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ABSTRACTS

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Prof. G. Schubiger
PD Dr. M. Schwöbel

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MEDICAL REPORT FOR THE PARENTS?

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Motivation: From our point of view the cooperation of parents of disabled children is especially important for rehabilitation. Our question is whether giving parents a medical report (MR) leads to better information and cooperation.
Methods: In a prospective study we conducted a structured interview with the parents after their child's consultation at the polyclinic for rehabilitation about their expectations concerning a MR. A copy of the MR was sent to the parents and responsible therapists. A questionnaire was used to find out how far parent's expectations were fulfilled. With a similar questionnaire practitioners and therapists were surveyed.
Results: 62 parents (P) were interviewed and 55 returned the questionnaires. The expectations of 48 (87%) P were fulfilled. 53 (96%) P would like a MR in the future. 14 (26%) P would prefer a MR written for them. 45 (82%) P understood most of the MR, 9 (16%) partly. 61 general practitioners (GP) and 55 therapists (T) filled in the questionnaire. 39 (64%) GP and 49 (89%) T find giving the MR to the parents is convenient, 8 (13%) GP do not think so. 47 (86%) P, 47 (77%) GP and 50 (91%) T believe that cooperation improves with MRs. 37 (67%) P, 31 (51%) GP and 34 (62%) T consider parents are better informed with a MR.
Conclusion: Giving the parents a MR is desirable in paediatric rehabilitation and in the opinion of parents, practitioners and therapists this direct information further improves cooperation.

Side EFFECTS OF THERAPY WITH RITALIN®: PREVALENCE AND ASSOCIATED FACTORS

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Introduction: Dosis and age are the only known risk factors for side effects of therapy with Ritalin®.
Question: Are psychosomatic reactions, neurotizism and emotional comorbidity risk factors for side effects of therapy with Ritalin®?
Methods: In a retrospective cohort study parents of 73 children with attention deficit hyperactivity disorder got questionaires for side effects and clinical effects of therapy with Ritalin®, emotional comorbidity (CBCL 4-18), the children got questionaires about neurotizism (HANES,KJ) and body complaints (GBB,KJ). The whole study group was divided with spli-half-method in a group with low side effect-index (group 1) and high side effect index (group 2).
Results: Decreased appetite (62%), prone to crying (44%), unhappiness (40%) and headache (40%) are the most called side effects. The children of group 1 are older and get a higher total drug dosis/day. They have a better compliance and in tendency more positive clinical effects. The children of group 2 show more associated psychiatric disturbance. No difference is found in neurotizism and body complaints.
Conclusion: Therapy with Ritalin® has a high prevalence of side effects, but mostly it was tolerably in relation to the good clinical effects. Associated mood and conduct disorders are risk factors for side effects.

EFFECT OF DETERGENT-COATED VERSUS NON-COATED SPACERS ON BRONCHODILATOR RESPONSE IN CHILDREN WITH ASTHMA

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Rationale: Previous studies have demonstrated that coating spacers with ionic detergents reduces the static charge and thereby improves *in vivo* pulmonary drug deposition. To date, little is known about the effect of coating on clinical outcome from inhaled bronchodilators.
Methods: Randomized, double blind study in children with asthma and an $FEV_1/FVC < 72$ % predicted. Spirometry was performed at baseline and 10 and 20 minutes after inhalation of 2 puffs salbutamol (100mcg/puff) via detergent-coated or non-coated spacer.
Results**:**

Group	n	Age	FEV_1 @ 0	ΔFEV_1 10	ΔFEV_1 20	FEV_1 @ 20
Non-coated	24	11.6 (8-17)	81% (65-106)	18.5% (3-35)	19.5% (9-35)	98% (77-122)
Coated	26	11.5 (7-18)	76% (45-101)	18.8% (5-50)	19.8% (0-50)	93% (56-119)

Conclusions: There was no improvement in bronchodilation from detergent coating the spacer in this group possibly because a maximal response had been achieved with the lower output.

INITIATION OF RESUSCITATION AND INTENSIVE CARE IN EXTREMELY PRETERM NEWBORNS IN SWITZERLAND

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Background: Survival rate among live births under 26 weeks' gestation has steadily increased. Unfortunately, this has not been followed by a parallel decrease in morbidity. On the contrary, there is mounting evidence for increasing major long-term morbidity in extremely premature infants surviving intensive care. The EURONIC study (11 European countries) showed a broad range of opinions regarding institution of intensive care in a paradigmatic case of a 560-g, 24 weeks' gestation newborn with a 1-minute Apgar score of 1. **Aim:** As nationality was the strongest determinant of whether resuscitation was provided, we were interested to examine how this situation would be approached by neonatal care professionals in Switzerland. **Methods:** The anonymous, self-administered EURONIC questionnaire (kindly made available by M. Cuttini, Trieste) was sent to all 11 neonatal intensive care units in Switzerland. So far, 60 physicians and 85 nurses completed the questionnaire. **Results:** Only 3% (EU 22%) of Swiss physicians and nurses would initiate resuscitation if intensive care could not be withdrawn once started. 66% (EU 64%) would agree on resuscitation provided that intensive care could be withdrawn in case of a poor prognosis. Finally, 25% (14%) would not initiate resuscitation. **Discussion:** Very few Swiss professionals start resuscitation in infants at the threshold of viability without the possibility to later withdraw intensive care. Only France (4%) and the Netherlands (1%) showed similar figures. There is much less difference regarding the decision to provide provisional care with the option to stop, should the infant's prognosis turn out to be very poor.

TRIAL OF LIFE APPROACH FOR INFANTS BORN AT LESS THAN 26 WEEKS OF GESTATION

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In Switzerland, there is an ongoing debate regarding active resuscitation of infants born at less than 26 weeks of gestation. For various reasons (uncertainty surrounding the precise gestational age, biologic variability at any given gestational age) infants born at ≥ 24 0/7 weeks of gestation are actively resuscitated in the delivery room at our institution after careful discussion with the parents. However, care can be redirected if the patient's evolution is unfavorable (*Trial of Life Approach*). Over a 4-year-period, 19 preterm infants with a gestational age of less than 26 weeks were actively resuscitated in the delivery room and transferred to the neonatal intensive care unit.

Gestational age (weeks)	Mortality	IVH (> 2)	PVL	ROP (> 3)	CLD (36 weeks)
24 0/7-24 6/7 (n=10)	50% (5/10)	20% (1/5)	0% (0/5)	0% (0/5)	0% (0/5)
25 0/7-25 6/7 (n=9)	22% (2/9)	0% (0/7)	0% (0/7)	14% (1/7)	14% (1/7)

Our results are comparable with published data from international tertiary neonatal intensive care units and have to be considered in the ongoing debate about the optimal management of infants born below 26 weeks of gestation.

TOXOPLASMOSIS AND PREGNANCY: FINDINGS FROM CORD BLOOD SCREENING IN 65.000 NEWBORNS

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Aims of the study: To analyse the changes in toxoplasmosis seroprevalence among pregnant women and in incidence of congenital toxoplasmosis over the past 20 years. Furthermore to assess the efficacy of toxoplasmosis infection treatment in pregnancy on the outcome of new-borns.
Methods: Cord blood screening for congenital toxoplasmosis has been performed prospectively on 65.000 samples collected between 1982 and 1999 in the region of Basel, Switzerland, covering up to 95% of all births. Congenital infection was suspected in cases of serum with specific anti-toxoplasma IgM or IgA or with a high level of IgG (>300 UI/ml) and an inquiry with the obstetrician in charge of the pregnant women was carried out. The children were then followed up for determination of antibody titers in peripheral blood and disease symptoms and eventually treated.
Results: A continuous decrease in toxoplasmosis seroprevalence among pregnant women and in incidence of congenital toxoplasmosis was observed. The implications of these findings regarding toxoplasmosis prevention strategies and the efficacy of treating women who seroconvert during pregnancy are discussed.

Swiss Minimal Neonatal Data Set: LENGTH OF HOSPITAL STAY OF VERY LOW BIRTH WEIGHT INFANTS

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Objective: To evaluate if the increased morbidity of very low birth weight (VLBW) infants has an impact on the length of hospital stay (LOS). **Methods:** In 1996, perinatal data of all infants born below 32 weeks of gestation and/or with a birth weight below 1500 g were collected nation-wide. LOS of all 619 VLBW infants who survived to discharge was analysed. **Results:** Independent of the degree of immaturity, 80% of all VLBW infants had been discharged by the time they reached their due date, and at a corrected age of 44 weeks, 95% of all patients were at home. However, 2.5% remained hospitalised for more than 12 weeks beyond term.

Gest. age (weeks)	n	LOS (weeks, mean ± SD)	Corr. age (weeks)
24 0/7 - 25 6/7	27	14.7 ± 2.7	39.6
26 0/7 - 27 6/7	72	14.0 ± 5.2	40.7
28 0/7 - 29 6/7	156	10.6 ± 4.8	39.1
30 0/7 - 31 6/7	258	6.9 ± 2.1	37.5

Conclusions: LOS of VLBW infants is largely determined by their physiological immaturity. The impact on LOS of the increased morbidity of VLBW infants is limited and significant only for a small subgroup of patients.

PULMONARY HEMORRHAGE IN NEONATES: Risk factors in preterm and term infants

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Objective: To determine risk factor profiles of preterm (PT) and near-term/full-term (NT/FT) infants with clinically significant pulmonary hemorrhage (PH).
Study design: Retrospective case-control study of all infants with PH cared for in three Harvard-affiliated neonatal intensive care units between 1987 and 1994.
Results: 50 cases of PH occurred in PT infants (GA≤34 weeks), and 26 cases occurred in NT/FT infants (GA>34 weeks). Among NT/FT infants, PH was observed significantly earlier than in PT infants (median age 6 hours and 46 hours, respectively). Mortality was 38% in PT infants and 23% in NT/FT infants with PH. Among PT infants, a GA between 24 and 26 weeks and antenatal glucocorticoid treatment reduced the risk of PH (odds ratios (ORs) 0.7 and 0.3, respectively), whereas requirement for resuscitation in the delivery room and thrombocytopenia increased the risk (ORs 4.3 and 4.0, respectively). Among NT/FT infants, meconium aspiration (OR 4.9), requirement for resuscitation in the delivery room (OR 2.9), and arterial hypotension (OR 3.5) increased the risk of PH.
Discussion: Antecedent factors and timing of PH differ between PT and NT/FT infants, suggesting that different mechanisms can lead to the postulated *stress failure of pulmonary capillaries* with resultant PH.

Neurodevelopmental follow-up of infants treated for a neonatal persistent pulmonary hypertension with magnesium sulfate or nitric oxide

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Introduction: Persistent Pulmonary Hypertension of the Newborn (PPHN) is a serious complication of neonatal adaptation with important mortality and morbidity. Previous therapies (Tolazoline, Prostacyclin, Hyperventilation, Alcalinization, Extra-corporeal membrane oxygenation) were associated with adverse outcome. High doses of magnesium sulfate (MgSO4) and nitric oxide (NO) are new therapies but long term follow-up is currently unavailable.
Methods: Neurodevelopmental follow-up at 18 months, 3 1/2, and 5 years of infants treated for a PPHN with MgSO4 or/and NO was compared to a contemporary control group. Infants with congenital malformations were excluded.
Population: 68 infants born between january 1992 and december 1998 were included into the study. 52 infants were treated with MgSO4, 8 with NO, and 8 with MgSO4 and NO.
Results: 5 infants/68 died (7.4%). Follow-up included 63 patients.

Evaluation at (n)	Normal infants	Neurodev. abnormalities	Major handicaps	QD/QI
18 months (45)	39 (86.7%)	4 (8.9%)	2 (4.4%)	107±9
Pre-school (28)	17 (60.7%)	9 (32.1%)	2 (7.1%)	112±15

Conclusions: Incidence of neurodevelopmental anomalies in infants treated for a PPHN with MgSO4 or NO is less than previously reported. Use of MgSO4 and NO for a PPHN appears to be safe.

INTEROBSERVER VARIABILITY (IOV) IN THE RADIOGRAPHIC DIAGNOSIS (XrDg) OF NEONATAL ENTEROCOLITIS (NEC).

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Objective: To test the hypothesis that IOV in the XrDg of NEC was considerable and could contribute to the variability in incidences reported by various Centers. Design : Abdominal Xrays were sent to 4 experts from 3 Perinatal Centers who were blinded to patient identity and clinical course. Each observer was asked to fill out a questionnaire asking for the presence or absence of 6 standard signs of NEC and an overall evaluation of the case as 1) NEC certain, 2) NEC possible, 3) NEC excluded. Population : 87 pairs of abdominal Xrays were arbitrarily chosen from 300 infants < 34 wks consecutively admitted to one of the 3 Centers, (Center C) irrespective of the indication for the Xrays. The incidence in NEC and NEC-related death per Center (Centers A, B, C) was compared in patients of < 1500 g. Statistical Method : To quantify the IOV we calculated the Kappa statistics for each individual characteristic and for overall XrDg of NEC between the 4 experts. Results : The prevalence of the XrDg of NEC certain varied from 7 to 51% (7, 16, 37 & 51% resp.) whereas the Dg of NEC excluded varied from 28 to 61% of cases among the 4 readers. Readers A and B (affiliated with Centers A & B) were significantly more prone to exclude a NEC than C & D, both affiliated with Center C ($p<0.0001$, McNemar test). Kappa values for concordance in overall Dg between the 4 readers was only slight, i.e. 0.16. Conclusions : IOV is sizeable and could in part explain the inter-center variations in incidence since the reference radiologists with the higher (C) and the lower (B) incidences of positive XrDg were affiliated to the Centers with respectively the highest (16%) and the lowest (5%) reported incidences of NEC (<1500 g.) whereas NEC-related mortality was constant (1.6 vs. 1.3% for Centers C vs. B).

