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Date: 19/06/2008
Time: 16:15–17:30

PED 1-1

Physical activity patterns in children with cerebral palsy measured by accelerometers – a pilot study

K. Sonderegger[1], F. Eugster-Büsch[1,3], E.D. De Bruin[1], R. Müller[1], N. Ruch[2], A. Meyer-Heim[3].

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Aims: Health problems related to inactivity in children from industrial countries are being increasingly investigated. Earlier research concerning children with cerebral palsy (CP) has focused on the quality, rather than the quantity of movements. The goal of this study was (1) to quantify the level and patterns of physical activity in everyday life for children with mild CP and to compare these results with both (2) the activity of healthy children and (3) the recommendations for daily activity of the Swiss Federal Office of Sport (BASPO).
Methods: The activities of six 10 to 12 years old children with mild hemiparesis (4 boys / 2 girls) due to CP (GMFCS Level I) were logged, wearing a uniaxial accelerometer on the hip of the non-affected side for one week during the day. Healthy children (n = 37, 10.92 ± 0.86 years) served as the age matched control group. By means of the acceleration data, the total physical activity, the total duration of different intensity levels and the number of continuous sequences in at least moderate intensity were determined.
Results: The average number of active minutes in the children of the study group was 70.6 min/day. There was a considerable difference between weekdays and weekends: 76.2 min/weekday and 56.6 min/weekend day. On a whole both groups spent the same amount of time in moderate (p = 0.53, 35.9 min in healthy children and 32.6 min in children with CP) and vigorous (p = 0.99, 38.0 min in both groups) activities. The analysis of the raw data over one week showed no difference between the groups (p = 0.92). A within-group analysis comparing the weekend with the weekdays showed no difference in children with CP (p = 0.79), whereas healthy children were significantly less active on the weekends (p = 0.08). There were no significant differences between the two groups in respect to the duration of the physical activity throughout the whole week (p = 0.78), nor on weekdays (p = 0.75) or on weekends (p = 0.98). With respect to the BASPO recommendations both groups move sufficiently.
Conclusion: Children with mild CP show similar activity levels as a healthy control group and, thus, may move sufficiently to fulfill the BASPO recommendations. This pilot study justifies more research with a greater number of participants to investigate the activity level of children with different diagnoses and degrees of disabilities to substantiate our findings and to, in the long run, develop recommendations adapted for children with motor disabilities.

Date: 19/06/2008
Time: 16:15–17:30

PED 1-2

Constrained-induced movement therapy of hemiplegia: our first experiences

P. Weber, S. Stock. University Children's Hospital, Basel

Aim: Pediatric hemiplegic cerebral palsy (CP) is one of the most frequently movement disorders of childhood. In contrast to symptoms of CP of the lower limbs therapeutic concepts for the treatment of the upper limbs are sparse. Recently, constraint-induced movement therapy (CIMT) has been shown promise for improving upper-limb function in children with cerebral palsy.
Methods: Between October 2006 and October 2007 4 children (mean age 7.1 yrs, range: 4.11–8.8 yrs) with hemiplegia were treated by CIMT. Three boys suffered from neonatal stroke, the girl from a stroke that had occurred when she was 2.8 yrs of age. One boy was treated twelve months later for a second time. In our approach the children were treated six hours a day for two consecutive weeks. During this time, the healthy hand was taped. Two hours a day, five days a week, the children were treated by an occupational therapist, and four additional hours a day, they were motivated to deal with a hand motor task at home. We controlled the effects of this treatment in two children by the standardized Assisting Hand Assessment, in two children by several tasks measuring speed, force, and accuracy of the hand movements by a modified Schilling test, a pegboard task, and a shape task with cubes. In addition, the parents gave their impression about the spontaneous activity of the hand before and after the treatment.
Results: The time and function measuring showed a small effect in some of the measured parameters. The two patients tested by the non-standardized test battery showed an improvement of force (one of 100%, one of 5%), the two children tested by AHA showed an increase of the global hand function value from 56% to 61%, respectively from 80% to 89%. The parents of all children reported an increased spontaneous use of the affected hand during various daily activities. Effects will be demonstrated by video sequences.
Conclusion: CIMT is a new therapeutic approach to treat children with a hemiplegic hand. Although our first experiences demonstrate only small effects, the parents report about a more intensive use of the hand after the training. An evaluation of the procedure with regard to the treatment duration per day and the number of therapeutic cycles has to be discussed.

Date: 19/06/2008
Time: 16:15–17:30

PED 1-3

Shaken baby syndrome (sbs) in Switzerland – results of the retrospective study 2002–2007

M. Fanconi, U. Lips. Child Protection Group, University Children's Hospital Zurich

Objective: Since the incidence of SBS in Switzerland is not known, we conducted a nationwide retrospective study covering the last 5 years.
Method: Data collection through SPSU system. Inclusion criteria: Presence of 1) ≥2 clinical symptoms (altered consciousness, convulsions, respiratory irregularities, bulging fontanel) or 2) 1 eye findig (retinal haemorrhages, vitreous haemorrhages) or 3) 1 MRI/CT finding (subdural haematoma, subarachnoid haematoma, parenchymatous lesions) or 4) history of shaking; all these findings in a child ≤6 years. Exclusion criteria: age >6 years or documented accident/disease explaining symptoms/findings. Outcome measurement following King's Outcome Scale for Childhood Head Injury (KOSCHI); (Crouchman M et al. Arch Dis Child 2001;84:120–4).
Results: Of 53 reported cases 50 met inclusion criteria. Cases reported from only 13 cantons; unexplained high number of reported cases in 2005. High male preponderance (31 m, 19 f), mean age 6.7 months (1–58). Clinical symptoms present in 42 patients, retinal/vitreous haemorrhages documented in 39, brain imaging pathologic in 46 patients. Shaking confessed in 14 cases. Outcome (n = 45 patients, outcome in 5 patients not yet known): Death (KOSCHI 1) 8 (17.7%), vegetative state (KOSCHI 2) 0, severe disability (KOSCHI 3) 10 (22.2%), moderate disability (KOSCHI 4) 14 (31.1%), good recovery (KOSCHI 5) 13 (28.8%).
Conclusions: According to these data incidence of SBS in Switzerland is 14 in 100'000 live births, which corresponds to incidences in other western countries.
1. Demographic characteristics and outcome of our patients are comparable to published studies.
2. Unexplained remains the lack of adequate reporting in half of Swiss cantons.

