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ABSTRACTS

10th Asian Congress of Pediatric Nephrology 2008 (ACPN 2008) Innovation in Prevention and Therapeutic Strategies August 28–30, 2008 Centara Grand & Bangkok Convention Centre at Central World, Bangkok, Thailand

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PLENARY LECTURE

PL-1	Pediatric nephrology in Asia: Bridging the great divide
PL-2	Renal Bone Diseases: Current understanding and therapeutic strategies
PL-3	Therapeutic strategies towards renal restoration in chronic kidney disease

SYMPOSIUM

SYM-1-1 to SYM-1-2	Advances in chronic kidney diseases
SYM-2-1 to SYM-2-3	Chronic renal failure
SYM-3-1	Peritoneal dialysis
SYM-4-1 to SYM-4-2	Hemodialysis
SYM-5-1 to SYM-5-3	Urinary tract infection/Vesicoureteral reflux
SYM-6-1 to SYM-6-2	Role of biomarkers in renal failure
SYM-7-1 to SYM-7-2	Acute renal failure
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SYM-9-1 to SYM-9-3	Renal tubular disorders
SYM-10-1 to SYM-10-3	Kidney transplantation
SYM-11-1 to SYM-11-2	Systemic diseases and the kidney
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MN-1	End stage renal disease in children: dialysis and renal transplantation
MN-2	Immunology and nephrotic syndrome
MN-3	Steroid-resistant idiopathic childhood nephrosis
MN-4	Renal tubular dysfunction in beta-thalassaemia
MN-6	The clinical and genetic features of Alport syndrome in Chinese population

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FP2-1 to FP2-7	General nephrology
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FP4-1 to FP4-4	Glomerular diseases
FP5-1 to FP5-5	Genetic diseases

POSTER

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AB-022 to AB-200	Genetic/molecular/tubular diseases
AC-020 to AC-079	Hypertension
AD-018 to AD-191	UTI
AE-188	KUB anomalies
AF-013 to AF-198	Acute/chronic renal failure
AG-152	Fluid and electrolytes
AH-053 to AH-207	Dialysis/transplantations
AI-044 to AI-202	Systemic disease and the kidneys
AJ-006	Renal microvascular diseases
AK-019 to AK-176	Miscellany

PL-1: PEDIATRIC NEPHROLOGY IN ASIA: BRIDGING THE GREAT DIVIDE

Hui-Kim Yap

Department of Pediatrics, Yong Loo Lin School of Medicine, National University of Singapore and Shaw-NKF Children's Kidney Center, University Children's Medical Institute, National University Health System, Singapore.

Asia is the largest continent in the world with an estimated population approaching 4 billion, of which children under the age of 19 years constitute more than 1 billion. Unfortunately, health care delivery across the Asian continent is very heterogeneous. The more urgent issues in child health in many Asian countries encompass the control of infections, malnutrition and diarrheal diseases. However, with improvements in public health especially in urban communities, the pattern of childhood disorders has changed in many countries, resulting in a shift in the causes of mortality from infections to other chronic diseases. Therefore, although the major challenges in pediatric nephrology practice still include the management of children with acute kidney injury due to severe dehydration, sepsis and toxins, the prevention and treatment of children with chronic kidney disease is emerging as the new challenge for this millennium. The first Working Group for Pediatric Nephrology in Asia was formed in 1986, holding its first scientific meeting in Tokyo, Japan in 1988. The Asian Society of Pediatric Nephrology (AsPNA) was officially inaugurated in 1996 with a mandate to promote the development of pediatric nephrology in Asia and foster regional cooperation amongst member countries. With regional education and training as one of the main focus, quality care including both acute and chronic renal replacement therapies, as well as strategies for the prevention of chronic kidney disease, will be available more widely to the pediatric population in Asia. Data on childhood end-stage renal disease in Asia is scarce. A survey on renal replacement therapy in 12 Asian countries by Chiu MC et al in 2005 revealed that peritoneal dialysis was the major modality of dialysis in many Asian countries, with chronic ambulatory peritoneal dialysis being the main modality (64%), followed by automated peritoneal dialysis (30%), and intermittent peritoneal dialysis (6%). Although some countries have specialized hemodialysis facilites for children, however, in many Asian countries, children and adolescents are dialyzed in adult hemodialysis centers. In Asia, the majority of pediatric renal transplants are live-donor related. Patient and graft survival rates at 5 years reported in the 2005 survey were approximately 92% and 81% respectively. On the whole, renal replacement therapy is expensive in many parts of Asia, therefore financial issues limit the availability of end-stage care, especially in the developing countries. In conclusion, the challenge of this millennium for pediatric nephrology in Asia is to address this great divide in health care for children. The main focus should be on training and education, focusing on identification of children at high risk of chronic kidney disease and providing early treatment to prevent progression and the need for renal replacement therapy. At the same time, reduction of morbidity and mortality in children with acute kidney injury can be achieved using evidence-based practice which should be made easily accessible to doctors even in remote regions of the continent.

PL-2: RENAL BONE DISEASES: CURRENT UNDERSTANDING AND THERAPEUTIC STRATEGIES

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Bone histomorphometry has been has been the gold standard in the evaluation and diagnosis of renal osteodystrophy (RO). The recent new definition of RO as Chronic Kidney Disease-Mineral and Bone Disorder has once again highlighted the use of bone biopsy as a powerful and diagnostic tool to determine skeletal abnormalities in chronic kidney disease. In addition, it has been recommend the inclusion of turnover (T), mineralization (M) and volume (V) in the assessment of bone histology. Thus, we apply the TMV classification in bone biopsies (BBx) obtained after double tetracycline labeling from 140 pts. aged 14±1.2 years on CCPD for 13±3 mos. All pts. were treated with Ca-based binders and daily oral calcitriol that was held for 4 weeks prior to BBx. S-PTH (1st Nichols, 1st generation), S-Alk p'tase, Ca, and P levels were determined at BBx. Twenty five percent of the patients had both normal T and M; 36% high T and M defect; 23% high T and 16% M defect alone. Sensitivity and specifity of PTH levels <400 pg/ml and alk p'tase levels <400 IU/L gave us a sensitivity of 80 and specivity of 77% for predicting patients with both normal T and M. PTH levels ≥600 pg/ml had a sensitivity of 78% and a specifity of 73 for predicting patients with high T and M defect. Current biochemical markers seemed to unhelpful in distinguishing patients with M defect alone and a bone biopsy is required for its evaluation. FGF-23 is associated with altered skeletal mineralization in individuals with normal renal function; its potential as a biomarker for mineralization in patients with chronic kidney disease remains to be defined. We demonstrated that bone formation rates correlated with PTH ($r=0.44$, $p<0.01$), but not with FGF-23. Higher FGF-23 concentrations were associated with improved osteoid thickness and osteoid maturation time. We conclude that FGF-23 may be a new biomarker for the non-invasive assessment of skeletal mineralization in patients with secondary hyperparathyroidism.

Calcium-containing phosphate binding agents and active vitamin D sterols have been widely recommended for the control of hyperphosphatemia and the prevention of secondary hyperparathyroidism. The administration of such compounds, however, contributes to the development of frequent episodes of hypercalcemia, hyperphosphatemia, adynamic osteodystrophy and vascular calcifications. On the other hand, untreated secondary hyperparathyroidism causes significant morbidity, including severe bone deformities, fractures and growth retardation in children. The availability of effective calcium-free-metal free phosphate binding agents, such as sevelamer combined with new active vitamin D sterols, such as doxercalciferol has widened the margin of safety for the treatment of secondary hyperparathyroidism. Although these agents can effectively control serum phosphorus and lower PTH levels, little is known about their effects on the skeletal lesions of secondary hyperparathyroidism. We have therefore prospectively compare the effects of calcium carbonate and sevelamer on the control of the biochemical and skeletal indices of secondary hyperparathyroidism during therapy with either calcitriol or doxercalciferol in pediatric patients treated with peritoneal dialysis. The data demonstrated that sevelamer allows for the use of higher doses of active vitamin D sterols without inducing changes in serum calcium levels and with adequate control of the skeletal lesions of

secondary hyperparathyroidism, but the mineralization defect persists. Serum PTH levels between 400-600 pg/ml are associated with indices of bone formation within the normal range in the vast majority of patients after treatment with intermittent doses of active vitamin D sterols.

PL-3: THERAPEUTIC STRATEGY TOWARD RENAL RESTORATION IN CHRONIC KIDNEY DISEASE

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Chronic kidney disease (CKD) is a new growing public health threat. Failure to prevent end-stage renal disease (ESRD) associated with a progressive increment in ESRD patients are relevant to 2 crucial issues namely (1) the insensitiveness of present diagnostic marker such as serum creatinine determination, or microalbuminuria which detects late CKD (stages 3-5; equivalent to CCr < 60 ml/min/1.73m2), but is unable to screen early CKD (stages 1,2; equivalent to CCr > 60-119 ml/min/1.73m2). (2) Present therapeutic target aiming at suppression of proteinuria or/and controlling blood pressure although is beneficial, but not perfect, since it does not correct the mechanism of renal disease progression. In order to establish an effective preventive strategy aiming to minimize ESRD, the above crucial and unanswered problems have been integrally addressed as follows. (1) With respect to the diagnostic marker, **fractional excretion of magnesium is a highly sensitive marker for screening early CKD stages 1 and 2**. FE Mg assists to initiate early therapeutic and preventive strategy under favourable environment towards renal regeneration. (2) With respect to the properly therapeutic target, we have studied **the role of renal microvascular disease inducing the development of tubulointerstitial fibrosis**. Accumulating evidence demonstrates oxidative stress, immunocirculating disturbance which are circulating toxins inducing glomerular endothelial cell injury and dysfunction. Glomerular endothelial dysfunction induces hemodynamic maladjustment at the efferent arteriole, a progressive reduction in peritubular capillary flow, a chronic ischemic injury to the tubulointerstitium, and eventually tublointerstitial fibrosis. Therefore, **correction of hemodynamic maladjustment** with multidrug vasodilators (ACEI, AII receptor blocker, calcium channel blocker, antiplatelet) would be the appropriate therapeutic target.

The present failure in treating late stages of CKD (3-5 patients) is likely to be explained by the altered vascular homeostasis observed in this group of CKD patients. We observed deficiencies in the mechanism of vascular repair namely deficiencies in VEGF, flt-1 (VEGF R1) angiopoietin-1, nitric oxide, and endothelial progenitor cell, which are essential factors for vascular angiogenesis; whereas factors opposing normal vascular repair such as VEGF R2 (KDR), angiopoietin-2 are elevated. Taken together, such alterations encompass in a default angiogenesis and renal microvascular rarefaction. In contrast, a normal or slightly impaired vascular homeostasis is encountered in early stage of CKD associated with favourable environment for renal regeneration. Thus, correction of hemodynamic maladjustment with multidrug vasodilators in the early stage of CKD can enhance peritubular capillary flow, improve renal perfusion to the tubulointerstitium, and effectively prevent the development of tubulointerstitial fibrosis. This coincides with the improved glomerular filtration rate and hence, prevents the ESRD.

In conclusion, an innovative strategy to effectively prevent ESRD can be implemented by (1) screening early stage of CKD with a novel diagnostic marker FE Mg (2) appropriately correcting the hemodynamic maladjustment with multidrug vasodilators to improve renal perfusion at the early stage of CKD.

SYM-1-1: RENAL MICROVASCULAR ABNORMALITIES IN CHRONIC KIDNEY DISEASE

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It is well known that the maintenance of renal vasculature is crucial for preservation of renal function. Maintenance of glomerular capillary would help maintain glomerular filtration rate (GFR), whereas maintaining peritubular capillaries in the interstitium would be essential for providing oxygen and nutrition to the tubules and interstitial cells. Afferent and efferent arterioles also play a key role in determination of renal hemodynamics, activation of renin-angiotensin system and intra-glomerular pressure. Recent studies demonstrate a substantial repair capacity of renal microvasculature in various spectrum of renal diseases, however this recovery process of the kidney is compromised in chronic kidney disease (CKD). Although an early proliferative response of the glomerular and peritubular capillary endothelium was noted with a loss of functioning nephron mass, the proliferation was not sustained with a progressive loss of the endothelium due to unchecked apoptosis over time. The loss of the glomerular endothelium predisposes to activation of platelets and the coagulation system that favors capillary collapse and the development of glomerulosclerosis. The degree of glomerular and peritubular capillary loss in CKD correlates with the severity of glomerulosclerosis and interstitial fibrosis. These changes in renal microvasculature were preceded by an altered expression of vascular survival factor and/or anti-angiogenic factors, such as vascular endothelial growth factor (VEGF) and thrombosponsin-1. Interestingly, macrophage-dervied cytokine and vasoactive mediators II was known to play a role in regulation of the expression of angiogenesis-modulating factor in the kidney.

There have been several lines of evidence suggesting that stimulation of endothelial proliferation might be beneficial in kidney disease. Glomerular endothelial cell proliferation plays a key role in the repair of capillaries and microaneurysms in the Thy-1 nephritis model whereas blockade of the capillary repair resulted in an aggravation of renal damage. In addition, VEGF supplement enhanced capillary repair and improved renal function in animal model of CKD. A key role for VEGF in glomerular capillary formation has also been suggested from studies of glomerular development in neonatal rats and mice. However, although angiogenesis-modulating therapy seems to be an attractive therapeutic modality of CKD, not all studies would support the view that increased angiogenesis prevents renal disease progression. For example, in diabetic nephropathy, the most prevalent cause of CKD in these days, an implication of modulating renal microvascular angiogenesis as therapeutic tool seems to depend on the stage of diabetic nephropathy and associated complication in individual patients. Further studies on the role of the microvasculature in progressive renal disease are necessary, and the insights may lead to new therapeutic approaches for the treatment of CKD, which shows the most remarkable worldwide epidemic nowadays.

SYM-1-2: ROLE OF HEMATOPOIETIC STEM CELLS IN RENAL REPAIR

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Acute kidney injury (AKI) carries high morbidity and mortality rates. The only treatment option that is currently available for AKI consists of supportive measures while patients are waiting for renal function to recover. Unfortunately, renal recovery is often incomplete. The long-term consequences from AKI include hypertension, chronic renal insufficiency and end-stage renal disease. Stem cells may offer new and more effective treatment. We and others have shown that transplanted hematopoietic stem cells can be incorporated into the regenerating tubules and express epithelial markers. Fusion between bone marrow cells and renal epithelial cells is not the primary mechanism of cell conversion. However, in our hands, the rate of incorporation of the transplanted cells into renal tubules is low. No improvement in renal function is observed when freshly isolated bone marrow cells are injected intravenously into mice with renal ischemia-reperfusion injury. To enhance the therapeutic potential of the bone marrow stem cells, we induce hematopoietic stem and progenitor cells to differentiate into renal cells prior to transplantation. Hematopoietic-to-renal conversion is observed after the sequential induction in vitro. Moreover, transplantation of the induced cells accelerates kidney recovery from ischemic injury. These results indicate that hematopoietic cells can be reprogrammed to convert into functional renal cells. Bone marrow cells can be a potential source for development of a stem cell-based therapy for kidney disease.

SYM-2 -1: PROGRESSION OF THE CHRONIC KIDNEY DISEASE IN CHILDREN

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Introduction: It has been appreciated that once glomerular filtration rate (GFR) has decreased to below a point for a certain period, chronic kidney disease (CKD) tends to progress relentlessly to end-stage. However, the rate of progression varies in patients, and investigations have elucidated various factors associated with the progression. Accordingly, it is presumed that manipulation of the modifiable factors may retard the progression of CKD. In addition, attenuation of renal fibrosis by a phosphodiesterase inhibitor, pentoxifylline (PTX) has been confirmed in animal studies.

Methods: Clinical and laboratory parameters associated with the progression were analyzed retrospectively in children with CKD. GFR was estimated by Schwartz formula and the rate of progression was measured by GFR decline per year. Effect of PTX (700 mg/m2/day p.o.) in combination with ACEI or ARB on the progression of pediatric CKD was also evaluated.

Results: Compared to the stationary group, progressive group of CKD had older age, higher initial GFR, more proteinuria, lower hematocrit, lower serum calcium, higher serum phosphorus, and lower total CO2 levels. Decrement of GFR during PTX treatment in combination with ACEI/ARB was less than that on ACEI/ARB only.

Conclusion: Correction of anemia, reduction of proteinuria, adjustment of calcium, phosphorus and total CO2 levels are associated with more stationary renal function. More attention is necessary at earlier stage and in older children to obviate the progression of childhood CKD. Multimodal approach is expected to hinder the progression more effectively.

SYM-2-3: NUTRITIONAL MANAGEMENT IN CHRONIC RENAL FAILURE

Alan R Watson

University of Nottingham, UK

The paediatric imperative is to promote growth and development. Nutrition is important for that objective, particularly during the infantile phase. Recent research has moved away from considering adequate intake resulting in protein calorie malnutrition to use OF the term "cachexia". There may be a loss of lean body mass combined with normal, or even, increased fat mass, high resting energy expenditure and inadequate response to nutrient supplementation. Research has also highlighted that nutritional factors may be implicated in progression of cardiovascular disease which is known to be accelerated in CKD. Hence, the need to consider the nutritional status of the child with regular assessment and support as CKD advances.

The nutritional management requires expert dietetic support and a multidisciplinary team approach. Basic anthropometric measurements should be taken at every clinic visit, regular clinic assessment combined with dietary assessment via diaries or dietary recall in clinic. The nutritional prescription should be based upon growth, biochemical and haematological data as well as consideration of relevant medications such as phosphate binders, iron and micronutrient supplements and salt balance. Each child has individualised nutritional requirements which should be given by the oral route whenever possible. Inadequate intakes to maintain adequate growth velocity require the early introduction of enteral feeding via the gastrostomy or nasogastric route. Intensive nutritional support requires dietetic and medical time and needs to take into account the psychosocial needs of the family. This may all help to promote normal growth without the need for the routine use of recombinant human growth hormone.

SYM-3-1: THE IMPACT OF INTERNATIONAL COLLABORATIONS ON THE PROVISION OF PERITONEAL DIALYSIS

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The International Pediatric Peritonitis Registry (IPPR) and the International Pediatric Peritoneal Dialysis Network (IPPN) are collaborative efforts that were established to collect data related to the provision of peritoneal dialysis (PD) to children and to use that data to improve patient care on a global basis. The IPPR created an online database to record information on the etiology and treatment of peritonitis in 47 centers from

14 countries. Global variation was noted in the 501 peritonitis cases identified in terms of causative organisms, frequency of staphylococcal methicillin resistance, incidence of culture-negative peritonitis and in the antibiotic susceptibility patterns. Gram-negative peritonitis was associated with the poorest outcome and was most often seen in patients < 5 years old and in those conducting frequent exit-site care with mupirocin. In-vitro antibiotic susceptibility data revealed only 69% of gram positive organisms and 80% of gram negative organisms to be susceptible to a first generation cephalosporin and an aminoglycoside, respectively, prompting consideration for alternative recommendations regarding empiric antibiotic therapy. In 89% of cases, there was full functional recovery with less frequent complete resolution occurring among the 52 patients who experienced a relapsing infection. The IPPN has expanded the goals of the registry to monitor overall PD performance and complications and to assess the association between PD practice and outcome. To date, nearly 600 patients have been enrolled in the Network from 76 centers in 27 countries. Preliminary data has revealed global differences in the distribution of PD modality selection, the frequency of growth retardation and growth hormone usage and in the management of secondary hyperparathyroidism. Current analyses are being conducted to assess the correlation between PET transport capacity, dialysis prescription and cardiovascular disease, in addition to the reasons for hospitalization. Continued data collection and analysis should help define global standards for pediatric PD care.

SYM-4-1: OPTIMIZING THE HEMODIALYSIS PRESCRIPTION

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Introduction

The goal of hemodialysis is to optimize patient well-being by appropriate solute and fluid removal. The prescription for each session should, a)minimize dialysis related symptoms, b) maximize fluid and solute removal, and c) reduce inderdialytic morbidity.

Methods

Review of the literature and clinical experience describing measures of dialysis adequacy,and options available to minimize symptoms related to solute and fluid removal.

Results

The average weekly plasma urea, the % urea reduction, the clearance (Kt/V) of urea, the plasma albumin,have each been suggested as markers of dialysis adequacy, but none should be used in isolation. Nutritional status,as measured by normalized protein catabolic rate (nPCR) as well as clinical anthropometry and growth must also be considered. Similarly, hypertension and abnormal cardiac function, anemia, calcium/phosphate and PTH disturbances must be minimized. Achievement of these goals, within the limitations of thrice weekly HD, may be improved by use of sodium ramping, ultrafiltration profiles, and non-invasive blood volume monitoring.

Conclusion

This review will consider the value and limitations of sodium ramping, ultrafiltration profiles, and blood volume monitoring for prescription of pediatric hemodialysis. Also, simple estimates of nPCR and Kt/Vurea, which have been validated in children will be included. Finally, the limitations of thrice weekly HD will be considered.

SYM-4-2: DIALYSIS AND CARDIOVASCULAR DISEASE (CVD)

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Treatment with dialysis and transplantation is now highly successful, but despite this the incidence of death from CVD has not improved, and many young patients will die from cardiovascular events; the risk of death for a such a child is 30 times that of the normal population, and the chances of death from CVD for a young adult on dialysis is equivalent to that of an 85 year old. Lifespan is reduced by 40-60 years in children on dialysis and by 20 to 30 years if transplanted, with about 50% of deaths due to CVD.

There are many factors contributing to this high incidence of CVD. One of the most important is fluid overload and LVH. Another cause, about which there is currently much interest, is vascular calcification. Abnormal bone mineral metabolism (CKD-metabolic bone disorder [MBD]) is an important cause of vascular damage and calcification, and there is now plenty of evidence to link this to mortality. We have undertaken studies of vascular structure (carotid intima-media thickness [IMT]), function (pulse wave velocity) and coronary calcification in children on dialysis and correlated the results with plasma phosphate, PTH, vitamin D and calcification inhibitors. We have found significantly worse results for these vascular parameters in children with PTH levels > 2 × the upper limit of normal and when levels of 1,25(OH)2D are either above or below the normal range.

To address this further, we obtained arteries that would normally be discarded from children undergoing insertion of peritoneal dialysis catheters or transplantation. We quantified the calcium (Ca) load in the arteries and correlated it with clinical, biochemical and vascular measures. We found that Ca accumulation begins pre-dialysis, but it is the induction of VSMC apoptosis in dialysis that is the key event by disabling VSMC defence mechanisms thus leading to overt calcification, eventually with clinically detectable vascular damage. Therefore, the identification of factors that lead to VSMC death in dialysis will be of prime importance in preventing vascular calcification.

A new area of research is that of myocardial dysfunction in association with intradialytic hypotension. Areas of myocardium develop regional motion wall defects, called myocardial stunning which, although initially temporary, may lead on to permanent myocardial damage.

Unravelling the processes of vascular injury in children with CKD will enable new treatments and even preventative therapies that will help reduce the burden of CVD when these children reach adulthood

SYM-5-1: VESICOURETERAL REFLUX: GENETIC CONSIDRERATION

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Renal parenchymal damage after urinary tract infection (UTI) and reflux nephropathy may lead to chronic renal failure in children and young adults. Vesicoureteral reflux has known to be related with genetic background. Several genetic mediators including renin angiotensin system (RAS), VEGF and TGF-β1 have been implicated in initiating and regulating parenchymal damage leading to renal scarring. This study was designed to investigate whether the genetic polymorphisms of RAS, VEGF and TGF-β1, were associated with the susceptibility to UTI, vesicoureteral reflux (VUR), and subsequent renal scarring.

RAS

There was no difference in the distribution of the ACE II, ID and DD genotype and the AT1 A1166C transition between the patients with primary VUR and the controls. However, the incidence of AT2 A-1332G transition was significantly lower in primary VUR patients ($p=0.047$). Furthermore, in the case of combination of ACE and AT2 gene, a significantly lower incidence of primary VUR was seen with II genotype of ACE and A-1332G transition in the AT2 receptor gene($p=0.003$). Concerning the risk factors of primary VUR, the grade of reflux was significantly higher in AT2 A-1332G transition group compare to the group without transition ($p=0.044$, mean grade of reflux 3.54 ± 1.17 vs 2.83 ± 1.43). Also, ACE ID/DD genotypes with AT2 A-1332G transition group had higher grade of reflux than ACE II genotype without AT2 A-1332G transition ($p=0.025$, mean grade of reflux 3.48 ± 1.21 vs 2.47 ± 1.36). Other risk factors did not show much difference in the distribution of ACE, AT1 and AT2 genotypes. These findings indicate that low rate of incidence but high grade of primary VUR is seen in AT2 A-1332G transition group, at least in the Korean population.

VEGF and TGF-β1

In both UTI and VUR groups, there was an increase in frequency of the VEGF -460 CC (control, 4.3; UTI, 15.9; VUR, 17.8%; $P<0.05$), TGF-β1 -509 CC (control, 8.7; UTI, 34.6; VUR, 35.1%; $P<0.001$), and TGF-β1 -800 GG genotypes (control, 19.1; UTI, 40.5; VUR, 40.4%; $P<0.05$). An increase in the TGF-β1 +869 CC (scar-positive, 35.4; scar-negative, 10.3%; $P<0.05$) and a decrease in the +869 TC genotype (scar-positive, 29.2; scar-negative, 55.2%; $P<0.05$) were observed in the scar-positive subjects. There were no differences in +405 VEGF genotype frequencies. The VEGF T-460C and the TGF-β1 C-509T, G-800A, and T869C polymorphisms could be genetic markers of the process of UTI and VUR.

SYM-5-2: THE ROLE OF ANTIBIOTIC PROPHYLAXIS IN UTI

YN Lim

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Urinary tract infection (UTI) is still one of the most common bacterial infections in children. Prior to the advent of antibiotics, about 20% of newborns and infants died from acute UTI. Fortunately, mortality today is very low. However the acute and long term morbidity of UTI particularly in this group of children is still considerable.

The standard of care for children especially very young children diagnosed to have UTI was to perform imaging procedures mainly ultrasound, micturiting cystourethrogram (MCUG) and DMSA scans to look for urinary tract abnormalities and renal scarring. The most common abnormality found was vesicoureteric reflux (VUR). VUR was said to predispose to recurrent UTI especially recurrent acute pyelonephritis which may lead on to progressive renal scarring. Progressive renal scarring may result in hypertension or kidney failure or both. This formed the basis of the therapeutic modalities to avoid further renal parenchymal damage, namely antibiotic prophylaxis to maintain urine sterility or surgery to eliminate the reflux.

However evidence thus far has not clearly shown the benefits of either antibiotic prophylaxis or surgery in the prevention of further renal damage. Newer randomized studies albeit not placebo controlled have thrown doubt as to the effectiveness of antibiotic prophylaxis in the prevention of symptomatic UTI and renal scarring.

Hence, treating every UTI promptly and adequately may be the way forward. We however, eagerly await the results of the RIVUR study (Randomized Intervention for Children with Vesicoureteral Reflux) - a randomized, double blind placebo control study on whether prophylactic antibiotics prevents renal scarring and UTI in children with VUR.

SYM-5-3: NON RETRACTILE PHYSIOLOGIC PHIMOSIS AND INFANTILE URINARY TRACT INFECTION

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Urinary tract infection (UTI) is a common ascending infection of fecal uropathogens. First UTI commonly develops in infancy and frequently recurs. Thus, infants with first UTI should be investigated for the host risk factors and managed properly to prevent recurrent UTI and subsequent renal scarring. A series of host risk factors for childhood UTI are listed up in Nelson Textbook of Pediatrics. Female gender is on top of them, followed by uncircumcised males.

Female preponderance of childhood UTI has been a well-known phenomenon but infantile UTI has developed predominantly in the male gender. It was noted in mid 1980s that a majority of infantile UTI developed in male infants who were not circumcised. In several large cohort studies, the incidences of UTI in male infants without neonatal circumcision were 10-20 times more prevalent than in male infants with neonatal circumcision. This high incidence of UTI in uncircumcised male infants is attributed to male preponderance and high male to female ratio of infantile UTI. Despite the definite protective effect of neonatal circumcision against UTI in early infancy of male infants, AAP (1999) still adopted a neutral or anti-circumcision stance. Although others were concerned about the AAP policy (2003), a recent systemic review (2005) also did not recommend routine neonatal circumcision.

A majority of uncircumcised neonates have nonretractile prepuces (physiologic or congenital phimosis), which makes penile hygiene difficult. Preputial sac of a nonretractile prepuce becomes a reservoir of uropathogens which can be a source of infantile UTI, until a nonretractile prepuce became spontaneously retractile.

In prepubertal children with phimosis who were referred for circumcision, topical application of moderate and highly potent steroids for 4-8 weeks has been proved as successful (65-95%) in resolving phimosis. Even in infants, who were diagnosed with first UTI and found to have nonretractile physiologic phimosis, hydrocortisone cream of lowest potency and physiotherapy (gentle retraction) were highly successful (96.1%). In infants whose prepuces became retractile after topical hydrocortisone, recurrent UTI developed less

frequently for the following 1 year than in infants with persistently nonretractile prepuces (7.1% vs 29.6%). A majority of male children, who were referred for aseptic pyuria and asymptomatic bacteriuria, were found to have nonretractile prepuces. Successful topical application of hydrocortisone with physiotherpy eliminated aseptic pyuria and asymptomatic bacteriuria.

Topical application of hydrocortisone with physiotherapy is a simple, economical, safe and effective therapy for a nonretractile physiologic phimosis and it should be considered as an ideal alternative to the much debated neonatal circumcision.

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SYM-6-1: Biomarkers for the Early Diagnosis in Acute Renal Failure

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Introduction: Acute kidney injury (AKI) represents a common and devastating problem in clinical medicine. The lack of early biomarkers for AKI has lead to a delay in initiating potentially effective therapies. The objective of this presentation is to review the status of novel urinary biomarkers for AKI that have progressed to the clinical phase of the biomarker discovery process.

Methods: Literature review (PubMed, MedLine) from 2000 to the present.

Results: The most promising AKI biomarkers include neutrophil gelatinase-associated lipocalin (NGAL), interleukin-18 (IL-18), and kidney injury molecule-1 (KIM-1).

Conclusions: It is likely that the AKI biomarkers will be useful for timing the initial insult and assessing the duration and severity of AKI. Based on the differential expression of the biomarkers, it is also likely that the AKI panels will help distinguish between the various types and etiologies of AKI, and predict clinical outcomes. Studies to validate the sensitivity and specificity of these biomarkers in clinical samples from large cohorts and from multiple clinical situations are currently in progress, facilitated by the development of commercial tools for the reproducible measurement of these biomarkers across different laboratories. The availability of such personalized and predictive information could revolutionize renal and critical care medicine in the not-too-distant future.

SYM-6-2: BIOMARKERS ON MECHANISM OF KIDNEY DISEASE PROGRESSION

Frederick J Kaskel

Definitions of a Biomarker:

1) a characteristic that is objectively measured and evaluated as an indicator of normal biologic processes, pathogenic processes, or pharmacologic responses to a therapeutic intervention.

2) Clinical end point: a direct measure of how a patient feels, functions or survives.

3) Intermediate end point: a biomarker which is intermediate in the causal pathway between an intervention and a clinical endpoint.

4) Surrogate: a laboratory measurement or physical sign that is used in therapeutic trials as a substitute for a clinically meaningful endpoint that is expected to predict the effect of the therapy.

CKD is characterized by glomerulo- and vascular sclerosis and tubulointerstitial fibrosis regardless of the etiology. This finding suggests that after acute kidney injury, adapative changes in nephrons initiate a chronic maladaptive process cumulating in scarring. Factors identified in mediating these changes are: hemodynamic, the renin-angiotensin-aldosterone system, cytokines, chemokines, growth factors, podocyte loss, dyslipidemia, specific mechanisms of tubulointerstitial fibrosis, low birth weight and low nephron number, genetic predisposition to CKD, and, proteinuria.

It is well established that proteinuria has been implicated in causing CKD progression via mesangial toxicity and tubular injury with interstitial fibrosis. It is part of the natural history of CKD and serves as a biomarker of kidney damage, a clue to the diagnosis of CKD, a risk factor for progression (causal in animal models), a modifier for efficacy of ACE inhibitor therapy in non-diabetic kidney disease, a hypothesized marker of vascular permeability (generalized endothelial dysfunction), a risk factor for cardiovascular disease (CVD) at lower levels than defined as CKD, and a hypothesized surrogate outcome for kidney disease progression and CVD reduction.

In addition to proteinuria, the cytokine, TGFß was found in urine to differentiate between histologically proven minimal change and focal segmental glomerulosclerosis pediatric patients (11.5 vs 148 pg/mg Cr, respectively, Woroniecki, RP. et al. Am J Nephrol 2008;28(1):83-90). Furthermore, application of urinary proteomics combined with bioinformatics, predicted the response to treatment in these patients with steroid sensitive or resistant nephrotic syndrome (Woroniecki RP, et al. Am J Nephrol 2006;26(3):258-67). Thus, the need for identification and measurement of potential biomarkers in prospective longitudinal cohort studies such as the Chronic Kidney Disease in Children will lead to better patient-specific targeted therapies to slow or halt the progression.

SYM-7-1: SPECTRUM OF ACUTE KIDNEY INJURY

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Acute renal failure (ARF) is a common condition, often under recognized with severe consequences. The condition has diverse etiologies, with features ranging from mild elevation in serum creatinine to anuric renal failure. In most instances, the decline in function is secondary to tubular injury that leads to functional or structural changes in the kidney. In the absence of a standard definition & recognizing that ARF includes a spectrum of conditions, the term acute kidney injury (AKI) has been proposed to reflect the entire range of the syndrome. The term includes functional or structural abnormalities or markers of kidney damage, present <3 months. AKI is associated with increased mortality that is worsened when dialysis is needed. An unrecognized effect of AKI is the later development and progression of chronic kidney disease.

AKI is increasingly prevalent in both developed and developing countries, and is associated with considerable morbidity and mortality. There is a lack of data regarding the true incidence of AKI, its epidemiology and outcome in children. In developed countries, AKI occurs chiefly in ICUs and is associated with multiorgan failure, sepsis and high mortality. Important causes of AKI in developing countries include severe sepsis, dehydration due to gastroenteritis, infections (severe malaria, leptospirosis, hemolytic uremic syndrome) and postinfectious glomerulonephritis. Many causes of AKI in developing countries can be prevented by low-cost interventions at individual, community and regional levels.

Therapy of AKI is supportive with attention to fluid and electrolyte status, ensuring adequate nutrition and treatment of complications. There is limited role for treatment with low-dose dopamine or frusemide. Patients with AKI should be followed up to examine for evidence of chronic kidney disease (renal dysfunction, proteinuria or hypertension).

SYM-7-2: EMERGING PROTEOMIC TECHNOLOGIES IN ACUTE RENAL FAILURE

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Introduction: Acute kidney injury (AKI), previously referred to as acute renal failure (ARF), represents an important problem in clinical medicine. Despite significant improvements in therapeutics, the mortality and morbidity associated with AKI remain high. The reasons for this include (a) an incomplete understanding of the underlying pathophysiologic mechanisms, and (b) the lack of early markers for AKI, and hence an unacceptable delay in initiating therapy. Fortunately, the application of innovative technologies such as proteomics to human and animal models of AKI has uncovered several novel proteins that are emerging as biomarkers and novel therapeutic targets.

Methods: Literature review (PubMed, MedLine) from 2000 to the present.

Results: Recent advances in proteomics have resulted in the identification of biomarkers in the plasma (NGAL) and urine (NGAL, KIM-1, IL-18) for the investigation of AKI, as well as identification of proteins that inform the pathophysiology of AKI and offer novel therapeutic targets.

Conclusions: Proteomic studies will likely yield additional sensitive and specific biomarkers for the investigation of AKI resulting from diverse etiologies. Such tools will be indispensable for the early diagnosis and initiation of timely therapeutic measures.

SYM-8-1: TITLE OF STUDY: THE FOCAL SEGMENTAL GLOMERULOSCLEROSIS IN CHILDREN AND YOUNG ADULTS CLINICAL TRIAL (FSGS-CT)

FUNDING AGENCY: NIH/NIDDK

FSGS Steering Committee: Richard Fine, Aaron Friedman, Jennifer Gassman, Tom Greene, Debra Gipson, Ron Hogg, Frederick Kaskel, Marva Moxey-Mims (NIH/NIDDK), Melina Radeva, Howard Trachtman, Sandra Watkins, Norm Siegel** (deceased)*

Goals and objectives: Primary FSGS is a leading cause of end stage renal disease (ESRD) in both children and adults, with loss of kidney function in 50% of patients over 10-years. The FSGS -CT was organized and implemented by the NIH in response to this poor prognosis, and the need to rigorously test treatment modalities and to delineate the cellular, molecular and genetic basis of this complex disorder. In evaluating the therapeutic interventions for the FSGS-CT, it was noted that no evidence based medicine has designated a specific therapeutic intervention for steroid-resistant FSGS that significantly reduces proteinuria or preserves renal function in a substantially large proportion of patients. However, the following 4 factors were taken into consideration in the ultimate design of this clinical trial:

- an established role for CSA in the treatment of FSGS
- the potential, but unproven benefit, of intermittent high-dose corticosteroid therapy in combination with another immunosuppressive agent
- the efficacy of either therapeutic intervention to induce sustained reduction in proteinuria after withdrawal of a therapeutic agent
- the side effects and consequences of any long term therapeutic intervention, if withdrawal of medication is unsuccessful

The FSGS-CT is a Phase III randomized trial of patients greater than 2 years and < 40 years of age being conducted at over 100 participating sites in North America which are divided among 3 core networks with principal investigators, study coordinators, and a central data-coordinating center (http://fsgstrial.org). Eligibility criteria

include: 1) an estimated GFR > 40 ml/min/1.73m2, 2) a first morning urine protein/creatinine ratio (Up/c) > 1.0, 3) biopsy-confirmed primary FSGS, 4) corticosteroid resistance as defined as failure to achieve sustained Up/c <1.0 based on a course of steroid treatment for 4 wks and/or a minimum cumulative dose of 56 mg/kg or 1,680 mg of prednisone or its equivalent. Major exclusion criteria include: 1) prior therapy with cyclosporine (CsA), tacrolimus, mycophenolate mofetil (MMF) or rapamycin, 2) BMI> 97th %tile for age or >40 kg/m2, 3) uncontrolled hypertension. Target sample size is 207 randomized patients to achieve 80% power to detect17.9% absolute difference in remission rate.