Simulated necrotizing enterocolitis by transplacental passage of Iopentol

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Background: The diagnosis of necrotizing enterocolitis may be very difficult. Radiological signs are: Pneumatosis of the bowel wall and, if perforation occurs, free gas in the abdominal cavity. The so-called "rigler sign" is typical for pneumoperitoneum: the bowel wall appears contrasted because of intra- and extraluminal gas.
Case report: Twin pregnancy after in vitro fertilisation. Twin B, female, born at 30 weeks gestational age, APGAR 8/9/10 at 1,5 and 10 minutes respectively. The child was intubated for RDS and developed a pneumothorax. Clinical picture for neonatal sepsis. On radiographs obtained at the second day of life appearance of "rigler sign" and thick bowel wall on the abdominal ultrasound. Clinically no evidence for necrotizing enterocolitis. A control radiograph of the brother's abdomen showed the same finding. Further history taking revealed that the mother had undergone a computed tomography (twice) with contrast medium (iopentol) for suspected pulmonary embolism 12 days before delivery.
Hypothesis: We propose, that iopentol may have passed into the fetal circulation and rapidly excreted by the fetal kidney into the amniotic fluid and then swallowed by the twins. Finally it was concentrated into the bowel lumen.
Conclusion: By positive "rigler sign" without clinical correlates for necrotizing enterocolitis, the clinician should think of this probably rare situation.

CONGENITAL PULMONARY LYMPHANGIECTASIA

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Case report: Shortly after birth, this full-term female infant (gestational age 40 3/7 weeks, birth weight 2970g) developed severe respiratory distress requiring intubation and mechanical ventilatory support. Chest x-ray revealed diffuse bilateral nodular interstitial markings. As ventilation and oxygenation became increasingly difficult, the patient was switched from conventional to high frequency oscillatory ventilation. A trial of exogenous surfactant was unsuccessful. Cultures of tracheal aspirate and blood remained sterile. Congenital heart disease was ruled out by echocardiography. After exclusion of surfactant protein B deficiency (ELISA of bronchoalveolar lavage fluid, DNA analysis of blood), an open lung biopsy was performed on day 18 of life. On histology, there was extensive subpleural and septal cystic lymphangiectasia. Following redirection of care, the infant rapidly expired.
Discussion: Congenital pulmonary lymphangiectasia leading to respiratory failure in the neonatal period is always fatal. In contrast, pulmonary lymphangiectasia secondary to pulmonary venous obstruction or as part of generalised lymphatic abnormalities has a better prognosis. Congenital pulmonary lymphangiectasia is a rare disease of unknown aetiology. Diagnosis can only be established histologically.

SURVEY OF CHILDREN DEPENDENT ON LONG-TERM MECHANICAL VENTILATION IN SWITZERLAND

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Background: Advances in the treatment of chronic respiratory failure in pediatric patients have resulted in a new patient population: children dependent on long-term ventilation. Data on the use of long-term ventilation in Swiss children is not available. We aimed to identify number, diagnoses and medico-technical management of this patient population in Switzerland. **Design**: Questionnaires sent to all pulmonologists, intensive care specialists, neurologists, health care organizations, rehabilitation services and ventilator companies known or suspected to be involved in pediatric long-term ventilation. **Results:** Detailed information was obtained on 32 children: congenital central hypoventilation syndrome (N=13), neuromuscular disorders (N=12), spinal cord injury (N=2), craniofacial anomalies (N=2), others (N=3). 10 children were ventilated by tracheostomy, 19 children by nasal mask, 2 children by phrenic nerve pacing and one child with a pneumatic belt. Ventilation was necessary 16-24 hrs a day in 5 children, during sleep in 24 children and only episodically in 3 children. All but 2 children were cared for at home; 20 families were supported with professional home care. **Conclusions**: The number of ventilator-dependent children in Switzerland is small. Most of them are cared for at home and followed by a few specialized centers. There is a need for establishing a continuing data collection on this particular patient population to assess outcome, quality of home care and changing incidence figures.

OUTBREAK OF COXSACKIE VIRUS B5 MENINGITIS IN A SCOUT SUMMER CAMP

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Introduction: Aseptic meningitis during the summer months is caused most commonly by enteroviruses (e.g., Coxsackie virus B2 and B5, Echo virus 4,6,9,11,16,30). Predominantly fecal-oral transmission favors the spread of these viruses among children and in closed communities. **Patients/Methods:** 2 days after returing form a scout summer camp in Campra, Ticino, in August 2000, 10 (8 boys) of the 32 participants (range 10-15 Years) were hospitalized at our institution with complaints of acute onset, high-grade fever, severe headache, and vomiting. In all patients, clinical signs included nuchal rigidity, and other signs of meningeal irritation. On admission, the peripheral white blood cells count was elevated in 1 patient ($18x10^9$/L), the range of plasma C-reactive protein was 0.3 to 12 mg/L. Median cerebrospinal fluid (CSF) cell count was 1026,5/µL (range, 169-2270). enteroviral RNA PCR in CSF was positive in 4 of 4 patients. Stool culture was positive for Coxsackie virus B5 in 10 of 10 patients. Clinical management was supportive. Fever and signs of meningeal irritation abated within 4 to 7 days. No complications were observed. **Conclusion:** This outbreak among adolescents of aseptic meningitis was caused by Coxsackie virus B5. Enteroviral PCR of cerebrospinal fluid was a useful tool for management of the outbreak because it allowed rapid identification of its cause, accurate information of local authorities, and reassurance of the families involved.

FAMILIAL MEDITERRANEAN FEVER (FMF): A CLINICAL AND MOLECULARBIOLOGICAL DIAGNOSIS

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Introduction*:* FMF is an inherited multisystem disease, characterised by recurrent, painful, self-limited episodes of peritonitis, pleuritis arthritis or erysipelas-like erythema usually accompained by fever. The desease is common in the middle east and mediterranean basin. In Switzerland FMF is rare, but with migration of the people could become a frequent desease. The Genelocalisation (16p13.3) and its correlated protein (Marenostrin) are now known. **Case history***:* A 4 year old Turkish child since she was 2 years old had periodic fever attacks. Her parents are close relatives (first degree). She was many times hospitalized because of episodes of high fever, severe abdominal pain (peritonitis), unilateral chest pain, and one episode of monoarthritis of the elbow with maculo-papulo exanthem. Laboratory data showed high inflammatory parameters (CRP between 80-220 mg/l). The leucocytes were normal. Labor findings for other rheumatological systemic deseases were negative. The molecular investigations in Geneva show the presence of two alleles (Genotype M694V/M694Y) by both parents and the homozygote inheritance by the child. The colchicine-therapy (1 mg/day) can remove the symptoms after six months. **Conclusion***:* FMF is a rare desease, but can also be diagnosed in Switzerland. Today Molecular investigations are the gold standard for the diagnosis of FMF. The colchicine therapy can prevent secondary complications, such as renal amyloidosis.

VARICELLA-ZOSTER VIRUS (VZV) AND INVASIVE MENINGOCOCCAL DISEASE

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Motivation: Chickenpox is considered a benign disease in children. Secondary bacterial infections with organ involvment due to group A betahaemolytic streptococcus are well known. Meningococcal disease complicating varicella is a recently reported association.
Case reports: Pat. 1 Two days after onset of chickenpox a 6-month old girl was admitted to hospital with fever and a petechial rash. Blood culture grew N. meningitidis serotype B. VZV was isolated from the vesicular rash. Pat. 2 A 5-year old girl presented two days before admission to hospital to the general practitioner with purulent conjunctivitis. With fever, pain in the arms and a petechial rash she was admitted by urgency. Blood, cerebrospinal fluid and conjunctival swab were positive for N. meningitidis serotype C. On the fourth day of hospitalization she developed chickenpox confirmed with a positive culture of VZV from a vesicle.
Conclusions: Exposure to varicella may lead to invasive disease by meningococci. Infants and toddlers are susceptible to both infectious agents. Active immunization is available for both VZV and N. meningitidis serotype C (conjugate vaccine).

Diagnosis of Acute Hematogenous Osteomyelitis and Septic Arthritis: A 20 Year Experience at the University Children's Hospital Basel

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Objective: To review the diagnostic experience with acute hematogenous osteomyelitis (AHOM) and/or septic arthritis (SA) at our institution. **Methods**: Retrospective (January 1980 and July 2000) case history review of patients with bacteriologically and/or radiologically confirmed diagnosis. **Results:** 90 patients (61% males), 4 weeks to 14 years of age, met the inclusion criteria. Median duration of disease prior to hospitalization was 3 days (range 0-14); 88% were admitted during the first week after onset of complaints. 81 patients did not receive antibiotics prior to hospitalization and are the subject of this presentation. In these ESR (1st hour; median 36; range 11-124), CRP (mg/l; median 64; range 0-221) and WBC (x 10^{-9}/l; median 13; range 5-34) were elevated in 99% (>10), 78% (>20) and 66% (>12), respectively. One child was normal for all three parameters. Blood cultures (BC) and/or tissue cultures (TC) were performed in 79 patients (98%). Bacteria were isolated from 54 patients (66%) with S. aureus as the most frequent organism (N=27; 51%). BC were performed in 67 patients and yielded 35(52%) positive cultures; TC (N=47) yielded 29 (61%) isolates. In 34 patients with both, BC and TC performed, only 12 (35%) were positive in both tests. Diagnostic findings were observed in 23 (58%) conventional radiographs, 31 (56%) sonographs, 39 (90%) 99mTc- labeled bone scans and 4 (100%) MRIs. In 41 patients with diagnostic radiological findings TC was performed and yielded 30 (73 %) bacteriological isolates. The overall median duration of hospitalization was 15 days (range 2-66). **Conclusion:** Our data suggest that the diagnostic procedures of choice should be 1) early bone scan (or MRI), 2) BC and 3) TC. Of supportive laboratory parameters, ESR followed by CRP were most valuable in our hands.

GR. A beta HEMOLYTIC STREP. OSTEOMYELITIS (GABHS) RARE COMPLICATION OF CHICKENPOX

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Introduction: in childhood, the GABHS causes various infections, of which the most frequent are tonsillopharyngitis and impetigo. Rarely, we may face invasive infections such as bacteremia, meningitis, necrotising fasceitis or osteomyelitis. The last 15 years, we have witnessed an important increase in the incidence of invasive GABHS infections as a complication of chickenpox. **Case:** 6 years old girl in good general health, presenting with a 3 days history of chickenpox, was hospitalized for fever and painful tumefaction of the right ankle. Lab investigations showed WBC 19,9 G/L (NSN 8,8), CRP >200, ASAT 174 U/l, ALAT 102 and a positive Strep throat. Parenteral antibiotherapy was started. Due to persisting fever, progressive swelling, rise in CK max. at 830 U/l and GABHS positive blood cultures, a bone scan was performed. Described first as normal, it revealed a hypoactivity of the right fibula. Two biopsies (antero-int/ext) of the right leg abnegated the presence of any germs or inflammatory lesions. Clinical improvement having failed despite an iv. Penicillin, Clindamycin and Acyclovir therapy, we proceeded to a MRI, which showed a right fibula osteomyelitis, a sub-periostic abscess and a myositis. A surgical drainage allowed GABHS to be isolated. Thereafter, the clinical and paraclinical evolution was harmonious. A bone scan after 10 days showed a revascularisation of the fibula. **Discussion:** this case illustrates a rare, but known, complication of an invasive GABHS infection. Nevertheless, to our surprise, the first bone scan has revealed a hypocaptation of the bone, corresponding to an absence of vascularisation. Only a deep surgical drainage has allowed a favorable evolution. **Conclusion:** every child suffering from chickenpox, whose evolution is not satisfactory, should benefit from further investigation, in order to exclude an invasive SBHGA infection, the prognosis being subject to a prompt diagnosis and early treatment.

VISCERAL LEISHMANIASIS: A CAUSE FOR HEPATO-SPLENOMEGALY, ALSO IN SWITZERLAND

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Motivation: Hepatosplenomegaly is a symptom of various diseases. Among them, visceral leishmaniasis is rare in Switzerland. The differential diagnosis and the respective tests will be discussed based on the case of a toddler. **Methodology:** Discussion of the differential diagnosis of hepatosplenomegaly with respect to our case report.
Patientenbericht: A 2 year 5 month old girl, resident of Elba, presented with deterioration for a few weeks, increasing hepato-splenomegaly, as well as normochromic normocytic anemia (Hb 81 g/l) and thrombocytopenia (106 G/l). Acute leukemia was excluded by bone marrow aspiration. Serology was positive for Leishmania infection (*Leishmania infantum* by PCR of bone marrow aspirate). Furthermore, hypergammaglobulinaemia was present, presumably caused by polyclonal stimulation of B-lymphocytes. Surprisingly, serology for EBV indicated chronic infection, which is probably due to the same process. Treatment was performed with liposomal Amphotericin B i.v. (3mg/kg/d for 5 days and on day 10, total dose: 18mg/kg) and led to improvement of the child's general condition, including regression of hepatosplenomegaly and normalization of blood parameters. **Conclusion:** Visceral leishmaniasis (Kala-Azar) is rarely observed in Switzerland. Our patient lives on Elba, where this disease, caused by *Leishmania infantum*, is endemic. Treatment with liposomal Amphotericin B is preferable to treatment with pentavalent antimonial compounds, because costs as well as duration of hospitalization can be reduced. With appropriate treatment prognosis is good and parasites get eliminated in 85-90% of the patients.