Date: 19/06/2008
Time: 16:15–17:30

PED 1-4

Partial splenectomy in pediatric hematology/oncology: a single-institution experience

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Splenectomy can improve chronic hemolytic anemias, such as pyruvate kinase deficiency (PK) and hereditary spherocytosis (HS). However, total splenectomy (T.S) results in partial immune defect and increases risk of overwhelming post-splenectomy infection (OPSI) especially in young children. Partial splenectomy (P.S) has emerged

as an alternative to T.S and successfully used in HS. It can be done at younger age, and is not associated with OPSI. Reports on P.S in pediatric population are still sparse, and possible benefit in other types of hemolytic anemia unclear. Aim. Evaluate our local experience with P.S.
Method: Retrospective chart review between 1995–2005. Need to perform 2ary T.S used as an outcome measure. Patients requiring T.S within 1 yr of P.S (early T.S group) were compared with those having a T.S performed at a later time or not (late or no T.S group). T-test used for statistical analysis.
Results: 7 patients (2 PK and 5 HS) had a P.S done at a mean age of 4.4 yrs (27m.-9 yrs). 2/5 HS patients also had congenital heart disease (CHD). Surgical procedure was laparoscopy in all but one. All patients were followed by ultrasound 2x year. There were no surgical or infectious complications. In 1/2 PK patients, no decrease of transfusion requirements was observed and T.S performed within 12 months; the 2nd patient showed transient improvement but eventually had T.S 2½ yrs later. In 3/5 HS patients, T.S was performed within 12 months because of persistent hemolytic anemia. 2 HS patients had clear improvement: one with complex CHD needed no transfusions for 4 yrs, but hemolytic anemia worsened during the 5th year, and eventually had T.S. The 5th HS patient is still doing well almost 5yrs post P.S. Overall, 6/7 patients required T.S of which 4 were done <1 year after P.S. All 3 patients <3 yrs belonged to the "early T.S" group. While all patients had spleen reduction by 50–70% by surgery, spleen size in the "early T.S" group returned more rapidly to pre-surgery size than those of "late or no T.S" group (spleen size within 1st year of P.S: 92% compared to 54% respectively; P = 0.001).
Conclusions: P.S is a safe procedure. It can permit a gain of several months with reduced PRBC requirement, but rapid spleen regrowth seems to predict the need for T.S. While P.S benefited the most in H.S patients, its role in PK and in patients operated at a very young age remains questionable. Indication and benefit of P.S. need to be determined in a larger prospective study.

Date: 19/06/2008
Time: 16:15–17:30

PED 1-5

Follow-up care in long-term childhood cancer survivors in Switzerland: who is missed out?

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Purpose: As the numbers of survivors of childhood cancer is increasing the question how to structure medical follow-up to detect, mitigate or prevent late effects of the tumour or its therapy might become gradually more important. Traditionally, in Switzerland this follow-up has been provided by paediatric oncologists. With growing numbers of survivors this becomes impossible. We aimed to study the current status of follow-up care for adult childhood cancer survivors in Switzerland.
Methods: Eligible for the study were all former childhood cancer patients registered in the Swiss childhood cancer registry (SCCR), who had survived at least five years, were aged ≥20 years at the time of the survey and had a valid address (N = 885). We sent a detailed questionnaire on somatic and psychological health, which included a number of questions on health care utilisation, particularly follow-up care. Prospectively collected clinical information on the tumour and its therapy was available from the SCCR.
Results: By Jan 03 we received 551/885 questionnaires (62%), of which 484 have been entered and analysed. The study is ongoing. Only 127 survivors (26%) had received a discharge summary or an individualised checklist that include recommendations for future check-up visits. At the time of survey, 116 (24%) survivors went to regular follow-up visits, 166 (34%) went irregularly and 202 (42%) never. In a multivariable logistic regression, female sex, shorter time since diagnosis, radiation therapy and perceived late effects, but not educational level were associated with having follow-up visits.
Conclusions: Contrary to recommendations, only a minority of adult survivors of childhood cancer in Switzerland go to regular follow-up visits, and very few possess a discharge summary or checklist. It needs to be determined if this is due to a lack of suitable health service facilities, or lack of awareness from the part of the survivors.

Date: 19/06/2008
Time: 16:15–17:30

PED 1-6

Growth arrest, senescence and apoptosis induction in brain tumour cells by a novel telomestatin derivative, a potent c-myc g-quadruplex interactive agent

T. Shalaby[1], A. Von Büren[1], M.L. Hürlimann[1], G. Shinya[2], I. Jelezarov[3], M. Grotzer[1].

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A significant proportion of children with medulloblastoma (MB), or atypical teratoid/rhabdoid (AT/RT) brain tumours continue to fail therapy hence identification of novel therapeutic strategies remains a major goal. Small molecules that stabilize the G-quadruplex structure (G4) in c-myc promoter sequences have shown potential as a new opportunity for cancer therapy. Exploiting this innovative line of attack to cancer cells, we investigated the effects of S2T1-60TD (TD), a novel telomestatin derivative, on a representative panel of MB and atypical AT/RT children's brain cancer cell lines. In this study the CD spectroscopy experiments demonstrated a high-affinity physical interaction between TD and the guanine rich promoter region sequence of c-myc. In the presence of TD the c-myc G4 structure significantly stabilized against thermal melting. These results were further supported by the PCR-stop assay. In contrast to control cells, treatment (72 h) with TD exhibited a clear anti-proliferative effect – as measured by MTS assay – in all MB and AT/RTcell lines tested, in a dose and time dependent manner and with IC50 at submicromolar levels (0.25 μM–0.39 μM). Importantly, TD demonstrated significant less cytotoxic effects on normal (not cancer) fibroblast MCR-5 cells (IC50 for MCR-5 was 5 folds more than the IC50 of MB and AT/RT cells). Through its potential to recognize, c-myc promoter sequence, 72 h treatment with TD reduced the mRNA and protein expressions of c-myc and hTERT – that is transcriptionally regulated by c-myc – and decreased both genes activities. Under conditions where inhibition of both proliferation and c-myc activity were observed, TD treatment decreased the protein expression of the cell cycle activator CDK2, and resulted in cell cycle arrest. Long-term treatment (5weeks) with non-toxic concentrations of TD resulted in time dependent telomere shortening which was accompanied by cell growth arrest (starting in day 28) and was followed by cell senescence and induction of apoptosis at day 35 in all of the five cell lines investigated. We demonstrated here the specific recognition of the G rich sequence in c-myc promoter by a novel small molecule. Through its potential to stabilize a G4 structure TD was able to inhibit c-myc, disrupt telomere maintenance, force cells to senescence and induce growth arrest and apoptosis in MB and AT/RT cell lines.

Date: 19/06/2008
Time: 16:15–17:30

PED 2-1

Enteral tube-weaning in infants in Switzerland – a successful interdisciplinary approach of the children's hospital lucerne

R. Hotz, R. Straub, P. Imahorn, M. Maurer, S. Schupp, J. Spalinger. Department of Pediatrics, Children's Hospital Lucerne

Aims: Long-term naso-gastric or gastrostomy-tube feeding is common in infants and children with feeding disability, chronic disease or impaired nutritional status. Exclusive tube feeding in contrast to normal eating behaviour is associated with complications and psychosocial impairment. Children fed by long-term enteral tube loose their ability to drink and eat. Programs of tube weaning are not well established in Switzerland. As tube weaning includes behavioural treatment and nutritional support, a specialized and competent multidisciplinary team is needed. This is a retrospective analysis of 5 children being weaned at the children's hospital of Lucerne, Switzerland.
Methods: To wean the children an adapted Graz model was applied. This consists in an interdisciplinary approach by a team of paediatricians, psychologists, nurses, physiotherapists, ergo-therapists, and a dietician. The weaning procedure starts with a few days of adaptation, followed by a period of intensive observation of the child's behaviour. During the third phase, nutritional supply through the tube is progressively reduced and completely stopped, in order to stimulate the "self regulation" of thirst and hunger of the child. When the child starts to eat and drink, parents are trained in behavioural techniques to support their child during the meals. Before tube weaning procedure is started, criteria to discontinue the tube weaning are defined, such as severe dehydration, metabolic disorders, renal failure or severe parental destabilisation.
Results: Five children (3 boys and 2 girls) were admitted to follow the weaning program. They were between 12 and 66 months old. The duration of the hospitalisation was between 6 and 10 weeks. The weight loss was between 9 and 17% of body weight. There was no severe dehydration or other complication. At discharge all children were weaned completely from gastrostomy feeding. During follow-up (6 months to 5 years) there were no relapses, all children are thriving.
Conclusions: Long term gastrostomy or enteral tube feeding is common in the paediatric age group. An intensive multidisciplinary behavioural treatment is successful in improving oral intake and weaning from tube feeding in children. To our knowledge, there is no established weaning program in Switzerland. Lucerne's children's hospital has successfully weaned 5 children from the gastrostomy tube without any complications using an adapted program of the well-known Graz tube weaning model.