Primary outcome

- Attainment of partial or complete remission

Main secondary outcome

- Relapse after withdrawal of immunosuppressive agents

Secondary outcomes

- Treatment failures
- Change in eGFR
- Side effects
- Declining quality of life
- Receipt of biological materials

Eligible patients are randomly assigned to one of two active treatment arms: CsA OR pulse dexamethasone and MMF. The target period for each therapeutic intervention is 12 months. Both study groups are also treated with either Lisinopril or Losartan for 18 months and low dose alternate day steroids for 6 months. The outcome measures have been defined to reflect clinical practice and to allow patients to be withdrawn if no therapeutic effect is evident within six months of randomization. The primary outcome is based on achievement of remission of proteinuria: complete remission (CR) Up/c < 0.2; partial remission (PR) Up/c < 50% of baseline value and < 1.0; or no remission (NR). The pattern of response to both therapeutic interventions will be assessed at 6, 12 and 18 months. Patients who do not achieve a CR or PR after 6 months of therapy in either therapeutic arm are defined as treatment failures for the primary outcome, and exit the study. For patients who achieve a CR or PR at six months, the primary outcome will be assessed again at 12 months. If patients have not maintained at least a PR at 12 months, they are considered a therapeutic failure and exit the study. The main secondary outcome is the persistence of remission following withdrawal of the CsA or MMF/dexamethasone after 12 months on therapy. This secondary outcome will be assessed after 6 months off therapy (i.e., 18 months after enrollment) only for patients who have achieved a CR or PR after 12 months on therapy.

Randomization began in November 2004 and ends on May 31st, 2008. 187 patients were enrolled and 136 randomized with 50 excluded for biopsy readings inconsistent with primary FSGS, Up/c <1.0, and eGFR <40 mL/min/1.73m2. Entry characteristics of the enrolled patients are: 57 < 24 years of age; 38% African Americans, 57% white, 5% other; 54% male, 46% female; eGFR 135 mL/min/1.73 m2 and Up/c >6.44. To date, 17 ancillary studies have been approved of which 11 received NIH support.

The FSGS-CT is the largest controlled trial of FSGS in North America and will establish a standard of therapy for corticosteroid-resistant primary FSGS. Additional benefits of the trial are the establishment of an infrastructure for the study of FSGS, the creation of a national repository of biospecimens for investigations on the pathogenesis of FSGS and the role of histological subclassifications of FSGS in the response to therapies, and the evaluation of the efficacy of withdrawing immunosuppressive drugs while maintaining ACE inhibitors/ARBs. A logical extension of the trial is the development of a Pediatric Nephrology Clinical Trials Group to facilitate new investigations, innovations and faculty development in translational research.

SYM-8-2: PROTECTIVE ROLE OF IMMUNOSUPPRESANTS ON DEFECTIVE NEPHRIN BIOGENESIS

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Introduction: Nephrotic syndrome (NS) is a common kidney disorder with multiple etiologies. The mechanisms by which therapeutic agents induce remission in susceptible individuals with NS are not well understood and remain one of the most important questions in nephrology today. We previously demonstrated that glucocorticoid receptor and glucocorticoid inactivating enzyme, 11㪈hydroxysteroid dehydrogenase type 2, are expressed in the glomerular podocyte (Kidney Int 1999; J Clin Endocrinol Metab 2002). Recent compelling evidence indicates that therapeutic agents may exert the antiproteinuric effect by restoring nephrin biogenesis because the loss of nephrin expression and altered cellular localization of nephrin in the podocyte foot processes results in proteinuria.

Method: To gain a direct crosstalk between therapeutic agents action and altered nephrin biogenesis, we focused on the role of the intracellular energy homeostasis in maintaining nephrin trafficking machinery. To inhibit nephrin biogenesis, we established a model of the endoplasmic reticulum stress (ER) induced by glucose starvation in nephrin expressing cell line.

Result and Discussion: Glucose starvation induced hypoglycosylated nephrin which retained in the ER, not be transported to the plasma membrane. Interestingly glucocorticoid partially rescued plasma membrane localization of nephrin through recovery of ER chaperone (calreticulin) function via increase of ATP by upregulation of the mitochondrial genes (Kidney Int 2006). On the other hand, mizoribine, which was discovered in Japan and frequently used for steroid dependent NS, restored the intracellular energy balance by salvaging the ATP levels and completely rescued expression of the mature, fully-glycosylated, plasma membrane nephrin by a mechanism dependent on the inhibition of the Inosine 5'-monophosphate dehydrogenase activity (J Am Soc Nephrol 2007). We also determined that the energy imbalance alters not only nephrin biogenesis but also other N-glycoprotein 㪈1-integrin biogenesis. These data suggest that immunosuppressants may directly act on the diseased podocyte through interacting with energy system, results in the rescue of pathological proteinuria of NS.

SYM-8-3: CYCLOSPORINE TREATMENT FOR FREQUENT-RELAPSING NEPHROTIC SYNDROME

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Introduction: Children with frequent-relapsing nephrotic syndrome (FRNS) experience the serious side effects that result from continuous corticosteroid therapy. In Japan, most of the pediatric nephrologists treat children with FRNS with cyclophosphamide, mizoribine or cyclosporine (CyA). And more than half of children with FRNS are treated with CyA. CyA is a relatively new agent that is useful in the management of FRNS. The beneficial effects of CyA, however, often are accompanied by side effects. Of greatest concern is chronic CyA nephropathy, characterized by arteriolar lesions and tubulointerstitial (TI) lesions.

Methods: We examined clinical course of chronic CyA nephropathy and risk factors for the development of CyA-induced TI lesions. We conducted a prospective, open-label multi-center randomized controlled trial with trough level monitoring of CyA (Sandimmune) to develop a safe and effective CyA treatment for FRNS. Patients were randomly divided into two groups with both initially receiving CyA for 6 months to maintain a whole-blood trough level between 80 and 100 ng/ml. Over the next 18 months, the dose was adjusted to maintain a slightly lower (60-80 ng/ml) trough level in Group A, while Group B received a fixed dose of 2.5 mg/kg/day. The primary end point was the rate of sustained remission with analysis based on the intention-to-treat principle.

Results: The arteriolar lesions were improved by discontinuation of CyA, but the TI lesions did not regress with drug discontinuation(J Am Soc Nephrol 11:2265-2271, 2000, Pediatr Nephrol 16:723-727, 2001). Thus, prevention of the development of CyA-induced TI lesions is the most important issue in CyA treatment for children with FRNS. Duration of CyA treatment (>24 months) and duration of heavy proteinuria (>30 days) were independent significant risk factors for the development of CyA-induced TI lesions (Kidney Int 61:1801-1805, 2002). After 2 years, the rate of sustained remission was significantly higher in Group A as compared with Group B. Mild arteriolar hyalinosis of the kidney was more frequently seen in Group A than in Group B, but no patient was diagnosed with CyA-induced TI lesions.

Conclusions: CyA for 2 years in a dosage that maintains the trough level between 80 and 100 ng/ml for the first 6 months and 60 and 80 ng/ml for the next 18 months is an effective and relatively safe treatment for children with FRNS (Kidney Int 73: 1167-1173, 2008). I also introduce an on-going prospective multi-center randomized controlled trial with C2 monitoring of CyA (Neoral) (JSKDC 03).

SYM-9-1: RENAL TUBULAR ACIDOSIS.

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Background: The inability of lowering urinary pH to less than 5.5 and raising net acid excretion to normal concentrations in response to metabolic acidosis are cardinal features of type 1 renal tubular acidosis (RTA). The bicarbonate wasting from the proximal tubules characterize type 2 RTA. Aldosterone deficiency or resistance gives rise to type 4 RTA.

Clinical features: The case vignettes of 4 children with RTA illustrate the diverse clinical presentations, the consequences of missed diagnosis, non-compliance and treatment. Guidelines for diagnostic workup of RTA types 1, 2 and 4 follow.

Growth Failure: The key experiments on the mechanisms of growth failure in metabolic acidosis are reviewed: impaired growth hormone pulsatile secretions, altered IGF-1 expressions, reduced nutritional intakes.

Genetics and Molecular Genetics:

Major studies on clinical associations and genetic loci are presented.

Recommendations: This review ends with the author's recommendations on therapeutic strategy in overcoming non-compliance.

SYM-9-2: DENT DISEASE

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Dent disease is an X-linked renal tubular disorder characterized by low molecular weight (LMW) proteinuria, hypercalciuria, nephrocalcinosis and progressive renal failure. Patients usually go into end-stage renal failure when they are 49 years old in average. Most of the patients are detected as proteinuria by a school urinalysis screening in Japan. About 60% of the Japanese patients with Dent disease are due to the mutations in chloride channel 5 (ClC-5) gene (CLCN5). CLCN5 encodes a 746 amino acid ClC-5 that has about 12 transmembrane domains. Mutational analysis of CLCN5 revealed mutational "hot spots" at codons 279, 648, and 704 (nonsense mutations) and at codon 506 (missense mutation) in British and Japanese patients. Three missense mutations (S270R, L278F and R280P) on the putative loop between domains 5 and 6 of ClC-5 protein suggest that this putative loop may have a regulatory role in ClC-5 function. ClC-5 protein is expressed in the proximal tubule, thick ascending limb of Henle, and the intercalated cells of the collecting duct. ClC-5 protein is localized to the subapical endosomes with H + -ATPase in the proximal tubule. Acidic endosomal pH formed by ClC-5 and H+-ATPase is necessary for normal endosomal function, including endocytosis, trafficking, and recycling to the surface. Two knocked-out mouse models revealed that megalin's expression was reduced. Moreover, histological analysis of the kidney specimen from the patient revealed reduced expression of megalin on the luminal membrane of the proximal tubules. Urine concentration of megalin is decreased in the patients with Dent disease. These observations suggest defective binding of protein with megalin in the luminal membrane of the proximal tubule and resulted proteinuria. Approximately 10% of patients with Dent disease have OCRL1 (causative gene for Lowe syndrome) mutations. OCRL1 encodes a protein that regulates the function of endosome. Mutations in OCRL1 can occur with isolated renal phenotype of Dent disease in patients lacking cataracts, renal tubular acidosis, and neurological abnormalities that are characteristic of Lowe syndrome. However, the precise pathogenesis is still unknown. This also suggests genetic heterogeneity in the pathogenesis of Dent disease. Unlike the patients with typical Lowe syndrome, none of patients have metabolic acidosis. These observations and findings suggest that OCRL1 mutations can cause the isolated renal phenotype of Dent disease and affected individuals lack the cataracts, typical facial features, renal tubular

acidosis, and neurologic abnormalities that are characteristic to Lowe syndrome. It is difficult to explain that OCRL1 mutations can cause the isolated renal phenotype of Dent disease. However, it is possible that another phosphatase, INPP5B, which shares amino acid homology with OCRL1, can compensate phosphatase activity in patients with Dent disease due to OCRL1 mutations.

There are no specific interventions at present that will change the natural course of renal manifestations and progressive renal failure in patients with Dent disease. Hypercalciuria is dysfunction corrected by thiazide diuretics therapy in doses similar to effective doses for idiopathic hypercalciuria, presumably by stimulating the reabsorption of calcium in the distal convoluted tubule, where ClC-5 channel is not expressed. However, this is not a long-term study which provides the evidence that it is effective to prevent or delay the progression of end stage renal failure. In animal experiment using clcn5 (mouse chloride channel 5 gene) knock-out mice, high citrate diets can delay the progression of nephrocalcinosis and end stage renal failure. Treatment with an angiotensin-converting enzyme inhibitor or angiotensin receptor blocker may delay progression of end stage renal failure.

SYM-9-3: PHYSIOLOGICAL, PHARMACOLOGICAL AND PATHOPHYSIOLOGICAL SIGNIFICANCE OF OAT FAMILY IN THE KIDNEY

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The kidney plays a pivotal role in elimination of the toxic compounds, e.g. endogenous metabolites, drugs, environmental compounds, and their metabolites from the body. Elimination of these toxic compounds via kidney is performed not only by glomerular filtration but also tubular secretion. Over the last decade, tremendous knowledge on the role of organic anion transporters in tubular secretion has been clarified. The organic anion transporters include several families; namely, OAT family, OATP family and MRP family. Among these, the members of OAT family play the central in the kidney.

In the kidney, OAT1 and OAT3 are expressed in the basolateral membrane of the proximal tubular cells, and take up a variety of drugs and their metabolites from the blood. In the brain, oat3 is expressed in the blood-brain barrier and blood-spinal fluid barrier, and play a role as a "functional BBB or BBBCF". These OAT members transport ㉾-lactam antibiotics, diuretics, NSAIDs, ACEIs, ARBs, methotrexate and so on. OAT members also transport endogenous compounds, such as cyclic AMP and GMP, prostaglandins and dicarboxylates.

OAT family includes two members, each of which transports important endogenous compound, e.g., urate and carnitine. hURAT1 (human uric acid transporter 1) was identified as the fifth member of OAT family and was revealed to transport urate. hURAT is located in brush border membrane of proximal tubular cells and take up urate from glomerular filtrate. Mutations in hURAT1 gene cause "hereditary renal hypouricemia". OCTN2 (CT1) is a carnitine transporter expressed ubiquitously in various tissues. Carnitine is an essential compound in the ㉾-oxydation of fatty acids, and genetic defects in OCTN2 gene causes a "systemic carnitine deficiency".

In my talk, I would like emphasize significance of OAT family in its physiological, pharmacological and pathophysiological aspects.

SYM-10-1: THE NAPRTCS TRANSPLANRT REGISTRY: 20-YEAR ANNIVERSARY

Bradley A Warady

Children's Mercy Hospital, USA

The registry of the North American Pediatric Renal Trials and Collaborative Studies (formerly the North American Pediatric Renal Transplant Cooperative Study) was initiated in 1987 to capture information about current transplant practice and trends in immunosuppressive therapy with a goal of improving the care of the pediatric renal allograft recipient in North America. Over the past two decades, data has been collected from 10,762 transplants (9,854 index) in recipients <21 years and has revealed substantial changes in patient management and outcome with time. Whereas the percentage of young (<6 years) recipients has remained constant at 20%, the percentage of young (<10 years) deceased donors has decreased from 35% in 1987 to only 7% in 2007. The height SDS at the time of transplant has increased from -2.43 to -1.37 and the mean number of hospitalization days during the first transplant month has decreased by 50%. Substantial changes in induction and maintenance immunosuppressive therapy have been associated with a decreased probability of a first rejection by 12 months for living-donor (54.1% to 13.7%) and deceased-donor (69.3% to 17.9%) recipients. In addition, 1 year graft survival rates have improved from 89.4% and 75.2% for living-donor and deceased-donor recipients, respectively in 1987-1990, to current rates of 95.7% and 95.0%. This improvement has, however, been complicated by a malignancy rate that has increased from 1.05 in 1987-1990 to 2.45 more recently. Nevertheless, 5 year patient survival has significantly improved for all deceased-donor recipients and the 3 year survival of infants receiving living-donor grafts has improved from 89.8% to 95.5%. Ongoing data collection in the transplant registry and the continued development of prospective clinical trials based on the analysis of registry data will likely result in continued improvement in the outcome of pediatric transplant recipients.

SYM-10-2: KIDNEY TRANSPLANTATION: JAPANESE EXPERIENCE

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We have two registries regarding renal transplantations in children: Japanese National Pediatric ESRD Registry and Japanese Kidney Transplantation Registry. I would like to introduce the recent data from these registries in this paper.

In Japan, 50 to 60 new patients younger than 15 years old became ESRD every year. In 402 children who became ESRD from 1998 to 2004, 83% were introduced to PD, 9% were introduced to HD and 9% received pre-emptive transplantation. The frequency of pre-emptive transplantation was a small number(s), but it has recently increased up to 22% in 2004. Sixty five percent of all ESRD received transplantation within 5 years after becoming ESRD.

A total of 2,031 children younger than 20 years old received transplantations from 1964 to 2004 in Japan. Of all, only 9.4% received cadaveric transplantation. 1,124 patients were younger than 15 years old and 132 patients were younger than 5 years old. After 2000, the number of the patients who received transplantation increased from 60/year up to about 90/year. We investigated patients and graft survivals in 1,751 children whose outcomes can be traced. We divided the patients into 3 groups: 440 children in 1964–1985, 646 patients in 1986-1995, 413 patients in 1996-2001. Patients survival rate in 5 years improved from 82% to 96% and 98%, and graft survivals improved from 63% to 78% and 90% in each 3 groups, respectively. Ten years graft survivals changed from 48% to 66% after 1985. Five years graft survivals in the group of 1996-2001 were 93% in 1-5 years old, 91% in 6-12 years old and 89% in 13-19 years old. These results showed the excellent data.

In addition to the data from the registries, I showed the data in our hospital (TMCH) as to graft survival in patients with ABO incompatibility and FSGS. Their survivals were not different from those of other transplant children in TMCH.

SYM-10-3: LONG-TERM COMPLICATIONS AND OUTCOME OF TRANSPLANTATION IN CHILDHOOD

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Studies of outcome post renal transplant over 20 years are now beginning to emerge. Such information is of considerable importance in order to counsel patients and families.

Mortality

The overall mortality rate remains high, with a relative risk of death post transplant that is 12.7 times higher than that of the age–related normal population, with little sign of improvement since the 1980s. Young age at transplant is an important factor that has been demonstrated to affect patient survival, although there is evidence that its effect has declined in recent years. All reports show a small but consistent benefit of living donation (LD).

The major causes of death post transplant are cardiovascular disease (CVD), infection and malignancy, variously reported as 30–36% for CVD, 24–56% for infection and 11–20% for malignancy. Two other important factors that contribute to death are non-concordance with medications or treatment withdrawal and obesity. All studies show a survival advantage for patients who are transplanted in comparison to dialysis. It might be expected, therefore, that patient survival after pre- emptive transplant would be superior, and studies have shown this to be the case.

Transplant survival

Transplant survival has shown a steady improvement over the years. It used to be that young recipient and donor age adversely affected outcome, but in the last decade, this no longer seems to be so. Adolescents, however, have the worst transplant survival of all ages, mainly due to non-concordance. LD and preemptive transplantation and changes in immunosuppression to more powerful agents also benefit outcome.

Other causes of transplant loss

The incidence of diabetes has been increasing post transplant, and is associated with increased graft loss. Diseases that recur post transplant include FSGS, MPGN and HUS. Oxalate will continue to be deposited in the transplanted kidney if liver transplant is not undertaken in patients with hyperoxaluria. Nephrotic syndrome can recur in patients with congenital nephrotic syndrome, and anti-GBM nephritis in patients with Alport's syndrome, both due to the development of antibody to the 'missing' protein. Hypertension is very common and is a significant and independent predictor of poor long-term transplant function,

Psychosocial outcome

Final height is improving, but so is obesity. Despite a high retransplantation rate and the presence of significant morbidity, renal transplantation in children can lead to attainment of a productive and satisfying life, with a high degree of rehabilitation in adulthood.

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SYM-11-1: SYSTEMIC LUPUS ERYTHEMATOSUS IN THAI CHILDREN

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In Thailand SLE is not an uncommon disease. The ratio of pediatric SLE to pediatric out- patients in tertiary care hospitals is 1: 700. The Thai Pediatric Nephrology Association has conducted a multicenter study in 13 tertiary care hospitals all over the country. The objective of the study is to determine the clinical outcomes of Thai children with SLE and risk factors for chronic renal insufficiency.The data of 500 SLE children, age at diagnosis less than 15 years, newly diagnosed during 1 January 2002- 31 December 2006 were reviewed. Severity and damage of the disease were assessed using ECLAM index and SLICC/ACR damage index respectively. The ratio of female: male is 5.5: 1. Kidney involvement is found in 82% of the patients. Kidney biopsy is done in 351 patients. The most common pathology found is diffused proliferative lupus nephritis. Prednisolone plus pulse intravenous cyclophosphamide is the standard treatment for these patients. There has been increasing use of mycophenolate mofetil in recent years. Majority of patients achieve favorable outcome. The mean ECLAM index at initial is 6 + 2.7 and decreases significantly after treatment. Chronic renal insufficiency is found in 11 patients while 7 of these patients require renal replacement therapy. Risk factor for developing chronic renal failure is initial GFR < 60 ml/min/1.73 m2. Concernig complication and damage, the frequently found organ damages are eyes (22.5%), kidney (10.5%), neurologic (8.5%) and musculoskeletal (16%) systems respectively. In this study, 14 patients died and the most common cause of death is infection. Quality of life which is one of the important outcome measures is being studied to determine the quality of life of Thai children with SLE.

SYM-11-2: IS IGA NEPHROPATHY INCREASED?

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Introduction: Primary IgA nephropathy is the most common primary renal disease in the world wide. Since it was reported in 1968 by J Berger and N Hinglais, the disease of IgA nephropathy is emphasized more and more in the aspect of incidence, clinical characters, prognosis and treatment. To understand the incidence as well as the clinical features in IgA nephropathy a retrospective study on IgA nephropathy was finished and reported by the Chinese Society of Pediatric Nephrology in 2007, the relevant data was compared with that reported previously by other groups.

Method: Totally 1349 cases of IgA nephropathy from 33 hospitals of different cities from January 1995 to December 2004 were collected and analyzed retrospectively. More data on childhood IgA nephropathy from published reports in Chinese along with data from other countries were compared.

Result: IgA nephropathy in children under 14 years accounted for 1.37% of the hospitalized cases with urological-kidney diseases, and 11.18% of cases with renal biopsies. The cases of IgA nephropathy increased from 327 (0.83%) to 1022 (1.73%) in the periods of 1995 to 1999 and 2000 to 2004, respectively ($P<0.01$). Whereas, the previous survey conducted by the Chinese Society of Pediatric Nephrology demonstrated that IgA nephropathy accounted for 7.3% of cases with renal biopsies, and ranked forth in the renal diseases in children.

Conclusion: The proportion of IgA nephropathy in renal dieases or in cases with renal biopsies seemed increased in recent years in China, which might attribute to the improvement of knowledge on renal diseases including IgA nephropathy, the increased case numbers performed renal biopsy and more available medical resources to take care patients. Nevertheless the real incidence of IgA nephropathy still need to carry out epidemiology investigation.

SYM-12-1: OBESITY AND RELATED RENAL DISEASE

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The global epidemic of obesity is largely responsible for the high prevalence of metabolic syndrome, which is characterized by central obesity, hypertension, dyslipidemia, insulin resistance, and elevated proinflammatory biomarkers. Metabolic syndrome adversely affects kidney structure and function. Increased body mass index has been shown to be associated with increased risk for developing end-stage renal disease. The mechanism by which obesity leads to renal disease is not fully understood at the present time. This presentation will summarize some human and animal studies conducted in the United States and Asian countries on the subject of obesity and related kidney disease. Obese patients have glomerular hyperfiltration and are at higher risk for developing glomerulopathy and focal segmental glomerulosclerosis. Investigations on adipokines, which include leptin, resistin and adiponectin, demonstrate an important role of adipokines in the pathogenesis of obesity-related renal disease. In children with chronic renal disease the level of a cardiovascular protective factor adiponectin is relatively low in overweight patients as compared to lean patients. An inverse correlation between albuminuria and plasma levels of adiponectin has also been shown in non-diabetic adults with obesity. Transgenic mice lacking adiponectin (Ad-/- mice) exhibit an increased urinary albumin excretion and effacement of podocyte foot processes. Treatment of adiponectin to the Ad-/- mice reduces albuminuria and restores podocyte morphology. The effect of adiponectin is largely due to the activation of 5'-AMP activated protein kinase (AMPK). Since albuminuria is a strong predictive factor for renal disease progression, adiponectin and its signal transduction pathway may represent a therapeutic target for potential treatment of obesity-related kidney disease.

SYM-12-2: RENAL DIABETIC EMBRYOPATHY AND EGR-ALPHA GENE IN ANIMAL MODEL AND HUMAN SONOGRAPHIC FINDINGS

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Introduction: The uncontrolled hyperglycemia of maternal diabetes has been found to negatively affect pregnancy, including renal agesis, small kidney, hydronephrosis and fused kidneys. Furthermore, the number of nephrons was significantly reduced in the pups of diabetic female rats. We hypothesized that children of gestational hyperglycemic mothers develop defects of renal development similar to those observed in our animal model.

Methods: We observed the defects of renal development of diabetic pregnancy from late phase of embryogenesis to postnatal period in animal model (C57BL/6J mice). We also used 2- (2-D) and 3- (3-D) dimensional power Doppler renal ultrasound (US) to measure and compare the kidney/liver echogenic ratio in 76 children born to gestational hyperglycemic mothers versus 240 age-matched healthy children. The age of 20 babies was < 6 months throughout their follow-up, while 56 children were followed between 6 months and 4 years of age. Whole kidney volume, mean grayness (MG) and vascularization (VI), flow (FI) and vascularization-flow (VFI) indices were calculated.

Result: In our preliminary studies, defects and abnormalities of mice fetal kidney development under the environment of maternal hyperglycemia induced by streptozotocin injection were established. Histologically, large glomerular size, reudced nephron number, and ureteric tubular detachment were observed. Cell apoptosis, but not cell proliferation, was significant increased in fetal kidney of diabetic group. In the molecular level, we noticed that glial cell line-derived neurotrophic factor (GDNF) and early growth response alpha (EGFα) expressions are down-regulated at peri-tubular region of developing fetal kidney of diabetic group. In > 6-month-old children from hyperglycemic mothers during pregnancy, the 2-D US echogenic kidney/liver ratio was higher than in age-matched controls ($P<0.01$). Furthermore, higher incidence of non-obstructive hydronephrosis was detected in 20 children (26.3%) born to gestational hyperglycemic mothers then no- hyperglycemic mothers (1%). 3-D US demonstrated lower mean renal volume, VI, FI and VFI in children of hyperglyce-

mic mother then in children of normoglycemia mothers beyond six months of age.

Conclusion: GDNF-dependent activation of GFRα-1 and Ret in fetal renal tissue is mediated by a complex intracellular signal network. The kidney/liver echogenic ratio on 2-D US and indices of renal volume and vascularization on 3-D power Doppler scan may enable clinicians to establish an early baseline and follow the evolution of these renal characteristics in high-risk children.

MN-1: END STAGE RENAL DISEASE IN CHILDREN: DIALYSIS AND RENAL TRANSPLANTATION

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Dialysis initiation

Children who reach chronic kidney disease (CKD) stage 4 (GFR < 30 ml/min/1.73 m2) should receive education about kidney failure and options for its treatment, including peritoneal dialysis (PD), hemodialysis (HD) and renal transplantation1. A systemic plan of monitoring basing upon the combination of clinical, biochemical, and psychosocial assessment prior to dialysis initiation is required. Dialysis initiation should be considered when GFR is 9 to 14 ml/min/1.73 m2 (CKD stage 5) or at greater GFR levels in the presence of malnutrition, fluid overload, hypertension, hyperkalaemia, growth failure or neurological consequences of uremia1.

Choice of dialysis modalities

While renal transplantation remains the optimal therapy for children with end-stage renal disease (ESRD), the majority of children require maintenance dialysis because of lack of a suitable donor. The choice of dialysis modality has to take into account of patient size with difficulties of vascular access in small children, patient/family choice, medical co-morbidities, and family support 1,.2. PD is usually the favored modality of pediatric renal replacement therapy (RRT) especially for young children. While PD continues to be the most prevalent modality selection for all children in some countries, there has been increase in HD utilization in the last decade in the United States. The utilization of PD in pediatric patients is approximately 50% worldwide 3. In the United States, PD is the most frequent dialysis modality (60% of dialysis children) according to the NAPRTCS Registry, whereas HD is more common according to the USRDS data which reflects that there is a preference for HD for adolescents patients that are cared in adult dialysis unit (4). Contraindications to PD include omphalocele/gastrochisis, bladder extrophy, diaphragmatic hernia and lack of an appropriate care giver. Contraindications to HD include very small infants, lack of vascular access, contraindications to anticoagulation and cardiovascular instability 5.

Peritoneal dialysis

Automated PD (APD) is the most frequently used and advocated PD modality in children as it gives greater freedom to the children and caregivers during the day for school and social activities5. However, continuous ambulatory PD (CAPD) is commonly used in countries that lack finances and technical support4.

There is recent advancement in automatic cyclers with computer chips that provide recording of treatment provision and remote access that resulted in improved patient adherence 6. The peritoneal dialysis adequacy recommendation has recently been changed due to re-analysis of the CANUSA data which showed residual renal function is the most important predictor for patient mortality and in the ADEMEX trail that increasing peritoneal small solute clearances did not lead to improved PD patient survival 6. The recent recommended minimal "delivered" dose of total (peritoneal and kidney) small-solute clearance should be a Kt/V urea of at least 1.8/wk 1.

Many children have long-term dependence on a functioning peritoneal membrane due to long waiting time for renal transplant in some countries. The use of biocompatible new PD solutions such as Physioneal (lacatate/bicarbonate mixed buffer pH 7-7.4), Extraneal (7.5% icodextrin), Nutrineal (1.1% amino-acid) not only preserve the peritoneal membrane but also increases the possibility of tailoring the APD prescription to the clinical, metabolic and nutritional needs of children and adolescents 7

Hemodialysis

During the past two decades, children have benefited from major improvements in both technology and clinical management of HD. Newer machines provide more precise control of ultrafiltration by volumetric assessment and continuous blood volume monitoring. More biocompatible synthetic membranes and specific small size material dialyzers and tubing make it possible to put infants on HD. However, the provision of adequate vascular access remains the greatest obstacle to successful HD, especially in infants 8. HD usually requires 4-h thrice- weekly in-hospital treatment sessions. Home nocturnal HD in adult studies reveals improved biochemical control and reported health-related quality of life. Frequent home HD can be provided by installing a water treatment system in the patient's home, or with machines that use sterile dialysate in bags (NxStage systems) 6,.9. Preliminary experience with frequent HD show that it leads to improved growth, decreased fluid and dietary restriction, excellent metabolic and blood pressure control, and the lack of need for phosphate binders. 10

Renal transplantation

Renal transplantation is the goal for children with ESRD. However, the chronic immunosuppression exposes children to multiple complications and drugs side effects. With the newer generation of powerful induction and maintenance immunosuppressants, immunosuppression minimization is a realistic objective for the pediatric patients which reduces morbidity from the toxicities associated with immunosuppressive drugs. Steroid minimization and avoidance have shown early promise. Elimination of Calcineurin-Inhibitors (CNI) carries the potential to minimize the chronic graft 'non-immune' injury which is one of the main causes of chronic allograft nephropathy (CAN), one of the leading cause of transplant kidney loss 11.,12. Randomized, prospective studies of steroid and CNI minimization and /or avoidance are needed to clearly confirm their short and long term safety and efficacy.

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MN-2: IMMUNOGLOBULIN AND NEPHROTIC SYNDROME

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Nephrotic syndrome (NS) is a common kidney problem among hospitalized pediatric patients. Basic pathology is related to leaking of proteins through glomerulus. Abnormality of immunoglubulins occurs because of gloumerular leakage, decrease synthesis and abnormal metabolism. Immunoglobulin is lost in urine along with albumin leading to hypogammaglobulinaemia. Deposition of IgM in the kidney is associated with longtime prognosis. Nephrotic syndromes (NS) with atopic child are associated with raised level of immunoglobulin E. Deceased level with reduced functional capacity of immunoglobulin is responsible for increase prone to infection in already immunocompromised patients with nephrotic syndrome. Low IgA level in patient with NS is associated with respiratory tract infection and diarrhea, and are frequently relapser. Studies showed that significantly reduced level of IgG is found in steroid resistant nephrotic syndrome in compare to steroid sensitive nephrotic syndrome. Persistent low or low normal serum IgG level during period of remission is predictor of frequent relapse in children with idiopathic nephrotic syndrome. So, immunuglobulins play an important role in the pathogenesis, in the management plan, and as a predictor of the outcome of nephrotic syndrome.

MN-3: STEROID-RESISTANT IDIOPATHIC CHILDHOOD NEPHROSIS.

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Introduction: Steroid-resistant nephrotic syndrome (SRNS) with focal and segmental glomerulosclerosis (FSGS) is a heterogeneous disorder and the most severe and frequent type of all glomerulopathies in children leading to end stage renal failure. The podocyte is at the centre of development and progress of FSGS since this unique cell type plays a major role in the integrity of glomerular structure and permeability.

Results: The rate of complete remission of SRNS after induction therapy using different immunosuppressive agents is reported to range between 30-84%, depending on the treatment schedule and on the underlying defects of FSGS. Children with genetic types of FSGS barely respond to immunosuppressive therapies and overtreatment prior to transplantation should be avoided. The response of children with an idiopathic type of FSGS to immunosuppressants is superior to those with genetic FSGS. However, many children with idiopathic FSGS do not enter complete remission if they are undertreated, e.g. by short term immunosuppressive monotherapies. If immunosuppressive treatment fails, these patients will have to undergo renal transplantation. However, as the unknown pathogenetic mechanisms may persist, more than one third of these patients with idiopathic FSGS develop a rapid recurrence of SRNS responding poorly to further long term therapeutic attempts.

Conclusion: The aim of this review is to show that, by contrast with previously published data, new treatment options taking into account recently identified genetic etiologies of SRNS, and in idiopathic forms superior results of an intensified treatment mode, currently lead to a different, more effective approach of childhood SRNS with lesions of FSGS.

MN-4: RENAL TUBULAR DYSFUNCTION IN BETA-THALASSAEMIA

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Advances in the treatment of Beta-Thalassaemia including iron-chelating agents and Hematopoietic stem cell transplantation (HSCT) make long term outcome of these patients much more important. Despite being one of the most common hereditary hemoglobinopathies in many parts of the world, data on long term renal involvement in Beta-Thalassaemia were scant.

Glomerular filtration is usually normal but recent studies showed that proximal tubular dysfunction is common in patients with Beta-Thalassaemia. This was demonstrated by uricosuria, aminoaciduria, low urine osmolality, increased urine levels of low molecular weight protein, N-Acetyl-Beta-D-glucosaminidase (NAG) and Beta2-microglobulin. Urine NAG, a sensitive marker for proximal tubular damage, was found to be correlated with proteinuria, patient's age, serum ferritin, duration of deferoxamine, and duration of receiving blood transfusion. The causes of tubular abnormalities are most likely

chronic anemia and oxidation reaction from iron deposition as increased level of urine malondialdehyde, an indicator of lipid peroxidation, was also reported.

Although rarely resulting in clinical disorders that warrant treatment, prolonged repeated tubular damage may cause tubulointerstitial fibrosis and chronic kidney disease. These patients may also be more at risk of acute renal failure from hypovolemia and nephrotoxic substances. Acute and chronic kidney diseases were reported in children and adult receiving HSCT but our long term study in a small group of Beta-Thalassaemia patients post HSCT showed low incidence of chronic kidney disease and a better renal tubular parameters than children of the same age and disease severity but without HSCT.

MN-6: THE CLINICAL AND GENETIC FEATURES OF ALPORT SYNDROME IN CHINESE POPULATION

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Introduction: Alport syndrome is a hereditary glomerulonephritis caused by COL4A3, COL4A4 and COL4A5 gene mutations. It is manifested by hematuria, sensorineural hearing loss, ocular lesions and progressive renal failure. In order to understand the disease via clinical aspect and genetic aspect, a retrospective study was carried out based on data from Chinese Alport syndrome patients.

Method: Clinical data including manifestations, renal pathology, type IV collagen alpha chains staining, cochlear and ocular abnormalities were collected and analyzed. PCR amplification of mRNA or genomic DNA of ⍺(IV) chains was applied to detect mutations. COL4A gene mutations were confirmed by DNA sequencing. Data or parameters obtained from this study were compared to others published previously by other investigators based on Caucasian population.

Result: There were 146 un-related index cases of Alport syndrome enrolled in this study, which included 104 males and 42 females. Among them, 134 patients were diagnosed with X-linked Alport syndrome and 12 patients (8.2%) with autosomal recessive Alport syndrome. No autosomal dominant hereditary mode of Alport syndrome was identified. Almost half patients (58%) presented with hematuria as the onset symptom, the rest part of patients (42%) presented with hematuria and proteinuria at the onset of the disease. Around 20% of patients manifested with nephrotic proteinuria. Non-glomerular hematuria was reported in about 20% of Alport patients. It seemed that 17% of patients was de novel mutation of COL4A5. There were 48% of Alport patients with hearing loss and 57% of patients with ocular lesions. One confirmed X-linked Alport syndrome patient was also diagnosed with IgA nephropathy. Mutation analysis revealed that, in this study, 51% of X-linked Alport syndrome patients with point-mutation and, only 4% of patients with large deletion in COL4A5 gene.