"POTT'S PUFFY TUMOR"

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Since the introduction of antibiotics, the incidence of complications of frontal sinusitis has diminished. We describe a rarity: A 13-year-old girl complained about headache and rhinitis. The initial therapy was symptomatical. After a few days, a indolent, hard, subcutaneous swelling of the forehead occurred. Except headache the girl showed no other symptoms. The diagnosis of a frontal sinusitis was made by a conventional X-ray. An oral therapy of antibiotics was introduced. The headache disappeared but the swelling of the forehead remained. A CT scan of the head showed a subperiosteal abscess of the frontal bone, a complication of the frontal sinusitis.
This rare entity was first described by Percival Pott in 1775 ("Pott's puffy tumor"). In most cases, a intracerebral infection is associated. The procedure of choice for diagnosis is the CT scan using contrast medium enhancement. In addition to intravenous administration of antibiotics, the indication to surgical drainage has to be made with the ORL or neurosurgeon

PARTIAL TRISOMY 8

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We report on an actually four month old infant with a partial trisomy 8. This chromosomal anomaly is associated with multiple malformations like congenital diaphragmatic hernia (CDH) and psychomotoric delay.
Case report: The girl (3170 g (P50), 51 cm (P50-90), 36,0 cm (P>90)) was born spontanously after an uncomplicated pregnancy without additional risks. Prenatal ultrasounds did not show any pathologic findings. After birth the child suffered from severe respiratory distress due to CDH, which was closed on the 3rd day. Other clinical findings were: dysmorphic face with hypertelorism, megalocornea, ASD and VSD. The actual echocardiography demonstrates typical signs of pulmonary hypertension. A systemic hypertension could be succesfully treated. Head MRI showed a general brain atrophy and a hypoplasia of the sella turcica. Since birth there was no growing of head circumference. Crying fits occured since the age of two month, fixation was not possible until yet. Feeding was always done by a nasogastric tube because of swallowing problems.
Conclusion: Our patient emphasizes the need for cytogenetic diagnostics in case of CDH, especially when associated with dymorphic features. Although only two other cases are published we suggest that partial trisomy 8 is associated with an unfavorable outcome.

Pathologic Galactosemia Screening due to porto-caval shunt in Trisomy 8 Mosaicism

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A term male newborn (gestational age 41 3/7 weeks, birthweight 3240g) presented with: camptodactyly left PIP III, deformed auricles, bilateral pes adductus with deep plantar dermatoglyphs, pear-shaped nose, down-slanting palpebral fissures, sucking weakness. Imaging revealed agenesis of corpus callosum, ectasia of renal pelvis, small ASD II, additional thoracic and lumbal vertebrae, butterfly vertebra C5. Unsuspicious clinical course. Screeningtest for galactosemia showed twice elevated galactose (max. 0.5mmol/L) despite normal galactose-1-phosphate-uridyltransferase [GALT]- activity. After unobtrusive challenging test with breast-milk (galactose 0.02 → 0.11 mmol/L bzw. NH_3 51 → 71 µmol/L) no diet was introduced. Ultrasound imaging revealed porto-caval shunting in the left hepatic lobe, no patent ductus venosus. Karyotype showed trisomy 8 mosaicism. At 2 months of age no intrahepatic shunts were visible.
Hypergalactosemia with normal GALT activity in newborns is mostly due to porto-caval shunting, hepatic encephalopathy should not be feared if serum NH_3 stays <80 µmol/L. Agenesis of corpus callosum is a major finding in trisomy 8 mosaicism. Porto-caval shunting has yet not been reported with this chromosome aberration. Its distinct clinical picture is thus broadened by a new feature.

INFANTILE MYOFIBROMATOSIS AND BIRTH DEFECTS

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Infantile myofibromatosis (IM) is a rare disease characterized by fibromatous tumors. Unless vital organs are involved, its prognosis is good as nodules tend to spontaneously regress.
We report a very rare case of syndromic neonatal generalized IM, ie associated with birth defects. At birth, examination revealed more than 50 skin and subcutaneous firm lesions of 0.2-4 cm diameter, a posterior cleft palate, a tail-like caudal appendix and an anteriorly placed anus. On X-Rays the lungs were considered unsuspicious but multiple osteolytic lesions of the long bones were visualized. MRI showed brain involvement and confirmed myocardial tumors diagnosed by echo. Karyotype was 46,XX. Biopsy of a skin lesion allowed diagnosis of IM.
At 10 weeks, the child was readmitted for a rapidly life-threatening pericardial effusion. Treatment with Interferon alpha 2a did not stop progress and was changed to vincristine/actinomycin after 7 days. The patient died of respiratory failure at 12 weeks of age. Autopsy revealed severe diffuse myofibromatoid lung infiltration, not suspected on X-ray and MRI.
This case, with only two other reports of IM associated with birth defects and only few cases of familial recurrence, suggests that the hitherto unknown etiology of IM might be genetic. In addition, we want to draw attention that imaging techniques may underestimate diffuse lung infiltration, described as an important prognostic factor in IM. Aggressive lung investigation with biopsy followed by early treatment may influence the course of generalized IM.

A NOVEL SYNDROME INVOLVING PRIMARY SKELETAL GROWTH AND RETARDATION IN SIBLINGS

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An identical pattern of malformations was found in two brothers both having microcephaly and severe developmental delay. Additionally, they had hypothelorism, epicanthic folds, and convergent strabismus. There was shortening of either the radius or the tibia and shortening of the first metacarpals. A persistent dorsally flexed finger and toes all of which are unusually long. Both boys had a high-pitched voice and were unable to communicate verbally at the age of 4.5 years. They both developed short stature. One brother has anal atresia; the other had a pulmonary artery atresia, VSD, ASD, and an over-riding aorta. This apparently new syndrome is possibly an autosomal, or a X-linked recessive trait.

WHAT DO ADOLESCENTS WITH CHRONIC CONDITIONS THINK ABOUT QUALITY OF LIFE ? FIRST STEPS IN THE CONSTRUCTION OF A NEW QUESTIONNAIRE.

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The number of children with chronic conditions who reach adolescence increases due to more performing care. These young people have specific needs and set new complicated questions about their quality of life and the adequacy of their medical care.

In a qualitative approach, we conducted semi-structured interviews with adolescents, in order to know their point of view about different domains related to their experience of patient. We plan to integrate these data in an existing questionnaire about quality of life, elaborated in Marseille and currently in validation. Our project is supported by the Unity of quality of care of the University Hospital of Geneva.

We present the results of 18 interviews of adolescents with various chronic conditions (average age 15.9 years). The domains pointed out by them in response to our questions are the following :

QUESTIONS	DOMAINS
Quality of life	Family, friends, school, health
Adolescence	Health, growth, physical appearance
Chronic condition	Announcement of diagnosis, beginning of disease, treatment, compliance, manifestation, evolution

Two third of the adolescents wish « not to be sick anymore ». The announcement of the diagnosis is lived through very intensely, especially when the chronic condition begins during adolescence. More than half of the young people has school difficulties, the great majority of these even repeated a year.The notions of self-esteem, freedom and mood seem more difficult to understand.

Conclusion : The integration of the adolescent as an active participant is a necessary step. The emphasis he lays on particular domains as well as the way he expresses himself are precious tools in the elaboration of a questionnaire.

PRENATAL DIAGNOSIS OF CONGENITAL HEART DISEASE BY FETAL ECHOCARDIOGRAPHY: GENEVESE EXPERIENCE

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Introduction : Fetal echocardiography (FECG) allows to detect early in pregnancy congenital heart disease (CHD), which is the most common form of congenital malformation. Thus, it becomes possible to prepare a high risk delivery and to improve the management of the newborn infant.

Patients and Method : This retrospective study counted 596 fetuses examined by FECG between 1990 and 1998 by an obstetrician and a pediatric cardiologist. The medical files of the Children's Hospital of 408 among these patients were reviewed, while 188 (30%) fetuses were lost to follow-up. The indication for FECG was in 299 cases (50%) a family history of CHD, with 158 (52%) of these scans detecting a structural cardiac anomaly. In 116 cases, the FECG was performed due to a suspicion of CHD during the obstetrical screening and 103 (89%) of these scans revealed an anomaly.

Results	Arrhythmias	46 (8 %)
	Structural Anomalies	326 (55%)
	among which 214 (65%) are ventricular	*septal defects (VSD)*
Evolution	Spontaneous resolution of the anomaly among 408 children followed-up	122 (30%)
	In utero deaths	6 (1%)
	Pregnancy interruption for severe CHD	35 (6%)
	Postnatal deaths	16 (3.9%)

We observed 17 (3.5%) false negatives and 3 (0.7%) false positive results.

Conclusion : A family history of CHD is the leading indication for FECG. Nonetheless, the proportion of scans detecting a cardiac anomaly is much higher if the indication is a suspicion of CHD during the general screening. The parents choose early termination of pregnancy in a in cases of severe CHD. Numerous structural structural anomalies show a spontaneaous resolution. The mortality in our series is low due to the improved management and the interrupted pregnancies with the severest CHD. The efficiency of FECG is good and the proportion of false positives and false negatives is consistent with the numbers found in the literature.

Superiority of Amplatzer Duct Occluder over traditional Coil-Occlusion in moderate to large sized Persistent Arterial Duct

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Background: So far the persistent arterial duct (PDA) was usually closed with catheter interventional detachable coils (CO). In moderate to large sized PDA the occlusion rate with CO is unsatisfying. The Amplatzer Duct Occluder (ADO) is an alternative to the CO. We compared the efficacy of both methods.

Method: Until 1999 the PDA occlusion was performed with CO in 10 patients (age 1.2-10.6y/PDA 2-3 mm). Since January 2000 ADO were implanted in 8 patients (age 2.2-9.3 y/PDA 2.5-6mm). The complete closure, a possible obstruction of the pulmonary artery or descending aorta was initially examined with angiography and during the follow up period with Doppler echocardiography. The complication rate and complete closure immediately , 3 and 12 months after intervention were compared.

Results: Immediate closure of the PDA after implantation of the ADO could be demonstrated in all patients. There were no complications. Only one out of 10 patients with CO demonstrated immediate closure, while after one year of follow up the arterial duct was closed only in 6 of 10 patients. In one patient a coil embolised into the pulmonary artery. Adequate positioning of the coil was not possible in one patient.

Conclusion: Amplatzer Duct Occluders demonstrate a higher successful occlusion rate immediately after intervention and after one year of follow-up.

BALLOON VALVULOPLASTY OF PULMONARY VALVE STENOSIS: MIDTERM RESULTS AND RISK FACTORS FOR THERAPY FAILURES

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Background: The balloon pulmonary valvuloplasty (BVP) is the treatment of choice for congenital pulmonary valve stenosis (PVS).

Method: Between 1988 and 2000 99 BVP were performed in 96 patients (P) aged 0-18.6 years (median 29.5 month). This retrospective study analyses the short and midterm results of the BVP as well as risk factors for a poor result. **Results**: The BVP led to a significant reduction of the systolic peak to peak pressure gradient from 59.8±24.8 mmHg to 21.5±13.0 mmHg ($p<0.001$), the instantaneous systolic pressure gradient from 74.70±22.54 mmHg to 30.85±16.3 mmHg ($p<0.001$). During follow-up a further reduction of the instantaneous pressure gradient to 19.9±15.7 mmHg ($p<0.001$) could be demonstrated. An excellent short and midterm result was shown in 86/96P (= 89,6%), 10/96 P (= 10,4%) needed a reintervention. A second BVP was performed in 3 P and showed a good result. The other 7 P underwent an operation due to a BVP failure. Risk factors for an unsuccessful intervention were: the presence of a supravalvar pulmonary stenosis ($p<0.001$), the severity of dysplasia of the pulmonic valve ($p<0.001$) as well as the absence of a poststenotic dilatation ($p<0.001$). Age was not a risk factor for a reintervention.

Conclusions: The balloon valvuloplasty of the congenital pulmonary valve stenosis shows an excellent short and midterm result. Risk factors for a balloon valvuloplasty failure were the presence of a supravalvar stenosis, a pronounced valvar dysplasia as well as the absence of a poststenotic dilatation.

ACUTE THROMBOPHLEBITIS OF BOTH FEMORAL AND ILIAC VEINS 5 YEARS AFTER REPAIR OF TOTAL ANOMALOUS PULMONARY VENOUS RETURN.

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A 5 year old boy ,who had had repair of total anomalous pulmonary venous return to the portal vein at 5 weeks of age, is hospitalised for an RSV and Mycoplasma pneumonia. On the sixth day of hospitalisation he complains of pain in his right thigh. The thigh is red and swollen and he limps upon walking. There is a leucocytosis at 25.8 G/l without left shift and the CRP is 89 mg/l. Coagulation studies, cardiolipines, IgG, IgA, IgM and protein S are normal. An angio MRI of the legs showed bilateral femoral and external iliac thrombosis. The IVC was free of thrombosis. He was placed on iv heparine then on dicoumarine with good clinical response and complete repermeabilisation of his venous axis within a few weeks.
He had undergone 2 cardiac catheterisations, the first at 1 month of life through right femoral venous puncture, the second at 15 months of life through puncture of the left femoral vein.
Conclusions: complete partial thombosis can be a complication of cardiac catheterisations through femoral venous puncture in infancy. The veins tend however to repermeabilise with time. In case of a hypercoagulable state (for example Mycoplasma infection in our patient), alterations in the venous walls at the sites of puncture or on the routes of the catheters can favor the development of new thombosis or thrombophlebitis.

