MAG-3 scintigraphy was performed in 66 patients, partial and complete obstruction at the ureteropelvic junction was observed in 21 and 15 patients, respectively. Median follow-up period was 60 months (6-204). Surgery was performed in 19 patients, all of whom were in the severe hydronephrosis group. At the last visit of patients who were followed with conservative therapy, hydronephrosis was resolved in 68 patients (31.1%), regressed in 45 (20.5%), remains stable in 83 (37.9%), and progressed in only 4 patients. Recovery rate was higher in the mild hydronephrosis group (37.4%-19.1% p:0.008). Recovery of hydronephrosis was observed less frequently in patients with enlarged kidney (p:0.002), parenchymal thinning ($p<0.001$), and obstruction on scintigraphy (p:0.004).
Conclusions: The prognosis of hydronephrosis is quite good in the pediatric age group. However, patients with severe hydronephrosis, enlarged kidneys, parenchymal thinning should be followed more closely.

EP-174 PERITONITIS AND URINARY TRACT INFECTIONS FOLLOWING INTRAVESICAL BOTULINUM TOXIN INJECTION IN CHILDREN ON PERITONEAL DIALYSIS: CASE REPORT

Ece Demirci Bodur[1], Nurdan Yildiz[1], Zubeyde Seker[2], Özde Nisa TÜrkkan[1], Serim Pul[1], Sadik AbidoĞlu[3], SerÇin GÜven[1], Neslihan ÇiÇek[1], İbrahim GÖkÇe[1], Ahsen KaragÖzlÜ AkgÜl[3], Harika Alpay[1]

[1]Marmara University Pediatric Nephrology, [2]Marmara University Pediatrics, [3]Marmara University Pediatric Surgery, Pediatric Urology

Introduction: Intradetrusor Botulinum toxin A injection(BTI) is a treatment modality to reduce detrusor overactivity and improve bladder capacity in children with neurogenic bladder(NB).The most common local side effects are macroscopic hematuria, difficult urination, urinary retention and urinary tract infection(UTI).In this report,two peritoneal dialysis(PD) patients who developed peritonitis,pyelonephritis and urosepsis after BTI were presented.
Material and methods: Case-1: A 14-year-old male,who was on PD due to ESKD secondary to PUV and NB,was admitted with abdominal pain,vomiting,fever,and turbidity in the urine and dialysate three days after intradetrussor BTI.His body temperature was 38.5°C and he had abdominal tenderness with cloudy urine and dialysate.Laboratory investigation revealed white blood cell(WBC):14.000/mm^3, CRP:127 mg/dL and procalsitonin:7.6 mg/dL.Urinalysis showed 3+ leukocyte esterase, 734/hpf WBC,and 69/hpf red blood cells.Dialysate WBC was

2000/mm^3 with 70% polymorphonuclear cells indicating peritonitis.After urine and dialysate cultures were obtained,the patient was initially treated with intraperitoneal(ip) ceftazidime and cefazolin for peritonitis and intravenous(iv) ceftriaxone for pyelonephritis. On the third day,both urine and peritoneal dialysate cultures revealed Klebsiella pneumoniae which were sensitive to ceftazidime,and cefazolin was discontinued.IP treatment was completed in 21 and iv 14 treatment in 10 days with complete recovery.

Case-2: A 10-month-old male with ESKD due to PUV and NB,treated with PD,admitted with fever of 40°C, weakness and loss of appetite 24 hours after intravesical BTI.Laboratory tests showed CRP:250 mg/dL, procalcitonin:100 mg/dL and pyuria.Dialysate was clear without cells.After obtaining blood,urine and dialysate cultures,iv vancomycin and meropenem were started.Blood and urine cultures showed Klebsiella pneumoniae and treatment was continued with meropenem for 21 days and he was discharged with recovery.

Results: Intravesical BTI may lead to urosepsis in addition to a known complication of uncomplicated UTI in children with NB.To our knowledge, concomitant UTI and peritonitis after intravesical BTI have not been reported.Multiple injections into the detrusor during intradetrussor BTI may cause damage to the subepithelial mucosa and initiate an inflammatory process.In addition, colonization of the bladder by microorganisms may facilitate transmural migration of bacteria leading peritonitis.

EP-175 URINE BIOMARKERS AND REFLUX NEPHROPATHY (RN) IN CHILDREN WITH VESICOURETERAL REFLUX (VUR)

Zaikova Natalia, Vladimir Dlin, Daria Penkina

Pirogov University, Moscow

Introduction: The available modern diagnostic methods often do not allow to establish the initial structural and functional changes of the kidneys. 99mTc-DMSA renal scan reveals fibrous foci that have already formed, and does not allow detecting initial and potentially reversible fibrous changes. Therefore, there is a need to develop sensitive methods for early diagnosis of kidney damage, inflammation and fibrosis.

The aim was to evaluate the relationship between urine biomarkers and renal scarring in children with VUR.

Material and methods: 117 patients aged 3 to 16 years (mean age 10.2 ± 4.5, 70.1% of girls) with VUR were examined. The control group consisted of 40 healthy children. All children underwent a complete nephrological examination. The levels of transforming growth factor (TGF-β1) and angiotensin II (Ang II), microalbumin (MA) were determined in morning urine using the ELISA method. To identify the severity of the lesion of the renal parenchyma, a static DMSA scan was performed. According to DMSA, the children were divided into 3 groups: 1 gr. – VUR without signs of sclerosis (15.4%), 2 gr. – VUR+1-2 foci (44.74%) and 3 gr VUR+ > 3-4 foci of sclerosis (40.1%).

Results: All patients with VUR had a high urinary excretion of all biomarkers when compared with the control group ($p<0.05$). The concentrations of all urine biomarkers were significantly higher in the gr.2 and gr.3 than gr.1 ($p<0.0001$). TGF-β1, Ang II and MA were correlated with renal scars. The same urine biomarkers also correlated with GFR.

Conclusions: DMSA renal scan, showed a direct correlations with the severity of VUR. We established a reliable dependence of the excretion biomarkers in the urine on the severity of RN according to DMSA scan in children with VUR. The excretion of biomarkers in the urine as non-invasive markers can be used as a criterion for the development and progression of nephropathy in VUR.

EP-176 THE CLINICAL AND LABORATORY DIFFERENCES BETWEEN MONOSYMPTOMATIC AND NONMONOSYMPTOMATIC ENURESIS

YaŞar Kandur[1], Zeynep Arslan[2], Aysegul Alpcan[2]

[1]Department Of Pediatric Nephrology, School Of Medicine, Kirikkale University, Kirikkale, Turkey, [2]Department Of Pediatrics, Faculty Of Medicine, Kirikkale University, Kirikkale, Turkey

Introduction: The present study aimed to determine the differences between monosymptomatic and non-monosymptomatic enuresis in the light of clinical and laboratory variables.

Material and methods: We retrospectively reviewed the medical records of pediatric patients with enuresis who were followed up between 2010 and 2021 at Kırıkkale University Hospital.

Results: One hundred and sixty-one patients with ;monosymptomatic enuresis (MNE) and 86 patients with non-monosymptomatic enuresis (NMNE) were enrolled in this study. The patients with MNE were significantly older than the patients with MNE (9.0 ± 2.5 vs 7.6 ± 2.4 years; $p=<0.001$). The proportion of females was significantly higher in the NMNE group (54.7% vs 40.4%; $p=0.032$). The hemoglobin level was significantly lower in the NMNE group (12.8 ± 0.8 vs 13.4 ± 1.0 g/dl; $p=0.05$). The univariable analyses using the above-identified parameters showed that a relatively low mean hemoglobin level was a risk factor for NMNE (OR=-0.603, 95% CI 0.346-0.867; $p=0.01$).

Conclusions: Making the differential diagnosis of MNE and NMNE and determining the risk factors earlier in disease course are essential tasks to be accomplished in the initial evaluation of patients with enuresis. Relatively low Hb levels may be novel risk factor for NMNE.

EP-178 TRENDS OF ACUTE KIDNEY REPLACEMENT THERAPY (KRT) MODALITY PREFERENCES IN PEDIATRIC INTENSIVE CARE UNIT (PICU): SINGLE CENTER EXPERIENCE OVER 20 YEARS

Austeja Stankute-kolosova, Karolis Azukaitis, Vilmanta Burokiene, Augustina Jankauskiene

Clinic Of Pediatrics, Institute Of Clinical Medicine, Faculty Of Medicine, Vilnius University

Introduction: The choice of KRT modality for acute kidney injury (AKI) in PICU depends on various factors. We aimed to review the preferences for KRT modality and their trends among AKI patients with different underlying diseases over last 20 years in a tertiary care PICU.

Material and methods: A retrospective chart review of initial KRT choice for all patients <18 years who treated in PICU of Vilnius University Hospital Santaros klinikos during 2000 – 2021 (decade 1: 2000-2010; decade 2: 2011-2021).

Results: 93 patients (n=49 decade 1; n=44 decade 2) with a mean age of 6.95 years (5 days – 17 years) were included. Most patients initiated KRT for sepsis (n=33; 35.5%), followed by hemolytic uremic syndrome (HUS) (23; 24.7%), AKI on chronic kidney disease (CKD) (23, 24.7%) and oncohematologic disease (14; 15.1%). Continuous veno-venous hemodiafiltration (CVVHDF) became the predominant KRT modality for sepsis and AKI on CKD patient groups, while peritoneal dialysis (PD) remained the mainstay for HUS patients. Mortality rates decreased in sepsis but remained high in oncohematology group.

	Sepsis 6.32±6.48 years		AKI on CKD 7.07±6.0 years		HUS 3.6±3.59 years		Oncohematology 9.64±5.5 years	
Decade	1 (n=19)	2 (n=14)	1 (n=14)	2 (n=9)	1 (n=10)	2 (n=13)	1 (n=6)	2 (n=8)
CVVHDF	8 (42.1%)	10 (71.4%)	3 (21.4%)	6 (66.7%)	-	5 (38.5%)	3 (50%)	4 (50%)
PD	9 (47.4%)	4 (28.6%)	9 (64.3%)	3 (33.3%)	9 (90%)	8 (61.5%)	3 (50%)	4 (50%)
HD	2 (10.5%)	-	2 (14.3%)	-	1 (10%)	-	-	-
Mortality	9 (47.4%)	4 (28.6%)	0	0	0	0	4 (66.7%)	6 (75%)

Conclusions:
Overall, the preference for CVVHDF increased during the last decade, while PD continued to be the therapy of choice in HUS group. Mortality rates of children with sepsis and KRT improved over the last decade but remained high in oncohematological patients.

EP-179 ACUTE SEVERE OLIGOHYDRAMNIOS IN A FEMALE FETUS WITH BILATERAL UPJ -OBSTRUCTION: A CASE REPORT

Lien Dossche, Evelien Snauwaert, Agnieszka Prytula, Joke Dehoorne, Ann Raes, Noortje Van Oostrum, Caroline Jamaer, Erik Van Laecke, Anne-franÇoise Spinoit, Johan Vande Walle

Ghent University Hospital

Introduction: Ureteropelvic junction obstruction (UPJO) is a common cause of antenatal detected persistent hydronephrosis. However, only in 2-5% of cases UPJO is bilateral, the majority occurs in boys (male-to-female ratio = 13:1).
Material and methods: Case: A female fetus followed for antenatal bilateral hydronephrosis suspected to be caused by bilateral UPJO, presented at 33 weeks gestation with acute severe oligohydramnios and no bladder filling. At the previous antenatal scan at 28 weeks gestation, there was no oligohydramnios and normal bladder filling noted. She was delivered at 34 weeks by Cesarean section, and bilateral UPJ obstruction was confirmed, together with anuria and renal failure. Percutaneous bilateral nephrostomies were performed with the eventual recovery of the diuresis. Both right and left kidney pyeloplasty were performed successfully with the recovery of renal failure. Moreover, on the left side beside an instrinsic UPJ-obstruction, a crossing blood vessel causing extrinsic UPJ-obstruction was identified.
Results: Antenatally detected bilateral UPJO is rare and not well described in the literature. Acute severe oligohydramnios in these cases is unexpected. Since urine production recovered completely once the nephrostomies were in place, the reason for this sudden phenomenon of oligohydramnios remains speculative and can only be related to an increasing degree of obstruction relative to poor or decreasing excretory pressure.
Conclusions: In conclusion, late-onset acute severe oligohydramnios can occur with bilateral UPJ obstruction.

EP-180 FAMILIAL HINMAN SYNDROME: RADIOLOGICAL AND URODYNAMIC FEATURES

Abir Boussetta[1], Amina Karray[2], Aicha Turki[1], Farah Krifi[1], Manel Jellouli[1], Tahar Gargah[1]

[1]Pediatric Nephrology Department, Charles Nicolle Hospital Tunis, Tunisia, *[2]Pediatric Surgery Department A, Béchir Hamza Hospital Tunis, Tunisia*

Introduction: To study the main radiological and urodynamic features in related children with Hinman syndrome
Material and methods: This was a retrospective study conducted in the pediatric nephrology department in Charles Nicolle hospital over a period of 20 years (January 01, 2000 to January 01, 2020). children belonging to the same family with vesicosphincter dysfunction (VSD) without anatomical lesions on cerebral-medullary MRI were included in our study.
Results: A total of 14 children were included in our study including 10 sisters, 2 brothers and 2 first cousins. The mean age at diagnosis was 9.9 ±4.6 years (4-16 years). Three patients had ureterohydronephrosis on ultrasound, a diverticular bladder was found in 28.5% of cases, and a vesico-ureteral reflux (VUR) was found in 21.42% of cases on retrograde urethrocystography. The debimetry curve was polyphasic in 78.57% of patients, the mean flow rate was 14.02± 5.7 ml (7.4 ml to 28.9 ml). 57.14% of the patients had a post-void residual (PVR) with a mean of 166.35±222.78 ml (70-800). Bladder sensitivity was increased in 8 patients and reduced in 6 patients. Half of the patients had a small bladder capacity and 42.85% had a large bladder capacity. The bladder compliance was normal in 5 patients (35.7%), reduced in 6 patients (42.8%) and increased in 3 patients (21.4%). Bladder contractility was normal in 6 cases, it was decreased in 7 patients and increased in one patient.
Conclusions: The association of Hinman syndrome with a hereditary factor should lead to the identification of a genetic support in order to identify populations at high risk of severe forms and to improve the outcome of the disease.

EP-181 TSC2/PKD1 CONTIGUOUS GENE DELETION SYNDROME – CASE REPORT

Đurđica Košuljandić[1], Igor Prpić[1], Ivana Kolić[1], Damir Miletić[2]

[1]Pediatric Clinic, Clinical Hospital Center Rijeka, Kresimirova 42, 51000 Rijeka, Croatia, *[2]Department Of Radiology, Clinical Hospital Centre Rijeka, Rijeka, Croatia*

Introduction: Tuberous sclerosis complex (TSC) is an autosomal dominant neurocutaneous syndrome associated with mutations or deletions of tumor suppressor genes: the TSC1 and TSC2. The syndrome presents with convulsions and psychomotor retardation. The TSC2 gene on chromosome 16p13.3 is neighbor to the PKD1 gene (tail-to-tail) responsible for autosomal dominant polycystic kidney disease-ADPKD. TSC2 / PKD1 CGDS, contiguous gene deletion syndrome, is caused by the

deletion of both genes and presented as polycystic kidney phenotype in early childhood.
Material and methods: We present a patient of 16.5 years with TSC, pervasive developmental disorder, and multiple renal cysts. The diagnosis of TSC existed since the age of six months, manifesting clinically as West syndrome. Neuroimaging showed subependymal nodules and infra and supratentorial cortical tubers, partially calcified. At 13 years old, kidney cysts up to 2 cm in size and a liver right lobe subcapsular solitary cyst were sonographically detected and confirmed by MRI. At 14.5 years of age, the parameters of global renal function were normal, without proteinuria and hypertension. Based on the polycystic kidney disease imaging finding, ADPKD type without angiomyolipomatosis, the TSC2 / PKD1 contiguous gene deletion syndrome was suspected. A genetic center analyzed the TSC1, TSC2, and PKD1 genes and confirmed the diagnosis.
Results: The result for the TSC1 gene detected no changes; for the TSC2 gene, Ex8: c.711_713delGCT, p. L238del (het); for PKD1 gene, Ex39: c.9499A> T, p.I3167F (het). Both mutations are classified as probably pathogenic (class 4), and that one, for the TSC2 gene, has never been described to date. Monitored by a multidisciplinary approach, there are no signs of renal function damage, hypertension, or extrarenal symptoms.
Conclusions: Patients with TCS require evaluation of renal damage and recognition of rare pathological conditions such as TSC2 / PKD CGDS to detect and control the possible progressive renal impairment on time.

EP-182 RENAL INJURY IN CHILDREN WITH CONGENITAL SOLITARY KIDNEY – A SINGLE CENTER EXPERIENCE

Jera Grabnar[1], Rina R Rus[2]

[1]University Medical Centre Ljubljana, Childrens Hospital, *[2]University Medical Centre Ljubljana, Childrens Hospital, Department Of Nephrology, Faculty Of Medicine, University Of Ljubljana*

Introduction: The aim of our study was to evaluate risk factors for renal injury in children with congenital solitary kidney (CSK).
Material and methods: The medical charts of 95 children with CSK (January 1980 to December 2017) were reviewed. We collected the following data: gender, causes for first US, appearance of kidney on US, kidney length measurements, other congenital anomalies of kidney and urinary tract (CAKUT), urinary tract infections (UTI), birth weight, age at last US, proteinuria, blood pressure, renoprotective treatment and glomerular filtration rate (GFR).
Results: Children with CSK were predominantly male (61%). Solitary kidney was discovered prenatally in 10/95 (10.5%) children, US due to UTI in 8/95 (8%), neonatal screening in 31/95 (32.3%), US as part of examinations for other congenital malformations, enuresis, urinary incontinence or abdominal pain in 31/95 (32.3%) children.
Abnormal US appearance of solitary kidney was found in 9/95 (%) children. 11/95 (12%) children had length of kidney below 75pc.
CAKUT was identified in 24/95 (25%) children. 26 (28%) children had UTI, 7/95 (7%) were born with low birth weight. Proteinuria was present in 14/91 (15%), arterial hypertension in 10/95 (11%) children (mean age of diagnosis 12.4 ± 2 years). 28/95 (29%) children were treated with ACE inhibitor. GFR (<70 mL/min/1.73m2) was found in 3 patients.
Twenty-seven (28.6%) patients met the criteria for renal injury (presence of hypertension and/or proteinuria and/or significantly impaired eGFR). In univariate analysis, only age at last US check was a significant risk factor for renal injury (OR 1.19, 95% CI 1.08 - 1.31, p<0.0001) and retained statistical significance in the multiple model (OR 1.20, 95% CI 1.07 - 1.33, p=0.001).
Conclusions: Our findings confirm the importance of lifelong regular follow-up of CSK patients with clinical monitoring of hypertension, proteinuria and renal function, since aging is a significant risk factor for renal injury.

EP-183 GENOTYPE-PHENOTYPE CORRELATIONS IN COL4A-RELATED DISEASES - A SMALL COHORT OF PEDATRIC PATIENTS

Anca-elena Marin[1], Florina Badea[1], Cristina Oprea[1], Diana Stoica[1], George-claudiu Costea[1], Adrian Lungu[1], Ovidiu Limoncu[1], Cristina Stoica[2]

[1]Fundeni Clinical Institute, Bucharest, Romania, *[2]Carol Davila University Of Medicine And Pharmacy, Bucharest, Romania*

Introduction: The widespread use of genetic tests has identified pathogenic variants in the genes affected in Alport syndrome(AS) in cohorts with other kidney phenotypes, including focal and segmental glomerulosclerosis(FSGS), steroid resistant nephrotic syndrome(SRNS), kidney failure of unknown cause, familial immunoglobulin A glomerulonephritis, and possibly cystic kidney disease.
Material and methods: Thirteen pediatric patients with COL4A mutations from 11 families were recruited; 8 patients have a family history of renal disease; in 8 of the families other members were tested for the same variants found in the index patients. We analyzed the genotype-phenotype correlations in our pediatric cohort and in the affected family members by indentifying the causative mutation and clinical and biological findings (hematuria, proteinuria, renal function, extrarenal features), but also histological data in these patients.
Results: Mutations were present in COL4A3-30.7%(4), COL4A4–30.7%(4) and COL4A5–38.4%(5 total,1 male). The male patient and his adult brother with X-linked AS have a severe phenotype (renal and extrarenal features) which correlates with the nonsense mutation identified in these cases, whereas the female carriers of X-linked AS demonstrate a phenotypic variability from microscopic to macroscopic hematuria, proteinuria and renal impairment. Individuals with 2 heterozygous mutations(30.7%-4 patients of the pediatric cohort and one adult family member) had a more severe phenotype than the ones with one heterozygous mutation(30.7%), presenting with nephrotic proteinuria(50% of the pediatric patients and the affected adult) and early-onset renal failure(ESKD at 17y and 27y respectively). Identified truncating variants(nonsense 7.69%, frameshifts 23.07%) and carboxi terminal missense mutations(7.69%) showed a more severe pattern of phenotypic manifestations, both renal and extrarenal. Variants affecting splicing apperead to cause a more severe renal outcome only when associated with additional heterozygous variants in COL4A genes. In our group, missense variants affecting glycine(30.7%) associated with early-onset renal failure only in one patient who had an additional heterozygous variant in COL4A3 gene. Heterozygous missense COL4A4 variant was identified in one patient with isolated FSGS and SRNS.
Conclusions: Knowledge of the causative mutation often indicates the likely clinical course. Both genetic and non-genetic factors are likely to contribute to the observed phenotypic variability and they provide predictive information about disease severity and prognosis.

EP-184 AUTOIMMUNE LYMPHOPROLIFERATIVE SYNDROME WITH GLOMERULONEPHRITIS; CASE REPORT

Demet Tekcan Karali[1], Canan Albayrak[2], HÜlya NalÇacioĞlu[1], HÜlya GÖzde Önal[1], Özlem AydoĞ[1]

[1]Ondokuz Mayis University, Faculty Of Medicine, Departments Of Pediatric Nephrology, Samsun, Turkey, *[2]Ondokuz Mayis University Faculty Of Medicine, Pediatric Hematology And Oncology, Samsun, Turkey*

Introduction: Autoimmune lymphoproliferative syndrome (ALPS) is a genetic disease caused by a defect in Fas apaptosis pathway. It is

characterized by chronic non-malignant lymphoproliferation, cytopenias and autoimmune pathologies. Autoimmune pathologies that can be seen in patients with ALPS are hepatitis, uveitis, encephalitis, and less commonly, glomerulonephritis.
Material and methods: Case: A 15-year-old girl, who was followed up with the diagnosis of ALPS, was consulted to the pediatric nephrology department because of swelling in the body. It was learned that the patient first presented with hemolytic anemia at the age of seven. Hepatosplenomegaly, thrombocytopenia, and recurrent lung infections were added to her clinic in the follow-up. She was diagnosed with ALPS, she used steroid, plasmapheresis, mycophenolate mofetil (MMF) and cyclosporine treatments. When she applied to the nephrology outpatient clinic with the complaint of diffuse swelling lasting for 15 days, she had not been taking immunosuppressive therapy for one year. In the physical examination of the patient, diffuse edema was found, and hypoalbuminemia (1.9 g/dl) and proteinuria at the nephrotic level (246 mg/m2/hour) were found in the examinations. Renal functions, complement 3 and 4 levels were normal, and there was no hematuria. Full house immune complex mediated glomerulonephritis (30% global sclerosis, 18% crescent) was found in the kidney biopsy which performed with the diagnosis of nephrotic syndrome. Repeated ANA and anti ds-DNA tests for Systemic Lupus Erythematosus (SLE) were negative, and the patient had no additional clinical findings suggestive of SLE. Pulse methylprednisolone (1 gram/day) was given for 3 days to the patient who did not respond to full-dose steroid treatment for four weeks. Intravenous cyclophosphamide was started to the patient who was still unresponsive to steroids at the sixth week, and enalapril was added to her treatment. In the follow-up, frequent albumin and furosemide infusions were required due to persistent edema and abdominal ascites, and severe pneumonia, cellulitis, and sepsis attacks occurred. Her treatment was continued with MMF. In the sixth month of MMF, she was unresponsive. A total of four doses of rituximab treatment were given with an interval of one week. MMF was continued after rituximab, but despite all treatments, remission was not achieved, severe proteinuria and hypoalbuminemia persisted. Therefore, immunosuppressive therapy was changed to cyclosporine. The patient was in partial remission (24 hour urine protein 40 mg/m2/hour, albumin 3.5 g/dl) in the third month of cyclosporine treatment, and in complete remission (24 hour urine protein 4 mg/m2/hour, albumin 4.1 g/dl) in the sixth month.
Results: Our patient is still under cyclosporine and low-dose prednisolone treatment, and is in complete remission and her kidney functions are normal.
Conclusions: Glomerulonephritis is one of the autoimmune pathologies that rarely accompanies ALPS and has been rarely reported in the literature. Although full house pattern glomerulonephritis was detected in kidney biopsy, clinical and laboratory findings of SLE were not observed in our patient. In the literature, there are ALPS cases associated with the SLE clinic, as well as SLE cases with Fas-Fas Ligand mutations. In addition, unlike most of the cases of glomerulonephritis diagnosed with ALPS in the literature, our patient was steroid resistant and responded to cyclosporine treatment. Our patient is presented because it is a rare case.

EP-185 RECURRENT ACUTE KIDNEY INJURY DUE TO THROMBOSIS OF THE VENA CAVA INFERIOR

Ozde Nisa Turkkan[1], Ibrahim Gokce[1], Birsen Barlas[2], Serim Pul[1], Ece Demirci Bodur[1], Sercin Guven[1], Neslihan Cicek[1], Nurdan Yildiz[1], Omer Dogru[2], Harika Alpay[1]

[1]Marmara University Pendik Training And Research Hospital Division Of Pediatric Nephrology, [2]Marmara University Pendik Training And Research Hospital Division Of Pediatric Hematology And Oncology

Introduction: Vena cava inferior thrombosis(VCIT) is among the rare causes of acute kidney injury(AKI). Hereby, a case with recurrent AKI secondary to VCIT is presented.
Material and methods: A 15-year-old male with no pre-existing disease was admitted to emergency outpatient clinic with abdominal pain and oedema at the lower extremities. In abdominal ultrasonography, thrombosis in the portal veins has been seen. MRI venography showed a mass approximately 8cm in diameter in the vena cava inferior (VCI) continued in the supradiaphragmatic area superiorly and cause a complete filling defect at the intrahepatic level, which is consistent with thrombosis.
Results: A combined heterozygous mutation in the methylenetetrahydrofolate reductase(MTHFR) gene was detected in thrombosis panel. CD values for Paroxysmal Nocturnal Hemoglobinuria, coagulation factors and C3, C4 were within the normal limits. ENA profile, lupus anticoagulant, hepatitis serologies, ANA and ANCA were negative. Anti-dsDNA was weakly positive. Folic acid and vitamine B12 levels were below the normal range at the time of admission. Enoxaparin sodium was started at treatment dosage. Homocysteine level obtained after replacement of folic acid and vitamine B12 was within the normal limits. Peripheral thrombolysis was performed by using tissue plasminogen activator(tPA) by interventional radiology. After the procedure, homogeneous hypoechoic nonulcerous smooth contoured thrombus echogenicity, approximately 35mm in diameter, persisted throughout the VCI suprahepatic segment. In 5month follow-up after the patients presentation, elevation of creatinine levels were observed 3times at approximately in one-month intervals. Creatinine levels were elevated up to 2.01mg/dl(GFR:61ml/m2/min) and decreased to basal levels within 24-48 hours. In renal, abdominal MR and CT angiography and venography images; no thrombus was detected in renal veins or arteries and renal venous and arterial flows were normal. Grade1 renal parenchymal echogenicity was detected during the third AKI episode in renal doppler ultrasonography and not persisted in follow-up imagings. The 24-hour urinary protein excretion was 12.86mg/m2/h during the first AKI episode, 6.9mg/m2/h at the last visit.
Conclusions: It should be noted that recurrent AKI may be observed in VCIT.

EP-186 URINARY PROTEOLYSIS FACTORS AS A PROGNOSTIC MARKER FOR THE PROGRESSION OF X-LINKED ALPORT SYNDROME IN CHILDREN

Zilya Bashirova, Ismail Osmanov

Pirogov Russian National Research Medical University, Moscow, Russian Federation

Introduction: Alport syndrome is a glomerulopathy with typical pathological changes in the GBM. However, studies have shown that tubular damage and interstitial fibrosis contribute to the progression of Alport syndrome. In this study, we wanted to assess whether urinary protheolis factors are associated with disease progression and to determine their prognostic value in children with X-linked Alport syndrome
Material and methods: 32 children (15M/17F) with X-linked Alport syndrome and normal renal function (CKD gr.1) were examined. All children received therapy with ACE inhibitors. The median age was 10.5 (IQR: 7.5;15). The median follow-up period was 5.5(IQR:3.5;6.5) years. The control group consisted of 12 age-matched healthy children with normal renal function. Laboratory tests included serum creatinine, MMP-2, MMP-3 and MMP-9 and their inhibitors TIMP-1 and 2, PAI-I in urine (were corrected for urinary creatinine excretion), determined by ELISA. A decrease in eGFR of ≥30% over 2 years from baseline was chosen to represent the rapidly progressive course. 28.1% of children had a rapidly progressive course of the disease (7M/2F), 71.9%-a slowly progressive course (8M/15F). The association of baseline urinary levels of MMPs and their inhibitors with eGFR and progression of patients with Alport syndrome to a later stage of CKD during the follow-up period was used to assess the prognostic value of the marker

Results: The chances of detecting a rapidly progressive course with a decrease in the level of MMP-9 (100%vs.47.8%(p=0.012), OR=1.82(95%CI:1.23-2.71)) and an increased level of TIMP-1 (88.9%vs.30.4%(p=0.005), OR=18.2(95%CI:1.96-175)) in the urine is statistically significantly more common in children with a rapidly progressive course of the disease than with a slowly progressive one. Couldnt find relationship between MMP-2, TIMP-2, PAI-I and disease progression
Conclusions: Our study suggests that urinary MMP-9 and TIMP-1 are a promising biomarker for accelerated decline in kidney function in children with X-linked Alport syndrome.This may help identify patients at high risk of progression for targeted clinical management and improve patient stratification in future studies

EP-187 OUTCOMES OF ACUTE KIDNEY INJURY IN CHILDREN DEPENDS ON AGE DISEASE ONSET

Svitlana Fomina, Olga Lavrenchuk, Galina Suslova, Ingretta Bagdasarova

Si "institute Of Nephrology Of Nanm Of Ukraine"

Introduction: The aim of study was to analyze outcomes of Acute Kidney Injury (AKI) in children from different age groups.
Material and methods: The disease course in 242 children was studied for the period from 2002 to 2021 years. Age stratification identified groups depends period of disease onset: before 12 months (n=33), 1-3 years (n=112), 3-10 years (n=56), 10-18 years (n=41). AKI outcomes were determined according to Chronic Kidney Disease (CKD) classification and assessed with survival techniques during follow up to 120 months.
Results: It was documented complete recovery of Glomerular Filtration Rate (eGFR>90) in 38.2% causes, up to the level of CKD 2 - in 33.6%, CKD 3- in 11.2% at the 6th month of follow up. simultaneously14.6% of patients continued the Kidney Replacement Therapy (dialysis). Cumulative probability of children with eGFR>15 was 84.6±2.32% on the 12th month with minimal decreasing further (to 76.8±3.41% on the 120th month).
The probability of CKD5 was differed in the age groups with the worst indicators after 10 years old ($\chi2$=23.4, p=0.00003; at the age before 12 months: 97.0% throughout the observation period; at 1-3 years: decrease the cumulative probability of those with GFR>15 from 91.1% to 81.7%; at 3-10 years: from 78.2% to 75.3%; over 10 years: from 63.4% to 42.8%).
The main quantity of negative outcome was formed in the first 12 months after AKI ($\chi2$=22.5, p = 0.00005) with the deterioration after 36 months in patients aged 1-3 years old and after 60 months in adolescents. However, 19.2% of CKD 5 cases were documented at 120 months.
Conclusions: Our data confirmed the significance of the age variable in pediatric AKI outcomes and with accent on 12 months and prolonged follow-up.

EP-188 KIDNEY DISEASE IN KEARNS-SAYRE SYNDROME. BEYOND TUBULAR INVOLVEMENT

Pedro Arango Sancho, Yolanda Calzada Baños, Marta Jiménez Moreno, Ana Cristina Aguilar Rodríguez, Elena Codina Sampera, Raquel Jiménez García, Marina Pons Espinal, Álvaro Madrid Aris

Hospital Sant Joan De Déu

Introduction: Mitochondrial diseases are characterized by presenting a wide range of clinical manifestations,mainly affecting those organs that are most dependent on aerobic metabolism.Kearns-Sayre syndrome (SKS) is a multisystem entity characterized by the triad of bilateral ocular ptosis,retinitis pigmentosa,and cardiac conduction abnormalities.Kidney involvement in this disease is rare and usually presents in form of tubular alteration,being more frequent its appearance as Fanconi Syndrome of variable severity,although other forms of manifestation have been rarely described
Material and methods: 14-year-old male patient,the result of a multiple gestation (first triplet,two healthy sisters) with a history of prematurity (35.5 weeks) and low birth weight (1500 g).Non-consanguineous parents.Father and paternal grandfather with a history of early acute myocardial infarction (46 years).He started a history of pancreatic insufficiency and short stature since he was 6 years old (treated with growth hormone),together with difficulty walking and retinopathy starting at 12 years old,at which time he was diagnosed with SKS.Referred at this age to our consultation for screening for renal pathology associated with the disease,presenting only dysplastic and hyperechoic kidneys together with adequate renal function (creatinine 0.9 mg/dl and GFR Schwartz (0.413):92 ml/min/1,73 m2) and high blood pressure,for which treatment with enalapril was started.Two years later,he presented rapidly progressive renal dysfunction (GFR Schwartz (0.413): 35-40 ml/min/1.73 m2) together with mixed proteinuria that reached the nephrotic range (urinary index Pr/Cr 7.5 mg/mg)
Results: At this time,it was decided to start treatment with prednisone and cyclosporine with a partial response (Pr/Cr index of 7.5 to 2 mg/mg) together with persistence of rapid progression towards end-stage chronic kidney disease.The renal biopsy performed revealed a pattern of focal segmental glomerulosclerosis with a significant chronicity component.Only 8 months after the onset of renal involvement,a cadaveric donor kidney transplant was performed with good adaptation without complications and with a normal glomerular filtration rate at present
Conclusions: Mitochondrial diseases can manifest with various renal presentations.Despite tubular involvement being the most frequent,other manifestations,such as focal segmental glomerulosclerosis can overshadow the renal prognosis of patients and should be considered in the differential diagnosis

EP-189 EVALUATION OF THE CLINICAL FINDINGS OF PEDIATRIC PATIENTS WITH VESICOURETERAL REFLUX TO ASSESS DISEASE SEVERITY

YaŞar Kandur[1], Aysegul Alpcan[2], Serkan Tursun[2]

[1]Department Of Pediatric Nephrology, School Of Medicine, Kirikkale University, Kirikkale, Turkey, [2]Department Of Pediatrics, Faculty Of Medicine, Kirikkale University, Kirikkale, Turkey

Introduction: The purpose of this study is to investigate how we can benefit from clinical and laboratory methods for being more selective during the decision process to perform a cystogram and to assess vesicoureteral reflux (VUR) severity.
Material and methods: We retrospectively reviewed the VCUG, ultrasound (US), DMSA findings, and medical records of pediatric patients with VUR.
Results: Sixty-three pediatric patients with VUR were enrolled in this study. The median age of the patients (F/M=37/26) at the time of diagnosis was 62.0±6.5 months (range 1-195 months). Seventeen (26.9%) patients had high-grade vesicoureteral reflux, and 46 (73.1%) patients had low-moderate-grade VUR. The differential renal function(DRF) of the more severely affected kidney was significantly lower in the highgrade VUR group than the low-moderate grade VUR group (18±5 vs 34±2; p=0.038). The mean potassium level was significantly higher and the Na/K ratio was significantly lower in the high-grade VUR group (4.7±0.5 vs 4.3±0.4 meq/L; p=0.022; 29±3 vs 32±3; p=0.029, respectively) in the high - grade VUR group. The proportion of patients with severe

AP diameter dilation was significantly higher in the low-grade VUR group than the high-grade VUR group (4 (23.5%) vs 35 (76.5%); p=0.005). There was a negative correlation between VUR grade and DRF (p=0.038, r=- 506).
Conclusions: We conclude that sintigraphic DRF and low N/K ratio allows us to predict the VUR grade. Therefore, in suspicious cases, radionuclide scans should not be delayed. The effect of VUR on the renin-angiotensin aldosterone system can be demonstrated by prospective controlled studies.