Conclusion: The clinical features revealed in this study in Chinese patients were similar to the previous reports, in addition to the prompt that higher portion of patients with proteinuria at the disease onset and small portion of patients with non-glomerular hematuria. While large deletion of COL4A5 gene was less than the other reports.

FP1-1: ANTICOAGULATION MANAGEMENT IN PEDIATRIC RENAL TRANSPLANT RECIPIENTS USING UNIVERSAL THROMBOPHILIA SCREENING AND RISK STRATIFICATION APPROACH

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Introduction: Vascular thrombosis accounts for 10.5% of graft failures in pediatric kidney transplant recipients (NAPRTCS 2006 annual report). This study was done to identify pediatric renal transplant patients with high risk for allograft thrombosis and develop a risk stratification protocol for post-operative anticoagulation.

Method: We screened every patient at the pretransplant evaluation for thrombophilic risk factors including Antithrombin III, Protein C or Protein S deficiency, Activated Protein C resistance, Factor V Leiden, MTHFR or Prothrombin gene mutation, Hyperhomocysteinemia, High Factor VIII, high Lipoprotein (a), Dysfibrinogenemia, and Antiphospholipid Antibodies. The patients were assigned a risk category (Level I-IV) based on the presence of thrombophilic risk factors. Their post-operative anticoagulation plan was pre-determined based on their risk level [table1]. Heparin drip was initiated a few hours after the transplant and titrated to achieve a 1.5 fold aPTT level. Anticoagulation was changed to low molecular weight heparin (LMWH) on day 2-4, when the risk of major bleed was over, and serum creatinine had normalized.

Result: Ten patients (aged 2-17 years) have been screened and 8/10 were found to be at high risk for thrombosis (3 were in Level II or 5 were in level III). The thrombophilic factors (number of patients) identified were prothrombin gene mutation (2), MTHFR (8), Lupus anticoagulant (2) and low protein S (2). Six patients have undergone renal transplant. All were successfully treated with LMWH post-operatively. No patient has developed allograft thrombosis. 1/5 patients developed a peri-renal hematoma requiring evacuation 7 days after the surgery, but completed prophylactic LMWH therapy.

Conclusion: Thrombophilia is markedly common in pediatric kidney transplant recipients. Universal screening can stratify risk and allow an effective intervention to reduce that risk. Our Institution has developed a unique risk stratification protocol to identify patients at high risk for allograft thrombosis and prevent this complication by post-operative anticoagulation. However; larger scales, prospective randomized control trial using this protocol are needed.

FP1-2: CURRENT STATUS OF CHILDREN ON PERITONEAL DIALYSIS IN KOREA

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Introduction: Peritoneal dialysis (PD) is the major form of dialysis in use for infants and children with chronic renal failure (CRF). We evaluated the current status of children on PD in Korea.

Method: In May 2008, questionnaires were sent to the pediatric nephrologists via e-mail. The questionnaires were about epidemiology, PD modes and adequacy, growth, blood pressure, laboratory findings and medications. Four centers replied and those data were reviewed.

Result: A total of 103 patients were included in this study. Male to female ratio was 1.6:1. Mean age was 11.5±4.9 years (0~19 years). Primary renal diseases diagnosed were as follows: primary glomerular disease (34%), chronic pyelonephritis-reflux nephropathy (14.6%), systemic disease (9.7%), renal dysplasia-hypoplasia (8.7%), heredofamilial disease (6.8%), vascular disease (3.9%), drug-induced nephropathy (1.0%), and unknown (12.6%). PD modalities were as follows: CAPD (42.7%), CCPD (27.2%), NIPD (11.7%), and Hybrid (18.5%). Weekly Kt/V was 2.1±2.0 (0.3~4.1). Results of peritoneal equilibrium test (PET) were as follows: low 36.8%, low average 31.6%, high average 19.7%, and high 11.8%. Z-score for weight was -1.00±1.20 (-4.54~2.50). Z-score for height was -1.55±1.65 (-9.42~1.87). Growth hormone was administered in 24.3% of patients. Systolic blood pressure (BP) for their ages and heights was as follows: < 50 percentile 27.2%, 50~90 percentile 33.0%, 90~99 percentile 10.7%, and > 99 percentile 13.6%. Anti-hypertensive drugs were administered in 62.1% of patients. Laboratory findings were as follows: hemoglobin 10.5±1.4 g/dL, calcium 9.7±0.7 mg/dL, phosphorus 5.4±1.4 mg/dL, parathyroid hormone 324.2±342.8 pg/ml.

Conclusion: Primary glomerular disease was the most common cause of CRF. CAPD was the most prevalent PD modality. Dialysis doses were variable. Low and low average peritoneal transport type were common. Growth disturbance were noted in many patients. Some patients had hypertension even with anti-hypertensive drugs. Hemoglobin level was slightly low. Calcium-phosphorus levels were maintained adequately, but many patients had secondary hyperparathyroidism.

FP1-3: POOR OUTCOMES OF TENCKHOFF CATHETERS WITH PERITONITIS IN CHILDREN ON CHRONIC PERITONEAL DIAYSIS

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Introduction: Peritonitis remains one of the major complications of peritoneal dialysis (PD) despite advances in Tenckhoff (TK) catheter design and dialysis techniques. This study examined factors associated with peritonitis and TK outcomes in children with end-stage renal disease (ESRD) over the past two decades.

Method: All TK catheter insertions performed in 78 paediatric patients with ESRD (median age at diagnosis at end-stage renal failure of 12.2 years, range 0.07-20 years) between December 1985 to May 2007 were reviewed retrospectively. Peritonitis rate was calculated as number of episodes/patient-year. Kaplan-Meier survival analysis was done using the log-rank test. Poor outcome was defined as catheter failure and removal. Factors associated with peritonitis studied include age at TK insertion, coiled vs straight catheters and exit site infection. Multivariate logistic regression analysis was used to determine correlations.

Result: A total of 124 TK insertions were performed in 291 patient-years. The incidence of peritonitis decreased from 1.2 episodes/patient-year in 1997 to 0.29 episodes/patient-year in 2006. Staphylococcus aureus was the most common isolated organism (19.2%) followed by coagulase-negative Staphylococci (9.6%), Enterobacter cloacae (8.4%), Pseudomonas aeruginosa (7.2%), and Candida albicans (6.0%). Logistic regression analysis showed that use of straight-catheters was significantly correlated with peritonitis (p= 0.022 OR=2.37 95%CI 1.13, 4.95). Median catheter survival was 50.2 months (range 0.3-120.2 months). Peritonitis was the primary cause of TK catheter removal (46.7%). Kaplan-Meier survival analysis revealed a significantly shorter lifespan of TK catheters in those affected with peritonitis (48.7 months, 95% CI 9.6, 56.4) compared to those without peritonitis (63.7 months, 95% CI 28.0, 99.5). Logistic regression analysis showed that only peritonitis was predictive of poor TK catheter outcomes (p<0.001, OR=4.62, 95%CI 1.13, 4.95). Probability of poor TK outcome=1/(1 + e-r) where r=1.53(peritonitis)-0.71.

Conclusion: Peritonitis is an important predictor of poor TK outcome with a shorter lifespan.

FP1-4: HEALTH-RELATED QUAILTY OF LIFE IN CHINESE CHILDREN AND ADOLESCENTS IN ESRD PROGRAM

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Introduction: Advanced medico-technological support has enhanced patient survival; other than medical outcome, health-related quality of life (HRQOL) becomes also an important outcome measure for chronic illnesses like those in ESRD program.

Method: This study was conducted at our Paediatric Nephrology Centre in June-July 2006 and March-April 2008. In 2006, all Chinese patients aged < 21 years enrolled in ESRD program for more than three months were recruited. In 2008, we recruited new eligible patients, and interviewed again those patients switching from automated peritoneal dialysis (APD) to renal transplantation (TX). A standardized Chinese questionnaire with subsequent transformed scores of 0-100 was used to reflect the HRQOL in 7 aspects, including general well being, physical ability, school/work performance, peer activities and relationships, family activities & relationships, sleep problems, worries and concerns.

Result: Twenty-three patients undergoing APD and 27 transplanted patients were interviewed. APD and transplanted patients and their parents gave highest scores to family relationships (83–93) and peer activities & relationships (79–90) and lowest scores to physical activities (44–71). Comparing with APD patients, transplanted patients reported significantly less sleeping problems (92 vs. 80; p=0.019). In parents' perspective, transplanted patients had significantly less sleeping problems (91 vs. 71; p=0.02), better physical activities (67 vs. 44; p=0.045) and higher total scores (80 vs. 68; p=0.042). Six eligible patients switched from APD to TX. After TX, patients and their parents gave higher total and individual scores but all did not reach statistical significance except parents viewed significantly improved peer relationship (89 vs. 80; p=0.011).

Conclusion: APD and transplanted patients have not much problems in peer and family relationship despite disease burden. With good dialysis care and support, patients undergoing APD have comparable HRQOL as transplanted patients except significantly more sleeping problems that may be explained by the procedures of APD during sleep.

FP1-5: OUTCOMES OF PAEDIATRIC KIDNEY TRANSPLANTS AFTER CONVERSION TO SIROLIMUS

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We report results of paediatric kidney transplant recipients converted from calcineurin inhibitors (CNI) to sirolimus (SRL).

Since 2003, 15 child/adolescent recipients have received SRL at our centre. All had initially received basiliximab induction with maintenance CNI, prednisone, and either azathioprine or mycophenolate. The median age at time of transplant was 11 years (range 23 m-16 y). SRL was introduced at a median time of 16 months posttransplant (range 2–90 months). CNIs were discontinued in 14 and reduced in 1. All were biopsied prior to conversion. Complete follow up was available for 353 patient-months of SRL treatment (mean 23±13 months/recipient). SRL was discontinued in one because of upper limb oedema and one graft failed due to non-adherence. SRL was otherwise well tolerated. Mean cholesterol levels 12 months after SRL were similar to baseline (4.78±1.25 vs 4.86±0.77 mmol/L, baseline vs 12 months post conversion respectively) but there was an increase in the use of statin therapy (mean atorvastatin dose 1.7 vs 11.3 mg/day; baseline vs 12 months. $P<0.01$). No adverse effects of statin therapy occurred. The mean triglyceride level increased with SRL use (2.0±1.7 vs 2.7±1.4; baseline vs 12 months. $p<0.05$). Haemoglobin levels were stable over 12 months (115±12 vs 117±19; baseline vs 12 months). Four recipients have received darbopoetin therapy. The most recent ten recipients started SRL within 20 months of transplant; all had histological evidence suggestive of either acute or chronic CNI nephrotoxicity. Eight of this group have been followed for at least 12 months. The mean GFR was unchanged during the first 12 months after conversion (64±12 vs 62 ±12 ml/min/1.73m2; baseline vs 12 months after conversion) but had fallen in those seven patients followed for 24 months (55±15 ml/min/1.73m2. $p<0.05$ vs baseline). Four recipients had episodes of steroid reversible acute rejection (AR) while taking SRL. We compared the number of AR episodes in the 9 recipients treated by SRL within 20 months post-transplant to an age-matched historical cohort who had been treated with a CNI based regimen (n=9). All in the control group had been treated for suspected AR between 2 and 5 years after transplant. Rates of rejection after SRL conversion were less than in controls during years 2 to 5 after transplant (0.01 vs 0.1 rejection episodes per month in SRL vs control; $p<0.01$).

We conclude that SRL conversion is well tolerated by paediatric kidney transplant recipients and offers at least as good a degree of protection form acute rejection as CNI based regimens.

FP1-6: SPECIALIST PEDIATRIC DIALYSIS NURSING IMPROVES OUTCOMES IN CHILDREN ON CHRONIC PERITONEAL DIALYSIS (PD)

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Introduction: Chronic PD for children in Singapore was instituted in 1988 at the National University Hospital. However, adult renal nurses provided the dialysis services during the first 10 years. As children with end stage renal disease have special needs, in 1998, a dedicated pediatric renal nursing team was recruited to manage the programme. This study was conducted to determine the impact of the specialist pediatric renal nursing service on PD-related outcomes during the 2 nursing periods.

Method: Data from all children (n=82, mean age 12.10±5.33 years) who entered the dialysis program during the two 10-year dialysis periods were analyzed: Period 1 from January 1988 to December 1997 (n=21,) and Period 2 from January 1998 to December 2007 (n=61). Rates of peritonitis, exit site infection, and modality change per patient-year of dialysis were used as indicators of nursing practice. The difference between the 2 study periods was compared using Poisson Regression.

Result: The mean ages of the patients at start of dialysis in Period 1 (10.36±5.12 years) and Period 2 (12.70±5.32 years) were similar. The peritonitis rate per patient-year was significantly higher in Period 1 (1.10) compared to Period 2 (0.30), with a relative risk of 1.68 (95% CI 1.13-2.50) ($p=0.01$). The exit site infection rates per patient-year was also worse in Period 1 (1.21) compared to Period 2 (0.25), with a relative risk of 2.09 (95%CI 1.39-3.13) ($p<0.001$). Although 38% of children in Period 1 compared to 28% in Period 2 had modality change due to peritonitis, there was no significant difference in the rate of modality change per patient-year between Period 1 (0.21) and Period 2 (0.09).

Conclusion: Establishment of a specialist pediatric renal nursing team for the management of children on chronic PD resulted in a significant improvement in the outcomes as measured by the decrease in technique-related complications.

FP2-1: RITUXIMAB IN PATIENTS WITH DIFFICULT NEPHROTIC SYNDROME

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Introduction: While most patients with idiopathic nephrotic syndrome (INS) achieve remission with daily corticosteroid therapy, 10-20% are resistant (SRNS) and about 30% show steroid dependence (SDNS). We examined the efficacy of treatment with rituximab, a chimeric monoclonal antibody against CD20 antigen on B lymphocytes, in patients with SRNS or SDNS refractory to standard therapy.

Method: Following ethical approval and parental consent, patient demographics, renal diagnosis, previous therapeutic regimens and biochemical parameters such as serum albumin, creatinine, immunoglobulins and urinary protein to creatinine ratio were recorded. Rituximab was administered by IV infusion at a dose of 375 mg / m2 once weekly for 4 weeks in SRNS and for 2 weeks in SDNS. All patients continued to receive therapy with calcineurin inhibitors, alternate-day prednisolone, or both; cotrimoxazole prophylaxis was given for 6 months. For patients with SDNS, rituximab was administered while in remission.

Result: Sixteen patients (9 boys) including 11 with SRNS (5 initial resistance; 6 late resistance) and 5 with SDNS, between the ages of 4 and 14 years were included. Renal histology showed minimal change

disease (MCD) in 5 and focal segmental glomerulosclerosis (FSGS) in 7 of the patients biopsied. The duration of nephrotic syndrome was 4± 1.2 years prior to inclusion and the mean follow-up after treatment was 1.2±0.4 years. All patients had received conventional immunosuppressive therapy (including cyclophosphamide and calcineurin inhibitors) prior to receiving rituximab. In patients with SRNS, the mean ratio of urinary protein to creatinine decreased from 7.2 at baseline to 1.8 at last follow-up. The mean serum albumin level rose from 1.7 g/dl (1.7 in initial; 1.6 in late resistance) to 2.6 g/dl (1.9 in initial; 3.1 in late resistance) and the mean cholesterol level declined from 482 to 278 mg/dl at last follow-up. The rise in serum albumin was more in MCD group than in those with FSGS, although the difference was not statistically significant. Following treatment, the one-year outcome was as follows: complete remission (1), partial remission (9), non-response (1).In the sub-group with steroid dependence, remission was sustained during a mean follow-up of 0.7±0.1 years. The differences in leukocyte counts and levels of IgG were not significant, serum creatinine levels remained stable throughout the study and none of the patients had serious infections.

Conclusion: Rituximab was used in difficult NS with mostly favourable outcome and no serious adverse events. However, the long-term efficacy and safety of rituximab in patients with NS needs further evaluation and prospective randomized controlled trials.

FP2-2: LONG VERSUS SHORT COURSE CORTICOSTEROID THERAPY FOR NEPHROTIC SYNDROME IN CHILDREN

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Introduction: This study aims to compare the effect of 2-month steroid treatment (short course) according to International study of Kidney Disease of Children (ISKDC) versus 6-month treatment (long course) on the clinical course of relapses during a two-year follow up in a single centre.

Method: This is a retrospective cohort study. Medical records of all patients aged 1 to 18 years with first episode steroid sensitive nephrotic syndrome (SSNS) seen in Princess Margaret Hospital between 1997 to 2006 were reviewed. Patients who were previously treated were excluded from the study. Patients were divided into two groups. Group I included patients treated with short course steroid, while group II included patients treated with long course steroid. Clinical parameters including age, sex, relapse rate, rate of sustained remission, mean duration of hospital stay within the two-year follow up were compared.

Result: A total of 46 patients were included in the study. Group I consisted of 22 patients (mean age of 5, M:F=17:5). Group II consisted of 24 patients (mean age of 6, M:F=15:9). The demographics were comparable between the two groups. Patients treated with long course steroid had significantly lower relapse rate (58% versus 90%, P=0.018, fisher's exact test. Odds ratio=7.1, 95%CI 1.35–37.7), higher rate of sustained remission (log rank value=10.5, P=0.0012), and shorter mean duration of hospital stay (16d versus 23 d, P=0.017) at the end of two-year follow up. No significant side effects of long term steroid such as growth retardation and hypertension were identified.

Conclusion: In our experience, 6-month therapy is superior to 2-month therapy for the treatment of first episode SSNS in children. It reduces subsequent relapse rate without increasing the frequency of severe side effects related to steroid use.

FP2-3: URINARY MONOCYTE CHEMOATTRACTANT PROTEIN-1 IS A POTENTIAL BIOMARKER FOR CELLULAR CRESCENT IN RENAL DISEASE

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Introduction: Cellular Crescent is a common pathological change of renal disease, which indicates that the renal disease is severe. Since cellular crescent can reverse after timely aggressive therapy, earlier detection of it is of great help for therapy. Although renal pathology is a golden standard for assessment of crescent, its application is limited because of the inherent disadvantage. Glomerular expression of monocyte chemoattractant protein-1 (MCP-1), β-catenin and cytokeratin19 (CK19) have been confirmed to associate with the severity of cellular crescent, their potential clinical applications were investigated in this study aimed to find non invasive urinary crescent biomarker.

Method: Firstly, 124 renal disease patients including 52 with cellular crescent and 72 without cellular crescent, and 20 healthy controls were studied. The urinary level of MCP-1, β-catenin and cytokeratin19 (CK19) were detected by ELISA method. The severity of cellular crescent was reflected by crescent index which evaluated by morphometric method. The sensitivity and specificity of the three molecules in urine for diagnosis of cellular crescent were evaluated by receiver operating characteristic (ROC) analysis. Then, the results were tested by another 80 patients with renal disease including cellular crescent group (n=41) and non cellular crescent group (n=39), and 25 healthy controls.

Result: For the first 124 patients, urinary MCP-1, β-catenin and CK19 levels correlated with crescent index significantly ($r=0.75$, $r=0.21$, $r=0.63$, $p<0.01$).By ROC analysis, the area under curve (AUC) of urinary MCP-1, β-catenin and CK19 were 0.691, 0.686 and 0.501, respectively. For the second patients group, the AUC of urinary MCP-1, β-catenin and CK19 were 0.83, 0.678 and 0.569, respectively.

Conclusion: The urinary level of MCP-1 and β-catenin seemed to be potential non invasive marker for cellular crescent, especially for MCP-1. The finding might help guiding the timely aggressive treatment for patients with cellular crescent.

FP2-4: LONG-TERM EFFECTS AND OUTCOMES OF CYCLOSPORINE IN CHILDREN WITH STEROID-RESISTANT IDIOPATHIC NEPHROTIC SYNDROME

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Introduction: To observe the effects of long-term cyclosporine A (CsA) treatment in 20 children with steroid-resistant idiopathic nephrotic syndrome (SRNS) and analyze the relevant influencing factors of CsA therapy.

Method: Twenty children with SRNS received CsA therapy for 2 years between 2001 to 2006 in the Department of Nephrology. The

mean age of children at initiation of CsA therapy was 5.5 years. Initial renal histology showed minimal change (MCNS) in 15 patients, focal segmental glomerulosclerosis (FSGS) in 4 patients and one mesangial proliferative glomerulonephritis (MsPGN). The starting dose of CsA was 3-5 mg•kg-1•d-1, adjusted to maintain a trough level of 100~200 ng/ml in the first 6 months. After one year, a low dose of CsA(1~3 mg•kg-1•d-1)with a a trough level of 40~70 ng/ml was accepted to maintain remission for 1 year.

Result: (1) Complete remission, partial remission and resistant to CsA were observed in 65%, 20% and 15%, respectively. Eleven patients who was complete remission discontinued CsA, 5 (45%) patients relapsed. (2) MCNS showed a better response to CsA than non-MCNS, but the difference showed no significant (P>0.05). (3) Hypertrichosis, gingival hyperplasia and hypertension occurred in75%, 25% and 10% of the patients, respectively. Two patients were found to have renal impairment (> 30% rise of serum creatinine) and recovered in 2 weeks. Post-therapy biopsies performed in 3 patients (2 with FSGS and one with MCNS) didn't show relevant tubulointerstitial fibrosis. 2 patients with FSGS of the twenty cases progressed into end-stage renal failure.

Conclusion: Long-term CsA treatment was confirmed to be effective in children with SRNS. Renal fibrosis was rare in our patients treated with a low dose of CsA for 2 years.

FP2-5: EARLY RELAPSE POST-CYCLOPHOSPHAMIDE THERAPY IS A PREDICTOR OF POOR LONG-TERM OUTCOME IN STEROID-DEPENDENT AND RESISTANT MINIMAL CHANGE NEPHROTIC SYNDROME (MCNS)

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Introduction: Cyclophosphamide remains a significant option in treating children with steroid-dependent and steroid-resistant MCNS despite associated side effects. This study aimed at identifying factors associated with long-term relapsing disease post-cyclophosphamide therapy.

Method: Clinical and histopathological data of all children with steroid-dependent or steroid-resistant MCNS who underwent renal biopsy and received 12-weeks of cyclophosphamide as second-line treatment were reviewed. Poor long-term outcome was defined as presence of any immunosuppressive medication 2 years post-cyclophosphamide therapy. Parameters studied included gender, race, age at diagnosis, hypertension, hematuria, and serum creatinine at initial presentation, global sclerosis, mesangial expansion, tubular atrophy and IgM deposits on renal biopsy, and early poor response, defined as no response or relapse within 2 months post-cyclophosphamide therapy. Factors predictive of poor long-term outcome post-cyclophosphamide therapy were analysed by multivariate logistic regression.

Result: Forty-six children had MCNS, 21 (45.6%) were steroid-dependent and 25 (54.4%) steroid-resistant. Median age at disease onset was 2.9 years (range 1-12.1 years). Mean follow-up period from start of cyclophosphamide therapy was 11.53±6.35 years. Twenty-nine (63%) children responded to cyclophosphamide initially, however 2 (6.9%) relapsed within 2 months post-cyclophosphamide therapy. Seven (15.2%) were hypertensive, 7 (15.2%) had hematuria and 4 (8.7%) had elevated creatinine at presentation. Thirty-one (67.4%) of biopsies were positive for mesangial IgM, 10 (21.7%) had global sclerotic lesions and 11 (23.9%) had tubular atrophy. Regression analysis showed that only early poor response to cyclophosphamide was predictive of poor long-term outcome (p<0.018, OR=5.8, 95%CI=1.4-24.4). Using this model, the probability of poor long-term outcome post-cyclophosphamide therapy was expressed by the equation: P=1/(1+exp–r), where r=1.75(early poor response)-0.74. Early poor response to cyclophosphamide had a positive predictive value of 84.7% in predicting poor long-term outcome.

Conclusion: Steroid-dependent or resistant MCNS patients with early poor response to cyclophosphamide therapy were more likely to have long-term relapsing disease requiring immunosuppressive therapy.

FP2-7: A COMPARATIVE STUDY OF EFFICACY AND SIDE EFFECTS OF PREDNISOLONE AND DEFLAZACORT IN THE TREATMENT OF CHILDHOOD NEPHROTIC SYNDROME

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Introduction: Glucocorticoids are mainstay of management of nephrotic syndrome. Deflazacort is a newer glucocorticoid which not only have similar efficacy but also lower incidence of side effects compared to prednisolone, however its high cost is the major drawback. In order to assess whether Deflazacort can replace prednisolone we compared effectiveness and the adverse effects of prednisolone and Deflazacort.

Method: 60 patients (30 on deflazacort therapy & 30 on prednisolone therapy) of Nephrotic syndrome in Nephrology Unit, Pt.J.N.M.Medical College & GBG Kidney care Hospital, Raipur from april2007 to april2008 were studied. All patients were subjected to routine investigations.

Result: •Among the patients treated with prednisolone 66.67% were having hypocalcemia whereas it was 40% among the patients treated with Deflazacort (p value<0.001). •Hyperphosphatemia was present in 40% of the patients on prednisolone while it was 20% among the patients on deflazacort (p value<0.005). •40% of the nephrotic syndrome patients treated with prednisolone had raised calcium phosphate product while it was raised in 20% of the patients treated with Deflazacort (p value<0.005). •70% of the patients on prednisolone therapy had hypercholesterolemia whereas it was 53.33% with Deflazacort therapy (p value<0.02). •Posterior subcapsular cataract developed in 6.66% of the patients treated with prednisolone while none of the patients with deflazacort developed it. •80% of the nephrotic syndrome patients treated with prednisolone had steroid induced side effects like cushing's habitus (moon like face), purple striae, muscular weakness, psychiatric disturbances while it was seen only in 40% of the nephrotic syndrome patients treated with Deflazacort (p value<0.001). •Relapse rate was 30% among the patients treated with Deflazacort while it was 43.33% with prednisolone (p value<0.1).

Conclusion: This study demonstrate that although deflazacort is mild expensive than prednisolone it was as effective as prednisolone and further more it had fewer steroid induced side effects.

FP3-2: Fever Duration and Renal Scar in Pediatric Urinary Tract Infection

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Introduction: Urinary tract infections(UTIs) are not uncommon findings in febrile pediatric patients and approximately one third of patients of UTI may have renal scars. This research was intended to establish the relationship between duration of fever and renal scars.

Method: 143 patients medical records were reviewed retrospectively. Inclusion criteria were as follows: 1) fever as defined by an axillary temperature ≥37.5°C, 2) accurate history of fever duration and the use of antibiotics 3) no previous history of UTI and 4) positive urine culture. We observed whether the longer fever duration could be associated with the development of initial renal defects and subsequent renal scars, increased C-reactive protein(CRP), leukocytosis and the presence of vesicoureteral reflux(VUR).

Result: 1) Patients with longer fever duration after antibiotics showed more frequent initial renal defects(P=0.014). However, fever duration before antibiotics was not associated with the development of initial renal defects(P=0.244). 2) Incidence of renal scar was increased with fever duration before antibiotics(P=0.006) and fever duration after antibiotics(P=0.015). 3) CRP was correlated with the fever duration after antibiotics(r=0.287, P=0.003). 4) There were no relationships between fever duration and VUR(P>0.05).

Conclusion: Our data suggest that fever duration before/after antibiotics are significantly associated with the increased development of renal scars in pediatric UTI.

FP3-3: IMAGING STUDIES FOR FIRST URINARY TRACT INFECTION IN INFANTS LESS THAN 6 MONTHS OLD: CAN IT BE MORE SELECTIVE?

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Introduction: Universal agreement in investigating urinary tract infection (UTI) in children with imaging is lacking. This study serves to evaluate the selective approach of imaging infants < 6 months old with UTI according to risk features suggested in UTI Guidelines by National Institute for Health and Clinical Excellence (NICE) 2007.

Method: This is a retrospective study of infants < 6 months old with first UTI from Jan 02 to Dec 06 having complete set of imaging studies including ultrasound kidneys (US), micturating cystourethrogram (MCU) and DMSA scan at > 6 months (those with known urological abnormalities and antenatal hydronephrosis were excluded). They were evaluated against a set of risk features according to NICE UTI guidelines i.e. serious illness, poor urine flow, abdominal mass, raised SCr, septicaemia, non-Ecoli UTI and failure to respond to treatment with suitable antibiotics within 48 hrs. Those having any one of these were classified as atypical and those having none as typical.

Result: 165 cases were identified. 115 infants having all three imaging studies completed were reviewed. Atypical group (32) had risk features of non-Ecoli UTI in 16, septicaemia 7 and failure to respond to antibiotics 9. None had other risk features. There were 6 scars, 26 refluxing ureters and 6 hydronephrosis (> 5 mm). In typical group (83), there were 23 scars, 22 refluxing ureters, 5 hydronephrosis, 2 hydroureter-hydronephrosis and 1 PUJ. 76 infants had normal US; and if only those (7) with abnormal US were further investigated as suggested by the Guidelines, 18 refluxing ureters and 23 scars would be left undiagnosed.

Conclusion: According to the NICE UTI Guidelines, typical UTI will only be investigated with US, and only abnormal US will be followed by MCU. For the present group, the suggested selective imaging approach would leave a significant number of VUR and renal scars undiagnosed. Not until the significance of long term outcome of scars & VUR is clarified definitely, it may not be an optimal practice for infants < 6 months old with first UTI.

FP3-4: KIDNEY BIOPSY IN CHILDREN - ? A DAY CARE PROCEDURE

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Introduction: Introduction: Percutaneous renal biopsy is a safe procedure. Fear of complications require hospitalization adding to treatment cost, which may be avoided. Aim: To study the timing of complications associated with a percutaneous kidney biopsy in children in order to determine the feasibility of it being performed as a day-care procedure.

Method: Material and Methods: Children undergoing kidney biopsy over 2 yrs included prospectively. Coagulation studies and renal parameters noted. Procedure not carried out under real time ultrasonography. Children sedated with midazolam and ketamine. Vital parameters monitored as a baseline and complications noted on a 3 hrly basis for 24 hours post-procedure, for timing of onset and duration. Statistical analysis: Univariate analysis using chi-square test.

Result: Results: 90 children (M:F::2:1) age11 m-12 y underwent a kidney biopsy. Nephrotic syndrome with atypical features was the commonest indication. Hematuria noted in 27% but only 3.3% required a transfusion. No child required any surgical or radiological intervention. All complications when occurred had onset < 6 hrs but most resolved by 12 hrs. Only 6 (6.6%) children continued to have hematuria beyond 12 hrs and this continued beyond 24 hrs. The occurance of a complication had no bearing to the age, gauge of needle used, baseline serum creatinine or the underlying kidney disease. Monitoring of vital parameters could not predict hematuria before it was manifest.

Conclusion: Conclusions: Age, underlying renal disease, baseline investigations and monitoring of vital parameters could not predict significant hematuria early. Complications after a kidney biopsy are obvious by 6 hrs, resolving in most within 12 hrs. All patients undergoing kidney biopsies should be kept under observation for minimum 12 hours. Those with macroscopic hematuria beyond 12 hrs or requiring intervention may be admitted. In others the kidney biopsy can safely be carried out as a daycare procedure.

FP3-5: FOLLOW-UP OUTCOME OF CHILDREN WITH ABNORMAL RESULTS IN URINE SCREENING IN SHANGHAI

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Introduction: Between 2003 and 2008 urine screening was carried out in some kindergartens and schools in Shanghai. We hope to discuss the characteristics of kidney diseases detected in urine screening.

Method: In Shanghai 46,171 school children (2003-2005) and all children in kindergartens (2006-2007, about 270,000 children/year) underwent urine screening. Children with abnormal results were asked to be followed up in designated hospitals. We documented and analyzed the follow-up records.

Result: During 2003-Feb 2008, 419 children (207 boys and 212 girls, from 2-year to 18-year) with abnormal screening results went to the hospitals, among whom 367 were hematuria, 36 were proteinuria, 5 were hematuria combining with proteinuria, 11 were leucocyturia and 4 cases diagnosed as nephrotic syndrome. 39 cases underwent renal biopsies which consisted of 32/39 hematuria, 3/39 proteinuria, 3/39 nephrotic syndrome and 1/39 hematuria with proteinuria. The pathologic diagnosis showed as following: 20/39 showed minor change, 2/39 minimal change disease (MCD), 7/39 IgA nephrology (IgAN), 7/39 thin basement membrane nephrology (TBMN), 2/39 mesangial proliferative glomerulonephritis (MsPGN), 1/39 focal segmental glomerulosclerosis (FSGS). We noticed that four patients with nephrotic syndrome were detected in kindergarten by urine screening. Seven children were diagnosed with IgAN by renal biopsy. 4/7 were long term hematuria, 2/7 proteinuria and 1/7 hematuria associating with proteinuria.

Conclusion: Most problem detected in urine screening was mild in our study but some may have poor outcome which needed close follow-up for a long time. Urine screening was well done in kindergartens in Shanghai.

FP3-6: OBESITY- RELATED GLOMERULOPATHY IN CHILDREN

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Introduction: The prevalence of obesity all over the world has been rising. It is well documented that obese patients are at greater risks to develop hypertension, coronary vascular disease, and insulin resistance, and more attention has been paid to the impact of obesity on kidney, recently. Since the first description regarding the relationship between massive obesity and proteinuria in 1974, increased evidence demonstrated that obesity- related glomerulopathy (ORG) should be identified as an isolated complication of obesity in adults. It is, however, unclear whether this condition exists even in children with obesity. This study was conducted to know whether obese children have higher risk of developing to ORG as well as in adult.

Method: Eighteen hundred and thirty nine children aged 6–14 years with abnormal urinary findings by the screening program in Tokyo in 2007 were enrolled. Tentative diagnoses were made based on the criteria for this program (Kaneko K, et al. Pediatr Nephrol 19:499; 2004): [group A] healthy in 592; [group B] orthostatic proteinuria in 112; [group C] asymptomatic proteinuria in 28; [group D] minimal hematuria in 572; [group E] asymptomatic hematuria in 255; [group F] nephritis/ suspected nephritis in 27. An obesity index based on the standard weight was compared among the groups using Kruskal-Wallis test. P value less than 0.05 was considered significant.

Result: The obesity index (%) was significantly higher in group F (nephritis/ suspected nephritis) than in other groups: (mean ± standard deviation:group A 2.1±15.4, group B -3.6±12.6, group C -3.3±18.1, group D 2.7±15.3, group E 3.9±15.0, group F 8.8±23.1, $P<0.05$). The number of obese children defined as an obesity index of 20% or more was greater in group F (18.5%) than in group A (10.0%).

Conclusion: ORG develops even in childhood and early intervention to lose weight should be considered, as the benefit of weight loss in ORG in adult is reported to be limited.

FP4-1: AUTOANTIBODIES TO COMPLEMENT FACTOR H ARE AN IMPORTANT CAUSE OF ATYPICAL HEMOLYTIC UREMIC SYNDROME (HUS)

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Introduction: Mutations in genes encoding complement regulators predispose to development of atypical HUS. We examined for abnormalities of the complement pathway in patients with atypical HUS.

Method: EDTA samples were collected from index patients presenting with atypical HUS and their families, before instituting plasma therapy or blood transfusion. Plasma concentrations of C4, C3, factors B, H and I, CD46 and anti-factor H antibodies were measured. Genes encoding factor H, I and CD46 were sequenced. Screening by multiplex ligation–dependent probe amplification for complement factor H-related (CFHR) 1 and CFHR3 genes was performed. Patients with anti-factor H antibodies received double-volume plasmapheresis daily till activity subsided, followed by 4 alternate-day and 4 twice-weekly plasmapheresis. They also received IV immunoglobulin (2 g/kg), IV cyclophosphamide (500 mg/m2 q 3-weekly) and prednisolone (1 mg/kg).

Result: Fourteen patients (12 boys) from 7 months to 15 yrs were studied. Renal histology showed features of thrombotic microangiopathy involving arterioles and capillaries. One patient had complete deficiency of factor H due to a homozygous mutation. Eight patients tested positive for anti-factor H antibodies (titer 194-32,000 UA); 7 had antibody titer above 1500 UA and were dialysis dependent. The antibody titer and disease activity reduced in all patients; 7 progressed to end stage renal disease. Genetic studies showed homozygous deletion of CFHR1 and CFHR3 genes in 4 patients with anti-factor H antibodies.

Conclusion: Antibodies to factor H appears to be an important cause of atypical HUS in India. The antibody titer correlated directly with the disease severity. Deletion of CFHR1 and CFHR3 genes might genetically predispose to development of anti-factor H antibodies, and occurrence of HUS.

FP4-2: HIGH-DOSE MIZORIBINE THERAPY FOR CHILDREN WITH SEVERE TYPE 1 MEMBRANOPROLIFERATIVE GLOMERULONEPHRITIS

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Introduction: To determine the safety and effectiveness of oral high-dose mizoribine (MZR) for children with severe Type 1 membrano-proliferative glomerulonephritis (MPGN).