Holter recording BEFORE AND after CISAPRIDE (C) IN infants

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Objective: To study prospectively the effects of C on heart rhythm using 24 hour ECG recording in term (T) and preterm (P) infants. Methods: We studied 31 subjects with gastroesophageal reflux disease (ALTE, apneas, bradycardias) before and three days after starting cisapride (0.8 mg/kg/day in 4 doses). Were recorded: Standard ECG for determination of corrected QT interval (QTc, ms) and QT dispersion (QTD, ms), and Holter 24h recording using a Marquette digital device. Results (table): 17 P (gestational age 28 to 36 weeks) and 14 T subjects were studied at a median chronological age of 29 days. Subjects were free of electrolyte disturbances and were not receiving medications recognized to interact with C. Maximal and mean heart rate (HR) decreased after C in all subjects, whereas minimal HR increased in preterms. In two preterm infants, both with normal QTc and QTD, a significant increase in supraventricular extrasystoles (37 to 860) occurred after C in one, and ventricular extrasystoles (0 to 148) in the other.

Mean ± SE	Preterm (n=17)		Term (n=14)	
	before C	after C	before C	after C
QTc	408 ± 7	433 ± 7*	400 ± 11	410 ± 6
QTD	31 ± 3	27 ± 3	30 ± 3	31 ± 2
maximal HR	213 ± 4	205 ± 4*	218 ± 3	206 ± 3†
mean HR	158 ± 2	149 ± 2*	145 ± 3	138 ± 3†
minimal HR	66 ± 5	76 ± 5*	86 ± 3	85 ± 2

*†paired t test, before vs. after C in P(*) and T(†): $p<0.05$
Conclusion: C significantly affects the peak (T+P), mean (T+P) and minimal HR (P). No relevant arrhythmia were documented however in preterms a proarrhythmic drug effect can not be excluded even in the presence of a normal QTc/QTD.

Lethal neonatal hypertrophic cardiomyopathy presenting with wolff-parkinson-white syndrome (WPW).

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Introduction : Neonatal hypertrophic cardiomyopathy (HCM) is commonly presents in infants of diabetic mother. HCM of other etiology are rare. We discuss a case of progressive biventricular HCM, with development of an associated Wolff-Parkinson-White syndrome, which opens a larger differential diagnosis at the age of presentation.
Case report : First child of unrelated parents. Mother G2P1, known for systemic hypertension subsequent to chronic renal failure and presenting at 22W gestation with severe ketoacidosis as inaugural manifestation of gestational diabetes. Delivery was by a cesarean section at 34W, because of fetal heart failure. The cardiogenic shock was stabilized under standard treatment. A bulky biventricular hypertrophia was the striking feature, which was rapidly worsening. Severe rhythm disturbances on an intermittent WPW complicated the course of the disease. Although a skeletal muscular biopsy showed a glycogen overload, the extensive work-up for a storage disease was normal. The infant died at 8 weeks of age due to terminal cardiac failure. Autopsy revealed renal microcysts and a massive HCM. On histology giant mitochondriae were demonstrated in myocardium and renal parenchyma, as a mutation in the mitochondrial DNA was documented on cultured fibroblasts, strongly suggesting a diagnosis of mitochondrial disease.
Conclusion : neonatal HCM is usually self-limited, when consecutive to gestational diabetes. In case of an evolutive disease, associated with rhythm disturbances and/or WPW, another etiology has to be suspected. This latter situation warrants extended workup, with multiple biopsies, in particular of the myocardium, in order to establish a final diagnosis.

DEVELOPMENT OF REST-ACTIVITY RHYTHMS DURING THE FIRST YEAR OF LIFE

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The development of rest-activity rhythms during the first year of life reflects maturational processes of infant sleep-wake behavior. The aim of this study was to determine the temporal evolution of rest-activity patterns by monitoring motor activity with actigraphs worn on the left ankle. Six healthy full-term infants were continuously recorded from the first days of life until 2 (n=1), 6 (n=4) or 12 months of age (n=1).
Mean 24h activity profiles of consecutive 30 day intervals and periodogram analysis of 10 day intervals revealed clear ultradian (4, 6 and 8h) and diurnal (24h) components. The 24h day-night rhythm was the strongest component in all infants and was present already in the first weeks of life. However, it appeared later in first born infants compared to later born infants. An 8h ultradian period indicated high activity in the morning and afternoon with a midday rest phase.
The development of miniaturized actigraphs has provided an ideal tool to perform long-term longitudinal studies of rest-activity patterns in infants. Ultradian and diurnal periodicities develop early after birth and are influenced by the social environment.

Epileptic falls and gait disturbance: a variant of benign partial epilepsy of childhood ?

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Epileptic falls usually occur in severe epileptic syndromes with multiple seizure types (myoclonic astatic epilepsy, Lennox-Gastaut syndrome), but can also be observed in isolation in a normal child.

We describe 2 boys with normal development, evaluated since 18 months and 2 years of age for gait disturbance and repetitive falls. History (home video) and clinical examination revealed jerks and drops of legs and trunk, suggestive of epilepsy. Their EEGs showed a midline epileptic focus, activated by sleep. Brain MRIs were normal. Their course was fluctuating, with seizures remission since 4 year old in case 1, and still active epilepsy at 4.5 year old in case 2 (transient effect of valproate, sustained effect of ethosuximide).

The jerks, drops and falls represent increasing severity of myoclonic and/or focal atonic seizures (negative myoclonus), predominantly affecting one or both legs, disturbing gait. Epileptic discharges at the midline might directly activate or inhibit mesial motor or premotor cortex, that control inferior limbs movements, posture and gait. The normal neurologic examination and certain EEG features suggest in these 2 patients a variant of benign partial epilepsy of chidlhood.

TREATMENT OF ACNE WITH CONSEQUENCES – PSEUDOTUMOR CEREBRI DUE TO HYPERVITAMINOSIS A

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Background: Pseudotumor cerebri (PTC) is an entity characterized by elevated intracranial pressure of probably multifactorial origin, but most of the cases remain idiopathic. Hypervitaminosis A as a causative agent in children is known, but only a few cases were published. **Case report:** A 15 years old girl complained of headache for 5 weeks, dizziness, nausea and epistaxis for 3 weeks and acute diplopia and blurred vision were reported. Physical examination revealed a reduced general condition, cracked lips, abducens paresis, papilloedema and impaired visual acuity. To treat her acne, she had taken Arovit® (Retinol) 200000 IE Vitamin A_1/die orally during 6 month until 3 weeks before admission. An intracranial mass was excluded and a lumbar puncture revealed an opening pressure of 62 cm H_2O (Norm <15 cm) with normal cell counts and chemistries. Because of a hypercalcaemia of 3.24 mmol/l (Norm 2.1-2.7) hypervitaminosis A was suspected, and confirmed by an increased level of vitamin A 1.8 mg/l (Norm 0.3-0.7). The patient was treated with lumbar punctures and acetazolamide and became clinically asymptomatic. The elevated serum levels of vitamin A and calcium returned normal within 2 month. **Conclusion:** PTC due to hypervitaminosis A is a serious complication, which can cause permanent visual loss. Patients treated for acne with retinoids require proper surveillance. The elevated serum level of retinoids after withdrawal can persist for weeks.

PARENTAL ANXIETY AND FEBRILE SEIZURES – IMPORTANCE OF INFORMATION

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Purpuse: The aim of the study is to investigate febrile seizures (FS) and analyse the effect on parents in order to improve our attitude towards them in the future.
Methods: Our retrospective study (questionnaire) with parents of 213 patients was evaluated with the Fisher's Exact Test and the Logistic Regression Model.
Results: We evaluated 135 questionnaires. At the first FS 121 parents (91%) admitted severe anxiety. In 69% anxiety was that strong, the parents believed their child would die. To 44% of the parents FS were unknown. In 79% of these parents, anxiety was on the highest level in comparison with 59% in the group that knew about FS. Severe anxiety was strongly associated with not knowing about FS. Although 66% of the parents were reassured by the first information, 87% of them actually still are afraid of new seizures.
Conclusions: Our study shows that the knowledge about FS is still insufficient among concerned parents. The strong association between severe anxiety and knowledge about FS proves that parental information before the first FS is necessary for anxiety reduction. We suggest to provide preventive education about FS before the age of the main frequency of FS and repeated information after the first seizure, preferably adjusted to the psycho-social background.

CONTINUOUS SUBCUTANEOUS GLUCOSE MONITORING IN CYSTIC FIBROSIS

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Background: There has been a dramatic increase in the survival of people with cystic fibrosis (CF). CF-related diabetes mellitus has evolved as one of the complications and the prevalence and incidence is raising. Annual use of the Oral Glucose Tolerance Test (OGTT) as a screening method for diabetes is standard in patients with CF aged above 10 years in most of the paediatric centers.
Patient: 14 y old girl with CF was screened for diabetes with the OGTT and was diagnosed with diabetes. In contrast the continuous subcutaneous glucose monitoring (CSGM) measurements over 72-hours showed in over 800 blood glucose values only one glucose level above 11.1 (11.2) mmol/l.
Discussion: The results of the CSGM in CF patients can be easier analysed and interpreted in respect to the patients clinical condition than the OGGT. With this information the regimen can be tailored to the specific patient needs (diet, medication).
Conclusion: The CSGM is a valuable alternative to the OGGT as the annual screening procedure for diabetes in CF patients.

Positive Effect of a training program on lung function and aerobic capacity in CF

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Background: Regular physical activity might improve aerobic capacity (Wmax) and might lessen the decrease in lung function in CF.
Method: 28 patients were divided into 3 groups: strength training (KT, n=10), endurance training (AT, n=10) and controls (KO, n=8). KT and AT performed a supervised training of 3x30 min./week. Before, after 3 and 6 months lung function and Wmax were measured.
Results: FVC and FEV1 increased in KT compared to KO. AT showed an increase in FVC, FEV1 and Wmax compared to KO. KT decreased RV/TLC compared to AT. AT lead to an increase in Wmax compared to KT (all $p<0.05$).
Conclusion: Endurance and strenght training lead to an improvement in lung function and aerobic capacity in CF. Training should be part of every CF patient's treatment regimen.

TREATMENT OF CHRONIC HEPATITIS B IN CHILDREN WITH A COMBINATION OF LAMIVUDINE AND α-INTERFERON IS NOT BETTER THAN WITH α-INTERFERON ALONE

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Background: Treatment of chronic hepatitis B (CHB) in children with α-interferon (IF) alone gives unsatisfactory results with seroconversion of HBe antigen in only about 20-30% of patients.
Aim: To improve seroconversion rate by combining the nucleoside analogue lamivudine with IF in the treatment of CHB in children.
Methods: From 1999-2000 12 children with CHB were treated with lamivudine 4mg/kg once daily for 1 month, followed by subcutaneous IF 5MU/m^2 body surface, 3x per week for 6 months. CHB was defined as presence of HBsAg, HBeAg and HBV DNA in serum for > 6 months. Study endpoint was the loss of HBeAg and HBV DNA and the appearance of HBeAb. The results of these 12 children were compared with the results of 28 children treated with IF alone from 1992-97.
Results: The two groups of children were comparable regarding age, sex and ethnical origin. Seroconversion rate in the group of children treated with IF alone was 17.8% compared to 25% in the group treated with the combination of lamivudine and IF ($p=0.6$).
None of the 8 children who responded to one of the two treatments showed loss of HBsAg during treatment. HBV DNA load was significantly reduced after 1 month of treatment with lamivudine, but increased again in the non responders under treatment with IF. No side effects of lamivudine were observed.
Conclusion: Reduction of hepatitis B viral load with lamivudine before treatment with IF doesn't improve the rate of seroconversion compared to the treatment with IF alone.