EP-190 A RARE HISTOPATHOLOGY: GRANULOMATOUS BK VIRUS NEPHROPATHY

İbrahim GÖkÇe[1], Ece Demirci Bodur[1], Burcu Tufan TaŞ[3], Deniz Filinte[2], Serim Pul[1], Özde Nisa TÜrkkan[1], SerÇin GÜven[1], Neslihan ÇiÇek[1], Nurdan Yildiz[1], NurŞah Eker[3], Harika Alpay[1]

[1]Marmara University Pediatric Nephrology, [2]Marmara University Pathology, [3]Marmara University Pediatric Hematology And Oncology

Introduction: BK Virus(BKV)usually causes infections in kidney transplant and bone marrow transplant(BMT) patients receiving immunosuppressive therapy and can cause asymptomatic hematuria,hemorrhagic cystitis and interstitial nephritis.Granulomas are rarely seen in the histopathological examination of BKV nephropathy (BKVN).In this case report,a patient who presented with acute kidney injury(AKI) developing 2 months after BMT and diagnosed with granulomatous BKVN will be presented.
Material and methods: Case: A 14-year-old male patient who had BMT from his brother 2 months ago due to acute lymphoblastic leukemia,presented with vomiting and diarrhea. He showed signs of mild dehydration and laboratory findings showed urine density of 1031,protein of 2(+)on dipstick,creatinine of 0.57 mg/dL,BUN of 11 mg/dL,urea of 24 mg/dL and normal electrolytes.Vomiting and diarrhea was stopped but the gradual increase in creatinine persisted.Ultrasonography revealed bilateral increased echogenity in kidneys.Direct microscopic examination of urine showed morphic erytrocytes.24-hour urine test showed nephritic range,mixed type proteinuria of 575 mg/day(protein:17 mg/m^2/h,microalbumin:163 mg/day).Cyclosporine treatment,which was planned to be discontinued one month later,was discontinued early due to the gradual increase in creatinine.C3,C4 were normal.EBV,CMV PCR tests and ANA was negative.Urinary BKV load was 52.891 copy/ml,blood BKV load was 91 copy/ml.Renal biopsy was performed and while waiting for the pathology results,oral ciprofloxacin was started.Biopsy showed inflammatory cell infiltration including eosinophils and granulomas in the tubulointerstitial area and diffuse tubulary injury which were consistent with BKVN.SV40 was positive in immune-histochemical staining.After 15 days,creatinine was 1.26 mg/dL,blood BKV was 27 copy/ml,urine BKV load was 198 copy/ml and urinary protein excretion decreased to 4 mg/m^2/h.
Conclusions: In the histology of BKVN; cytopathic changes, tubulointerstitial inflammation and fibrotic changes are expected in BKV-infected areas.Classical granuloma formation is rare and reported in 1 out of 46 biopsies of BKVN.We wanted to draw attention to the fact that granulomatous nephropathy detected in the histopathological examinations of immunosuppressed patients who had kidney biopsy for AKI and/or proteinuria may be associated with BKVN.

EP-191 EPITOPIC EVALUATION IN PEDIATRIC KIDNEY TRANSPLANTATION - EXPERIENCE OF A ROMANIAN SINGLE CENTER

Rachisan Andreea Liana, Santionean Diana, Aldea Paul Luchian, Bulata Bogdan, Delean Dan, Aldea Cornel, Elec Florin Ioan

University Of Medicine & Pharmacy Iuliu Hatieganu Cluj-napoca

Introduction: HLA epitope-based matching offers the potential to improve immunological risk prediction and management in children receiving renal grafts; however, studies demonstrating the association between systems for defining epitope mismatches and clinical end-points are lacking in this population.
Material and methods: We systematically reviewed the electronic patient record for all consecutive renal transplantations from January 1995 to December 2020. We included 55 pediatric recipients (female/male ratio = 30/25, mean age at transplantation 12.07 ± 3.44 years) of a kidney transplant with HLA typing (HLA-A, B, and DR). All the patients underwent an HLA matchmaker algorithm score in order to determine the epitopic load.
Results: The cohort included first kidney transplantations mainly from deceased donors (33 patients). Transplants were allocated to recipients with a mean cumulative ABDR mismatches of 4. The HLA matchmaker score ranged between 0 and 25, with a mean of 10.94±5.98. There was a significant correlation between the number of HLA antigenic mismatches and HLA matchmaker score with a p<0.001; r^2=0.36. The mean HLA matchmaker epitopes for patients with ABDR mismatches <4 was 7.10 ±4.48 and for patients ≥4 was 12.97 ±5.98, with a p=0.001.
Conclusions: This is a single center analysis of 55 kidney pediatric recipients on the impact of HLA epitopic load approaches. These findings are inconclusive but suggest that HLA epitopic load may provide one tool for assessing long-term risk in this population while highlighting the need for further clinical studies.

EP-192 EFFECTS OF SIROLIMUS ON RENAL FUNCTIONS AND GROWTH IN PEDIATRIC RENAL TRANSPLANT RECIPIENTS

Nesrin Tas, Bora Gulhan, Gulsah Ozdemir, Tuba Tastemel Ozturk, Demet Baltu, Eda Didem Kurt Sukur, Diclehan Orhan, Fatih Ozaltin, Ali Duzova, Rezan Topaloglu

Hacettepe University Childrens Hospital

Introduction: Calcineurin inhibitors (CNI) are the main treatment strategy of pediatric renal transplantation, however, there are known side effects in the short and long term. In this study, we investigated the efficacy of sirolimus (SRL) and its effects on growth in pediatric renal transplant recipients.
Material and methods:
We performed a retrospective analysis of 26 renal transplant recipients who underwent sirolimus/everolimus conversion (mTORi) during their follow-up period.
Results: A total of 26 patients (18 boys, 8 girls) who converted to mTORi treatment were included in the study. The most common etiology of ESRD was congenital anomalies of the kidney and urinary tract. The mean age of transplantation was 10.6 ± 3.9 years. The duration from transplantation to mTORi conversion was 17.2 ± 19.6 months. The most common indication for SRL conversion was biopsy-proven IF/TA (n=10, 38.4%). Other reasons for mTORi conversion were; BKVAN (n=7), cyclosporine toxicity (n=5), progressive decline of renal function (n=3), PTLPD (n=1). The mean duration from SRL conversion to the last visit was 26.5 ± 27.5 months. Height, weight, and BMI z scores were not different at the time of conversion and 2nd year after conversion. Graft survival at 24. months was 100.0 % in IF/TA group who were converted to SRL but 60.0% in non-mTORi group with IF/TA (p=0.02). Side effects in mTORi group were as follows; only hyperlipidemia (n=6), only proteinuria (n=6), hyperlipidemia and proteinuria (n=9).
Conclusions: Our study encourages using SRL in pediatric transplant patients. In 24 months follow-up we did not find an adverse effect on growth.

EP-193 LITHIUM POISONING: AN UNCOMMON INDICATION FOR HEMODIALYSIS IN PEDIATRICS

Elena Codina Sampera, Ana Cristina Aguilar Rodríguez, Pedro Arango Sancho, Marta Jiménez Moreno, Raquel Jiménez García, Yolanda Calzada Baños, Álvaro Madrid Aris

Hospital Sant Joan De Déu

Introduction: The most common causes of acute hemodialysis (HD) in pediatrics are:hydrosaline overload or correction of alterations in the internal environment.Certain intoxications would also be an indication,although in pediatrics they are a rare cause.We present a case of acute lithium poisoning in a pediatric patient
Material and methods: A 15-year-old adolescent who reported having ingested 60-90 tablets of 400 mg of lithium carbonate (507 mg/kg) and 7-8 tablets of 20 mg fluoxetine two hours before his arrival at the emergency room (autolytic)
Results: After an initial evaluation,he was transferred to the Pediatric Intensive Care Unit (PICU) for placement of a central venous catheter and initiation of purifying treatment with HD,even without knowing the serum lithium levels.Once the catheter was placed,we received the initial blood lithium level of 2.37 mEq/L (moderate toxicity).The serum lithium level just at the time of starting HD was 3.65 mEq/L (severe toxicity).A 6-hour session was held,with flows scheduled for the technique higher than usual.The patient vomited several times during the technique (about 30 lithium tablets were recovered),similarly presenting mild neurological symptoms (bradypsychia in the first hours together with mild dysmetria and tremor of the mandible and distal extremities).During the first 24 hours,he required a new HD session,with subsequent blood lithium levels of 0.9 mEq/L (outside the toxic level).The patient presented a good subsequent evolution with resolution of the neurological symptoms and without the need for new sessions of renal replacement therapy
Conclusions: The chemical and pharmacological characteristics of lithium (low molecular weight,low plasma protein binding,low volume of distribution and total excretion in the urine) make HD the best extrarenal treatment in case of need for lithium clearance.It would be indicated in cases of serum lithium level >4mEq/L together with impaired renal function and/or decreased level of consciousness,seizures or life-threatening arrhythmias (regardless of the plasma level).In our case,the programming of the HD was a challenge due to its marked differences with respect to usual regimens:long duration with high flows.Despite the abundant bibliography described in adults,in pediatrics it is a very infrequent cause of the need for acute HD

EP-194 FAMILIAL RENAL GLUCOSURIA DUE TO MUTATIONS IN THE SLC5A2 GENE IN A MALE ADOLESCENT;

Martine Docx[1], Nathalie Segers[1], Johan Vande Walle[2]

[1]*Queen Paola Childrens Hospital Antwerp Belgium*, [2]*University Hospital Ghent Belgium*

Introduction: Familial renal glucosuria (FRG) is an inherited disorder mostly caused by mutations in the SLC5A2 gene, mapped to 16p11.2 and coding for the sodiumglucose co-transporter 2 (SGLT2) in the proximal tubule.
Material and methods: A 17-year old Macedonian male presented since the age of 1.5 years with an isolated renal glucosuria in the absence of hyperglycemia. Renal function was normal. No bedwetting, polyuria-polydipsia or polyphagia and no intellectual disability. Renal glucosuria varies from 8 g/L (1.5 years) until now 41.2 g/L (17 years). Genetic analysis was performed. He was heterozygous for two variants of the SLC5A2 gene:(1)c571A>C p(Thr191Pro) and (2) c1405G>A p (Ala469Thr). The familial mutations have been looked for by Sanger Sequencing.The mother has a heterozygosity for mutation 1 and absent for mutation 2. The father has a heterozygosity for mutation 2 and absent for mutation 1. Both parents have a normal renal function and no glucosuria.
Results: The variant c1405G>A p(Ala469Thr) of the SLC5A2 gene was reported by Calado et al. (2008 NDT 23:3874-3879) in the index case (Family 15). The index case is a 2 year old Macedonian girl with also a compound heterozygosity. She had a glucosuria of 14.2 g/L.
Conclusions: Our patient have similar characteristics with the index case in the literature. These are: ethnicity, compound heterozygosity, mild-moderate glucosuria and the young age of onset. FRG is mostly a benign disorder with no longterm effects on renal function. Exceptional failure-to thrive, postprandial hypoglycemia as well as chronic urinary and genital infections are described.

EP-195 METABOLIC ALKALOSIS IN INFANTS TREATED WITH PERITONEAL DIALYSIS

Shimrit Tzvi-behr, Efrat Ben-shalom, Yaacov Frishberg, Rachel Becker-cohen, Choni Rinat, Jenny Weinbrand- Goichberg, Sapir Choshen

Shaare Zedek Medical Center, Jerusalem, Israel

Introduction: Acid base balance is maintained by the kidney, via excretion of ammonium and titratable acids and bicarbonate reabsorption. Metabolic acidosis develops as kidney function deteriorates. Peritoneal dialysis is considered efficient in controlling metabolic acidosis. Metabolic alkalosis is uncommon in dialysis treated patients. The aim of this retrospective study was to assess the rate of metabolic alkalosis in pediatric patients treated with peritoneal dialysis.
Material and methods: Medical records of children treated with peritoneal dialysis in Shaare Zedek Medical Center from January 2000 to June 2021 were reviewed and compared with young adults currently treated with peritoneal dialysis. Demographic, clinical and peritoneal dialysis characteristics were extracted from the medical records.
Results: Thirty chronic peritoneal dialysis patients were included in our study. Seven under 2 years, 13 between 2 and 18 years and 10 adults. 90.3% of the measurements in infants showed metabolic alkalosis compared to 32.3% in the 2-18 years group and none in the adult group. Higher size-adjusted daily exchange volume, lack of urine output and high lactate containing dialysate were associated with metabolic alkalosis. Alkalosis was not explained by vomiting, diuretic therapy or carbonate containing medications. High transport membrane, low dietary protein and malnutrition, all previously reported explanations for metabolic alkalosis, were not found in our study.
Conclusions: Metabolic alkalosis is common in infants treated with peritoneal dialysis as opposed to older children and adults. High lactate containing dialysate and higher size-adjusted daily dialysate exchange volume are possible predictors. This finding, coupled with the higher mass transfer area coefficients previously found in infants, may reflect a higher solute transport capacity including bicarbonate absorption in this age group. Acid-base status should be closely followed in infants and using a dialysis solution with lower bicarbonate or lactate level should be considered.

EP-196 CLINICAL PHENOTYPE OF NEPHRONOPHTHISIS11 CAUSED BY MUTATION IN THE TMEM67 GENE IN INFANT

Nadezhda Savenkova, Elvira Andreeva

Saint-petersburg State Pediatric Medical University, Russian Federation

Introduction: M.Rasmussen et al (2018) identified Nephronophthisis3 and autosomal recessive Polycystic Kidney Disease (PKD) are most often phenocopies of TMEM67 gene mutation.
Material and methods: The proband with the clinical diagnosis of PKD was underwent examination including assessment of renal function, ultrasound (US) with the definition total kidney volume (TKV), blood pressure control and genetic testing.
Results: The child was born from a 2nd normal pregnancy in family with a negative history of PKD. Parents are not related. After birth identified arterial hypertension, increased blood renin and enlarged (TKV100sm3) hyperechoic kidneys with diffuse small cysts (max size 0.56-0.43sm) in medulla by US. Kidney function is preserved. Clinical physical examination at the age of 3 months the child is developed according to age, confirms metabolic acidosis, anemia, hepatomegaly. By whole exome sequencing analysis identified a homozygous variant of mutation in the TMEM67 gene (*c.1843T>C:p.Cys615Arg)* in infant. The involvement of the mutant TMEM67 gene is known to be associated with a broad range of clinical presentations in JBTS6, Nephronophthisis11, BBS, COACH, MKS3 syndromes. At the age of 3 months, no data were obtained for liver fibrosis, eyes pathology, heart defects, skeletal, central nervous system anomalies and other extrarenal manifestations described mutation in the TMEM67gene. The child is interpreted as Nephronophthisis11 caused by mutation of the TMEM67 gene, requires continued treatment.
Conclusions: We describe the features of phenotype and early diagnostic of the Nephronophthisis11 caused by mutation TMEM67 gene in an infant with enlarged hyperechoic kidneys and arterial hypertension from birth.

EP-197 EVALUATION OF RISK FACTORS FOR ACUTE PYELONEPHRITIS AND PERMANENT RENAL DAMAGE (RENAL SCARRING) IN CHILDREN UNDER 2 YEARS OF AGE WITH A FIRST FEBRILE URINARY TRACT INFECTION

Aikaterini Gkrepi[1], Vasileios Giapros[2], Spyros Tsiouris[3], Anastasios Serbis[1], Vasileios Xydis[4], Ekaterini Siomou[1]

[1]*Department Of Pediatrics, University Hospital Of Ioannina, Greece*, [2]*Neonatal Intensive Care Unit, University Hospital Of Ioannina, Greece*, [3]*Nuclear Medicine Department, University Hospital Of Ioannina, Greece*, [4]*Radiology Department, University Hospital Of Ioannina, Greece*

Introduction: To study possible predictive factors for acute pyelonephritis (APN) and renal scarring in children ≤2 years of age hospitalized with a first documented febrile urinary tract infection (UTI).
Material and methods:
Forty-five patients ≤2 years of age and 45 age-matched controls were prospectively included in the study. On admission, the neutrophil count, serum levels of creatinine, CRP, ESR, cystatin C and 25OHD were evaluated. The patients were divided in two subgroups based on acute DMSA (technetium 99m-dimercaptosuccinic acid) scan, those with a normal DMSA and those with a DMSA indicating APN. The subgroup of patients with abnormal acute DMSA had a follow-up DMSA scan after 6 months to evaluate for renal scarring.
Results: APN was found in 24/45 (53.3%) children. The children with APN had significantly higher CRP ($p<0.01$) and ESR ($p<0.05$) and lower 25OHD ($p<0.01$) levels compared with those with normal DMSA. Cystatin C levels did not differ significantly between the patient and control groups. The correlation between 25OHD level and APN was independent of age, fever duration and CRP/ESR level, implying that low vitamin D level is an independent predictor of acute renal damage in children with febrile UTI with a relatively high specificity (81.2%) and sensitivity of 54%. Renal scaring was found in 7/19 (36.8%) of children with APN. CRP levels >100 mg/L during APN were found more frequently in children with renal scarring, comparing to those with no renal scarring ($p<0.01$). VUR grade ≥3 was found only in children with renal scarring.
Conclusions: Vitamin D levels were found to be an independent predictor of acute renal damage in children with APN. Cystatin C was not found to be a predictive factor for renal damage. CRP levels >100 mg/L during APN and the presence of VUR grade ≥3 were associated with renal scarring.

EP-198 AN UNCLEAR CASE OF NEPHROGENIC DIABETES INSIPIDUS

Malgieri Gabriele, Bruno Minale, Daniela Molino, Vittorio Serio, Luigi Annicchiarico Petruzzelli

Aorn Santobono-nephrology And Dialysis Unit

Introduction: RF,a female ELBW (800 g),with antenatal history of moderate polyhydramnios and transient neonatal diabetes,polyuria,hypernatremia,dismorphic phenotype and clinical diagnosis of Silver Russell syndrome,not confirmed by SNP-Array.At 16 days of post natal age,she developed AKI stage III (creat max 3.43 mg/dl).Renal echography was negative for pathological findings.Infusional program was started and creatinine value of 0.4 mg/dl was achieved.
Material and methods: Polyuria,hypernatremia and poor growth was persistent.To differentiate CDI from NDI we applied Desmopressine Infusion Test.The test findings were compatible with diagnosis of NDI.
At 6 months of life,first access at our Nephrology Unit.Persistent poor growth,polyuria (12/ml/kg/h),hypernatremia,mild hypokaliemia (3.2-3.4 mEq/lt) was present.Venous EGA was normal.Renal echography was negative.
The inusual persistence of hypokaliemia and antenatal history of mild polyhydramnios imposed a diagnostic reassessment.Bilateral hypoacusia was diagnosed.
Genetic test,highlighted the presence of genomic variants c.1038_1042 of the CATTC and c.1432G>A,in gene SLC12A1,both in eterozygous condition (double heterozygosity).
Results: SLC12A1 gene encode for NKCC2 protein (NaK2Cl cotransporter in TAL),which is mutated in BS type 1.
Bartter Syndrome,is a salt losing tubulopathy, characterized by polyuria, hypokalemia, hypochloremic metabolic alkalosis, failure to thrive, growth retardation, and a medical history of polyhydramnios with premature birth.
Indomethacine treatment was started.Polyuria decreased,and growth improved,but value of natremia of 138-140 mEq/lt are persistent,despite the increase of dosage of indomethacine.
Conclusions: In our patient BS1 hasn't a classical presentation:absence of nephrocalcinosis,metabolic alkalosis and hyponatremia delayed the diagnosis.
We don't forget that the NDI may be secondary not only primitive.
The BS1 with secondary NDI represents a diagnostic and treatment challenge in acute and chronic phase.

EP-199 FOCAL GLOMERULOSCLEROSIS WITH IGA DEPOSITS IN A PATIENT WITH ULCERATIVE COLITIS TREATED WITH MESALAMINE

Federica Zotta, Marina Vivarelli

Irccs, Division Of Nephrology And Dialysis Bambin Gesu Pediatric Hospital, Rome Italy

Introduction: Drug-induced interstitial nephritis and nephrotic syndrome (NS) mainly secondary to minimal change disease have been reported in patients with ulcerative colitis (UC) treated with mesalamine. We report the first case of a pediatric patient with UC and focal segmental glomerulosclerosis (FSGS) associated with IgA deposits.
Results: A 16-year-old with a recent history of UC and without extraintestinal manifestations, presented with a bilateral swelling in lower extremities and weight gain of 10 kg during two months. For UC, the patient was initiated on 5-aminosalicylate mesalamine and metronidazole for seven days. After 2 months of this regimen, he developed the edema.Further laboratory investigation showed significant hypoproteinemia (4,6 g/dl), hypoalbuminemia (2,3 g/dl) and the urine protein-to-creatinine ratio was 3,59 mg/mg associated with microscopic hematuria (8-10 pf). Blood pressure and renal function were normal. Renal ultrasonography disclosed no abnormalities. Normal levels of C3 and C4 were observed and no autoimmune factors were detected. Renal biopsy revealed a FSGS associated with mesangial IgA deposits. No signs of interstitial nephritis were found.Prednisone (PDN) 60 mg/m2 /day and ramipril 1,25 mg/day were initiated and mesalamine was stopped immediately. A rapid remission of the nephrotic syndrome (NS) was observed within 4 weeks of steroid therapy. After 6 months, PDN therapy was discontinued but it was transiently reintroduced due to relapse of gastrointestinal symptoms.
Conclusions: In our patient, the appearance of NS during clinical remission of UC appears to exclude an extra-intestinal involvement of the UC and may suggest a role of Mesalamine. Mesangial IgA deposits could be secondary to chronic intestinal inflammation in this UC patient. It is unknown how mesalamine triggers FSGS, but the pathogenesis could be related to reversible podocyte toxicity. Six months of steroids therapy, ACE-i along with cessation of mesalamine induced remission of NS.

EP-200 DENT DISEASE TYPE 2 AS A CAUSE OF FOCAL SEGMENTAL GLOMERULOSCLEROSIS IN A 6-YEAR-OLD BOY

Jakub Zieg[1], Jan Langer[2], JaromÍr HÁČek[1], Martin Bezdíčka[1]

[1]Motol University Hospital Prague, Czech Republic, *[2]First Faculty Of Medicine, Charles University And General University Hospital In Prague, Prague, Czech Republic*

Introduction: Dent disease is an X-linked recessive renal tubular disorder characterized by proximal tubule dysfunction. Typical features include low molecular weight proteinuria, hypercalciuria, nephrocalcinosis, nephrolithiasis, rickets, and chronic renal failure. The disease is caused by mutations in either the CLCN5 (Dent disease 1) or OCRL1 (Dent disease 2) genes.
Material and methods: We present a case of a 6-year-old boy with nephrotic proteinuria without hypoalbuminemia or edema. His renal biopsy revealed focal segmental glomerulosclerosis (FSGS), some of the glomeruli were globally sclerotic. Hypercalciuria was present intermittently and urine protein electrophoresis showed low molecular weight protein fraction of 50%. The family history revealed that his maternal uncle was being followed-up for proteinuria of unknown etiology.
Results: The next generation sequencing identified pathogenic variant in OCRL gene causing Dent disease type 2. The patient was prescribed hydrochlorothiazide at the dose of 1 mg/kg/day, ramipril 5 mg daily and phosphate supplementation. The patient has been followed up in good clinical condition for 3 years in total.
Conclusions: This case report highlights the importance of a complex view of FSGS as a heterogeneous entity. The presence of only mildly elevated albuminuria in a child with nephrotic proteinuria should point to tubular disease. We report an uncommon histologic finding of FSGS in Dent disease type 2 and highlight the importance of protein content examination and genetic analysis for the proper diagnosis in these complicated cases.

EP-201 BARDET-BIEDL SYNDROME CAUSED BY COMPOUND HETEROZYGOSITY OF BBS12 GENE IN ONE FAMILY: A CASE REPORT

Ana Simičić Majce[1], Adela Arapović[1], Sandra Prgomet[1], Marko Todorović[2], Bernarda Lozić[1], Mirna Saraga-babić[3], Marijan Saraga[1]

[1]University Of Split, School Of Medicine, University Hospital In Split, Pediatric Department, *[2]General Hospital Šibenik, Department Of Pediatrics*, *[3]University Of Split, School Of Medicine, Department Of Anatomy, Histology And Embriology*

Introduction: Bardet-Biedl syndrome (BBS) is a rare autosomal recessive ciliopathy, caused by at least 24 genes. It is manifested by various signs visible at birth, such as polydactyly, brachydactyly, and syndactyly. Other signs include obesity, metabolic syndrome, retinopathy, kidney, heart, gastrointestinal, neural, and genital abnormalities which usually develop over time. The prevalence of BBS is 1:100000 -1:13000, depending on geographic location. The most affected genes are BBS1 (51%), BBS10 (20%).
Material and methods: Our patients were a male newborn, his brother, and his sister. They were tested with a gene panel for ciliopathic genes.
Results: The proband was newborn, born in 38th gestational week, with Apgar score 10/10, BW 3200 g, and BL 49 cm. During pregnancy, 1st gynecological ultrasound revealed renal pelvis dilatation, hexadactyly of the right hand and both feet, and syndactyly of 2nd and 3rd fingers. In addition, patent foramen ovale, enlarged and hyperechoic kidneys with a large number of small macrocysts, liver hyperechoic bile ducts, and mildly condensed left lung with lots of B-lines were observed. Based on those findings, we suspected autosomal recessive cystic kidney disease. On the 15th day of life, 2nd ultrasound revealed that only changes in kidneys remained present. His older brother (18 months) also had polycystic kidneys and the same type of hexadactyly and syndactyly, while their older sister (5 years) additionally had hydrometrocolpos and hydronephrosis at birth. We suspected that proband and his siblings have clinical phenotypes of overlapping symptoms of BBS and McKusick Kauffman syndrome. The testing of their panel genes confirmed two pathogenic missense variants, c.1277G>A (p.Cys426Tyr) and c.940A>G (p.Arg314Gly) in the BBS12 gene (each on opposite chromosomes) and supported a diagnosis of autosomal recessive BBS-12 (BBS12, MIM 615989).
Conclusions: We presented a rare case of a family with BBS, caused by compound heterozygosity of the BBS12 gene.

EP-202 NOCTURNAL ENURESIS IN CHILDREN: WHAT PARENTS KNOW ABOUT?

Selsabil Nouir, Sameh Mabrouk, Houda Ajmi, Fadwa Majdoub, Miniar Tfifha, Jalel Chemli, Noura Zouari, Saoussen Abroug

Pediatric Department, Sahloul Hospital, Tunisia

Introduction: Enuresis is defined as a lack of control of urination, most often nocturnal, involuntarily and unconsciously, occurring in children after 5 years, the age of physiological sphincter maturity.
Parents knowledge about enuresis are still insufficient in our Tunian context. The aim of pour study is to determine prevalence of enuresis in primary schools, to study parents knowledge about the trouble and to propose an assistance strategy to enuretic children
Material and methods: A descriptive, observational cross sectional study carried out in school environment in the region of sousse and

studying enuretic children parents knowledges and attitudes about the trouble.
all parents of schooled children aged of 6 to 12 years old and studying in 3 schools randomly chosen in the region of sousse were included
Results: 1200 parent had answered the questionnaire, and 350 children suffered of enureis, the mean age of children was equal to 8.08 +/- 1.86 years. male to female ratio of 1.5
72% of enuretic children in our study had associated symptoms (bladder instability,constipation, behavioral disturbances), the most frequent associated sign was constipation (42.59%) and most frequent reason of consultation was the impact on childs life (91.7%) and family disturbance (54.4%). Parentsconsultation was positively correlated to parents age (p=0.002), marital status (p=0.04) and existance of family history of enuresis (p=0.00).
Parents behaviour regarding eneuresis was studied, 94.9% had the sens of understanding their children and the pathological situation, 52.7% of them reassure their children wheras 21.3% of parents exhibited angry behaviour, 17% compare their children to others
Faced to enuresis all parentss tryed to reduce episods, most of them ensure their child had complete urination before sleep (81.5%), other observed conduct were wearing diapers, fluid restriction and nocturnal awackning.
Conclusions: Eneuresis is a common condition witch can affect child psychology and lower him self esteem. thus, it requires adequate care involving parents and children through an improvment in the level of knowledge

EP-203 EARLY CYSTEAMINE TREATMENT FOR NEPHROPATHIC CYSTINOSIS: RENAL OUTCOME OF RUSSIAN CHILDREN

Valentina Maltseva, Petr Ananin, Tatyana Vashurina, Kirill Savostyanov, Alexander Pushkov, Olga Zrobok, Andrey Fisenko, Alexey Tsygin

Federal State Autonomous Institution "national Medical Research Center For Childrens Health" Of The Ministry Of Health Of The Russian Federation

Introduction: Nephropathic cystinosis is an inherited autosomal recessive disease that leads to early-onset chronic renal failure in consequence to accumulation a lysosomal cystine in cells caused by mutations in the CTNS gene. Early initiation of cysteamine delays progression to end stage kidney disease (ESKD).
Material and methods: Retrospective analysis of renal function of 32 children with nephropathic cystinosis (17 male, 53%) diagnosed in our Center in the period 2008-2021. We analyzed the progression to ESRD in initiated cysteamine treatment groups A (1.0-2.5 years; n=13, 40.6%), B (2.6-5.0 years; n=5, 15.6%), C (after 6.0 years; n=4, 12.5%) and D (without cure; n=10, 31.3%). Renal survival probability rates were calculated according to Kaplan-Meier, log-rank test to compare survival curves.
Results: Median age at initiating of cysteamine therapy was 1.7 years in A group (IQR: 1.1 – 2.1; range: 0.8 – 2.4); median was 3.0 years in group B (IQR: 2.8 – 3.1), median was 5.9 years in group C (IQR: 5.7 – 6.3), median was 8.7 years (mean 11.1 years; IQR: 7.8 – 12.5, range 6.0 - 26.5) in group D. Twenty one (66%) children reached ESRD at mean 10.5 years (median: 9.6; IQR: 8.2 – 13.3; range: 6.5 – 15.4), of which 16 (50%) patients had a kidney transplantation, not including 3 deaths. Log-rank analysis showed that early starting cysteamine therapy significantly delayed the ESRD onset (p = 0.032), median survival time in A group was 11.8 years (95%Cl 8.0 – 15.6) vs. 8.9 years (95%Cl 2.5 – 15.3) in B group vs 7.7 years (95%Cl 5.4 – 10.1) in C group vs. 7.8 years in group D (95%Cl 7.2 – 8.3), respectively.
Conclusions: Orally initiation of cysteamine significant the delays ESKD in children with cystinosis.

EP-204 THE COMBINATION OF SERUM CYSTATIN C, DOPPLER RENAL RESISTANCE INDEX AND KIDNEY INJURY MOLECULE-1 IN EARLY DETECTION OF DIABETIC KIDNEY DISEASE IN CHILDREN WITH DIABETES MELLITUS TYPE 1

Ivana Trutin[1], Gordana Stipančić[1], Lea Oletić[1], Mario Laganović[2]

[1]*Clinical Hospital Center Sestre Milosrdnice Zagreb*, [2]*University Hospital Centre Zagreb, Zagreb*

Introduction: Diabetic kidney disease is the leading cause of end-stage renal disease. Regression to normoalbuminuria in already albuminuric children calls into question albuminuria as an early indicator of diabetic kidney disease development. The aim of this study is to develop a clinically useful model based on the association of serum cystatin C, Doppler renal resistance index and kidney injury molecule-1 in urine with standard indicators of renal function in order to determine the optimal model for early detection of diabetic kidney disease.
Material and methods: The study included 75 children with type 1 diabetes(T1D), normoalbuminuria and normal renal function aged 10-18 years, and 75 healthy children in the control group. In both groups, we determined serum cystatin C, KIM- 1 in urine, renal resistance index(RI), lipidogram, thyroid hormones, serum creatinine, anthropometric parameters, blood pressure, glycated hemoglobin, estimated glomerular filtration rate, serum uric acid, urine albumin / creatinine ratio ,and degree of pubertal development.
Results: The results suggest that RI and urinary KIM-1 are higher in patients with T1D compared to the control group. In this study, RI is higher in affected children and is useful for assessing early hemodynamic changes in the preclinical phase of early renal impairment. Dyslipidemia, higher values of serum creatinine and insulin dose are associated with the increased risk of developing microalbuminuria and diabetic kidney disease.
Conclusions: combination of serum cystatin C, KIM-1 in urine and RI plays a significant role in future preventive and therapeutic action.

EP-205 POLYMORPHISM OF STAT4, PTPN22, VEGF, TGF-B, PDCD1 AND PD-L1 IN CHILDREN WITH HEREDITARY NEPHRITIS

Hanna Bialkevich[1], Ina Kazyra[1], Aleksandr Sukalo[1], Natalia Nikitchenko[2], Roza Goncharova[2]

[1]*Belarusian State Medical University*, [2]*Institute Of Genetics And Cytology Of The National Academy Of Sciences Of Belarus*

Introduction: To study the genetic polymorphism of STAT4, PTPN22, VEGF, TGF-B, PDCD1 and PD-L1 in children with hereditary nephritis (HN).
Material and methods: 39 patients with HN aged 6 to 16 years (m:f 25:14) and 416 children without kidney diseases aged 13 to 16 years (m:f 236:180) were included. We use a standard method of phenol-chloroform extraction to isolate genomic DNA. Polymorphic variants of genes were determined using such methods of polymerase chain reaction (PCR) as estriction fragment length polymorphism PCR and real-time PCR.
Results: the distribution of frequencies of allele among the entire sample subjected to the Hardy-Weinberg law and was similar to frequency values in European populations. In children with HN the frequencies of genotypes and alleles by polymorphic loci of STAT4 rs7574865 and rs3821236, PTPN22 rs2476601, TGF-B rs1800469, PDCD1 rs11568821, VEGF rs699947 and rs2010963 didnt differ from the control group. However, association of the locus rs2297136 of PD-L1 with HN was established. GA genotype was the most common

for HN (in 14/39 (35.9%) cases), carriers of the G allele were 22/39 (56.4%) patients. According to recessive and log-additive models, homozygous genotype GG and minor allele G were significantly more often detected in children with HN in the boys group compared to the control group (OR=2.74 (95%CI 1.16 – 6.49), p=0.027 and OR=1.94 (95%CI 1.03 – 3.65), p=0.037 respectively).
Conclusions: GG genotype and allele G of the polymorphic locus rs2297136 of the PD-L1 are associated with the risk of HN in the group of male patients. These data can be used to develop new methods of HN diagnosis.