Date: 19/06/2008
Time: 16:15–17:30

PED 2-2

Malposition of feeding tubes in neonates

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2 Abteilung für Bilddiagnostik, Kinderspital Zürich

Objectives: Accurate tube placement of orogastric and nasogastric feeding tubes in neonates is important to ensure safe and effective enteral feeding. Errors in placement and position of feeding tubes are described in literature, but there is little evidence of the exact prevalence of improperly placed tubes.
Aims: The aim of the study was to evaluate the prevalence of improperly placed feeding tubes in standard radiographs of the neonate.
Methods: A retrospective review of 381 x-rays of 172 neonates (mean gestational age 30 3/7 weeks, 25–42 weeks) was performed and exact positions of the feeding tubes on x-rays were defined. Before placement the insertion length and depth of the feeding tubes was predicted by measuring the distance from the nose to the earlobe and then to the termination of the xiphoid process. To describe tube position on the radiographs, we used a scoring system according to the description of T. Weibley et al. (1987). Anatomical structures like the diaphragm or the stomach were used as orientation to define tube position. On this basis 5 different positions were defined. Tube placement error was defined as tubes whose tip or orifices were not in the body of the stomach.
Results: On 21% (78 / 381) of the radiographs exact tube position could not be defined. The visualisation of the stomach was due to small amount of air in the stomach as part of the dynamic digestive tract in the a.p. radiographs not possible. These 78 radiographs were excluded from further analysis. Investigation of the 303 remaining radiographs showed that only in 41% of the placements was the optimal position of the feeding tubes achieved. We regard this percentage low, considering that feeding tube placement is a very common procedure in neonatal intensive care units and several complications caused by placement errors are described in the literature (Metheny et al. 2007). Overall 59% (179 / 303) of the feeding tubes had been placed incorrectly. Out of these incorrect placements 61% were located to deep in the stomach, bending along, touching lightly or grazing the greater curvature of the stomach. Deep placement is therefore the most frequent placement error in neonates. We assume that this is important especially in very low birth weight infants, because tube tips might cause bleeding or perforation of the stomach. No significance was found for the possible interacting factors: gestational age, birth weight and mechanical ventilation.
Conclusion: Our study showed a high rate of wrongly placed feeding tubes in neonates. There is a need for both better rules to estimate the distance between nose (lips) and the body of the stomach and improved methods to confirm correct tube position.

Date: 19/06/2008
Time: 16:15–17:30

PED 2-3

Alcohol intoxication among adolescents: Evaluation of a new Ambulatory consultation

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2 Service d'abus de substances, Département de psychiatrie, Hôpitaux Universitaires de Genève

Aims: Evaluation of the impact of a multidisciplinary ambulatory consultation (specialists in substance abuse and paediatricians of adolescent medicine) for the adolescents having consulted for an acute alcohol intoxication at the paediatric emergency unit (max. age: 16 years).
Methods: Collected data stems from anonymous questionnaires sent to patients seen at our consultation from June 2006 to December 2007. The monthly delay between consultation and survey ranges from 1 to 20 with an average of 10.
RESULTS: Out of the 87 patients seen at the Emergency unit for an acute alcohol intoxication and convoked, we evaluated 82 (94%) at our multidisciplinary consultation within 10 days following their intoxication. More than half of them returned the questionnaire. The items concerning the consultation itself are summarized in table 1 and those concerning the alcohol consumption since the consultation in table 2.

Table 1: summary of items about the consultation

Learnt something during the consultation (alcohol effects, risks, control of consumption …)	72%
The consultation was useful	60%
The consultation improved the communication with their parents	32%

Table 2: summary of items about alcohol consumption

No new alcohol intoxication after the consultation	68%
• once or twice	16%
• more than twice	16%

When they answered the questionnaire, none of them think to have an alcohol problem. 40% were affected by social difficulties (problems with the police, fights, school failure, and accidents).
Conclusion: Given the high rate of participation to the consultation and the fact that two out of three patients said they learnt something then, the result is very encouraging. Yet, the rate of new alcohol intoxication after the consultation has to be reduced. However, this kind of ambulatory consultation remains a good alternative to the systematic hospitalisations, especially in terms of health costs. Moreover, it gives a chance to get an early contact with a specialist and a follow-up, if necessary.

Date: 19/06/2008
Time: 16:15–17:30

PED 2-4

Effects of late cord-clamping on cerebral and systemic circulation in premature infants (vlbw)

M. Nelle[1], O. Linderkamp[2], C. Höcker[2].
1 Div. of Neonatology, Univ. Childrens Hosp. Berne; 2 Div. of Neonatology, Univ. Childrens Hosp. Heidelberg, Germany

Aim: Was to investigate the effects of late vs. early cord clamping on systemic circulation and cerebral blood flow velocity (CBFV) in prematures <1500 g (VLBW).
Subjects: 34 neonates were studied 4 h after caesarean section. In 18 neonates (birth weight: 1240 ± 290 g; gestational age: 30.0 ± 2 wks) the umbilical cords were clamped after 30 seconds and the infants were placed 30 cm below placenta level (Late), and in 16 (1231 ± 350 g; 28.6 ± 2 wks) the cords were clamped immediately (Early). Volume expansion (serum or plasma) were given to keep the mean blood pressure (MBP) >30 mm Hg.
Methods: MBP (mm Hg), left ventricular output (LVO, ml/kg/min), mean cerebral blood flow velocity (CBFV) in the Arteria carotis interna (ACI, m/s; Doppler-ultrasound), hemoglobin (Hb, g/dl), and hematocrit (Hct, %) were measured. Systemic and cerebral hemoglobin transport (HbT), and systemic vascular resistance (SVR; mm Hg/kg/min^{-1}) were estimated. Statistic: *unp. t-test.
Results (4 h after birth)

	Early cord clamping (n = 16)	Late cord clamping (n = 18)	*p-value
MBP	36 ± 4	44 ± 7	0.03
LVO	241 ± 38	245 ± 25	NS
ACA	0.16 ± 0.03	0.20 ± 0.05	NS
SVR	123 ± 40	145 ± 30	0.05
Hct	0.46 ± 0.4	0.55 ± 0.5	0.002
cerebral HbT	7.4 ± 1.8	11.1 ± 4.1	0.04
systemic HbT	154 ± 27	181 ± 24	0.05

Conclusions: Late cord clamping improves MBP, systemic vascular resistance, hemoglobin, systemic and cerebral hemoglobin transport in prematures <1500 g. The early cord clamped group required more volume expansion in the first 24 h (Early: 12/16, 14 ± 7 ml/kg; Late: 4/18, 5 ± 4 ml/kg; p <0.03).