Cystic fibrosis (CF) and family planning: from adoption to reproductive medicine

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Motivation: Male and female CF patients frequently suffer from infertility. Illustrated by three clinical cases we present different options for the family planning process.
Case descriptions: Pat.1: 37-yrs. old patient (ΔF508/1717-1G→A) with normal weight, actual FEV1 of 50.3% and CF-related diabetes mellitus. After documentation of azoospermia adoption of two children, 4 and 2 years ago according to formal regulations in Switzerland. Pat.2: 28-yrs. old patient (compound-heterozygous for G542X/deletion intron 1 to 3) with mild underweight. First pulmonary symptoms at age 18 years. Partner non-heterozygous for CF. FEV1 in 8^{th} week of gestation: 53.1%. Cesarean section in 34^{th} gestational week for maternal reasons. Pat.3: 34-yrs. old patient (ΔF508/1717-1G→A) with normal weight, and actual FEV1 of 93.9%. After documentation of azoospermia, pregnancy is induced by intracytoplasmic sperm injection (ICSI) after exclusion of a CF-heterozygous status in the female partner. So far the course of the pregnancy is without any problems.
Conclusions: Today CF patients have various options for their family planning. In every situation individual counseling is essential, as there is still a lack of solid prognostic long-term data on life expectancy and pregnancy

pH monitoring in the distal and proximal oesophagus in symptomatic infants

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Objective: Standard distal esophageal pH monitoring data are sometimes within normal ranges in children with clinically suspected gastro-oesophageal reflux disease(GERD). Therefore, we hypothesized that the amount of acid reflux reaching the proximal esophagus may be higher in some subgroups of patients than in healthy controls or in other subgroups of patients. **Methods:** The parameters of a 24 hours pH monitoring in the proximal part of the esophagus were analyzed in 120 symptomatic infants (mean age 4.5 mo, range 0.5-17) in whom the reflux parameters in the lower esophagus were clearly within normal ranges. The infants were divided in four patients groups : excessive regurgitation (n :41) ; inconso-lable crying (n :31) ; apparent life threatening event (ALTE, n :18) and chronic respiratory disorders (CRD, n :30). The control group consisted also of 120 infants. **Results** : The patients with CRD were significantly older than the other patients groups and the controls. In the distal esophagus, there was no statistically significant difference between the reflux parameters. As could be expected, every parameter was statistically significantly smaller in the proximal than in the distal esophagus (paired t-test, Wilcoxon test, one-way ANOVA, Mann-Whitney test). The number of reflux episodes in the upper esophagus was significantly higher in the group with CRD than in the other patients groups and controls. **Conclusion**: Our results do not support an advantage of dual simultaneus esophageal pH monitoring in the distal and proximal esophagus in infants, especially not in infants presenting with ALTE, excessive regurgitation or inconsolable crying. In the subgroup of patients with CRD, the number of reflux episodes reaching the proximal esophagus with normal distal pH monitoring data may be increased. But more data are needed.

The use of alternative Medicine by children with asthma

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Epidemiological data on the use of alternative medicine (AM) in children are scarce. Our aim was therefore to determine the prevalence of AM in Swiss children with asthma and to assess possible interactions with conventional therapy.
A questionnaire on respiratory symptoms and use of alternative and conventional medicine was sent in 1998 to all members of the Swiss Association of Parents of Asthmatic and Allergic Children (SEAAK).
786 questionnaires (85%) were returned, 739 (aged 1 to 20 years) were analysed. Asthma severity was comparable to a random sample of wheezing children. 221 children (30%) used AM, applied by a physician (12%) or an AM practitioner (18%). Most children (24%) used only one method, most commonly homeopathy (18%), bioresonance (5%), kinesiology (3%) and acupuncture (1%). AM use was predicted by higher maternal education and age between 4 and 12 years, but not by gender nor by asthma severity. Adjusting for age and asthma severity, children receiving AM reported similar use of bronchodilators, but significantly lower use of inhaled steroids (odds ratio 0.41, $p<0.0001$), more frequent visits to the physician ($p<0.01$), more chronic symptoms (61% vs. 46%, $p=0.04$) and more severe asthma attacks (22% vs. 13%, $p=0.07$), compared to non users.
A substantial proportion of Swiss children with asthma use alternative medicine, usually in combination with conventional asthma therapy. AM use is associated with lower use of inhaled steroids and unsatisfactory asthma control.

Pilote-recherche about the enhancement of the quality of life in asthmatic children : evaluation of the program of the Centre de Médecine d'Exercice of the SSJ in Genèva.

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This study has shown the clinical and evaluative pertinence of the use of a health related quality of life (HRQL) measure called Paediatric Asthma Quality of Life Questionnaire (PAQLQ) (Juniper 1997). These data have pointed out the functional health of asthmatic children. The measure of quality of life was also used to evaluate the multidisciplinary program of the CME. The children taking part in the program have a medical follow-up and have gymnastics courses adapted to their pathology.
In our study, 15 children (age 11,46 +/-2,37) took part in the program and were compared to a paired group (age 11,25 +/-2,74). It was shown that a regular adapted sports activity reduces the inadaptation to physical activities and increases physical performances, the quality of the social and emotional life and the quality of life of asthmatic children.
Comparing the two groups a significant difference in the quality of life was observed confirming the efficacy of the program.

ESOPHAGEAL DUPLICATION: A RARE CAUSE OF TRANSIENT ACUTE RESPIRATORY DISTRESS IN CHILDEN

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Introduction*:* Esophageal duplications are rare congenital anomalies and may present at any age with non-specific respiratory or gastrointestinal symptoms. They are also known to cause severe respiratory distress by compression of the trachea and the main bronchi. **Case history:** We report a 7-year old boy who complained of recurrent episodes of acute respiratory distress since the age of 4 years. Respiratory symptoms were transient and accompanied by upper respiratory tract infections. The initial suspicion of asthma was supported by positive allergy testing against house dust mites and a good response to treatment with inhaled bronchodilators. A chest radiograph taken because of recurrent atypical symptoms revealed a mediastinal mass. Further work-up by CT scan lead to the differential diagnosis of a bronchogenic cyst or an esophageal duplication. Surgical resection was performed without complications and the esophageal duplication was confirmed by pathology.
Conclusions*:* Chest radiography is an important part of the initial work-up of recurrent or atypical wheezing in children, because not all cough and wheezing is asthma. Prompt surgical resection is the treatment of choice for bronchogenic cysts or esophageal duplications to prevent possible complications such as infection or obstruction of the esophagus or the large airways.

Community acquired pneumonia in children: are WHO guidelines applicable in Switzerland ? Preliminary study and perspectives.

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When faced with a respiratory tract infection (RTI) in a child, the practitioner must answer 3 questions: 1. Is it pneumonia and does it require antibiotic treatment ? 2. Does the child require hospitalisation ? and 3. Is a blood sample and/or a chest X-ray useful in order to answer the first 2 questions ? WHO promotes an algorithm based on careful examination of 5 clinical parameters: respiratory rate (the absence of tachypnea ruling out pneumonia), nasal flaring, cyanosis, feeding difficulty and absence of wheezing. Without investigations, one can thus classify cases according to 3 stages of severity each corresponding to an antibiotic treatment, either ambulatory or in hospital.
Subjects and methodology: we have followed a similar decisional process at l'HEL. All children between 2 months and 5 years of age who presented with a RTI during the winter 99-00 were examined; only those with at least tachypnea (stage 1 pneumonia) were enrolled. All children with wheezing were excluded. All children with mild (stage 1), moderate (stage 2) and severe (stage 3) pneumonia underwent chest X-rays. Only those with moderate (stage 2) and severe (stage 3) pneumonia underwent a blood sample (white cell count, C-reactive protein (CRP) and blood culture).
Results: 50 children were enrolled. All had a favourable outcome. The chest X-ray confirmed the diagnosis of pneumonia in 95% of the children and showed pleural effusions in 25% of cases. 3 patients who were initially treated with a normal chest X-ray had an abnormal chest X-ray 24 hours later. At the time of diagnosis, 22 patients (43%) had leucocytosis of more than 15 000 G/l, 34 (68%) had a CRP greater than 60 mg/l and 3 (6%) had positive blood cultures (each time for Streptococcus pneumoniae). 15 patients (30%) had Respiratory syncythial viral infections. All other etiologic investigations including mycoplamsa were negative.
Discussion:1. The application of the WHO algorithm is possible in Switzerland; tachypnea is the cardinal clinical sign.
2. Blood samples are not useful in the initial clinical workup.
3. The chest X-ray has a good sensitivity but can be normal at the beginning of the disease in which case the antibiotic treatment should not be delayed if the diagnosis is clinically suspected.
4. A large scale study must be undertaken to validate this process.

HYPERSPLENISM AS COMPLICATION DURING ALL-TREATMENT

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Background: The peripheral pancytopenia caused by hypersplenism impedes an effective leukemia treatment. Case report: A 7 ½ year old girl fell ill of ALL (HR-group) in June 99 (initial Hb 109 g/l, Lc 25,2 G/l with 74 % atypical cells, Tc 10 G/l; spleen 10 cm below costal margin in MCL, liver 6 cm below costal margin in MCL). During the induction according to ALL-BFM 95 there was massive bleeding after port-à-cath-implantation caused by persisting thrombopenia, so that multiple thrombocyte-transfusions were necessary. In the following months (HR-blocks) weekly transfusions of erythrocytes and thrombocytes were given. After remission of ALL hepatosplenomegaly persisted. The frequency of transfusions increased and therapy had finally to be stopped. A radiation treatment of the spleen with 330 cGy was unsuccessful (spleen volume after radiation 1200 ccm), so that a splenectomy was performed after successful vaccinations (HiB,Strept. pneumoniae) in Sept. 2000 (histology: extended bleeding sequelae, aggregats of foam cells and extramedullary hemopoiesis; weight: 578 g). Since then, therapy could be continued according to study protocol without transfusions, moderate hepatomegaly and reasonable good health of the girl.
Conclusion: Treatment of ALL according to study protocol with hypersplenism is possible after total splenectomy. Other treatment modalities are not found in the literature.

LOCALIZED PEDIATRIC PRIMARY SKELETAL NON-HODGKIN'S LYMPHOMA, AN OVERTREATED SUBGROUP WITH AN EXCELLENT PROGNOSIS?

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Background: Non –Hodgkin's lymphoma (NHL) is a common highly malignant disease in children involving the mediastinum and abdomen. The skeleton is the primary tumor site in only 5% of pediatric patients with NHL. The major complaint is bone pain often in absence of other symptoms. Bone biopsy is the main diagnostic tool. The management of these tumors includes chemotherapy, radiotherapy and surgery. Recently published studies tend to show that chemotherapy alone is sufficient to achieve a complete and lasting remission. Prognosis of localized skeletal NHL seems to be excellent with an event-free survival of > 90% at 5 years.
Case: We report a 15-year-old boy suffering from constant bone pain in the right hip for several months. Clinical and laboratory investigations were normal. Osteomyelitis in the proximal femur has been suspected according to bone scan and MRI. Bone biopsy was performed and revealed a diffuse large B-cell-lymphoma (REAL-classification). Staging investigations revealed no other skeletal sites and no evidence of disseminated disease. The boy underwent chemotherapy according to the NHL-BFM 95 protocol for a duration of 3 months. Bone biopsy after treatment showed complete remission. Follow-up examinations including MRI for almost two years after completion of chemotherapy exclude a relapse. There are no residual symptoms.
Conclusion: Primary skeletal NHL represents a rare but mostly curable tumor in children and adolescents. Although a malignant high grade lymphoma , primary skeletal NHL seems to have been overtreated. Prognosis is excellent and radiotherapy probably does not improve event-free survival. Bone pain in children and adolescents is of different origin. Diffential diagnosis must always include malignant diseases.

Myelodysplastic Syndromes in Childhood: A diagnostic Challenge

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Myelodysplastic syndromes (MDS) in childhood are rare clonal stem cell disorders. Characteristic is a protracted course of cytopenia or pancytopenia with disturbed hematopoiesis. In most cases, an acute myelogenous leukemia typically will develop within months to years.
Cases: Since August 1999 we have diagnosed seven children with MDS, age range 0-15 years (3 refractory anemia (RA), 2 with myelofibrosis; 2 RA with excess blasts (RAEB); 2 RAEB in transformation (RAEB-t)). The initial findings were thrombocytopenia in 5 cases and one severe neutropenia, together with mild changes of other cell lines. Four of the children initially presented with either ITP (3 children) or chronic neutropenia (1 child). The 2 patients with RAEB-t had cytogenetic findings (-7 or +21). Three children have received an allogeneous HLA-identical stem cell transplantation. Presently, two are in remission (6 and 2.5 months) and the other patient, with RAEB-t, relapsed after 7 months.
Conclusion: The diagnosis of MDS in childhood is often made late during the disease course. MDS must be excluded in cases presenting with chronic cytopenias and atypical findings, such as an involvement of a second hematopoietic lineage. The therapy of choice is stem cell transplantation where 50% disease free survival can be expected.

MYC mRNA EXPRESSION PREDICTS SURVIVAL OUTCOME IN CHILDHOOD MEDULLOBLASTOMA (MB)

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To identify biologic prognostic factors in childhood MB, we determined expression levels of MYC mRNA in fresh frozen tumor samples from 26 MB patients utilizing semiquantitative RT-PCR. MYC mRNA expression levels were compared with clinical variables, biological variables (TrkC mRNA expression, GFAP expression) and survival outcomes in univariate and multivariate Cox regression analysis. MYC mRNA expression levels in primary MB showed a wide range with a 22-fold difference between the highest and lowest values and did not correlate with *MYC* gene amplification. MYC mRNA expression was an independent significant prognostic factor for progression-free survival outcome and more predictive than standard clinical factors. The combination of low MYC mRNA expression and high TrkC mRNA expression identified a good-outcome group of MB patients (n=7) with 100% progression-free survival after a median follow-up time of 55 months (range, 15 - 91 months). Three of these 7 good-outcome patients survived without radiotherapy. Accordingly, low MYC mRNA expression is a powerful independent predictor of favorable clinical outcome in MB. The combination of low MYC mRNA expression and high TrkC supports the existence of a good-outcome group of MB patients for whom substantially less toxic therapeutic strategies might be employed.