Method: Between 2003 and 2007, 5 children diagnosed with primary Type 1 MPGN by renal biopsy were treated with intravenous high-dose methylprednisolone pulse therapy, followed by alternative-day prednisolone that was slowly tapered off over several years. However, 2 of them continued to show signs of proteinuria and hypocomplementemia or disease flare despite steroid therapy, and they were both were diagnosed with severe Type 1 MPGN. Each patient underwent a repeat renal biopsy at 23 or 19 months after the first biopsy. Repeat biopsy findings revealed no histological improvement in either of them. Therefore, they were both administered with daily oral MZR due to unsatisfactory steroid effects. The dose of MZR was adjusted to achieve a peak blood level of around 3.0 mcg/ml to maximize clinical efficiency.

Result: Urinary protein excretion decreased from 3+ to trace in one of the patients at 18 months and from 2 + to negative in the other at 13 months after starting MZR therapy. The serum C3 level increased to the normal level in the patient with persistent hypocomplementemia. The dose of prednisolone was also tapered in both. Neither of them developed adverse reactions.

Conclusion: Our results suggested that MZR at an optimal blood concentration is effective and safe for patients with severe Type 1 MPGN. Further evaluation is warranted for the use of MZR therapy in a long-term follow up study, and children with severe type 1 MPGN should be included in a prospective-randomized trial of MZR therapy.

FP4-3: PREVALENCE OF VITAMIN D INSUFFICIENCY AND THE EFFECT OF CHOLECALCIFEROL SUPPLEMENTATION IN CHILDREN WITH CHRONIC KIDNEY DISEASE (CKD)

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Introduction: Vitamin D insufficiency (serum 25-hydroxyvitamin D (25,OHD) level < 30 ng/ml) is common in patients with CKD and may contribute to mineral bone disease. We estimated the prevalence of vitamin D insufficiency in children with CKD, and examined the effect of cholecalciferol supplementation on serum levels of 25,OHD, intact parathyroid hormone (PTH), calcium and phosphorus.

Method: In a prospective interventional study children with estimated GFR between 15 to 89 ml/min/1.73 m2 were given 6, 00,000 IU of cholecalciferol. Children on vitamin D supplements and those with serum calcium level >10.5 mg/dl and phosphorus > 6.0 were excluded. Primary (25,OHD, PTH levels) and secondary (calcium, phosphorus, alkaline phosphatase levels and urinary protein excretion) outcomes were measured at baseline and 6 weeks.

Result: 40 children (85% boys) with a mean age of 7.7±3.8 (range 2-15) yr were studied. 33 (82.5%) children had vitamin D insufficiency at baseline. Of these, 15 children had 25,OHD levels < 15 ng/ml. The median serum 25,OHD levels increased from 18.6 (95% confidence interval [CI], 15.4–23.4) ng/ml at baseline to 48.5 (42.1–56.8) ng/ml at 6 weeks (P=0.000) while the PTH levels declined from 55.3 (47.1–77.8) pg/ml to 41.4 (31.2–56.8) pg/ml (P=0.01). The calcium, phosphorus and alkaline phosphatase levels did not change significantly from the baseline. The median urinary protein to creatinine ratio decreased from 0.30 (0.03–1.5) to 0.61 (0.08–2.26) mg/mg (P=0.07). There was no correlation between serum 25,OHD and PTH levels.

Conclusion: Vitamin D insufficiency is highly prevalent in children with CKD and cholecalciferol supplementation appears to be an effective treatment in correcting vitamin D status and reducing PTH levels.

FP4-4: INFUSION OF HUMAN ADIPOCYTE DERIVED STEM CELLS HELP THE RECOVERY OF ACUTE RENAL FAILURE

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Introduction: The aim of this study was to explore the role of human adipocyte derived stem cells (hADSCs) in a repair of acute renal failure.

Method: For renal failure, acute ischemic tubular necrosis was induced Spraque-Dawley rats by clamping bilateral renal arteries for 40 min. hADSCs obtained from elective liposuction procedures. After steps of isolation, stem cells were seeded at 1~2×105/cm2 in complete medium. Dextran-coated iron was used as a labeling agent and Poly-l-lysine (MW 70~150 kDa) as a transfection agent. Before releasing the clamps, three shots of iron labeled stem cells (1×106/0.2 cc) for left kidney or PBS for right kidney were injected at upper, middle and lower part of renal cortex directly. MRI images, one day after reflow, confirmed presence of iron labeled cells in the renal cortex. The examination on both kidneys was done after 1, 3 days, 1,2,4,6,10 and 15 weeks(n=5 in each). The evaluation of the degree of tubular necrosis (PAS stain) and the locations of stem cells(prussian blue stain) was done by light and electron microscopically.

Result: Serum creatinine on day 1 was 2.7±0.4 mg/dL, on day 3, 2.2 ±1.4 mg/dL and on the first week 0.8±0.3 mg/dL. After then, it has no significant change among groups. Tubular injury was scored by counting the number of damaged tubules in random selected 100 cross sectioned tubules. On day 1, 47±16 of left side and 76±11 of right; day 3, 11±17 and 55±21 were scored. From 1 week group, the tubular necrosis had been recovered well. Cells positive stained for prussian blue located at peritubular area. However, tubular epithelial cells or glomerular cells were not positive. Electron microscopically, fine iron granules with electron density were found in their cytoplasm, which were thought as injected stem cells. FISH for anti-human chromosome (CEP) 17 probe were positive to those cells.

Conclusion: We demonstrate stem cells contribute to repair the tubular epithelium in acute renal failure model, not by tubulogenesis directly but by indirect effects, which should be evaluated.

FP5-1: PODOCINR168H, NOT PODOCINV165X, PLAYS MUCH MORE IMPORTANT ROLE ON PODOCYTE INJURY

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Introduction: A composite heterozygous NPHS2 mutation was verified for the first time in a Chinese familial FSGS, which resulted in a truncated (podocinV165X) and a missense mutant podocin (podocinR168H). Both mutant sites were localized to a same domain in podocin C-terminu. Nevertheless, our previous studies showed that the binding characteristics of both mutant podocins with N- and C-terminal antibodies were obviously different, which suggested that the two mutant podocins might play a different role and involve different pathogenesis. This study here aimed to explore the different roles of the two mutant podocins on podocyte injury via in vitro experiment.

Method: Immortalized mouse podocyte was transfected with the vectors containing wild or mutant podocins. Podocyte injuries were evaluated by FITC-Annexin V. Effects of wild and both mutations on nephrin, CD2AP and the recently identified podocyte ion channel TRPC6 were displayed by immunofluorescence staining and western blot. Cytosolic free Ca2+ was measured with fluo-3AM. To evaluate TRPC6 role in cell injuries, podocyte was co-transfected with TRPC6 siRNA and wild or mutant vectors.

Result: Both wild and mutant podocins significantly increased podocyte apoptosis. Wild and V165X was partially targeted to membrane, whereas R168H was only distributed around nuclei. Both wild and mutant podocins showed co-localization with nephrin, CD2AP and TRPC6. Nephrin, CD2AP and TRPC6 were obviously up-regulated in R168H-podocytes, while TRPC6 and CD2AP was evidently increased in wild and V165X-podocytes, respectively. A cortical cytoskeleton was observed in wild and V165X-podocytes, whereas R168H seriously disturbed cytoskeleton arrangement. Both wild and mutant podocins increased cytosolic free Ca2+ level. Podocyte apoptosis, Ca2+ level and expressions of other slit diaphragm molecules did not change in just TRPC6 knockdown cells. With the knockdown of TRPC6, Ca2+ level and podocyte apoptosis decreased significantly only in wild and V165X-podocytes.

Conclusion: PodocinR168H caused much more significant changes of podocyte molecules and cytoskeleton, which prompted that podocinR168H plays more critical role on podocyte injury.

FP5-2: ACE GENE POLYMORPHISM CAN NOT PREDICT THE PROGRESSION RATE IN IRANIAN CHILDREN WITH FSGS (A TWO-CENTER STUDY).

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Introduction: Focal segmental glomerulosclerosis (FSGS) is one of the most common glomerulonephritis leading to end stage renal disease (ESRD). A few clinical and paraclinical factors such as hypertension, percentage of cortical interstitial fibrosis,the value of glomerular filtration rate (GFR) at the onset of disease and angiotensin converting enzyme (ACE) gene polymorphism are considered as promoting factors. However, there are controversial reports on relationship between ACE gene polymorphism and rapidity of progression of FSGS to ESRD in different population. AIMS: we investigated the relationship between Insertion/Deletion (I/D) ACE gene polymorphism and rapidity of progression of Focal Segmental Glomerulosclerosis (FSGS) to End Stage Renal Disease (ESRD) in Iranian children.

Method: Forty-one children aged 1 year to 18 years admitted at St AlZahra Hospital, Isfahan and St Ali Asghar Hospital, Tehran, Iran with biopsy proven idiopathic FSGS were enrolled. They were divided into two groups according to the time of progression to renal death. Renal death was defined as glomerular filtration rate (GFR) less than 50 ml/min/1.73m^2 or decreasing GFR more than two times compare to the baseline. Reaching renal death in less or more than two years was assumed as rapid progressors (RP) or slow progressors (SP) respectively. The intron 16 of ACE gene was amplified by PCR technique.Statistical significance was regarded as $P<0.05$.

Result: Twenty-eight patients were male and 13 were female.In 15 RP patients the genotype distribution was DD-26.6%,II-6.7%,ID-66.7%.In 26 SP patients the genotype was similar (DD-38.6%,II-7.6%,ID-53.8%, $P>0.05$). There were no statistically significant differences for ACE I/D gene polymorphism between two groups of patients ($p>0.05$).

Conclusion: Our study revealed no correlation between ACE I/D gene polymorphism and rapidity of progression of FSGS to ESRD in Iranian children.

FP5-3: ANGPTL3 MODULATES GLOMERULAR ENDOTHELIAL CELLS BARRIER PROPERTIES VIA A POSSIBLE SIGNALING PATHWAY INVOLVING IN INTEGRIN αVβ3 AND PI3K/Akt

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Introduction: Some studies have shown that molecules produced by podocyte may influence glomerular endothelial barrier properties. we found the level of Angiopoietin-like protein 3 (Angptl3) mRNA was significantly increased in kidneys of children with minimal change nephrotic syndrome and the level of Angptl3 was increased in the glomerulus of adriamycin rats along with the development of proteinuria. We also confirmed expression of Angptl3 in cytoplasm of podocyte. Experiments confirmed that Angptl3 could bind to integrin αvβ3 of umbilical venous endothelial cell. Integrin αVβ3 is also one of the integrin heterodimers that the glomerular endothelial cells (GEnCs) express. So, we postulate Angptl3 secreted by podocyte have effect on GEnCs and may take part in development of proteinuria. The aim of this study was to investigate the effect of Angptl3 on the barrier properties of GEnCs and the possible signaling pathway.

Method: Permeability of GEnCs was assessed by transendothelial resistance (TEER) and by the diffusion of fluorescein isothiacyanate-labeled bovine serum albumin (FITC-BSA). The level of phospho-specific Akt were delected by Western blot.

Result: We demonstrated that Angptl3 induced decrease in TEER and increase in permeability rates for FITC-BSA, and phosphorylation of Akt was markedly evaluated after GEnCs was treated with Angptl3. We showed pretreatment of LY294002, a PI3K inhibitor, prevented Angptl3-induced decrease in TEER and inhibited increase in FITC-BSA.

We observed pretreatment of LM609, a specific integrin αVβ3 antibody, could significantly blocked Angptl3-induced Akt phosphorylation increase, and inhibited decrease in TEER and increase in FITC-BSA.

Conclusion: Theses studies indicate Angptl3 could significantly increase the permeability of GEnCs monolayer via integrin αVβ3 and PI3K/Akt signaling pathway.

FP5-4: INTRAUTERINE GROWTH RESTRICTION AND POSTNATAL OVERNUTRITION AFFECT THE PROTEOMES OF THE KIDNEYS IN ADULT RATS

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Introduction: Intrauterine growth restriction (IUGR) is associated with hypertension, diabetes and chronic kidney disease in adulthood. Postnatal overnutrition following IUGR may be of pathogenic importance for the development of diabetes and cardiovascular disease. Our previous animal studies have shown IUGR adults have higher urinary excretion of protein and blood pressure than controls and postnatal overnutrition following IUGR cause more severe hypertension and proteinuria than IUGR itself. The aim of this study was to identify the possible pathogenesis of kidney disease in IUGR and the effect of postnatal overnutrition by comparative proteomic approach.

Method: IUGR was induced in rats by isocaloric protein restriction in pregnant dams. IUGR pups were divided into two groups, fed either standard-protein diet (IUGR group) or high-protein diet (HP group). At the age of 12 weeks, kidney proteins were obtained from each group. 2-DE, staining, mass spectrometry and database searching were used. The 2-DE test was repeated three times in each group.

Result: The 2-DE image analysis detected average 727 +/- 58 spots in IUGR group, 740 +/- 43 spots in HP group and 758 +/- 53 spots in control group. The differential proteomic expression analysis between IUGR and control group found 12 proteins had significantly differential expression, which were transcription regulators including prohibitin and ribonuclease UK114, structural molecules including capping protein, enzymes including glutathione s-transferase alpha-1, retinal dehydrogenase1, transketolase and so on. Subsequently, the differential proteomic expression analysis between IUGR and HP group found 16 proteins had significantly differential expression, which were transcription regulators including ribonuclease UK114 and NADH-ubiquinone oxido-reductase, structural molecules including ezrin and gamma-actin, enzymes including catalase, isocitrate dehydrogenase and so on.

Conclusion: Data from this study may provide, at least partly, valuable experimental evidence of proteins involved in the pathogenesis of kidney disease in IUGR and the effect of postnatal overnutrition.

FP5-5: NPHS2 PROMOTER POLYMORPHISMS ARE ASSOCIATED WITH SUSCEPTIBILITY TO SPORADIC PRIMARY NEPHROTIC SYNDROME IN SINGAPORE CHINESE CHILDREN

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Introduction: NPHS2 exonic mutations have been reported extensively in sporadic primary nephrotic syndrome (NS) in Caucasian populations, but are uncommon in Asians. This study examined the NPHS2 promoter region for gene variants and analysed their association with NS in Singapore Chinese children.

Method: Direct sequencing of the 5' promoter and all 8 exons of NPHS2 was performed on genomic DNA from 58 patients with primary sporadic NS (age of diagnosis ranging from 1-20 years) and 55 normal controls. Statistical correlation of genotype with disease was performed using SNPStats (http://bioinfo.iconcologia.net/index.php?module=Snpstats).

Result: We identified five NPHS2 single nucleotide polymorphisms (SNPs) in our patients and controls - three were located in the 5' promoter region (-670C/T, -116C/T and -51G/T); with two in exon 8 (954T/C and 1038A/G). No missense mutations were found. The genotype frequencies were consistent with Hardy-Weinberg expectations. The SNPs in the promoter region were found to be associated with NS under different model fits (-670C/T, recessive model, p=0.042; -116C/T, over-dominant model, p=0.019; -51G/T, additive model, p=0.039). No association with NS was observed for the SNPs in exon 8. Haplotype analysis was performed subsequently for the three promoter region SNPs, which were only weakly linked (-670C/T and -116C/T: r2=0.346, p=0; -670C/T and -51G/T: r2=0.215, p=0; -116C/T and -51G/T: r2=0.098, p=7e-4). Four haplotypes: TCG, CTG, CCT and CCG were observed. The haplotype CCT, was significantly associated with NS (OR: 2.4, 95%CI 1.05-5.50) (p=0.04). This haplotype was responsible for 20% of nephrotic patients, but was only present in 9% of healthy controls. Thirty of the 58 patients (51.7%) were steroid-resistant. Further analysis revealed that the haplotype frequency was 25% in steroid-resistant patients, two-fold that of steroid-dependent patients (14%).

Conclusion: The promoter haplotype of NPHS2 may confer susceptibility to sporadic NS. Functional studies are needed to validate this hypothesis and elucidate possible disease-causing mechanisms.

FP5-7: ANGIOTENSINII AFFECT F-ACTIN IN MOUSE PODOCYTES VIA ERK1/2

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Introduction: The actin cytoskeleton is thought to play a critical role in maintaining podocyte structure and function. Some evidences have shown that angiotensin II (AngII) could induce the cytoskeleton changes in podocytes. Nevertheless, the relevant signal transduction pathway is not fully clarified. Some studies suggested that ERK1/2 MAPK pathway might be connected with the cytoskeleton of some types of cell. Here, it was explored whether ERK1/2 pathway is involved in the effects of AngII on podocyte actin cytoskeleton.

Method: Mouse podocyte clone (kindly provided by Prof. Peter Mundel) was cultured at 33°C for proliferation,then transferred to 37°C and cultured for 1 week to be differentiated. After podocyte was treated with AngII (10-7M) for 2, 5, 10, 15, 30, 45 and 60 min, the protein expressions of ERK1/2 and phosphorylated ERK1/2 were detected by western blotting. TRITC-phalloidin was directly used to stain the actin filaments (F-actin) of podocytes. To verify the key role of ERK1/2 in the podocyte cytoskeleton alterations resulted from AngII, U0126, the specific inhibitor of ERK1/2 pathway was applied for 1 h before AngII treatment.

Result: Phosphorylated ERK1/2 increased markedly ($P<0.05$) at 15 min and persistently to 60 min after AngII treatment. Normal podocytes displayed an obvious and parallel actin stress. In AngII treated podocytes, a cortical pattern of F-actin mainly on cell membrane was observed at 30 min, and marked depolymerization of which occurred obviously at 45 min and persisted to 120 min. Pre-treated with U0126 for 1 h, the ERK phosphorylation was completely inhibited, and the abnormal arrangement of F-actin was partially attenuated in AngII treated podocytes.

Conclusion: AngII activated the ERK1/2 MAPK pathway, which at least partially contributed to the rearrangement of podocyte actin cytoskeleton induced by AngII.

AA-002: LIPID PROFILE IN CHILDHOOD IDIOPATHIC NEPHROTIC SYNDROME AND ITS CORRELATION WITH RELAPSE

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Introduction: Nephrotic syndrome is one of the most common problems among the renal diseases in children. Many patients, about 40% of whom responded well previously to prednisolon, however, relapse frequently, when treatment with prednisolon is tapered off or stopped. Factors for predicting such relapse are yet to be established. Dyslipidaemia during remission could be a predictor of relapse in nephrotic syndrome

Method: A total of 26 hospitalized children of 1-8 years age, suffering from nephrotic syndrome 1st attack were included as cases. Another 22 hospitalized children of same age group, not suffering from any kind of renal diseases, were included as controls. Lipid profile were measured in both cases and controls after initial enrollment. All the cases were treated with oral prednisolon adequately according to ISKDC protocol. During remission, lipid profile were again measured and on the basis of presence of abnormal lipid profile at that time, cases were divided into 2 groups. All the cases were followed-up every monthly for subsequent 6 months to observe development of subsequent relapse.

Result: Among the cases, group-I consisted 16 patients who had normal lipid profile during remission and group-II consisted 10 patients who had abnormal lipid profile during remission. Both groups of cases showed higher mean level of s.cholesterol($p<0.001$), s.LDL ($p<0.001$), s.TG ($p<0.001$) and s.Lp(a) ($p<0.001$) than those of controls during initial diagnosis. During remission, group-II patients showed higher mean serum cholesterol (332.9±105.19 mg/dL vs. 183.13±16.89 mg/dL; $p<0.001$), serum LDL (252±101.67 mg/dL vs. 119.19±21.33 mg/dL; $p<0.001$), and serum TG (182.8±73.83 mg/dL vs. 93.31±20.95 mg/dL; $p<0.001$). 5 patients out of 10 patients of group II (19% of total case) developed subsequent relapse within 6 months follow-up. Among the relapsers, mean cholesterol (334±46 vs. 232±34 mg/dL; $p<0.05$) was significantly higher than that of non-relapsers of group-II patients. On the other hand, no patient of group-I developed relapse within 6 months follow-up.

Conclusion: It may be concluded from the present study that elevated lipid profile, specially serum cholesterol during initial remission, may be a predictor of subsequent relapse in idiopathic childhood nephrotic syndrome.

AA-005: ATTACK RATE OF SUBCLINICAL POST-STREPTOCOCCAL GLOMERULONEPHRITIS IN SIBLINGS

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Introduction: To estimate the incidence of subclinical disease, thirty five (35) families (siblings) of index cases with clinical post-streptococcal glomerulonephritis were screened for the presence of subclinical disease.

Method: One hundred and five siblings were investigated 1-7 days after the admission of the index cases. The diagnosis of subclinical disease was based on the presence of abnormal urine analysis, hypocomplementaemia and increased antistreptolysin O titre.

Result: Abnormal urine analysis were found in 20.95%(total 22) of sibling contacts. The incidence of nephritis among 105 siblings was 12.38%(13 siblings). This was clinical in 5(4.76%) and subclinical in 9(8.57%) cases and the calculated ratio of subclinical \clinical disease was 1.8.There were 5 siblings (4.76%) whose urine analysis could not be explained by appropriate test.

Conclusion: Sibling contacts have risk for the development of clinical \subclinical post-streptococcal glomerulonephritis.These contacts with unexplained urinary abnormalities might have subclinical nephritis in evolution.

AA-017: TWO CASES OF IgG-ASSOCIATED MESANGIAL GLOMERULONEPHRITIS IN CHILDREN

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Introduction: Rare cases of IgG –associated mesangial glomerulonephritis(IgG GN) defined by exclusive or predominant mesangial IgG deposits were reported first by Sato et al. (1993). and subsequently 10 pediatric cases were reported by Yoshikawa et al. (1994). Previous reports suggested that the prognosis of IgG GN is relatively benign but recent report suggested that prognosis of IgG GN is highly variable. Also the recurrence of IgG GN in a renal transplant was reported by Fakhouri et al. (2002). Such a recurrence highlights the specificity of this type of glomerulonephritis. We experienced two pediatric cases of IgG GN proven by renal biopsy with different clinical manifestations.

Method: Case 1 (Nephrotic syndrome) A 4-year-old girl with nephrotic syndrome admitted because of general edema. The patient`s urinalysis showed proteinuria and microscopic hematuria. Renal biopsy was performed because of relapsed nephrotic syndrome. Light microscopic finding was nonspecific with almost normal histology. Immunofluorescent findings showed diffuse segmental IgG(+) and IgM(+) deposits in the capillary walls, and focal segmental spotty C4

(trace), C1q(trace) deposits. Electron microscopic findings showed focal portion of mesangial electron dense deposits without mesangial widening. After diagnosed as IgG GN, ACE inhibitor, steroid and cyclosporin were admitted with a stable clinical course more than one and half years. Case 2 (Asymptomatic microhematuria) An 11-year-old girl admitted for evaluation of microsopic hematuria detected through mass school urinary screening program. Renal biopsy was performed for exact diagnosis. Immunofluorescent findings showed focal segmental IgG(+), IgM(+/-) and C3(+/-) deposits. Electron microscopic findings showed focal portion of mesangial electron dense deposits without mesangial widening. She is doing well without any clinical symptom with normal blood chemistry but persistent microhematuria for last two years.

Conclusion: We report two pediatric cases of IgG-associated mesangial glomerulonephritis proven by renal biopsy with distinct different clinical manifestations.

AA-021: CORRELATION BETWEEN ANGIOTENSIN CONVERTING ENZYME GENE INSERTION/DELETION POLYMORPHISM AND ACUTE POSTSTREPTOCOCCAL GLOMERULONEPHRITIS IN CHILDREN

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Introduction: This paper presents a study on the correlation between angiotensin converting enzyme (ACE) gene insertion/deletion (I/D) polymorphism and the susceptibility and clinical characteristics of acute poststreptococcal glomerulonephritis (APSGN) in children.

Method: 58 APSGN child patients were collected as case group and 100 normal children were adopted as control group, who are all Han nationality. Salting-out method was used to extract the genomic DNA. Polymerase chain reaction (PCR) technique was performed to identify the ACE (I/D) gene polymorphism in all subjects. Renal function, complement component 3 (C3), the serum ACE, 24 hours urine total protein were measured and X-ray film of chest and echocardiography were examined in APSGN group. All statistical analysis were performed using the statistical package for social science 11.5 (SPSS11.5). $P<0.05$ was considered statistically significant.

Result: The frequencies of the DD genotype and D-allele in the case group were higher than the control group (41.4% vs. 22.0%, $P<0.05$; 54.3% vs. 43.0%, $P<0.05$, respectively). In the case group with albuminuria, the frequencies of D-allele and the DD+DI genotype were higher than that without albuminuria (62.5% vs. 42.9%, $P=0.047$; 81.8% vs. 50.0%, $P=0.018$). There were no relationship between ACE genotypes and sex, age at onset of APSGN, hematuria, hypertension, C3 levels and complications.

Conclusion: ACE-D allele is correlated with the inheritance susceptibility of APSGN and with the occurrence of albuminuria, which implies a worse renal prognosis. It is suggested to further study the pathogenesis of APSGN in the context of ACE-D for early treatment and better prognosis as well as prevention and alleviation of complications.

AA-034: B7-1 AND TOLL-LIKE RECEPTOR-4 (TLR-4) SIGNALING IN AN IL-13 OVEREXPRESSION RAT MODEL OF MINIMAL CHANGE-LIKE NEPHROPATHY

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Introduction: Our previous studies have shown that over-expression of IL-13 gene resulted in podocyte injury with downregulation of nephrin, podocin and dystroglycan and concurrent upregulation of B7-1 in the glomeruli, inducing minimal change-like nephropathy (MCN). This could either be due to a direct effect of IL-13 signaling on the podocyte, or a consequence of IL-13-induced B7-1 danger signaling function. This study examined the relationship between upregulation of B7-1 expression and TLR-4 gene expression in the IL-13 overexpression model of MCN.

Method: Recombinant rat IL-13 gene was inserted into a mammalian expression vector, pCI, and transfected into Wistar rat quadriceps by in-vivo electroporation weekly till sacrifice at day 72. Glomerular gene expression of nephrin, podocin, dystroglycan, IL-13 receptor subunits, B7-1 and TLR-4 were examined using real-time PCR and expressed as an index against β-actin. Glomerular B7-1 and TLR-4 protein expression were also examined by immunofluorescent staining.

Result: While glomerular gene expression was significantly upregulated for IL-4Ra, and IL-13Ra2, and downregulated for nephrin, podocin and dystroglycan, B7-1 gene expression index (mean±SEM) was also upregulated in the IL-13 transfected rats (0.011 ± 0.001) compared to controls (0.005 ± 0.001) ($p<0.05$). The nephrotic rats showed strong patchy glomerular staining for B7-1 on immunofluorescence. Concurrently, glomerular gene expression of TLR-4 was significantly higher in the IL-13 transfected rats (0.0011 ± 0.00004) than in controls (0.0008 ± 0.00005) ($p<0.001$). This was further confirmed with immunofluorescence which showed strong glomerular staining for TLR-4. B7-1 gene expression in IL-13 transfected rats correlated significantly with serum IL-13 levels ($r=0.57$, $p<0.01$), as well as TLR-4 ($r=0.50$, $p=0.001$).

Conclusion: The strong expression of B7-1 associated with TLR-4 in the glomeruli of nephrotic rats suggests that this could be one of the signaling process by which IL-13-induced podoycte injury occurs. Further studies will be performed to explore the potential relationship between IL-13, B7-1 and TLR-4 using siRNA.

AA-037: STUDY ON PATHOLOGIES AND CLINIC OF 73 CASES OF ASYMPTOMATIC HEMATURIA IN CHILDREN

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Introduction: Hematuria is one of the most common symptoms in glomerular disease in children. Nephrologist and urologists are frequently faced with patients with asymptomatic haematuria that remains unexplained after cystoscopy and renal imaging. We performed renal biopsy for 73 cases who were diagnosed asymptomatic hematuria in our admission patients from January 1997 to March

2008 and wanted to analyze the kidney histological categories of asymptomatic hematuria in children.

Method: 73 cases were diagnosed asymptomatic hematuria in our study. All 73 cases were performed by renal biopsy.All renal tissues were examined by light microscopy, electron microscopy and immunofluorescence.

Result: All the 73 patients were identified glomerular diseases by renal biopsy. In our study the most common histological categories of asymptomatic hematuria were as follows:24 cases of IgA nephropathy,19 cases of MsPGN (mesangial proliferative glomerulonephritis), 8 cases of EcPGN (endocapillary proliferative glomerulonephritis), 5 cases of IgM nephropathy(6.8%),5 cases of focal segmental glomerulosclerosis(FSGS)(6.8%),5 cases of membranous nephropathy(MN) (6.8%),4 cases of membranoproliferative glomerulonephritis(MPGN) (6.0%),2 case of minimal change disease(MCD) (2.7%),1 case of thin basement membrane nephropathy(TBMN) (1.4%).

Conclusion: In our study IgA nephropathy was the most common pathological category in asymptomatic hematuria, the second is MsPGN, the third is EcPGN.

AA-039: SUCCESSFUL TREATMENT OF A PATIENT WITH LIPOPROTEIN GLOMERULOPATHY

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Introduction: Lipoprotein glomerulopathy (LPG) is a rare disease demonstrating considerable thrombi material consisting of lipoprotein in the glomerular capillary lumen. Mutation of apoprotein E (APOE) was identified as a cause of LPG, and these patients have been considered to show poor prognosis leading to chronic renal failure even after renal transplantation. We present the successful treatment of a 14-year-old Japanese girl with an 8-year history of LPG.

Result: Case: Proteinuria was incidentally detected when this patient was 3 years old in 1998. To characterize her nephrotic syndrome, she underwent renal biopsy at age 4. The pathological specimen showed dilated capillary lumina with massive lipoprotein thrombi in every glomerulus. Thereafter she was referred to our hospital and followed from June 1999. Based on pathologic findings and genetic analysis, we diagnosed her as having LPG. Her father and younger sister also showed the same mutation of apo E, ?APOE Sendai?, but were not affected by LPG. Initially, we administered probucol, enalapril, and dipyridamole. Since the response was not satisfactory, we changed probucol to bezafibrate and atorvastatin calcium hydrate in March 2003, and added valsartan in December 2004. Since then, her clinical and laboratory findings showed the following improvements: serum albumin increased from 2.6 to 4.1 g/dl, serum APOE decreased from 11.0 to 7.1 mg/dl, and urinary protein disappeared. However, creatinine clearance deteriorated from 160.8 to 69.2 ml/min/1.73 m2 over 9 years, but she has remained above 60 ml/min/1.73 m2, which is within normal limit. Under informed consent, we performed re-biopsy for pathological re-evaluation in April 2007, and confirmed that nearly all intraglomerular lipoprotein thrombi had vanished.

Conclusion: LPG has been recognized as a refractory and progressive disease, but we have successfully treated this patient with antilipidemic drugs, antiplatelet drug, and angiotensin II receptor blocker.

AA-041: THE OCULAR COMPLICATION IN CHILDREN WITH NEPHROTIC SYNDROME RECEIVING STEROID THERAPY

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Introduction: The present study aimed to investigate ocular complication arising from nephrotic syndrome and its treatment in children.

Method: Retrospective data were collected from the medical records of 62 patients with nephrotic syndrome who were treated at the East-West Kidney Disease Research Institute of Kyung Hee University from January 2000 to December 2007. The treatment consisted of intravenous methylprednisolone 20 mg/kg/day administration for 3 consecutive days and followed by oral MPD 0.8 mg/kg/day for 2 weeks. This was taken as one course and was carried out until remission. Among the 62 patients, 13 patients had ocular complications. We compared the age, sex, duration of treatment, time to attaining remission and initial laboratory findings between the two groups.

Result: Among the 13 patients, cataract was found in 6 patients (4.5%) and glaucoma in 7 patients. (5.2%) The mean duration of treatment was 24.83±18.5 months in the case group and 20.52±11.56 months in the control group. There was no statistical difference (p-value 0.61) between the two groups. In correlation, there was no statistically significant difference among the duration of treatment, laboratory profile extracted at the time of the first hospital admission and occurrence of ocular complications. On the other hand, it took 235±299 days to the remission in the case group and 89±175 days in the control group and it was statistically different (p-value 0.03).

Conclusion: In consequence, the occurrence of ocular complications in children who had nephrotic syndrome was related with the time to remission, and the total dose of corticosteroid was in proportion to the time to remission. In other words, the total dose of administered corticosteroid is key factor in the occurrence of ocular complications. Therefore, pediatricians should be aware of the potential risk of developing ocular complications and its early detection.

AA-046: CASE REPORT: EBV-CAUSED MEMBRANOUS NEPHROPATHY IN CHILD

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Introduction: EBV is a common pathogen of infectious diseases in pediatrics that is able to induce multi-organ impairment, especially that in kidney.

Method: The EBV-infected patient's clinic history in our department was analyzed, the clinic characters, diagnosis and treatment were reviewed being companied with the review of pertinent literature.

Result: The patient was a 4 years and one month old boy. After infected with EBV, the swollen eyes and the hematuria under microscope (5-8 RBC /HP) appeared. The urinary protein was (+), renal function was disorder and blood urea nitrogen increased significantly (8.36 mmol/L). The histologic diagnosis by the renal biopsy was membranous nephropathy. He was treated by Ganciclovir. Prednisone, dipyridamole, piperazineFerulateTablets and ACEI were also given. The clinical symptoms disappeared in two years. The

number of RBC in urine was 0-1/HP, the proteinuria was negative, the renal function and BUN were normal.

Conclusion: A patient with edema, hematuria and proteinuria should be paid attention by pediatrician to avoid a missed-diagnosis or a misdiagnosis. A renal biopsy should be performed as soon as possible to identify the type of nephropathy. The proper treatment is also necessary.

AA-047: A PROSPECTIVE MULTICENTER CLINICAL CONTROL TRIAL COMPARING MYCOPHENOLATE MOFETIL DISPERSIBLE TABLETS AND PULSE INTRAVENOUS CYCLOPHOSPHAMIDE FOR REFRACTORY NEPHROTIC SYNDROME IN CHILDREN

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Introduction: The present study was a prospective multicenter clinical control trial to evaluate the efficacy and safety of Mycophenolate Mofetil Dispersible Tablets(MMF) in addition to prednisone on refractory nephrotic syndrome.

Method: 142 patients from nine clinical trials centers were divided into two groups,where there were no significant differences between the two groups on age, gender, and clinical categories. They were treatment group(87 patients) treated with MMF and control group(55 patients) treated with pulse intravenous cyclophosphamide(IVC) in addition to prednisone. 87 patients of MMF treatment group were treated with full-dosage(30~40 mg/Kg.d) MMF for at least six months, then gradually tapered persisted till one year if with efficacy, otherwise stop MMF; 55 patients were treated with IVC(10 mg/Kg.d×2 d/2 w) for 3 months, then with IVC(500 mg/m2) at 4, 7, 10 month repectively.Both groups were combined prednisone(0.5~1 mg/Kg.d) for 2~3 months,then gradually tapered. Urine protein, liver function, renal function and drug side effect were observed regularly for one year.

Result: Of the 87 patients who received the MMF protocol,complete remisson was achieved in 58 patients,partial remission in 16 patients, early effect in 9 patients,and no improvement in 4 patients,total remission rate [complete remission(CR) and partial remission(PR) and early effect(EE)] were 95.45%,negative proteinuria rate was 77%; while 35 patients, 9 patients, 1patient,10patients, 81.8%, 65.4% respectively in CTX protocol group.There were no significant difference in negative proteinuria rate between two groups.While effective rate of MMF protocol group was significantly higher than CTX protocol group.Furthermore MMF was found more effective in reducing proteinuria,improving hypoproteinemia,oliguria,edema than CTX. MMF was better tolerated with lower adverse reaction rate, including transient elevated AST or ALT (three), infections (thirty two),gastrointestinal symptoms (eleven), menstrual disorder (one), muscle waver (one).Side effects of 55 patients who received CTX protocol included transient elevated AST or ALT (nine),infections (thirty),gastrointestinal symptoms (fifteen),hypohemoglobinemia (four),hypoleupenia (one),phalacrosis (one).

Conclusion: The results demonstrated that combined therapy of MMF (20~35 mg/Kg.d) and prednisone (0.5~1 mg/Kg.d) show similar negative proteinuria rate with CTX protocol in treatment of RNS, Furthermore the former can shorten the induced remission time,and has lower side-effect rate.

AA-057: RESPONSE TO CYCLOSPORINE A IN PAKISTANI CHILDREN WITH PRIMARY FOCAL SEGMENTAL GLOMERULOSCLEROSIS

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Introduction: Nephrotic Syndrome (NS) is the most common chronic renal disease in children. The hallmark of this disease is characterized by multiple relapses, which are usually responsive to steroids. A subgroup of children require steroids to be in remission and hence develop steroid dependence(SDNS) Infrequently less than 10% are resistant to steroid therapy(SRNS) Primary Focal Segmental glomerulosclerosis (FSGS) is the most common cause of steroid resistance. Cyclosporine A (CsA) have been found effective in FSGS. In Pakistan use of CsA is limited therefore we did this study to see the response to CsA in primary FSGS in children suffering from NS.

Method: Total of thirty cases were included in the study. Indication for starting CsA was steroid resistance (n=18) and steroid dependence (n=12) with steroid toxicity. CsA was given in a dose of 4-5 mg/kg body weight per day in 2 divided doses for a period of 12—24 months. Response to CSA seen after three months of therapy was categorized into: complete remission, partial remission or no response. Follow-up was continued for two years. Urine analysis, renal function and monitoring of CsA level was done. Any side -effects that developed were noted.