EP-206 HEPATIC-ASSOCIATED IGA NEPHROPATHY IN A CHILD WITH PORTAL HYPERTENSION

Emre LeventoĞlu[1], Bahar BÜyÜkkaragÖz[1], Bahriye Uzun Kenan[1], Sinan Sari[2], Sevcan A. BakkaloĞlu[1]

[1]Gazi University Faculty Of Medicine Pediatric Nephrology, [2]Gazi University Faculty Of Medicine Pediatric Gastroenterology

Introduction: IgA nephropathy is a commonly reported glomerular disease in adults with liver disease, especially alcoholic liver cirrhosis, chronic hepatitis or portal hypertension. Although its prevalence in children is not known clearly. The pathogenesis is characterized by the disruption in the clearance of circulating immune complexes by Kupffer cells which cause the immune complexes to accumulate in the kidneys.
Material and methods: We present a patient with portal hypertension who developed IgA nephropathy.
Results: 8-year old boy who was diagnosed as non-cirrhotic portal hypertension at the age of 4 and using propranolol applied to hospital because of recurrent gross hematuria. Physical examination showed only splenomegaly. Laboratory analysis showed bicytopenia, normal kidney functions and normal liver enzymes. Urinalysis was positive for blood and dysmorphic red blood cells were seen with no red cell casts on microscopy. There was minimal range proteinuria. Ophthalmological examination and hearing test were normal. His serum complement levels were mildly decreased; C3 and C4 were at 71.9 mg/dl (79-152) and 11 mg/dl (16-38), respectively. Ultrasonography showed splenomegaly, but kidneys were normal in size and echogenicity, and renal doppler was normal. In portal system doppler, cavernous transformation in the portal vein was detected with the hepatopedal direction of the portal vein.
Cystoscopy was performed and no focus was observed. Repeated liver biopsy showed near-normal histology; the diagnosis of non-cirrhotic portal hypertension was confirmed. Kidney biopsy was performed; mesangial expansion, hypercellularity and mesangial IgA, IgG, IgM, and C3 staining were seen. Therefore, due to the presence of non-cirrhotic portal hypertension, full-house immune complex storage and mesangial hypercellularity in kidney biopsy, the patient was diagnosed to have secondary IgA nephropathy associated with liver disease.
Our patient had already been treated with propranolol but the dose of propranolol was increased because of some improvement in IgA nephropathy can occur with control of portal hypertension. Although microscopic hematuria persisted in the follow-up period of approximately three years, no further macroscopic hematuria attack was observed.
Conclusions: The clinicians dealing with children with chronic liver disease should be aware of hepatic-associated IgA nephropathy and renal-related complications should be taken into consideration.

EP-207 RELATIONSHIP BETWEEN THE RENAL FUNCTION AND THE RENAL PELVIS ANTEROPOSTERIOR DIAMETER IN CHILDREN WITH URETEROPELVIC JUNCTION OBSTRUCTION

Erkam Yildirim, Gizem Yildiz, Meral Torun Bayram, Alper Soylu, Salih Kavukcu

Dokuz EylÜl University Medical Faculty, Department Of Pediatric Nephrology, Izmir, Turkey

Introduction: Ureteropelvic junction obstruction (UPJO) may lead to renal parenchymal damage. Kidneys with large intrarenal pelvis have been reported to be more resistant to obstruction damage than those with small intrarenal pelvis. We evaluated the relationship between renal function and renal pelvis anteroposterior diameter (APD) in UPJO.
Material and methods: Children with unilateral SFU grade 4 hydronephrosis were enrolled. Demographic data, antenatal/postnatal diagnosis, ultrasonographic and radionuclide imaging data were noted. Antenatal cases were grouped as <20 vs ≥20 mm, <30 vs ≥30 mm, while postnatal cases as <30 vs ≥30 mm based on APD. These groups were compared for parenchymal thickness and differential function. Correlation of APD with parenchymal thickness and differential function was also evaluated.
Results: There were 28 patients [21 (75%) male; 17 (61%) antenatal diagnosis]. Median age at diagnosis was 83 (24-198) months in postnatal cases who presented with flank pain (8), urinary tract infection (2) or coincidentally (1). Pyeloplasty was performed in all patients except one antenatal case. Median age at pyeloplasty was 6 (3-31) and 81 (43-180) months in antenatal and postnatal cases, respectively. Parenchymal thickness and differential function of hydronephrotic kidneys were not different in both antenatal and postnatal case groups. APD was not correlated with parenchymal thickness and differential function in antenatal and postnatal cases. Only 6 patients (5 postnatal) had <30% differential function in the hydronephrotic kidneys. APD of the postnatal cases with <30% differential function was ≥37 mm, but mean APD of these patients was not different from the other postnatal cases. APD of the antenatal case with <30% differential function was relatively small (20 mm).
Conclusions: Our findings did not confirm that the smaller the intrarenal pelvis the higher the renal damage in UPJO. The single antenatal case with higher renal damage despite relatively small APD suggests that this damage is due renal dysplasia in association with UPJO.

EP-208 CONGENITAL NEPHROTIC SYNDROME: A CASE OF AN UNUSUAL FAVORABLE DISEASE COURSE

Eline Hermans, Johan Vande Walle, Agnieszka Prytula, Joke Dehoorne, Evelien Snauwaert, Lien Dossche, Ann Raes

Ghent University Hospital

Introduction: Congenital nephrotic syndrome (CNS) is a rare kidney disease characterized by the typical triad of edema, proteinuria and hypoalbuminemia within the first 3 months of life. Most cases are due to defects in the NPHS1 gene, resulting in CNS of the Finnish type. This report discusses the unusual disease course of a boy with CNS of the Finnish type.
Material and methods: We present the case of a boy of 2 ½ years old with CNS of the Finnish type. He was diagnosed at the age of 2 months after being admitted to the hospital with vomiting and ascites. Genetic analysis of the NPHS1 gene revealed compound heterozygosity for the mutation C.1868G>T p.(Cys623Phe) and c.1913A>G p.(Tyr638Cys). The boy was treated with intermittent regular albumin infusions, diuretics and angiotensin-converting-enzym inhibition (ACE-I). The course of this CNS was favorable with tapering of the albumin infusions and complete withdrawal at the age of 11 months. Diuretics were discontinued 7 months later. To this date, his renal function remains normal, with stable mild proteinuria, stable low-normal serum albumin and normal blood pressure under ACE-I.

Results: The typical CNS of the Finnish type, caused by Finmajor (p.Leu41fs*91) and Finminor (p.Arg1109*) mutations, demonstrates relatively little phenotypic variation with massive proteinuria at birth and rapid progression to end-stage renal disease. This boy showed partial remission of CNS, probably linked to the different NPHS1 mutations. Only eight other isolated cases of CNS of the Finnish type with a favorable outcome are reported in the literature. Additionally, one specific family with multiple cases of CNS and a specific ethnic group have been described to have a milder phenotype.
Conclusions: CNS of the Finnish type can rarely present with a mild disease course with prolonged preservation of renal function.

EP-209 DIFFICULT CASES OF PERITONITIS WITH UNUSUAL MICROORGANISMS IN CHILDREN WITH PERITONEAL DIALYSIS

Ece Demirci Bodur, Nurdan Yildiz, Serim Pul, Neslihan ÇiÇek, SerÇin GÜven, Özde Nisa TÜrkkan, İbrahim GÖkÇe, Harika Alpay

Marmara University Pediatric Nephrology

Introduction: We present three peritonitis episodes caused by Sphingomonas melonis,Stenotrophomonas maltophilia and Finegoldia magna resulted in catheter loss in two patients on chronic peritoneal dialysis(PD).
Material and methods: Case 1 A 11-year old girl receiving PD for two years admitted with abdominal pain and cloudy effluent with white blood cell(WBC) count of 880/mm^3(69% neutrophils) . After obtaining dialysate culture, intraperitoneal cefazolin and ceftazidime were started. On fifth day,dialysate WBC decreased to 50/mm^3.Sphingomonas melonis,a gram negative plant pathogen that was isolated from melons,grew on culture and was susceptible to sefotaxime.On further inquiring, she reported that she often played in the garden near their house. On 13th day,dialysate WBC increased and intravenous meropenem started based on antibiogram. On 16th day,she had fever and increased dialysate WBC count.Three other dialysate cultures were still positive for Sphingomonas melonis, and PD catheter was removed.
Results: Case 2 A 7-year old girl recieving PD for 11 months admitted with abdominal pain and cloudy effluent with WBC count of 720/mm^3(65% neutrophils).Intravenous meropenem was started based on antibiograms of previous peritonitis. Stenotrophomonas maltophilia, a gram negative multidrug resistant opportunistic pathogen was isolated and treatment switched to intravenous trimethoprim-sulfamethoxazole and levofloxacin.After 2 weeks,WBC count did not decrease and catheher was replaced.One year later, she came with severe peritonitis attack.Intravenous vancomycin and piperacillin-tazobactam was started.Finegoldia magna, an anaerobic gram(+) bacteria, was isolated and the treatment switched to intravenous vancomycin and meropenem based on antibiogram.However,peritonitis did not resolved and PD catheter was removed.
Conclusions: Stenotrophomonas maltophilia,Sphingomonas melonis and Finegoldia magna are unusual pathogens for PD-associated peritonitis, the last two of which have not been reported before. Peritonitis with uncommon microorganisms are potentially resistant to treatment and PD catheter removal is required to cure the infection.Strict hygiene rules must be followed by patients/caregivers and the patient's technique and habbits should be reassessed especially for such rare cases.

EP-210 COULD NEUTROPHIL-LYMPHOCYTE RATIO BE USED AS A PROGNOSTIC FACTOR IN HEMOLYTIC UREMIC SYNDROME?

Nurdan Yildiz[1], Ceren Bİlgun[2], Sercin Guven[1], Ece Demirci Bodur[1], Neslihan Cicek[1], Serim Pul[1], Ozde Nisa Turkkan[1], Ibrahim Gokce[1], Harika Alpay[1]

[1]Marmara University, Medical School, Division Of Pediatric Nephrology, *[2]Marmara University, Medical School, Division Of Pediatrics*

Introduction: Neutrophils to lymphocytes ratio (NLR) is a marker of inflammation that has been associated with chronic conditions with low-inflammation, poor clinical outcomes in malignancies and cardiovascular disease.We aimed to assess the value of NLR at the time of diagnosis in predicting clinical course including need for dialysis and development of chronic kidney disease(CKD), and distinguishing atypical/typical hemolytic uremic syndrome(aHUS) in the first attack of the patients.
Material and methods: Forty eight patients with HUS and 50 healthy children were included in this cross-sectional retrospective study.Demographic, clinical and laboratory findings, need for dialysis, genetic analysis if any, stool analysis results were recorded from medical files. Neutrophil/lymphocyte ratio and estimated glomerular filtration rates (eGFR) at first admission were calculated.The relationship between NLR,mean platelet volume(MPV) and clinical course,development of hypertension,proteinuria and chronic kidney disease were evaluated. Statistical Package for the Social Sciences (SPSS Inc,Chicago,IL,USA) version 21.0 program was used for the statistical analysis.
Results: The mean NLR value of the patients and controls were 1.98 ±1.46 and 0.95±0.65, respectively. Neutrophil/lymphocyte ratio were significantly higher and the MPV were significantly lower in patients than the controls (p=0.000 and p=0.000 respectively). Moreover, higher NLR was significantly associated with need for dialysis at the time of diagnosis (p=0.02).On the other hand,NLR was not different in patient with typical and atypical HUS(p=0.95). In long-term follow-up, NLR at the time of diagnosis was significantly higher in patients with eGFR<60 than the patients with eGFR >60 ml/min/1.73 m^2(p=0.025).Higher NLR values were not associated with sequelae proteinuria and hypertension.
Conclusions: Our results suggested that a high NLR value at the time of diagnosis could be a useful predictor of worsening clinical course,need for dialysis and CKD development.However,it is insufficient in distinguishing typical and atypical HUS and predicting the development of hypertension and proteinuria in long-term.

EP-211 VALIDATION AND CHARACTERIZATION OF A NOVEL DIGENIC MOUSE MODEL OF ARPKD

Claudia Dafinger[1], Sebastian Brähler[2], JÖrg DÖtsch[1], Bernhard Schermer[2], Max Liebau[1]

[1]Department Of Pediatrics, University Hospital Cologne, *[2]Department Of Internal Medicine Ii, University Hospital Cologne*

Introduction: Autosomal recessive polycystic kidney disease (ARPKD) is one of the most severe kidney diseases in childhood and adolescence. ARPKD is characterized by enlarged cystic kidneys and obligatory fibrotic changes in the liver. In most cases, ARPKD is caused by variants in the gene Pkhd1, encoding the transmembrane protein fibrocystin (FC). The function of FC is poorly understood. Our previous data suggest dysregulation of metabolic and inflammatory pathways (e.g. SRC-Stat3).
Material and methods: Existing orthologous mouse models do not fully reflect the renal pathology of ARPKD patients. A novel digenic mouse model, which carries a hypomorphic variant of Pkd1 (R3277C) in addition to a Pkhd1 deficiency, was recently described. Similar to ARPKD patients this model shows early-onset cystic changes in the kidneys. Using various immunhistological and immunofluorescence stainings we

are now focusing on the validation and characterization of this new model in our group.
Results: After importing the new digenic mouse model we were able to successfully reproduce the described hepatic and renal phenotype. To further investigate the underlying pathophysiological mechanisms we analyzed components of metabolic and inflammatory pathways. We did not observe altered distribution of immune cells. However, we could show that the activation of Stat3 is increased in tubular epithelial cells and tubulointerstitial areas of digenic animals.
Conclusions: In summary, we confirm the phenotypical findings of this novel ARPKD mouse model. To gain deeper insight in the causing cellular and molecular mechanisms, we are currently further characterizing this model using high-throughput methods. The new digenic mouse model can further be used for the development of new potential therapeutic strategies.

EP-212 HYPOMAGNESEMIA WITH HYPERCALCIURIA AND NEPHROCALCINOSIS IN AN ADOLESCENT GIRL

Hulya Nalcacioglu[1], Aysegul Yilmaz[2], Demet Tekcan Karali[1], H.gozde Onal[1], Ozlem Aydog[1]

[1]Ondokuz Mayis University Faculty Of Medicine, Pediatric Nephrology Department, Samsun, Turkey, [2]Ondokuz Mayis University Faculty Of Medicine Pediatric Genetic Department, Samsun, Turkey

Introduction: Familial hypomagnesemia with hypercalciuria and nephrocalcinosis (FHHNC) is a rare tubular disorder. The typical features of the disease are hypomagnesemia, hypercalciuria, and nephrocalcinosis, and progression to renal insufficiency is common in this syndrome.
Material and methods: Herein, we report a case of a 16-year-old female who presented clinically with recurrent nephrolithiasis and was diagnosed with FHHNC.
Results: A 16-year-old girl was referred for investigation after a documented renal colic and passage of calculi. Her past medical history was characterized by episodes of the passage of calculi and urologic interventions. The ultrasound examination of the kidneys demonstrated bilateral diffuse medullary nephrocalcinosis. Serum and urine biochemistry revealed hypomagnesemia (0.4 mmol/ l), increased iPTH (415 pg/ml) and hypercalciuria (5 mg/kg/day). Based on these clinical and laboratory parameters, the diagnosis of FHHNC was established. Evaluating for extrarenal symptoms, no hearing abnormalities were found, but she had nystagmus. Magnesium citrate supplementation and hydrochlorothiazide were given and well-tolerated. Renal function was mildly impaired in our patient and was relatively stable during the 1-year follow-up. Medical treatment aiming to control hypomagnesemia and hypercalciuria was partially efficient because of no regular use. The diagnosis of familial hypomagnesemia with hypercalciuria and nephrocalcinosis (FHHNC) was confirmed by mutational analysis.
Conclusions: FHHNC may present with the clinical features with mild hypomagnesemia leading to secondary hyperparathyroidism and should be considered in the presence of nephrocalcinosis with hypercalciuria and hypomagnesemia.

EP-213 THROMBOEMBOLIC DISEASE AS THE FIRST MANIFESTATION OF NEPHROTIC SYNDROME ASSOCIATED WITH GENETIC AND ANATOMICAL THROMBOSIS RISK CONDITION

Mar Espino-Hernández[1], Paloma Gutierrez Medina[2], Julia Vara Martín[1]

[1]Hospital Universitario 12 De Octubre, [2]Hospital Universitario De Getafe

Introduction: Thromboembolic disease is an important and severe complication that occurs in nephrotic syndrome (NS) in approximately 3% of children, associated with a hypercoagulable that leads to an increased risk of thrombotic events.
Material and methods: Review of medicals records of a girl with a first episode of NS that at diagnosis presented as severe venous thrombosis in left lower limb (LLL) with pulmonary thromboembolism (PTE).
Results: A 15-year-old girl was admitted to the hospital with a 3-days history of shortness of breath, asthenia, edema and painful in LLL. She has tachycardia with normal cardiovascular and respiratory physical examination. Rx thorax, ECG were normal. Doppler ultrasonography of lower limbs showed thrombosis involving common and left femoral veins. Computer-tomography-pulmonary-angiography demonstrated PTE affecting both the main pulmonary arteries and some segmental arteries. Laboratory investigation was incidentally detected to have NS, proteinuria (urine P/Cr 9), hypoproteinemia (4,7 mg/dl), hypoalbuminemia (2 mg/dl). Platelet count, creatinine, electrolytes, prothrombin, partial thromboplastin time, protein C and S, antithrombin were norma. Anticardiolipin antibodies and lupues anticoagulant were negative. But heterozygous mutation was identified in prothrombin gene G202210, which increases thrombotic risks. Anticoagulant treatment was started with low-molecular-weight heparin and steroid treatment for NS. Additional investigation by abdominal angio-magnetic-resonance identified compression of the left common iliac vein by the right common iliac artery (May-Thurner syndrome) that also constitutes a thrombotic risk. The patient recovered successfully, developed a completed remission of NS. She receives thromboprophylaxis treatment.
Conclusions: Thromboembolic disease is associated with high mortality, therefore early diagnosis and treatment are essential for patient survival. Screening for hereditary thrombophilia may be necessary for NS patients with thromboembolic events. May-Thurner syndrome should be investigated in patients with venous thrombotic events in left lower limb.

EP-214 PREDICTIVE FACTORS OF URINARY TRACT INFECTIONS AFTER PEDIATRIC KIDNEY TRANSPLANTATION

Abir Boussetta[1], Farah Krifi[1], Nesrine Abida[1], Taieb Ben Abdallah[2], Manel Jellouli[1], Tahar Gargah[1]

[1]Pediatric Nephrology Department, Charles Nicolle Hospital Tunis, Tunisia, [2]Research Unit Of Immunopathology And Immunology Of Renal Transplantation Lr03sp01

Introduction: To determine the predictive factors of urinary tract infections (UTIs) after kidney transplantation (KT) in Tunisian children.
Material and methods: This was a cross-sectional, descriptive, retrospective study carried out in the pediatric and internal medicine A departments of the Charles Nicolle hospital in Tunis. Transplanted patients of age less than or equal to 20 years were included during a 31-year period (from January 1989 to December 2019). A multivariate study was conducted to determine the independent predictive factors for the occurrence of UTIs after KT. A p-value of less than 0.05 was considered significant.
Results: A total of 115 kidney transplantations were included in our study during this period (69 boys and 46 girls). The average age was 15.5 years old. The transplantation was done from a living donor in in 67.8 % of cases. Infectious complications were frequent and dominated by UTIs in 37.4 % of cases. The mean time to onset of this UTI was 504.8 days. Its average number per patient was 3.4. The most implicated germ was Escherichia coli.The identified predictive factors of these UTIs were: Recipient's male gender, recipient's age, presence of vesicoureteral reflux prior to KT, presence of urological complications especially ureteral stenosis, the presence of urinary lithiasis, vesico-renal reflux, and the number of hospitalizations during the first year after KT ($p<0.05$). There was

no statistically significant difference between the survival rate of transplant recipients and grafts according to the presence or absence of post-transplant UTI.
Conclusions: Urinary tract infection is the most common complication in children after KT. Several predictive factors have been identified in our study. Monitoring of patients at high risk of UTIs after KT would help to anticipate these infections.

EP-215 TWO RARE CASES OF INCIDENTALLY DETECTED "PANCAKE" KIDNEY

Chrysoula Kosmeri[1], Eleni Papastergiou[1], Ioanna Aggeli[1], Evangelia Gkika[2], Chrissa Sioka[2], Ekaterini Siomou[1]

[1]Department Of Pediatrics, University Hospital Of Ioannina, Ioannina, Greece, [2]Department Of Nuclear Medicine, University Hospital Of Ioannina, Ioannina, Greece

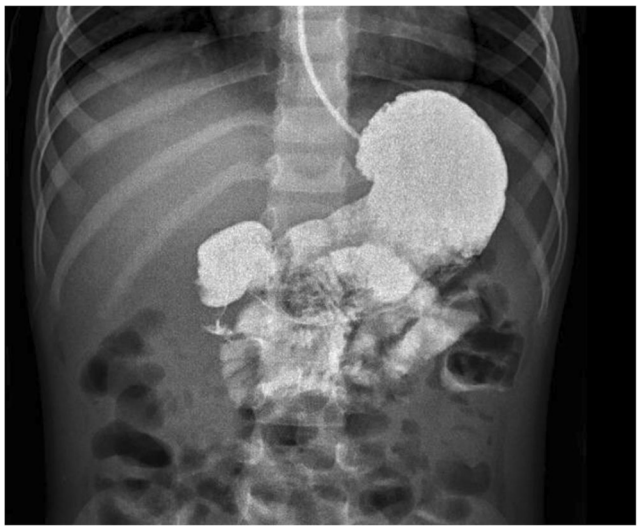
Introduction:"Pancake" kidney is one of the rarest structural renal anomalies and is a rare subtype of crossed fused renal ectopia. We present two cases of "pancake" kidney detected accidentally.
Material and methods: A case of a male neonate 15 days old and of a female infant 4.5 months old that were diagnosed and followed up in our department are described.
Results: In both cases, the evaluation was conducted due to abnormal findings in antenatal and postnatal ultrasound (findings of possible horseshoe kidney in the first case and crossed fused renal ectopia in the second). The infants were asymptomatic, and the prenatal history was unremarkable. Technetium 99m-dimercaptosuccinic acid (DMSA) scintigraphy was conducted in both cases and ectopic kidneys that were fused in their upper, medial, and lower poles were pictured representing a "pancake" kidney. Each kidney had its excretory system and, in each case, both kidneys had normal uptake with no signs of parenchymal lesions. Both patients were asymptomatic, had no history of urinary tract infections, had a normal renal function, and had no other congenital anomalies.
Conclusions: Children with "pancake" kidney usually remain asymptomatic and the diagnosis is often made accidentally, as in our cases. However, the clinician should be aware that the "pancake" kidney may be accompanied by other congenital anomalies of the urinary tract and an increased risk for malignancy. Therefore, these patients should be followed-up systematically by ultrasound and renal function tests.

EP-216 HEMOLYTIC UREMIC SYNDROME COMPLICATED BY DUODENAL INTRAMURAL HEMATOMA

Mahnaz Sadeghian[1], Elham Zarei[1]

[1]Aliasghar Children Hospital, Department Of Pediatrics, School Of Medicine,iran University Of Medical Sciences, Tehran Iran, [2]Aliasghar Children Hospital, Department Of Radiology, School Of Medicine,iran University Of Medical Sciences, Tehran Iran

Introduction: Hemolytic-uremic syndrome (HUS) is the most common cause of acute renal failure in young children, characterized by the triad of microangiopathic hemolytic anemia, thrombocytopenia, and uremia. Whereas Serious gastrointestinal complications like gut necrosis, hemorrhagic colitis, pancreatitis, transient diabetes, hepatitis, cholestasis, peritonitis, and rectal prolaps have already been reported. We report a case of HUS complicated by duodenal intramural hematoma in a previously healthy 5 year old girl.
Results: A 5-year-old, previously healthy girl presented with a history of fever, dysentery, and hematuria admitted in a remote local hospital with diagnosis of SARS- Covid19 and treated with prednisolon 50mg /day, metronidazole, dimeticone, metoclopramid. Subsequently she developed severe anemia (received packed cell) and tonic clonic generalized seizure. Ten days later, she referred to our center for HUS management. The course of hospitalization was complicated with intermitent fever (urosepsis E. coli, *Klebsiella,* candida), encephalitis, pericarditits, and hyper reactive airway disease.SARS- covid19, immune deficiency investigations were negative(Figure1) .The patient developed protracted vomiting and persistent abdominal pain that was associated with pancreatitis, anicteric hepatitis. After ten days supportive therapy she presented with massive bilious vomiting, abdominal distention, and pain with no tenderness. Bile stained duodenal mucosa was seen on endoscopy. Abdominal scan and upper GI series revealed two hypoechoic heterogeneous collection with 15*14 and 40*30 mm in the wall of proximal part of 2nd and 3rd portion of duodenum suggesting intramural hematoma that was treated by fasting, parenteral hyperalimentation ,and low dose octerotide. Ultimately, resolution occurred in 3 weeks. After four months of follow up, she was completely recovered.



Conclusions: Conclusion: Although Intramural hematoma is rare, it can complicate the course of HUS.

EP-217 BK VIRUS NEPHROPATY PATIENT TREATED WITH LEFLUNOMIDE

BeltİnGe DemircioĞlu KiliÇ, Mehtap Akbalik Kara, Mithat BÜyÜkÇelik, AyŞe Balat

Gaziantep University, Department Of Pediatric Nephrology,gazİantep/ Turkey

Introduction: BK virus is one of the most important infectious disease which can be seen 1-10% after kidney transplantation and may cause graft loss 30-80% of cases. Reducing immunosuppressive therapy remains the first step of treatment. The efficacy of treatments such as cidofovir, leflunomide and intravenous immunoglobulin (IVIG) is still controversial. We wanted to present a patient of BK virus nephropaty which we diagnosed early with close follow-up and responded to leflumonide treatment.

Material and methods: Case presentation: While the patient was followed-up on chronic dialysis due to posterior uretral valve and neurogenic bladder, cadaveric transplantation was performed in our center. Anti-thymocyte globulin and steroid were given for the induction and mycophenolate mofetil (MMF), tacrolimus and steroid were given for the maintenance therapy. He was discharged on the 23rd day after transplantation with a level of creatinine 1.1 mg/dl. On the 35th day after transplantation, creatinine was 1.46 mg/dl due to pyelonephritis, CRP level was 94.9 mg/L and BK virus was 1928 copies/ml. Meanwhile MMF was reduced in first step. As the BK virus continues to increase in weekly follow-up, tacrolimus dosage was also reduced and ciprofloxacin treatment was given. BK virus was detected 14000 copies/ml, tacrolimus was changed to cyclosporine-A (Cyc-A) and IVIG treatment was initiated. Unfortunately BK virus copy number reached to 43000 copies/ml then MMF was discontinued and leflunomide was started. Finally BK virus became completely negative in the fourth month of the follow-up. The patient creatinine level is 0.9 mg/dl at the 24 th month after transplantation and he is maintained with Cyc-A, leflunomide and steroid.
Results: No
Conclusions: Although there is no effective antiviral agent for the causative agent, early detection and close follow-up of virus are important. Leflunomide USAge instead of MMF can be effective and safe in pediatric patients who did not respond other therapies.

EP-218 RANK/RANKL EXPRESSION IN FETAL DEVELOPING, POSTNATAL AND NEPHROTIC HUMAN KIDNEYS

Marija Juric, Ivona Kosovic, Katarina Vukojevic, Mirna Saraga-babic, Natalija Filipovic

Department Of Anatomy, Histology And Embryology, School Of Medicine, University Of Split, Split, Croatia

Introduction: RANKL, the member of the tumor necrosis factor (TNF) superfamily, binds to receptor activator of NF-kB, RANK, who belongs to the TNFR superfamily. RANK/RANKL pathway plays critical role in bone metabolism and immunity. Pathological kidney conditions are linked to changes in RANK/RANKL expression and localisation. The aim of this study was to describe changes in spatiotemporal expression of RANK and RANKL proteins within fetal developing, postnatal and nephrotic human kidneys.
Material and methods: The expression of RANK and RANKL were examined on paraffin sections by immunofluorescence of 8 to 10- and 38-week old developing, postnatal 1.5-year old and nephrotic syndrome patient human kidney tissue.
Results: In 8-10^{th} developmental week (dw), RANK was weakly expressed in all structures, but during further development, in 38^{th} dw, expression was absent. In postnatal healthy kidneys, expression was visible only in glomeruli, at the visceral layer of the Bowman's capsule and in few glomeruli cells, while in nephrotic kidneys the expression at the same locations was much stronger. Regarding RANKL, the expression was more prominent in all presented specimens. Between 8^{th} and 10^{th} dw RANKL was expressed strongly in renal vesicle, C and S shape bodies, but was absent from collecting tubules. In 38^{th} dw RANKL was expressed strongly in all cells of distal convoluted tubules (DCT), moderately in cells of proximal convoluted tubules (PCT) and weakly in glomeruli. In postnatal healthy kidneys, RANKL was expressed strongly in PCT and DCT, as well as in glomeruli, especially parietal layer of the Bowman's capsule. In nephrotic kidneys moderate expression of RANKL in glomeruli, PCT and DCT was visible, with the strongest expression in affected, enlarged tubules.
Conclusions: Expression pattern of RANK and RANKL implicates their role in different stages of kidney development and may provide us with better understanding of RANK/RANKL pathway in pathological conditions.

EP-219 NEUTROPHIL-TO-LYMPHOCYTE AND PLATELET-TO-LYMPHOCYTE RATIO IN PERITONEAL DIALYSIS ASSOCIATED PERITONITIS IN PEDIATRIC PATIENTS

Ece Demirci Bodur[1], SerÇin GÜven[1], Pinar Zeytun[2], Serim Pul[1], Neslihan ÇiÇek[1], Özde Nisa TÜrkkan[1], Nurdan Yildiz[1], İbrahim GÖkÇe[1], Harika Alpay[1]

[1]Marmara University Pediatric Nephrology, [2]Marmara University Pediatrics

Introduction: Neutrophil to lymphocyte ratio (NLR) and platelet to lymphocyte ratio (PLR) have been evaluated for an inflammatory marker in many of diseases lately. We aimed to evaluate the relationship between these parameters and the characteristics of (PDAP) episodes in children.
Material and methods: A total of 44 episodes of PDAP experienced by 20 patients were included in this study. From complete blood count (CBC), initial NLR and PLR were calculated and leukocyte count of direct microscopic examination of peritoneal effluent, C-reactive protein (CRP), hemoglobin (Hb), creatinin, MPV, albumin values on admission were detected. Age, gender, peritonitis rate and duration, time to each peritonitis episode, treatment failure (loss of peritoneal catheter or transfer to hemodialysis) were recorded. Median value of 3.8 and 134,75 were used for NLR and PLR, respectively. We divided the patients into two groups according to the median values of NLR and PLR as low and high NLR and PLR groups.They were compared to biochemical and clinical parameters.
Results: Eleven patients were female (55%). Median age was 67 months (min-max:5-205 months).Time on peritoneal dialysis therapy was median 12 months (min-max:1-65 months). Median peritonitis duration was 5 days (min-max:2-27 months). Eighteen (40.9%) episodes were caused by Gram negative microorganisms.Sixty-three percent of peritonitis episodes were presented with more than 1000/mm^3 leukocytes on direct microskopic examination whereas 6.8% were below 100/$mm^{3.}$ Nine (6.8%) episodes were resulted with treatment failure. Median CBC values were as following: white blood cell (WBC) of $11*10^3\mu L$, neutrophil of $8*10^3\mu L$, lymphocyte of $2*10^3\mu L$, platelet of $290*10^3\mu L$, NLR of 3.8, PLR of 134.75,Hb of 9.8 g/dL, creatinine of 4.43 mg/dL, CRP of 18.3 mg/L, albumin of 3.25 g/dL. Older age, higher values of , Hb, creatinine,MPV,albumin and leukocyte count on CBC were found to be associated with higher NLR (>3.81). Older age, higher Hb and higher platelet counts were associated with higher PLR (>134.75). NLR and PLR were not associated with gender, peritoneal effluent leukocyte count on direct examination, type of microorganism, multiple peritonitis episodes or treatment failure.
Conclusions: NLR and PLR are not associated with the severity or prognosis of PDAP in pediatric patients.

EP-220 A NOVEL MISSENSE MUTATION IN SLC12A3 GENE IN TWO SIBLINGS WITH GITELMAN SYNDROME

Asli Çelebİ Tayfur[1], Zehra Meral[2], Meyrİ Arzu YoldaŞ[2]

[1]Bolu Abant İzzet Baysal University, Faculty Of Medicine, Department Of Pediatric Nephrology, [2]Bolu Abant İzzet Baysal University, Faculty Of Medicine, Department Of Pediatrics

Introduction: Gitelman syndrome (GS) is s a rare autosomal recessive hereditary salt-losing tubulopathy characterized by hypokalemic

metabolic alkalosis with hypomagnesemia and hypocalciuria. GS is caused by mutations in the SLC12A3 gene encoding the thiazide-sensitive sodium chloride contrasporter.
Results: We report here 2 siblings (patient 1: a 2.5 years old boy, patient 2: a 8.5 years old girl) with Gitelman syndrome presenting with the complaints of muscle weakness, fatigue, cramps and polyuria. There was a consanguinity between parents . On physical examination at admission the siblings were normotensive but both had growth retardation. Laboratory analysis showed hypochloremic metabolic alkolosis, hypomagnesemia, hypokalaemia, hyperreninemia, hypocalciuria. Serum sodium, serum creatinine and blood urea nitrogen levels, 24 hour urine chloride level and renal ultrasonography findings were normal in both patients. Genetic testing showed a compound heterozygous mutation [c.488 C> (p. Thr163Met) and c.426delG (p. Met143*)] in transcript NM_000339.2 in SLC12A3 gene in the proband. The heterozygous c.426delG mutation was a novel missense mutation that has not previously been reported in the literature in association with GS. The patients were was treated with magnesium and potassium supplements and and a diet rich in nutrients with high potassium nd high magnessium.
Conclusions: Patients with electrolyte disturbances may present with muscle weakness, fatigue cramps and polyuria. Gitelman sydrome should be considered in the differential diagnosis if hypokalemia and hypomagnesemia are detected.

EP-221 ENTEROBACTER RELATED INFECTIVE ENDOCARDITIS IN A PEDIATRIC PATIENT ON MAINTENANCE HEMODIALYSIS

Ruveyda Gulmez[1], Gulcin Unlu[2], Ayse Agbas[1], Reyhan Dedeoglu[3], Esra Karabag Yilmaz[1], Ebru Burcu Demirgan[1], Seha Saygili[1], Nur Canpolat[1]

[1]Department Of Pediatric Nephrology, CerrahpaŞa Faculty Of Medicine, Istanbul University-cerrahpaŞa, Istanbul, Turkey., [2]Department Of Pediatrics, CerrahpaŞa Faculty Of Medicine, Istanbul University-cerrahpaŞa, Istanbul, Turkey., [3]Department Of Pediatric Cardiology, CerrahpaŞa Faculty Of Medicine, Istanbul University-cerrahpaŞa, Istanbul, Turkey.