METASTATIC OSTEOSARCOMA AND MULTIPLE LUNG RESECTIONS: A LONGTERM FOLLOW UP

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Background: The existence of pulmonary metastases in patients with osteosarcoma represents a poor prognosis. We report a $11^1/_2$-year-old girl who underwent multiple lung resections. **Methods:** Case report and review of the literature as basis for repetitive thoracotomy. **Case report:** In September 1997 a $11^1/_2$-year-old girl was admitted with a high-grade metastatic osteosarcoma of the proximal metaphysis of the right tibia with bilateral pulmonary metastases (6 on the left and 3 on the right side). She was given combination therapy according to the protocol COSS-96 with neoadjuvant and adjuvant chemotherapy, tumorresection and bilateral thoracotomy for resection of metastatic lesions. Chemotherapy response assessed on the tumor sample according to Salzer-Kutschik revealed a grade III regression (< 10% vital tumor cells). Six months after completion of therapy a relapse of pulmonary metastases was found on the left side. Two metastases were resected and an adjuvant chemotherapy with carboplatin and etoposid was given. Twelve months later a second pulmonary relapse was diagnosed and a single metastasis was resected on the right side. After 5 months the third pulmonary relapse with a single metastasis was found and resected on the right side. Complete remission was maintained during 16 months. The multiple lung resections were well tolerated and the patient lives without affecting pulmonary function with normal school attendance. **Conclusions:** Multiple resections for recurrent pulmonary metastases of osteosarcoma appear to correspond to a curative management with an acceptable morbidity. However, clinical trials are needed to prove this observation.

Stem Cell Transplantation (SCT) in 22 patients with Chronic Granulomatous Disease (CGD)

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CGD is the most frequent inborn phagocyte function defect with a mortality of 2-5% per year, despite regular therapy with antibiotics and granulocyte transfusions. Causes of death are severe fungal/bacterial infections and secondary organ insufficiencies due to granulomatous inflammation. SCT is curative, but not yet fully accepted. The outcome of 22 transplants (20 HLA-genoidentical, 2 unrelated donors; 20 myeloablative, 2 myelosuppressive) performed in 11 European centers was analyzed according to risk factors present at time of SCT.

Patients with fungal infections refractory to treatment: 6 pat were transplanted under ampho B/granulocyte coverage. 3/6 were cured from both CGD and infection. 3/6 died from pulmonary inflammatory reactions at take and/or from graft-versus-host disease grade IV.

Pat with severe pulmonary restriction following recurrent pneumonias: 5/5 pat were cured from CGD and have improved/normalized their lung function (pulmonary restriction, clubbing, oxygen dependency)

Pat without the above risk factors: 11/11 pat were cured from CGD and are alive and well.

Conclusion: SCT for CGD is curative in 16/16 (100%) pat without risk factors or with pulmonary restriction only, but remains experimental in patients with uncontrolled fungal infection. SCT should be considered early in pat with severe clinical course, if an HLA genoidentical donor is available.

Successful therapy in a child with relapsed B-ALL with anti CD20 antibodies.

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The survival rate of highly aggressive B-cell ALL and the related Burkitt lymphoma in childhood has dramatically improved within the last 20 years and approx. 75%-80% of children will survive with intense chemotherapy. Relapses usually occur within a few months after end of treatment and are often fatal. **Case Report:** A 12-years old boy with a mature B-ALL, t(8;14), CD20 positive and CNS involvement, was treated according to the NHL-BFM 95 protocol. He achieved a prompt remission but suffered bone marrow relapse immediately following end of therapy. Two blocks of FLAG chemotherapy (Fludarabine, ARA-C and G-CSF) failed to achieve a 2nd remission. Following one intravenous infusion of the single agent Mabthera, a chimeric human/mouse antibody against CD20, a complete remission of the bone marrow was achieved. After three further infusions with Mabthera an isolated CNS relapse was diagnosed three months later. A repeated course of intrathecal and systemic chemotherapy combined again with Mabthera lead to a 3rd remission that was subsequently followed by an autologous stem cell transplantation (conditioning regimen with Bu/Cyclo and Mabthera). Currently (02/01) the patient is in complete remission 3 months after CNS and 8 months after bone marrow relapse respectively. **Discussion**: To date, it has been unusual to achieve a long lasting second remission after relapsed B-cell leukemia /lymphoma. The new antibody Mabthera is also effective in relapsed B-ALL in childhood. The application of an anti CD20 antibody labeled with a radioactive component might even increase the anti tumor effect in such a disease.

EOSINOPHILIC FASCIITIS: RARE DISEASE IN THE DIFFE-RENTIALDIAGNOSIS OF PERSISTING EOSINOPHILY

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Motivation: Eosinophilia can be observed in many deseases. The eosinophilic fasciitis is a very rare disorder. Diagnosis and management are discussed based on a case report of an infant. **Methods:** case report and review of the literature.

Results: a 12 month-old girl presented eosinophilia since 2 month, with a hyperpigmentation, a maculouse exanthem and edema of her hands and ankles as well as therapy resistant pain of her extremities. The patient was admitted to assess the eosinophilia. The highest value of the eosinophiles was 30.88×10^9/l. Infectious causes were excluded. The bone marrow aspirate revealed a dysplastic megakaryopoiesis with normal morphology of the other cell rows. Acute leukaemia was excluded. A myelodysplastic syndrome could not be excluded completely. With time the swelling of the soft tissue increased. An ultrasound and an MRI showed a cellulitis, fasciitis, and perimyositis, which may suggest eosinophilic fasciitis. The diagnosis was confirmed by biopsy. A treatment with Prednisolon (2mg/Kg/day) resulted in an improvement of her general condition and pain and soft tissue swelling disappeared. **Conclusions:** Eosinophilic fasciitis is a desease of unknown origin affecting the collagen, characterized by a painfull induration and edema oft soft tissues, sometimes accompanied by a severe eosinophilia. According to the literature the desease develope in a localized sclerodermy in aproximatly 2/3 of the patients. The younger the patient the wors the prognosis. Similar cases are very seldom in the literature. To our knowledge a child of the age of our patient has never been reported.

SUCCESSFULL STEM CELL TRANSPLANTATION (SCT) IN 2 CHILDREN WITH PURINE-NUCLEOTIDE-PHOSPHORYLASE (PNP)-DEFICIENCY

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PNP-deficiency is an autosomal recessively inherited disease of the purine metabolism. Accumulation of toxic metabolites, e.g. Deoxy-GTP, leads to progressive combined immunodeficiency, autoimmunehaemolytic anaemia (AIHA) and a varying neurologic disorder. Stem cell transplantation (SCT) is the only curative therapy. However, SCT`s performed world wide so far had a high transplantation related mortality. 2 children (2 y und 8 mo) with complete PNP-deficiency were treated for the first time using a myelosuppressive conditioning regimen with ATG, Melphalane and Fludarabine. One had a haploidentical, the other one an HLA-phenoidentical SCT with maternal stem cells. Chronic Rotavirus- and RSV-infections, mucocutaneous candidiasis, AIHA and RSV, CMV-infections, respectively, were complicating both transplantations. The patient with the haploidentical SCT (10 months after SCT) has a mixed donor chimerism (50%) with PNP-concentrations of 25 % of normal (the donor was heterozygous for the disease). The AIHA could be successfully managed with anti-CD 20 monoclonal antibody treatment. The other (8 mo after SCT) has got an almost complete donor chimerism after donor lymphocyte infusions (95 %). In both patients, immunological reconstitution after SCT is normal. **Conclusion:** Flu/Melph/ATG is a mild and efficient conditioning regimen in multiply infected patients with PNP-deficiency. The reversibility of neurological disease after SCT has to be carefully investigated in the future.

BEHAVIOUR DISTURBANCES, CONVULSIONS AND HYPERINSULISM IN A TEENAGE GIRL.

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In teenagers, behaviour disturbances are common and rarely due to organic causes. In this age group, convulsions are usually explained by neurological disorders, so that glycemia (calcemia) may be underevaluated / forgotten at admission. CASE : a 14 yr old teenage girl is referred for headaches, behaviour disturbances with verbal agressivity, automatic responses and periods of memory loss ; episodes of pallor, fatigue and dizziness were also described. Based on EEG bradydysrythmia, valproate therapy was started, without improvement. Therapy discontinuation was followed by 2 epileptic fits, one generalized, the other partial simple with left motor hemiyndroma (EEG recording) ; glycemia (3.2 mmol/l) was missevaluated. Therapy was resumed using phenytoin : behaviour disturbances did not improve so that psychotherapy was initiated. One year and a half after onset, the mother reported that symptoms vanished following intake of sugary beverages : a fasting test with repeated insulin/glucose determinations showed hyperinsulinism but without pancreatic lesion (abdominal IRM and CT). Awaiting labelled octreotide scintigraphy and arteriography, diazoxide therapy (~10 mg/kg/d) led to clinical and metabolic normalization. CONCLUSION : hypoglycemia should be suspected on the early signs of neuroglucopenia (undue agressivity, concentration and/or memory difficulties) mimicking common behaviour disturbances. These signs may precede adrenergic signs of hypoglycemia (tachy-cardia pallor, etc). Unrecognized, hypoglycemia proceeds to un-specific neurologic pictures inducing, in turn, diagnostic and therapy errors.

EARLY CARE OF MULTIPLE ENDOCRINE NEOPLASIA 2A (MEN 2A) IN THE CHILD.

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MEN 2A is caused by mutations of the RET protooncogene (chr. 10, exon 10 or 11), comprising medullary carcinoma of the thyroid [MCT] (100%), pheochromocytoma (50%) and hyperparathyroidism (5-20%). CASES : the mutation of exon 11 (codon 634) has been detected in the mother of 4 children ; thyroidectomy was performed 8 years ago for CMT. Among her 4 asymptomatic Children aged 13 yrs (C1), 11 yrs (C2), 5 yrs (C3) and 3,5 yrs (C4), three (C1, C2, C4) have this mutation. Pheochromocytoma and hyperparathyroidism have been ruled out. The results of the pentagastrin test led to early thyroidectomy in these 3 cases :

	Basal calcitonin	Peak calcitonin	Histopathology
	(no < 10 ng/l)	(no < 30 ng/l)	
C1	7	247	under way
C2	40	856	in situ carcinoma
C4	7	81	under way

Peak calcitonin levels over 100 ng/l indicate at least in situ CMT and therefore total thyroidectomy with avec extended lymph node excision (C1, C2). Between 30 and 100 ng/l (C4), there is C cell hyperplasia implying total thyroidectomy with lymph node biopsy.
CONCLUSION : in keeping with recent reports, total thyroidectomy should be performed early in childhood in mutated cases. The extent of lymph node excision is based on the results of the pentagastrin test. This strategy improves survival rate at 10 yrs (80% vs 30% in cases without metastases). Cases without mutation do not need endocrinological follow up.

WHY MEN 2A HAS BECOME A PEDIATRIC PROBLEM : CASE REPORT AND REVIEW.

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Background: MEN 2A is a familial cancer syndrome with the triad of medullary thyroid carcinoma (MTC), pheochromocyto-ma and hyperparathyroidism. The molecular basis of the MEN 2A are missense mutations in the RET proto-oncogene, transmitted in an autosomal dominant manner. The identification of these specific mutations allows detection of gene carriers in kindred of MEN 2A families for whom total thyreoidectomy at 5 years of age is recommended to prevent development of MTC.
Objective: To evaluate evidence for therapeutic recommendations.
Methods: Case report and review of the literature for pediatric patients. Inclusion criteria: age <21 years, histological degree of C-cell-disease (normal=N, C-cell-hyperplasia=CCH, medullary thyroid carcinoma=MTC, metastatic MTC=MMTC).
Results: In 122 cases (own case plus review) median age at thyreoidectomy was 10 (2-20) years. Histological analysis showed 6% N; 30% CCH; 53% MTC; and 11% MMTC. In the first decade MTC occurs as frequently as CCH (45 vs. 44%) and may already be found at the age of 2 years.
Conclusions: 1. MTC can occur already before 5 years of age. 2. Patient Screening should be performed in the first year of life to detect gene carriers. 3. Prophylactic total thyreoidectomy at 5 years of age prevents development of MMTC and leads to definite healing of C-cell-disease in MEN 2A patients.

STEM CELL TRANSPLANTATION (SCT) IN 4 CHILDREN WITH MUCOPOLYSACCHARIDOSIS (MPS) TYPE IH

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MPS type IH (Hurler disease) is a relatively frequent autosomal recessive lysosomal disease with absent α-iduronidase activity. The natural course comprises progressive intellectual impairment, visceromegaly, joint contactures, bone dysplasia, heart valve thickening, corneal clouding and hearing impairment. If performed within the first 2 years of life, SCT can arrest mental regression and reverse organ dysfunction (except for bone dysplasia). We report on 5 SCTs performed in 4 children with MPS type IH (age at SCT 9 to 30 months). HLA-identical family donors were used in 4, matched unrelated donors (MUD) in one patient. Four patients received myeloablative conditioning including Busulphane and i.v.Cyclophosphamide (in 3 patients i.v. Busulphane encapsulated in liposomes). In one child myelosuppressive conditioning with Melphalane and Fludarabine was administered. No significant transplantion-associated side effects were observed. The oldest patient had orthopaedic spinal surgery at age 7 yrs and is attending normal school with good results at age 8 yrs. The MUD-transplanted patient is alive and well with full donor chimerism 2 months after SCT. The other two children had autologous reconstitution despite donor lymphocyte infusions; one of them was successfully re-transplanted with a myelosuppressive conditioning using the same donor. **Conclusion:** SCT is a curative therapy option in patients with MPS type IH: early diagnosis is important.