Result: Maximum number of cases were seen in 6-10 years age group 13/30 (43.33%) Male to Female ratio was 1:0.9. Complete remission was seen in 17/30 cases (56.67%) while 9/30 cases showed partial remission (30%). No response was seen in 4/30 cases (13.33%). Three out of this 4/30 cases went into chronic renal failure. Side effects noted were; gingival hyperplasia in 14/30 and hypertrichosis in 11/30 cases.

Conclusion: CSA seems to be effective in the treatment of FSGS in at least 50-60% of the cases in Pakistan.

AA-060: EFFECTS OF PUROMYCIN AMINONUCLEOSIDE ON THE GLOMERULAR EPITHELIAL CELLS

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Introduction: Puromycin aminonucleoside (PAN) nephrosis is a well-known experimental model for human nephrotic syndrome. We wish to clarify the mechanism of proteinuria in nephrotic syndrome patients by using in vitro PAN nephrosis model.

Method: Following the administration of various concentrations of PAN and antioxidants we observed the changes of podocyte cytoskeletons in cultured rat glomerular epithelial cells (GEpC) by scanning electron microscope, reactive oxyten species (ROS) analysis, permeability assay, confocal microscope, and Western blot assay.

Result: PAN not only induced the ultrastructural changes of GEpC, such as shortening and fusion of microvilli, but also separated the intercellular gaps and linear ZO-1. PAN induced oxidative stresses in

time- and dose-dependent manners and increases of intercellular permeability which inhibited by anti-oxidants. High concentration of PAN induced not only actin polymerization and disorganization, but also the conglomerulation and internal dislocation of α-actinin protein. The intensities of fluorescences of ZO-1 protein were diminished and internalized by PAN in a dose-dependent manner, which were also prevented by anti anti-oxidants.

Conclusion: PAN induced the changes of podocytes cytoskeleton and hyperpermeability with the changes of junctional protein ZO-1 by oxidative stresses in GEpC. Glomerular hyperpermeability induced by PAN through oxidative stresses is thought to be the mechanism of proteinuria in experimental nephrotic syndrome.

AA-067: EVALUATION OF THE EFFECT WITH LIGHT MICROSCOPE AND URINE FLOW CYTOMETER TO DETECT THE SOURCE OF HEMATURIA IN CHILDREN

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Introduction: Since the etiology of hematuria can be quite complex, applying a more effective and accurate method to early orientate the diagnosis towards a glomerular or a non-glomerular disease is very important. This study aims at evaluating the effect of the light microscopy(LM) and the automated urine flow cytometer(UFCM) in detecting the source of hematuria in children.

Method: (1) 125 children who had been diagnosed as having glomerular diseases underwent renal biopsy. (2) The morning urine specimen from all 125 patients with glomerular diseases as well as 46 patients with non-glomerular were collected for erythrocyte morphological examination by LM. In addition, the samples from 47 patients with glomerular diseases and 36 patients with non-glomerular diseases were examined by using UFCM. The sensitivity and specificity for the diagnosis of glomerular hematuria according to different criterions by LM were tested. In this study, the positive rates of glomerular hematuria based on different urine specimen were also contrasted, which included as following: ① morning urine and random urine specimen from 32 cases; ② urine specimen containing a great deal proteinuria or without proteinuria; ③ urine samples containing different number of RBCs.

Result: (1) This group of 125 children with clinical diagnosis of glomerular diseases was confirmed by renal biopsy. (2) The diagnostic criteria for glomerular hematuria were those with severe dysmorphic RBC≥30% or dysmorphic RBC≥70%.The sensitivity and specificity of diagnosing glomerular hematuria via LM was 92.8% and 97.8%, respectively. Whereas the corresponding rate from UFCM was 58.5% and 93.8%. (3) The positive rate of glomerular hematuria with the two methods in testing the same day's morning urine and random urine samples from the 32 cases with glomerular diseases was respectively 78% VS. 71.9%(LM) and 65.6% VS. 62.5%(UFCM). P>0.05. (4) By employing LM on the urine specimens from 22 patients with massive proteinuria and from 67 patients with isolated hematuria, the sensitivity reached respectively 81.8% VS. 91.4% (P>0.05). (5) The positive proportion of glomerular hematuria in RBC <100/ul group was lower than that of RBC 100-2000/ul group with LM, while no such differentiation was found in RBC<100/μl group when using UFCM.

Conclusion: (1) This series of data indicated that if we use the diagnostic criteria for glomerular hematuria that dysmorphic RBC≥70% or severe dysmorphic RBC≥30%, the sensitivity and specificity are both higher. (2) Both specificity reached over 90%, but the sensitivity of the UFCM was lower. (3) The random urine specimen could be accepted with the two methods in children.

AA-098: PATHOLOGICAL CHARACTERISTICS AND OUTCOMES OF ASYMPTOMATIC IgA NEPHROPATHY IN CHILDREN

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Introduction: To study the pathological characteristics and outcomes of IgA nephropathy (IgAN) presented with asymptomatic proteinuria and/or microscope haematuria in children.

Method: Clinical and pathological characteristics of 54 children with IgA nephropathy confirmed by renal biopsy were analyzed. 54 children with IgAN were divided into two groups according to their clinical features at the first onset: asymptomatic IgAN group (AsIgAN) and symptomatic IgAN group (SIgAN). Histologic changes were classified by Lee SM and Katafuchi semiquantitative scoring system.

Result: 18 children were in AsIgAN group and 36 children were in SIgAN group. The degree of proteinuria in SIgAN group (2.3±2.2 g/d) was higher than that in AsIgAN group (0.4±0.3 g/d) at the time of biopsy (P<0.05). Although asymptomatic IgAN children were mainly of Lee's type I~II, there were 11% with Lee's grading IV~V and 27% of children with interstitial injury. Symptomatic IgAN children were mainly of Lee's type II~III, there was no significant difference between two groups(P>0.05).Urine microablumin increased in 87% children presented with microscope haematuria. After an average of (26.9±8.8)months follow-up, only one case of Lee's V progressed into renal failure and the others maintained normal renal function.

Conclusion: Although children with Asymptomatic IgAN have tiny clinical symptoms, severe renal pathological lesion and poor outcomes also occurred. Our results suggest that urine screening help to detect renal diseases and renal biopsy should be administered in the patients with increasing urine microalbumin.

AA-127: LONG VERSUS STANDARD INITIAL PREDNISOLONE THERAPY IN CHILDREN WITH IDIOPATHIC NEPHROTIC SYNDROME

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Introduction: Although it is well accepted that corticosteroid induce a complete remission in most children with new onset idiopathic nephrotic syndrome,the optimal duration of initial steroid treatment for children with this disorder is unclear.To compare the efficacy of longer initial course of prednisolone with standard regimen in patients with idiopathic nephrotic syndrome (INS) in Children.

Method: randomized controlled trial.Patients with INS who responded to initial course of prednisolone, either standard or long regimen were included and followed for one year. The standard regimen consisted of

prednisolone 60 mg/m2/day for four weeks followed by 40 mg/m2 every alternate day single morning dose for further four weeks. The long regimen consisted of prednisolone 60 mg/m2/day for six weeks followed by 40 mg/m2 every alternate day single morning dose for further six weeks.There were 93 children who fulfilled the criteria of the study, 47 from long group and 46 from standard group. The two groups did not differ in age, blood pressure, serum albumin, serum cholesterol or serum Creatinine prior to the initial prednisolone therapy

Result: Cumulative prednisolone dose was significantly higher in the long regimen group than the standard group ($p=0.001$). Relapse within one year was noted in 73.2% of long group and 58.1% of standard group. The odd ratio for relapse within one year was 0.51 (95% confidence interval 0.17, 1.53). This did not reach the statistical significance ($p=0.634$). The side effects of prednisolone between two groups were also not statistically significant ($p>0.05$).

Conclusion: Our data suggest that prolongation of prednisolone therapy for initial episode of steroid-sensitive idiopathic nephrotic syndrome does not have a beneficial effect on outcome in next one year.

AA-149: THROMBOTIC COMPLICATIONS IN NEPHROTIC SYNDROME (NS)

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Introduction: Abnormalities of coagulation are known in NS, but thrombosis of major vessels is rare. We report 4 cases with such complications, seen over a 10-year period at a tertiary pediatric renal unit.

Method: Detailed laboratory evaluation and imaging were done. Anticoagulant therapy was used as indicated

Result: Case 1. 1.5 year-old-girl with steroid responsive NS (SRNS) was in remission with prednisolone (PRED) 1.5 mg/kg on alternate days (AD), when she had vomiting, generalized tonic–clonic seizures and right hemiparesis. MRI of brain showed thrombosis of sagittal and left transverse sinuses. She was managed with supportive care and gradually made a complete recovery. Case 2. 7 year-old-boy had SRNS with frequent relapses since age 1.5 years. While in remission with 20 mg PRED on AD, he developed coldness of left leg and bluish discoloration of toes. Doppler study showed narrowing of anterior and posterior tibial arteries in middle and distal parts. Anticoagulant therapy lead to gradual, complete recovery without tissue loss. Case 3. 5 year-old-boy with steroid dependent NS was in remission with PRED 20 mg on AD. He acutely developed severe headache and vomiting with papilledema. CT scan was normal but MRI revealed sagittal sinus thrombosis. He fully recovered with supportive care. Case 4. 10 year-old-boy had steroid resistant NS with membranoproliferative glomerulonephritis. Treatment with enalapril and PRED on AD lead to partial response (absence of edema, increase in serum albumin, mild proteinuria). He developed increasing swelling of the right upper extremity, without pain and fever. Doppler studies showed thrombosis of right jugular and subclavian veins. He was treated with anticoagulants and made a complete recovery. None of the patients had a preceding acute illness or taken diuretics. Renal function parameters were normal.

Conclusion: Thrombosis of major vessels, mostly venous but occasionally arterial, may rarely occur in NS without any precipitating illness (eg hypovolemia, sepsis). Early detection and prompt treatment are crucial.

AA-153: IS HYPERHOMOCYSTEINEMIA AN INDEPENDENT RISK FACTOR IN NEPHROTIC SYNDROME PATIENTS OF PAEDIATRIC AGE GROUP

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Introduction: Homocysteine is a non-protein forming amino acid, whose metabolism is dependent on the vitamins (folic acid, B12 and B6), and the enzymes (methylenetetrahydrofolate reductase and cystathionine-synthetase). Hyperhomocysteinemia is an independent risk factor for cardiovascular complications & atherothrombosis in several clinical settings in which renal function is impaired, but its prevalence in nephrotic syndrome has not been investigated in detail eventhough this syndrome provides an excellent model to study a possible link between albuminuria, proteinuria & hyperhomocysteinemia.

Method: The study was conducted on 30 patients of Nephrotic syndrome of pediatric age group admitted in Nephrology Unit, Department of Medicine, Pt.J.N.M. Medical College & GBG Kidney care Hospital, Raipur (C.G.). All patients were subjected to serum Homocysteine levels and all other routine investigations.

Result: •Mean age of the patients was 11.46±4.64 years. •Males were 73.33% & females were 26.67%. •Mean value of serum Homocysteine was 15.69±1.92 mmol/dl. •Among the patients with hyperhomocysteinemia 72.72% were males and 27.28% were females. •Hypercholesterolemia was present in 53.33% of patients of which 81.25% of the patients had hyperhomocysteinemia. •80% of the total patients had Hypoalbuminemia (S.albumin<3.5 gm/dl), of which 79% had hyperhomocysteinemia. •Hypothyroidism was present in 40% of patients, Out of them 75% had hyperhomocysteinemia. •Mean value of homocystein was more among the patients with thromboembolism (18.69± 0.82 mmol/dl) as compared to non-thromboembolic patients (15.69± 1.92 mmol/dl).

Conclusion: •Majority of patients with Hyperhomocysteinemia were males. •Hyperhomocysteinemia was more prevalent in 6-12 years of age group. •Hyperlipidemia was associated with higher levels of Homocysteine. •A significantly higher proportion (79%) of subjects with hypoalbuminemia (Serum albumin < 3.5 gm/dl) had raised Homocysteine levels. •75% of the Children with Hypothyroidism had Hyperhomocysteinemia. •Mean value of Homocysteine was more among the patients with vascular complications.

AA-154: STUDY OF ANTIOXIDANT STATUS AMONG THE PATIENTS OF NEPHROTIC SYNDROME OF PEDIATRIC AGE GROUP FROM TRIBAL AREA OF DEVELOPING COUNTRY-INDIA

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Introduction: The children with nephrotic syndrome are frequently associated with decrements in antioxidant levels which can be

measured by serum copper, zinc, vitamin C, apolipoprotein A, and total antioxidant status.

Method: The study was conducted on 35 patients of nephrotic syndrome admitted in Nephrology Unit, Pt.J.N.M. Medical College & GBG Kidney care hospital, Raipur from April 2007 to April 2008. All patients were subjected to serum copper, serum zinc, vitamin C, apolipoprotein, Total anti oxidant status {estimation of Superoxide dismutase, Glutathione peroxidase & Catalase}, fasting lipid profile, thyroid profile & all routine investigations.

Result: •Males were 74.29% & females were 25.71%. •Mean age was 11.46±4.64 years. •Levels of all the antioxidants were markedly reduced in age group 6-12 years. •Mean value of serum copper, Zinc, vitamin C, Homocysteine, apo A and total anti oxidant status was 59.64±1.01 µg/dl, 66.59±5.61 µg/dl, 0.189±0.039 mg/dl, 15.69±1.92 mmol/dl, 182.81± 4.75 mg/dl, and 0.536±0.071 mmol/l. •Dyslipidemia was present in 63.16% males and 37.84% females. •Antioxidant levels were significantly reduced among most of the patients with dyslipidemia. •Serum albumin < 2 gm/dl was found in 51.43% of children and most of them had reduced antioxidant level. •Reduced antioxidant status was present in 74% males and 26% females. •Dyslipidemia was present in 54.29% of patients. •51.43% of the children had nephrotic syndrome relapse and antioxidant status was significantly reduced in them.

Conclusion: •Reduced antioxidant was more in males as compared to females. •Children between 6-12 years of age had markedly reduced antioxidant levels. •Dyslipidemia was more common in males than females. •Dyslipidemia and Low levels of serum albumin were associated with markedly reduced antioxidant levels. •All the patients experiencing relapse of nephrotic syndrome had markedly reduced antioxidant levels.

AA-155: FAMILIAL NEPHROTIC SYNDROME – A RARE CASE REPORT

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Introduction:

Method: A 6 year old girl admitted with complaints of facial puffiness, pedal oedema, gradual abdominal distension, decreased urine output, off & on since last 3 month. She was taking treatment irregularly in suburb. Patient has significant family history, had 9 siblings, Out of which 3 female children suffering from nephrotic syndrome died at age of 8-10 years due to nephrotic syndrome & its complications. On investigating 2 sisters showed nephrotic range proteinuria (24 hr urinary proteins 3.2 gm/24 hr& 3.6 gm/24 hr), deranged thyroid profile and Hyperlipidemia. Both sisters were treated on the line of nephrotic syndrome with Deflazacort, Torasemide, Losartan, Ranitidine, Atorvastatin. Patient's step mother died after 6 month of diagnosis of nephrotic syndrome. Investigation of the patient's mother also revealed nephrotic range proteinuria, Hypothyroidism & Hyperlipidemia. Male siblings were asymptomatic and their investigations were within normal limit. On general examination, her weight was18 kg (>100 percentile), height-110 cm, pulse-100/min, BP-110/70 mm of Hg in lying down position, facial puffiness, mild pedal oedema. Per abdominal examination showed abdominal distension, stretched umbilicus, shifting dullness. Other systemic examinations are within normal limit. On investigating her Hb-11.5 gm%,TLC-8900/ mm3,ESR-35 mm/hr,Blood sugar-96 mg%, Urea-96 mg%, creatinine-1.5 mg%,serum protein-4.5 gm%,s.albumin-2.4 gm%,s.globulin-2.1 gm%, 24 hr urinary proteins-4.5 gm/24 hr, S. cholesterol-355 mg/dl,S.triglyceride-155 mg/dl, S.LDL-276 mg/dl, S. HDL-45 mg/dl, S.VLDL-34 mg/dl, Urinalysis showed – Albumin +++ , pus cell 3–4/hpf. Ultrasound abdomen showed– bilateral medical renal disease with moderate ascitis with bilateral pleural effusion. Renal biopsy was done which showed minimal change disease.She was treated with Deflazacort, Torasemide, Losartan, Ranitidine, Atorvastatin. Patient showed improvement & was discharged with urinary albumin trace & weight reduced from 18 kg to13.5 kg.

Result:

Conclusion: Numerous inflammatory & non inflammatory diseases affects the glomerules & lead to alteration in glomerular permeability, structure,& function. A genetic basis has been identified in number of renal diseases. This is a case of Familial nephrotic syndrome, a genetic disorder with rare presentation seen in paedriatic age group.

AA-164: THE INTEGRITY OF SLIT DIAPHRAGM REGULATED BY TYROSINE PHOSPHORYLATION - THE ROLE OF PHOSPHORYLATION OF TRPC6

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Introduction: Recent investigations have identified several indispensable components of Slit Diaphragm (SD) such as Nephrin, Neph1 and Podocin. This complex is now known to serve as signaling platform through their phosphorylations, as well as filtering barrier structure. TRPC6 was also identified as an SD-component, and its activity is regulated by diacylglycerol, a product of phospholipase C (PLC). Some of the TRPC6 mutations enhance its channel activity, and elevated level of TRPC6 is also observed in acquired nephrotic syndromes. TRPC6 activity can also be regulated by phosphorylation by a tyrosine kinase, Fyn. We hypothesized the role of TRPC6 phosphorylation in the interaction with SD-components.

Method: Phosphorylation of TRPC6 was examined by stimulation such as EGF or by coexpression with Fyn. The interaction between TRPC6 and SD components was investigated by co-immunoprecipitation using transfected HEK293T cells and GST-pull-down analysis.

Result: Tyrosine phosphorylation of TRPC6 was observed upon EGF-stimulation or coexpression with Fyn. Among several SD-proteins, Nephrin was found to bind to TRPC6 in a phosphorylation-dependent manner. Pull-down analysis using GST-Nephrin cytoplasmic domain revealed that phosphorylated TRPC6 but not non-phosphorylated TRPC6 bound to Nephrin. This interaction was observed irrespective of Nephrin phosphorylation, indicating that phosphorylation of TRPC6 but not Nephrin is crucial for this interaction. Phosphorylated TRPC6 also bound to PLCgamma and N-terminal portion of TRPC6 was responsible for this interaction. By mutational analyses using a series of single phenylalanine substitutants of TRPC6, we identified distinct tyrosine residues crucial for the interactions with Nephrin or with PLCgamma.

Conclusion: Tyrosine phosphorylation of TRPC6 regulates its association with Nephrin and PLCgamma. Phosphorylation-dependent

trimeric complex of TRPC6/Nephrin/PLCgamma may regulate the integrity and function of SD.

AA-167: URINARY LEVEL OF TRANSFORMING GROWTH FACTOR B-1 IN DIFFERENT PROTEINURIA STAGES IN CHILDREN WITH IDIOPATHIC NEPHROTIC SYNDROME

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Introduction: Proteinuria is the basic problem in nephrotic syndrome which leads to raise morbidity and mortality in such patients. Massive proteinuria in nephrotic syndrome is likely to cause major complications, i.e. acute renal failure, severe infection as peritonitis, anemia, growth disturbance, thrombosis due to hypercoagulability state, and hypovolaemic shock. Although, the majority of children with classic idiopathic nephrotic syndrome respond to steroid treatment, and called steroid sensitive nephrotic syndrome, but more than half children develop frequently relapsing or frequently relapsing with steroid dependency and some get steroid resistant. Urinary TGF-β1 excretion correlates with the amount of filtrated protein and duration of exposure protein to tubular cells. TGF-β1 is a pluripotent fibrogenic cytokine which through some pathomechanisms causes interstitial fibrosis and glomerulosclerosis, ending with nephron loss.

Method: This study was a cross sectional study to compare urinary TGF-β1 and protein levels in children with nephrotic syndrome remission, relapse, and steroid resistant to its level in children without kidney disease. Urinary protein level was measured as urinary protein and creatinine ratio in spot urine sample taken in the morning. Urinary TGF-β1 level was determined using Elysa (Quantikine kit for human TGF-β1 immuno assay)

Result: A hundred twenty children with idiopathic nephrotic syndrome were enrolled in this study, consisted of 34 children with steroid sensitive nephrotic syndrome remission, 31 children with steroid sensitive nephrotic syndrome relapse, and 55 children with steroid resistant nephrotic syndrome. Thirty-five children without kidney disease were recruited as the control group. The highest urinary protein level was found in children with nephrotic syndrome relapse, significantly higher than the level in steroid resistant nephrotic syndrome. But, urinary TGF-β1 level in children with nephrotic syndrome relapse was as high as its level in children with steroid resistant nephrotic syndrome, significantly much higher compared to the level in nephrotic syndrome remission and those in children without kidney disease. There was a moderate positive correlation between urinary TGF-β1 and protein excretion.

Conclusion: Urinary TGF-β1 level is high in children with sensitive steroid nephrotic syndrome relapse and steroid resistant nephrotic syndrome, significantly higher compared to the level in children with steroid sensitive nephrotic syndrome remission and those in children without kidney disease

AA-168: RENAL BIOPSY FINDINGS IN CHILDREN WITH GLOMERULONEPHRITIS

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Introduction: The glomerular diseases pose a common diagnostic and therapeutic challenge in pediatric nephrology practice. Percutaneous renal biopsy (PRB) is commonly performed to obtain renal tissue for histological diagnosis, decide about prognosis and monitoring of response to treatment. The most common histological lesions in primary nephrotic syndrome are minimal change disease (MCD), focal segmental glomerulosclerosis (FSGS), membranoproliferative or mesangio proliferative glomerulonephritis (MPGN/MesPGN) and membranous GN. There are specific histopathological findings in secondary GN like lupus /HSP nephritis and in different forms of congenital nephrotic syndrome (CNS). The objective of this study was to determine the indications, types of histological lesions and complications of renal biopsy in children with glomerulonephritis

Method: This is a hospital based prospective study of 75 children of 1 mo to 15 yrs age who underwent renal biopsy from June 2005 to April 2008 at National Institute of Child Health and The Kidney Centre, Karachi. Ultra sound guided automated Monopty Bard gun technique was used in all children. Coagulation profile and renal function assessment and complete blood count were done prior to biopsy. B.P was controlled if high and acute infection was treated before biopsy. Children were kept NPO 4-6hrs before and after biopsy. Patients were monitored clinically during and 24 hrs after the procedure. Children received short-acting anesthesia and few of them underwent general anesthesia. All samples were taken from the left kidney and studied under light microscopy and immuno-fluorescent (IF) stain. Children with persistent gross hematuria after 24 hr had a post-biopsy ultrasonography.

Result: A total of 75 children underwent PRB during the study period. There were 39 (52%) boys and 36(48%) were girls. Age group ranged from 0.12 to 15 years (mean age 7.43 yrs). Majority of children (45/60%) were of 5-15 years age group. The most common indication for PRB was primary nephrotic syndrome (46/61.33%) followed by secondary GN (10/13.33%), congenital NS (9/12%) and nephritic nephrotic syndrome (8/10.66%). Among the primary NS(46), steroid-resistant nephrotic syndrome were 27(36%), steroid dependent nephrotic syndrome in 14 (18.66%) and there was partial response in 5(6.66). Mean number of passes was 2.3(2–4). PRB was successful in 71 patients (96.08%) and failed in 4(5.3%) cases. It was cortico-medullary tissue in 41 and cortical in 27 samples. Mean number of glomeruli was 24.8(2–88). Among the 71 diagnostic samples, the most common histology was FSGS (25/35.21%), followed by MCD (20/26.66%%), MPGN/Mes PGN (11/15.49%) and membranous nephropathy (6/8.45%). Diffuse mesangio proliferative (post infective) and HUS/acute cortical necrosis was seen in 3 cases. Finish type of CNS in 2 and IgM nephropathy in one. Immuno-flourescent stain was positive in 33 out of 67 samples (49.25%)stained. It was positive in all MPGN/Mes PGN (11), membranous (6), Lupus (2/3)/ HSP (2) and 52% of FSGS. Over all, complications were observed in 4 patients (5.33%). Transient gross hematuria (<24 hrs) was observed in 3 and self-resolving perirenal hematoma in one. No patient experienced severe pain or infection at the biopsy site.

Conclusion: Steroid resistant primary NS was the most common indication for biopsy and FSGS was the most common histological lesion followed by MCD and MPGN. PRB under US guided was successful in >96% with minor complication of gross hematuria.

AA-171: COMPARISON OF THERAPEUTIC EFFECT OF LEVAMISOLE AND CYCLOPHOSPHAMIDE IN FREQUENTLY RELAPSING NEPHROTIC SYNDROME.

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Introduction: Although levamisole or cyclophosphamide have been used widely for long time to reduce relapse rate and dose of steroid in nephrotic syndrome, there was no study comparing the effects of two drugs.

Method: With levamisole or cyclophosphamide, authors had treated 49 patients of frequent relapsing nephrotic children who had visited BNU Hospital from Jan., 2001 until July, 2007. We compared therapeutic effects of levamisole (L-group, 23 patients) and cyclophosphamide(C-group, 26 patients) by relapse rates and cumulative dose of steroid before and after trial of these drugs.

Result: Pathological type: In L-group, eleven patients had renal biopsy in which ten patients showed minimal change and one patient showed FSGS. In C-group group, seventeen patients had renal biopsy in which eleven patients showed minimal change, five FSGS, one IgA nephropathy. Reduction in relapse rate: In L-group, relapse rate decreased from 4.28±2.16 relapse/patient/year before levamisole trial to 2.34±3.07 relapse/patient/year on levamisole for one year($p=0.02$). In C-group, relapse rate decreased from 6.95±4.86 relapse/patient/year before cyclophosphamide to 4.81±5.25 relapse/patient/year for one year after cyclophosphamide($p=0.04$). Reduction in prednisolone dose: In L-group, the cumulative dose of prednisolone decreased from 704.76±296.42 mg/m2/month to 490.63±204.44 mg/m2/month($p< 0.01$). In C-group it decreased from 896.35±632.46 mg/m2/month to 688.79±369.57 mg/m2/month($p=0.20$).

Conclusion: Both levamisole and cyclophosphamide treatment effectively reduced relapse rate and cumulative dose of prednisolone in frequent relapsing nephrotic syndrome in children. For the comparison of efficiency of both drugs, more studies including many of patients are needed in the future.

AA-174: BONE MINERAL DENSITY IN CHILDHOOD NEPHROTIC SYNDROME WITH NORMAL RENAL FUNCTION

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Introduction: Children with nephrotic syndrome may be at risk for metabolic bone disease because of alterations in bone and mineral metabolism and steroid-induced osteoporosis. This study aims to determine the occurrence of metabolic bone disease in children with nephrotic syndrome with normal renal function.

Method: Design: Cross Sectional Study Setting: Pediatric Nephrology Clinic at University of Santo Tomas Hospital Participants: Children aged 5-18 years with nephrotic syndrome with normal renal function, at least on 6 months steroid therapy Intervention: (a) Clinical and biochemical tests to determine risk factors for low bone mineral density (BMD) (b) Dual-energy x-linked absorptiometry (DXA) of the lumbar spine (L1 to L4) and whole body Main Outcome Measures: Bone mineral density Z-scores and risk factors that would correlate with BMD

Result: A total of 15 patients were included in this study (8 boys, 7 girls). Mean age at diagnosis was 6.7±3.9 years (1.6–12 years) while mean age at the time of DXA was 10.5±3.6 years (5.8–18 years). Only 2 patients (13.3%) have low BMD, both were males with younger age at diagnosis and at the time of DXA (3.2±0.5 years and 6.8±1.3 years, respectively) compared to those with normal BMD (7.3±4.0 years and 11.1±3.6 years, respectively). Duration of disease was the same between the 2 groups (3.8±2.7 years, normal BMD; 3.6±0.8 years, low BMD). None of those with low BMD had symptoms of hypocalcemia nor history of fracture. One patient with low BMD have regular intake of coffee (25–30 mg caffeine/day). Mean calcium intake for the 2 groups were within the recommended daily allowance (RDA) while mean vitamin D intake were below the RDA for both groups. All patients have normal serum ionized calcium except for one with low BMD who have slightly high level (1.39 mmol/L). Serum phosphorus was low in patients with low BMD while 2 patients with normal BMD have high levels. There was no difference in the mean cumulative steroid dose between the 2 groups (792.9±531.4, normal BMD; 859.4±571.1 mg/kg, low BMD). Of the factors evaluated, only serum phosphorus statistically correlate with low BMD.

Conclusion: Only 13% of the patients were found to have low BMD. Low serum phosphorus was the only factor found to significantly correlate with low BMD. Majority of our patients with nephrotic syndrome with normal renal function on ≥ 6 months of steroid therapy have normal bone mineral density.

AA-178: ATYPICAL PRESENTATION OF NEPHROTIC SYNDROME AS ASYMETRICAL PERIPHERAL GANGRENE

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Introduction: Multiple proteins of coagulation cascade have altered level in nephrotic syndrome, in addition platelet aggregation is enhanced. Only 2% of children with nephrotic syndrome have thrombosis. Thromboembolic event is most likely if serum albumin is less than 2 gm/dl.

Method: Multiple proteins of coagulation cascade have altered level in nephrotic syndrome, in addition platelet aggregation is enhanced. Only 2% of children with nephrotic syndrome have thrombosis. Thromboembolic event is most likely if serum albumin is less than 2 gm/dl. A 15 year old female patient came with complaints of facial swelling since 1 month which gradually progressed to generalized anasarca followed by burning pain in left hand fingers since 15 days associated with blackish discoloration of fingers since 12 days. Gradually she developed nasal twang & nasal regurgitation since 12 days, recurrent hiccoughs since 6 days, weakness & inability to move her left hand & feet since 3 days. On examination, her weight was 27 kg, pulse-98/min, BP160/90 mmof Hg in lying down position. Patient was pallor with no cyanosis, icterus & clubbing. On investigating her Hb-10.4 gms/dl, TLC-11800/mm3, platelet count-4.68lac/mm3, Blood urea-22 mg/dl, Creatinine-0.9 mg/dl, Serum Sodium-135 mmol/dl, Serum Pottassium-4 mmol/dl,s.protein-3.4 gm/dl, S. Albumin-1.95 gm/dl, Globulin-1.49 gm/dl, S.cholesterol-590 mg/dl, Triglicerides-350 mg/dl, HDL-40 mg/dl, LDL-480 mg/dl, VLDL-70 mg/dl, Urinalysis- albumin 2+, Pus cells-2-3/hpf, RBC-nil/hpf, Epi. Cells-3-4/hpf, 24 hr urinary protein- 2.4 gm, CRP-negative, HbsAg and HIV were negative. Other immunological tests C3,C4 level, RA factor, anti ds DNA,pANCA,cANCA were negative, Malaria card test-negative, BT-1min15 sec., CT-1min 45 sec., Fundus- mild arterial attenuation. Color Doppler of left upper hand showed thrombosis of middle 1/3 of ulnar artery, ECG showed sinus tachycardia with inferolateral ischaemia. MRI Head revealed infarct in the left lateral aspect of lower medulla. Patient was started on torsemide, cefoperazone, sulbactum, deflazacort, low

molecular weight heparin & albumin infusion. Her facial swelling diminished & her serum protein & albumin showed improvement.

Conclusion: Hypercoagulability is a complication seen in NS. Not only venous thrombosis is very common at any site but also spontaneous arterial thrombosis may occur. In nephrotic syndrome, thrombosis of major limb arteries is uncommon but feared complication.

AA-182: IGG ASSOCIATED MESANGIAL PROLIFERATIVE GLOMERULONEPHRITIS: A CASE REPORT

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Glomerular deposits are a major pathologic feature of a wide range of human glomerulonephritides. Various patterns of mesangial glomerular deposits have been reported. Glomerular Ig deposits are encountered in a large number of primary and secondary glomerulonephritis, such as lupus or cryoglobulinemic nephropathy. Rare cases of primary glomerulonephritis defined by predominant mesangial IgG deposits.

We report on a case of a 7-year-old boy diagnosed IgG mesangioproliferative glomerulonephritis by kidney biopsy. He was found gross hematuria suddenly and then hematuria improved not having medical treatment gradually. But microscopic hematuria continued for several months. Hypertension was no observed and physical examination was unremarkable. The first visit, In urine microscopy RBC was detected above 100 cells/HPF, and pyuria, proteinuria were not detected. A spot urine creatinine, calcium, microalbumin were 44 mg/dL, 24.1 mg/dL, 8.14 mg/dL, respectively. Serum protein, albumin, glucose, BUN, creatinine were unremarkable. Serum IgG was 561 mg/dL, and IgA, IgM, C3, C4 were normal. Ultrasonography showed mild chronic hydronephrosis in right kidney. Abdominal CT revealed isolated dilated upper polar calyx in right kidney. He underwent percutaneous renal biopsy, and 70 glomeruli were available for study. In light microscopy, the glomeruli showed mild increased mesangial matrix and cellularity. No crescentic or sclerotic glomerulus was found. In immunofluorescent examination, the patient showed a deposition of IgG in the glomerular mesangium. IgA and IgM were completely negative. Electron microscopic examination showed that two glomerulies revealed mild degree of scattered mesangial and paramesangial electron dense deposits, and effacement of foot processes were significantly noted. He has no symptom at now, and we follow up the patient on a regular basis.

AA-192: COMMON SERIOUS INFECTIOUS COMPLICATIONS OF CHILDHOOD NEPHROTIC SYNDROME AT CHILDREN'S HOSPITAL N01

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Introduction: Infection is one of the most common complications and still remains a main cause of mortality in children with nephrotic syndrome. This article aimed to evaluate the epidemiology, clinical and laboratory characteristics, and pathogens of common serious infectious complications of children with nephrotic syndrome in order to contribute to a reduction of mortality rate of the disease.

Methods: From November 2004 to August 2005, 41 episodes of serious infections of 35 children with nephrotic syndrome at Department of Nephrology, Children's hospital N01 were enrolled in a prospective case series study. Serious infections were the ones which can promptly cause death and need to be treated with intravenous antibiotic.

Results: Mean age of the patients was 8 +/- 4 years, male to female ratio was 1.86/1. 24.4% of infectious episodes occurred at the first onset of nephrotic syndrome while 75.5% of the others occurred at recurrent or steroid resistant nephrotic syndrome. 70.7% of the patients were being treated with only steroid or in combination with other immunosuppressant drugs. 100% of the infectious episodes occurred simultaneously in an active phase of nephrosis. Symptoms implying infectious complication include fever, leukocytosis above 15000/mm3, neutrophil count above 7500/mm3 and CRP above 20 mg/l which were in turn 68.3%, 68.3%, 75.6% and 51.2%. Three kinds of common serious infectious complications were primary peritonitis (34.1%), pneumonia (34.1%) and cellulitis (31.7%). Bacterial pathogens were isolated in 13/41 (31.7%) of severe infectious episodes: S. pneumoniae 17.1%, E. coli 4.9%, Pseudomonas spp 4.9%, Klebsiella spp 2.4% and Enterobacter spp 2.4%. 80% of isolated Gram negative bacteria were pathogens of infectious cases admitted hospital after 48 hours while 75% of isolated Streptococcus pneumoniae related to pre-admitted hospital infections. 80.5% of serious infectious episodes were successfully treated with Cefotaxime +/-Aminoglycoside and the mean therapeutic time was 9.3 +/- 2.2 days. Death occurred in 2 cases of pneumonia, accounted for 4.9% of the overall mortality rate.

Conclusions: Serious infections are common complications with a high mortality rate in the active stage of childhood nephrotic syndrome. Cefotaxime +/- Aminoglycoside is an appropriate initial antibiotic choice for treatment of serious infectious complications in nephrotic syndrome. Alternative therapeutic protocol should be a combination of Vancomycin for coverage of Penicillin resistant S. pneumoniae and a Fluoroquinolone for coverage of Gram negative bacteria before available results of bacterial isolation.

AA-196: THE EFFECTS OF ANGIOTENSIN CONVERTING ENZYME INHIBITOR AND ANGIOTENSIN II RECEPTOR BLOCKER ON STEROID RESISTANT NEPHROTIC SYNDROME IN CHILDREN

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Introduction: Steroid resistant nephrotic syndrome (SRNS) in children is very difficult for treatment and often has unfavorable outcome. Reduced proteinuria has delay worsening of renal function and remission of proteinuria predicts a good long-term prognosis in children with nephrotic syndrome.

Method: The aim of this study was to reduce proteinuria in persistent proteinuria patients. This prospective descriptive study was designed

to study the effectiveness of angiotensin converting enzyme inhibitor (ACEI) and angiotensin receptor blocker (ARB) on proteinuria reduction in steroid resistant nephrotic syndrome in children. Enalapril was given at starting dose of 0.1 mg/kg/day and stepped up 0.1 mg/kg/day every 4 weeks until 0.6 mg/kg/day (maximum 40 mg/day) and losartan 1 mg/kg/day was added and stepped up to 2 mg/kg/day (maximum 100 mg/kg/day) for another 4 weeks.