Date: 19/06/2008
Time: 16:15–17:30

PED 2-5

Effects of a 24h light-dark rhythm in neonatal care on sleep and crying patterns of preterm infants

C. Guyer[1], J. Fontijn[2], H.U. Bucher[2], O.G. Jenni[1].
1 Child Development Center, University Children's Hospital Zurich; 2 Clinic of Neonatology, University Hospital Zurich

Aims: The influence of a 24h light-dark rhythm in neonatal care on sleep and crying patterns of preterm infants was examined at corrected ages of 5 and 11 weeks.
Methods: 41 preterm infants born before 32 weeks gestational age were either nursed in the standard 24 h dim light condition (control group; n = 21) or in a cycled condition with light exposure between 7 am–7 pm and dark between 7 pm–7 am (intervention group; n = 20). Parents filled out a standardized sleep and crying diary over 3 consecutive days at 5 and 11 weeks corrected age. Variables for sleep, wake and content, fussing, crying and unsoothable crying were analysed in four 6h intervals starting at midnight.
Results: Lighting conditions differed significantly between the two groups (intervention vs control group: 511.8 lux ± 166.7 vs 99.9 lux ± 45.3). Both groups did not differ in gestational age, weight, length and head circumference at birth or discharge, or in duration of lighting exposure (intervention vs control group: 36.19 days vs 31.2 days). A preliminary analysis by repeated measure ANOVA including 29 preterm infants (15 controls, 14 intervention group) showed a significant interaction between interval and age for the variables sleep and awake and content. At age 5 weeks sleep was clustered between midnight and noon, whereas at 11 weeks sleep was shifted to the time between 18:00 and 6:00 o'clock. An opposite effect was found for awake and content. Fussing and crying showed an age (fussing: P <0.008, crying: P <0.029) and interval (fussing: P <0.00, crying: P <0.003) effect with more events at 5 weeks of age and in the evening hours. Furthermore, for fussing a significant interaction between age and intervention (P <0.024) was found with less fussing episodes at 5 weeks of age for infants nursed in the cycled condition (0.26 hours vs 0.5 hours).
Summary and conclusion: The findings of this study showed the expected redistribution of sleep and wake over the day with increasing age. Fussing and crying demonstrated the expected peak at age 5 weeks and a clustering in the evening hours. This result reflects the maturation of sleep homoeostasis during infancy with the ability of falling asleep earlier at night and therefore decreasing the amount of fussing periods in the evening hours. While no differences in sleep, wake and content, crying and unsoothable crying were found between the two conditions, nursing in a cycled light condition reduced fussing episodes at the peak age of 5 weeks.

Date: 19/06/2008
Time: 16:15–17:30

PED 2-6

Elevated cord blood levels of mannose-binding lectin-associated serine protease-2 in infants with necrotising enterocolitis – a case-control study

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Aims: Necrotising enterocolitis (NEC) continues to cause significant morbidity and mortality in premature infants. The role of innate immunity in the pathogenesis of NEC remains unclear. Mannose-binding lectins (MBL) recognize microorganisms and activate the complement system via MBL-associated serine protease-2 (MASP-2). Deficiency of either MBL or MASP-2 due to single nucleotide polymorphisms is common, but their role in NEC is unknown. The aim of this study was to investigate whether cord blood MBL and MASP-2 levels are associated with the development of NEC.
Methods: Case-control study including 32 infants with radiologically confirmed NEC and 64 controls. MBL and MASP-2 were measured in cord blood using ELISA. Uni- and multivariate logistic regression was performed.
Results: Of the 32 NEC cases (median gestational age, 30.5 weeks; range, 25 to 39), 13 (41%) were operated and 5 (16%) died. MASP-2 cord blood concentration ranged from undetectable (<10 ng/mL) to 277 ng/mL. 18 of 32 (56%) NEC cases had high MASP-2 levels (>30 ng/mL) compared to 22 of 64 (34%) controls (univariate odds ratio, 2.46; 95%-confidence interval, 1.03 to 5.85; p = 0.043). High cord blood MASP-2 levels were significantly associated with an increased risk of NEC in multivariate analysis (odds ratio, 3.00; 95%-confidence interval, 1.17 to 7.93; p = 0.027). MBL levels were not associated with NEC (p = 0.64).
Conclusions: Infants developing NEC had significantly more often high MASP-2 cord blood levels compared to controls. Elevated MASP-2 may favour complement-mediated inflammation and could thereby predispose to NEC. In contrast, the extremely low levels of MASP-2 found in most premature infants suggest that the lectin pathway of complement activation is often not fully functional at birth which may represent a protective mechanism against NEC.

Date: 19/06/2008
Time: 16:15–17:30

PSY 1-1

Childhood autism: an interdisciplinary approach

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Aim: To document the interdisciplinary approach in diagnostic examination and therapeutic treatment for children with autism spectrum disorder in Northwest Switzerland ("Basler Model")
Methods: In an expert panel we retrospectively and selectively reviewed the charts of children with autism spectrum disorders, who had been seen by the neuropediatricians and child psychiatrists during the last five years. In context of this expert discussion an interdisciplinary approach for children with autism spectrum disorder was developed.
Results: By means of presenting four representative cases we disclose both the indication for and the possibilities of an interdisciplinary approach to children with autism spectrum disorder. Cases with epilepsy, severe sleep disorders, and a metabolic disease document the indication of neurological examination and management of autistic children. First, we present a boy suffering from fragile X-syndrom and autism, who developed epileptic seizures. In this boy, a bilateral hippocampal sclerosis could be documented. Second, depict a girl with features of a global developmental retardation with autistic behaviour. A metabolic examination disclosed in this patient a dihydro-pyrimidine-dehydrogenase deficiency. Third we portray several autistic children with severe difficulties to fall asleep and the therapeutic effects of giving melatonin. Fourth we describe report about a boy, who was born preterm at 28 weeks. In the neurodevelopmental follow up autism was disclosed. Intensive early education and treatment distinctively improved the behaviour of this patient. In summary a diagram will be presented that provides an overview of interdisciplinary approach in the diagnosis and management of patients with autism spectrum disorders.
Conclusion: Autism spectrum disorder is a pervasive developmental disorder sometimes associated with neurological comorbidity. Children with neurogenetic or neurodevelopmental disorders have an increased risk to develop autistic behaviour. Both constellations situations demonstrate the need for an interdisciplinary approach to treat these children. The "Basler Model" accommodates this request.