Repeated hypoglycemias in an infant: metabolic investigations including acyl-carnitine

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Blaise has been admitted twice (at 18 and 27 months) in the hospital as emergency because of neurological symptoms (apathy, drowsiness, tonic-clonic seizures) after fasting triggered by an acute infection (rhinitis and pharyngitis). During the two admissions a hypoglycaemia has been found ; the neurological symptoms disappeared after glucose infusion.
The biochemical and molecular work-up led to the diagnosis of very long chain acyl-CoA deshydrogenase deficiency. Based on the case report we propose to give a priority to collecting blood initially on filter paper (Guthrie cards) in critically ill hypoglycemic patients before the biochemical work-up. Obtaining the samples early can spare the patient a subsequent fasting test. Caracteristic acyl-carnitine profiles are not necesseraliy obtained on blood obtained after recovery. The analysis of acyl carnitines on the intitial sample stored in the patients file can later be requested when the most of usual causes of hypoglycaemias have been excluded through targeted and standardised investigations (experimented metabolic center). This case report shows that assecuring material for a further testing for rare disorders during the phase of critical illness before doing a careful biochemical work-up can alleviate the load of investigations for a patient.

URETHRALITHIASIS – A RARE CAUSE FOR ACUTE URINARY RETENTION

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Motivation: During the year 2000 5 children 2 to 5 years old (4 ♀/1 ♂) consulted our emergency departement because of an acute urinary retention (AUR). Cystitis, vulvovaginitis (2x) and constipation were observed as reasons for AUR in girls. A very rare cause for AUR was found in the boy.
Case report: The 5-year old boy was referred because of a painful AUR with concomitant pain in the penis. Due to a resistance in the penile urethra it was not possible to introduce a transurethral catheter into the bladder. A suprapubic catheter (SC) was placed and 1000 ml of urine (with negative urine culture) were drained. The voiding cystourethrography (VCUG) performed through the SC showed a stop auf the constrast media in the fossa navicularis. A cystoscopy was done and a distal urethral stone could be identified and removed. The stone measured 6 mm in diameter and consisted of 50% Ca-oxalate and 50% NH_4-urate.
Conclusions: In Switzerland urethralithiasis is a rare manifestation of stone disease. Stones in the bladder and urethra are endemic in warm climate zones. Obstructive causes for AUR must be considered especially in boys. The diagnostic work up always has to include palpation of the penile urethra, a sonography of the abdomen , a VCUG and an urethrocystoscopy.

CEREBRAL VENOUS THROMBOSIS: COMPLICATION OF THE IDIOPATHIC NEPHROTIC SYNDROME

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Background: Children with idiopathic nephrotic syndrome (INS) are at high risk of thromboembolic complications. Contributing factors include a high red blood cell count, hypovolemia, drugs (diuretics, steroids) and hypercoagulability.
Case report: A boy with INS suffered the 12th relapse at the age of 13 years. After one week of daily prednisone (75 mg), he went into remission (albustix negative). Polyuria developed with loss of 7 kg of weight and he had severe headache. One day later he complained of double vision. Physical examination showed bilateral papilledema; blood pressure was normal (110/70 mm Hg). A cerebral CT scan demonstrated widespread thrombosis of the right sinus sagittalis superior, transversus and sigmoideus incl the jugular internal vein. Hematocrit (42%) and fibrinogen (3,3 g/l) were normal, fibrin D-dimers (1,3 mg/l) were elevated and anti-thrombin III (58%) was slightly diminished. On anticoagulation - heparin iv for 9 days, followed by daily dalteparine (2500 U sc) for 8 weeks - and prednisone, clinical, laboratory and CT-scan findings all normalised.
Conclusions: A cerebral venous thrombosis has to be considered in all children with INS and neurological symptoms. The risk of thrombosis is not only increased during proteinuria, but also at the beginning of remission. Anticoagulation is advocated in patients with normal blood pressure.

ARTHROSCOPIC HANDLING OF HIP JOINT INFECTION IN CHILDREN

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Introduction: Arthritis is not very common in children. Out of this uncommon diseases the bacterial coxitis is the most frequent one. This can also occur in babies. Material and method: Since 1996 we treated 9 children between 3 months and 14 years of age. All patients had a follow up until they were pain free. Results: All patients showed an infection of the hip joint clinically together with abnormal laboratory parameters. The children had 1 to 2 arthroscopies of the hip to clear out the joint and to get a bacteriological diagnosis. According to their antibiogram the children were treated with antibiotics. They were mobilised immediately after surgery. After an average of 3 - 4 weeks all patients were pain free and had normalised laboratory parameters. Conclusion: The arthroscopic flushing of the hip joint is clearly an improvement compared to the pure flushing of the hip joint with a needle and clearly an improvement compared to a open joint revision with a shorter

FIRST EXPERIENCES ON THE MINIMAL INVASIVE TECHNIQUE OF NUSS FOR THE CORRECTION OF PECTUS EXCAVATUM.

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Purpose: Outcome of the analysis of first cases of MIRPE.
Methods: Since February 2000, 7 Nuss-procedures have been performed. There were 6 boys and 1 girl of a median age of 16 years (range 13,8 - 17,8). Preoperatively, we observed a chest wall asymmetry in 6 cases (left 3, right 3). Under thoracoscopic view a retrosternal transversal bar was placed transthorathically from the right side in 6 cases and from the left side in 1 case. The average operation time was 138min., (range 110 - 180). There was no significant loss of blood. All patients received an epidural catheter (3 - 5 days) and per os analgetics for post operative pain control. Because of the epidural controlling, the patients remained at the intensiv care unit for 24 hours. **Results:** For all patients and surgeons the cosmetic results were very good. **Conclusion:** The treatment of pectus excavatam following the Nuss procedure is a simple, quick, cosmetically satisfaying and reasonable operation. From our point of wiew, the pain controll as well as the only secondary complete spontaneous correction of the remaining thorathic asymmetry should be improved.

ARTHROSCOPY AND HYDRAULIC MOBILISATION OF THE HIP JOINT WITH BUMP RESECTION IN CHILDREN

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Introduction: The movement restriction of the hip joint after aseptic and posttraumatic necrosis of the femoral head is not only a functional problem. It is also a problem for the remodelling of the hip due to the joint congruence. **Material and method:** Since 1997 children with movement restriction of the hip joint after aseptic and posttraumatic necrosis of the femoral head were treated arthroscopically and were sequentially documented with an at least 1 year follow up. We treated 15 patients, 6 girls and 9 boys between 10 and 17 years of age. 13 patients had a hydraulic distension followed by a gentle mobilisation of the hip joint. The arthroscopy was done without distraction. 2 patients had an arthroscopic resection of a lateral ossification of the femoral head (Bump Resection). **Results:** All patients had a clear improvement postoperativly. The average flexion - improved up to 110° with 40° - 0° - 40° adduction – abduction. **Conclusion:** The hydraulic mobilisation of the hip joint is a standardised method for the improvement of the joint function and thus a good possibility for the improvement of the joint congruence after femoral head necrosis and it can be done without distraction of the hip.

MANAGEMENT OF THE IMPALPABLE TESTIS : THE ROLE OF LAPAROSCOPY

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Aim: Nonpalpable testes represent 20 % of all cases of cryptorchidism (CR). The aim of this study is to present the results of 50 patients who underwent diagnostic laparoscopy (LA). The definitive surgical procedure was decided by the surgeon after the laparoscopic status.
Materials and methods: Between 1993 to 2000, 50 patients underwent laparoscopy for CR with nonpalpable testis (left: 32; right: 14; bilateral: 4). Mean age: 3,2 years old. Diagnostic LA was performed with or without other surgical procedure: LA n=8 ; LA and inguinal exploration n = 21 ; LA and orchidopexy n=13 ;
LA and 1-stage of Stephens-Fowlers laparoscopic procedure n=8.
Results: No operative complication was observed. 18 boys were outpatients. Median hospital stay: 24 hours (0 to 3 days). Definitive diagnostic was performed in all the cases: 21 high CR, 29 testicular atrophy. 8 post-operative complications: 3 cutaneous abscess, 1 subcutaneous haematoma, 1 testicular atrophy, 1 failure of orchidopexy, 1 stitch granuloma and 1 umbilical hernia. Mean follow-up time: 3.6 years (2 months to 8 years).
Discussion : Diagnostic LA is a safe and simple procedure. A diagnostic was performed in all the cases during laparoscopy (specificity 100 %). In case of intraabdominal testicular atrophy, an inguinal surgical exploration can be avoided.

THE SIGNIFICANCE OF BOTH AN EARLY ORCHIDOPEXY AND GERM CELL MATURATION FOR FERTILITY IN CRYPTORCHIDISM

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Twenty-seven adults who underwent an orchidopexy before two years of age have recently had their spermiograms analyzed. Sixty-three percent of them had a normal sperm count; their germ cells had undergone the second stage of maturation and Ad spermatogonia were present at the time of surgery 20 - 25 years earlier. In contrast, 37% of them were infertile despite a seemingly successful orchidopexy; they had impaired second-step germ cell maturation and Ad spermatogonia were absent at orchidopexy. Thus, the transformation of gonocytes into Ad spermatogonia is crucial for fertility. This underscores the importance of a testicular biopsy at orchidopexy to identify those who could benefit from early LH-RH treatment after orchidopexy.

CONGENTIAL DIAPHRAGMATIC HERNIA (CDH) - HAS ANYTHING CHANGED IN THE LAST 12 YEARS?

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CDH remains a challenge for pediatric intensive care and surgery. The aim of the present study was to analyse our results, and, possibly, draw conclusions. Patients: 1988-99 48 newborns with high-risk CDH (clinical manifestation within 6 h after birth) were treated, 18 from 1988-91 (conventional and high-frequency jet ventilation), 17 from 1992-95 (high-frequency oscillation), and 13 from 1996-99 (NO). Results: In 44 instances CDH was left-sided, in 4 right-sided. 23 patiens had associated malformations including 4 with chromosomal abnormalities. Survivors (n = 25) had significantly better 5 min. Apgar Scores, less cardiovascular dysfunction and coagulation abnormalities, and less often pneumothorax. First arterial pH, P_aCo_2, oxygenation index (OI) and alveolo-arterial oxygen gradient (P_{A}-$_aO_2$) were significantly better in survivors. 31 patients were operated with 25 survivors, 17 neonates never reached a stage of operability. There was no difference in outcome during the 3 abovementioned periods. P_{A}-$_aO_2$ < 200 - 250 mmHg and OI < 5 - 10 were robust criteria for successful surgery. Conclusions: Despite refinements of support, at least statistically, the results have not improved over the years. However, the concept of gentle ventilation, waiting for good gas exchange and delayed repair seems to be correct.

A NIGHTMARE AFTER A CIRCUMCISION

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Aim : To present a boy who suffered from severe ischemic glans after circumcision and dorsal penis nerve block.
Case Reports: A 3 year old boy with phimosis had a CC in day care surgery with standard anesthesia with Sevorane. A dorsal penis nerve block was performed with Bupivacain 0.5% before the procedure, using a 25G caudal needle. The intervention and first postoperative period were uneventful. 6h later, before discharge, the boy complained about severe pain (pain scale 8 of 10) and a dark blue glans with petechiae was observed. A vascular problem was suspected and the decision taken to perform a caudal block for sympaticolysis. 20 min later the situation improved clearly and the caudal block was continued for anothers 48h. No other therapy was necessary and the nightmare was gone without lasting consequences.
Discussion: The dorsal penis nerve block is a safe and common procedure, that provides an effective analgesia for surgery of the penis. Complications are usually absent or only minor. However single cases with severe complication have been described, but usually due to trauma or accidental lokal administration of vasoconstrictive agents. Caudal-epidural sympaticolysis is an effective measure in this situation and can prevent nightmares.

SURVEY OF THE MANAGEMENT OF UNILATERAL INGUINAL HERNIA BY PAEDIATRIC SURGEONS, MEMBERS OF THE SSCI

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Aim: Infant and child presenting with unilateral hernia continue to provoke controversy among paediatric surgeons about the management of the "other side". The aim of this study was to obtain a survey of the management of the unilateral inguinal hernia by the paediatric surgeons, members of the SSCI (Swiss Society of Paediatric Surgery). **Materials and methods:** A case-scenario questionnaire was sent to all ordinary members of the SSCI. The questionnaire was elaborated to determine the management of the unilateral inguinal hernia (premature, full-term infants, ovary hernia, pre- and per-operative investigation).
Results: Responses were received from 39 of 61 members (64%). All responder surgeons based the operative indication on the clinical status. No paraclinic investigation was recommended. 31% of surgeons treated the symptomatic side only. 69% performed controlateral exploration decided by : - age, - sex, - prematurity,
- side of hernia. Laparoscopic evaluation through ipsilateral sac was performed by 15 % of responders. If the ovary was nonreducible all surgeons performed surgery urgently in the next 3 days.
Discussion : All the surgeons consider that the inguinal hernia treatment is surgical and no paraclinic investigation is necessary. The operation is urgent if the ovary is nonreducible. One third of surgeons treats the symptomatic side only.