Result: During September 2003 through December 2007 we diagnosed 8 SRNS 5 male (26.5%) and 3 female (37.5%) with mean age at diagnosis 8.3+4.1 years (2.05–13 years) and age at enrollment 11.7+3.8 years (6–16 years). Renal histology were 7 FSGS (87.5%) and 1 IgM nephropathy (12.5%). 75% of them were treated with oral cyclophosphamide. The result of combine treatment with high dose enalapril and losartan showed significant reduction on mean spot urine protein: creatinine ration from 9.85+2.25 to 3.64+1.59 ($P<0.05$) and 24 hr. urine protein from 182.79 +59.56 to 28.68+8.23 mg/m2/hr ($P<0.05$). Serum cholesterol and albumin showed improvement but not statistic significance. Blood pressure and renal function were not significant change. No clinical and laboratory side effect were reported during treatment.

Conclusion: Combine renin anigiotensin blockade with high dose ACEI and ARB was safe and effective proteinuria reduction in childhood SRNS.

AA-197: DAILY CORTICOSTEROIDS PREVENT INFECTION ASSOCIATED RELAPSES IN PATIENTS WITH FREQUENTLY RELAPSING NEPHROTIC SYNDROME (FRNS)

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Introduction: Patients with FRNS are at risk for complications during relapses and side effects of immunosuppressive agents. Since 50-70% of relapses follow minor infections, interventions that reduce the risk of infection associated relapses are valuable. In a randomized controlled trial, we examined if short-term (7-day), daily administration of prednisolone was effective in preventing infection associated relapses in patients with FRNS treated with alternate-day (AD) prednisolone with/without levamisole.

Method: Following approval from the Ethics Committee & parental consent, 100 patients, between 1 and 16 yr-old with idiopathic FRNS (>2 relapses in 6 months) were included and stratified into those receiving long-term AD prednisolone (0.5–1 mg/kg) or AD prednisolone & levamisole. During intercurrent infections, patients were randomized to either continue the above therapy (non-intervention group) or receive the same dose of prednisolone every day for 7-days (intervention group). The duration of the study was 12-months.

Result: Results for the first 6-months are presented. Patients in the intervention (n=50) and non-intervention (n=50) groups had similar baseline clinical & laboratory features. The frequency of infections was similar in the two groups. Mean (95% confidence intervals) relapse rates in the first 6-months in the intervention and non-intervention groups was 0.8 (0.3, 1.2) and 1.0 (0.5, 1.5) respectively ($P=0.1$). A higher proportion (62%) of patients in the former group had sustained remission compared to the latter (44%). The mean (SD) frequency of relapses following infections in the intervention and non-intervention groups was significantly different at 0.38 (0.49) and 0.96 (1.0) respectively ($P=0.009$).

Conclusion: Short-term (7-day) daily administration of prednisolone, at the same dose as that being taken on alternate-days, at the onset of infections significantly reduces the subsequent risk of relapses. The benefit of this approach seems promising in reducing infection precipitated relapses in patients with FRNS.

AA-199: NEPHROTIC SYNDROME WITH IGM NEPHROPATHY: RESPONSE TO TREATMENT AND LONG-TERM OUTCOME

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Introduction: Minimal change or mild mesangial proliferation with IgM deposit (IgM nephropathy) are common pathological findings in frequent relapsing or steroid dependent nephrotic syndrome in Thai children. Response to alkylating agents and long-term outcome are not well elucidated.

Methods: A retrospective study was performed in 48 nephrotic children with relapsing, dependence or resistance to steroid and IgM nephropathy at the Department of Pediatrics, Ramathibodi Hospital during January 1990-December 2007.

Results: There were 30 males and 18 females (M:F=1.67:1), median age at onset of nephrotic syndrome (NS) was 4.33 years (0.1–14), hematuria 23.4%, hypertension 35%, mean serum albumin 15.8+6.22 g/L, and eGFR 146+58 mL/min/1.73 m2 (6.25% with CKD II). Prior to the biopsies, 33 were steroid dependence, 6 were relapsing, 1 was partially response and 8 were resistance. Kidney biopsies were performed 18.5 months (0–153) after the onset of NS. Forty-five patients were treated with cyclophosphamide (34), chlorambucil (6), cyclosporine (3) and prednisolone (2). At the last follow-up (median time after the biopsies 4.41 years), 24 patients (53.3%) were in remission, 20 patients (44.4%) were relapsing, and one was not responded to above treatments. The eGFR were increased to 162+50 mL/min/1.73 m2 with only one patient in CKD II.

Conclusions: Relapsing, steroid dependence or resistant NS with IgM nephropathy were responded to alkylating agents with good long-term outcomes.

AA-203: CLINICAL MANIFESTATION OF MEMBRANOUS LUPUS NEPHRITIS IN CHILDREN

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Introduction: Membranous lupus nephritis (MLN), classV lupus nephritis, accounts for about 20% of renal involvement in systemic lupus erythematosus. The different classifications used in various reports resulted in inconsistent clinical manifestation and outcome in this group.

Method: Medical records of pediatric patients with LMN diagnosed by using the International Society of Nephrology/Renal Pathology Society (ISN/RPS) 2003 classification of lupus nephritis at Chiang Mai University hospital were retrospectively reviewed.

Result: From 2004 to 2007, the total number of 92 kidney biopsies were performed in children with SLE. Twenty eight patients (30%) had LMN, twelve of them (43%) was mixed-type LMN (combination of class V and III or class V and IV). The mean age at diagnosis was 11.1 years old. The clinical of nephrotic syndrome was recognized in 36%. Hypertension was noted in 57%. The mean 24-hr urine protein and urine protein to creatinine ratio were 105 mg/kg/day and 6.0, respectively. The mean serum creatinine and glomerular filtration rate (GFR) were 0.8 mg/dL and 107 ml/min/1.73 m2. The GFR of less than 90 ml/min/ 1.73 m2 was noted in 37% of patients. Most patients (86%, 79%) had microscopic hematuria and nephrotic range proteinuria, respectively. The mixed-type LMN had tendency to have lower initial GFR and higher serum creatinine than the pure membranous type.

Conclusion: Based on the ISN/RPS 2003 classification, the incidence of MLN in this study is higher than the previous reports. The mixed-type group presents with lower GFR and higher serum creatinine than the pure type.

AA-206: CLINICAL OUTCOME OF PEDIATRIC SLE AND LUPUS NEPHRITIS: A MULTICENTER STUDY OF 500 CASES IN THAILAND

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Objective: To determine the clinical outcome of Thai children with SLE.

Background Children with SLE especially those with nephritis are not uncommon among children with kidney diseases in Thailand. Due to its chronic nature, the disease can lead to growth and developmental impairment.The cost of care especially if kidney failure occurred is very high. Data collection for the whole country is needed in order to improve the care and prevent serious complication in these patients.

Methods: The data of patients with SLE, age at diagnosis less than 15 years, diagnosed during 1 January 2002 to 31 December 2006 in thirteen tertiary care hospitals were reviewed. Severity and complication of the disease were assessed every six to twelve months period using ECLAM index and SLICC/ACR damage index respectively.

Results: There were 500 patients,77 (15.4%) male and 423 (84.6%) female. The mean age at diagnosis was 11.1 + 2.5 years (range 1-15 years). The patients were followed for a mean period of 21.2 months (range 0-61 months). Renal involvement was found in 410 patients (81.8%). WHO class 4 was the most common renal histopathology, which was found in 185 patients (52.7%). The mean ECLAM index at diagnosis was 6 + 2.7 and decreased significantly after treatment. Chronic renal insufficiency (CCr < 60 ml/min/1.73 m2) was found in 11 patients (2.2%). Risk factor for chronic renal insufficiency was initial GFR< 60 ml/min/ 1.73 m2. Mean SLICC/ACR damage index in 324 patients who were followed for more than one year (mean follow up time 26.4 months) was 1.35.The damage frequently found in ocular (22.5%), renal (10.5%), neuropsychiatric (8.5%) and musculoskeletal (6%) systems. All patients received prednisolone. 62% received prednisolone plus other immunosuppressive drugs. Complete and partial renal remission were found in 60 and 27% of patients respectively.14 patients (2.8%) died in 1-39 months. The most common cause of death was infection.

Conclusion: Children with SLE especially those with nephritis are not uncommon in Thailand. Renal pathology of WHO class 4 is frequently found. With current immunosuppressive treatment, majority of patients achieve favorable outcome. However, infectious complication is the leading cause of death.

AB-022: ASSOCIATION OF INVS (NPHP2) MUTATION IN AN ADOLESCENT EXHIBITING NEPHRONOPHTHISIS (NPH), AND COcomplete situs inversus

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Introduction: Cystic kidney disease has been linked to mutations in the Invs gene in mice with an inversion of embryonic turning (inv/inv) and the INVS (NPHP2) gene in human infantile nephronophthisis (NPH). NPH is one of the most important kidney disorders which can cause ESRD in children and young adults. NPHP2 maps to chromosome 9q21-q22 and recent studies have suggested that NPHP2 is associated with INVS. Several mutated alleles in INVS were identified in patients with infantile NPH. Some individuals exhibited extrarenal anomalies such as situs inversus, cardiac ventricular septal defect, and arterial hypertension. Infantile NPH shows marked cyst formation in contrast to other forms of NPH, and rapidly progresses to end-stage renal failure (ESRD) before 5 years of age.

Method: In this report, we describe an adolescent with a mutation in INVS who had preservation of his renal function beyond infancy. The patient showed findings of NPH with mild renal insufficiency together with situs inversus. He also exhibited a series of features consistent with Jeune syndrome involving asphyxiating thoracic dystrophy, heart failure, and hypertension prior to advanced renal insufficiency. NPH also has been associated with Jeune syndrome. Based upon these features, our patient is likely to have the combined clinical features of infantile NPH with Jeune syndrome.

Result: Genetic analysis for INVS disclosed a heterozygous mutation of T→G at position rs7024375 in the 5'UTR of INVS in the patient and his mother, while no abnormalities were found in any of the 17 exons of INVS or NPHP1, 3, and 4. This substitution in 5'UTR of INVS might influence the stability of INVS mRNA, which could compromise inversin protein transcription. The instability of inversin protein synthesis due to this heterozygous mutation may be responsible for the slowly progressive course of renal insufficiency in our patient.

Conclusion: To our knowledge, this is the first patient possessing a genetic alteration in INVS who had preservation of renal function past childhood. This study may suggest that our patient is a compound heterozygote for infantile NPH and Jeune syndrome, and is presumably a novel clinical variant of infantile NPH.

AB-048: EXPERIMENT OF TRANSPLANTING METANEPHRIC MESENCHYMAL TRANSFECTED BY UPA GENE TO THERAPY CHRONIC KIDNEY DISEASE

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Introduction: Study regeneration role and possible mechanism of metanephric mesenchymal and metanephric mesenchymal transfected by uPA gene in CKD.

Method: 1. Construction and package of recombinant plasmid of uPA (urokinase-type plasminogen activator) gene and pAAV-IRES- hrGFP vector. 2. The viral particles of rAAV and AAV were transfected into rat′s metanephric mesenchymal (rat-inducible metanephric mesenchymal-18, RIMM-18) in vitro and the positive RIMM-18 cells that expressed GFP were detected using the inverted fluorescence microscope. 3. Study regeneration role and possible mechanism of metanephric mesenchymal and metanephric mesenchymal transfected by uPA gene in CKD that were transplanted via caudal vein.

Result: 1. (1) uPA cDNA obtained from immature rat kidney by reverse transcription polymerase chain reaction method were correct. (2) The results suggested the recombinant AAV vectors carrying uPA gene were constructed successfully. The sequence of the recombinant plasmid of pAAV-hrGFP-uPA were proved identical to the reported cDNA sequence by DNA sequence analysis. (3)The viral particles titer of recombinant AAV and uPA gene were 1.12×1012 vg/mL by dot hybridization and AAV were 1.0×1012 vg/mL. (4)AAV-293 cells were transfected by rAAV viral particles and AAV viral particles and the ratio of positive cells after 48 h were 80% and 95% respectively when the MOI=1×105. 2. (1) The rate of RIMM-18 cells that were transfected by viral particles of rAAV and AAV were 60%; (2)The expression of GFP were decreased with the passage of transfected RIMM cells; (3)The transfected RIMM-18 cells still have the original characteristic and were vimentin positive and cytokeratin negative; (4) There were uPA gene in RIMM-18 cells transfected by rAAV and no uPA gene in RIMM-18 cells transfected by AAV by RT-PCR; (5) There were no uPA expression in RIMM-18 cells transfected by rAAV or AAV by Western blotting. 3. (1) At 14d, 21d, 28d, renal interstitial injury of rAAV group, AAV group and RIMM-18 group's rats were more relieved versus those of model group and culture medium group. (2)Urine protein in 24h, blood urea nitrogen, serum albumin and endogenous creatinine clearance rate were not different between five groups. At 14d, 28d, serum cretinine of rAAV group, AAV group and RIMM-18 group's rats were less than those of model group and culture medium group. (3) The expression of uPA by semiquantitive analysis of immunohistochemical staining of rAAV group, AAV group and RIMM-18 group's rats were stronger than those of model group and culture medium group.The expression of PAI-1 and TGF-β1 of rAAV group, AAV group and RIMM-18 group's rats were less than those of model group and culture medium group. (4) The uPA gene by RT- PCR analysis of rAAV group's rats were stronger than those of AAV group, RIMM-18 group, model group and culture medium group.There were no different between rAAV group's rats at 7d, 14d, 21d, 28d. The PAI-1 and TGF-β1 gene of rAAV group, AAV group, RIMM-18 group's rats were less than those of model group and culture medium group. (5) The expression of uPA by Western blotting of rAAV group, AAV group and RIMM-18 group's rats were stronger than those of model group and culture medium group.The expression of PAI-1 and TGF-β1 of rAAV group, AAV group and RIMM-18 group's rats were less than those of model group and culture medium group. (6)There were GFP in the obstructed renal tissue of rAAV group and AAV group's rats at 1.5d, 3d, 7d, 14d, 21d, 28d and at the same time there were GFP in the contralateral renal tissue. There were GFP in the liver and spleen tissue at 1.5d, 3d and GFP became less at 7d, 14d, 21d, 28d. There were no GFP in the heart tissue and blood smears at every time.

Conclusion: 1.The recombinant plasmid of AAV and uPA gene were successfully constructed and packaged. 2.The transfection rate of rAAV and AAV viral particles to RIMM-18 was as high as 60%. AAV vector can deliver uPA gene effectively. 3. (1)RIMM-18 cells could improve renal function and reduce fibrosis of UUO rats by transplantation via caudal vein. (2)The transferred uPA gene could not express.AAV vector had no effect on RIMM-18 cells. (3)RIMM-18 cells transfected by rAAV and AAV viral particles could home to injured renal by obversing the expression of GFP under the fluorescence microscope.GFP positive cells settled in tubular and RIMM-18 cells could repair injured renal. (4) There were GFP positive cells in contralateral renal, liver and spleen tissue and no GFP positive cells in heart tissue and blood smears. (5)There were no immunological rejection and tumor in cell transplantation groups.

AB-073: EXPRESSION OF Non-cleavable Membrane-anchored HB-EGF enhances Cell Spreading in MDCK cells

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Introduction: Heparin-binding EGF like growth factor (HB-EGF) is synthesized as a membrane-anchored precursor and is capable of activating EGF receptors in a juxtacrine manner. As epithelial cells attach and spread, they may make contact with neighboring cells, and focal complex processes including lamellipodia are considered premature cell-cell contact sites. In MDCK cells, stable expression of a non-cleavable and membrane-anchored human HB-EGF construct (MDCKH5AA cells) resulted in enhanced cell spreading and formation of significantly extended lamellipodia and filopodia compared to wild type (WT) cells. There was increased tyrosine phosphorylation of paxillin and FAK in the MDCKH5AA cells compared to the control cells, while Src tyrosine phosphorylation was not different between the two cell lines. In addition, there was a marked increase in phosphorylation of Akt in MDCKH5AA cells compared to the control cells whereas ERK 1/2 phosphorylation was no different between the two cell lines.Membrane-anchored HB-EGF associates with glycoproteins and ECM through its proximal (heparin binding) domain and with the tetraspanin, CD9 through its EGF-like domain. Chimeric proteins with substitution of either the TGF-a proximal domain, which has no heparin-binding capability, or EGFs receptor binding domain, which does not interact with CD9, both decreased the level of cell spreading, although the soluble, cleaved form of both of these constructs retained full capacity to activate EGFR. Taken together, our findings suggest that membrane-associated HB-EGF enhances cell spreading and cell-cell adhesion by juxtacrine activation of EGFR. During spreading, optimal interaction of HB-EGF with EGFR may require the participation of other accessory proteins.

Method: In MDCK cells, stable expression of a non-cleavable and membrane-anchored human HB-EGF construct (MDCKH5AA cells) resulted in enhanced cell spreading and formation of significantly extended lamellipodia and filopodia compared to wild type (WT) cells.

Result: There was increased tyrosine phosphorylation of paxillin and FAK in the MDCKH5AA cells compared to the control cells, while Src tyrosine phosphorylation was not different between the two cell lines. In addition, there was a marked increase in phosphorylation of Akt in MDCKH5AA cells compared to the control cells whereas ERK 1/2 phosphorylation was no different between the two cell lines. Time-lapse video showed that the formation of the long lamellipodia and filopodia in MDCKH5AA cells was a late event (at least 3 hours post-plating) and

was more prominent with increasing cell density. Cells expressing a non-cleavable HB-EGF construct mutated to prevent EGFR activation (MDCKH5aa cells) had no change in cell spreading compared to WT, suggesting a necessary role for EGFR activation.

Conclusion: Our findings suggest that membrane-associated HB-EGF enhances cell spreading and cell-cell adhesion by juxtacrine activation of EGFR. During spreading, optimal interaction of HB-EGF with EGFR may require the participation of other accessory proteins.

AB-074: THE SLIT DIAPHRAGM ASSOCIATES WITH PROTEINURIA IN ALPORT SYNDROME

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Introduction: Alport syndrome is a hereditary glomerulonephritis caused by primary glomerular basement membrane deficiency. Proteinuria is an important risk factor for the disease progression, but the mechanism is still on debate. The slit diaphragm which plays key role in other proteinuric renal disease also changes in Alport syndrome patients with proteinuria, however, the roles of the slit diaphragm is not clear. This study aimed to disclose the association between the slit diaphragm and proteinuria in Alport syndrome.

Method: Twenty one Alport syndrome children were studied with age of 2 to 17 years old and the proteinuria duration ranged from 20 days to 11 years. According to the degree of 24 hrs urinary protein (UP), the patients were divided into mild group (UP <30 mg/kg), moderate group(UP 30~50 mg/kg) and heavy group (UP>50 mg/kg). The podocyte foot process width (FPW) was assessed by morphometric analysis. The relationship between FPW and UP was analyzed. By immunoperoxidase staining on paraffin embedded renal sections, slit diaphragm molecules nephrin, podocin and actin associated protein synaptopodin were studied.

Result: The FPW correlated with UP significantly($r=0.765$, $P<0.01$). The FPW of the mild group was significantly narrower than that of heavy group (551+174 nm vs. 1513+776 nm, $P<0.05$). Unexpectedly, two patients with heavy proteinuria did not show obvious foot process effacement, their durations of proteinuria were relative short (1 year). The staining of nephrin and podocin changed both in heavy proteinuria patients with and without foot process effacement, but synaptopodin preserved in the latter.

Conclusion: The extent of slit diaphragm disappearance reflected by FPW associated with proteinuria in Alport syndrome. Slit diaphragm molecules nephrin and podocin were involved in the mechanism of heavy proteinuria irrespectively of foot process effacement. Early intervention might contribute to delay podocyte dramatic change and disease progression

AB-133: TWO CASES WITH EPSTEIN ANOMALIES: GENETIC AND CLINICAL ANALYSES

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Introduction: May-Hegglin anomaly (MHA) is a rare autosomal dominant type of hereditary disease characterized by thrombocytopenia, large size of platelet and inclusion bodies in WBC. Clinically May-Hegglin anomaly is further divided into several clinical subgroups, one of which is Epstein syndrome. In Epstein syndrome, progressive renal insufficiency and sensorinuronal deafness are present. Recently, the gene encoding for type A myosin heavy chain 9 (MYH9) was identified as a responsible gene for MHA. In this paper, we describe the genetic and clinical analyses of two patients with Epstein syndrome.

Result: Case 1 was diagnosis as having MHA in infancy by thrombocytopenia and large size of platelet. Proteinuria gradually developed after school age, and had been treated with ACEI and ARB. He developed ESRD at 17. Genetic analysis revealed a heterozygous R702C mutation in MYH9 gene. Case 2 is a 3-year-old girl. Thrombocytopenia was noticed during neonatal period and she was referred to our hospital. MHA was suspected because of large size of platelet and inclusion bodies in WBC. Genetic analysis revealed heterozygote R702C mutation in MYH9 gene. Proteinuria was apparent as early as 2 years and 8 months, and now is treated with ARB.

Conclusion: The precise mechanisms by which mutations in MYH9 gene caused nephropathy is still unknown. The fact that proteinuria is dominant in Epstein syndrome might be explained by the fact that type A myosin heavy chain is expressed mainly in podocytes and endothelial cells, while hematuria is cardinal abnormality in Alport syndrome, which is cased by structural changes of GBM. This study also indicates that R702 mutation in MYH9 is a hot spot causing Epstein syndrome.

AB-136: Novel phenotype and mutations in Dent's disease

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Introduction: Background: Dent\'s disease (X-linked nephrolithiasis) is a tubulopathy secondary to inactivating mutations in the chloride channel in proximal tubular cells. Its features vary in ethnic groups. Aim: We examined 3 unrelated Indian families with an index patient with Dent's disease for CLCN5 mutations.

Method: After approval from the Ethics Committee and parental consent, EDTA and urine samples were collected from families of patients presenting with Dent's disease. Total urine protein was measured using VITROS microslide technology (Ortho-Clinical Diagnostics GmbH, Germany). For detection and characterization of proteins from urine, samples were run by SDS-PAGE on a SEBIA Hydragel 5 kit. Low molecular weight proteins were measured by immunonephelometry. Leukocyte DNA was amplified by polymerase chain reaction using primers directed to all 12 exons of the CLCN5 gene, followed by automated sequencing. DNA samples from 50 unrelated healthy subjects served as controls.

Result: Three boys, with age of onset between 1-2 yr (present age 4-yr, 19-yr and 21-yr) were studied. All presented with failure to thrive, polyuria, polydipsia, salt craving and refractory rickets. All had recurrent episodes of night blindness, responsive to vitamin A therapy. There was no family history of similar disease. Investigations showed hypophosphatemic rickets, low molecular weight proteinuria and hypercalciuria (12-20 mg/kg/day). They had no evidence of nephrolithiasis or nephrocalcinosis, renal dysfunction, aminoaciduria or

glucosuria. Genetic evaluation showed M504K, W58C and L729X mutations on CLCN5 gene respectively. Their mothers were carriers in all cases.

Conclusion: We report novel mutations of the CLCN5 gene in three patients presenting with polyuria, polydipsia, refractory rickets, hypercalciuria, low molecular weight proteinuria and recurrent night blindness in early childhood, in the absence of nephrolithiasis or nephrocalcinosis. Our findings expand the spectrum of features reported with Dent's disease.

AB-151: THE ASSOCIATION OF AT2R GENE POLYMORPHISM A190G WITH THE DEVELOPMENT OF PRIMARY VESICOURETERAL REFLUX IN TAIWANESE CHILDREN

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Introduction: Primary vesicoureteral reflux (VUR) is a common pediatric disease that may lead to reflux nephropathy and end-stage renal disease in some patients. Current knowledge indicates that the evolution of VUR is not equal in all patients. It suggested the influence of various factors including genetics. The aim of this study is to investigate the association of the Type 2 Angiotensin II Receptor (AT2R) gene polymorphism with the development and severity of primary VUR.

Method: We studied the AT2R gene polymorphisms A190G for association with development of primary VUR and disease severity in 111 VUR children and 60 healthy controls. Sixty of the 111 VUR patients had low-grade VUR (grade I-III) and 51 had high-grade VUR (grade IV and V). To analyze the polymorphisms in the A190G of AT2R gene, the SNP genotyping assay were performed. The genotypic frequency and allele frequency were analyzed to detect the correlation between the patients with mild, severe VUR, and healthy control.

Result: We found that the A190G of AT2R gene was associated with the development of VUR in both male and female patients. Significantly higher G and lower A allele frequencies were presented in male VUR patients (G allele 0.39 and A allele 0.61) compared with control (G allele 0.30 and A allele 0.70, p=0.011). The AG genotype was higher in female VUR patients compared with controls (P=0.005).There is no association with the severity of VUR in both male and female patients.

Conclusion: AT2R A190G gene polymorphism was associated with the development of the VUR in both male and female patients. There is no association with the severity of VUR in Taiwanese children. It raises the possibility to utilize the genotype of AT2R as a risk factor to evaluate the development of primary VUR.

AB-161: INVOLVEMENT OF TRPC6 IN ANGIOTENSINII-INDUCED PODOCYTE APOPTOSIS

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Introduction: Some evidences prove angiotensin II can lead to podocyte injury, and Ang IIcan affect slit diaphragm complex molecules. Transient receptor potential protein 6 (TRPC6) is a non-selective cation channel, which represents a component of the glomerular slit diaphragm. AngII can enhance Ca2+ entry via mutant TRPC6. These studies explore whether TRPC6 is involved in podocyte injury induced by AngII.

Method: Mouse podocyte clone (kindly provided by Prof. Peter Mundel) was cultured at 33°C for proliferation and at 37°C for differentiation. After treatment with angiotensinII(10–7M) for 12 h, 24 h, 48 h and 72 h, the protein expression of TRPC6 and other SD molecules were detected by western blotting. Podocyte apoptosis was analyzed with Annexin V-FITC and PI staining by flow cytometry. Cells were loaded with Fluo-3am calcium indicator and stimulated by OAG plus 2 mM Ca2+. The alteration of intracellurla Ca2+ was detected by using FLX 800 spectrophotofluorometer. To verify the role of TRPC6 in these alteration induced by AngII, TRPC6 siRNA was transfected according to the standard protocol.

Result: After treatment with AngII, the protein expression of TRPC6 was increased at 72 h, while the podocin protein was degreased at 24 h, 48 h and 72 h. Other SD molecular expressions (nephrin, ㄹ-actinin-4, and CD2AP) didn't change. The percentage of apoptotic podocytes was significantly increased at 48 h and 72 h. After transfection with TRPC6 siRNA in AngII-treated podocytes, the apoptosis was decreased, but, it was higher than that of control group. The intracellular Ca2+ was increased in podocytes treated with AngII for 72 h. After transfection with TRPC6 siRNA, The increased influx of Ca2+ was lower than that of AngII-treated group.

Conclusion: AngII can increase the protein expression of TRPC6 and decrease that of podocin. TRPC6 is involved in AngII-induced podocyte apoptosis, in part. The increased influx of Ca2+ in podocytes is a component of podocyte injury, mediated by TRPC6 channels.

AB-200: ASSOCIATION BETWEEN CELL SPREADING AND JUXTACRINE MANNER OF NON-CLEAVABLE MEMBRANE-ANCHORED HB-EGF IN MDCK II CELLS

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Introduction: Heparin-binding EGF like growth factor (HB-EGF) is synthesized as a membrane-anchored precursor and is capable of activating EGF receptors in a juxtacrine manner, which associated with cell survival and epithelial integrity. As epithelial cells attach and spread, they may make contact with neighboring cells, and focal complex processes including lamellipodia are considered premature cell-cell contact sites.

Method: We constructed MDCK II (wild type) cells stably expressing a non-cleavable and membrane-anchored human HB-EGF (H5AA cells) and H5AA cells mutated to prevent EGFR activation (mH5AA cells). Using these cells, we evaluated morphological differences and expression of foxal adhesion complex in western blot analysis.

Result: H5AA cells resulted in enhanced cell spreading and extension of lamellipodia compared to wild type (WT) cells. Western blot analysis shows there was increased tyrosine phosphorylation of paxillin and FAK in the H5AA cells compared to WT cells, while Src tyrosine phosphorylation was not different between the two cell

lines. In addition, mH5AA cells had no change in cell spreading compared to WT.

Conclusion: The membrane-associated HB-EGF enhances cell spreading and cell-cell adhesion by juxtacrine activation of EGFR. During spreading, optimal interaction of HB-EGF with EGFR may require the participation of other accessory proteins.

AC-020: HYPER-RENIN HYPERTENSION – REPORT OF TWO RARE TREATABLE CAUSES.

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Introduction: Hypertension is relatively uncommon in the pediatric age group affecting less than 5% of children. Severe hypertension is usually due to a secondary cause. The likelihood of identifying a cause is inversely related to the age of the patient and directly related to the degree of elevation of blood pressure. We report here two adolescents with severe hypertension and hyper-renin secretion in whom two rare remediable causes were found.

Method: Case 1: An 11 year old presented with severe headaches and was found to have severe hypertension requiring 3 anti-hypertensive medications. Her father was also known to have hypertension from the age of 30 years and a cause for this was not found. Our patient manifested persistent hypokalaemia (serum K 3.1-3.5 mmol/L) and high plasma renin levels. Further investigation led to the diagnosis of a reninoma. Following resection, she has mild hypertension (3 months post-surgery) and is controlled on monotherapy. Case 2: A 7 year old boy being treated with potassium supplements for hypokalaemic periodic paralysis developed hypertension 7 years later. The blood pressure required two antihypertensives for control. There was no family history of hypertension. Evaluation revealed hyper-renin hyperaldosteronism. A long drawn out search for a renovascular cause then ensued and revealed a dual supply to the right kidney with a long aberrant supply from the bifurcation of the aorta to the lower pole. The patient refused surgical intervention and is currently aged 19 years and on 2 antihypertensive drugs

Result:

Conclusion: Although essential hypertension is increasingly recognized in the adolescent age group, the young age of a patient and severe elevations of blood pressure should trigger the search for an identifiable and potentially treatable cause for this condition which has potentially high risk of morbidity and mortality.

AC-076: A RETROSPECTIVE ANALYSIS OF 13 CHILDREN WITH RENAL VASCULAR HYPERTENSION

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Introduction: Renal vascular hypertension (RVH) is a common secondary hypertension in children, earlier intervention can improve the prognosis. We analyzed 13 children with RVH in our department aimed to find their characteristics.

Method: Thirteen children diagnosed with RVH during 2000-2008 were included. The data including etiology, onset symptom, the injury of renal, renal arterial, heart, eye ground and brain, plasma renin and serum potassium level, and treatment were analyzed.

Result: There were 8 boys and 5 girls aged from 8 months to 15 years old. The mean blood pressure was 173/125mmHg. Seven children had Takayasu arteritis, 1 neurofibromatosis, 1 after surgery of tumor, 1 ectopic kidney, 1 renal dysplasia, 1 fibromuscular dysplasia and 1 foreign body in renal arterial. Four children presented with headache, vomiting and convulsion, 4 with polyuria and/or foamy urine and/or edema, 4 with palpitation, and 2 no symptoms. Proteinuria occurred in 80% and alleviated with the normalization of hypertension, most of them were selective proteinuria. Eight children had heart injury, 3 had eye ground involvement. Plasma renin increased in 80% children. High plasma renin with hypokalemia occurred in 45%. Eight patients were diagnosed by renal angiography, 5 by ultrasonography, or computerized tomographic angiography or magnetic resonance angiography. The positive rate of renal vascular Doppler ultrasonography was 67%. The bilateral renal length difference exceeded 1.5 cm in 72% children. Bilateral renal vascular stenosis occurred in 4 children, and single in 9. Eleven children received at least 2 kinds of drugs, 7 undergone renal angioplasty or nephrectomy due to poor effect to drugs.

Conclusion: Takayasu arteritis is the most common etiology revealed by this study. There was a higher ratio of involvement on heart, brain, and kidney. Renal arterial Doppler ultrasonography, the renal size difference exceeds 1.5 cm, higher plasma renin with hypokalemia will support the diagnosis of RVH.

AC-079: A CASE WITH RENAL VASCULAR HYPERTENSION CAUSED BY FOREIGN BODY IN RENAL ARTERIAL

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Introduction: Renal vascular hypertension (RVH) caused by foreign body in renal arterial is rare. The classical disease history from the onset to the nephrectomy of a boy with RVH due to foreign body in renal arterial was studied.

Method: The clinical data were analyzed to find the characteristics.

Result: A 10 years old boy was enrolled in our hospital because of high blood pressure averaged 150/100 mmHg for 3 month, he did not respond to calcium channel blocker combined with beta receptor blocker, no headache, vomiting and convulsion complained. Four month before the onset, he underwent an operation on brain tumor. There is no family history of hypertension. No specific signs were found on physical examination. No involvement of heart and eye ground was found. Laboratory examination found normal urine analysis, clearance rate of creatinine of 57.5 ml/min.1.73 m2. A small sized right kidney of 5.6×1.8×2.0 cm and a normal left kidney of 9.4×3.7×3.6 cm were revealed by ultrasound. By DTPA scan, the GFR was 66 ml/min and 0 ml/min for left and right respectively. Renal angiography showed a 50% stenosis and a tube shaped foreign body in right renal arterial. Four months after onset, he underwent

operation for removing the foreign body from the renal arterial, the hypertension could be controlled with calcium channel blocker, 22 months after the operation, the filtration function of left kidney was damaged, he underwent nephrectomy and the blood pressure normalized. Ischemic renal injury was revealed by renal pathology examination.

Conclusion: It is a rare RVH case caused by foreign body in renal arterial. The prominent finding is the seriously damaged renal function. No presentation was complained, and no heart and eye involvement. For kidney without filtration function, nephrectomy was the best choice to prevent the damage to another kidney.

AD-018: WHICH ANTIBIOTICS SHALL WE CHOOSE FOR THE PATIENTS WHO HAVE FEBRILE URINARY TRACT INFECTION IN THE ANTIBIOTICS-RESISTANT ERA?

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Introduction: It is well known that antibiotics resistance in the microorganism of increased in the adult patients of UTI. However, there are few reports of antibiotics resistance in the childhood patients. We performed a prospective study for the antibiotics responsiveness of childhood patients who have febrile UTI.

Method: A total 110 febrile UTI patients who admitted in the department of pediatrics of Cheongju St. Mary's Hospital from April 2007 to January 2008 were investigated. All the patients had suspicious UTI because of fever higher than 380C and leukocyturia >5/HPF. UTI was confirmed by overgrowth of single strain organism at least 1×105 colony forming unit/mL in the cultured urine. The patients were divided into two groups according to antibiotics used (group A (N=60): ampicillin-sulbactam + tobramycin vs. group B (N=50): cefotaxime). Mean febrile days, needs to change antibiotics, and other laboratory data were evaluated. If fever was persisted until the 3rd day after initial antibiotics started, it was changed to another one.

Result: There were no significant differences in age, sex and mean febrile days before admission. The pathogenic bacteria grown in urine culture were not different between two groups. The incidence of antibiotics failure which means persisted fever until the 3rd day of the initial antibiotics was significantly higher in the group A than in the group B (35% vs. 6%, p=0.004). The mean febrile days after initial antibiotics treatment were longer in the group A compare to the group B (2.2 days vs. 1.8 days, p=0.01). Other laboratory findings and the defects in DMSA scan were not different between two groups.

Conclusion: In this study, we showed the 3rd generation cephalosporine is superior to the conventional ampicillin-sulbactam + aminoglycoside in the treatment of childhood UTI patients.

AD-036: PROBIOTICS DURING BREAST FEEDING MIGHT CONFER PROTECTIVE EFFECTS AGAINST URINARY TRACT INFECTION IN INFANTS

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Introduction: Urinary tract infections are common clinical problems occurring in infants and pediatric patient groups, most frequently caused by uropathogenic E. coli. Breast feeding has known to have protective effect to urinary tract infection, so many clinicians encourage mothers to breastfeed. It has also known that probiotics have protective effect against urianry tract infection. In this study, we examined the synergistic effect of breast feeding and probiotics in breastfed infants.

Method: Double-blinded, placebo-controlled study of 60 mother-infant pairs was performed. The probiotics and control(placebo) groups are statistically no difference in characteristics of infants and mothers. No statistical difference in total breast feeding period in both groups. Mothers of both groups were administered probiotics or placebo and the urines of infants were taken every month. All infants had no urinary tract infection at all in the initiation time.

Result: In probiotics group, urinary tract pathogens were detected in three infants, but in placebo group, urinary tract pathogens were detected in eight infants. The risk of developing urinary tract infection during the first 1 year of life in infants whose mothers received probiotics was reduced in comparison with that in infants whose mothers received placebo(10% and 26.7%, respectively).

Conclusion: Administering probiotics during breast-feeding offers a safe and effective mode of promoting the immunoprotective potential of breast-feeding and provides protection against urinary tract infection during the first 1 year of life.

AD-100: SIGNIFICANCE OF MICTURATING CYSTOURETHROGRAPHY IN DIAGNOSIS OF VESICOURETERIC REFLUX IN CHILDREN

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Introduction: To evaluate the time of micturating cystourethrography (MCU) which is used to detect and grade vesicoureteric reflux (VUR).

Method: Between Jan. 2003 to Dec. 2007, 392 patients who were admitted in our hospital for urinary tract infection (UTI) underwent MCU. The results of B ultrasound, urine microproteinuria, urinalysis, urine culture, DMSA and renal function were evaluated.