Introduction: Gram-negative organisms are a rare cause of infective endocarditis, but hemodialysis (HD) patients with central venous catheters are at increased risk for infective endocarditis due to bacteremia.
Material and methods: We present an adolescent patient who underwent chronic HD with an indwelling catheter and was diagnosed with Enterobacter-associated infective endocarditis.
Results: A 14-year-old girl on maintenance HD developed fever (40°C) during a dialysis session. She was receiving standard 4-hour HDF three times a week for 8 months. She had an indwelling catheter in her right internal jugular vein that had been placed 5 months earlier. She had no cardiac abnormalities or history of rheumatic fever. On physical examination, she was cachectic and looked ill. Her heart rate was 130 beats/minute with a 3/6 murmur in the mesocardiac area. There were no obvious foci of infection, rashes, or petechiae. The catheter exit site was clean. Laboratory tests revealed leukocytosis (14600/μL), neutrophilia (12700/μL), thrombocytopenia (124000/μL), elevated CRP (183 mg/L) and procalcitonin (>100 μg/L). A blood culture was obtained and treatment with teicoplanin was initiated with a suspicion of catheter-related bacteriemia. On the second day, meropenem was added to treatment because of positive signals for a gram-negative microorganism from the blood culture. Transthoracic echocardiogram showed a vegetation below the posterior leaflet of the tricuspid valve (20*10 mm). Enterobacter was detected in blood cultures (peripheral veins and HD catheter). Treatment was changed as vancomycin, meropenem and colistin. Her HD catheter was removed, and a left femoral catheter was placed. Her fever continued for fourteen days, but blood cultures remained negative. By the sixth week of treatment, vegetation regressed to 5x5 mm.
Conclusions: Infective endocarditis should be considered in HD patients with persistent fever. Enterobacter-related infective endocarditis is associated with high morbidity and mortality; however, early diagnosis and appropriate treatment ensure a favorable outcome.

EP-222 PHEOCHROMOCYTOMA AND CEREBELLAR TONSILLER HERNIATION IN AN ADOLESCENT

Emre LeventoĞlu[1], Bahar BÜyÜkkaragÖz[1], Bahriye Uzun Kenan[1], Arzu Okur[3], Esra DÖĞer[2], Sevcan A. BakkaloĞlu[1]

[1]Gazi University Faculty Of Medicine Pediatric Nephrology, [2]Gazi University Faculty Of Medicine Pediatric Endocrinology, [3]Gazi University Faculty Of Medicine Pediatric Oncology

Introduction: Pheochromocytoma is a rare catecholamine-secreting tumor. Frequent symptoms include hypertension (60-90%), headache (67%), followed by nausea, sweating, palpitations, and flushing. And neurologic complications such as hypertensive encephalopathy or herniation can be seen due to severe hypertension.
Material and methods: We present a patient with pheochromocytoma who developed a reversible tonsiller herniation.
Results: A 15-year-old girl was admitted to the hospital with neck pain and sudden onset of severe headache. Intermittent sweating attacks had been present. Her blood pressure was 180/110 mmHg (>99th percentile) and tendency to sleep was noted. Kidney function tests, serum electrolytes and glucose, blood gas analysis, thyroid function tests, renin-aldosterone levels and urinalysis were normal. Cranial computerized tomography (CT) demonstrated herniation in bilateral cerebellar tonsils. Intravenous sodium nitroprusside was started. Then, enalapril and amlodipine were added.
Excretion of normetanephrine were significantly increased as 10952 μg/24h (0-549 μg/24h)]. In abdominal CT a mass lesion with 76x53x68 mm diameter, compressing the right kidney anteriorly was detected in the right adrenal gland. Positron emission tomography (PET) revealed pathologically increased gallium Ga-68 DOTATATE uptake in the 5.5x5 cm mass located in the right adrenal gland.
Doxazosin was added to the treatment after previous antihypertensives. Esmolol infusion was implemented after alpha-blockade. After a short-term infusion of esmolol, it switched to metoprolol. On the 27th day of hospitalization, the patient was operated upon providing optimal blood pressure regulation. The need for antihypertensive treatment decreased as early as four days after the operation, and metoprolol treatment was discontinued primarily. All antihypertensive doses were gradually reduced and discontinued within one month. Control cranial MRI was normal at the post-operative third month.
Conclusions: Pheochromocytoma should be considered in the etiology in patients presenting with severe systemic hypertension. It should also be remembered that although very rare, hypertension may be associated with increased intracranial pressure by increasing cerebral or cerebellar perfusion, causing tonsillar herniation and accordingly inducing symptoms like neck pain and vomiting.

EP-223 PSEUDO-BARTTER SYNDROME AND STAGHORN CALCULI IN AN INFANT WITH TUFTING ENTEROPATHY

Emre LeventoĞlu[1], Bahar BÜyÜkkaragÖz[1], Demet Teker DÜztaŞ[2], ÖdÜl EĞritaŞ GÜrkan[2]

[1]Gazi University Faculty Of Medicine Pediatric Nephrology, [2]Gazi University Faculty Of Medicine Pediatric Gastroenterology

Introduction: Tufting enteropathy is rare genetic disorders that occur in the first weeks of life and are characterized by chronic and severe diarrhea. It can increase the frequency of nephrolithiasis due to malabsorption as well as chronic dehydration. Also, chloride deficiency secondary to chronic severe diarrhea causes pseudo-Bartter syndrome.
Material and methods: We present a patient with tufting enteropathy who followed-up as pseudo-Bartter syndrome and developed staghorn calculi in kidney.
Results: 8-month-old girl presented to hospital for poor feeding, diarrhea and failure to thrive which started right after birth. At presentation, body weight was 4700 g (<3rd percentile). Laboratory results showed metabolic alkalosis (pH: 7.54, bicarbonate: 29.7mmol/L), hyponatremia, hypokalemia and hypochloremia. Hyperreninemic hyperaldosteronism was present. Urine output was in the upper range. There was hyperoxaluria and hypocitraturia. Stool electrolyte measurements showed increasing in chloride. In the abdominal ultrasonography, echogenicities consistent with the stones filling the whole pelvicalyceal systems were noted in the middle and lower parts of the both kidneys. Considering the possibility of Bartter syndrome initially, indometacin was started. Pyridoxine and potassium citrate were added to the patients treatment.
Severe diarrhea persisted in the follow-up. Indomethacin was discontinued at the end of three months of use considering the clinical picture of pseudo-Bartter syndrome. Genetic analysis demonstrated c.163T>C p.Cys55Arg homozygous mutation in epithelial cell adhesion molecule (EpCAM) gene. The definite diagnosis of congenital tufting enteropathy was made.
Staghorn calculi persisted in the follow-up, however, she did not have any urinary tract infections, urinary outflow obstruction or progression into chronic kidney disease. At the age of 3 years of age, she still had significant growth and developmental retardation and worsening in the fluid-electrolyte balance due to severe diarrhea. She was commenced on permanent total parenteral nutrition (TPN). However, she developed catheter-related sepsis on the 3rd month of TPN, and died on the 21st day of hospitalization in the intensive care unit.
Conclusions: Clinical and biochemical picture of severe diarrhea may be very similar to Bartter syndrome. It should also be remembered that nephrolithiasis or staghorn calculi may develop even in infancy due to chronic diarrhea syndromes due to malabsorption or dehydration on a pathophysiological basis.

EP-224 RAPIDLY PROGRESSIVE PEDIATRIC PULMONARY-RENAL SYNDROME OF ILL-DEFINED ETIOLOGY AND PROMPT RESPONSE TO IMMUNOSUPPRESSIVE TREATMENT

Vanja Ille Matić[1], Nastasia Kifer[2], Filip Rubić[1], Toni Matić[1], Sandro Dessardo[1], Miran Cvitković[1], Slobodan Galić[1], Kristina Vrljičak[3], Ivanka Kos[3], Lovro Lamot[3]

[1]Intensive Care Unit, Department Of Pediatrics, University Hospital Center Zagreb, Croatia, [2]Division Of Pediatric And Adolescent Rheumatology, Department Of Pediatrics, University Hospital Center Zagreb, Croatia, [3]Division Of Nephrology, Dialysis And Transplantation, Department Of Pediatrics, University Hospital Center Zagreb, Croatia

Introduction: Pulmonary-renal syndrome (PRS) refers to the combination of acute kidney injury (AKI) caused by rapidly progressive glomerulonephritis, and lung involvement with severe respiratory failure, which often involves diffuse alveolar haemorrhage. It implies different rare conditions such as various forms of systemic vasculitis, Goodpasteur syndrome and systemic lupus erythematosus. Nevertheless, nonspecific conditions without vasculitis can also cause PRS.
Material and methods: We describe a child with unknown cause of PRS who was successfully treated with immunosuppressive therapy.
Results: A 16-year-old girl presented with abdominal pain, vomiting, high fever, cervical lymphadenopathy and splenomegaly. Initially, her laboratory findings indicated systemic inflammation, with increased inflammatory markers and transaminases. Urinalysis was positive for leukocytes, erythrocytes, nitrites and proteins with active sediment and significant number of E. coli in microbiology. Subsequently, nephrotic range proteinuria and AKI were observed. Seven days later she developed dyspnoea with respiratory insufficiency necessitating mechanical ventilation. Computed lung tomography revealed ground-glass opacities and crazy paving pattern. The broad differential diagnosis of PRS was considered, with alternative complement pathway activation, negative ANCA, anti-GBM and ASO titre. Renal biopsy was suggestive of possible infectious cause, while microbiology revealed high circulating EBV DNA load with positive EBV serology. Steroid pulse therapy led to prompt improvement of lung and kidney function, with further amelioration and no relapses during the six months follow-up.
Conclusions: Even though PRS is mostly caused by specific well defined immunological disorders, unknown and atypical causes can also lead to this life-threatening condition. Therefore, early clinical recognition and prompt immunosuppressive treatment are of greater importance than defining the cause which might remain inconclusive long after the full remission has been achieved.

EP-225 EVALUATION OF CAKUT IN CHILDREN OF A SINGLE CENTER

Ganna Zvenigorodska[1], Oleksandr Moskaliuk[1], Galina Guminska[2]

[1]National Pirogov Memorial Medical University, [2]Vinnytsya Regional Clinical Childrens Hospital

Introduction: Half of all children with end-stage renal disease were born with malformed renal tract. Congenital anomalies of kidneys and the urinary tract (CAKUT) form the main cause of CKD in children. Around 5% of adults with ESRD were born with CAKUT. Aim: To determine the structure of the CAKUT-syndrome in children at Vinnytsya Regional Childrens Clinical Hospital, Ukraine in 2018-2020 y.
Material and methods: A retrospective cohort study was conducted using medical chart data from patients primarily treated in a single center in Ukraine during 2018 – 2020 years. Clinical, biochemical and radiological data were obtained.
Results: CAKUT-syndrome was diagnosed in 610 children in Vinnytsya Regional Childrens Clinical Hospital during 2018 – 2020 years. The initial presentation of CAKUT-syndrome was in 112 patients (18.3%) and included signs of urinary tract infections and structural changes of kidneys. Most patients were boys. High number of CAKUT-syndrome was diagnosed prenatally. Less than 10% of all CAKUT were diagnosed in adolescence. Most of them presented with isolated changes in urinalysis (74%), rarely - with arterial hypertension. Among all CAKUT-syndrome in 2018, half of all cases belonged to congenital hydronephrosis, VUR, obstructed megaureter). In subsequent years 2019 and 2020, these defects predominated in the structure of the congenital anomalies, (45.3% and 42.3% respectively). CAKUT-syndrome including ectopia of the kidney, horseshoe kidney met with different frequency in 2018 (45 children), 2019 (37 children) and 2020 (32 children). The last place in the structure of the CAKUT-syndrome belonged to epispadia, diverticulum of the bladder, strictures - 5.3% in 2018, 3.3% in 2019, and 4.5% in 2020.
Conclusions: Number of children with CAKUT-syndrome increased each year and may form a CKD in future. Early diagnosis, adequate surgical treatment, using renoprotective therapy prevents the development of CKD in children with CAKUT-syndrome.

EP-226 VESICOURETERAL REFLUX AND RENAL SCARRING - 18 YEARS OF EXPERIENCE

Beatriz Vieira[1], Sofia Branco[1], Inês Mazeda[1], Sara Catarino[2], Ana Luísa Santos[1], Célia Madalena[1]

[1]Centro Hospitalar Da Póvoa De Varzim/vila Do Conde, Póvoa De Varzim, Portugal, [2]Centro Materno Pediátrico, Chusj, Porto, Portugal

Introduction: Vesicoureteral reflux (VUR) is the commonest urological abnormality in children and has been associated with an increased risk of urinary tract infection (UTI) and reflux nephropathy (RN). In children, RN is diagnosed mostly after UTI or during follow-up for antenatally diagnosed congenital anomalies of kidney and urinary tract (CAKUT).
Objective: To analyse the characteristics of a pediatric population with RN.
Material and methods: This is a retrospective study of children with RN, diagnosed after an UTI (group 1) or during follow-up of CAKUT (group 2), followed in our hospital between 2004-2021. Renal scars were diagnosed by dimercaptosuccinic acid (DMSA) renal scan. Statistical analysis were performed using SPSS®. A P-value of less than 0.05 was accepted as significant.
Results: Group 1 included 140 children (201 units) with VUR of which 51 had RN (36,4%). Group 2 included 39 children (58 units) with VUR of which 18 had RN (46,2%). In children with RN, there was a female predominance (67%) in group 1 and a male predominance (66,7%) in group 2. In group 1, the incidence of renal scarring was 29.9% in children with the first UTI before age 2, 14.9% between ages 2 and 4 and 13.8% in children over 5 years of age. There was a significative association between recurrent UTI and RN in both groups. Furthermore, the risk of renal scarring seemed to be increased at children with constipation. In both groups, there was a significant association between VUR severity and the presence of renal scarring (RN in group 1: IV-V: 100%, I-III: 23,4%; in group 2: IV-V: 84,6%, I-III: 40%).
Conclusions: Higher grade for VUR and higher number of UTI proved to be determinant for renal scarring. The challenge is to identify the children with higher VUR and at risk of recurrent UTIs in order to prevent future kidney damage.

EP-227 VERY EARLY ONSET OF SYSTEMIC LUPUS ERYTHEMATOSUS

Samantha Innocenti, Silvia Bernardi, Liloie De La Guillonniere, Maud Prevot, Camille Menvielle, Olivia Boyer

Pediatric Nephrology Department, Marhea Reference Center, Ap-hp, Hôpital Universitaire Necker-enfants Malades, Paris, France

Introduction: Childhood-onset systemic lupus erythematosus(cSLE) is rare, especially before five years of age. Symptoms are commonly overlooked,despite the severity of certain clinical manifestations.
Material and methods: A three-year-old Guianese girl, born to unrelated parents, presented with macrohematuria. No family history of renal nor rheumatological diseases. She had presented scarlet fever two months earlier and upper respiratory tract infections.
Results: Clinical examination was unremarkable. Laboratory tests showed nephrotic syndrome, brown macrohematuria, normal GFR and ultrasound. Initial suspicion of post-streptococcal glomerulonephritis(PSGN) was sustained by the hypocomplementemia. However, atypical presentation emerged with non-hemolytic anemia,positive direct Coombs test,thrombocytopenia, hypergammaglobulinemia and high ESR:CRP ratio. Active EBV replication and IgG anti-SARS-Cov2 were found. Subsequently a malar rash, palatal petechiae and oedema appeared with rapid worsening of anemia and thrombocytopenia. Differential diagnosis focused on hematological malignancies,viral infections or autoimmune diseases. Bone-marrow aspiration and immunophenotyping excluded malignancies or EBV-induced cytopenia; immune-deficiency and SARS-Cov2 associated multisystemic-inflammatory-syndrome were ruled-out. Autoimmune screen revealed high ANA, anti-dsDNA, ENA and p-ANCA titer with anti-C1q, anti-platelet and anti-beta2GP1 antibodies associated with LAC positivity, but no anti-FB antibodies.Thus, cSLE was highly plausible, kidney biopsy was mandatory but postponed due to the hemorrhage risk. Thereafter, normalisation of CBC with three methylprednisolone boluses and oral prednisone,mycophenolate and hydroxychloroquine,enabled it.Histologically, class V lupus nephritis with NIH-score of 0/24 for activity and 0/12 for chronicity was observed. Considering the very early-onset and suspected monogenic cSLE, genetic study is ongoing.
Conclusions: PSGN is the leading cause of acute nephritis in children.Kidney biopsy is not required in typical cases with acute nephritic syndrome and C3 hypocomplementemia. However, red-flags such as hypocomplementemia persistence, rapidly progressive glomerulonephritis or extra-renal manifestations, indicate a kidney biopsy to identify conditions associated with a negative renal outcome and requiring prompt appropriate treatment like C3-glomerulopathy, membranoproliferative glomerulonephritis or cSLE.

EP-228 PHASE 3, RANDOMIZED, MULTICENTER STUDY TO EVALUATE THE EFFICACY AND SAFETY OF PEGCETACOPLAN IN TREATMENT OF C3G OR IC-MPGN

Marina Vivarelli[1], Bradley P. Dixon[2], Fadi Fakhouri[3], Matthew C. Pickering[4], Terence Cook[4], David Kavanagh[5], Giuseppe Remuzzi[6], Patrick Walker[7], Christoph Licht[8], Gerald Appel[9], Zhiqun Zhang[10], Li Li[11], Hetal Kocinsky[11]

[1]Division Of Nephrology And Dialysis, Bambino Gesù Pediatric Hospital, Rome, Italy, [2]Department Of Pediatrics, University Of Colorado School Of Medicine, Aurora, CO, USA, [3]Lausanne University Hospital And University Of Lausanne, Lausanne, Switzerland, [4]Imperial College, London, Uk, [5]National Renal Complement Therapeutics Centre, Newcastle Upon Tyne, Uk, [6]Mario Negri Institute For Pharmacological Research, Milan, Italy, [7]Arkana Laboratories, Little Rock, Ar, USA, [8]The Hospital For Sick Children, Toronto, On, Canada, [9]Columbia University, New York, NY, USA, [10]Former Employee Of Apellis Pharmaceuticals, Waltham, MA, USA, [11]Apellis Pharmaceuticals, Waltham, MA, USA

Introduction: Complement 3 glomerulopathy (C3G) and immune complex membranoproliferative glomerulonephritis (IC-MPGN) are rare diseases characterized by excessive deposition of C3 breakdown products in renal glomeruli leading to proteinuria and progressive renal disease. Pegcetacoplan is a targeted C3 investigational therapy for diseases related to complement overactivation. This is a phase 3, randomized, placebo-controlled, double-blind, multicenter study of the efficacy and safety of pegcetacoplan in individuals with C3G or IC-MPGN.
Material and methods: Approximately 90 patients (age, ≥12 years; weight, 20-100 kg) diagnosed with C3G or IC-MPGN, either as primary disease or posttransplant disease recurrence, will be recruited. Inclusion criteria include 2+ staining for C3c, global glomerulosclerosis <50%, urine protein-to-creatinine ratio (uPCR) ≥1000 mg/g, and estimated glomerular filtration rate (eGFR) >30 mL/min/1.73 m^2. Exclusion criteria include previous pegcetacoplan exposure, C3G/IC-MPGN secondary to other conditions, and significant infection/malignancy. Patients will be randomized 1:1 to receive subcutaneous infusions of pegcetacoplan (1080 mg/20 mL) or matching volume of placebo twice weekly for 26 weeks (in addition to standard care). Thereafter, in the open-label period, all participants will receive pegcetacoplan twice weekly for 26 weeks. Assessments include first-morning uPCR every 4 weeks and renal

biopsies at baseline/screening and weeks 26 and 52. Primary endpoint is proportion of participants with reduction in uPCR ≥50% relative to baseline at week 26. Secondary endpoints include proportion of participants with eGFR scores that are stable or improved from baseline; change in C3G histologic index activity score; and proportion of participants with decreased C3c staining on renal biopsy from baseline at week 26. Safety outcomes will also be monitored throughout the study. Participants may enter a subsequent 8-week follow-up period or long-term extension study.
Results: This is a study design abstract.
Conclusions: This study will evaluate the safety and efficacy of complement protein C3 inhibitor pegcetacoplan in treating C3G and IC-MPGN.

EP-229 THE LATE DIAGNOSED CASE OF FANCONIS SYNDROME ASSOCIATED WITH A MUTATION IN THE HNF4A GENE.

Zaikova Natalia, Marina Aksenova, Vladimir Dlin, Elena Toszliyan

Pirogov University, Moscow

Introduction: Renal Fanconis syndrome (FS) comprises a heterogeneous group of disorders characterized by generalized proximal tubular dysfunction. Early diagnosis may facilitate therapy in some cases.
Material and methods: The aim of the study was to report the late diagnosed case of FS associated with a mutation in the HNF4A gene.
Results: Case report. A 6-years girl from a unrelated healthy parents was presented in our department with polyuria (3-5 l/m2/day), inconsistent hyperglycemia (6.7-4.6 mmol/l), hypophosphatemia (0.89 mmol/l), metabolic acidosis (pH - 7,29, HCO3 - 25 mmol/l), low molecular weight proteinuria (b2 – 35408 mcg/day), proteinuria (0.317-0.543 g/day), albuminuria (80 mg/l), glycosuria (69.1-72.4 mmol/day), hyperphosphaturia (TmP 0.41-0.59); eGFR - 58.6 - 60.1 ml/min/1.73 m2; systemic osteoporosis, increased blood levels of AP (1932 IU/l), PTH (75.4 pg /ml), Ca/Cr (2.1-1.4 mmol/mmol). The blood activity of the 1,4a-glucosidase and cysteine's concentration in leukocytes were normal.She did not have the signs of liver dysfunction. Ultrasound revealed medullar nephrocalcinosis gr.1. Whole exome sequencing revealed heterozygous pathogenic variant in HNF4A gene (transcript NM_000457.4, cDNA s.253C>T, AA replacement of the R.g85Trp r.). The therapy with sodium bicarbonate 4% (2 mmol/kg/day), Alfacalcidol 0.25 mcg/day (0,14 mcg/kg /day), Reducto special 55 mg/kg/day, enalapril - 0.15 mg/kg /day.
Conclusions: FS type 4 should be suspected in children with macrosomia, postprandial hyperglycemia, liver dysfunction since early diagnosis and treatment are important in improving the quality of life and reducing complication. Given the high risk of early development of diabetes in patients with HNF4A gene mutation, constant monitoring of glycemia is necessary.

EP-230 SURVIVAL OF PATIENTS WITH FOCAL SEGMENTAL GLOMERULOSCLEROSIS: A TUNISIAN SERIES

Abir Boussetta, Khouloud Ben Njima, Nesrine Abida, Farah Krifi, Manel Jellouli, Tahar Gargah

Pediatric Nephrology Department, Charles Nicolle Hospital Tunis, Tunisia

Introduction: To study the global survival and the renal survival in children with Focal Segmental Glomerulosclerosis (FSGS)
Material and methods: This was a retrospective study conducted in the pediatric nephrology department of Charles Nicolle hospital, Tunis, over a period of 20 years, from January 2001 to December 2020. Inclusion criteria were: patients less than 18 years of age at the time of diagnosis of the disease, regularly followed up with renal biopsy-proven FSGS. Exclusion criteria were: children with suspected or proven secondary FSGS. For the purpose of studying survival, the Kaplan-Meier nonparametric estimation was used.
Results: A total of 35 patients were included in our study, the median age at diagnosis of the FSGS was 4.5 years. The mean survival of our patients was 14.7 years. The global survival at 1 and 3 years was 100% and at 5 years was 75%. The average renal survival of our patients was 13 years, renal survival was estimated at 100% at 1 and 3 years, 85% at 5 years and 73% at 10 years. At five years, renal survival in the 1–3-year, 3–5-year, 5-10 year and >10-year age groups were 88%, 100%, 75% and 67%, respectively. This association was not statistically significant (p=0.105). Children with hypertension at diagnosis had a 100% renal survival rate at 5 years compared to 90% for children without initial hypertension (p=0.610). Patients with the tip-lesion variant had the best survival at 5 years (100%) followed by those with the NOS type (80%). Patients treated with Ciclosporin A had a mean of 13.12 years survival, those treated with Cyclophosphamide had a 11.17-year survival, and those treated with MMF had a mean of 13.62 years survival. The difference in survival rates was not statistically significant.
Conclusions: FSGS is less good prognosis than other variants of nephrotic syndrome in children.

EP-231 DIABETES FOLLOWING PEDIATRIC KIDNEY TRANSPLANTATION: A TUNISIAN SERIES

Abir Boussetta[1], Farah Krifi[1], Nesrine Abida[1], Taieb Ben Abdallah[2], Manel Jellouli[1], Tahar Gargah[1]

[1]Pediatric Nephrology Department, Charles Nicolle Hospital Tunis, Tunisia, [2]Research Unit Of Immunopathology And Immunology Of Renal Transplantation Lr03sp01

Introduction: To describe the clinical and evolutive features of post-renal transplant diabetes in Tunisian children.
Material and methods: This was a cross-sectional, descriptive, retrospective study carried out in the pediatric and internal medicine A departments of Charles Nicolle hospital in Tunis. Transplanted patients of age less than or equal to 20 years were included during a 31-year period from January 1989 to December 2019. Patients who developed diabetes after renal transplantation were included in our study.
Results: A total of 115 transplanted were including in our study during this period (69 boys and 46 girls). The average age was 15.5 years old. The transplantation was done from a living donor in in 67.8 % of cases. Immunosuppression protocols were based on: anti-lymphocyte serum prescribed in all cases, anticalcineurin (ciclosporin in 52 patients and tacrolimus in 48 patients), corticosteroid therapy prescribed in all patients, Mycophenolic acid (MMF) in 79 patients (68.7%). Azathioprine was prescribed in 30 patients who were transplanted before 2000. The immunosuppressive treatment was adjusted according to the function of the graft and the residual levels of the different molecules. Diabetes was noted in 8.6% of patients 10 cases) after kidney transplantation. All these children were on tacrolimus before the onset of diabetes. Patients were put on oral antidiabetics in 3 cases and on insulin therapy in 7 cases. The outcome was favorable in 30% of cases with transient diabetes. No degenerative complications following diabetes were noted in our series
Conclusions: Diabetes is the most frequent metabolic complication in post pediatric kidney transplantation. It can be induced by immunosuppressive therapy, hence the importance of strict clinical and biological monitoring of these patients.

EP-232 A RARE SIDE EFFECT OF A FREQUENT MEDICINE IN A CHILD: QUARTZ STONE

Emre LeventoĞlu[1], Selin Kuzucu[2], Kibriya Fidan[1], OĞuz SÖylemezoĞlu[1]

[1]Gazi University, Faculty Of Medicine, Department Of Pediatric Nephrology, [2]Gazi University, Faculty Of Medicine, Department Of Pediatrics

Introduction: Urolithiasis is a non-malignant condition that can affect any part of the urinary tract. Kidney stone formation is affected by factors such as climate, dietary habits, drugs, occupation, fluid intake, genetic predisposition, urinary tract infections and malformations in the urinary tract.
Material and methods: With this case report, the surprising reason for the formation of silicate stone which is extremely rare was explained.
Results: A 7-year-old female patient who had a bone marrow transplant due to aplastic anemia two years ago was admitted to hospital with the complaint of stone in the urine. The patient with left flank pain noticed a 0.4x0.5mm gray-beige stone in her urine.
Upon presentation, her physical examination was normal. Laboratory examination revealed normal kidney function test and blood gas analysis. For urine; tubular phosphate reabsorption was 95.8%, fractional excretion of sodium was 0.2%; urine protein/creatinine was 0.08 mg/mg, calcium/creatinine was 0.01 mg/mg, oxalate/creatinin 0.01 mg/mg, citrate/creatinin 0.35 mg/mg. Urine density was measured as 1007, and urine pH was measured in fresh urine by dipstick as 6. The urine output was 3.6 ml/kg/hour. Abdominal ultrasonography was normal. She was using only ursodeoxycholic acid as a drug for 18 months because of post-transplant hyperbilirubinemia.
In the stone analysis performed by X-ray diffraction method, dolomite ($CaMg(CO_3)_2$) mineral accompanied by calcium was observed in addition to the quartz mineral (SiO_2). The ursodeoxycholic acid tablet form that the patient was using was changed to the syrup form which is silicon dioxide free. Also hydration and a salt-restricted diet are recommended in the treatment and no new stone formation was observed in the follow-up.
Conclusions: The fact that the disease is very rare causes curiosity on this subject. The importance of performing stone analysis as short as possible becomes clear once again. When prescribing drugs to the patient, not only the side effects of the active substance should be taken into consideration, but also the possible side effects of the excipients in the drug content should be evaluated.

EP-233 NEUROFIBROMATOSIS TYPE 1 AND SYSTEMIC VASCULOPATHIES

Emre LeventoĞlu[1], Kibriya Fidan[1], Bahar BÜyÜkkaragÖz[1], Esra SerdaroĞlu[2], Tuba Atalay[3], Merve Yazol[4], OĞuz SÖylemezoĞlu[1]

[1]Gazi University, Faculty Of Medicine, Department Of Pediatric Nephrology, [2]Gazi University, Faculty Of Medicine, Department Of Pediatric Neurology, [3]Gazi University, Faculty Of Medicine, Department Of Ophthalmology, [4]Gazi University, Faculty Of Medicine, Department Of Radiology

Introduction: Neurofibromatosis type 1 (NF1) is an autosomal dominant disease caused by a mutation in NF1 gene. In NF1, a wide range of pathological conditions such as vascular aneurysms, stenosis, or arteriovenous malformations can be seen.
Material and methods: We present an adolescent with NF1 who developed various vascular pathologies.
Results: A 4-year-old boy was admitted to hospital due to hyperpigmented macules in 2008. Lish nodules were detected. The p.R304* pathogenic mutation (also known as c.910C>T), located in coding exon 9 of the NF1 gene, was detected in the genetic examination of NF1. Patient was diagnoses as neurofibromatosis type 1 (NF1).
After a year, his blood pressure (BP) was 170/100 mmHg. Plasma renin activity was elevated, >500 (1-6.5 ng/mL/hr). Kidney doppler ultrasound showed bilateral renal arterial stenosis. Amlodipine and propranolol was initiated. Percutaneous transluminal renal angioplasty was performed; contour irregularity and minimal stenosis in the the ostium of right renal artery, perivascular diaphragmatic collateral arteries at the level of left renal artery orifice were observed. Coronary balloon catheter was used for right renal artery. For left renal artery, surgery was performed, occluded segment was removed and anastomosis was made. Therefore, BP returned to normal levels. Antihypertensive drugs were discontinued.
After a few years, BP became moderately high. Ophthalmic examination showed tiny veins twisted into spiral shape and corkscrew retinal vessels. Antihypertensive treatment was initiated as amlodipine and metoprolol. In 2021, Conventional angiography was performed due to hypertension. Abdominal aorta was low in caliber. The celiac truncus was markedly dilated. It was occluded at the ostium of the superior mesenteric artery and was filled from celiac truncus via the dilated pancreaticoduodenal arch. In the right carotid system, the communicating segment of internal carotid artery was obstructed. The anterior cerebral artery A1 segment was hypoplastic. There was an aneurysm of 2 mm in size on the posterior wall of the ophthalmic segment of left internal carotid artery. Because of cerebral vasculopathy, close follow-up was planned for cerebral ischemia risk.
Conclusions: NF1 is a systemic disease, micro and macrovascular complications should always be taken into consideration, and the necessity of targeting early detection and symptomatic treatment of emerging complications.

EP-234 INVASIVE RENAL FUNGAL INFECTIONS IN PRETERM- REPORT ON 3 CASES

Stroescu Ramona[1], Gafencu Mihai[1], David Vlad[1], ChiŞavu Flavia[2], Steflea Ruxandra[1], Doros Gabriela[1]

[1]University Of Medicine And Pharmacy "victor BabeŞ" TimiŞoara, România, [2]"louis Ţurcanu" Emergency Hospital For Children, TimiŞoara, România

Introduction: Preterm newborns have immature immune systems; there is a reduced production of cytokines which limits T cell activation that explain the increased risk of infection. Invasive fungal infections in preterm is more common and can lead to sever multiorgan affection with poor outcome.
Material and methods: We present 3 cases with atypical urinary fungal infections.
Results: Case 1: Triplet II born at 28 weeks referred from the Neonatal unit with clinical picture of acute abdomen at the age of 2 weeks. Imaging was inconclusive therefore he proceeded to an exploratory laparotomy, where a diagnosis of urinary ascites was made. Candida albicans infection was confirmed on urine culture. He was treated with antifungals and made a full recovery. Case 2: Triplet I presented to ED one month later with symptoms of respiratory infection. Commenced on empirical antibiotic medications but clinically deteriorated. Urine culture was positive for fungal infection so antifungal medication was commenced as well. Ultrasound scan demonstrated a right perinephric urinoma. This was drained percutaneously. Case 3: Preterm who received antibiotherapy for 3 weeks and developed fungal pyelonephritis with typical image on ultrasound, urine culture was negative but also with a good response after antifungal therapy.
Conclusions: Fungal ball formation in urinary tract can cause obstruction leading to extravasation. Extravasation of urine and formation of urinoma is rare. Invasive fungal infections in preterm are common and extremely difficult to diagnose. Urinary obstruction due to fungal ball is a rare and atypically that shall be diagnosed and treated immediately. Empirical

treatment with antifungal therapy should be considered in high-risk, low-birth-weight infants who fail to quickly respond to empirical antibacterial treatment. Risk factors to consider when deciding to administer empirical antifungal therapy include: prior exposure to antibiotics, extreme prematurity, long term hospitalisation

EP-235 A CHILD WITH ABDOMINAL PAIN AND ANURIA

Zehra Aydin[1], Yusuf Kenan Cetinoglu[2], Fatma Semsa Cayci[3], *Sare Gulfem Ozlu*[3]

[1]Batman Training And Research Hospital, Department Of Nephrology, Batman, Turkey, [2]Batman Training And Research Hospital, Department Of Radiology, Batman, Turkey, [3]Ankara City Hospital, Department Of Pediatric Nephrology, Ankara, Turkey

Introduction: The imperforate hymen is an unusual obstructive congenital anomaly that results from the lack of resorption of the hymen membrane [1]. It can cause hematocolpos that is the retention of menstrual blood from hymen to the cervix.The accumulation of blood with vaginal distension leads to mechanical pressure on the urethra, bladder, uterus, and rectum.
Material and methods: Here we report a girl with imperforate hymen and presented with abdominal pain and anuria.
Results: A 13 years old girl was admitted to our pediatric nephrology department with abdominal pain and anuria. She had abdominal pain four days ago, and she noticed decreased urine output two days after the first complaint, so they applied to the emergency department of a local hospital. After urinary catheterization, she had sufficient urinary output, abdominal pain transiently improved, and she was discharged to home. Because her abdominal pain gradually increased for the following two days and did not respond to painkillers, and she had no urine output for the last twelve hours, she was admitted to our hospital.A detailed genital, rectal examination was performed. Rectal examination was normal. On examination of the external genitalia, pink-colored protruding imperforate hymen was identified. After the genital examination, ultrasonography was performed as the initial diagnostic modality [4]. Transabdominal ultrasound showed a suprapubic homogeneous and urine-filled bladder with right-sided moderate hydronephrosis. The uterus was also distended.She was consulted with a gynecologist and, the diagnosis of the imperforate hymen was confirmed. Hymenectomy was planned as a curative treatment.
Conclusions: Imperforate hymen should be kept in mind in peripubertal female patients with acute urine retention, abdominal pain, abdominal distension.