Date: 19/06/2008
Time: 16:15–17:30

PSY 1-2

Network therapy: a systemic model

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Definition: The network therapy includes professionals, family and child; it presentifies all third parts, gives time to share the eperiences and thoughts, and looks for consens, in respective mediation, coaching, as well framing for multiproblematic and underorganised situations. It limits overreaction and overinvolvment, isolation and helpless, gossip and power positions.
Illustrations: Are illustrated two long term multidisciplinary psychopediatric guidance networks, one of of an anorectic infant, another of a a preschooler battered child.
Modelisation: The systemic model proposes a enmeshed hierarchy where the crisis within the team are complementarly monitored.
Training: An european postgraduate training in network interventions is presented in Torino, Barcelona and Nice, considering risks and resilience, intra-institutional and inter-institutional interactions, parenthood, migration, aso
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Date: 19/06/2008
Time: 16.15–17:30

PSY 1-3

Troubles anorexiques en milieu psychiatrique intrahospitalier. Quel sens et quels soins donner aux symptômes dans l'articulation du passage de l'adolescence à l'âge adulte?

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Notre communication s'articule sur quatre points: 1. Poser l'hypothèse de compréhension du symptôme anorexique comme un «frein» dans le passage à l'âge adulte. 2. Faire part de notre expérience clinique dans la prise en charge de patients présentant un trouble des conduites alimentaires (anorexie) au sein d'une unité intra hospitalière (Hôpitaux Universitaires de Genève, Service de Psychiatrie de Psychiatrie de l'enfant et de l'adolescent), sous la direction du Pr. F. Ladame jusqu'en 2006, actuellement du Pr. F. Ansermet. 3. Présenter la genèse du protocole de soin utilisé au sein de l'unité. 4. Présenter la spécificité des thérapies à médiations dans un cadre de prise en soins multidisciplinaires.
En référence à une lecture psychodynamique des troubles anorexiques, un protocole de soins a été élaboré dans une unité psychiatrique intrahospitalière. La réflexion menée en équipe pluridisciplinaire s'est orientée sur le protocole clinique à mettre en place. Elle s'est également attachée à formuler des hypothèses de mise en sens du symptôme dans la dynamique du passage de l'adolescence à l'âge adulte. La spécificité des soins s'organise autour de l'accueil de patients admis en volontaire et sous contrainte. Le travail thérapeutique repose sur des prises en soins à médiation (psychomotricité, ergothérapie, artthérapie) et groupales. Le suivi familial est un élément important dans le cadre de soins.

Date: 19/06/2008
Time: 16:15–17:30

PSY 1-4

The network, abuses, and failures: 2 clinical cases

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The objective of this paper is to reflect on the failures of the network and some of their root causes. By now it is well-known the importance of many intervening causes surrounding serious cases (e.g., autism, generalized developmental handicaps, borderline pathologies) or complicated ones (e.g. social-environmental deficiencies, psychiatric parental pathologies) that work together in a network, therapeutic and otherwise. The network for whose constitution requires as the first condition the collaboration and absence of differences, shown to be latent or hidden among the collaborators. In addition, it is well-seen when the conditions allow for it to those who are involved in these clinical cases even though it is in reality the intricacies of these networks are difficult. What is the role of the psychiatrist in these cases? Two clinical situations (cases taken from the psychological medical services of Lugano) will illustrate our thoughts on the matter. It seemed to us in effect to essentially analyze the dynamics in play in the two cases, rather than explain the problem from the quantitative point of view: basically the experience shows that the abuses and failures of the network are rather common or frequent.
In the first case, the failure in helping the parental request did not allow the needed therapeutic intervention to begin early enough, and on the other hand it impeded the formation of the professional network necessary to treat the child. In the second case, we observe the formation of a false network, whose connections are not integrated to help in the development of the child as an individual, and that wind up favouring the division of the patient into isolated compartments. Our analysis has led us to believe that on the one hand, it is the same psychopathology in play that brings the subjects to function in a divided or fragmented way, and on the other, the ambivalence and the over-projected parental attitude all play to the disadvantage of the subject, and because of this, there is a tendency to cut and divide requests for help, and it is to separate the interventions of the various professionals involved. For these reasons, many networks do not make up or do not function notwithstanding their existence.
In these conditions, it is necessary that the psychiatrist play a role in these aspects of divisions and projections present within the network, not only in his therapeutic role with the patient, but also in order to give key understanding to all of his or her colleagues involved, considering their anticipation albeit unintentional, damaging as well as taking into consideration the examples of salvation illusions.

Date: 19/06/2008
Time: 16:15–17:30

PSY 1-5

Children and adolescents with parents suffering from mental illness: facts and needs

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Introduction: Mental illness is a burden for individuals themselves, but also for partners, families and especially for children of affected parents. Those children out of families with parents suffering from mental illness are often not recognized, unless they suffer from illness themselves or get symptomatic in school or other places.
Objectives: In order to assess and then improve the living situation of children and families affected by mental illness of a parent, as a first step we sought to determine the prevalence of this condition in a reasonable large urban community in Switzerland.
Methods: Cross-sectional study in a Swiss city with 100'000 inhabitants, greater area of 220'000 people, pretested questionnaire, postal interviews (n = 455), sent to general practitioners, psychiatrists, psychotherapists, psychologists in private praxis, child psychiatrists, in-patient psychiatric clinics, community health centres, day-care-centres and others. The over-all response rate was 55 %.
Results: Final results of a recently performed representative cross-sectional study in a Swiss population will be presented, an overview will be given to demonstrate the importance of an early recognition, support and therapy of those children and their parents. The amount of affected children and adolescents (up to the age of 18 yrs.) in Switzerland was estimated around 20'000. Even though psychiatrists seem to know whether there are children in a client's family, they often don't know much about their health, quality of life and wellbeing. The cooperation of the care systems of children and adolescents with the social and health care providers of the adult system was found to be on a rather low level (over-all max. 36%).
Conclusion: Multimodal and multidisciplinary efforts are required to administer to children and parents sufficient knowledge and support to cope with partial or chronic dysfunctions in personal life, intra- and extrafamiliar interactions and in daily life activities.

Date: 19/06/2008
Time: 16:15–17:30

CHI 1-1

Bedside primary placement of spring-loaded preformed silo in laparoschisis: experience in a single center

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Aim: To share our experience with primary management of laparoschisis using spring-loaded preformed silicone silos followed by secondary closure of the abdominal wall.
Background: Traditional management of laparoschisis includes prompt reduction of the bowels and surgical abdominal wall closure within hours after birth. Staged reduction using a silo aims at reducing the risks of abdominal compartment syndrome and barotrauma from mechanical ventilation. Since 1995, preformed spring-loaded transparent silicone silos have allowed spontaneous gradual reintegration of the abdominal content, obviating the need for urgent surgery. As a consequence, secondary abdominal wall closure could be performed decreasing the risks of prolonged post-operative pulmonary ventilation secondary to increased intra-abdominal pressures.
Method: In a tertiary care university hospital, 4 children with laparoschisis have been treated with staged bowel reduction in a silo followed by secondary surgical abdominal wall closure. Children's demographics, length and type of mechanical ventilation, time to full-feed, local and general complications and length of stay have been recorded.
Results: In each child, the silo was placed at the bedside in the delivery room without anaesthesia. Initial mechanical ventilation was not required. Intestinal reintegration into the abdomen was complete within in a median of 7 days (range 6–9). Secondary surgical closure of the abdominal wall was always possible. No sign of abdominal compartment syndrome was noted and no secondary laparotomy was required. Low-pressure mechanical ventilation without inotrope support was necessary for 4 days in one child. Median time to full enteral feeding was 28 days (range 17–35). Median hospital stay was 43.5 days (range 21–112).
Conclusion: Staged intestinal reduction following primary bedside placement of a preformedspring-loaded silo was safe and well tolerated in 4 newborns with laparoschisis. While avoiding abdominal compartment syndrome, it allowed easy secondary surgical closure with no further specific complications.