Result: 392 patients with UTI were enrolled in this study whose mean age was 2.43+/-3.10 years (range 1 month- 14 years). The mean period before treatment was 2.27+/-7.52 months (range 0.03–72 months). In all of the patients, 162 children were proved to have VUR by MCU, in which 150 patients were primary VUR. There were 83 males and 67 females in the group, the ratio of male: female was 1.23: 1. All patients were divided into two groups (VUR and no VUR). There were significant difference in acute DMSA, renal scar and urine microproteinuria between two groups ($P<0.05$). In our study the incidence of primary VUR in all patients with UTI was 41.33% (150/392). 93 patients(62%) of all patients with VUR were younger than 2 years old, and 39 patients (26%) were 2-7 years old, 18 patients (12%) were older than 7 years.

Conclusion: The UTI patients with abnormal results in DMSA (acute DMSA and renal scar) and urine microproteinuria should be given more attention. Boys who were younger than two years old have more possibility of suffering VUR. We should choice the optimum time for patients with UTI undergoing MCU, early diagnosis and treatment are clinically significant. DMSA, microproteinuria, age and gender are useful markers to identify those patients at high risk for VUR.

AD-190: THE RELATIONSHIP BETWEEN VESICOURETERIC REFLUX (VUR) AND RENAL SCARRING (RS) IN INFANTS WITH URINARY TRACT INFECTION (UTI): ASSOCIATION OR DISSOCIATION

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Introduction: The association of "UTI & VUR" with RS has been well established. However, this relationship tends to dissociate. This review aims to study the relationship between VUR and renal scarring in infants with UTI.

Method: Infants ≦6 months with first UTI presented between Jan 02 to Dec 06 having both MCU & late DMSA scan and having no known urological abnormalities were included. The relationship between VUR and renal scarring were evaluated statistically as follows.

Result: 115 infants having both MCU and late DMSA scan were reviewed. There were 48 refluxing ureters (48/230=20.9%): 10 associated with RS (20.8%) and 38 (79.2%) with no RS. RS occurred in 15.0% of kidneys with grade II VUR, 22.2% for grade III, 71.4% for grade IV, and absent in grade I. The sensitivity, specificity, positive predictive value (PPV) and negative predictive value (NPV) for VUR to identify RS were 34.5%, 77.6%, 20.8% and 87.4% respectively. There were 29 scarred kidneys (29/230=12.6%): 10 (34.5%) (4 with ≧ 2 scars) associated with VUR (grade II–IV) and 19 (65.5%) with no VUR (18 with single scar, 1 with 2 scars). The sensitivity, specificity, PPV and NPV for RS to identify VUR were 20.8%, 87.4%, 34.5% and 77.6% respectively.

Conclusion: In the current review, grade II-IV VUR was associated with RS in different proportions (15.0%-71.4%) while grade I dissociated from RS; and RS with ≧ 2 scars also associated with VUR while single scar appeared to dissociate from VUR. The association/dissociation relationship between VUR and RS persisted with denotation.

AD-191: A 20 YEAR- RETROSPECTIVE ANALYSIS OF SPECTRUM AND ANTIBIOTIC RESISTANCE OF UROPATHOGENS ISOLATED FROM PEDIATRIC INPATIENTS WITH COMMUNITY-ACQUIRED URINARY TRACT INFECTIONS

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Introduction: Urinary tract infections (UTIs) are one of the most frequent infectious diseases in children, with an increasing resistance to antimicrobials. The aims of this study were (1) to investigate the spectrum of microbial etiology and the factors that may impact on the results of urine culture; (2) to determine the change in antimicrobial susceptibility of uropathogens isolates from children with community-acquired UTIs and the clinical outcomes of patients with an isolate resistant to the antibiotic received.

Method: In this prospective study over a period of 20 years (1987.1~2006.12), we enrolled 514 pediatric inpatients whose discharged diagnosis were community-acquired UTIs and urine samples had been cultured for bacteria when hospitalized. The histories, including the medications before the urine samples were collected, the results of urine culture and the therapeutic effect were analyzed.

Result: (1)514 urine samples of which 259 (50.4%) grew bacterial isolates were analyzed. Among the total of 259 isolates, most common isolates were: Escherichia coli, accounting for 63.7% of isolates, followed by Enterococcus faecalis (16.6%), bacillus proteus(6.6%). Other bacteria, including Klebsiella pneumoniae, Pseudomonas aeruginosa and Streptococcus accounted 13.1% in total. (2)The rate of the patients who had received antibiotic before urine sample being collected among cases whose culture results were negative was higher than the rate for those with positive results.(3) Comparing the range of resistance of E. coli isolated in recent 3 years(2004~2006) to earlier 3 years (2001~2003) to following antibiotics: the resistant to Piperacillin was 75.3 vs 48.0%, to Cefotaxime was 45.2 vs 20.0% and to Ceftriaxone was 45.2 vs 16%,($P < 0.05$). (4) 21 cases in 26 whose isolated uropathogens with ESBLs positive were cured by the antibiotics with resistant in vitro tests.

Conclusion: These data show the higher level of antimicrobial resistance among the uropathogens causing community-acquired urinary tract infection in recent years. The clinical therapeutic effect may incongruity with the results in vitro tests. The positive rate of urine culture can be influence by the antibiotic treatment.

AE-188: MR UROGRAPHY IN CHILDREN: A PICTORIAL EXHIBIT

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Introduction: Magnetic Resonance (MR) urography is an evolving technique with the potential for allowing optimal noninvasive evaluation of many abnormalities of the urinary tract.

Method: We have performed MR urography in the evaluation of suspected urinary tract obstruction, hematuria, and congenital anomalies, as well as surgically altered anatomy over last 2 years.

Result: This pictorial exhibit shows the potential of MR urography in the anatomic evaluation of the urinary tract, hydronephrosis, obstructive uropathy, congenital malformations, pyelonephritis and renal scarring. MR urography was particularly useful in children because it does not use ionizing radiation. MR imaging has inherently greater soft-tissue contrast than other imaging techniques. When used in conjunction with dynamic scanning after administration of a contrast agent, it provided non-invasive analysis of the perfusion, concentration and excretion of each kidney.

Conclusion: MR urography allows a reliable assessment of renal and ureteral anatomy and of dysplastic or ectopic renal buds, even in non- or poorly functioning systems. MR urography therefore has the potential to replace the currently used excretory urography and scintigraphy.

AF-013: OUT COME OF CHILDREN OF ACUTE RENAL FAILURE WITH SEIZURE- IN A TEACHING HOSPITAL

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Introduction: Acute Renal Failure (ARP) in children is very common and associated seizure is responsible for fatal outcome. This study was conducted in the Pediatric Nephrology unit of Dhaka, Shishu (children) Hospital over a period of one year.

Method: In this study 35 children with convulsion positive ARF were compared with similar number of convulsion negative ARF children.

Result: Age of convulsion positive children were significantly lower compared to convulsion negative group (24±37 Vs 43±41 mo.). Incidence of fever, unconsciousness and septicemia were higher in convulsion positive group. Duration of ARF before referral was also much higher among convulsion positive group. Aetiologically Haemolytic Uremic syndrome and septicemia were the major causes of ARF in convulsion positive group while ARF following diarrhoea was the major cause in convulsion negative group. Hyponatremia, hypoglycemia, pyogenic meningitis & leucocytosis were the significantly important risk factors of convulsion. Significantly higher number of patient died due to ARF in convulsion positive group. Outcome of neonatal age group is worst & those who survived developed neurological sequelae.

Conclusion: Convulsion in ARF patient is more common in lower age group. Metabolic abnormalities & infections are the major cause of seizure. Mortality is higher in convulsion positive ARF. It appears that early referral & early treatment will reduce the mortality.

AF-033: RANDOMIZED CONTROLLED TRIAL COMPARING THE EFFICACY OF SEVELAMER VERSUS CALCIUM ACETATE IN CHILDREN WITH CHRONIC KIDNEY DISEASE STAGE III-IV

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Introduction: Hyperphosphatemia is associated with an increased morbidity and mortality risk in patients with chronic kidney disease. The relative effectiveness and safety of sevelamer, a mineral-free phosphate binder, for treatment of hyperphosphatemia in children with chronic kidney disease is uncertain. This study was designed to compare the efficacy and acceptability of sevelamer hydrochloride with calcium acetate as a phosphate binder in pediatric patients with chronic kidney disease.

Method: A 12-week open- label trial of sevelamer hydrochloride vs. calcium acetate was initiated in 22 patients, aged 2 to 18 years, with CKD stages III and IV. Dietary phosphate and calcium intake were assessed and baseline serum calcium, phosphate and PTH levels recorded. After a 2-week washout of phosphate binders and vitamin D, patients were randomized to receive sevelamer hydrochloride or calcium acetate.

Result: Serum phosphate decreased from 6.6 mg/dl to 5.8 mg/dl at 12-weeks in the calcium acetate group (P 0.7) and 6.2 mg/dl to 6.0 mg/dl in the sevelamer group (P 0.2). Serum calcium changed from 8.4 mg/dl to 8.5 mg/dl at 12-weeks in the calcium acetate group (P 0.2) and 9.4 mg/dl to 8.6 mg/dl in the sevelamer group (P 0.8). The serum calcium x phosphate product was not significantly different between the two groups at baseline as well as at 3-months (49.4 mg2/dl2 and 51.8 mg2/dl2 in the calcium acetate and sevelamer groups respectively (P 0.7). After adjusting baseline biochemical parameters in the two groups using ANCOVA, the serum phosphate at 12 weeks in either group did not change significantly {Calcium acetate group (Normal mean; Adjusted mean) (5.88; 5.30), Sevelamer group (6.07; 6.18) [P (adjusted means) 0.57]}, however, the serum calcium at 12 weeks was altered significantly{Calcium acetate group (Normal mean; Adjusted mean) (8.43; 9.21), Sevelamer group (9.34; 8.21) [P (adjusted means] 0.008) [Mean difference 0.8]}.The adjusted means for Ca X P product at 12 weeks were not significantly different {Calcium acetate group (Normal mean; Adjusted mean)(49.41; 47.84), Sevelamer group (51.18; 50.28) [P(adjusted means) 0.42]}. There was no significant difference between the PTH levels of the two groups at baseline or at 3-months. In the sevelamer group there was a non-significant decrease in serum bicarbonate whereas the total and LDL cholesterol significantly decreased at 12-weeks (P 0.04). Sevelamer hydrochloride was well tolerated and without adverse effects related to the drug.

Conclusion: Compared with calcium-based phosphate binders, use of sevelamer in patients with chronic kidney disease is associated with similar to slightly higher phosphate levels, similar calcium phosphate product, reduction in serum calcium levels and a significant improvement in levels of total and LDL cholesterol. CRG number CRG030600043

AF-077: REPORT OF A BOY WITH RICKETS AS A PRONOUNCED SYMPTOM OF VESICOURETERAL REFLUX

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Introduction: To find the underlying disease of persistent rickets in a boy.

Method: Analysis of the presentation and laboratory investigations.

Result: A 13 years old boy was admitted because of X shaped legs accompanied by growth stunting for 6 years. The height was 141 cm. No anemia, hypertension or leukocyturia were revealed. The serum calcium was slightly decreased to 2.06 mmol/l, the phosphate decreased to 0.7 mmol/l. The urinary phosphate was normal. Alkaline phosphatase (ALP) increased to 1449 U/l, while PTH was normal. Both proximal and distal renal tubule functions were compromised. X ray of wrist showed fraying and widening of the metaphysis, extensive cupping of the distal ends of the radius and ulna. Small sized kidneys, dilated ureters and pelves were revealed by MRI. Grade V vesicoureteral reflux (VUR) was found by micturating cystourogram examination. DTPA scan revealed GFR of 10 ml/min of both kidneys. By DMSA scan, renal scar was found on the right kidney, while the left kidney was small. No abnormality was revealed in the spinal cord on MRI. Urinary dynamic analysis showed normal function of the bladder. He was diagnosed as rickets, chronic renal failure (uremic stage), VUR (primary), and reflux nephropathy. Some uncommon points existed: 1. persistent rickets was pronounced, the reason was not very clear. Hypophosphatemia and normal serum PTH were uncommon in renal osteodystrophy; there were no risk factors for adynamic bone lesion, such as dialysis or diabetes mellitus. Normal urinary phosphate and chronic renal failure did not support familial hypophosphatemic rickets. The compromised renal tubules caused by reflux nephropathy might be the main cause. 2. Chronic renal failure without anemia and hypertension. 3. VUR without evidence of UTI.

Conclusion: For persistent rickets in younger boy, reflux nephropathy should be considered as a possible cause.

AF-083: CLINICAL USE OF VALUE LEVEL URINARY N-ACETYL-BETA-D-GLUCOSAMINIDASE ACTIVITY AS AN INDICATOR OF ONGOING TUBULAR INJURY AND NEPHROSCLEROSIS

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Introduction: Search of markers of progressing of chronic diseases of kidneys is an actual problem. N-acetyl- beta -D-glucosaminidase (NAG) is a high molecular-weight lysosomal enzyme found in many tissues of the body. This enzyme shows high activity in renal proximal tubular cells, and leaks into the tubular fluid as the ultrafiltrate passes through proximal tubules.

Method: The purpose - to study level of activity urinary NAG in depending from duration of kidneys illness. We followed 26 patients (16 males, 10 females, range in age from 3 to 16 years old) with chronic pyelonephritis. Duration of disease was from 1 till 15 years. The control group has been presented by 38 healthy children same age. Each test of urine has been investigated twice (urine pH - 7,4 and 5,5) with spectrophotometer.

Result: Level of activity urinary NAG at healthy children 0,805±0,05 su/ml or 1,089±0,243 su/ml /gr Cr. Patients with inflammatory process of a various activity degree had different parameters of activity urinary NAG. Values have made accordingly from 0,687 to 2,992 su/ml., mean - 1,462±0,162 or 1,851+0,23 su/ml /gr Cr, $p< 0,001$ in comparison with parameters of control group. Strong decrease of investigated enzyme parameters at 4 patients with a severe kidney pathology (congenital - 2 patients) and scars (0,196, 0,236, 0,223, 0, 233 su/ml, age - 7,11, 14, 15 years) was revealed. Positive strong correlation is established with urinary albumin excretion(UAE), R=0,88, p - 0, 00004. It has allowed to assume obtained data as a reflection of proximal tubular cell necrosis.

Conclusion: Activity urinary NAG, impairment NAG/UAE are a sensitive markers of progressing CRD and nephrosclerosis.

AF-124: AN UNUSUAL CAUSE OF ACUTE RENAL FAILURE – BURKITT'S LYMPHOMA

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Introduction: We report a 4-year-old boy who presented to us as hypertensive encephalopathy and acute renal failure requiring renal replacement therapy. Renal biopsy diagnosed Burkitt's lymphoma, which is rare in presenting as acute renal failure

Case report: A 4-year-old boy complained of fever without localizing sign for 1 week. He had abdominal pain and vomiting for 3 days, and noticed a gradual decreased in urine output. Physical examination showed blood pressure of 135/92 mmHg, pallor, periorbital and ankle edema. Both kidneys were enlarged and palpable down to the lower quadrates of abdomen. Blood test showed haemoglobin 10.1 g/dL, serum creatinine 312 μmol/L, urea 11.8 mmol/L, hyperuricemia (1.14 mmol/L). Creatinine clearance (CrCl) was 10 ml/min/1.73 m2. CT brain showed hypodense area involving grey-white matter in the left occipital and the left higher parietal region. Hypertension was controlled with labetalol infusion. Continuous veno-venous haemodiafiltration (CVVHDF) was commenced. Computerised tomography of abdomen showed bilateral enlarged kidneys of 13cm in length with loss of corticomedullary differentiation and poor renal perfusion, the pancreas was diffusely enlarged and there was focal bony destruction of right ilium. Renal biopsy showed that the renal cortex was diffusely infiltrated by medium size lymphoid cells with coarse chromatin, with active mitosis and karyorrhexis. The tumor cells are CD 3-, CD 20+, CD10+, Tdt- and bcl-2. Ki-67 index is 100%. The glomeruli and tubules were widely spaced out and normal looking. The diagnosis was Burkitt's lymphoma. Cerebrospinal fluid examination showed the presence of lymphoma cell. Chemotherapy of protocol HK-NHL98 including cyclophosphamide, vincristine, prednisolone and intrathecal methotrexate, hydrocortisone, cytarabine was started. CVVHDF could be withdrawn at Day 7 after commencement of chemotherapy. The renal function improved with serum Cr 78 μmo/L and CrCl of 42 ml/min/1.73 m2 at Day 10. Follow up computerized tomography of abdomen at Day 20 showed decrease in size of both kidneys and pancreas, and improvement in renal perfusion.

Discussion: Burkitt's lymphoma is a rare cause as initial presentation with acute renal failure, though acute renal failure is more commonly seen during treatment of malignancy or when there were complications like sepsis. With prompt diagnosis and commencement of therapy, the tumour regressed with significant shrinkage in the kidney sizes and normalization of renal function.

AF-166: IMPAIRMENT OF RENAL FUNCTION FOLLOWING PEDIATRIC LIVER TRANSPLANTATION AND IMMUNOSUPPRESSIVE DRUGS

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Introduction: Although pediatric liver recipients face the risk of chronic renal failure as a result of the nephrotoxicity of the commonly used immunosuppressive drugs such as cyclosporine (CyA) and tacrolimus, there are few reports that describe the incidence and progression of renal failure. We aimed to assess the incidence of renal failure in our hospital.

Method: We identified 16 patients (4 males, 12 females) who received liver transplants because of biliary atresia between 1988 and 2007. We examined their renal function, the term of using immunosuppressive drugs, and the blood concentration of CyA and tacrolimus by clinical records retrospectively.

Result: As of December 2007, 15 patients used tacrolimus, and one patient used CyA. The ages of the patients were 5.9±5.9 years (range, 2 to 20 years). The follow-up time after using immunosuppressive drugs was 3.2±5.4 years (range, 1 to 19 years). The blood concentration of tacrolimus was 3.0±2.3 ng/ml (range, 0.5 to 6.3 ng/ml). There were no patients with hypertension. However there were two patients with renal dysfunction. One was treated with tacrolimus, and the other was treated with CyA. One patient with tacrolimus had proteinuria and hematuria. Renal biopsy showed IgA nephropathy

combined with tublointerstitial lesions and hyalinizing of arterioles. We thought that using tacrolimus may cause those complications for IgA nephropathy. The other patient with CyA had proteinuria and relatively high blood trough levels of CyA (130 ng/ml).After reducing the dose, she recovered her renal function and proteinuria vanished.

Conclusion: Both of the patients with renal dysfunction have used immunosuppressive drugs for more than 15 years. The majority of other patients have used them for less than 5 years. Therefore we think that some of them will progress to renal failure in the future. We are going to follow-up having found early renal dysfunction.

AF-169: CLINICAL PROFILES OF CHILDREN WITH ACUTE RENAL FAILURE AT DR. SOETOMO HOSPITAL, SURABAYA - INDONESIA

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Introduction: Background: Acute renal failure (ARF) is characterized by an abrupt and sustained decline on glomerular filtration rate. The process is multifactorial involving alteration of haemodynamics, specific nephronal susceptibilities, intratubular obstruction and cellular metabolic fluxes. Objective: The aim of the study was to describe the characteristics of children with acute renal failure.

Method: Methods: A retrospective and descriptive study was conducted on 60 children with ARF from January 2000 to April 2008 in Dr. Soetomo Hospital, Surabaya-Indonesia. The characteristics of the patients included age, sex, aetiology and outcome were recorded.

Result: Results: A total of 60 children with acute renal failure were included, consisted of 40 boys (66.7%) and 20 girls (33.3%), with median age of 8.5 years (mean age 7.1±4.3 years). The aetiology of ARF was mainly renal disorders in 50 (83.8%) cases then followed by prerenal in 8 (13.3%) and post renal disorders in 2 (3.3%) cases. Glomerulonephritis was the major cause (58%) of ARF, followed by septicemia (16%), bilateral hydronephrosis (10%), and others (16%). Overall, 49 (81.7%) patients survived and 11 (18.3%) patients died. Of these 11 patients, 72.7% had a decrease of glomerulofiltration rate (GFR) >75%.

Conclusion: Conclusion: Acute renal failure occured mainly in school age children with glomerulonephritis as the major cause. Keywords: acute renal failure, children, characteristics

AF-173: CLINICAL FEATURES OF CHRONIC RENAL FAILURE IN CHILDREN IN THE CHILDREN HOSPITAL 1, HCM CITY

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Introduction: This study is a prospective, case study with the aim of assessing the epidemiology and the clinical features of patients with chronic renal failure (CRF) in the children hospital 1, HCM city, Vietnam

Method: Prospective, case study. All patients who were diagnosed CRF with the GFR < 50 ml/min/1.73 m2 bsa (GRF, according to Schwartz\' s formula) admitted to Nephrology department from 6/2003 to 6/2005 were selected.

Result: During 2 years, there were 41 patients (21 boys) with CRF admitted to the hospital.The mean age was 11,4 + /-3.5 year old. The mean GFR was 11.28 +/-7.21 ml/1.73 m2 bsa/min (min: 2.29;max: 30.47).The leading cause was glomerulonephritis (20 cases). The others were obstructive uropathy (4 cases), polycystics kidney disease (3 cases), renal hypoplasia (2 cases), SLE (1 case), and no cause identified (11 cases). 13 patients have known had kidney disease before. The most common symptom was edema (28 case, 68.3%), 24 cases hypertension (58.5%), 3 cases OAP (7.3%). The others symptoms were vomitting (22 cases), dyspnea (17 cases), loss of appetite (20 cases), 7 cases had blood in the vomit or in stools. Assessing of the development, 31 cases (75.6%) were malnutrition, 34 cases (82.9%) were short for ages. The mean hemoglobin concentration was 6.7 +/-2.1 g/dl, 3 cases had the platelet < 100 000/mm3. 26 cases had metabolic acidosis. The mean potassium was 5.23 +/-1.62 mEq/l (min 2.1; max 9.6 mEq/l). 34 cases had proteinuria, 24 cases had hematuria when the urine was examined. 24 cases had small kidney size in abdominal ultrasound.

Conclusion: This study provides some informations concerning CRF in children in the Children hospital 1, HCM city, Vietnam. The leading ages were the school ages (over 6 years old) 90.3%. The leading cause was glomerulonephritis. The common symptom were edema, vomitting, loss of appetite,dysnea, hypertension, anemia. 52.5% cases were ESRD.

AF-198: OUTCOME OF CHILDREN WITH ACUTE RENAL FAILURE REQUIRING RENAL REPLACEMENT THERAPY: A SINGLE CENTER EXPERIENCE

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Introduction: Acute renal failure (ARF) is a life-threatening complication in critical ill pediatric patients. Recently, renal replacement therapy (RRT) modality has shifted from peritoneal dialysis (PD) and hemodialysis (HD) to continuous veno-venous hemofiltration (CVVH).The objectives were to review etiology, treatment modalities and outcomes of children with ARF requiring RRT, and also to identify the risk factors of mortality.

Method: The medical records of 36 children diagnosed with ARF requiring RRT who were admitted between March 2004 and February 2008, were reviewed.

Result: The median age at diagnosis of ARF was 4.2 years. Eighteen were males. The main causes of ARF were hypoxic/ischemia ATN (47%) and multifactor (25%). The initiated modalities of RRT were PD 64%, HD 17% and CVVH 19%, respectively. Unfortunately, 4 cases initiated with PD received inadequate treatment which must be changed or combined with other modalities. The median duration of RRT was 7.5 days (interquatile range 3–17 days). The mortality rate at 30 days after the treatment was 64%. Multivariate analysis revealed that sepsis (p=0.033, HR 8.94, 95%CI 1.20–66.58) and duration of RRT less than 5 days (p=0.019, HR 4.27, 95% CI 1.27-14.39) were associated with mortality. According to survival curve, the probability

of survival in the first 10, 30 and 60 days after the initiation of RRT was 72, 44 and 30%, respectively. The renal statuses of 11 survivors at day 90 were completely recovery of 54.5%, chronic renal insufficiency of 27.3% and dialysis dependence of 18.2%.

Conclusion: Despite the increasing use of RRT in the tertiary care hospital, the mortality rate of children with ARF remained high. ARF was a significant co-morbid problem of critically ill patients. Sepsis was the significant risk factor of death. No specific modality was better than others.

AG-152: STUDY OF ELECTROLYTE ABNORMALITIES AMONG THE PATIENTS OF NEPHROTIC SYNDROME OF PEDIATRIC AGE GROUP FROM TRIBAL AREA OF DEVELOPING COUNTRY-INDIA

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Introduction: Despite of various homeostatic processes in living beings which regulates the concentration of various electrolytes such as sodium, potassium, calcium, phosphate, copper, zinc in different compartments of the body, the nephrotic syndrome make a child vulnerable for disturbance in electrolyte balance.

Method: 45 Nephrotic syndrome patients from tribal areas of Chhattisgarh admitted in Nephrology Unit, Department of Medicine, Pt.J.N.M.Medical College & GBG KIdney care hospital Raipur (C.G.) from april 2007 to april 2008 were studied. All patients were subjected to all routine investigations.

Result: •The mean age of the patients was 12.13 + 4.48 years. •73.3% were males & 26.7%were females. •Most common electrolyte abnormality observed was reduced copper and zinc level. •Hyponatremia was present in 60% of the patients of which 88.88% were males and 11.12% were females. •Serum calcium was <8.8 in 80% patients, of which 75% were males and 25% females. •Serum phosphate was more for respective age in 40% of the patients, of which 88.88% were males and 11.12% were females. •Electrolyte abnormalities were more common in 6-12 year of age group.

Conclusion: •Majority of patients with dyselectrolytemia were males. •Dyselectrolytemia was more prevalent in 6–12 years of age group. •Reduced copper and zinc level was the most common electrolyte abnormality. •In children with nephrotic syndrome reduced antioxidant production from the vital elements like copper and zinc are one of the factor leading to renal injury. •Among patients having lipid abnormalities, almost all patients had low copper and zinc level.

AH-053: ANALYSIS OF PERITONEAL EQUILIBRATION TEST AND VEGF GENETIC POLYMORPHISM IN KOREAN PEDIATRIC DIALYSIS PATIENTS

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Introduction: The peritoneal equilibration test (PET) is helpful in measuring peritoneal permeability in peritoneal dialysis (PD). The data reported by Warady et al. have been used as the global reference in pediatric patients. We evaluated and analyzed the peritoneal transport characteristics of Korean pediatric patients on PD. In addition, we studied the association between genetic polymorphism of vascular endothelial growth factor (VEGF) and peritoneal membrane transport.

Method: We investigated the data from 32 pediatric patients undergoing PD within 12 months at Seoul National University Children Hospital and Samsung Medical Center. PET was performed by instillation of 1,100±50 mL/m2 body surface area of a 2.5% glucose solution according to a published protocol. Results of PET were compared with the published data. We investigated the influence of the single nucleotide polymorphism (SNP) of VEGF (-2578C/A, -14978T/C, -1154G/A, -634G/C, +936C/T) on the peritoneal membrane transport.

Result: The mean 4-hour D/PCr and the mean 4-hour D/D0 glucose were 0.56±0.12 and 0.43±0.09, respectively. In comparison to the results published by Warady et al., our patients had higher D/PCr and lower D/D0 glucose. Analysis of VEGF SNP showed the haplotype CTGGC carrier was associated with higher D/P Cr and lower D/D0 glucose.

Conclusion: These results suggest that individual race or country may need their own reference values for interpretation of PET. The VEGF SNP may influence the peritoneal membrane transport. Further studies of larger scale are necessary to confirm the results.

AH-068: PERITONITIS IN CHILDREN WITH CONTINUOUS AMBULATORY PERITONEAL DIALYSIS: 5-YEAR EXPERIENCE AT DR.SOETOMO HOSPITAL SURABAYA INDONESIA

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Introduction: Peritonitis is a serious clinical problem commonly occurs in patients with end-stage renal disease (ESRD) treated by continuous ambulatory peritoneal dialysis (CAPD) that may require hospitalization, cause damage to peritoneum and death. A variety of etiologic microorganisms may cause CAPD peritonitis, mostly Gram-positive organisms. Many different antimicrobial agents, intraperitoneally or intravenously, have been used to treat CAPD peritonitis. The objective of this paper was to study the incidence, etiology and outcomes of peritonitis episodes in children with CAPD.

Method: A retrospective and descriptive study was carried out on 15 patients who had been treated by CAPD between 1 March 2003 and 29 February 2008 in Dr.Soetomo Hospital Surabaya Indonesia. Peritonitis diagnosis was based on at least 2 of the following: abdominal pain, cloudy PD effluent, white blood cell (WBC) count in the effluent at least 100/μL and positive culture of effluent.

Result: Mean age at onset of CAPD patients was 8.9±3.5 years, most were boys (73.3%) and only 4 (26.7%) were girls. During the period of 5 years, 9 (60.0%) out of 15 ESRD patients with CAPD remained peritonitis-free. There were 10 episodes of peritonitis among 6 patients in 185 patient-months, which equates to an overall peritonitis rate of 1

episode every 18.5 months (0.6 episodes/year at risk). All patients experienced abdominal pain and had cloudy effluents. Most of dialysat fluid cultures were negative (44.4%) with Staphylococcus coagulase negative as the most frequent microorganism isolated (22.2%) and treated with intraperitoneal Cefazoline, followed by Staphylococcus aureus in 1 patient treated with intravenous Ceftriaxone and yeast cell in 1 patient treated with intraperitoneal Fluconazole. Cure occurred in 70.0% of episodes while death occurred in 30.0% of episodes.

Conclusion: Peritonitis rate in our population is acceptable although there are several areas for improvement. Its etiologies and outcomes follow the international descriptions.

AH-089: COOPERATION OF CONTINUOUS RENAL REPLACEMENT THERAPY WITH EXTRACORPOREAL MEMBRANE OXYGENATION IN CRITICALLY ILL PATIENTS

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Introduction: Extracorporeal membrane oxygenation(ECMO) has been proved successful in the treatment of refractory cardiac or respiratory insufficiency. However, acute renal failure in such patients is frequently encountered and then continuous arterial-venous hemofiltration(CAVH), continuous veno-venous hemofiltration(CVVH) or continuous veno-veno hemodialysis(CVVHD) may be beneficial. Due to the experience of cooperation of ECMO with CRRT is limited in Taiwan. We report the frequency of usage, and outcomes in children treated with one unit providing ECMO and CAVH, CVVH or CVVHD.

Method: Between February 2007 and December 2007, five critically ill patients(3 male, 2 female), aged from 4 moth to 23 year-old, were treated with ECMO in our pediatric intensive care unit due to the cardiac and/or respiratory failure. All of them were complicated with acute renal injury and/or fluid overload. According to RIFLE classification, they were all classified as RIFLE class F and continuous renal replacement therapy was applied immediately. Of the five patients, three patients, who was diagnosed as ARDS with pulmonary hemorrhage, cardiac temponade and cardiogenic shock respectively, underwent CVVHD. One, who was diagnosed as pneumonia with respiratory failure, underwent CVVH and one, who was diagnosed as dilated cardiomyopathy with left heart failure, underwent both CVVH and CAVHD.

Result: The mean duration of CRRT is 6 days(range 2 days to 11 days). The mean blood urea nitrogen before CRRT is 43 mg/dL (range 29 to 63 mg/dL) and the mean serum creatinine before CRRT is 2.54 mg/dL(range 1.3 to 3.8 mg/dL). After CRRT, the mean blood urea nitrogen fell to 21.2 mg/dL (range 5 to 34 mg/dL) and the mean serum creatinine fell to 1.4 mg/dL (range 0.4 to 2.3 mg/dL). Three patients died eventually due to irreversible cardiac and/or respiratory failure and two patients survived(one male, one female).

Conclusion: Performing CAVH, CVVH and CVVHD in critical-ill children who underwent ECMO therapy with large age and weight ranges still poses significant clinical and technical challenges. Our experience demonstrates the effectiveness of CRRT in the restoration of renal function during ECMO, but there is still need to define a more practical protocol for cooperation of CRRT with ECMO and to avoid possible complications.

AH-102: RELAPSING SEVERE ANEMIA DUE TO PARVOVIRUS B19 INFECTION AFTER RENAL TRANSPLANTATION IN CHILDREN: A CASE REPORT

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Introduction: Few reports describe severe anemia due to Parvovirus B19 (PB19) infection after renal transplantation in children. Intravenous immunoglobulin (IVIg) is successfully used to treat PB19-related anemia; however, several reports have shown PB19-related anemia can recur despite IVIg therapy.

Method: We studied a case of PB19 infection in a renal transplantation child from our center.

Result: A 16-year-old boy with ESRD due to IgA nephropathy received a cadaveric renal transplantation. Initial immunosuppression consisted prednisolone, mycophenolate-mofetil, tacrolimus and basiliximab. Seven weeks after transplantation, he experienced acute rejection (Banff grade III), which was successfully treated with high dose steroids, ATG and plasmapheresis. Subsequently, his hemoglobin dropped to 54 g/l and bone marrow biopsy showed hypoplasia of red cells. He was treated with blood transfusion, EPO, Vitamin B12 and folate. However, his hemoglobin ranged from 55 to 65 g/l. Further investigation revealed positive PB19 IgM-antibodies in serum. After therapy with IVIg (0.4 g/kg/d for 4 days) and conversion from mycophenolate-mofetil to rapamycin, anemia resolved. But two months after treatment with IVIg, anemia recurred and PB19 IgM-antibodies was still positive in serum. A second course of IVIg (0.2 g/kg/d for 10 days) was given, which was again followed by the resolution of anemia and recurrence. Subsequently, one course a month of IVIg (0.4 g/kg/d for 5 days) was repeated four times and bigeminal immunosuppressants with low doses including prednisolone and tacrolimus were used. Within the next seven months, anemia did not recur and serum PB19 IgM-antibodies kept negative. Renal function remained stable, with an actual serum creatinine of 93 μmol/l (eGFR 108 ml/min/1.73 m2) and no proteinuria.

Conclusion: Although rare, PB19 infection should be considered in differential diagnosis of anemia after renal transplantation. IvIg is useful to control PB19-related anemia. Furthermore, baseline immunosuppression should be carefully adjusted to control PB19 infection.

AH-132: THE IMPACT OF PMX-DHP TREATMENT IN AN INFANT WITH NON-ENDOTOXIN-RELATED SEPTIC SHOCK AND MOF

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Introduction: Endotoxin hemoabsorption therapy with a polymyxin B-immobilized column (PMX-DHP) has been used to treat mostly adult

patients with severe sepsis or septic shock with Gram-negative bacterial infection. The mechanism has been thought to reduce circulating endotoxin.

Method: Case report: The patient was a 2-year-old boy and was admitted to our hospital under the diagnosis of septic shock and MOF including acute kidney failure. He had neuroblastoma stage IV with matastases to liver, bone and bone marrow. His pediatric logistic organ dysfunction (PELOD) score was forty-two points, which means his predicted death rate was 99.1%, on admission. The value of endotoxin was within normal range and the level of inflammatory cytokines, endogenous cannabinoides and HMGB-1 were significantly elevated, before PMX-DHP. After admission, he was immediately treated with endotoxin absorption thrapy by PMX column. Significant improvement of hypotension and increase in urine output were observed right after PMX-DHP treatment. The value of inflammatory cytokines, endogenous cannabinoides and HMGB-1 decreased after PMX-DHP treatment, subsequently.

Result: Discussion: Several studies have demonstrated that PMX-DHP therapy was effective for septic shock without endotoxemia. On the other hand, the role of paracrine mediators of hypotension such as anandamide (ANA), 2-arachidonyl glyceride (2-AG) and HMGB-1 has been emphasized in recent investigations in septic shock. We successfully treated an infant case of severe septic shock and MOF without endotoxemia, using PMX-DHP. In this case, PMX-DHP was effective in reducing serum levels of the inflammatory cytokines as well as ANA, 2-AG and HMGB-1, which might lead to Significant improvement of hypotension, urine output and the PELOD score.

Conclusion: This case report suggests that PMX-DHP is an effective treatment for childhood septic shock and MOF without endotoxemia. The mechanism of PMX-DHP for septic shock seemed to be a reduction of ANA and 2-AG, causative mediators of cytokine storm.

AH-159: PERITONEAL DIALYSIS ASSOCIATED PERITONITIS IN KOREAN CHILDREN WITH CHRONIC RENAL FAILURE

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Introduction: The purpose of this study is to present our experience of peritoneal dialysis associated peritonitis in Korean children and to propose guidelines for empirical treatment by the recent regional data of organisms and their sensitivity to antibiotics

Method: We have collected data on pediatric patients from the Korean Pediatric CRF Registry between January 1, 2007, and April 30, 2008.

Result: Of the 108 patients, peritonitis was occurred in 24 patients(32 episodes). The rate of peritonitis was one episode over 41.88 patient-months. Gram-positive organism accounted for 71.4% of all episodes. Of the gram-positive organism, most common pathogen was Staphylococcus aureus (20%), next common pathogens were coagulase negative Staphylococcus(15%), Staphylococcus epidermidis (15%) and Streptococcus viridans(15%). 85% of the gram-positive pathogens was sensitive to 1st generation cephalosporin(cephalothin). However, in patients younger than 4 years old, 67% of gram-positive pathogens was resistant to 1st generation cephalosporin. Of the gram-negative organism, most common pathogens were Acinetobacter baumanii(28.6%) and Serratia marcescens(28.6%). All of the gram-negative pathogens were sensitive to ceftazidime. 88.5% of all pathogens were sensitive to cephalothin or ceftazidime. The rate of peritonitis was higher in continuous ambulatory peritoneal dialysis patients(one episode over 35.28 patient-month) than in automated peritoneal dialysis patients(one episode over 50.36 patient-month) ($P<0.05$).