EP-236 HISTOLOGICAL CHARACTERISTICS OF LUPUS NEPHRITIS IN TUNISIAN CHILDREN

Abir Boussetta, Delia Louati, Aicha Turki, Farah Krifi, Manel Jellouli, Tahar Gargah

Pediatric Nephrology Department, Charles Nicolle Hospital Tunis, Tunisia

Introduction: To describe the main histological characteristics of lupus nephritis (LN) in Tunisian children
Material and methods: This was a descriptive, retrospective, multicenter study conducted in five departments of pediatric and nephrology in Tunisia. Patients with systemic lupus erythematosus (SLE) less than 18 years of age at the time of renal biopsy (RB), were included in our study. Exclusion criteria were: children with SLE who do not have RB that proven LN. Our study was conducted over 28 years, from January 01, 1990 to December 31, 2018. The histological classification of lupus nephropathy was done according to the International Society of Nephrology/Renal Pathology Society 2003.
Results: All our patients had an initial RB. Sixteen patients (40%) had a second one and one patient had a third one. Class IV LN was found in 40% of cases, class IV+V was found in 20% of cases, class IV+III in 12.5% of cases. Class III was found in 15% of cases, class II in 10% of cases and class VI in one case. The mean number of glomeruli was 25.6 (10-100), vascular involvement was specified in 35 patients (87.5%), thrombotic microangiopathy was noted in 6 cases (17.1%). Activity signs were present in 80% of cases, presence of hyaline deposits was the most frequent sign (62.8%), followed by fibrinoid necrosis in 48.5% of cases. Chronicity signs were present in 14 patients (40%) and included glomerular sclerosis in 64.2% of cases, and interstitial fibrosis in 57.1%.
Conclusions: The histological features in LN are important to identify. Although there is no correlation between clinical and histological findings, they have a definite prognostic value.

EP-237 INCIDENCE AND OUTCOMES OF CHILDHOOD HEMOLYTIC-UREMIC SYNDROME IN BELARUS IN 2021

S.v. Baiko[1], E.o. Samoilovich[2], G.v. Semeiko[2], M.d. Charadnichenka[1]

[1]Belarusian State Medical University, Minsk, Belarus, [2]Republican Research And Practical Center For Epidemiology And Microbiology, Minsk, Belarus

Introduction: The annual incidence of hemolytic uremic syndrome (HUS) in Belarus for the period 2015-2019 was 5,0 and 2,3 cases per 100,000 children aged <5 and <15 years with a significant increase in 2021.
Material and methods: Of the 77 children diagnosed with HUS 64 were hospitalized in Minsk pediatric dialysis center and divided into 2 groups: 29 without (1) and 35 children during the outbreak (2). In 51 children underwent stool analysis for Shiga toxin-producing Escherichia coli by real-time PCR.
Results: The incidence of HUS in 2021 was 10,0 (56 cases) and 4,8 (77 cases) per 100,000 children aged <5 and <15 years. The boys were 52%, children <5 years old – 73,7%, patients with atypical HUS – 3,9%, requiring dialysis – 56,6%. During the outbreak (27.09.–29.10.2021) HUS was diagnosed in 44 children from 3 regions of the country: Minsk city – 17, Minsk region – 16, Vitebsk region – 11. Patients of groups 1 and 2 did not differ by age: 2,5 (1,6; 5,1) vs 3,6 (2,2; 5,1) years, frequency of hemocolitis: 62,1% vs 68,5%, initial levels of hemoglobin and platelets, need for dialysis: 79,3% vs 57,1% and mechanical ventilation: 20,7% vs 5,9%, duration of anuria 13 (7; 16) vs 12 (8; 15) days, mortality 3,4% (aHUS) vs 2,9% (child with COVID-19), the % of detection of Shiga toxin DNA: 33,3% vs 37,0%. CNS disorders were more often observed in the 1st group (24,1% vs 0%, p=0,003).
Conclusions: The incidence of HUS in children in Belarus remains one of the highest in Europe. Over the past 17 years of observation the first outbreak of HUS was recorded in 2021.

EP-238 24-MONTH KIDNEY OUTCOME IN AN INTERNATIONAL COHORT OF 382 CHILDREN AND ADOLESCENTS WITH LUPUS NEPHRITIS

Chiara De Mutiis[1], Kjell Tullus[2], International Lupus Nephritis Study Group In Children[2]

[1]Pediatric Department, Maggiore Hospital, Bologna, Italy, [2]Renal Unit, Great Ormond Street Hospital For Children Nhs Ft, London, Uk

Introduction: Children with lupus nephritis (LN) have higher risk for end stage renal disease and higher mortality compared with age-matched healthy children. We present kidney outcome of a large multi-national cohort of children with LN.
Material and methods: 382 patients (≤18 years old) with LN class ≥III diagnosed and treated in the last 10 years in 23 international centers were studied up to 24 months of follow-up. We defined complete remission as urine-protein-creatinine ratio ≤0.2 mg/mg and eGFR ≥90 ml/min/1.73m2 or serum creatinine increased less than 15%. Partial remission was defined as urine-protein-creatinine ratio <2 mg/mg or a 50% reduction if UPCR ≥2 at baseline and eGFR ≥90 ml/min/1.73m2 or serum creatinine increased less than 25%. Stable remission was considered as the persistence of complete remission from 6th months to 24th months of follow up and assessed in a subgroup of 351 patients.
Results: 57% and 34% of patients achieved complete and partial remission at 24-month follow-up, respectively. Only 25.4% maintained stable remission. The reasons why patients didn't achieved stable remission were eGFR <90 ml/min/1.73m2 in 16.8%-30.5% of cases, increased serum creatinine in 6.8%-13.1% and urine-protein-creatinine ratio >0.2 mg/mg in 29.3%-45.6%. Patients with biopsy class III achieved complete remission more often than other biopsy classes.
Complement 3, serum creatinine, urea and biopsy class at diagnosis were predictive parameters of stable renal remission.
No difference in achieving stable remission was found between children who received mycophenolate or cyclophosphamide as induction treatment.
Conclusions: In our cohort the rate of stable complete remission in patients with LN is still low. Severe kidney involvement at diagnosis was the most important risk factor for not achieving stable remission, while no difference were found between induction treatments. Randomized treatment trials in children with LN are needed to improve kidney outcome.

EP-239 URINARY SYSTEM INVOLVEMENT IN MUCOPOLYSACCHARIDOSIS DISEASE: TWO CASES WİTH CHRONİC KİDNEY DİSEASE

Serim Pul[1], Neslihan ÇiÇek[1], Özge GÜnal[2], Emel Yilmaz GÜmÜŞ[3], Cagri Akin Sekercİ[4], Ece Demirci Bodur[1], SerÇin GÜven[1], Özde Nisa TÜrkkan[1], SelÇuk YÜcel[4], Burcu ÖztÜrk HiŞmi[3], Nurdan Yildiz[1], İbrahim GÖkÇe[1], Harika Alpay[1]

[1]Marmara UnİversİTy, PedİatrİC Nephrology, [2]Marmara UnİversİTy, Pediatrics, [3]Marmara UnİversİTy, Pediatric Nutrition And Metabolism, [4]Marmara University, Urology

Introduction: Mucopolysaccharidosis(MPS) is a rare inherited metabolic disease group that leads to many organ and system dysfunctions as a result of excessive glycosaminoglycan(GAG) accumulation in lysosomes.Although the musculoskeletal, cardiopulmonary, gastrointestinal and central nervous system can be affected frequently in MPS, urinary system involvement is rare.In this report, two MPS cases with chronic kidney disease(CKD) are presented.
Results: Case-1: An eleven-year-old male was diagnosed as MPS type-2 eight years ago.He was consulted to the nephrology clinic for decreased urine output.His serum creatinine, electrolytes, urinalysis, blood gas were normal and his ultrasonography revealed no pathology in kidneys but 120 cc post-void residue in bladder.Clean intermittent catheterization was started and urodynamic evaluation was planned.A significant increase in urine output was observed after furosemide administration, then his kidney function was re-evaluated.His serum cystatin-c level was persisted high for three months and estimated-glomerular filtration rate(eGFR) measured with cystatin-c based formula was 51.8 ml/min/1.73m^2.His urological evaluation is ongoing currently.
Case-2: A fourteen-year-old female was diagnosed with MPS type-3 eight years ago.She was consulted to the nephrology clinic due to decreased urine output and recurrent urinary tract infection.Her serum creatinine, albumin, electrolytes, blood gas and urinalysis were normal her ultrasonography revealed increased renal echogenicity and in scintigraphic imaging there was increased background activity but no cortical lesion.Urodynamic evaluation showed a low-capacity, low-filling pressure bladder with sphincter insufficiency.Her serum cystatin-c level was persisted high for three months and eGFR was 43 ml/min/1.73m^2.
Conclusions: Cystatin-c is a better marker to evaluate kidney functions because of overerestimated creatinine based eGFR levels in MPS patients.Detrusor-sphincter insufficiency due to muscular involvement is not a surprising finding in these patients.It should be kept in mind that MPS patients are at risk for urinary system dysfunction and investigations should be performed in the presence of clinical suspicion.

EP-240 PHOSPHATE METABOLISM IN NON-KIDNEY DISEASE

Anastasia Yudina, Svetlana Vanyakina, Elena Tush, Tatyana Eliseeva, Anna Obukhova, Andrey Stroganov, Olga Khaletskaya

Privolzhsky Research Medical University, Ministry Of Health Of The Russian Federation, Nizhny Novgorod

Introduction: Phosphorus is a vital component of the cell and organism, playing critical roles in many essential processes. But other hands, phosphate toxicity is a well-established phenomenon, especially in chronic kidney disease.
Aim our investigation is evaluate phosphate metabolism in chronic non-kidney disease — bronchial asthma in children.
Material and methods: We examined 26 children and adolescents with bronchial asthma hospitalized in the pediatric department of the Childrens Clinical Hospital No. 1 in Nizhny Novgorod at the age of 4 to 17 years; the median age was 11.5 [9.0; 14.0] years, boys 69.2 (18/26)%. The study was retrospective and did not require ethical committee approval. Examination and treatment of children was carried out in accordance with accepted standards and clinical recommendations according to the nosological diagnosis. Determination of calcium, phosphate and creatinine was performed using standard biochemical laboratory methods in samples obtained from 24-hour urine collections and in venous blood samples. Spirometry studies were performed using a MasterScreen Pneumo spirometer (Jaeger, Germany) in accordance with international recommendations. Statistical analysis was performed using the Statgraphics Centurion, v. 18. Differences were considered statistically significant at $p<0.05$.
Results: We found a statistically significant negative correlation between the concentration of phosphorus in the urine and in the blood with the FEV1/FVC index (r=-0.4, p=0.04 and r=-0.44, p=0.02, respectively) and MEF 25 (r=-0.42, p=0.03 and r=-0.37, p=0.05 for blood and urine phosphorus concentrations, respectively).There is also a a statistically significant correlation relationship between MEF 25 and the calcium-phosphorus urine ratio (r=0.48, p=0.01), as well as the concentration of calcium in the blood (r=-0.39, p=0.04). But statistically significant correlation between tubular phosphate reabsorbtion and FEV1/FVC index and MEF 25 did not found (r=-0.27, p=0.18 and r=-0.32, p=0.12).
Conclusions: We found negative influence phosphate levels on bronchial conduction. So, need a futher investigation phosphate toxicity in non-kidney disease.

EP-241 A NOVEL AQP2 MISSENSE GENE MUTATION IN A FEMALE GIRL.

Martine Docx[1], Bart Loeys[3], Nathalie Segers[1], Johan Vande Walle[2]

[1]*Queen Paola Childrens Hospital Antwerp Belgium*, [2]*University Hospital Ghent Belgium*, [3]*University Hospital Antwerp Belgium*

Introduction: Nephrogenic Diabetes Insipidus (NDI) is a rare disease and is characterized by production of hypoosmolar urine (> 50-60 ml/kg) (age: 0-10 year) despite concurrent hypovolemia, due to the inability of renal collecting tubules (CT) to absorb water in response to antidiuretic hormone (ADH). In extreme rare occasions, the disease is caused by mutations in the AQP2 gene located on chromosome 12q13.12.
Material and methods: A Syrian family including 5 family members was investigated. The proband was first admitted at the age of 4 $^{9/12}$ years, presenting polydipsia, polyuria and signs of failure to thrive. Testing the proband upon admission revealed a low spot urine osmolality of 134 mOsm/kg (NL: 300-900 mOsm/kg) and a normal plasma osmolality of 285 mOsm/kg (NL: 275-295 mOsm/kg). The diagnosis of nephrogenic diabetes insipidus was retained and DNA analysis was performed. The patient is treated with a combination of indomethacin and amiloride together with an adequate supply of fluid and sodium restriction.
Results: The genetic diagnosis is a novel AQP2 missense gene mutation (homozygous variant c490A>C(pIIe164leu)) in the proband.Testing of both parents and brother and sister gave the same variant (all were heterozygous) and all were since now symptom-free. The AQP2 monomer consists of five loops. According to our opinion the missense mutation of our proband is located in the fifth transmembrane segment.
Conclusions: DNA sequence analysis revealed a novel missense mutation in the AQP2 gene. We suggest in nephrogenic diabetes insipidus that adequate diagnosis and treatment is important for improving the prognosis of these children.

EP-242 AUTOSOMAL RECESSIVE TYPE I PSEUDOHYPOALDOSTERONISM: A SEVERE AND INFREQUENT CAUSE OF HYPERKALEMIA

Raquel Jiménez García, Pedro Arango Sancho, Ana Cristina Aguilar Rodríguez, Yolanda Calzada Baños, Marta Jiménez Moreno, Elena Codina Sampera, Álvaro Madrid Aris

Hospital Sant Joan De Déu

Introduction: Pseudohypoaldosteronism type 1 is a rare hereditary syndrome characterized by resistance to mineralocorticoids.They manifest with sodium loss, hypovolemia,hyperkalemia,metabolic acidosis,and markedly elevated plasma aldosterone and renin levels.There are two phenotypes associated with the type of inheritance:autosomal recessive or dominant.The first affects the genes of the subunits of the epithelial sodium channel (ENaC),being the defect systemic,serious and permanent; the second form,and the most frequent,is due to a heterozygous mutation of the NR3C2 gene that encodes the mineralocorticoid receptor with affection limited to the kidney and that is usually associated with a slight loss of salt, improving with age
Material and methods: A 7-day-old male with hours of evolution of hypoactivity,two vomiting and dyspnea.Parents of Pakistani origin,positive consanguinity (first cousins),report death of their first daughter of unexplained cause at 8 days of life.On admission,he presented supraventricular tachycardia in context of severe hyperkalemia associated with hyponatremia
Results: He received antihyperkalemic measures,hydration and intravenous corticosteroids due to suspected congenital adrenal hyperplasia.The subsequent hormonal study shows normal levels of 17-OH-progesterone, dihydroepiandrosterone-sulphate,and androstenedione,along with elevated cortisol.These results rule out the suspicion of adrenal hyperplasia, suggesting possible hypoaldosteronism, mineralocorticoid resistance,or renal tubular acidosis.After confirming elevated aldosterone and renin,it was oriented as Pseudohypoaldosteronism, complementing the diagnosis with an electrolyte test in sweat,which is positive.The genetic study detects a variant in SCNN1A of uncertain significance in homozygosity,which,associated with clinical symptoms and extension studies,is considered a confirmatory test.The management has been complicated,but it has been possible to send home
Conclusions: In the case of our patient,the history,evolution and complementary studies lead us to pseudohypoaldosteronism with autosomal recessive inheritance with systemic involvement,which determines early presentation and its severity.It is probable that the first daughter was also affected by this syndrome and the cause of death was similar to the one presented by our patient.The diagnosis of certainty through genetic confirmation in cases of dehydration and salt loss of neonatal presentation and possible tubular origin is essential since the management and prognosis changes radically according to the associated pathology

EP-243 RITUXIMAB NEEDS TO BE CONSIDERED "LICENSED" ALSO IN NEPHROTIC SYNDROME

Mario Giordano, Diletta Domenica Torres, Vincenza Carbone, Luisa Santangelo, Marida Martino, Alessandro Mascolo

Pediatric Nephrology And Dyalisis Unit- Pediatric Hospital Giovanni Xxiii Bari, Italy

Introduction: Rituximab was approved in 1997 by the Food and Drug Administration for treatment of some tipes of B-cells neoplasm as refractory leucemias or relapsing lymphoma. In subsequent years, its use has been progressively extended to other various non nephrological diseases, as multiple sclerosis or neuromyelitis optica or immune thrombocytopenia and others more. Despite the proven efficacy and safety, currently in Europe, RTX appears to be authorized by EMA (European Medicines Agency) exclusively for non-Hodgkin Limphoma (NH), Chronic lymphocytic leukaemia (CLL), rheumatoid arthritis, Granulomatosis with polyangiitis (GPA) and microscopic polyangiitis and Pemphigus vulgaris.
From the early 2000s, many reports have suggested that RTX is effective for children with frequently relapsing or steroid-dependent nephrotic syndrome (FRNS/SDNS).On the other hand, in the steroid-resistant nephrotic syndrome (SRNS), in the absence of certain effective therapies, efficacy of RTX seem to be controversial to control the disease . In spite of the now common use of RTX in nephrotic syndrome, this therapeutic option still remains "off-label", with various regulations in each European country
Material and methods: We report our experience about use "off - label" of RTX in a cohort of 33 children affected by NS (21 males, 12 females). After the usual steroid therapy, 25 were labeled as SDNS, 8 as SRNS. As second line therapy we used other drugs variously combined (Cyclosporine or tacrolimus in 30, cyclophosphamide in 11, Micofenolate in 9). Due to the persistence of NS, we prioposed to parents use "off-label" of RTX . It is a procedure governed by laws that, at least in Italy, requires the treating clinician to forward 1) authorization to the hospitals Health Department, supported by scientific literature 2) informed consent 3) communication of a therapeutic plan on the specific reasons involved in the use off-label (age, different therapeutic indication, route of administration or dosage)
Results: Altogheter, in our patients we administered RTX (at dosage di 375 mg/mq) 68 times. Each individual patient received a variable number of administrations from 1 to 5.
Only 3/8 pts with SRNS had total remission of proteinuria after infusions of RTX (1 - 5, mean 3 infusions). 4 of the remaining patients underwent one or more sessions of Plasma Exchange (PEX) but just one of them witnessed complete remission.
19/25 pts affected by SDNS or FRNS had a complete remission, after one o more administration, while one showed partial remission. 3 pts were lost to follow up, one was considered unresponsive.

Conclusions: All medicines must be authorised before they can be marketed and made available to patients. In the European Union (EU), there are two main routes for authorising medicines: a centralised route (EMA) and a national route (AIFA, in Italy). In light of the now common use of RTX in infantile NS, with efficacy and safety demonstrated by numerous trials and controlled clinical studies, it would be appropriate for the EMAor AIFA, urged by the scientific societies, to declare RTX "*AUTHORISED*" also in FRNS/SDNS.
The purpose of this work is not to make known our experience on the use of Rituximab but rather to underline how this drug is now commonly used in NS. Continue to consider it as an "off-label drug" is now outdated in light of the multiple clinical experiences.

EP-244 RENAL MALFORMATIONS IN CHILDREN IN WEST PART OF ROMANIA

Raluca Isac, Mihai Gafencu, Ruxandra Maria Steflea, Cristina-ioana Olariu, Ramona Florina Stroescu, Andrada Mara Ardelean, Gabriela Doros

University Of Medicine And Pharmacy „victor Babes" Timisoara

Introduction: Congenital Anomalies of Kidney and Urinary Tract (CAKUT) represent a heterogenous group of diseases with variable evolution, clinical manifestations and complications. CAKUT is the main cause of chronic kidney disease (CKD) in children, while renal defects (hypo/dysplasia) are responsible for over 50% of adolescents needing renal replacement therapy. Diagnosing CAKUT is relatively facile due to non-invasive ultrasound examination screening, both prenatal and postnatal. Early diagnosis and monitoring may improve life quality and reduce the risk of complications, such as urinary tract infection (UTI) or CKD.
Material and methods: We analysed retrospectively 42020 observation files for a period of 30 months and identified 252 children with CAKUT, while 73 patients had 82 renal malformations. Malformations of interest were: renal agenesia, renal hypoplasia, renal dysplasia, rotation/number or fusion anomalies. Data collection included: demographical data (age, sex, rural/urban origin), clinical data (symptoms, occasion of diagnosis, bilateral/unilateral defect, associated pathology) and biological data (renal function, microscopic urine exam, urine culture). Data was statistically analysed using SPSS v22 for Windows, MS Excel.
Results: Out of 82 renal malformations, 48 were solitary renal malformations, 9 of them double, 21 associated with ureteral defects, one with vesical defect, while 3 patients had complex malformation. Renal displazia was the most frequent encountered defect, (0.057% incidence), followed by renal agenesia (0.049%) and renal hypoplasia (0.036%). Bilateral defects were present in 17 patients (20.73%), with a predilection for right side in unilateral malformations (1,4:1). Equal sex repartition and equal repartition in urban/rural areas. Most patients were aged over 6 years (54.79%) and toddlers (26.02%). Diagnosis was mainly random (38.35%) or simultaneously with an UTI episode (32.86%). Thirty-two patients (43.83%) with renal malformations developed UTI, while 21.91% of patients developed CKD.
Conclusions: Early diagnosis of renal malformations is mandatory for monitoring renal function.

EP-245 A CASE OF CLINOSTATIC PROTEINURIA

Maria Cristina Mancuso, Laura Lucchetti, Francesco Emma, Marina Vivarelli

Division Of Nephrology And Dialysis, Department Of Pediatric Subspecialties, Bambino Gesù Pediatric Hospital Irccs, Rome, Italy

Introduction: Orthostatic or postural proteinuria is a common condition in children, represented by proteinuria while in the upright position only, due to increased pressure on the renal glomeruli, which forces the passage of proteins through them. In the supine (clinostatic) position, pressure decreases and protein loss is absent
Material and methods: An 11-year-old female patient was admitted to our department for occasional finding of proteinuria. No microhematuria was detected. She had normal serum creatinine levels, normal serum C3, C4 and immunoglobulin levels, negative family history for nephropathies. To discern orthostatic proteinuria, separate urine samples collected first-morning after at least 6 hours supine and at the end of an active day were collected.
In the first urinalysis, UPCR was 0.71, in the second the ratio was negative (0.1). Assuming an error in the collection of the sample, the test was repeated 2 more times. However, the results were the same.
Results: An ultrasound was performed showing normal kidneys and urinary tract. Collaterally, an expansive formation of 8 x 10 cm was found on the spleen, with clear and regular margins and ecostructure compatible with a splenic cystic. An MRI confirmed the diagnosis. Given the large size of the cyst, we hypothesized that the spleen could put pressure on the left kidney in the supine position, to explain the pathogenesis of the patients purely clinostatic proteinuria. Scleroembolization of the cyst has been attempted without a substantial reduction of the cyst, therefore proteinuria is still present in this patient.
Conclusions: The finding of high-molecular weight non-nephrotic proteinuria in older children requires dosing in the orthostatic and clinostatic position to exclude a mechanical pathogenesis. In this particular case, the splenic cyst may cause a renal compression in the supine position only, reversing the frequent condition of orthostatic proteinuria and determining instead clinostatic proteinuria.

EP-246 CAN RENAL RESISTIVE INDEKS BE A USEFUL MARKER OF EARLY RENAL IMPAIRMENT IN CHILDREN WITH TYPE 1 DIABETES

Ivana Trutin[1], Gordana Stipančić[1], Lea Oletić[1], Mario Laganović[2]

[1]*Clinical Hospital Center Sestre Milosrdnice*, [2]*University Hospital Centre Zagreb*

Introduction: The aim of this study was to determine the Renal resistive index measured by the Doppler ultrasound method in children with type 1 diabetes who are normoalbuminuric and in normal age-matched controls. Renal resisitve index is an indicator of increased vasculature intrarenal resistance.
Material and methods: In this cross-sectional study 76 health children and 76 children with type 1 diabetes age 10-18 years were included, with a disease duration of 2-10 years with normal renal function and normoalbuminuria . The Doppler renal resistive index was measured in subjects in both groups.
In a study group a blood sample was taken for glycated hemoglobin and in both groups blood samples for creatinine, lipids,serum uric acid and first-morning urine samples for albumin/creatinine ratio were taken.
Results: Lower serum creatinin (<0.001) and serum uric acid (0.014) and higher albumin/creatinin ratio (p=0.004) were observed in type 1 diabetes group. Higher estimated glomerular filtration was also observed in type 1 diabetic group although not statistically significant. Higher renal resisitive index was found in type 1 diabetes group (p <0.001. No differences were found in age, gender distribution, blood pressure or pubertal development. An unsupervised machine learning approach was used to identify a possible marker for renal impairment within the diabetic group. With the expectation maximization method 2 groups were identified, the so-called clusters of patients with common characteristics. Common features of Cluster 2, compared to Cluster 1, are higher renal resisitve index

values, albumin/creatinin ratio, estimated glomerular filtration, lower serum creatinin and lower diastolic blood pressure.
Conclusions: Renal resistive index is increased early in type 1 diabetes mellitus and it may be helpful as an early marker in diagnosing preclinical stage of diabetic nephropathy in normoalbuminuric children with type 1 diabetes mellitus.

EP-247 CLINICAL SPECTRUM OF CONGENITAL ANOMALIES OF KIDNEY AND URINARY TRACT IN MACEDONIAN CHILDREN

Nadica Ristoska-bojkovska, Andrea Bojkovska
Kb Acibadem Sistina

Introduction: Congenital abnormalities of the kidneys and urinary tract present a family of deseases of various anatomic spectrum, including renal anomalies, and anomalies of the bladder and urethra.
Material and methods: Patients (700) with congenital anomalies had standard techniques to examine the kidneys and urinary tract: echosonographic examination, micturition cystography, diuretic scintigraphy, cortical scintigraphy (Tc-99 DMSA scan), direct radionuclear cystography, and in selected cases computerized tomography and nuclear magnetic resonance
Results: Positive familial history has been found in 12,42 % of the patients where no significant statistical difference has been identified in relation to ethnicity and type of malformation. Most common malformation in our series is vesicoureteral reflux (VUR- 39,92%). The molecular diagnosis of patients with renal hypo dysplasia and renal agenesis can be achieved with Copy number variation (CNV) analysis in 10% of the patients.
Conclusions: In general we can conclude that the prognosis of pediatric CAKUT in Macedonia is excellent. Adverse prognosis is associated with existence of obstructive anomalies (valvula of the posterior urethra), and bilateral affection (hypo dysplasia, VUR). Favorable results obtained from this study are due to the high non- selectiveness of our series.

EP-248 IMMUNOGLOBULIN A VASCULITIS IN CHILDREN AND ITS ASSOCIATION WITH NEPHRITIS- A SINGLE CENTER EXPERIENCE

Brankica Spasojević[2], Mirjana Cvetković[1], Ivana Gojković[1], Gordana Miloševski-lomić[1], Srđan Nikolovski[1], Ana Petronijević[1], Milica Vukanović[1], Bojana Veselinović[1], Dusan Paripović[2], Mirjana Kostić[2]

[1]*University Childrens Hospital Belgrade*, [2]*Faculty Of Medicine Belgrade*

Introduction: Henoch Schönlein purpura (HSP), recently called IgA vasculitis is a systemic vasculitis characterized by deposits of immunoglobulin A in blood vessels. Renal impairment of these patients, IgA vasculitis nephritis (IgAVN), is the main determinant of prognosis. IgAVN is a relatively benign disease in children, whereas, long-term cohort studies have shown high sustained rates of severe proteinuria and renal dysfunction in these patients during adulthood.
Material and methods: We described our experience of a single center with clinical presentation, kidney involvement, treatment modalities and outcome in children diagnosed as HSP. One hundred and eighty eight patients with HSP diagnosed from January 1, 2010, through December 31, 2021 were retrospectively identified.
Results: The median age of patients was 6.96 years (4.69-11.13) and female/male ratio 92/96. At presentation, 77.7 % of patients had arthritis/artralgia, 44.4% had diffuse abdominal pain (intestinal bleeding in 11.6 %), 27.1% had angioedema and 34.2% kidney involvement. During the mean follow up of 21.04±27.57 months, 10.7% of patients had hematuria, 9.1% hematuria and nonnephrotic proteinuria, 8.6% hematuria and nephrotic proteinuria, 4.3% isolated nonnephrotic proteinuria and 1.6% had the most severe onset presenting as nephritic-nephrotic syndrome. Kidney biopsy was done in 16 patients (9%). Those patients treated with steroids, five of them received cyclosporine due to no steroid response. Only one patient with rapid progressive nephritis successfully treated with cyclophosphamide and 11 plasma exchange sessions. Remission of hematuria experienced 77.2% of patients after a median time of 2.47 months (0.00-7.00). After a median 4.33 months (1.42-9.0), complete remission of proteinuria was achieved in 82.2% of patients and partial in 11.1% . At the most recent follow up, only one patient has CKD grade II.
Conclusions: IgAVN in children have generally good prognosis with minority of cases developing CKD. Despite this relatively benign disease in children, patients with IgAVN need lifelong care and observation.

EP-249 ANATOMOPATHOLOGICAL FEATURES OF PRIMARY FOCAL SEGMENTAL GLOMERULOSCLEROSIS IN CHILDREN: TUNISIAN SERIES

Abir Boussetta, Khouloud Ben Njima, Nesrine Abida, Farah Krifi, Manel Jellouli

Pediatric Nephrology Department, Charles Nicolle Hospital Tunis, Tunisia

Introduction: To describe the main anatomopathological characteristics found in a series of Tunisian children with focal segmental glomerulosclerosis (FSGS).
Material and methods: This was a retrospective study conducted in the pediatric nephrology department of the Charles Nicolle Hospital in Tunis, Tunisia over a period of 20 years, from January 2001 to December 2020. Inclusion criteria were: patients less than 18 years of age at the time of diagnosis of the disease, regularly followed up with renal biopsy-proven FSGS. Exclusion criteria were: children with suspected or proven secondary FSGS.
Results: A total of 35 patients were included in our study, the median age at diagnosis of the FSGS was 4.5 years. Renal biopsy (RB) was performed at least once in all our patients. The indications were as follows: steroid-resistant nephrotic syndrome in 16 cases, nephrotic syndrome with atypical presentations such as gross haematuria, renal impairment, hypertension and hypocomplementaemia in 5 cases, nephrotic syndrome with frequent relapses in 10 cases, isolated asymptomatic proteinuria in 4 cases. Not otherwise specified (NOS) variant was found in 77.1% of cases, tip lesion variant in 17.1% of cases, the perihilar variant and the collapsing one were found in 2.9% of cases each. Tubulointerstitial injury were found in 77.1% of cases. Interstitial fibrosis was found in 62.8% of cases with extensive fibrosis in one patient with tip lesion variant. Tubular atrophy was found in 13 patients, and tubular necrosis in 3 children.
Arterial and arteriolar nephrosclerosis were found in 40% of cases, there was no statistically significant correlation between histological features and response to treatment.
Conclusions: There is no correlation between the different anatomopathological aspects and the clinical characteristics and outcomes. For further identification of clinical significance, morphologic studies including tubulointerstitial changes and findings of immunohistochemistry and electron microscopy would be necessary.

EP-250 HYPOPHOSPHATASIA CHILDHOOD FORM PHENOTYPIC FEATURES: CASE REPORT

Nazi Levi, Zhanna Leviashvili

Saint Petersburg State Pediatric Medical University

Introduction: HPP (ORPHA 436) AR/AD is caused by a mutation in the ALPL gene, encoding a nonspecific tissue isoenzyme of alkaline phosphatase (ALP) - tissue-specific ALP, which in turn leads to the accumulation of phosphoethanolamine, pyridoxal-5-phosphate and inorganic pyrophosphate. HPP is clinically manifested by damage to the bone, pulmonary, nervous (vitamin B-dependent convulsions) and renal systems, a progressive course. HPP is classified: perinatal, infant, childhood, adult and odontohypophosphatasia. In the absence of timely enzyme replacement therapy for severe forms of HPP, the prognosis for life is unfavorable.
Material and methods: We present a case report of a 4year6month girl with HPP in childhood.
Results: A 1-year-old girl had manifested rickets and muscle hypotension. At 1year4 months' growth was 74cm, weight 8kg. Skeleton deformities: an increase in the frontal tubercles, brachycephalic skull, a large fontanel 1.5x1 cm, deformity of the chest, Garrisons groove, rickety rosary, an increase in the wrists, shortening of the right limb, deformity valgus. X-ray changes in the metaepiphyseal zones, expansion of growth zones, unevenness of the time zone of calcification and expansion of metaphysis, "tongues" of enlightenment in the projection of growth zones into the metaphysis, curvature and shortening of tubular bones. Premature loss of deciduous teeth. Low level of ALP (in 1-3 years 33-44U/l), hypercalcemia, low 25(OH)D, hypercholesterolemia, hypercalciuria, hypostenuria, Ca/Cr 1,0mg/mg, nephrocalcinosis, GFR 93.55 ml/min*1.73 m2. Genetic diagnostic detected homozygote variant of mutation in the ALPL gene.
Conclusions: The features of the phenotype and early diagnosis of hypophosphatasia in childhood form caused by a mutation of the ALPL gene in a girl are described. Enzyme replacement therapy in a patient with Asphotase-alpha (Strensiq) gave a positive effect.

EP-251 A RARE CAUSE OF THROMBOCYTOPENIA AND PROTEINURIA IN AN ADOLESCENT GIRL

Hulya Nalcacioglu[1], Aysegul Yilmaz[2], Demet Tekcan Karali[1], H.gozde Onal[1], Ozlem AydoĞ[1]

[1]Ondokuz Mayis University Faculty Of Medicine, Pediatric Nephrology Department, Samsun, Turkey, *[2]Ondokuz Mayis University Faculty Of Medicine, Pediatric Genetic Department, Samsun, Turkey*

Introduction: Myosin heavy chain-9-related disorders (MYH9-RDs) are a genetic disorder of autosomal dominant inheritance caused by mutations of the MYH9 gene. Most affected individuals develop other extrahematologic manifestations of the disease including sensorineural hearing loss, renal disease, cataracts, and/or elevation of liver enzymes.
Material and methods: Myosin heavy chain-9-related disorders (MYH9-RDs) are a genetic disorder of autosomal dominant inheritance caused by mutations of the MYH9 gene. Most affected individuals develop other extrahematologic manifestations of the disease over their lifetime, including sensorineural hearing loss, renal disease, cataracts, and/or elevation of liver enzymes.
Results: A 15-year old female was admitted to the nephrology clinic to evaluate proteinuria with a history of chronic thrombocytopenia since childhood. At first, ITP was suspected due to thrombocytopenia. When he was ten years old, hearing loss was detected along with micro/macrohematuria and nephrotic-range proteinuria. The audiogram revealed a profound high-frequency sensorineural hearing deficit bilaterally. Cataract was present by ophthalmological evaluation. Due to the clinical suspicion of MYH9-RD, genotyping of the patient was performed. MYH9 gene (c.4997C>T) heterozygous mutation was found. Enalapril (5mg/day) was initiated for renal protection. The patient was lost to follow-up.
Conclusions: MHY9RD is a rare syndrome that can end with end-stage renal disease and severe hearing loss. This rare diagnosis should take into consideration acquired and inherited forms of thrombocytopenia.