BEHAVIOR AND PSYCHOLOGICAL ADJUST-MENT IN CHILDREN AFTER ACCIDENTS

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Background: This study investigated psychological adjustment and prevalence of behavioral maladjustment in a group of children after accidents without severe head injury.
Method: 95 children (mean age=9.8 years), their mothers, and their teachers were investigated with standardized assessment instruments one month after the occurence of an accident which led to hospitalization of the child.
Results: 15% of the children showed posttraumatic stress reactions in a clinical range. Such reactions were most frequent in victims of road traffic accidents. Compared to a healthy reference group, mothers (28%) and teachers (24%) both reported elevated rates of behavioral maladjustment.
Conclusions: Psychological and behavioral sequelae after accidents are quite frequent in children. To clarify duration of these reactions longitudinal studies have to be conducted.

CONSERVATIVE HANDLING OF KIENBOECK DISEASE OF A 12 YEARS OLD GIRL

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Introduction: The aseptic necrosis of the os lunatum is a disease particularly of young adults, usually causing a disturbance of the blood circulation and a necrosis of the bone. **Material and method:** We treated a 12 years old girl suddenly suffering from pain at the adominant wrist without trauma. The x-rays and the MRI showed a necrosis of the os lunatum. We treated the girl with immobilisation of the wrist and we supported the revascularisation medicamentously. **Results:** Under this therapy the girl was pain free within 3 month. During the treatment the patient went to school without any problem. At the final check up the revitalisation of the os lunatum was proved. **Conclusion:** There is no doubt that a necrosis of the os lunatum can occur without trauma, and there is no doubt that the treatment must be compatible with the needs at school over several months. Because of this, the immobilisation has to be as minimal as possible but still have the possibility of a collapse of the carpal tunnel in mind.

DIAGNOSTIC AND THERAPEUTIC PROBLEMS OF SPONDYLODISCITIS WITH ABSCESS FORMATION IN CHILDREN

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Since the clinical symptoms of a spondylodiscitis are rather uncharacteristic, the time of the diagnosis is often delayed.
A 2 10/12y old girl fell downstairs unobserved in June 1999 and complained about back pains.
The investigations gave no signs of a fracture or other lesions.
Only one month later we saw the girl again.
The X'rays showed the first signs of a spondylodiscitis. The ESR was only minimally elevated. MRI indicated a spondylodiscitis L3/L4 with a small abscess formation.
After investigating the literature, we chose a conservative therapy with antibiotics, due to the slightness of the abscess formation and insignificant neurological deficits.
Spondylodiscitis should be a differential diagnosis in a child with persistent back pain following trauma.

EXTENSIVE IDIOPATHIC SEGMENTAL NECROSIS OF THE FEMORAL HEAD IN A 15 YEARS OLD BOY WITH OPEN GROWTH ZONE

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Introduction: The occurrence of an idiopathic femoral head necrosis in adolescents is very rare.
Materials and Methods: A 15 years old boy is presented with a-traumatic acute pain in the region of the right hip for a period of 3 weeks prior to the emergency consultation. The x-rays shows the picture of extensive segmental necrosis of the femoral head. In view of the extent of the condition as well as the still active metaphyseal involvement we performed an intravenous therapy with Iloprost accompanied by complete bed rest for 2 weeks. **Results:** In the additional controls a bone metaplasia was apparent in the conventional radiographs without accompanieing by an increasing loss of the round shape of the head. **Conclusion:** The problem how to treat idiopathic segmental necrosis of the femoral head has so far not been solved. In adults displacement osteotomies, drillings or prosthetic repair are undertaken. However such operations can not be performed on adolescents with an open growth zone particularly if an acute metaphyseal involvement has to be reckoned with. A conservative therapy with medicamentous support seems to be indicated.

«MERMAID SYNDROME»: REPORT OF A CASE OF SIRENOMELIA

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Symelia results from skin union of two inferior limbs, which appear otherwise complete. The birth of a living « mermaid » is very rare, its frequency being 1:60 000. Most of them are stillborn or die shortly after birth.

We report the case of a female neonate born to a III G III P mother, with an apgar score of 9-9-9 and still alive 2 months later. The baby had dipodic simelia (cf. Rx) with severe bilateral renal hypoplasia, absence of bladder and complex urogenital sinus. The complexity and severity of the malformations were of very poor prognosis and no further surgical treatment was applied.

The different types of sirenomelia are described, and the possible ethiologies reminded. The constantly associated orthopaedic, ano-rectal and urogenital malformations are described.

ONLY ABDOMINAL PAIN ?

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Aim : To demonstrate two teenagers with abdominal pain due to intussusception (IS) caused by a Meckel's diverticulum (MD).
Case Reports: The first boy, 11 years old, presented with a history of diffuse abdominal pain for the past 40h and one episode of non-bloody diarrhea. On examination no fever, a soft, but slightly tender abdomen without a palpable mass or peritonism. 6h later his abdominal pain and tenderness on examination were increasing, so he was taken to theatre for laparotomy. At the beginning of anesthesia he had a first episode of hematochezia. The second boy, 14 years old had diffuse abdominal pain for 8h, again slight tenderness of the abdomen but no peritonism. Because of suspicion of constipation on rectal examination he received an enema, and his abdominal pain improved after defecation. 12h later he presented again with increasing abdominal pain, but this time obstructed bowel sounds and blood stools on rectal examination. Both boys had a plain abdominal xray and ultrasound before laparotomy without specific pathologic findings. On laparotomy both boys showed ileo-colic IS with a long MD as the leading point. The first patient needed resection of 20cm of his ileum, the second patients ileum could be reduced without resection, only the MD was removed. Both boys had an uneventful postoperative course. **Discussion :** IS is a rare pathology in this age group and can be easily missed if not thought of.This two patients didn't show the classical signs of IS like intermittent intense abdominal pain, vomiting and abdominal mass. Bloody stools were found only late in the course. Xray and ultrasound were not helpful for diagnosis.

LAPAROSCOPIC TREATMENT FOR VARICOCELE

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Aim: The aim of this study is to present the results of laparoscopic treatment for varicocele in teenagers. The treatment consists in spermatic veins ligation, with or without spermatic artery ligation.
Materials and methods: Between 1992 to 2000, 30 patients (mean age 13,6 years old) underwent surgery for varicocele (29 left, 1 bilateral). The indications for surgery were symptomatic grade II or grade III varicoceles. Pneumoperitoneum was achieved by CO_2 insufflation through a Veress needle in 3 cases and with open laparoscopy technique in 27 cases. Spermatic vessels were ligated and sectionned by laparoscopy: **Gr 1.** Artery and veins (n=14); **Gr 2**. Veins (n=17). **Results:** No operative complication was observed. Mean operative time: 68'. Median hospital stay: 24 hours (0 to 2 days). 4 post-operative complications were observed: 2 hydroceles (Gr1), 1 haemorrhage and 1 incision abscess. Mean follow-up time: 44 months (1month to 8,3 years). No secondary testicular atrophy. 9 recurrences: **GR 1:** 2/14 (14%), **GR2:** 7/17 (41%). 3 patients of Gr2 underwent further surgery. **Discussion:** Laparoscopic treatment for varicocele is a safe and simple surgical procedure. In our experience spermatic artery ligation decreases recurrence rate without increasing complication rate.

INTUSSUSCEPTION AS A CAUSE OF ACUTE ABDOMINAL PAIN IN ADOLESCENTS

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Motivation: Intussusception is mostly an acute disorder of infants and toddlers.
In adolescents intussusception occurs rarely and is often associated with an anatomically identifiable lead point.
Case reports: Pat. 1 15-year old boy with colicky abdominal pain without vomiting or diarrhoea and tenderness on pressure in the left middle abdomen. Ultrasound (US) demonstrated a small-bowel intussusception. Laparatomy was performed with resection of an ileo-ileal intussusception with a length of 15 cm, induced by a Meckel diverticulum.The diverticulum showed invagination in itself. Pat. 2 2 days after the onset of a bloody diarrhoea admission of a 13-year old boy with colic in the para-umbilical region. A tumor was palpable in the right upper quadrant. US demonstrated an ileo-colic intussusception. Laparatomy was performed with reposition of an ileo-colic intussusception induced by lymphoid hyperplasia. Stool specimen was positive for verotoxin-producing E. coli (VTEC).
Conclusions: In 1.5-12% of children with an intussusception a re-lated lead point is detectable.
In case of unexplained abominal pain there is high grade of suspicion for intussusception. US is the first imaging modality. Surgery for exploration and revision is often necessary.

Congenital Diaphragmatic Hernia (CDH): Prognostic Value of Prenatal Diagnosis

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Introduction: According to the recent development of fetal surgery for congenital diaphragmatic hernia (CDH), particular interest was focused on prenatal ultrasound. Time of diagnosis, side of the hernia, associated malformations and the results of karyotyping represent factors which are essential for therapy and prognosis of these patients.

Methods and Results: We performed a retrospective analysis of 16 children resp. their mothers who were investigated resp. treated for CDH during the last 15 months at our institutions. Out of 11 intrauterine diagnosed patients, 7 died during the postnatal period (4x chromosomal anomaly with associated malformations, 3x isolated CDH on the right side). In 2 fetuses fetal lung volume measurement by 3D-ultrasound was performed which showed lung volumes reduced more than 50% to normal. Because of multiple major malformations, 2 pregnancies were terminated intrauterine (18./23. gestation week). 2 children were operated successfully on the first day of life. 5 children were referred from extern and presented CDH as incidental finding. All 5 children underwent successful operation.

Conclusion: Prenatal diagnosis, chromosomal anomalies and associated major malformations increase the mortality rate of patients with CDH significantly. The prognosis of postnatally detected CDH, esp. as incidental finding is good. Intrauterine diagnosed CDH requires careful prenatal sonographic examinations of all organ systems and karyotyping. Interdisciplinary assessment of prognosis and counselling of the parents is essential. An ongoing prospective study evaluates possible correlations of 3D-measured fetal lung volume and pulmonar vascular restistance with postnatal clinical course.

BILATERAL ADRENAL LYMPHOMA IN A CHILD

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We report a case of bilateral adrenal metastases of a Burkitt's lymphoma. A 15 year-old child, from Angola, without significant past-medical history presented with an acute abdomen due to a peritonitis. Emergency laparo-tomy revealed small bowel necrosis secondary to an ileo-ileal intussusception. Histologic examination of the resected gut (80 cm of small bowel) showed only necrotic tissues. Bilateral adrenal masses were seen on a post-op ultrasound (US). Endocrine and radiology work-up favored the diagnosis of non-secreting pheochromocytoma, although bilateral adrenal hemorrhages were not excluded. Surgery of the adrenal masses was postponed; they were monitored with periodic US. Two months later, a second intussusception required a new laparotomy. At that time, peritoneal and small bowel masses were biopsied and the left adrenal mass was excised. Histology showed Burkitt's lymphoma in all specimens analyzed. Chemo-therapy for a stage III Burkitt's lymphoma was admi-nistered according to the POG 9317 protocol. The child is in complete remission 6 months after the end of treatment. To our knowledge, the bilateral adrenal localization of a lymphoma has not been previously reported.

A COMBINED PEDIATRIC SURGICAL AND PEDIATRIC DAY-CARE CLINIC: EXPERIENCES WITH A NEW CONCEPT IN THE NORTHWEST REGION OF SWITZERLAND

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The first pediatric day-care clinic was established in Liestal (BL) in 1995. It combines surgical and medical services which filled a neglected gap between pediatricians, general practi-
tioners and the Children's Hospital. The results have been positive in many respects, especially in terms of the quality of life for the patients, the acceptance and contentment of the parents. More than 60% of today's surgical procedures requiring anesthesia are performed in day-care facilities. These include severe cases involving primary and secondary reconstructive procedures on male genitalia both of which are performed routinely in our Clinic. KTK offers two distinctive medical services: (1) extensive diagnostic examinations collaborating with pediatric surgeons and radiologists; (2) emergency medicine (i.e., infants who require i.v. medications). The average day-care hours correspond to 1.9 days compared to 5.5 days in pediatric hospitals for the *same* group of patients. This type of day-care center would be able to optimize the expenses for medical care provided that it is an integral part of a regional governmental health care plan. KTK is a novel service for children who would otherwise be hospitalized in a regional Medical facility. Pediatric health care of the future has to be constructed on three pillars: (1) pediatricians and practitioners (2) children's day-care clinics, and (3) university and regional hospitals. A sucessful integration will provide an optimal model for continuous medical education and clinical research.

Inflammatory response of neutrophil granulo-cytes and monocytes to cardiopulmonary bypass in pediatric cardiac surgery

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Background: Cardiopulmonary bypass (CPB) induces a systemic inflammatory response that may cause substantial clinical morbidity. Activation of inflammatory cells during CPB may contribute to this process. This study sought to determine whether the activation state of polymorphonuclear neutrophils (PMN) and monocytes correlates with myocardial damage after CPB.

Method: Phenotypic and functional analysis of circulating PMN and monocytes were done in 20 pediatric patients before and after open heart surgery with CPB.

Results: After CPB, the level of cardiac troponin-I (cTn-I) correlated with bypass time and duration of postoperative inotropic support. Plasma levels of myeloperoxidase (a marker of PMN degranulation) were significantly elevated after CPB. However, plasma concentrations of adhesion molecules sL-Selectin and sICAM-1 were lower after CPB. PMN function, as measured by surface expression of CD11b/CD18 and respiratory burst was reduced, too. In contrast, respiratory burst of circulating monocytes was increased in some patients and there was a correlation with the increase of the level of cTn-I after CPB.

Conclusion: After CPB, an inflammatory process including phenotypic and functional changes of PMN and monocytes was detectable and correlated with severity of clinical course. In particular, activation of monocytes might contribute to myocardial damage.