Date: 19/06/2008
Time: 16:15–17:30

CHI 1-2

Temporary vac dressing in a premature baby to avoid intra-abdominal hypertension and achieve skin coverage: report of a case

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Aims: Avoiding intra-abdominal hypertension by transient closure of the abdominal wall using a silo bag is a standard treatment in neonates with large congenital abdominal wall defects. The use of a vacuum-assisted closure (VAC) dressing for secondary wound closure of the abdominal wall has become standard treatment in adults and children but its use in newborns and premature babies has to be validated.
Method: We report the experience of a 28 weeks gestation quintuplet who suffered from severe necrotizing enterocolitis with secondary small bowel fistulas. Multiple laparotomies with small bowel resections, stoma formations, abscess drainages and adhesiolysis were necessary. After the last revision, primary closure was no longer possible because of persistent distended, inflammed bowel and jeopardized abdominal wall. This led to the use of a VAC dressing to allow for secondary healing of the abdominal wall over the exposed bowel. –Vitals signs were monitored and abdominal pressure was recorded through a bladder catheter.
Results: Intra-abdominal pressures varied from 4 to 14 mm Hg. Urine output always remained above 0.5 cc/ kg x hour and extubation was possible 10 days later. The baby was 3 months old and weighted 2.7 kg when a tailor-cutted VAC dressing was applied over the exposed small bowel applying a continuous negative pressure of 75 mm Hg. It was changed in the operation theatre on a weekly basis until the bowels were no longer exposed, which occurred after 32 days. Spontaneous abdominal wall epithelialisation was observed one month after termination of the VAC dressing. Short bowel syndrome with high-output ileostomy remained, as the continuity of the bowel has not been restored yet.
Conclusions: A VAC dressing with low suction pressure was suitable for secondary wound closure of the abdominal wall in a premature baby. Intra-abdominal hypertension was avoided and spontaneous small bowel skin coverage was achieved. Fistulas could not be avoided.

Date: 19/06/2008
Time: 16:15–17:30

CHI 1-3

Small-bowel volvulus without malrotation: a challenging diagnosis

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2 Neonatologie, Universitätsspital Zürich

Aims: We present 4 cases of small-bowel volvulus without malrotation. Progression of symptoms in these cases differs markedly from classic midgut-volvulus which is usually associated with malrotation, and they are discussed within.
Methods: We retrospectively reviewed 4 cases of small-bowel volvulus without malrotation from 2005 to 2008.
Results: All 4 cases suffered from a sudden deterioration of their general condition and the final diagnosis was made intraoperatively. In 3 premature neonates the preoperative provisional diagnosis was necrotizing enterocolitis. These 3 patients presented with pain, abdominal distention and tenderness, erythema of the abdominal wall, sudden high gastric residuals without vomiting and showed radiographic signs of ileus. Intraoperatively a small-bowel volvulus without malrotation and without any other anomalies was found. Only one case showed the classic sign of bilious vomiting. This was the only term born child and the only case in which an anomaly causing the volvulus (mesenteric cyst) was found.
A resection of intestine was required (14–120 cm) in all cases.
Conclusions: Small-bowel volvulus without malrotation is a challenging diagnosis, because the classical clinical sign of bilious vomiting is often absent. Three out of our 4 cases and multiple cases described in literature show that a volvulus should always be considered when a neonate presents with rapid clinical deterioration and signs of a mechanical ileus even when the classic symptom, bilious vomiting, is absent.

	Gestational age	Abdominal distention/ tenderness	Rectal bleeding	Erythema of the abdominal wall	Radio graphic signs of ileus	Provisional diagnosis preoperative	Gastric residuals	Intraoperative findings	Age at time of symptoms (days)
1	27 2/7	+/+	(+)	+	+	NEC	+	Volvulus, no malrotation	26
2	27 4/7	+/+	–	+	+	NEC	+	Volvulus, no malrotation	32
3	31 3/7	+/+	(+)	+	+	NEC	+	Volvulus, no malrotation	27
4	37 4/7	–/–	+	–	– (gasless abdomen)	Volvulus	Bilious vomiting	Volvulus, no malrotation, mesenteric cyst	1

Date: 19/06/2008
Time: 16:15–17:30

CHI 1-4

Laparoscopic repair of congenital diaphragmatic hernia in the post-neonatal period

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Introduction: Congenital diaphragmatic hernias (CDH) diagnosed after the neonatal period are uncommon and usually associated with a better prognosis. Moreover, minimal invasive surgery have shown to be useful in their correction in children.
Methods: The files of all children with the diagnosis of CDH established after the neonatal period and operated laparoscopically in two Swiss pediatric surgical institutions were analyzed.
Results: Five patients, 2 with a right-side and 3 with a left side CDH were included in the study. Age range was between 2 months and 3 years-old. All were operated through a laparoscopic approach. 4 patients have had a laparoscopic primary closure of the diaphragmatic defect. One conversion due to technical difficulties and a large diaphragmatic defect that required a Gore-tex patch was performed. Follow-up range was from 9 months to 7 years. To date all patients are doing well. No recurrences were observed and no re-intervention became necessary.
Conclusion: A laparoscopic surgical approach to treat children with a congenital diaphragmatic hernia diagnosed after the neonatal period is a safe and feasible option.

Date: 19/06/2008
Time: 16:15–17:30

CHI 1-5

Fixed for ever?

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Fixed for ever? A severe late complication in an adult patient operated for duodenal atresia in the neonatal period
We report the case history of a 33 years old patient with Down-syndrome who was operated for extrinsic and intrinsic duodenal obstruction on the first day of his life. The early postoperative period, childhood and adolescence were uneventful. At age 25 stagnation and obstruction in the proximal duodenum let to symptoms of complicated peptic ulcer disease and the patient was treated for bleeding gastroduodenal ulcers by conservative means for more then two years. Because the patient was never symptomfree and received several blood transfusions during this time period a gastrojejunostomy was performed at age 27. But bleeding from his duodenal ulcers never stopped. Therefore we decided to reoperate the patient. The megaduodenum was resected and the papilla Vateri directly anastomosed to a jejunal loop. Three years after the operation the patient is still symptomfree and has never had any bleeding again. In cases with stagnation and obstruction of the proximal duodenum recurrent bleeding from duodenal ulcers may occur. If the papilla Vateri is part of the megaduodenum, the bowel resection must include part I and II of the duodenum and to restore the continuity the papilla must be implanted in a jejunal loop. Because late complications may occur in patients operated as newborns for duodenal obstruction, follow up into adulthood is recommended.