EP-252 SECONDARY NEPHROGENIC DIABETES INSIPIDUS (NDI) IN BARTTER SYNDROME (BS) TYPE 1: TWO CASE REPORTS.

Maria Concetta Lonardo[1], Vittorio Serio[2], Luigi Annicchiarico Petruzzelli[2], Paolo Giannattasio[2], Angela De Luca[2], Enrico Zulli[2], Fiorella Migliaro[3], Gabriele Malgieri[2], Carmine Pecoraro[2]

[1]Emergency Department, Santobono-pausilipon Children's Hospital, Naples, Italy, *[2]Nephrology And Dialysis Unit, Santobono-pausilipon Children's Hospital, Naples, Italy*, *[3]Division Of Neonatology, Department Of Translational Medical Sciences, University Federico Ii, Naples, Italy*

Introduction: Bartter syndrome is a rare inherited salt-losing renal tubular disorder, whose primary pathogenic mechanism is defective salt reabsorption. BS is classified into five types, based on molecular genetics. There is significant variability in the clinical expression of the disease.
Material and methods: We report the cases of two infants with antenatal BS and a secondary form of NDI. The first patient was a female infant born at 35 weeks of gestation with history of polyhydramnios and severe IUGR, with bilateral nephrocalcinosis. At two month of age she was referred for fever, failure to thrive, dehydration and vomiting. The second patient was an extremely premature female newborn of 26 weeks of gestation, after a pregnancy complicated by a marked polyhydramnios. Among other prematurity-related complications, she had neonatal diabetes mellitus, hypovolemic acute renal failure and severe failure to thrive; renal function slowly normalized; she started to show severe hypernatremia and polyuria. Both of our patients had hypernatremia and a urinary concentration defect, consistent with NDI. Laboratory test also revealed mild-moderate hypokalemia, normotensive hyperreninemic hyperaldosteronism, inconstant metabolic alkalosis with normal serum chloride, hypercalciuria.
Results: Intravenous 5% glucose solution was administered in order to correct hypernatremic dehydration. After therapy with indomethacin was started, their growth and serum electrolytes levels promptly improved. Potassium chloride is orally supplemented. Genetic analysis yielded mutations in the SLC12A1 gene, consistent with the diagnosis of BS type 1.
Conclusions: A phenotypic variability has been reported in patients with BS; sometimes, type 1 and 2 BS may present with or develop a secondary NDI. In this case, despite BS is a salt-wasting disorder, salt restriction is crucial, in order to avoid polyuria and hypernatremic dehydration. Moreover, antenatal BS should always be considered in patients with polyuria and a history of polyhydramnios, prematurity and growth retardation.

EP-253 VITAMIN D-DEPENDENT RICKETS TYPE 1A (VDDR1A), BY MUTATIONS IN THE CYP27B1 GENE AND COAGULOPATHY DUE TO VITAMIN K DEFICIENCY IN SIBLINGS: FOLLOW-UP LONG-TERM

Nazi Levi, Zhanna Leviashvili, Nadezhda Savenkova, Karina Papayan

Saint Petersburg State Pediatric Medical University

Introduction: VDDR1A (OMIM: 264700) AR is caused by mutation, encoding the enzyme 1-alpha hydroxylase, manifests rickets, hypocalcemia, hypophosphatemia, high alkaline phosphatase (ALP), parathyroid

hormone (PTH), and acidosis. Vitamin K is essential for the synthesis of anticoagulant proteins, osteocalcin and GLA matrix-protein.
Material and methods: Introducing long-term follow-up of siblings (proband 16years and sibs 13years8months) with VDDR1A.
Results: Manifestation in proband and siblings of 1 and 1,5 years, respectively. The diagnosis of VDDR1A in siblings was established by phenotype and genetic testing of the CYP27B1 gene in a heterozygous state (compound-heterozygous).

Table

Clinical characteristics of patients

	proband 7years	with therapy 16years	sibs in 5years	with therapy 13years8months
Length(см)	110↓	166	99,5↓	150
Ca(n2,3-2,7mmol/l)	1,91↓	2,30	2,05↓	2,27
ALP(n141-460un/l)	736,0↑↑	155	800↑↑	
P(n1,3-2,3mmol/l)	1,20↓	1,46	0,80↓	1,45
PTH(n9-52pg/ml)	237.5↑↑	63,8	134,2↑↑	75
$25(OH)_2D$(n14-80ng/ml)	42	83,4	23↓	43
$1,25(OH)_2D_3$(n16-65pg/ml)	16,01↓	41,3	17,08	31,65
Ca/Cr(n<0,2mg/mg)	0,13↓	0,16	0,36↓	0,25
P/Cr(n0,97mg/mg)	1,76	1,05	1,44	1,11
TRP (n80-85%)	73,38↓	86,63	75,15	85,05
MTRP(n0,97-1,68mmol/l)	0,88↓	0,94	0,71↓	1,96
FPE(n5-20%)	26,63	13,37	24,85	21,01
MTRP/TmP(n1,15-2,44mmol/l)	0,73↓	0,67	0,52↓	0,98
TmP(ml/min*1,73м^2)	120	140,29	139,14	162,36
Ca-urine(n5mg/kg/day)	3,6	3,03	0,6	1,8
P-urine(22mg/kg/day)	30,5	8,26	19,35	9,2
Osteocalcin(n3,10-13,7ng/ml)	16,8↑	15,6	19,60↑	16,4
B-crosslaps(n0,15-0,82ng/ml)	1,86↑	1,01	2,03↑	1,58
Vit K1(n0,1-22ng/ml)	0,04↓	1,14	0,09↓	1,12
aPTT index(n0,8-1,1)	1,25↑	-	1,47↑	-
Fibrinogen(n1,8-4,0g/l)	1,66↓	-	2,3↓	-
Factor VII(n73-133%)	66,6↓	-	57,4↓	-
Factor XII(n67-143%)	40↓	-	63↓	-

Conclusions: We described siblings with phenotype and genotype of VDDR1A and coagulopathy associated with vitamin K deficiency. The therapy (calcidiol, calctriol, phosphate buffer, calcium) is effective and prevents the progression of the disease.

EP-254 VASCULAR COMPLICATIONS AFTER PEDIATRIC KIDNEY TRANSPLANTATION

Abir Boussetta[1], Farah Krifi[1], Nesrine Abida[1], Taieb Ben Abdallah[2], Abderrazak Bouzouita[3], Manel Jellouli[1], Tahar Gargah[1]

[1]Pediatric Nephrology Department, Charles Nicolle Hospital Tunis, Tunisia, [2]Research Unit Of Immunopathology And Immunology Of Renal Transplantation Lr03sp01, [3]Urology Department, Charles Nicolle Hospital Tunis, Tunisia

Introduction: To study the clinical and evolutive characteristics of vascular complications after kidney transplantation in Tunisian children.
Material and methods: This was a cross-sectional, descriptive, retrospective study carried out in the pediatric and internal medicine A departments of the Charles Nicolle hospital in Tunis. Transplanted patients of age less than or equal to 20 years were included during a 31-year period from January 1989 to December 2019.
Results: A total of 115 transplantations were included in our study during this period, there were 69 boys and 46 girls. The average age was 15.5 years old. Renal transplantation was performed from a living donor in 67.5% of the cases and from a cadaveric donor in the rest of the cases. Hypertension was the most frequent cardiovascular complication after renal transplantation in our series in 51.3% of cases. Four children presented with venous thrombosis, immediately after transplantation (three boys and one girl). The average age was 12.3 years old. The outcome was poor with loss of the renal graft in all cases. Three patients presented arterial thrombosis after transplantation diagnosed by doppler of the renal vessels at the first day in two cases and after one year of the transplantation in the 3rd case. They were three boys, aged 11, 18 and 20, respectively. the outcome was poor in the three cases with a renal graft loss for two patients and the death of the third one. Three children presented with retroperitoneal hematoma that resolved well with surgery
Conclusions: Vascular complications after pediatric kidney transplantation are common and serious, and can affect the renal and life prognosis of transplant recipients.

EP-255 TYPICAL HUS AND PURPURA

Marta Giambrone, Ciro Corrado, Rosa Cusumano, Giovanni Pavone, Maria Chiara Sapia, Davide Vella, Clara Giambrone, Sofia Felice, Melania Guardino, Chiara Marino, Claudio Montante, Giulia Mincuzzi, Maria Michela D Alessandro

Pediatric Nephrology. G. Di Cristina Hospital. Palermo

Introduction: A 16 year-old boy was admitted to the emergency room due to the onset of asthenia, pallor, macrohematuria and diarrhea. For the finding of severe anemia, thrombocytopenia and AKI he was referred to our department.
Material and methods: At admission, the patient presented with sensory obnubilation, diplopia, dysarthria and mental confusion. Laboratory tests showed: hemoglobin 7.6 g / dL, platelet 5,000 / mm3, LDH> 1800, BUN 330 mg / dL, creatinine serum 16.7 mg / dL, AST/ ALT 96/36 IU/L, lipase 452 IU/L and amylase 415 IU/L. Brain CT was negative, while chest CT revealed bilateral pleural effusion with thickening. Abdominal CT showed terminal sigmoid stenosis. He was treated with hemodialysis and eculizumab.
Results: Two days later a stool culture positive for STEC O157 was documented. We couldnt test the plasma activity of ADAMTS-13 due to the lack of reagents. The patient continued eculizumab therapy, with clinical benefit. Two months later a NGS for MTA allowed to identify, two new variants in compound heterozygosity p.L115V and p.C347R in the ADAMTS-13 gene, inherited respectively from the mother and the father. We diagnosed a congenital TTP also called Upshaw – Schulman syndrome (USS). It is an extremely rare hereditary deficiency of ADAMTS13 activity cells. The clinical signs are usually mild during childhood, often with isolated thrombocytopenia, and could become more evident when patients have infections
Conclusions: This case is particularly interesting due to the initial positivity of the STEC, which played a role as a trigger for this rare late onset of TTP

EP-256 THE FREQUENCY OF HAEMATURIA IN CHILDREN DEPENDING ON THE SUMMER HEAT

Irina Balalaeva

Voronezh N.n.burdenko State Medical University

Introduction: The objective of the study was to determine the frequency of haematuria (HU) in children in relation to high summer temperature.
Material and methods:
We detected the frequency of HU in 3246 patients under the age of 17 years in 2018-2021 years.
Results: We noted increasing frequency of symptomless microscopic HU from 2,4% in 2018 year to 8,3% in 2021 year (P<0,001). Frequency of HU growth during 4 months after summer was associated with mean day summer temperature increase by 4,5° in 2021 year in comparison of 2019 year (P<0,05).
Conclusions: Larger frequency of HU during the 2 last years was associated with more summer temperature in relation of more concentrated urine.

EP-257 DIFFERENT PHENOTYPES IN ALPORT SYNDROME.

Mar Espino Hernández[5], Cristina Blazquez Gómez[2], Julia Vara Martín[1], Marina Alonso Riaño[3], Maria Teresa Sanchez Calvin[4], Jesús Ramirez[1], Pablo Bello Gutierrez[6]

[1]Pediatric Nephrology Hospital 12 De Octubre. Madrid. Spain., *[2]Pediatric Nephrology. Hospital Universitario Principe De Asturias. Alcala De Henares. Madrid. Spain.*, *[3]Pathology Deparment. Hospital 12 De Octubre. Madrid. Spain.*, *[4]Genetics Deparment. Hospital 12 De Octubre. Madrid.spain.*, *[5]Hospital 12 De Octubre. Universidad Complutense. Madrid. Spain*, *[6]Pediatric Nephrology Hospital Rey Juan Carlos. Mostoles. Madrid. Spain.*

Introduction: The availability of genetic studies has facilitated the diagnosis of patients with hematuria and has broadened the range in the spectrum of Alport syndrome, with different clinical manifestations and prognosis. We try to describe different phenotypes of patients with diagnosis of collagen IV disease.
Material and methods: We reviewed medical records of these patients: symptoms, family history, age at diagnosis, genetic mutation, biopsy, glomerular filtration rate (GFR) and proteinuria at diagnosis and follow-up.
Results: We included 10 patients, with no family ties to each other, 7 girls and 3 boys with a mean age of 12.5 years. Clinically, 5 patients presented microhematuria, 1 microhematuria and proteinuria, 2 recurrent macroscopic hematuria and 2 nephrotic syndrome. The diagnosis was made in 4 patients by renal biopsy, two of them no pathogenic mutation was found. Genetical studies showed mutations in COL4A5 in two patients, COL4A3 in 4 and COL4A4 in 2 patients. All our patients have normal GFR. One patient with nephrotic syndrome maintains proteinuria/creatinine ratio 0,7 and another responded to steroids without proteinuria at the present but with recurrent macroscopic hematuria. Patients with COL4A5 mutation have proteinuria even with angiotensin-converting enzyme inhibitors. No one else has proteinuria. One patient, with a COL4A3 mutation and C3 and C4 complement low levels, had a grandmother who died with chronic renal failure of unknown cause. The patient with heterozygous mutation in COL4A5 mutation has a mother who required transplant.
Conclusions: Clinical manifestations of Alport syndrome may be atypical and mimic other diseases. Renal biopsy with electronic microscopy is a proven diagnostic tool. However, genetic panel sequencing of collagen IV alterations should be performed before an invasive procedure in patients with glomerular hematuria since the genetic result is often conclusive.
Prognosis is good in short-term follow-up, but rather poor in long-term disease as suggested by the presence of proteinuria and family history.

EP-258 DISTAL RENAL TUBULAR ACIDOSIS – A RARE MUTATION: A CASE STUDY

Edita Petrosyan[1], Maria Proskura[2], Maria Molchanova[1], Valeria Gavrilova[2], Anastasia Ryzhova[2]

[1]Pirogov Russian National Research Medical University, *[2]Russian Children's Clinical Hospital*

Introduction: We report a case of a 4-year boy, with a rare autosomal recessive form of distal renal tubular acidosis.
Material and methods: The boy, was admitted to the hospital at age 1.5 months with fever (38°C), vomiting, food refusal, poor muscle tone and poor weight gain. It was sixth pregnancy (1st boy and 2nd girl died at the age of 3 and 2 months, had similar symptoms; 3rd, 5th – miscarriages; 4th - healthy boy, 2 y.o.). Blood tests: acidosis (pH -7.21), low potassium (1.7 mmol/l), low sodium (132 mmol/l). Serum calcium, phosphorus, urea and creatinine levels were normal. Parathyroid hormone (PTH) increased up to 136 pg/ml. Urine tests showed an alkalotic pH from 7.8 to 8.3, urine calcium-creatinine index 2.31. Genetic test: homozygous mutation in intron 19 of the ATP6V0A4 gene c.2257+3G>T, genotype A/A, leading to alternative splicing.
Results: Metabolic disorders were corrected using potassium citrate 1200mg/d, spiranolokton (12.5 mg/d), hypothiazide 6.75mg/d and alfacalcidol (0.25 μg/d). In result, feeding problems resolved, growth improved. Last hospitalization was at the age 3 y 10 months. Child's physical and mental development was normal, weight 14kg, height 100cm (50 centile). No hearing loss was observed. Serum potassium 4.3 mmol/l, sodium – 142 mmol/l, total serum Ca - 2.5 mmol/l, PTH- 62.1 pg/ml. Blood pH -7.38. Nevertheless, urine tests showed an alkalotic pH from 8.8 to 9.3 and there were mild ultrasound symptoms of nephrocalcinosis (small hyperechoic inclusions around the renal pyramids). Kidney function was preserved (GFR- 109ml/min/1,73m2).
Conclusions: This case demonstrated the importance of genetic examination, which would determine the correct treatment to prevent adverse complications and improve patients life prognosis.

EP-259 THE RARE CLINICAL CASE OF STEROID-RESISTANT NEPHROTIC SYNDROME AND DUCHENNE MYOPATHY.

Varvara Obukhova, Yulia Papina, Svetlana Artemieva, Vladimir Dlin

Veltischev Research And Clinical Institute For Pediatrics Of The Pirogov Russian National Research Medical University

Introduction: Steroid-resistant nephrotic syndrome (SRNS) is the second most frequent cause of end-stage kidney disease (ESKD) in the first 3 decades of life. In children SRNS is usually associated with podocyte genes variants. We would like to present a rare clinical case of SRNS associated with nuclear proteins genes mutations in child with Duchenne myopathy.
Material and methods: The boy with difficulty walking and increased volume and tone of the calf muscles (from 2 yrs of age) developed nephrotic syndrome at age of 4 yrs: edema (ascites, hydropericardium, hydrothorax), proteinuria 3+, hematuria, hypoalbuminemia 14-16 g/l, hypercholesterolemia 12-19 mmol/l, blood creatinine 22-24-16 μmol/l, ALT 151, phosphokinase 5549-14858, LDH 255, normal C3 level and negative antinuclear antibodies. Ultrasound revealed large kidneys with weakened blood flow, left ventricular hypertrophy and thrombus 15x7 mm in vena cava inferior. Treatment with methylprednisolone for 6 weeks was without effect. Kidney biopsy revealed FSGS. The therapy with cyclosporine for 10 months was uneffective. Genetic testing revealed a compound heterozygous mutation of NUP 93 (c.1954C>T) and NUP 205 (c.2243G>A), previously described in patients with SRNS, and 53-55 multi-exon deletion in DMD gene confirming the diagnosis of Duchenne myopathy.
Results: The boy is on treatment with deflazacort (during 7 yrs). He walks independently, but the muscle weakness progresses. The child receives ACE inhibitor (enalapril) 0.5 mg/kg, bisoprolol 5 mg/day, amlodipine 7.5 mg/day. He is in partial remission of NS (proteinuria 1+, 1.1 g/24h, blood albumin 33-36 g/l) and has CKD gr2 (creatinine 20 μmol/l, cystatin C 0.87, eGFR=78 ml/min/1.73m2). There are signs of steroid toxicity: growth retardation (SDS -1.4), arterial hypertension and mycardial remodeling, osteopenia (Z-score total = -1.3).
Conclusions: The case demonstrates the importance of genetic investigation in pts with SRNS and difficulty of management of pats with combined hereditary pathologies.

EP-260 ARE MEASLES, RUBELLA, MUMPS, AND VARICELLA SEROPOSITIVITY SUFFICIENT IN PATIENTS WITH NEPHROTIC SYNDROME?

Aykut ÖzÖn[1], SeÇil Arslansoyu Çamlar[2], GÖkÇen Erfidan[3], ÖzgÜr Özdemir ŞimŞek[3], Cemaliye BaŞaran[3], Demet Alaygut[2], Fatma MutlubaŞ[2], Dilek Yilmaz ÇiftdoĞan[4], Belde Kasap Demir[5]

[1]University Of Health Sciences Izmir Tepecik Training And Research Hospital Department Of Pediatrics, [2]University Of Health Sciences Izmir Faculty Of Medicine Department Of Pediatric Nephrology, [3]University Of Health Sciences Izmir Tepecik Training And Research Hospital Department Of Pediatric Nephrology, [4]Izmİr Katİp Çelebİ Unİversİty Faculty Of Medİcİne Department Of Pedİatrİc Infectİous DİseaseS, [5]Izmİr Katİp Çelebİ Unİversİty Faculty Of Medİcİne Department Of Pedİatrİc Nephrology And Rheumatology

Introduction: Nephrotic syndrome (NS) may cause a loss of protective antibodies against infectious agents, both due to the pathogenesis of the disease and the use of immunosuppressive treatments. It was aimed to evaluate the measles, mumps, rubella (MMR) and varicella immunoglobulin (Ig) levels in the cases followed up with idiopathic nephrotic syndrome (INS).
Material and methods: The patients aged 2-18 years who were followed up with INS between November 2018 and June 2021, who completed their national vaccinations and were in remission were included. Exclusion criteria were being under the age of two, having secondary NS/ proteinuria/ active infection/ known immunodeficiency, or failure to reach the vaccination schedule. Demographic and anthropometric data, follow-up period, number of attacks, USAge of immunosuppressive drugs, histopathological diagnoses, leukocyte and lymphocyte counts, serum urea, creatinine, albumin, c-reactive protein, Ig G-A-M, total cholesterol, and proteinuria levels were recorded. Positivity of IgG values was evaluated as >100 mIU/ML for varicella, >10 IU/ML for rubella, >250 mIU/ML for measles, >25 mIU/ML for mumps. Group comparisons were made according to treatment status and steroid response.
Results: Thirty-nine patients (age between 3 and 18 years) were included. 51.4% were male, the median age was 13 years (3.7-18), the follow-up period was 50 months (12-120 months), and the mean number of attacks was 5.38 (1-17). Seropositivity rates were 56.4% for measles, 69.2% for rubella, 43.6% for mumps and 71.8% for varicella. No difference was found between the seropositivity rates of the groups of steroid-sensitive and steroid-resistant groups, previous treatments and treatment status, pulse steroid treatment status, number of attacks, and histopathological diagnosis.
Conclusions: In this study, it was shown that the antibody levels of INS patients were low. Therefore, it may be helpful to evaluate vaccine responses at certain times in the follow-up of children with a diagnosis of INS.

EP-261 AUTOSOMAL RECESSIVE FAMILIAL HYPOMAGNESEMIA WITH HYPERCLCIURIA AND NEPHROCALCINOSIS (FHHNC) CAUSED BY MUTATION IN THE CLDN16 GENE IN GIRL

Nadezhda Savenkova, Zhanna Leviashvili, Elena Snezhkova, Sergey Laptiev

Saint-petersburg State Pediatric Medical University, Russian Federation

Introduction: Autosomal recessive FHHNC (OMIM# 248250) is hereditary tubulopathy caused by mutations in the CLDN16 and CLDN19 genes encoding the proteins claudin–16 and claudin–19, respectively.
Material and methods: We analyzed phenotype and genotype of renal tubular disease characterized by hypomagnesemia with hypercalciuria and nephrocalcinosis in a 16 years old white girl.
Results: Clinical presentation in a infant was with nephrocalcinosis, associated with hypercalciuria, recurrent urinary tract infection and normal kidney function. Clinical diagnostic includes medullar nephrocalcinosis, hypercalciuria 10 mg/kg, hypomagnesemia 0,6 mmol/l, increased UMg/Cr 0,24 mg/mg and UCa/Cr 2,0 mmol/mmol, early hyperparathyroidism. Genetic diagnostic detected heterozygote variant of mutation in CLDN16 gene - rs751959432 (c.217+5G>A). FHHNC in a child characterized by progression to Chronic Kidney Disease (CKD). Initial symptoms of CKD in a patient of 12 years included polyuria, polydipsia, hypostenuria, increased serum creatinine and urea, decreased glomerular filtration rate (GFR), arterial hypertension. Treatment included Mg supplements, thiazides, citrate, dietary restrictions, antibiotics, management of arterial hypertension. CKD stage 4 was diagnosed at the age of 16 years. Clinical characteristics CKD: serum creatinine 182mmol/l, urea 14mmol/l, GFR 28ml/min, renal tubular acidosis, high parathyroid hormone, anemia, arterial hypertension with left ventricular hypertrophy.
Conclusions: We described 16 years old white girl with the phenotype of autosomal recessive Familial hypomagnesemia with hypercalciuria and nephrocalcinosis caused by mutation in the CLDN16 gene and progressed in to CKD stage 4.

EP-262 SURGICAL COMPLICATIONS AFTER PEDIATRIC KIDNEY TRANSPLANTATION

Abir Boussetta[1], Farah Krifi[1], Nesrine Abida[1], Taieb Ben Abdallah[2], Abderrazak Bouzouita[3], Manel Jellouli[1], Tahar Gargah[1]

[1]Pediatric Nephrology Department, Charles Nicolle Hospital Tunis, Tunisia, [2]Research Unit Of Immunopathology And Immunology Of Renal Transplantation Lr03sp01, [3]Urology Department, Charles Nicolle Hospital Tunis, Tunisia

Introduction: To describe the clinical and evolutive features of surgical complications after kidney transplantation (KT) in Tunisian children
Material and methods: This was a cross-sectional, descriptive, retrospective study carried out in the pediatric and internal medicine A departments of the Charles Nicolle hospital in Tunis. Transplanted patients of age less than or equal to 20 years were included during a 31-year period from January 1989 to December 2019. The kidney that was removed from the donor was placed in the extraperitoneal iliac fossa. We anastomosed the renal vein with the external iliac vein and the renal artery with the internal iliac artery or the external iliac artery. We performed extravesical anastomosis using the Lich-Gregoir technique for all kidney transplant recipients.
Results: A total of 115 transplants were included in our study during this period; there were 69 boys and 46 girls. The mean age was 15.47 years. Surgical complications occurred in 28.6% of cases. These complications were dominated by vesico-ureteral reflux in 12 cases. We have recorded 5 cases of urinary fistula, 4 cases of lymphocele and 4 cases of renal allograft venous thrombosis. Two patients presented a right sural thrombophlebitis. We have recorded one case of ureteral stenosis after 12 months of renal transplantation leading to graft loss and a subsequent return to hemodialysis, and one case of transplant renal artery stenosis. Three children presented with retroperitoneal hematoma that resolved well with surgery. Only one case of urinary lithiasis was noted. One child presented with two different urological complications: a urinary fistula and vesicoureteral reflux.
Conclusions: Kidney transplantation is a complex surgical procedure, especially in children, which can lead to surgical complications that can affect the prognosis of the graft and even the patient

EP-263 KIDNEY MORPHOLOGICAL CHANGES OF THE DAB1-/- MICE

Anita Racetin, Nela Kelam, Mirela Lozić, Natalija Filipović, Snježana Mardešić, Mirna Saraga-babić, Katarina Vukojević

University Of Split School Of Medicine

Introduction: Examination of kidney size, nephron substructures diameters and podocyte morphology of the Dab1-/- (yotari) mice kidneys in comparison with wild type mice. We assumed that inactivation of Dab1 may cause the disorder in a broad spectrum of congenital anomalies of the kidney and urinary tract (CAKUT).
Material and methods: Wild type and yotari mice were sacrificed at postnatal days P4, P11, and P14. Paraffin-embedded kidney tissues were sectioned and analyzed by bright-field and electron microscopy. Mean kidney and nephron substructures diameter was calculated and analyzed by statistical t-tests.
Results: At the TEM microphotographs, foot process effacement in the glomeruli (G) of yotari mice was revealed, whereas aberrations in the structure of proximal convoluted tubules (PCT) and distal convoluted tubules (DCT) were not observed. Furthermore, yotari kidneys were smaller in size, with a reduced diameter of nephron segments, and thinner cortex.
Conclusions: Renal hypoplasia in conjunction with foot process effacement revealed CAKUT phenotype and loss of functional kidney tissue of yotari.

EP-264 PERITONEAL DIALYSIS/LAVAGE IN MULTIORGAN FAILURE FOLLOWING HEMATOPOIETIC STEM CELL TRANSPLANTATION

Katarzyna Gąsowska, Katarzyna Zachwieja, Aleksandra Krasowska-kwiecień, Jolanta Goździk, Dorota Drożdż

Jagiellonian University Collegium Medicum

Introduction: Peritoneal dialysis/lavage in multiorgan failure following hematopoietic stem cell transplantation
Material and methods: 2-month-old boy with familial hemophagocytic lymphohistiocytosis was admitted for allogenic bone marrow transplantation. Previously his treatment included etoposide, dexamethasone and cyclosporine – showing regression of symptoms. On admission he presented Cushing-like-appearance, mild hepatomegaly, required a feeding tube (poor swallowing). Echocardiography confirmed hypertrophic cardiomyopathy. From the beginning, the child showed fluid-retention tendency, requiring close monitoring of the water-electrolyte balance and use of diuretics
Results: The patient underwent HSCT after proper preparation. From 7th day after transplant, he presented with fever- empirical antibiotics were introduced. The childs condition did not improve- water and electrolyte disturbances, coagulation disorders, elevated inflammatory markers were observed. Intensive diuretic treatment, modification of antibiotic therapy and ATIII administration were necessary. Fever subsided, CRP decreased, coagulation status improved. Over next 5 days severe hepatitis and signs of liver failure developed: worsening of coagulation disorders, hyperbilirubinemia and increasing ascites. The child required daily supplementation of plasma coagulation factors and oxygen therapy 1-4 l, crackles over the lung fields occurred. Kidney damage was observed, therefore on 24 th day Tenckhoff catheter was implanted to decompress ascites. Peritoneal cavity was rinsed with 2.27% dialysis fluid every 4 hours. There was leakage of dialysate from the peritoneal cavity next to the catheter. Diuresis was continued to be forced (250-350 ml/day). There have been no auscultation changes over the lungs since Tenckhoff catheter insertion. The childs condition was stable, with moderate: dyspnea and generalized edema was observed. The fluid balance, renal function parameters improved. The patient died later because bleeding complications occurred.
Conclusions: Peritoneal dialysis/lavage proved to be an efficient treatment option in infants presenting AKI/multiorgan failure after HSCT.

EP-265 PERITONEAL DIALYSIS/LAVAGE IN MULTIORGAN FAILURE FOLLOWING HEMATOPOIETIC STEM CELL TRANSPLANTATION

Katarzyna Gąsowska, Katarzyna Zachwieja, Aleksandra Krasowska-kwiecień, Jolanta Goździk, Dorota Drożdż

Jagiellonian University Collegium Medicum

Introduction: Hematopoietic stem cell transplantation (HSCT) became treatment in numerous diseases – it is connected with severe complications including multiorgan disfunction, often acute kidney injury.
Material and methods: 2-month-old boy with familial hemophagocytic lymphohistiocytosis was admitted for allogenic HSCT. Previously his treatment included etoposide, dexamethasone and cyclosporine – showing regression of symptoms. He presented Cushing-like-appearance, mild hepatomegaly, required a feeding tube. Echocardiography confirmed hypertrophic cardiomyopathy. Initially the child showed fluid-retention tendency, requiring close monitoring of the water-electrolyte balance and use of diuretics.
Results: The patient underwent HSCT after proper preparation. 7th day after transplant, he presented with fever- empirical antibiotics were introduced with no clinical improvement – water-electrolyte disturbances, coagulation disorders, elevated inflammatory markers were observed. Intensive diuretic treatment, modified antibiotic therapy and ATIII administration were necessary. Fever subsided, CRP decreased, coagulation status improved. Over next 5 days severe hepatitis and liver failure developed: worsening of coagulation disorders, hyperbilirubinemia and increasing ascites. The child required daily supplementation of coagulation factors and nasal-cannula oxygen therapy, crackles in lungs occurred. Decreased kidney function was observed (serum creatinine 30umol/l, cystatin C 1,78mg/l, urea 22mmol/l, severe overhydration – with tendency to increase), therefore on 24 th day Tenckhoff catheter was implanted to decompress ascites. Peritoneal cavity was rinsed with 2,27% dialysis fluid every 4 hours. There was leakage of dialysate next to the catheter. Diuresis was continuously forced reaching its maximum level 250-350 ml/day. Auscultation changes in the lungs withdrew after Tenckhoff catheter insertion. Fluid balance, renal function parameters improved (creatine ~10 umol/). The childs condition remained stable, with moderate: dyspnea and edema. 6 days later the patient presented with bleeding to respiratory tract, therefore he was moved to ICU, where respiratory failure progressed fatally in few days. Peritoneal dialysis was continued until patient's death.
Conclusions: Peritoneal dialysis/lavage proved to be an efficient treatment option in infants presenting acute renal/multiorgan multiorgan dysfunction after HSCT

EP-266 HEMORRHAGIC CYSTITIS SECONDARY TO ADENOVIRUS AND BK VIRUS INFECTION AND ACUTE RENAL FALIURE IN A BOY WITH ACUTE LYMPHOBLASTIC LEUKEMIA

Danko Milosevic[1], Daniel Turudic[2], Ernest Bilic[1]

[1]*University Of Zagreb, School Of Medicine*, [2]*Department Of Pediatric Hematology And Oncology, University Hospital Centre Zagreb*

Introduction: A boy with acute lymphoblastic leukemia (common-ALL, standard risk) was admitted for treatment according to ALL-BFM 2009 protocol (induction phase).
Material and methods: Case report
Results: After 36 days of ALL treatment, the boy had gross macrohematuria, proteinuria (0.53 g/L) alongside mild renal insufficiency. The child was oliguric (less than 0.5 mL/kg/h) with elevated urea (13.8), and fast-rising creatinine (134) CT scan revealed mild hydronephrosis with left side nephrolit (4 mm) and a hyperdense content in the prevesical part of the of both ureters corresponding with clot, i.e., fresh hemorrhage. The bladder wall was trabeculated, circularly non-uniformly thickened, hyperechoic, up to 7.5 mm thick, in the sense of inflammatory changes. In the lumen of the bladder, for the most part along the wall, a hyperechoic content of up to 9 mm in size is seen; differential diagnosis may correspond to coagulation or hemorrhagic cystitis. PCR quantitative detection for BK Polyomavirus (226000 copies/ml) and Adenovirus (> 1000000 copies/ml) were positive. The child was treated with hyperhydration, a short course of hemodialysis, fresh frozen plasma, intravenous immunoglobulins (IVIG), antithrombin III drug (Atenetiv), and periodic bladder lavage. Cyclophosphamide treatment was not administered prior to macrohematuria and postponed until the resolution of the nephrological condition. Renal insufficiency quickly resolved, but macrohematuria persisted for 33 days, significant proteinuria (> 0.12 g/L) lasted for one week.
Conclusions: Acute renal failure and hemorrhagic cystitis were probably due to hematologic treatment and subsequent simultaneous BK and Adenovirus viral infections.

EP-267 FROM A HARMLESS IV CATHETER TO LIMB NECROSIS: A THROMBOEMBOLIC COMPLICATION IN AN INFANT WITH NEPHROTIC SYNDROME

Eline Hermans, Johan Vande Walle, Agnieszka Prytula, Joke Dehoorne, Evelyn Dhont, Evelien Snauwaert, Lien Dossche, Ann Raes

Ghent University Hospital

Introduction: Thromboembolism is a well-known complication in nephrotic syndrome (NS) patients. The reported incidence of thromboembolism in nephrotic children is lower than in adults, ranging from 1.8 to 4.4%. This report discusses a thromboembolic complication in an infant with congenital NS.
Material and methods: We present the case of a boy of two months old who had been recently diagnosed with congenital NS. During the initial in-hospital management of the severe proteinuria and hypoalbuminemia (including low-molecular-weight heparin), an transient episode of sudden paleness and coldness of the left forearm occurred. The day after this episode, the same signs reappeared with additional marbling of the skin. A pulsatile flow over the IV catheter in the left arm was noted, confirming its accidental intra-arterial position. A Doppler ultrasound showed a thickened arterial wall at the site of the catheter. Despite administration of a continuous heparin infusion, he developed compartment syndrome of the forearm for which multiple fasciotomies were performed. Necrosis of the skin of the forearm and the tip of the index finger occurred. Extensive wound care, silicone and pressure garment therapy were continued until 1 ½ year after the incident. The boy was treated with acetylsalicylic acid until the age of 22 months.
Results: This case describes an arterial thromboembolic complication in an infant with NS. Thromboembolic complications are generally venous, whereas the occurrence of (peripheral) arterial thromboembolism in NS is rare. However, multiple cases have been described in the literature, each associated with significant morbidity and/or mortality. The precedent of an arterial puncture appears to be an important risk factor.
Conclusions: Thromboembolisms are a serious complication in children with NS. A high index of suspicion is required as the clinical features may be subtle. In general, physicians should recognize the risk of (accidental) arterial punctures, especially in patients with additional risk factors.