Conclusion: The empirical therapy with 1st generation cephalosporine and ceftazidime can be also effective to peritoneal dialysis associated peritonitis in Korean children. But, in patient younger than 4 years old, glycopeptides(vancomycin or teicoplanin) should be considered as the first empirical therapy in Korean children.

AH-175: PREVALENCE OF HYPERTENSION IN POST-TRANSPLANT PAEDIATRIC PATIENTS

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Introduction: To investigate the prevalence of hypertension and the blood pressure (BP) pattern in children after renal transplantation using casual blood pressure and ambulatory blood pressure monitoring (ABPM), and to evaluate the control of hypertension in this paediatric population.

Method: This is a cross-sectional retrospective study. Patients with functioning renal graft and age less than 20 years old were included. ABPM was done by using a SpaceLabs 90121 oscillometric device during routine yearly assessment. The daytime, night time pattern and day-to-night BP variation were measured. The BP load measured was stratified into 3 grades: <35%; 35–50% and >50%. The mean 24-hour ABPM results are correlated with the casual BP during routine follow-up.

Result: A total of 20 patients were included in the study. The male-to-female ratio was 1:1. The mean age of patients was 15.1+/-4.1 years and ABPM was performed on average 3.4+/-2.1 years after transplantation. The mean GFR by Schwartz estimation was 73.3 ml/min/1.73 m2. Overall, 9 patients (45%) had nocturnal hypertension, 16 (80%) lost the nocturnal dip phenomenon and 2 of them had a reversed pattern. Ten patients required anti-hypertensive drugs, and calcium-channel-blocker was most commonly prescribed. One patient had a systolic BP load of 35–50%, four had a diastolic BP load of 35–50% and two had a diastolic BP load of >50%. Despite treatment, 3 patients were still hypertensive by casual BP measurement and all 3 were also shown to be hypertensive by ABPM (PPV: 100%). For those 17 patients with a normal casual BP, 16 of them had a normal ABPM (NPV: 94%). One patient was detected hypertensive by ABPM but was missed by casual BP.

Conclusion: Hypertension is prevalent among paediatric patients with renal transplantation. ABPM is useful in detecting hypertension, in particular nocturnal hypertension. More emphasis should be put on control of diastolic blood pressure in this group of patients

AH-194: PEDIATRIC RENAL TRANSPLANTATION: A SINGLE-CENTER EXPERIENCE IN NORTHEAST THAILAND

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Introduction: Renal transplantation is the preferred therapy for children with end-stage renal disease (ESRD).

Method: We reviewed the medical records of children with ESRD who received renal transplantation at Srinagarind Hospital, Khon Kaen, between August 15, 2001 and May 12, 2008.

Result: Eight male and seven female patients were identified. The mean age was 12.7±3.2 years (range, 5.0–17.6). The major cause of ESRD was a congenital anomaly of the kidneys (53%). All of the children received cadaveric transplantations and none received induction therapy. Triple immunosuppressive drugs comprising cyclosporine A, prednisolone and mycophenolate mofetil were administered to 12 patients. Tacrolimus instead of cyclosporine A was given to three patients who had received a renal transplant since January 2008. The median follow-up time was 13 months (1 to 80 months). The most frequent complication was urinary tract infection (40%). Acute graft loss was found in one patient (6.7%) due to graft infarction. Other complications included herpes viral infection (20%), chronic rejection (20%), acute rejection (13.3%), gum hypertrophy (13.3%), myopathy (6.7%), lymphocele (6.7%) and transitional cell carcinoma of the bladder (6.7%). Cyclosporine A was changed to tacrolimus in five patients diagnosed with gum hypertrophy (2 patients), chronic rejection (2 patients) and myopathy with chronic rejection (1 patient). Two patients returned to dialysis due to graft infarction and chronic rejection, respectively. Mean serum creatinine at last follow-up of the remaining cases was 1.2±0.5 mg/dL (0.6–2.3 mg/dL).

Conclusion: Our study demonstrates the potential for successful outcomes of pediatric renal transplantation in this resource-limited area if complications can be reduced and long-term outcomes improved.

AH-201: EPSTEIN-BARR VIRUS (EBV) INFECTION IS A RISK FACTOR FOR POST-TRANSPLANT LYMPHOPROLIFERATIVE DISORDER IN PEDIATRIC RENAL TRANSPLANT RECIPIENTS

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Introduction: Post-transplant lymphoproliferative disorder (PTLD) is a rare but potentially fatal complication post-solid organ transplantation. This study examined the incidence and course of EBV-associated PTLD in pediatric renal transplant recipients.

Method: A total of 43 pediatric renal allograft recipients transplanted at the University Children's Medical Institute from 1989-2008, were retrospectively studied.

Result: The incidence of PTLD was 9.3% (4 cases) with median onset of 2 years (4.8 months–12 years) post-transplant. Etiology of renal failure in the 4 patients included systemic lupus erythematosus, focal segmental glomerulosclerosis, chronic glomerulonephritis and juvenile nephronophthisis. Three patients had received IL-2 receptor monoclonal antibodies for induction, while 1 had anti-thymocyte globulin. Pre-transplant EBV serological testing of 32 recipients in our cohort showed that 10 (31%) were sero-negative and 8 of these developed positive EBV DNA titres on post-transplant surveillance. Clinical presentation of PTLD included rapidly progressive pneumonitis, gastro-intestinal bleed, sub-mandibular lymphadenopathy and infectious mononucleosis syndrome with adenotonsillar hypertrophy and airway obstruction. Histopathological diagnosis revealed B-cell PTLD in 3 and T-cell-rich Hodgkin-like disease in 1 patient. The 2 patients with relatively severe PTLD had been exposed to immunosuppression including alkylating agents for a mean period of 10.2 years prior to transplant. Treatment consisted of reduced immunosuppression, intravenous immunoglobulin and ganciclovir/acyclovir in all patients. Two patients received rituximab therapy, one received conventional chemotherapy and two were treated with resection of primary PTLD. One patient died shortly after diagnosis secondary to chemotherapy. At a median follow-up of 1.5 years, the other 3 patients had normal functioning grafts.

Conclusion: Pediatric renal transplant patients who are EBV sero-negative pre-transplant are at high risk of PTLD. Post-transplant surveillance for EBV DNA may further assist in identifying high risk patients so that early treatment can be instituted. The role of pre-transplant exposure to immunosuppression especially alkylating agents needs to be further defined.

AH-207: PEDIATRIC KIDNEY TRANSPLANTATION: TWELVE YEARS OF EXPERIENCE

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Objective: To describe the outcome of kidney transplantation in children

Method: We retrospectively reviewed a total of pediatric patients with end stage renal disease who underwent kidney transplantation at Siriraj hospital from May 1996 to April 2008.

Results: A total number was 19 patients who were 14 males and 5 females. Mean age at the transplant was 11.2±3.0 (range, 6.7–16.5) years. Mean duration after transplant was 4.3±3.3 (range, 0.1-11.7) years. Of all donors, 5 were cadaveric and 14 were living-related. Greater than 3 HLA mismatches were found in 3 pairs. There were 2 HLA-DR mismatches in 2 pairs and 1 HLA-DR mismatch in 14 pairs. Six pairs had positive CMV serology in donors but negative in recipients. Ten were positive in both donors and recipients. Only 2 patients received induction antibody. A combination of steroid and other two immunosuppressants was used in all patients. Initially, cyclosporin A was given in 13 and tacrolimus in 5 patients. Azathioprine was given in 7 and mycophenolate in 11 patients. 12% developed graft rejection and 17.5% had a glomerular filtration rate less than 60 mL/min/1.73 m2 during the first year after transplant. Serum creatinine seemed to maintain at a stable level after 2 years of transplant. 5-year patient survival and graft survival rate was 86% and 100%, respectively.

Conclusion: Our outcomes of renal transplant in children are comparable to those in developed countries.

AI-044: CLINICO-PATHOLOGICAL ANALYSIS OF 24 BOYS WITH LUPUS NEPHRITIS

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Introduction: Lupus nephritis(LN) is not a few in childhood, especially in adolescent girls.But there are different conditions in boy. So we analyzed the clinico-pathological characters of boy with LN.

Method: Retrospective study was taken by analyzing the data of clinical, renal pathology and follow-up survey of 24 boys with LN.

Result: All boys suffered from LN at the average age of 11.2±1.9 years old. The course of disease before diagnosis was 9.3±24.8 months(range from 1 to120 months). 4 cases of them were misdiagnosised for a long time as HSPN, NS or AGN. Among our boys, kidney damage was the first manifestation in ten cases(41.7%), and 2 cases(8.3%) only had LN. 23 cases(95.8%) were accompanied with proteinuria(24 h-Upro eduction ranged from 0.87 g to 16.98 g). 5 of 23 cases(20.8%) had decling renal function. Nephrotic syndrome was the main clinical types (45.8%). Renal biopsies were performed in 19 cases. Class IVLN (57. 9%) was the most frequent pathological findings, and "full house" was found in 68.4% of biopsies. All patients were taken by the treatment of steroid and immunosuppressive agents. Sixteen boys obtained clinical remission after average 3.4 months, and the remission rate was 66.7%. Six cases (25%) relapsed during follow–up survey, and two cases suffered from renal failure.

Conclusion: In boy's LN, nephrotic syndrome was the main clinical manifestation, and class IVwas the main pathology type. Lower morbidity and atypical clinical symptom were main reasons of misdiagnosis and missed diagnosis. The key point of improvement the prognosis of boy's LN was early diagnosis, regular therapia and follow-up survey.

AI-062: INTRAVENOUS DEXAMETHASONE FOLLOWED BY ORAL PREDNISOLONE VERSUS ORAL PREDNISOLONE IN THE TREATMENT OF CHILDHOOD HENOCH-SCHÖNLEIN PURPURA

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Introduction: The aim of this study was to evaluate the effectiveness of intravenous corticosteroid therapy when Henoch-Schönlein purpura (HSP) patients are unable to tolerate oral medications due to abdominal pain.

Method: We retrospectively analyzed 111 children with a diagnosis of HSP (Mean age 6.9±2.3 years, Male: Female=55:56) from the years 2000 to 2003. One hundred and eleven children with HSP were divided into the two groups; 49 patients who were treated with only oral prednisolone (PL group) and 62 patients with oral prednisolone after intravenous dexamethasone (Dexa+PL group).

Result: Palpable purpura was seen in all 111 patients (100%), abdominal pain in 55 (50%), and arthralgia in 65 (59%). There were no significant differences in the incidence of arthralgia between the two groups, but Dexa+PL group had significantly longer duration of fasting than PL group (0.7±1.2 vs. 0.02± 0.1 days, P<0.01) due to more severe and frequent abdominal pain (68% vs. 27%, P<0.01). Serum albumin levels were significantly lower in Dexa+PL group than in PL group (4.0± 0.4 vs. 4.3±0.4, P<0.01). However, the development of nephritis (21% vs. 32%, P=0.98), the number of relapse (4% vs. 11%, P= 0.167) and persistent nephritis at last follow-up (12% vs. 16%, P =0.563) were not much different between the two groups.

Conclusion: Intravenous dexamethasone followed by oral prednisolone may be a useful and effective therapeutic strategy in HSP children who cannot tolerate oral medications due to severe gastrointestinal manifestations such as severe abdominal pain or melena.

AI-064: COMPARISON OF CLINICAL OUTCOME ACCORDING TO THE DURATION OF CORTICOSTEROID THERAPHY IN CHILDHOOD HENOCH-SCHÖNLEIN PURPURA: A BICENTRIC STUDY

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Introduction: The aim of this study was to evaluate whether the incidence of relapse or nephritis might be influenced by the duration of corticosteroid therapy in children with Henoch-Schönlein purpura (HSP).

Method: We retrospectively analyzed 186 children with a diagnosis of HSP (Mean age 6.3±2.5 years, Male: Female=104:82) in two major hospitals in Ilsan, Korea from the years 2000 to 2007. One pediatric nephrologist from hospital A has used oral prednisolone in patients with mild symptoms and intravenous dexamethasone followed by oral prednisolone in those with severe symptoms (Group A, N=94). The other from hospital B has prescribed nothing in patients with mild symptoms and oral prednisolone in those with severe symptoms (Group B, N=92).

Result: There were no significant differences in age, sex, body weight, white blood cell count, hemoglobin, hematocrit, platelet count, serum protein and albumin levels between the two groups. The incidence of abdominal pain (45% vs. 50%, p=0.557) or arthralgia (61% vs. 61%, p=1.0) also did not differ between the two groups. However, the duration of steroid therapy was significantly longer in Group A than in Group B (21.8±8.4 vs. 6.1±7.4 days, p<0.0001) and the cumulative dose of prednisolone was also higher in Group A than in Group B (404.4±248.2 vs. 105.1±137.4 mg, p<0.0001). The development of nephritis was more frequent in Group A (23% vs. 10%, p=0.017), while the incidence of relapse was higher in Group B (10% vs. 23%, p= 0.017).

Conclusion: The longer duration of steroid use was associated with the lower incidence of relapse, but the higher rate of nephritis development. Therefore, corticosteroids should be used carefully in a selected group of HSP children, and be tapered rapidly after acute symptoms are controlled.

AI-082: GOOD OUTCOMES WITH MYCOPHENOLATE BASED INDUCTION PROTOCOL IN CHILDREN WITH PROLIFERATIVE LUPUS NEPHRITIS

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Introduction: Proliferative lupus nephritis (LN) is associated with poor renal outcome in children. Cyclophosphamide remains the gold standard for inducing remission, although infection, infertility and an association with secondary malignancy remain significant concerns with this therapy. This study examined the outcomes of children with proliferative lupus nephritis using a new mycophenolate (MMF)-based protocol comprising pulse intravenous methylprednisolone, MMF +/- cyclosporine for induction.

Method: Sixteen children with proliferative LN, WHO class III and IV (age range at start of treatment 3.7–14.8 years) who were treated between the years 1995 to 2007 were included in this retrospective study. MMF dose was 1200 mg/m2/day. We compared the clinical and laboratory parameters pre-induction, at 6 months and at 1 year. Treatment outcome was defined by Systemic Lupus Erythematosus Disease Activity Index (SLEDAI), renal function, proteinuria and serologic markers (complement C3, C4 and anti-dsDNA). Statistical analysis was performed using Wilcoxon signed rank test.

Result: At presentation, 50% had nephrotic syndrome, 31.2% had nephritic-nephrotic syndrome, while 25% had renal failure requiring dialysis. Renal biopsy classification (WHO) was IV in 68.8% and III in 31.2%. Comparing clinical and laboratory parameters at induction, 6 months and 1 year, respectively, SLEDAI (25.38±8.72 vs 1.45± 2.38 vs 2.43±2.62), serum complement C3 (46.55±21.05 vs 107.02± 27.42 vs 109.79±24.42 mg/dL), serum complement C4 (12.48±13.8 vs 23.03±14.33 vs 21.95±11.41 mg/dl) and urine protein (6.97±7.09 vs 0.98±1.56 vs 0.21±0.15 g/d/1.73 m2) all improved significantly ($p<0.03$). Anti-dsDNA positivity also improved from 93.8% (15/16) to 31.2% (5/16) ($p<0.001$). Additionally renal function normalized by 6 months in 66.7% (6/9) of patients with renal failure at onset, while 88.9% (8/9) had an improved estimated glomerular filtration rate (eGFR) of more than 60 ml/min/1.73 m2.

Conclusion: Combination MMF protocol resulted in significant clinical and serological improvement in patients with proliferative LN and can be used as an effective therapeutic alternative for induction.

AI-097: THE ROLE OF VASCULAR ADHENSION MOLECULE-1 AND INTERCELLULAR ADHENSION MOLECULE-1 IN CHILDREN WITH SLE

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Introduction: Former research has showed that over-expression of prohibitin (PHB) suppresses renal interstitial fibroblasts proliferation. ICAM-1 and VCAM-1 take part in the pathogenesis and progression of SLE. We sought to investigate ICAM-1 and VCAM-1in peripheral blood and the expression of ICAM-1and PHB in renal tissues of SLE children, and to explore the relationships between adhension molecules and active state of SLE.

Method: Soluble ICAM-1 (s-ICAM-1) and soluble VCAM-1(s-VCAM-1) were detected by using ELISA. CD54 (ICAM-1) and CD106 (VCAM-1) on peripheral blood mononuclear cells were detected by using flow cytometry. Expression of ICAM-1 and PHB in renal tissues were detected by using immunohistochemistry respectively.

Result: There was no significant difference between control and SLE group in the concentration of s-ICAM-1 and s- VCAM-1. While the concentration of s-ICAM-1 ((3.71+/-1.59)×105 pg/ml) in active state group (SLEDAI>9) of SLE was significantly higher than that (2.49+/ -1.04×105 pg/ml) in inactive group ($P<0.05$). CD106+ T cells were significantly higher in SLE group (1.05%±2.00%) than that (0.24%± 0.21%) in control group ($P<0.05$). The level of s-ICAM-1, s- VCAM-1, CD54 + and CD106 + T cells had no significant difference between renal injury and non-renal injury group. Correlation analysis showed that s-ICAM-1 had positive correlation with SLEDA I($r=0.393$, $P< 0.05$). Positive areas percentage of PHB protein (7.02%+/-4.91%) in renal tissues of lightly tubulointerstitial injury group were significantly higher than those (2.32%+/-1.27%) in seriously injury group ($P< 0.05$), while there was no significant difference of ICAM-1 expression in renal tissues between the two groups. Correlation analysis showed that the expression of ICAM-1 had positive correlation with PHB in SLE children($r=0.616$, $P<0.05$).

Conclusion: s-ICAM-1 might be a predictor to reflect the active states of SLE children. ICAM-1 and PHB may be another early monitoring factors in renal injury of SLE children.

AI-107: ANALYSIS OF FACTORS INFLUENCING THE PROGNOSIS OF CHILDREN WITH LUPUS NEPHRITIS IN SHANGHAI

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Introduction: To explore the factors affecting the long-term survival of children with lupus nephritis.

Method: The data of 101 patients who diagnosed as lupus nephritis from Jan. 1996 to Jun. 2007 in our hospital were investigated retrospectively. According to different outcomes, patients were divided into remission group (complete remission and partial remission) and ineffective group (nonresponse and death); according to different recipes in induction treatment, divided into cyclophosphamide (CTX) group, mycophenolate mofetil (MMF) group and other drugs group; according to whether receive renal biopsy and pathological classification, divided into type I and II group, type III and IV group, type V group and no biopsy group. Data was analyzed by univariate $\chi 2$ test and multivariate logistic regression model using SPSS 11.0.

Result: Univariate analysis showed that the following 4 variables were correlated to prognosis: different pathological types ($P=0.005$), heavy proteinuria at onset($P=0.003$), different recipes ($P=0.001$) and irregular treat ($P=0.000$). In multivariate analysis, it was confirmed that irregular treatment ($OR=9.955$) was significantly associated with treated outcome.

Conclusion: For all SLE patients with evidence of nephritis, we should do our possible to perform renal biopsy, in order to confirm pathological type, guide treatment and estimate prognosis. Regular

treatment plays an important role in prognosis of children with lupus nephritis.

AI-202: STUDY OF RENAL ABNORMALITIES IN RELATION TO CD4 LEVEL IN HIV POSITIVE PATIENTS OF PEDIATRIC AGE GROUP FROM TRIBAL AREA OF DEVELOPING COUNTRY-CENTRAL INDIA

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Introduction: Renal complications occur frequently in the course of HIV disease. Many of these complications are preventable & treatable, making early recognition & intervention essential.

Method: 200 HIV positive patients admitted/attending OPD were studied of which 20 were of pediatric age group.

Result: •Males were 60% & females were 40%. •Low CD4 percent (<25) was found in 75% of patients. •In patients with CD4 percent <25, Males were 53.33% while females were 46.67%. •90% of patients had proteinuria(>200 mg/24hours). •Among the patients with CD4 percent <25, 93.33% had proteinuria (>200 mg/24hours) while it was present in 60% of patients with CD4 percent>25. •95% patients had Hemoglobin(Hb) level <11 gm%. •Hb was <11 gm% in all the patients with CD4 percent <25. •In the patients with low CD4 percent (<25) with proteinuria, 35.71% had malnutrition (Mid arm circumference<13.5 cm) & among the patients with CD4 percent >25, 20% had malnutrition. •Among the patients with duration of illness >6months, 88.88% had low CD4 percent(<25) with renal insufficiency, while it was present in 63.63% of patients with duration of illness <6 months. •Tuberculosis infection was present in 15% of patients with low CD4 percent(<25) with renal injury.

Conclusion: •HIV infection with proteinuria was more common in male children. •Low CD4 percent(<25) with renal injury was more common in males as compared to females. •Most of the children with low CD4 percent had significant proteinuria. •Hb level was significantly low in patients with low CD4 percent & renal insufficiency. •Low CD4 percent with proteinuria had significant Hyperlipidemia. •Malnutrition was more common in patients with low CD4 percent & proteinuria. •Duration of illness had negative relationship with CD4 percent with renal insufficiency. •Tuberculosis infection was much more common in low CD4 percent with renal injury.

AJ-006: SEVEN YEAR FOLLOW UP OF MICROSCOPIC POLYANGIITIS PROGRESSED TO CHRONIC SCLEROSING GLOMERULONEPHRITIS: A CASE REPORT

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Introduction: Microscopic polyangiitis (MPA) is a systemic necrotizing vasculitis that involves many organ systems including the skin, joint, kidneys, and lungs. The diagnosis of MPA is often difficult to make, and delayed because of the variability of the clinical presentation. Also in spite of early diagnosis and intensive care, 5 year renal survival rate is 55%, and 5 year patient survival rate is 65%.

Method: We experienced a case in 7-year-old girl of microscopic polyangiitis with positive serum perinuclear antineutrophil cytoplasmaic autoantibodies (P-ANCA), associated with pulmonary hemorrhage and gastrointestinal bleeding. The patient was managed under intensive care and has been following up since March 2001.

Result: The diagnosis of patient's first renal biopsy was MPA, P-ANCA associated crescentric glomerulonephritis. The patient's second renal biopsy was done 5 years 6 months later since first renal biopsy, and pathologic diagnosis was chronic sclerosing glomerulonephritis, advanced, due to MPA. We began methylprednisolone pulse therapy, combined with a low dose of cyclophosphamide and plasmapheresis therapy in 2001, and ACE inhibitor, Angiotensin II receptor blocker, cyclophosphamide were used until now and the patient's current age is 14 years old. The patient's laboratory findings showed BUN 117 mg/dL/Cr 2.3 mg/dL at initiation of disease, BUN 30.5 mg/dL/Cr 1.6 mg/dL in 2004, BUN 30.7 mg/dL/Cr 1.6 mg/dL in 2006. But renal function was progressed to chronic failure with latest laboratory data BUN 51.7 mg/dL and Cr 3.2 mg/dL.

Conclusion: We report a case of long-term follow-up and survival for 7 years in microscopic polyangiitis with chronic sclerosing glomerulonephritis. ACE inhibitor, Angiotensin II receptor blocker and small dose of immunosuppressant with close observation is the key to maintain the patient survival.

AK-019: NOCTURNAL ENURESIS IN MALAYSIA – PARENTS' PERCEPTION AND REMEDIES.

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Introduction: Childhood enuresis is a common condition all over the world yet Malaysian parents rarely discuss it with their doctors. To understand the reasons for this, we interviewed parents of children who came for consultation.

Method: A questionnaire was designed to determine parents': (i) views on the cause of their child's bedwetting, (ii) previous coping strategies, (iii) reasons for seeking medical consultation and (iv) perception of the impact of enuresis on their child

Result: Thirty-two parents of children aged between 7 and 16 years attending the Enuresis Clinic consented to this study. Ninety-two percent of the children had more than 3 wet nights per week yet only 44% had ever seen a doctor. Deep sleep was thought to be a cause in 81% and "drinking too much" in 47% whilst genetics/familial in 19%. Only 6% thought their child would "outgrow" the problem. All parents had tried at least one of the 10 coping strategies itemized. The three most common practices were restricting pre-bedtime fluids (81%), waking the child to void (97%) and "lifting" the child (62%). Diapers were used in 41% and punishment in 25%. Reasons for seeking consultation included fear of the effects on the child (97%), fear of an underlying medical problem (65%), restriction of the child's social activities (56%), the child's embarrassment (59%) and tired of washing sheets (44%). Parents' perceived impact on the child included low self esteem (77%) and social stigma (63%).

Conclusion: Despite less than half the families seeking medical consultation for this self-limiting disease, children with nocturnal

enuresis in Malaysia and their families are clearly distressed by the problem and its psychosocial consequences, in particular the social stigma and perceived low self esteem. It is time that medical practitioners address these issues in a bedwetting child and offer appropriate therapy.

AK-051: OFF-LABEL USES OF DRUGS IN JAPANESE CHILDREN WITH KIDNEY DISEASES

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Introduction: Our ability to provide safe and effective drug therapy for infants and children is often hampered by a lack of dosing information derived from careful, well-controlled trials and the limited database available concerning drug safety in this patient population. This problem was first articulated 40 years ago that infants and children were becoming "therapeutic orphans". It is important to note that many of such medicines are frequently used off-label in relation to dose, indication, age, and route of administration in infants and children. We approach the problem of off-label uses of drugs in Japanese children with kidney diseases.

Method: We reviewed 90 drugs for children with kidney diseases derived from four Japanese references for pediatric nephrologists and revealed off-label uses of drugs in relation to dose, indication, age, and route of administration.

Result: There were 6 of 90 drugs with no product license in Japan but 2 drugs with product license in US or EU. 44 drugs (49%) had lack of pediatric drug formulations. 11 drugs (12%) were not licensed use in children with the kidney diseases but were licensed use in children with other diseases. 69 drugs (77%) were not existing dosing information for pediatric patients. 59 drugs (66%) were not existing explicit statements that the safety and efficacy in children.

Conclusion: Most drugs of the Japanese physicians' references entries have either no existing dosing information for pediatric patients or explicit statements that the safety and efficacy in Japanese children with kidney diseases. The progress made in the solution to the therapeutic orphan problem during 30 years in US and EU. We work for a general level of comfort with pediatric clinical trials and undertake the problem of off-label uses in Japanese children with kidney diseases.

AK-078: SUBCAPSULAR URINOMA WITH GLOMERULONEPHRITIS IN A GIRL

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Introduction: To make a diagnosis of a girl with kidney subcapsular hydrops, abnormal urine analysis and hypertension.

Method: Physical examination and laboratory investigations were analyzed.

Result: A 5 years old girl was admitted because of kidney subcapsular hydrops, proteinuria without edema, and hypertension (130/94 mmHg) for 50 days. No trauma or familial hypertension history were provided. No hydrops was found before the age of 1 year. Urinary RBCs were 5-7/Hp and protein was 77 mg/kg/24 hr. The serum albumin was normal. Ultrasound examination revealed normal sized kidneys, increased echogenicity in both kidneys, and subcapsular hydrops on the upper pole of the right kidney connected with an old renal fissure. UCG and fundus examinations were normal. GFR of the right kidney was slightly decreased as compared to the left (65 ml/min vs. 67 ml/min, by DTPA scan). By puncture of hydrops, yellow clear fluid was drained, the analysis showed similar composition to that of original urine, so subcapsular urinoma was diagnosed. Urine collection from two kidneys separately was performed by cystoscopy; nonselective proteinuria of 1+ was found in urine from the right and 2 + from the left kidney. Analysis revealed urea 36.8 mmol/l, potassium 6.92 mmol/l, creatinine 0.42 mmol/l in the right kidney urine compared to urea 77.2 mmol/l, potassium 11.19 mmol/l, and creatinine 1.29 mmol/l in the left, which suggested that the right kidney function was compromised. According to proteinuria from both kidneys with microscopic hematuria, without edema and hypoalbuminemia, glomerulonephritis was diagnosed.

Conclusion: The girl was diagnosed with glomerulonephritis and subcapsular urinoma. It was a rare case because of their co-incidence. Reasons for the hypertension, if caused by the glomerulonephritis or the pressure by subcapsular urinoma, as well as reasons for subcapsular urinoma need to be clarified during the follow-up.

AK-106: PSYCHOLOGICAL EVALUATION FOR CHILDREN WITH CHRONIC KIDNEY DISEASE

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Introduction: To evaluate the psychological conditions in children with chronic kidney diseases and the influence of their family.

Method: Totally 46 patients were analysed, of which 38 children were primary nephrotic syndrome with normal renal function, 8 patients were with chronic renal failure (GFR<15 ml/min/1.73 m2). Six psychical questionnaires were used to evaluate the conditions: Child Temperament Questionnaire, Elsenc Personality Questionnaire (EPQ), Children,s Self-consciousness Checkilst, Child Behavior Checkilst (CBCL), Medical Coping Modes Questionnaire (MCMQ) and Home Environment Checkilst.

Result: (1) Personality: 50% children age from 8–16 years old were introversive, and 40% children age from 3–7 years old were passive emotion; (2) Self- consciousness: Anxious score of children with chronic kidney diseases was lower than that of healthy children. In anxious group 40% children were introversive. All the children with abnormal conduct in CBCL were in anxious group. The mean period after treatment with steroid in anxious group was longer than that in non-anxious group. And the treatment in anxious group was more difficult than that in non-anxious group; (3) The behavior score was abnormal in seven children in CBCL; (4) In MCMQ, the parents of children with chronic kidney diseases would use the surrender or brave behavior; (5) In Home Environment Questionnaires: the scores of intimacy, perio knowledge, entertainment, morality and regulation were lower than that of healthy children. The knowledge score of

early group was lower than that of relapse and remission groups. The entertainment of remission group was more than that of early and relapse groups.

Conclusion: The children with chronic kidney disease had obvious anxious emotion, it was related with introversion, course, treatment and income. The anxious emotion will effect abnormal behavior, treatment and quality of life. We should pay more attention to the psychological conditions of children and their parents.

AK-176: URINARY CALCIUM TO CREATININE RATIO IN CHILDREN IN LIMESTONE AREA, KOREA

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Introduction: Random urine calcium to creatinine (U Ca/Cr) ratio has been used for screening hypercalciuria. Howerver, the U Ca/Cr ratio varies in different ages and in geographic areas. The purpose of this study aims at investigating the U Ca/Cr ratio and the effects of the types of drinking water to the U Ca/Cr ratio among primary school children in a limestone area.

Method: This study was performed on 398 healthy school children of age 7–12 in a limestone area. Random urine samples which were collected from all the children just before lunch hour in April 2008 were analyzed for calcium and creatinine. In a questionnaire, on the same day, the parents and caregivers filled out diet habit, disease history, family income, morning diet, and the type of drinking water of the children in research. As the control group, U Ca/Cr ratio of 350 healthy children in a non-limestone area was detected.

Result: The mean value for U Ca/Cr ratio of the limestone group and the control group were 0.09±0.076 and 0.11±0.133, respectively ($P < 0.001$). The 95th percentile for U Ca/Cr ratio of the limestone group and the control group were 0.25±0.076 and 0.30±0.134, respectively. The U Ca/Cr ratio >0.20 was 9% in the limestone group and 18.9% in the control group($P<0.05$). A negative correlation $R=-0.15$, $P=0.002$ was observed between age and U Ca/Cr ratio. There was no significant difference in sex. The U Ca/Cr ratio of the limestone group was irrelevant to diet habit, disease history, family income, morning diet, and BMI (B.wt/m2). No significant variations in U Ca/Cr ratio according to the type of drinking water such as filtered water, ground water and tap water were observed in the samples of the limestone group.

Conclusion: The expected value of U Ca/Cr ratio in the limestone area was not significantly high enough to compare with that in the non-limestone area. There were not different U Ca/Cr ratio according to the drinking water types.

A
Afroz Shireen AF-013
Akira Matsunaga AA-039
Aragon Eric Emmanuel AI-082
Asmaningsih Ninik AF-169
B
Bagga Arvind AA-197
Bagga Arvind SYM - 7-1
C
Cao Qi AA-098
Cao Li AD-191
Cao Qi FP2-4
Chan CM James SYM - 9-1
Chiou Yuan-yow AH-089
Chiou Yee-hsuan AB-151
Chiu Chun Man MN-1
Choi Hyun jin AH-053
Chu Mei AA-037
Chu Mei AA-067
D
Devarajan Prasad SYM - 6-1
Devarajan Prasad SYM - 7-2
Ding Jie MN-6
Ding Jie SYM - 11-2
E
Ehrich H. H. Jochen MN-3
Ejaz Iftikhar FP1-3
F
FAN Qingfeng FP5-1
Fang Xiao yan AD-100
Fang Xiao yan AK-106
Feng Shipin AA-046
Futrakul Prasit PL-3
G
Geary F Denis SYM-4-1
Gheissari Alaleh FP5-2
Guan Na AB-074
Guan Na AC-076
Guan Na AC-079
Guan Na AF-077
Guan Na AK-078
Guan Na FP2-3
Gulati Ashima AF-033
Gulati Ashima FP2-1
Gulati Ashima AH-201
Gunasekara Vindya FP1-6
Gupta Punit AA-153
Gupta Punit AA-154
Gupta Punit AA-155
Gupta Punit AA-178
Gupta Punit AG-152
Gupta Punit AI-202
Gupta Punit FP2-7
H
Ha Tae-sun AA-060
Ha Il soo SYM-2-1
Hanif Mohammed MN-2
Hari Smriti AE-188
Hari Pankaj FP4-3
Honda Masataka SYM-10-2
I
Igarashi Takashi SYM-9-2
Iijima Kazumoto SYM - 8-3
J
Jung Ji in FP3-2
K
Kanaheswari Yoganathan AC-020
Kanaheswari Yoganathan AK-019
Kanda Shoichiro AA-164
KANEKO Kazunari FP3-6
Kang Duk-hee SYM-1-1
Kanitkar Madhuri FP3-4
Kaskel Jeffrey Frederick SYM-6-2
Kaskel Jeffrey Frederick SYM-8-1
Khan Aamer FP2-5
Kim Pyung kil AA-017
Kim Sung-do AA-041
Kim Kee hyuck AI-062
Kim Kee hyuck AI-064
Kim Pyung kil AJ-006
Kim Su-yung AA-171
Kim Jung sue AD-018
L
Lai Wai ming AH-175
Lee Joo hoon FP1-2
Lee Seung joo SYM-5-3
Lee So hee AA-182
Lee Kwok wai AF-124
Li Yun ling FP5-3
Lim In seok AD-036
Lim Yam-ngo SYM-5-2
Lin Fangming SYM-1-2
Lin Fangming SYM-12-1
Lin Ching-yuang SYM-12-2
Liu Hai mei AI-097
Luchaninova Valentina N. AF-083
M
Ma Alison lap-tak FP2-2
Mahmud Shahabuddin AA-002
Moorani Khemchand AA-168
Muinuddin Golam AA-127
Namgoong Mee kyung AK-176
Ng Cherry fai ngo FP1-4
Ng Jun li FP5-5
Nguyen Duc quang AA-192
Nhu uyen Tran nguyen AF-173

O
Opastirakul Sauwalak AA-203
P
Paik Kyung hoon AH-159
Park Yong FP4-4
Pattaragarn Anirut AH-207
Perez-Reyes Ma. Jewellie AA-174
Prasetyo Risky vitria AH-068
Pudjiastuti Partini AA-167
R
Rees Lesley SYM-4-2
Rees Lesley SYM-10-3
Rianthavorn Pornpimol FP1-1
Rosenberg Andrew FP1-5
S
Saito Hiroshi AH-132
Salusky Benjamin Isidro PL-2
Sekine Takashi SYM-9-3
Sekine Takashi AB-133
Sethi Sidharth kumar AB-136
Sethi Sidharth kumar FP4-1
Shen Ying AA-021
Shen Qian AH-102
Shen Qian FP5-4
Sherali Afroze ramzan AA-057
Shimooka Takeshi AF-166
Srivastava Rajendra AA-149
Sugimoto Keisuke AB-073
Sugimoto Keisuke AB-200
Sultana Sabina AA-005
Sumboonnanonda Achra MN-4
Sun Li AI-107
T
Tanaka Ryojiro FP4-2
Tancharoen Worarut AA-196
Tangnararatchakit Kanchana AF-198
Tapaneya-Olarn Wiwat AA-199
Thirakhupt Prapaipim AA-206
Thirakhupt Prapaipim SYM-11-1
Tse Kei-chiu niko AD-190
Tse Kei-chiu niko FP3-3
W
Wang Qing ling FP3-5
Warady A. Bradley SYM-3-1
Warady A. Bradley SYM-10-1
Watson R Alan SYM-2-3
Wisanuyotin Suwannee AH-194
Y
Yan Kunimasa SYM-8-2
Yanagida Hidehiko AB-022
YAO Yong AI-044
Yap Hui kim PL-1
Yata Nahoko AK-051
Yeo Wee song AA-034
Yi Zhu-wen AA-047
Yi Zhu-wen AB-048
Yoo Kee hwan SYM-5-1
Z
Zhang Han AB-161
Zhang Han FP5-7