Date: 19/06/2008
Time: 16:15–17:30

CHI 1-6

Hematologic and subjecitve changes after subtotal splenectomy in children with hereditary spherocytosis

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2 Hämatologie/Onkologie, Ostschweizer Kinderspital, St. Gallen

Aims: To assess the hematologic and subjective effect of subtotal splenectomy in children suffering from symptomatic hereditary spherocytosis.
Methods: The study examined 13 children between 6 and 17 years with hereditary spherocytosis undergoing subtotal splenectomy between December 2002 and July 2007. The patients either suffered from symptomatic hypersplenism, sequestration, clincal signs of hemolysis or required transfusion therapy. Laboratory and clinical signs for hemolysis as well as splenic regrowth were measured and subjective changes before and after the operative treatment were monitored.
Results: Subtotal splenectomy was succsessful in 12 of the 13 children – in 1 boy, a total splenectomy had do be performed the day after the first intervention because of postoperative bleeding. In the postoperative course, the hemoglobin levels rose about 20 g/l in 3 months or about 40 g/l in 1 year in comparison with the preoperative value. Reticulocyte counts were about half of the preoperative value and Bilirubin and LDH-levels had clearly decreased. Although a regrowth of the splenic remnant was observed, hemolysis did not necessarily reoccur. Due to abdominal bleeding, one child required postoperative transfusion therapy but no re-operation. Other adverse complications such as severe infections (OPSI) were not observed. All children interviewed reported an improvement in their general condition. In the majority of the cases, these statements were affirmated by parents, teachers or relatives.
Conclusions: For children suffering from hereditary spherocytosis, subtotal splenectomy appears to be a good alternative to total splenectomy, aiming at controlling hemolysis and preserving the hematologic as well as the immunologic function.

Date: 19/06/2008
Time: 16:15–17:30

CHI 1-7

Management of patients after surgery for oesophageal atresia and other oesophageal disorders: a multidisciplinary approach

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Aim: Oesophageal Atresia (OA) encompasses a group of congenital anomalies comprising an interruption of the continuity of the oesophagus with or without persistent communication with the trachea. OA occurs in 1 in 2500 live births. Associated anomalies occur in 50% of cases. At the OstschweizerKinderspital we created a multprofessional management team taking care of the follow up for patients withoesophageal disorders, mainly for the treatment of anastomotic leaks, anastomotic strictures, gastrooesophagealreflux, tracheomalacia, dysmotility, respiratory function, scoliosis and nutritional problems. We wouldlike to present this multidisciplinary team and its management methods to pediatricians and pediatricsurgeons and show that the follow up of these often complicated patients in a setting of specialists working in a well organised team has an effect on the outcome of the disease and on the quality of lifeof the patients and their parents.
Methods: 19 patients after oesophageal atresia surgery and 11 with peptic stenosis of theoesophagus from all over Switzerland have been treated by the multiprofessional team consisting of a pediatric gastroenterologist (1), a pediatric pulmonologist (2), a pediatric surgeon (3), a pediatric psychiatrist (4), a pediatric radiologist (5) and a nutritionist (6). All patients were discussed at ourweekly conference, diagnosis and treatment depended on the major symptoms.
Results: All 30 patients were taken care of by the team, 13 patients had a significant dysphagia and were successfully treated by a total of 45 dilatations in 2 years. 14 patients were seen by our pulmonologist and bronchoscopy was performed. 16 patients had a full nutritional work up, 6 of them showing malnutrition due to selective eating or intestinal malabsorption. 2 patients are treated because of scoliosis.
Conclusion: We would like to show that it is of major importance for the management of patients with OA or other oesophageal disorders to introduce these patients to a team of different specialistsfrom the very beginning of the diagnosis and treatment. The outcome and the quality of life of thesepatients and their parents depends on the structured organization of such a team. A questionnaire isbeing created to evaluate the satisfaction of the patients and their parents. Preliminary results should be availablewithin a few weeks.

Date: 19/06/2008
Time: 17:45–19:00

CHI 2-1

Infantile systemic hyalinosis and hydrocephalus

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Aim: Systemic infantile hyalinosis (SIH) is a rare familial condition with clinical onset during the first weeks or months of life. It is characterised by deposition of hyaline material in skin and bone and severe involvement of the viscera. We would like to demonstrate a case in which a patient with infantile hyalinosis presented with hydrocephalus communicans at the age of 4 9/12 years.
Case report: After uneventful pregnancy a girl was delivered at term with a positive familiar history of infantile systemic hyalinosis. At the age of 2 months the girl developed symptoms and was clinically diagnosed to have SIH as well. A computertomography (CT) of the head at the age of 3 11/12 was done to investigate cutan findings on the scalp. It showed completely normal intracerebral findings. At the age of 4 9/12 years the girl presented with a history of repeated headache attacks for 5 days. The parents noted frequent vomiting and increased fatigue as well. On admission she showed GCS of 15, no abnormalities in her neurological findings except of a papillary edema in fundoscopy on both sides. CT head scan as well as magnetic resonance imaging of the head showed significant increase of the whole ventricular system and signs of elevated pressure without any other abnormal intracerebral findings. A ventriculo-peritoneal shunt was inserted. Postoperative CT-scan of the head showed a decrease in ventricular size and again no further abnormalities. Otherwise neurosurgical postoperative course was uneventful up to date.
Conclusion: As far as we know this is the first description of a patient with SIH and hydrocephalus. It remains unclear whether hydrocephalus can be part of the presentation of SIH, especially because so far in the literature CNS appeared normal in all patients with SIH. However this case clearly shows that hydrocephalus can be acquired in patients with SIH. We therefore recommend to have a low level of suspicion for hydrocephalus in children with SIH and to do further investigation with magnetic resonance if there is any clinical sign for possibly elevated intracranial pressure.

Date: 19/06/2008
Time: 17:45–19:00

CHI 2-2

Modified technique of mesenterico-rex shunt with insufficient length of the jugular vein graft

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3 Radiologie pédiatrique, Hôpital Civils de Lyon, France

Mesenterico-Rex Shunt (MRS) can relieve portal hypertension and restore a physiological portal flow in patients with portal vein thrombosis(de Ville de Goyet 1998). We describe a technical variant where the autologous internal jugular vein (IJV) was too short to bridge the superior mesenteric vein (SMV) and distal intra-hepatic left portal vein, also called Rex Recessus (RR).
Patient: 15 years old boy with portal cavernoma, several episodes of gastrointestinal bleeding despite repeated sclerotherapy. Pre-operative check-up, including retrograde trans-jugular portography, showed persistent esophageal and gastric varices, severe hypertensive gastropathy, obstructed portal vein, patent SMV and spleno-mesenteric confluence, patent intra-hepatic portal branches, and normal trans-hepatic pressure gradient. A MRS was planned. The left IJV was retrieved from its infra cranial part to its confluence with subclavian vein. Nevertherless, after performance of the RR to IJV anastomosis, the IJV graft proved to be too short for classical latero-terminal anastomosis on the SMV. After clamp testing, the proximal jejunal branches of the SMV were tied, the proximal SMV (first 4 cm below the pancreas) was mobilized, and a termino-terminal anastomosis between SMV and IJV was performed. Portal pressure decreased from 23 to 13 mm Hg, and intra-operative US-Doppler showed good flows in the shunt and intra-hepatic portal branches. Post operative course was uneventful and the child is alive and well, with patent shunt, 1 month after surgery.
Conclusion: This modified MRS technique may be useful in cases of insufficient length of the autologous venous graft, avoiding the need for prosthetic conduits.