EP-268 A CASE OF SECONDARY PSEUDOHYPOALDOSTERONISM CAUSED BY ACUTE URINARY TRACT OBSTRUCTION

Lieselot Peremans[1], Johan Vande Walle[1], Lien Dossche[1], Evelien Snauwaert[1], Agnieszka Prytula[1], Joke Dehoorne[1], Caroline Jamaer[2], Mieke Waterschoot[2], Anne-francoise Spinoit[2], Eric Van Laecke[2], Ann Raes[1]

[1]Department Of Paediatric Nephrology, Ghent University Hospital, C. Heymanslaan 10, Ghent, Belgium, [2]Department Of Paediatric Urology, Ghent University Hospital, C. Heymanslaan 10, Ghent, Belgium

Introduction: Congenital anomalies of the kidney and urinary tract (CAKUT) count for 20 to 30 percent of all anomalies identified in the prenatal period. Different defects often occur in one child, and defects can be both unilateral and bilateral. We present a case of a neonate with CAKUT presenting with pseudohypoaldosteronism due to obstructive uropathy.
Material and methods: Case description.
Results: Prenatal evaluation diagnosed a boy with multicystic dysplasia of the left kidney and hydroureteronephrosis of the right kidney. He was born full-term and ultrasound on day 2 confirmed the prenatal image. Cystography showed active vesico-ureteral reflux (grade I) on the left side and a more dilated hydroureter (grade I-II) on the right side. Due to suspicion of an obstructive megaureter combined with a declining kidney function, a ureterocutaneostomy was placed on the right side at the age of 23 days. One day after hospital discharge, the patient presented on the emergency department because of anuria since a couple of hours. Ultrasound showed increase of hydronephrosis of the right kidney suggestive for obstructive uropathy and stable multicystic dysplasia of the left kidney. Lab results showed severe hyponatremia (124 mmol/L), hyperkalemia (7,1 mmol/L), decreased bicarbonate (17,5 mmol/L) with relatively high chloride (98 mmol/L) and acute kidney injury (peakcreatinine 2,52 mg/dL). There were no signs of infection. After acute management of hyperkalemia, the patient was urgently transferred to surgery for a percutaneous nephrostomy. The child recovered with normalization of electrolyte disturbances and kidney function.
Conclusions: We discuss a male neonate presenting with a life-threatening condition with acute kidney injury and pseudohypoaldosteronism due to obstructive uropathy. Although this phenomenon is well-documented in children with obstructive uropathy and urinary tract infections, the risk is unneglectable after urological surgery in patients with unilateral functioning kidney.

EP-269 EARLY BIOLOGICAL THERAPY IMPROVES THE PROGNOSTIC IN COMPLEMENT ACTIVATION MEDIATED RENAL DISEASES? CASE REPORTS FROM A SINGLE CENTER IN WEST ROMANIA

Ruxandra Maria Steflea[1], Ramona Stroescu[1], Flavia Chisavu[1], Gabriela Doros[1], Mihai Gafencu[1]

[1]"victor Babes" University Of Medicine And Pharmacy, Timisoara, Romania, [2]Louis Turcanu Emergency Clinical Hospital For Children, Timisoara, Romania.

Introduction: We will present three cases of complement activation mediated renal diseases and the importance of early biological therapy.
Material and methods: Between 2020-2021 three patient, 1 male (age 17) and 2 female (age 5 and 6), where admitted in our hospital, from Timisoara Romania. At admittance all patients presented with acute renal insufficiency, severe anemia with the presence of schizocytes on the peripheral blood smear, high LDH levels, thrombocytopenia and sever hypertension. One girl had also bloody diarrhea. All patients underwent clinical evaluation and full blood work, in order to form a positive diagnosis.
Results: Renal replacement therapy was mandatory in all 3 cases with poor management of the malignant hypertension. Two patients were further investigated for atypical hemolytic uremic syndrome and chronic hemodialysis was necessary. The other patient was initially diagnosed with hemolytic syndrome and after 88 hours of CVVHDF was stopped and renal functions had resumed, but she had persistent bloody stools and needed several blood transfusions. Blood samples were sent to a specialized laboratory in order to analyze the complement pathway. The results concluded a complement dysregulation in the female patients secondary to sepsis and only one patient was confirmed with atypical hemolytic uremic syndrome. All patients started biological therapy with eculizumab which lead to the normalization of the blood pressure, resolution of the anemia and overall better quality of life. The male patient underwent a successful kidney transplant and no longer needs hemodialysis. One patient is currently waiting for a kidney transplant, the last one is in clinically stable.
Conclusions: Early diagnosis and prompt biological therapy improves the prognostic in complement activation mediated renal diseases. Further studies are mandatory to better understand the role of complement activation in severe infections.

EP-270 TWO CAUSES OF HYPONATREMIA; SYNDROME OF INAPPROPRIATE SECRETION OF ANTIDIURETIC HORMONE AND PSHYCOGENIC POLYDIPSIA

Asli Çelebİ Tayfur[1], Zulal Özdemir Uslu[2], Deniz Yilmaz[3], Didem ArdiÇli[3], Mine Tinmaz[2]

[1]University Of Health Sciences, Ankara KeÇiÖren Training And Research Hospital, Department Of Pediatric Nephrology, [2]University Of Health Sciences, Ankara KeÇiÖren Training And Research Hospital, Department Of Pediatrics, [3]University Of Health Sciences, Ankara KeÇiÖren Training And Research Hospital, Department Of Pediatric Neurology

Introduction: The syndrome of inappropriate secretion of antidiuretic hormone (SIADH) is a cause of euvolemic hyponatremia due to nonphysiologic stimuli for arginine vasopressin production in the absence of renal or endocrine dysfunction. Psychogenic polydipsia is a disturbance in thirst control not caused by impairment in production or release of ADH.
Material and methods: Two boys aged 12 years and 16 years (Case 1 and Case 2, respectively) admitted to Pediatric Emergency Department because of convulsion and impaired orientation and cooperation. Case 1 had been followed up for epilepsy and Tourette syndrome and receiving levetiracetam and oxcarbazepine. Case 2 had been receiving antibiotic treatment for dental abcess and he had drunk 5 lt of water in the last 4 hours before admission in order to alleviate his toothache.
Results: On physical examination an ecchymotic area with a diameter of 5 cm on the left parietal region was observed in Case 1. Laboratory analysis showed hyponatremia, decreased plasma osmolality and inappropriately concentrated urine in both patients. The cranial computed tomograpy images revealed a hematoma on left posterior parietal scalp region and density difference was consistent with hemorragic contusion in right lateral frontoparietal region in Case 1. SIADH was considered in both patients. Fluid intake was restricted and the antiepileptic drug treatment was rearranged in Case 1. Intravenous (IV) hypertonic saline was administered in Case 2 and fluid restriction and IV furosemide (1 mg/kg/dose) were ordered. Their symptoms, general status and serum sodium concentrations were gradually improved.
Conclusions: Head trauma, pain and antiepileptic drug treatment in Case 1 and psychogenic polydipsia, enfection and pain in Case 2 were considered as the causes of SIADH. Patient's symptom severity, serum sodium concentration and the status of the condition (acute/chronic) are the clues to determine the treatment modality in SIADH.

EP-271 REFRACTORY HYPERTENSION AS A COMPLICATION OF MEDICAL NEPHRECTOMY IN A CHILD WITH NEPHROTIC SYNDROME DUE TO NPHS2 MUTATION

Gizem Yildiz, Meral Torun Bayram, Alper Soylu, Salih Kavukcu

Dokuz EylÜl University Medical Faculty, Department Of Pediatric Nephrology, Izmir, Turkey

Introduction: Non-invasive medical nephrectomy by using NSAID, ACE inhibitors or calcineurin inhibitors decrease urine output and proteinuria resulting in rise of serum albumin in children with nephrotic syndrome before renal transplantation. Reported side effects of this approach include hypotension, elevated liver enzymes and reversible hypertension during treatment. We report a case with persistent refractory hypertension after medical nephrectomy.
Material and methods: Case Report
Results: Steroid resistant nephrotic syndrome was diagnosed in a 1.5-year-old boy. Renal biopsy revealed FSGS. Genetic evaluation showed homozygote pathogenic NPHS2 mutation (c.353C>T, p.P118L). End stage kidney disease developed at 10 years of age. As he still had proteinuria (11 g/day) and hypoalbuminemia (1.5 g/dL) at this stage, medical nephrectomy was performed by cyclosporine A (2x75 mg), indomethacin (2x25 mg) and enalapril (1x10 mg) for 7 days before pre-emptive living related renal transplantation. After renal transplantation proteinuria resolved, but he developed severe hypertension (160/100 mmHg) that was refractory to medical therapy with combination of three antihypertensive drugs. Plasma renin (298 pg/mL; range 6-58) and aldosterone (1180 pg/mL; range 35-300) levels were greatly elevated. Graft functions including MR angiography and DMSA scan were normal. Bilateral native nephrectomy was performed after 2.5 years of follow up. Pathologically there was sclerotic glomeruli, thickened arteriolar wall, microcalcification, tubular atrophy and chronic mixed inflammatory cell infiltrates. Blood pressure, renin and aldosterone levels decreased immediately after the operation and the patient did not need antihypertensive drugs anymore.
Conclusions: Medical nephrectomy for stopping proteinuria before transplantation has been used in patients with persistent nephrotic syndrome. Transient hypertension has been reported in a case given IV cyclosporine and IV angiotensin 2 infusion. Our case indicates that the drugs used for medical nephrectomy might cause injury in native nephrotic kidneys leading to high renin hypertension after transplantation.

EP-272 THROMBOSIS IN VASCULAR ACCESS OF HEMODIALYSIS IN CHILDREN

Elif BenderlioĞlu, Yunus Murat Akçabelen, Namık Yaşar Özbek, Umut Selda Bayrakçı

Ankara City Hospital

Introduction: Different methods have been considered to prevent complications such as infection and thrombosis in hemodialysis treatment. In long-term hemodialysis patients, arteriovenous fistula (AVF) is recommended to be preferred to central venous catheters (CVC) in adult and recently in pediatric patients because of less side effects.
The aim of this study is to analyze the thrombosis and affecting factors according to types of vascular access in pediatric hemodialysis patients.
Material and methods: Information of children who received outpatient or inpatient hemodialysis treatment in our hospital between 2020-2021 was reviewed retrospectively.
Results: Of the 80 patients, 46 (57.5%) were male, and the median age was 13±4.7 years. While 41 of the cases (51.3%) were acute renal failure, the others were chronic renal failure. Fifteen cases with a CVC for less than 5 days were excluded from the analysis. Thrombosis was detected in 8 of 54 patients with CVC and in 6 of 11 patients with AVF ($p<0.05$). Three of the AVF's had thrombosis within 3 months while the other 3 were seen in the later stages. Although 75% of cases with thrombosis of CVC were male the difference was not significant regarding the gender. 78% of thrombosis cases were patients who had an vasculer access for dialysis for more than 3 weeks. No significant association was found between age, location of CVC, acute-chronic renal failure, duration of the catheter USAge and the results of genetic analysis related to thrombosis.
Conclusions: Thrombosis in CVCs was found mostly in men and the risk is increasing when the duration of the catheter USAge was more than 3 weeks. Due to insufficient number of patients with AVF, further studies are needed.

EP-273 LATE ONSET OF ANCA POSITIVITY AND ARTHRITIS LEVAMISOLE-INDUCED:A CASE REPORT

Silvia Bernardi[1], Samantha Innocenti[2], Cassandre Cremades[3], Marina Charbit[3], Olivia Boyer[4]

[1]School Of Nephrology, Università Degli Studi Di Milano, Asst Papa Giovanni Xxiii, Bergamo, Italy, [2]School Of Nephrology, Università Degli Studi Di Firenze, Aou Meyer, Italy, [3]Pediatric Nephrology Department, Marhea Reference Center, Ap-hp, Hôpital Universitaire Necker-enfants Malades, Paris, France, [4]Centre Du Syndrome Néphrotique De L'enfant Et De L'adulte, Service De Néphrologie Pédiatrique, Institut Imagine, Hôpital Necker Enfants Malades, Ap-hp, Université De Paris, Paris, France

Introduction: Levamisole is effectively used in steroid-dependent nephrotic syndrome(SDNS).Recent studies demonstrated Levamisole-induced ANCA positivity and vasculitis associated with a broad spectrum of clinical manifestations,mostly occurring in case of prolonged use.
Material and methods: We present the case of a fifteen-year-old boy with history of SDNS treated with Levamisole as a steroid-sparing agent,who developed ANCA positivity and arthritis after almost 10 years of treatment.
Results: This boy was diagnosed with nephrotic syndrome in 2010 at the age of four years.In 2012,because of steroid-dependency,Levamisole was introduced at the dose of 2.5 mg/kg/48h,in association with low-dose alternate-day steroids.It was well tolerated.While on therapy only four uncomplicated relapses occurred.On December 2021 the patient complained of severe joint pain affecting hands and feet and peripheral edema with high fever.Urinary dipsticks were negative and neither respiratory nor dermatologic involvements were reported.Immunofluorescence tests showed high titer of ANA and ANCA confirmed with both MPO(37 UI/ml,N<3.5 UI/ml) and PR3 positivity(3.8 UI/ml,N<2 UI/ml) at FEIA method. Liver function and haematological results were normal. No positive history of cocaine consumption was reported.Levamisole was then interrupted, assuming that the manifestations were drug induced, obtaining prompt remission of symptoms.No relapse occurred after Levamisole cessation.ANCA antibodies will be monitored, since persistently elevated titers have been reported up to several months after discontinuation.
Conclusions: Our aim is to raise awareness on this rare complication and illustrate that Levamisole-induced ANCA vasculitis may have a late onset,even after ten years of treatment.Current literature lacks of univocal guidelines establishing timing of ANCA monitoring,leading to a huge variability between centers.Further studies are required to determine the appropriate duration of Levamisole therapy and frequency of titration.

EP-274 A RARE CASE OF RENOVASCULAR HYPERTENSION IN A CHILDREN WITH SCHIMMELPENNING SYNDROME

NilÜfer GÖknar[1], Sema Yildirim[3], Sabriye Gülçin Bozbeyoğlu[2], Diana Üçkardeş[1], Emre Keleşoğlu[1], Cengiz Candan[1]

[1]Istanbul Medeniyet University Department Of Pediatric Nephrology, [2]Istanbul Medeniyet University Department Of Radiology, [3]Istanbul Medeniyet University

Introduction: Schimmelpenning syndrome (OMIM 163200) is defined by association of nevus sebaceous with cerebral, ocular or skeletal defects. Ninety-five percent of cases with this syndrome has mutations in HRAS gene and 5% in KRAS gene.
Vascular anomalies (coarctation of aorta, aortic aneurysm, renal artery, and carotid stenosis, chylothorax, and lymphedema) were reported with a frequency of 12.6 to 33%. Renal artery stenosis was reported in only one patient in the literature up to our knowledge.
Material and methods: Herein we report a girl with Schimmelpenning syndrome and renovascular hypertension.
Results: A 12 years-old girl admitted to outpatient clinic with fatigue, headache and, palpitation. Her symptoms had started six months ago and aggravated in last one week. Physical examination included, heart rate was 98/min, blood pressure was 138/69mmHg, a 15cm length hyperpigmented lesion on left cervical lesion, hairless region on scalp and, sclerocornea on left eye. She was followed up from an ophthalmologist with a diagnosis of Schimmelpenning syndrome. No blood pressure measurement was done previously. ABPM revealed mean blood pressure was 132/74mmHg (>95th percentile) and 74% systolic and 40% diastolic blood pressure load. Echocardiographic investigation and renal doppler ultrasonography were normal. MRI and CT angiography showed left renal artery stenosis with a 19x12mm aneurysmatic dilatation. On follow up amlodipine was given and then blood pressure was normalized. Conventional angiography was performed and showed left renal artery narrowing. Balloon dilatation failed to relieve obstruction and she is still having antihypertensive treatment. Genetic analysis of H-RAS was negative. Biorad CFX Connect Real Time PCR revealed K-RAS mutation on fourth exome.
Conclusions: Schimmelpenning syndrome is a neurocutaneous syndrome and the knowledge on vascular anomalies is limited. Children with neurocutaneous syndrome has high risk of having renovascular hypertension and routine blood pressure controls must be done.

EP-275 A LATE ONSET UVEITIS AFTER TUBULOINTERTITIAL NEPHRITIS AND UVEIT (TINU) SYNDROME

Ece Demirci Bodur[1], İbrahim GÖkÇe[1], Sezin Bayraktar[2], Serim Pul[1], Özde Nisa TÜrkkan[1], SerÇin GÜven[1], Neslihan ÇiÇek[1], Nurdan Yildiz[1], Harika Alpay[1]

[1]*Marmara University Pediatric Nephrology*, [2]*Marmara University Paediatrics*

Introduction: Tubulointerstitial nephritis and uveitis(TINU) syndrome is a rare subgroup of acute tubulointerstitial nephritis(TIN).It is characterized by TIN with a benign course and uveitis with relapses.While uveitis may occur approximately 2 months before tubulointerstitial nephritis,there are new-onset uveitis cases reported in the literature up to 14 months after the onset of TIN.In this case report,a patient who developed uveitis 10 months after being diagnosed with TIN is presented.
Material and methods: Case:A 16-year-old male patient presented with weakness,loss of appetite and weight loss in one month.Medical history revealed that he started fitness 2 months ago and lost 10 kilograms during last month.He did not have history of taking NSAIDs,protein powder but he used vitamin C and multivitamin pills.On physical examination,his height, weight and blood pressure percentiles and systematic examinations were normal.On laboratory tests creatinine was 1.93 mg/dl,BUN was 29 mg/dl,albumin 4.5 g/L,complete blood count,liver function tests and serum electrolytes were normal and peripheral blood smear showed no atypical cells.Urinalysis showed dansity of 1026 and protein of 1(+) on dipstick.Creatinine increased up to 3 mg/dl,urine output and blood pressure were normal.24 hour urine analysis revealed tubular proteinuria(protein 27 mg/m2/h, b-2 microglobulin 41.925 mg/day,microalbumin 66.14 mg/day). Since kidney functions did not improve,renal biopsy was performed.Pathology showed eosinophil-dominated inflammation in the tubulointerstitial area which was consistent with TIN.There was no pathological finding in his eye examination.Pulse steroid treatment followed by oral 2 mg/kg/day prednisolone treatment was started.After steroid he was hypertensive so amlodipine was added to his treatment.At sixth month follow-up,creatinine was 0.94 mg/dL and proteinuria was 3 mg/m2/h. Steroid treatment was tapered and discontinued.At tenth month follow up,eye examination was performed for hypertensive retinopathy but anterior uveitis was detected in the left eye.
Conclusions: Patients presenting with TIN should be followed up for late uveitis,especially during the first 1-year period and less frequently afterwards,even if their kidney functions are normal.

EP-276 A CASE OF C3 GLOMERULOPATHY PRESENTING WITH HENOCH-SCHöNLEIN PURPURA-LIKE CLINIC

Ece Demirci Bodur[1], İbrahim GÖkÇe[1], Gizem Dikencik[2], Serim Pul[1], Özde Nisa TÜrkkan[1], SerÇin GÜven[1], Neslihan ÇiÇek[1], Nurdan Yildiz[1], Harika Alpay[1]

[1]*Marmara University Pediatric Nephrology*, [2]*Marmara University Pediatrics*

Introduction: Henoch Schönlein Purpura(HSP) is the most common vasculitis of childhood.It is a small vessel vasculitis and characterized with an IgA-containing immune complex deposition. It typically presents with palpable purpura,abdominal pain,arthralgia/arthritis and kidney involvement.C3 glomerulopathy(C3G) is a rare glomerulonephritis which results from the abnormal systemic activation of complement pathways.It is characterized with deposition of complement components in the glomeruli.In this report,a 10 year-old boy with HSP findings and diagnosed as C3G is presented to draw attention to C3G in the differential diagnosis of HSP.
Material and methods: Case: A previously healthy 10-year-old male patient admitted with arthralgia and rash on his ankles.He had history of cough and fever for the last two weeks.On physical examination his body weight was 31 kg(50-75p),height was 134 cm(50-75p)and blood pressure was 110/70 (50-90p),he had minimal edema and purpuric lesions on both of his ankles,other system findings were normal.Laboratory tests showed serum creatinine of 0.59 mg/dl,albumin of 3.2 g/L,total leukocyte of 11600/mm^3,hemoglobin of 11.2 g/dl,thrombocyte of 343.000/mm^3.Urinalysis showed 3(+) protein on dipstick and 23 erythrocytes, mostly dysmorphic, in microscopic examination.Nephrotic range proteinuria (54 mg/m^2/h) was detected in 24-hour urine.He did not have family history for kidney or rheumotological diseases.Anti Streptolysin-O antibody level was high(911),serum C3(1.17 g/L) and C4(0.16 g/L) levels were normal,ANCA was negative,ANA was weak-positive (1/100). Although the location of the rash is not typical,renal involvement due to HSP vasculitis was considered in the initial diagnosis with this clinical and laboratory findings.Kidney biopsy was performed before starting steroid therapy to demonstrate kidney involvement and exclude other possible etiologies.Light microscopy findings showed endocapillary proliferation,lobulation in glomeruli and mesangial expansion.Direct immunoflorescent examination revealed 2(+)C3, 1(+)IgG,1(+)fibrinogen,IgA,IgM,C1q,kappa,lambda were negative.No amyloid deposition was detected.All of these findings were consisted with C3 glomerulopathy.After one course of intravenous 20 mg/kg pulse steroid,2 mg/kg oral prednisolone treatment was started with enalapril.On follow up,proteinuria decreased and renal functions were normal. The patient, who is in complete remission, is still being followed up.
Conclusions: It should be kept in mind that C3G may be the etiology in children with nephrotic range proteinuria and vasculitis-like findings triggered by infection.

EP-277 RARE VARIANT OF THE PKD1 GENE IN A PATIENT WITH POLYCYSTIC KIDNEY DISEASE (PKD) – CASE REPORT

Joanna Milart, Malgorzata Placzynska, Katarzyna Jobs

Military Institute Of Medicine

Introduction: Autosomal dominant polycystic kidney disease (ADPKD) is caused by the mutations in PKD1 (16p13.3) and PKD2 (4q22.1) genes which account for approximately 78% and 15% of affected individuals. PKD1-linked ADPKD has the manifestations of renal cysts, liver cysts, and intracranial aneurysm. Other clinical symptoms include: early-onset hypertension, abdominal fullness and pain, nephrolithiasis, hematuria and urinary tract infections (UTIs) They are usually observed decades before the onset of renal insufficiency. The most serious renal complication is end-stage renal disease.
Material and methods: Case presentation Presented female patient was diagnosed with renal cysts at age of 11 by the ultrasound performed due to abdominal pain. Parents and other family members didnt have any history of policystic kidney disease. Renal cysts were confirmed by computed tomography and kidney scintigraphy. There was no evidence of liver cysts, nephrolithiasis, hematuria, urinary tract infections hypertension or renal insufficiency. The patient remained under the constant care of pediatric nephrologist.
Results: At the age of 17 the girl had a genetic test which showed no pathogenic or potentially pathogenic variants. However a very rare variant with unknown pathogenicity was found in the PKD1 gene. Next Generation Sequencing revealed a c.3161+53C>G variant located in intron_13/45. The estimated frequency of this variant in Europe is 0,0060.
Conclusions: Mutations of PKD1 are associated with most cases of autosomal dominant polycystic kidney disease. Many pathogenic variants are already known. There are still new variants detected that require further analysis.

EP-278 BIOMARKERS OF KIDNEY INJURY IN CHILDREN WITH LEUKEMIA AFTER ANTICANCER THERAPY

Mariya Skrylnikova, Olga Zhdanova, Tatiana Nastausheva, Elena Kulakova, Liliya Stahurlova, Inna Kondratjeva, Anna Khan

Voronezh State Medical University Named After N.n. Burdenko

Introduction: Early diagnosis of kidney disorder in children with leukemia who have received anti-cancer treatment is essential in prevention of development and progression of chronic kidney disease. The aim of our research was to study the markers of kidney damage after completion of polychemotherapy and radiation therapy.
Material and methods: A study of 39 children (22 boys and 19 girls, aged 10.7 ± 3.7y.) with acute lymphoblastic leukemia (ALL) who completed anticancer therapy was carried out. The patients were divided into 3 groups: 1-st group with a period of 2 weeks to 2 years from completion of the therapy; 2-nd group with a period of 2 years to 4 years from completion of the therapy, and 3-d group with a period of 4 to 6.5 years from completion of the therapy. The control group consisted of 50 children (25 boys and 25 girls) aged 10.7±4.8 years old. The results were presented as a median and interquartile range [IQR].
Results: Children of the 1-st group had the level of urinary KIM -1 - 323.19 pg / ml [150.43-888.55], which was significantly higher according to the control group: 162.35 pg/ml [95.85-253.95], p=0.009. Urinary β2-m /UCr - 0.85 mkg / mg [0.35-7.55], urinary IL -18 /UCr - 26.36 pg/mg [17.32-39.05] were even lower than in control group; β2-m /UCr - 4.63 mkg/mg [1.75-9.43], p=0.035, IL-18 /UCr - 44.86 pg/mg [35.03-58.15], p= 0.018. All urinary markers in children from 2-nd and 3-d groups did not differ from those in the control group. The level of cystatin C in blood serum in all three groups of patients was higher than in the control group 0.47 mg/l [0.43-0.53]: 1-st group - 0.62 mg/l [0.54-0.86], p<0.001; 2-nd group - 0.6 mg/l [0.48-0.64], p=0.024; 3-d group - 0.57 mg/l [0.51-0.63], p=0.004.
Conclusions: In the first two years after completion of anticancer therapy children with ALL had an increased level of urinary KIM -1. The level of cystatin C in the blood serum after completion of the polychemotherapy and radiation therapy remains elevated during 6 years.

EP-279 DGKE MUTATION: A RARE CAUSE OF STEROID RESISTANCE NEPHROTIC SYNDROME

Mehtap Akbalik Kara, BeltİnGe DemircioĞlu KiliÇ, Mithat BÜyÜkÇelik, AyŞe Balat

Gaziantep University, Department Of Pediatric Nephrology

Introduction: Steroid resistance nephrotic syndrome (SRNS) is a heterogeneous group of diseases in children which is divided into two as immune-based and monogenic SRNS. When compared to recent years, the prevalance of genetic studies in the form of larger panels ensure the diagnosis of patients and prevent unnecessary administration of immunsupressive treatments. Recent studies have shown that mutations in DGKE gene may cause either membranoproliferative glomerulonephritis (MPGN) or hemolytic uremic syndrome (HUS) in children. Here we wanted to present a patient with clinical and laboratory findings of MPGN without HUS.
Material and methods: Case presentation: The patient was the first child from consanguineous parents who admitted to our hospital at the age of five because of nephrotic syndrome. She did not respond to steroid therapy and renal biopsy confirmed MPGN. Cyclosporin A, mycophenolate mofetil and tacrolimus treatments were administered. The genetic analysis for podocin, nephrin and WT1 were all normal. Cyclophosphamide and rituximab therapies were suggested but could not be given owing to a lack of consent from her parents. At the age of thirteen she was brought to hospital with headache and visual impairment. Papilledema and high pressure of cerebrospinal fluid was obtained. Brain MR angiography revealed thrombus in the superior sagittal and in the left transvers sinus. Acetazolamide and low molecular weight heparin was initiated. Heterozygous MTHFR and homozygous PAI-1 mutation was detected. When glomerular genetic study re-evaluated a homozygous mutation in DGKE gene (c.433_433 del) was obtained. She is maintained with ACE-inhibitor and anti-aggregant therapy, the glomerular filtration rate is 117 ml/min/1.73 m^2, albumin level is 3.4 mg/dl with nephrotic proteinuria.
Results: No
Conclusions: We believe that it would be useful to re-evaluate the patients with wide genetic analysis methods who are thought to have genetic origin for nephrotic syndrome.

EP-280 USING CORTICOSTEROIDS IN THE TREATMENT OF ACUTE TUBULAR NECROSIS: EFFECTİVE OR NOT?

İbrahim GÖkÇe, Serim Pul, Ece Demirci Bodur, Özde Nisa TÜrkkan, SerÇin GÜven, Neslihan ÇiÇek, Nurdan Yildiz, Harika Alpay

Marmara UnİversİTy, PedİatrİC Nephrology

Introduction: Acute tubular necrosis (ATN) is the most common etiology of intrarenal acute kidney injury(AKI). Acute tubular necrosis is usually self-limited, has no specific treatment, and largely improves with the elimination of the underlying cause. Limited data suggest that steroid therapy may be beneficial in selected cases accompanied by tubulointerstitial inflammation. In this report, a boy with ATN secondary to sepsis and responsive to steroid treatment is presented.
Material and methods: Case A five-year-old male who had neurogenic bladder and normal kidney function, presented with fever, vomiting and bloody diarrhea. He was admitted to the intensive care unit due to septic shock. On the second day of admission, hemodialysis was started due to oliguric course, electrolyte imbalance and volume overload. In addition to AKI, signs of multiple organ failure(MOF) such as severe cholestasis and respiratory failure were also present. Despite improvement of clinical signs of septic shock and MOF, the need for hemodialysis continued after the eighteenth day. Kidney biopsy was performed to identify the etiology of prolonged and severe kidney damage. Light microscopy showed normal glomeruli, focal damage in the tubules and moderate inflammatory cell infiltration in the tubulointerstitial area. Because of acute severe renal impairment seen in our patient who had previously normal kidney function and ATN findings that accompanied by normal glomerular structure and modearate tubulointerstisial inflammation, 250 mg pulse methylprednisolone was given and treatment was continued with 2 mg/kg/day oral methylprednisolone. On the fifth day of treatment, a significant increase in urine output and an improvement in kidney function were observed. Hemodialysis was stopped at the end of the first month of treatment. Steroid treatment was tapered and discontinued within 2 months. He is still being followed up as stage-3 CKD.
Conclusions: We wanted to emphasize that short-term corticosteroid therapy may be beneficial in refractory and severe ATN cases, if the glomerular structure is intact, if the signs of chronicity are not evident and if moderate to severe tubulointerstitial inflammation is present in histopathological examination.

EP-281 SARS-COV 2 DISEASE IN A PEDIATRIC DIALYSIS UNIT

Gabriele Malgieri, Vittorio Serio, Luigi Annicchiarico Petruzzelli, Daniela Molino, Rosamunda Darcangelo, Bruno Minale

Aorn Santobono-nephrolgy And Dialysis Unit

Introduction: Data about COVID 19 in children on dialysis are scarce. A retrospective study was performed from March 2020 until January 10/ 2022, of childrens (17 pts) on chronic hemodialysis in our Dialysis Unit,were diagnosed with SARS-CoV-2.

Material and methods: For inclusion in the study,SARS-CoV-2 polymerase chain reaction (PCR) result had to be positive. Both inpatients and outpatients were included.A positive contact was present 5/7 pts.
7/17 children (41.1%), were found by PCR to be positive for SARS-CoV-2: 3 M and 4 F (median age 17.7 y).Underlying disease:2 Nephronophthisis,1 Alport S.,1 IgA Nephropathy,3 Renal dysplasia.
Results: 3 pts were asymptomatic;3 pts were febrile;4 pts with cough and rhinorrhea;1 pts with mild dyspnea and abnormal X-ray.None needed respiratory support.
Antibiotics and steroids were administered,respectively to 3 and 2 pts.
68 dialytic treatment were performed in isolated room;the target of Urea Reduction Rate >70% and kt/v >1.2 in all pts was achieved.
Mild intradialytic hypotension in 3 pts..None fluid overload was noticed.
Conclusions: In our experience,COVID 19 disease in children on dialysis show a similar course as in healthy children,with mild symptoms.
Our pts show no comorbidity (Lung and immune disease),and this could justify a better prognosis.

EP-282 CLINICAL AND EVOLUTIVE ASPECTS OF CHILDREN WITH PRIMARY FOCAL SEGMENTAL GLOMERULOSCLEROSIS: A TUNISIAN SERIES

Abir Boussetta, Khouloud Ben Njima, Nesrine Abida, Farah Krifi, Manel Jellouli, Tahar Gargah

Pediatric Nephrology Department, Charles Nicolle Hospital Tunis, Tunisia

Introduction: To evaluate the epidemiological, clinical, biological and evolutive features of primary Focal Segmental Glomerulosclerosis (FSGS) in Tunisian children.
Material and methods: This was a retrospective study conducted in the pediatric nephrology department of the Charles Nicolle Hospital in Tunis, Tunisia over a period of 20 years, from January 2001 to December 2020. Inclusion criteria were: patients less than 18 years of age at the time of diagnosis of the disease, regularly followed up with renal biopsy-proven FSGS. Exclusion criteria were: children with suspected or proven secondary FSGS.
Results: Au total of 35 patients were included in our study, there were 19 boys and 16 girls (sex-ratio:1.18). The median age at diagnosis was 4.5 ans, the disease started before the age of 3 in 34.3% of cases and after the age of 10 years in 25.7% of cases. A family medical history of kidney disease other than FSGS was reported in 33% of cases. The initial clinical presentation was pure nephrotic syndrome in 88% of cases, acute nephritic syndrome in 3% and an isolated asymptomatic proteinuria in 9% of cases. Initial treatment was corticosteroid therapy, a total of 22 cases of corticosteroid-sensitive nephrotic syndrome, 9 cases of corticosteroid resistant nephrotic syndrome and 4 cases of spontaneous remission were recorded. The outcome of our patients was as follows: 15 children (42.8%) had chronic renal failure (GFR<90ml/mn/1.73 m^2SBA) including end-stage renal disease (ESRD) in 4 cases (11.5%). One patient was transplanted without recurrence of the FSGS on the renal graft.
Conclusions: FSGS is a rare but under-diagnosed renal pathology in children. Early diagnosis allows to adapt therapeutic procedures to improve the prognosis of these patients.

EP-283 A RARE COMPLICATION OF CHILDHOOD NEPHROTIC SYNDROME: PANCREATITIS

Özgür Özdemir Şimşek[1], Gökçen Erfidan[1], Seçil Arslansoyu Çamlar[2], Demet Alaygut[2], Fatma Mutlubaş[2], Belde Kasap Demir[3]

[1]Department Of Pediatrics, Division Of Nephrology, University Of Health Sciences Izmir Tepecik Training And Research Hospital, Izmir, Turkey, [2]Izmır Faculty Of Medicine, Department Of Pediatrics, Division Of Nephrology, University Of Health Sciences Turkey, Izmir, Turkey, [3]Department Of Pediatrics, Division Of Pediatric Nephrology And Rheumatology, Izmir Katip Çelebi University, Izmir, Turkey

Introduction: Nephrotic syndrome (NS) has major complications such as infection, thromboembolism, renal failure, anasarca, and hypovolemia. Other potential complications that may occur in children with NS are persistent hyperlipidemia, anemia, abnormal endocrine tests with normal functions. We present acute pancreatitis due to dyslipidemia in a patient with NS.
Material and methods: A 9-year-old female patient was being followed up with the diagnosis of steroid-resistant NS for the last 8 months. She was on tacrolimus, lansoprazole, prednisolone, and furosemide treatment. She was hospitalized to transfuse albumin due to oliguria and edema. Patient growth was normal. She had ascites and (+1) pretibial pitting edema. The patient, who was scheduled to be discharged after albumin transfusion, suddenly developed chest pain, stomachache and vomiting. Laboratory tests revealed urea 32mg/dL (N:10-38); serum creatinine 0.5mg/dL (N:0.5-1.2); albumin 1.9g/dL (N:3.5-5.5); amylase 1017U/L (N:28-100); lipase 2524U/L (N:10-140); triglyceride 1799mg/dL (N:0-150); and total cholesterol 600mg/dL (N:110-169). Computed tomography was compatible with acute pancreatitis, which was attributed to hyperlipidemia due to NS in the patient. Viral markers were negative.
Results: The patient with normal liver function tests and no bleeding disorder was treated with fresh frozen plasma 3 times at a dose of 10cc/kg/dose to provide lipoprotein (a). Intermittent albumin infusions were administered to make the patient normovolemic. Oral gemfibrozil treatment was initiated. At the end of 72 hours, triglyceride level was 375mg/dL, amylase decreased to 238U/L, and lipase decreased to 342U/L. Physical examination was completely normal.
Conclusions: Acute pancreatitis should be kept in mind even in childhood for sudden onset abdominal pain in NS patients with hyperlipidemia, who have multiple medications, especially those using tacrolimus. Our case is the youngest patient with acute pancreatitis due to NS.

EP-284 PREVALENCE OF GROWTH RETARDATION AND UNDERWEIGHT IN CHILDREN WITH CHRONIC KIDNEY DISEASE IN SINGLE CENTER

NilÜfer GÖknar[1], Diana ÜÇkardeŞ[1], Elif Nur Akkoca[2], Serap Toprak[2], Mustafa Deveci[1], Emre KeleŞoĞlu[1], Cengiz Candan[1]

[1]Istanbul Medeniyet University Department Of Pediatric Nephrology, [2]Istanbul Medeniyet University

Introduction: Malnutrition is common among patients with chronic kidney disease (CKD), especially with end stage kidney disease. Prevalence changes between 5-80% in different patient populations. Inadequate nutritional intake, uremia, urinary protein loss, removal of amino acids in dialysate and chronic inflammation are the most important reasons of malnutrition in children with CKD.
Material and methods: A cross-sectional study was carried out in patients aged 10.82±4.78years (range; 1-17years) who were diagnosed CKD for at least for 3 months. From January 2021 to December 2021, 70 children (33 boy) were included. Eleven children were on hemodialysis and two were on peritoneal dialysis. The causes of CKD were dysplasia/hypoplasia (n:20), uropathy (posterior urethral valve and/or vesicoureteral reflux n:12), neurogenic bladder (n:9), asphyxia/hypoxia (n:4), hemolytic uremic syndrome (n:4), nephronophthisis (n:2), HNF1-B mutation (n:2), PAX-2 mutation (n:2), polycystic kidney disease (n: 6), primary hyperoxaluria (n:1), HUPRA (n:1), cystinosis (n:1), secondary

to chemotherapeutics (n:1), chronic tubulointerstitial nephritis (n:1) and, unknown (n:4).
Anthropometric indices of the patients including height and weight were measured. The standard deviation scores (SDS) were computed from the references for Turkish children. Also, body mass index (BMI) was calculated and SDS scores and percentiles were computed. Blood urea, creatinine, and serum albumin levels were evaluated.
Results: Mean height SDS was -1,28±1,87 (IQR -2,29- -0,10), mean weight SDS was -1,14±1,99 (IQR -2,5-+0,13), mean BMI SDS was -0,64±1,87 (IQR -2,75- +0,64). Sixteen children (27%) were underweight (<5th percentile) (4/13 with CKD stage 5, 6/12 with CKD stage 4); seven children were overweight (>85th percentile <95th percentile) and, three was obese (≥95th percentile). Nineteen children (5/13 with CKD stage 5, 4/12 with CKD stage 4) had short stature (<-2SDS). All children had normal serum albumin levels.
Conclusions: Children with chronic kidney disease are at high risk of malnutrition and short stature and routine anthropometric measurements must be done in each clinical visit.

EP-285 EARLY CYSTEAMINE TREATMENT FOR OF NEPHROPATHIC CYSTINOSIS: RENAL OUTCOME OF RUSSIAN CHILDREN

Valentina Maltseva, Petr Ananin, Tatyana Vashurina, Kirill Savostyanov, Alexander Pushkov, Olga Zrobok, Andrey Fisenko, Alexey Tsygin

Federal State Autonomous Institution "national Medical Research Center For Childrens Health" Of The Ministry Of Health Of The Russian Federation

Introduction: Nephropathic cystinosis is an inherited autosomal recessive disease that leads to early-onset chronic renal failure in consequence to accumulation a lysosomal cystine in cells caused by mutations in the CTNS gene. Early initiation of cysteamine delays progression to end stage kidney disease (ESKD).
Material and methods: Material and methods: Retrospective analysis of renal function of 32 children with nephropathic cystinosis (17 male, 53%) diagnosed in our Center in the period 2008-2021. We analyzed the progression to ESRD in initiated cysteamine treatment groups A (1.0-2.5 years; n=13, 40.6%), B (2.6-5.0 years; n=5, 15.6%), C (after 6.0 years; n=4, 12.5%) and D (without cure; n=10, 31.3%). Renal survival probability rates were calculated according to Kaplan-Meier, log-rank test to compare survival curves.
Results: Median age at initiating of cysteamine therapy was 1.7 years in A group (IQR: 1.1 – 2.1; range: 0.8 – 2.4); median was 3.0 years in group B (IQR: 2.8 – 3.1), median was 5.9 years in group C (IQR: 5.7 – 6.3), median was 8.7 years (mean 11.1 years; IQR: 7.8 – 12.5, range 6.0 - 26.5) in group D. Twenty one (66%) children reached ESRD at mean 10.5 years (median: 9.6; IQR: 8.2 – 13.3; range: 6.5 – 15.4), of which 16 (50%) patients had a kidney transplantation, not including 3 deaths. Log-rank analysis showed that early starting cysteamine therapy significantly delayed the ESRD onset (p = 0.032), median survival time in A group was 11.8 years (95%Cl 8.0 – 15.6) vs. 8.9 years (95%Cl 2.5 – 15.3) in B group vs 7.7 years (95%Cl 5.4 – 10.1) in C group vs. 7.8 years in group D (95%Cl 7.2 – 8.3), respectively.
Conclusions: Orally initiation of cysteamine significant the delays ESKD in children with cystinosis

EP-286 EARLY CYSTEAMINE TREATMENT FOR NEPHROPATHIC CYSTINOSIS: RENAL OUTCOME OF RUSSIAN CHILDREN

Valentina Maltseva, Petr Ananin, Tatyana Vashurina, Kirill Savostyanov, Alexander Pushkov, Olga Zrobok, Andrey Fisenko, Alexey Tsygin

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Conclusions: Orally initiation of cysteamine significant the delays ESKD in children with cystinosis

EP-287 COMPARISONS OF OFFICE AND 24-HOUR AMBULATORY BLOOD PRESSURE MONITORING IN CHILDREN WITH CHRONIC RENAL FAILURE

Serra Sürmeli Döven[1], Esra Danacı Vatansever[1], Dilek Er[1], Gülistan Kibar[1], Derya Karpuz[2], Ali Delibaş[1]

[1]Mersin University Faculty Of Medicine, Department Of Pediatric Nephrology, [2]Mersin University Faculty Of Medicine, Department Of Pediatric Cardiology

Introduction: Hypertension is a frequent complication in patients with Chronic Renal Failure (CRF). This study aimed to compare Office Blood Pressure Measurement (OBPM) and 24-Hour Ambulatory Blood Pressure Monitoring (ABPM) for diagnosis of hypertension in patients with CRF.
Material and methods: Twenty eight patients with CRF who were followed-up at Pediatric Nephrology Department of Mersin University Faculty of Medicine between 2017- 2022 were included in the study. Medical information and laboratory results at last admissions of the patients were garnered from records, retrospectively. All patients underwent OBPM and ABPM. Mean cystolic and daistolic blood pressures for ABPM and OBPM were compared by using Student's t-test.
Correlation between OBPM and ABPM were measured by Pearson Correlation analysis.
Results: In the 28 children enrolled, 22 were female (78.6%) and 6 (21.4%) were male. Mean age of the patients was 28±25.48 (117-208) months. The etiologies of CRF were glomerulonephritis (17.9%), neurogenic bladder (17.9%), cystic kidney diseases (14.3%), vesicoureteral reflux (14.3%), thrombotic microangiopathies (7.1%), cystinosis (7.1%)

and unknown etiology (17.9%). Mean cystolic and diastolic blood pressures for daytime ABPM were 124.96±14.94 mmHg and 59.00±12.84 mmHg. Mean cystolic and diastolic blood pressures for nighttime ABPM were 111.89±25.87 and 72.07±15.78 mmHg. Mean cystolic and diastolic blood pressures for OBPM were 115.17±14.60 mmHg and 78.64±13.48 mmHg. Mean cystolic blood pressure values for daytime ABPM were significantly greater than OBPM (p=0.004). But cystolic and diastolic blood pressure values were not correlated for ABPM and OBPM (r=0.372, p=0.051 and r=0.263, p=0.177). Cystolic, diastolic blood pressure values for daytime and sleeptime ABPM were correlated (r=0543, p=0.003, r=0.815, p<0.001).
Conclusions: Cystolic blood pressure values for daytime ABPM were greater than OBPM but there was no correlation between two. ABPM should be used for diagnosis of hypertension in patients with CRF.

EP-288 KIDNEY DAMAGE AT CHILDREN WITH DIABETES

Mariya Petrova, Tamara Makarova, Julia Melnikova

Kazan State Medical University

Introduction: Diabetes is the leading cause of chronic kidney disease and end-stage kidney disease in the worldwide.

Objectives: To determine the frequency and the nature of kidney damage in children with diabetes according to the Children's Republic Clinical Hospital of Republic of Tatarstan of Russian Federation (CRCH).
Material and methods: We performed a post-hoc analysis of 450 case histories of 160 patients over 5 years. The indicators were analyzed: CBC, urine analysis, biochemical tests. The glomerular filtration rate (GFR) was determined by the Schwartz formula.
Results: Age of patients: 16 children (10%) 8-11 years old, 38 children (30%) 12-13 years old, 96 children (60%) 14-18 years old. 120 girls (75%) and 40 boys (25%). In 48 children (30%) there were changes in urine tests depending on the duration of the disease: changes were observed in 3 (6.25%) children with an experience of up to 1 year, in 28 (58.33%) children with an experience of 1-5 years, in 13 (27.08%) - 5-10 years, in 4 (8.34%) - over 10 years. There were no changes in the tests in 112 (70%) patients. The average level of proteinuria in children with a disease experience of less than 1 year was 0.5 g/l, 1-5 years - 1.2 g / l, 5-10 years - 1.98 g / l, over 10 years - 3.1 g / l. GFR in children with disease experience less than 1 year was 158 ml/min/1.73m2, 1-5 years - 136 ml/min/1.73m2, 5-10 years - 73 ml/min/1.73m2, over 10 years – 29 ml/min/1.73m2.
Conclusions: It was found that kidney damage, accompanied by proteinuria, in diabetes is about 32.5% and is important in the prognosis of the disease. Early detection of kidney damage in the preclinical stage is necessary to prevent further progression.

EP-289 PROGNOSTIC FACTORS IN CHILDREN WITH PRIMARY FOCAL SEGMENTAL GLOMERULOSCLEROSIS

Abir Boussetta, Khouloud Ben Njima, Nesrine Abida, Farah Krifi, Manel Jellouli, Tahar Gargah

Pediatric Nephrology Department, Charles Nicolle Hospital Tunis, Tunisia

Introduction: To identify the prognostic factors in children with primary focal segmental glomerulosclerosis (FSGS).
Material and methods: This was a retrospective study conducted in the pediatric nephrology department of the Charles Nicolle Hospital in Tunis, Tunisia over a period of 20 years, from January 2001 to December 2020. Inclusion criteria were: patients less than 18 years of age at the time of diagnosis of the disease, regularly followed up with renal biopsy-proven FSGS. Exclusion criteria were: children with suspected or proven secondary FSGS. A univariate analysis followed by a multivariate analysis was done to identify the predictive factors of progression to chronic renal failure (CRF), end-stage renal disease (ESRD), and/or death. A p-value<0.05 was significant.
Results: A total of 35 patients were included in our study, the median age at diagnosis of the FSGS was 4.5 years. The univariate study found that failure to achieve remission on corticosteroids after the first episode of nephrotic syndrome (NS) was a predictor factor of progression to CRF. Family history of kidney disease, extra-renal signs, failure to achieve remission on corticosteroids after the first episode of NS, failure to achieve remission after 3 months of treatment with cyclosporine A (CsA) were associated with a risk of progression to end-stage renal disease (ESRD). Predictive factors for death were: creatinine clearance<60 μmol/l, and end-stage renal disease. After a multivariate study, only a family history of kidney disease was found to be predictive of progression to ESRD. No predictive factors for death were found.
Conclusions: It is important to identify the predictive factors of progression to renal failure in order to adapt the therapeutic strategies to the patients and thus improve their prognosis.

EP-290 UVEITIS IN THE FOLLOW-UP OF A PATIENT WITH TUBULOINTERSTITIAL NEPHRITIS

Özgür Özdemir Şimşek[1], Gökçen Erfidan[1], Seçil Arslansoyu Çamlar[2], Demet Alaygut[2], Fatma Mutlubaş[2], Cemaliye Başaran[1], Gamze Türe[3], Belde Kasap Demir[4]

[1]Department Of Pediatrics, Division Of Nephrology, University Of Health Sciences Izmir Tepecik Training And Research Hospital, Izmir, Turkey, *[2]Izmır Faculty Of Medicine, Department Of Pediatrics, Division Of Nephrology, University Of Health Sciences Turkey, Izmir, Turkey*, *[3]Department Of Ophthalmology,university Of Health Sciences Izmir Tepecik Training And Research Hospital, Izmir, Turkey*, *[4]Department Of Pediatrics, Division Of Pediatric Nephrology And Rheumatology, Izmir Katip Çelebi University, Izmir, Turkey*

Introduction: The entity with tubulointerstitial nephritis (TIN) and uveitis, first described in 1975, is called TINU syndrome. We present a rare case of TINU who presented with tubulointerstitial nephritis and developed uveitis during the follow-up.
Material and methods: A 17-year-old male patient was hospitalized with a creatinine level of 2mg/dL(N:0.5-1.2) after prescription of amlodipine and propranolol in another hospital due to high blood pressure. The creatinine level was 1.8mg/dL a week ago. He had a body weight of 98kg(95-99p), and a height of 172cm(25-50p). Physical examination was normal and the patient was normotensive. The tubular tests revealed FeNa: 2.4%, FeK: 5.7% and TPR:84%. Parathormone and serum C3-C4 were normal; and ANA was negative. Proteinuria in 24-hour urine analysis was 20mg/m^2/h. The kidney biopsy was consistent with acute TIN. The patient received 60mg/day of oral prednisolon, which was gradually tapered and discontinued at 12 weeks. The creatinine value decreased to 1mg/dL. On the 14th day of discontinuation of steroid therapy, the patient presented with uveitis. There was no increase in the creatinine value and tubular tests were in normal range. ANA and anti-dsDNA, Brucella serology, rheumatoid factor, p-ANCA, c- ANCA, HLA-B27 were negative. HLA-B51 was positive, but the pathergy test was negative. The patient had no history of oral and genital aphthae, skin findings, erythema nodosum, neurological pathology, joint pathologies and thrombosis in terms of Behçets Disease. Serum angiotensin-converting enzyme level for sarcoidosis was within normal limits.

Results: Low dose steroid and subcutaneous methotrexate was instituted at a dose of 20mg/week. At the end of the first month, ocular findings were under control.
Conclusions: In our patient with TINU, uveitis was not simultaneous with the TIN clinic. It should be kept in mind that uveitis may develop during the follow-up in patients diagnosed with TIN and eye examinations should be repeated at regular intervals.

EP-291 FREQUENCY AND STRUCTURE OF RENAL DAMAGE IN CHILDREN WITH SYSTEMIC LUPUUS

Guzel Karymova, Igor Zorin

Orenburg Medical University

Introduction: The aim of study was to determine frequency and structure of kidney pathology in children with systemic lupus erythematosus (SLE).
Material and methods: We had retrospective analysis of 9 case histories of patients with SLE hospitalized in rheumatology department of Orenburg regional clinic hospital in period 2010-2020.
Results: We established SLE most often was in girls - 77.7% (n=7). The debut of SLE was in age of 9-12 years. Kidney damage was found in 3 patients (33%). One patient had nephrotic syndrome, second patient had chronic tubulointerstitial nephritis as a result of thrombotic microangiopathy (TMA) with CKD I. This patient underwent nephrobiopsy, which revealed histological picture of TMA: mesangiolysis, endotheliosis and thrombosis of glomerular capillaries and arterioles, fragmentation of erythrocytes; diffuse focal acute tubular necrosis. The third patient had nephrotic syndrome with hematuria and arterial hypertension, CKD I. Morphological diagnosis based on the results of nephrobiopsy: diffuse immunocomplex glomerulonephritis with "full house" immunoexpression, with slight mesangial and pronounced endocapillary hypercellularity; without crescents, without glomerulosclerosis, tubulo-interstitial fibrosis and arteriolosclerosis. Lupus - jade, class IV (A). All children with SLE had arterial hypertension by data of ABPM in night.
Conclusions: Renal damage in children is formed in severe variant of SLE. It's need to morphological and functional examination of kidneys for diagnosis verification.

EP-292 MORPHOLOGICAL PATTERNS OF STEROID RESISTANT NEPHROTIC SYNDROME WITH ONSET IN EARLY CHILDHOOD

Svitlana Fomina, Valentin Nepomnyashchy, Ingretta Bagdasarova

Si "institute Of Nephrology Of Nanm Of Ukraine"

Introduction: The aim of study was to determine the dominant morphological patterns in children with steroid resistant nephrotic syndrome (SRNS) and disease manifestation before the age of 3 years.
Material and methods:
Transcutaneous nephrobiopsy was performed in 24 nephrotic children with disease onset in early childhood (number of glomeruli in the samples: 15 (9;29)). There were verified SR after 6 weeks of steroids in standard doses.
Results: The dominance of familial NS (8/33.3%) was determined: it was clarified in one family in relatives of the third degree of kinship, with the main pattern of focal-segmental glomerulosclerosis (FSGS) and focal-global glomerulosclerosis with tubular atrophy, and interstitial fibrosis, and arteriosclerosis in 2 cases (genetic testing was performed on one of the siblings, and NPHS2 mutation found). FSGS were also identified in 6/25.0% unrelated patients (3: tip lesion; 3: NOS); genetic testing has established LMX1B mutation in one boy from 4 examined.
In 2 patients (8.3%) diffuse mesangial sclerosis were detected (in one child genetic testing confirmed WT1 mutation and nephroblastoma was diagnosed in a two years period; in the second child the mutations were not proven). In 2 children with NS course more than 12 months global glomerulosclerosis and fibroplastic changes did not allow to identify the initial morphological patterns of the disease (but in one of them COQ2 mutation was defined, in the second case a genetic testing was not performed).
In other cases we identified minimal change nephropathy (4/16.7%) and mesangial proliferation (3/12.5%) without genetic examination.
Conclusions: The advantage of genetic determined NS have documented in SR patients with disease onset before age of 3 years. But the high incidence of idiopathic NS is confirmed with probability long-term, delayed treatment effect. NS heterogeneity in this cohort makes screening for the genetic determinants advisable.

EP-293 INFANTILE NEPHROTIC SYNDROME WITH ATYPICAL PRESENTATION

Lieselot Peremans[1], Goedele Philippe[2], Lien Dossche[1], Evelien Snauwaert[1], Agnieszka Prytula[1], Joke Dehoorne[1], Johan Vande Walle[1], Ann Raes[1]

[1]Department Of Paediatric Nephrology, Ghent University Hospital, C. Heymanslaan 10, Ghent, Belgium, [2]Department Of Pediatrics, Az West, Ieperse Steenweg 110, 8630 Veurne, Belgium

Introduction: Nephrotic syndrome is defined by the clinical triad of heavy proteinuria, hypoalbuminemia and generalized edema. Infantile nephrotic syndrome can be an insidious disease. Children with atypical features should be referred for specialist pediatric nephrology assessment, including renal biopsy and genetic analysis.
Material and methods: Case description.
Results: A Caucasian boy of 1,5 years old with a blank medical history was referred because of nephrotic range proteinuria in combination with haematuria. Clinical examination showed mild periorbital edema, present for one month, and significant hypertension (> P99). Urinalysis showed proteinuria (13,6 g/g creat) and haematuria (5261/μL). Lab results showed a normal kidney function and hypoalbuminemia (21 g/L). Clinical examination showed mild periorbital edema and significant hypertension. Kidney ultrasound showed no vascular abnormalities, but bilateral enlarged kidneys with increased echogenicity and swollen aspect of the cortex. Ophthalmic screening showed no abnormalities. Cardiac screening was negative. Infectious diseases were excluded. A kidney biopsy was performed and showed an image of mesangial hypercellularity and diffuse mesangial sclerosis (DMS) with negative immunofluorescence, suggestive for an underlying hereditary cause. Genetic analysis is pending. Treatment with oral corticosteroids was initiated, and until now, proteinuria is declining progressively in this patient.
Conclusions: We present a young boy with infantile nephrotic syndrome based on DMS. It is a rare disease with a poor prognosis and rapid progression to end-stage kidney failure. The etiology and pathogenesis of DMS is not well understood, but it is associated with dominant pathogenic variants in WT1 (23,1%) and biallelic pathogenic variants in PLCE1 (17,8%), LAMB2 (13,6%) and NPHS1 (4,9%). This case shows that it is essential that small children who present with atypical features of nephrotic syndrome need specific pediatric nephrological assessment.

EP-294 SEVERE KIDNEY DYSFUNCTION IN A CHILD WHO PRESENTED WITH CONSTIPATION

Emre LeventoĞlu[1], Bahar BÜyÜkkaragÖz[1], Kibriya Fidan[1], Serhat GÜrocak[2]

[1]*Gazi University, Faculty Of Medicine, Department Of Pediatric Nephrology*, [2]*Gazi University, Faculty Of Medicine, Department Of Urology*

Introduction: Constipation is a common childhood problem, accounting for about 3-5% of all visits to the pediatrician. Most of them are functional constipation; toilet phobia is one of the rare causes. Constipation is also a condition associated with bladder dysfunction and urinary tract infections. The relationship between this abnormal bowel and bladder function is called the dysfunctional elimination syndrome (DES).
Material and methods: We present a girl who has been diagnosed with chronic kidney disease due to severe constipation, in which toilet phobia is the triggering factor.
Results: A 4-year-old girl applied to the pediatrician because of constipation. She delayed her urination and defecation due to the fear of falling into the toilet. The patient with a history of recurrent urinary tract infection had a significant increase in serum creatinine.
In physical examination; growth retardation, pallor, abdominal distention, and fecal contamination in her underwear were observed. Laboratory analysis revealed blood urea nitrogen (BUN): 65 mg/dl, creatinine: 2.01 mg/dL, hyperkalemia, hyperphosphatemia, anemia and metabolic acidosis. There was no pyuria and hematuria in the urine analysis, there was no growth in the urine culture, but the protein/creatinine ratio was high.
Colonic distention and intense gas/stool appearance on abdominal X-ray; diffuse residual fecaloids and elongated bladder on computed tomography; reflux in the both kidneys on voiding cystoureterography; and decreased cortical functions in dimercapto succinic acid scintigraphy were detected.
Excessive trabeculation in the bladder was observed in cystourethroscopy. In the same session, fecaloids in the distal rectum were evacuated by digital rectal manipulation. There was rapid improvement in kidney function; creatinine dropped to 0.8 mg/dL after four days. In the video-urodynamic study, it was observed that detrusor pressures increased and urethral sphincter activity increased simultaneously during the voiding phase. Dyssynergic contraction of the pelvic floor muscles by the urethral and anal sphincters was demonstrated. Doxazosin was started for DES.
The patient, who overcame her toilet phobia and recovered from constipation, is being followed up with renal replacement therapy with the diagnosis of chronic kidney disease.
Conclusions: Constipation, which is common in childhood, may be associated with serious consequences such as urinary complaints and even progression to chronic kidney disease.

EP-295 RENAL DISEASE IN BARDET BEIDEL SYNDROME IN TUNISIAN CHILDREN

Sameh Mabrouk, Hajer Mokni, Selsabil Nouir, Miniar Tfifha, Fadwa Majdoub, Houda Ajmi, Jalel Chemli, Noura Zouari, Saoussen Abroug

Pediatric Department, Sahloul Hospital, Tunisia

Introduction: The ciliopathies are a growing number of disorders caused by mutations in genes involved in the function of the primary cilium. Bardet-Biedl syndrome (BBS) belongs to this group of disorders. In this setting, kidney dysfunction is highly variable an urine concentrating fefect is the most common feature. End stage renal disease in not frequent in BBS.
the aim of our study is to determine particularities of renal disease in our patients and to describe clinical, biological and radiological findings.
Material and methods: Retrospective study conducted in pediatric department of Sahloul hospital of Sousse, Tunisia between the period of 2010 and 2021
All children with BBS, presenting kidney disease were included. Patients without kidney damage were exclude
Results: 7 patients were included in the study, mean age at diagnosis was aqual to 6.5 years and boys were most affected (sex ratio=2).
Familial history of consanguinity was founs in all patients, similar cases was noticed in one patient. Diagnosis of BBS was retained Beales criteria. Obesity, ophtalmological abnormalities and intellectual disability were found in all patients. 2 children had Hurshpungs disease and were operated at neonatal period.
At diagnosis, 6 of our patients (85%) had end dtage renal disease, all patients had cystic kidneys at ultrasoud examination. in three cases associated CAKUT was found accelerting evolution to ESRD.one patient had congenital single kidney, the second had megauretar with vesico-ureteral reflux and the third had ureteropelvic junction obstruction.
All 6 patients were treated with renal replacement therapy type hemodialysis and none recieved renal graft.
Conclusions: Kidney malformation in BBS is heterogeneous and is a cause of morbidity and mortality through the development of CKD. Renal damage in BBS is the most predicting prognosis factor with a possible evolution to ESRD, hence the importance of establishing an early diagnosis.

EP-296 CASE OF NEPHROTIC SYNDROME IN SIBLINGS

Iuliia Kyslova, Oleksandr Mazulov, Olga Yablon

National Pirogov Memorial Medical University

Introduction: Nephrotic syndrome is the most common glomerular disorder of childhood. An estimated incidence of two to seven cases per 100,000 children and a prevalence rate of 16 cases per 100,000 children. It is characterized by combination of the clinical and biochemical presentation with edema, nephrotic-range proteinuria, hypoalbuminemia and hypercholesterolemia.
Material and methods: A 2,5-year-old girl presents with complaints of puffy eyes, edema of lower extremities. On physical examination, there is no erythema or evidence of trauma, insect bite, cellulitis conjunctival injection or discharge. No family history of kidney disease. Blood pressure is 90/50 mm Hg. Urinalysis: protein 7 g/l, erythrocytes 1-2 in field of vision. Protein/creatinine ratio 250 mg/mmol. Serum albumin 22 g/l, cholesterol 9.5 mmol/l, BUN 5.8 mmol/l, creatinine 64 μmol/l. She was prescribed with prednisolone 2 mg/kg and remission obtained at 2nd weeks. Later prednisolone dose was reduced to 0,5-0,7 mg/kg but relapse occurred, and patient received corticosteroids continuously. A girl was diagnosed with steroid-sensitive nephrotic syndrome, steroid dependent course, frequent relapsing. At age of 3 she was treated with cyclophosphamide for 4 months without effect. From 4 years old she was treated with mycophenolate mofetil (MMF) for 2 years, relapse of nephrotic syndrome was less frequent and faster response to prednisolone was achieved. At age of 7 she underwent a kidney biopsy – minimal histological changes in kidney cortex. Results of immunofluorescence: IgA, IgG – negative in glomeruli, IgM – slightly positive, segmental in the glomerular capillary walls. Complement C3, albumin - negative in glomeruli. At the ages of 11 she has slowed physical development, developed with Cushings syndrome and secondary cataract. At 12 years girl received Rituximab i/v infusion two times. Complete remission was maintained for three years. At the age of 15 a new relapse, treated with MMF, after 6 months new relapse and i/v infusion of Rituximab. CD19 and CD20 levels decreased significantly after Rituximab administration. Now girl has a complete remission. The younger brother of this girl at the age of 3 was diagnosed with nephrotic syndrome. In 1

month since prednisolone administration he developed a relapse of nephrotic syndrome. Now, the boy is receiving alternate-day prednisolone.

Results: There is early onset of nephrotic syndrome, complete remission after 1-2 weeks of prednisolone at standard dose and steroid-dependent form of nephrotic syndrome in both children. Steroid-sensitive nephrotic syndrome (SSNS) is the most common glomerular disease seen in the children usually has a favorable outcome despite its relapsing course. The genetic architecture of childhood SSNS remains poorly understood due to varying clinical course. About 3% of children with SSNS may have an affected sibling or first degree relative.

Conclusions: These patients need genomic studies. It may help to accelerate the discovery of further risk factors, to broad our understanding of the pathogenesis of SSNS.

EP-297 NON-STEROIDAL ANTI-INFLAMMATORY DRUGS TREATMENT AND RISK OF CHRONIC KIDNEY DISEASE IN PATIENTS WITH BARTTER SYNDROME AND PRIMARY NEPHROGENIC DIABETES INSIPIDUS

Francesca De Zan[1], Matko Marlais[1], Klaus Arbeiter[2], Gema Ariceta[3], Francesca Becherucci[4], Martine Besouw[5], Kathrin Burgmaier[6], Denis Morin[7], Zdeněk Doležel[8], Ismail Dursun[9], Laura Espinosa[10], Ann Christin Gjerstad[11], Leire Gondra[12], Matthew Harmer[13], Martin Konrad[14], Sandrine Lemoine[15], Leire Madariaga[16], Pierluigi Marzuillo[17], Davide Meneghesso[18], Svetlana Papizh[19], Nikoleta Printza[20], Karl Schlingmann[14], Francesco Trepiccione[17], Yincent Tse[21], Alexey Tsygin[22], Anna Wasilewska[23], Marcus Weitz[24], Jakub Zieg[25], Francesco Emma[26], Detlef Bockenhauer[1], Nsaid-ckd Consortium[1]

[1]University College London Great Ormond Street Hospital For Children And Institute Of Child Health, London, Uk, *[2]Division Of Pediatric Nephrology And Gastroenterology, Medical University, Vienna, Austria.*., *[3]Pediatric Nephrology, Hospital Vall Dhebron. Barcelona. Spain*, *[4]Meyer Childrens Hospital, Firenze, Italy*, *[5]Department Of Pediatric Nephrology, University Of Groningen, University Medical Center Groningen, The Netherlands.*., *[6]Department Of Pediatrics, University Hospital Cologne And University Of Cologne, Faculty Of Medicine, Cologne, Germany*, *[7]Service De Néphrologie Pédiatrique, Centre Hospitalier Universitaire Montpellier, Centre De Référence Maladies Rénales Rares Du Sud-ouest Sorare, Université De Montpellier-filière Orkid, Montpellier, France.*., *[8]Paediatric Clinic, University Hospital Brno, Medical Faculty Of Masaryk University, Brno, Czech Republic.*, *[9]Erciyes University, Faculty Of Medicine, Department Of Pediatrics, Division Of Nephrology*, *[10]Paediatric Nephrology Departament Hospital La Paz. Madrid*, *[11]Division Of Paediatric And Adolescent Medicine, Oslo University Hospital, Oslo, Norway.*., *[12]Pediatric Nephrology Department, Cruces University Hospital, Biocruces Bizkaia Health Research Institute, Barakaldo, Spain*, *[13]Department Of Paediatric Nephrology, Southampton Childrens Hospital, University Hospital Southampton, Southampton, Uk*, *[14]Department Of General Pediatrics, University Childrens Hospital MÜnster, MÜnster, Germany.*., *[15]Hospices Civils De Lyon Nephrogones Néphrologie*, *[16]Pediatric Nephrology Department, Cruces University Hospital, Iis Biocruces-bizkaia, University Of The Basque Country*, *[17]Department Of Woman, Child And Of General And Specialized Surgery, Università Degli Studi Della Campania "luigi Vanvitelli", Napoli, Italy*, *[18]Uoc Nefrologia Pediatrica, Dialisi E Trapianto, Dipartimento Della Salute Della Donna E Del Bambino, Università Degli Studi Di Padova*, *[19]Veltischev Research And Clinical Institute For Pediatrics Of The Pirogov Russian National Research Medical University*, *[20]Pediatric Nephrology Unit, Aristotle University Of Thessaloniki, Hippokratio General Hospital Of Thessaloniki, Thessaloniki, Greece.*, *[21]Great North Childrens Hospital, Newcastle Upon Tyne, Uk*, *[22]National Medical And Research Centre For Childrens Health. Moscow*, *[23]Department Of Pediatrics And Nephrology, Medical University Of Białystok, Poland.*, *[24]University Childrens Hospital Tuebingen, Tuebingen, Germany*, *[25]Department Of Paediatrics, Charles University In Prague, And Motol University Hospital, Prague, Czech Republic.*, *[26]Division Of Nephrology, Department Of Pediatric Subspecialities, Bambino Gesù Childrens Hospital, Ircss, Rome, Italy.*

Emphasis>Emphasis Type="Bold">Introduction: Primary nephrogenic diabetes insipidus (NDI) and Bartter syndrome are rare disorders. Patients affected by these diseases are often treated with non-steroidal anti-inflammatory drugs (NSAIDs) to reduce urinary losses. Long-term use of NSAID in the general population is associated with a risk of chronic kidney disease, but it is unclear, whether this also applies to patients with NDI or Bartter syndrome. We therefore set out to investigate the association between NSAID and eGFR in these patients.

Material and methods: This is a retrospective, cross-sectional cohort study conducted through the European Rare Kidney Disease Reference Network (ERKnet), the European Society of Paediatric Nephrology (ESPN) and the European Renal Association (ERA-EDTA). Clinicians were invited by email through these organisations to submit clinical information on NSAID exposure, as well as first and last known eGFR if patients with genetically confirmed Bartter syndrome or NDI via an online portal.

Results: This is an interim report, as data collection is still ongoing. As of 10th January, data on 185 patients were submitted. The patients were 60% male and 40% female. White ethnicity was predominant equal to 76%, followed by Asian 11%, Black 4% and Other 6%. Underlying genes were (KCNJ1 15%, SLC12A1 16%, CLCNKB 35%, AVPR2 22%, AQP2 7%). 83% children received NSAID treatment, whilst 17% did not. The median age was 9.9 (IR 8-17.9).

Conclusions: The large number of patients for whom data have been provided is encouraging and suggests that we will have statistical power to detect any substantial differences in eGFR associated with NSAID use.