Date: 19/06/2008
Time: 17:45–19:00

CHI 2-3

Kohler's disease of the tarsal navicular: a case report and review of the literature

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Case report: A healthy 6 year old boy presented for atraumatic pain and limp of the left foot. There was no swelling or bruising, and weight bearing, although possible, was painful. Plain films showed flattening, sclerosis and irregularity of the left tarsal navicular, and MRI was compatible with AVN. Diagnosis was Kohler's disease of the tarsal navicular.
Treatment consisted of a short leg walking cast for 8 weeks; the patient was pain free within 2 weeks of presentation, and remained so after cast removal.
Discussion: Kohler's disease is an idiopathic rare avascular osteonecrosis of the tarsal navicular that presents as atraumatic pain and limp in children typically between 5 and 6 years of age (1, 2), predominantly boys (3). The differential diagnosis includes unreported trauma, infection, tumor, tarsal coalition, rheumatological disease, overuse syndromes including stress fracture, and Sever's disease. In an otherwise healthy child, diagnosis may be made on history, physical examination and the characteristic aspect on plain films. Avascularity may be demonstrated, if necessary, by bone scan (3) or MRI. Further laboratory or radiological studies are warranted only for the relevant differential diagnoses. After a period of necrosis and resorption, the navicular revascularises and recovers with no functional or radiological sequelae (2, 7). Therapy is symptomatic. Eight weeks short leg casting reduces the average duration of symptoms from 15 to 3 months. (2). Similar presentation an older child (>8) are more probably Brailsford's disease, which can persist into adulthood and cause symptomatic structural changes in the tarsus. Only 2 cases of Brailsford's disease with a history of Kohler's disease have been reported (1, 4).
Epidemiology: Extrapolation of epidemiological and population data from (5, 6) shows that the number of cases in Switzerland must be on the order of 25–50/yr. Most practitioners may see only 1 or 2 cases in their careers and the rarity of this condition has therefore motivated this presentation.
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Date: 19/06/2008
Time: 17:45–19:00

CHI 2-4

Subtrochanteric rotational osteotomy for the treatment of reduced femoral anteversion

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Background: Reduced femoral antetorsion, acetabular retroversion and irregularities of the femoral head-neck-offset cause reduced internal rotation of the hip and may lead to femoro-acetabular impingement (FAI) which is nowadays regarded to be a prearthrotic condition of the hip. If the femoral antetorsion is clearly diminished, a proximal femoral osteotomy can correct the torsion and therefore reduce FAI. Newer fixed-angled implants may facilitate this procedure by allowing for a subtrochanteric osteotomy with immediate full-weight bearing with crutches instead of performing an intertrochanteric osteotomy with an angle-blade plate.
Aims: To evaluate bone healing and complication rate after subtrochanteric rotational osteotomy and fixation with 5.0 Locking Compression Plate (LCP; Synthes®) for reduced femoral antetorsion with immediate full weight-bearing.
Methods: This is a retrospective study investigating 21 consecutive children (33 hips) with a mean age of 13 years (age 9–18 years) treated for reduced internal rotation of the hip between July 2004 and October 2007 at the Children's University Hospital of Zurich. Four additional patients (six hips) operated during the same period were excluded due to concomitant surgeries prohibiting full weight bearing. Nine patients had unilateral, 12 patients single-stage bilateral correction. All patients underwent a subvastus approach for subtrochanteric osteotomy and Fixation with 5.0 LCP. Patients were allowed to bear full-weight from the first postoperative day on protected with crutches during 6 weeks. We investigated time to

union, implant failure, loss of reduction and complication rate as well as improvement of internal rotation clinically.
Results: After 12 weeks, all osteotomies were radiographically fully consolidated. None of the hips showed a loss of reduction or implant failure. Internal rotation improved from a mean of 8.6 (–5 to 20) degrees to 37.9 (30 to 60) degrees. We recorded 2 major complications (1 compartment syndrome of the lower leg, 1 incomplete sciatic paralysis), both in patients with another simultaneously performed surgical procedure.
Conclusion: Subtrochanteric rotational osteotomy is a reliable procedure to improve internal rotation of the hip. Fixation with 5.0 LCP allows simultaneous bilateral correction and full weight-bearing with crutches immediately postoperatively exhibiting a minimal risk of loss of reduction or implant failure.

Date: 19/06/2008
Time: 17:45–19:00

CHI 2-5

Psychological distress in patients with pectus excavatum as an indication for therapy

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Background: Adolescents with pectus excavatum (PE) are often affected by their body shape. The aim of our approach was to quantify the patients' individual psychological distress and to create a psychological indication for treatment.
Methods: 10 adolescents (8 male, median age 16 years, interquartile range 15–17 yrs.) with PE were examined at our psychological department. Using standardized psychological tests, projective tests and interviews psychologists validated the patients' individual psychological status. All patients were offered psychological therapy and correction of the deformity. In addition, the children were followed-up by a telephone questionnaire (median follow-up after starting therapy 12.8 months (5.9–18.0).
Results: No patient had a relevant physiological limitation. The median follow-up since presentation to our psychologists was 15.0 months (9.1–20.6). 8 patients (5 were operated, 2 used the vacuum bell, 1 will undergo surgery) had distinct psychological limitations especially concerning the dimensions attractiveness, self-esteem and somatisation. They demonstrated increased insecurity, anxiety and denegation of their body. Since all patients were within puberty the psychological distress due to the PE has to be interpreted as disadvantageous for their further development.
7 patients completed the follow-up questionnaire and reached a median score of 80.8% (76.4–86.8%), which indicates a good improvement in all patients.
Conclusions: We conclude that the psychological indication for treatment is justified, since our results support this indication.

Date: 19/06/2008
Time: 17:45–19:00

CHI 2-6

Latissimus dorsi muscle-flap over gore-tex patch for coverage of large thoracic defects in paediatric ewing sarcoma

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Aim: To introduce a multidisciplinary technique of complex chest wall reconstruction in children with Ewing sarcoma. Primary rib involvement accounts for 16% of paediatric Ewing sarcoma. Neo-adjuvant chemotherapy and surgical tumour resection may leave the patient with large thoracic wall defects needing complex reconstruction, taking the somatic growth potential into account.
Method and results: We report our experience in three consecutive children aged 3, 10 and 12 years, in whom single-stage resection and reconstruction were performed using a Gore-Tex Dualmesh patch, covered by a latissimus dorsi rotation flap harvested in continuity with the thoracolumbar fascia. This technique, which has not been previously described in children, can be easily adapted to the local conditions, and the surgeon can chose which vascular pedicles have to be preserved. The latest of our patients, who had vertebral hemicorporectomies, also had a vertical expandable prosthetic titanium rib anchored to stabilize the chest and help prevent subsequent scoliosis throughout growth. These three cases illustrate our collaborative efforts to keep with oncological principles while offering the patients acceptable function and aesthetic quality. The surgical strategy involved paediatric, orthopaedic and reconstructive surgeons.
Conclusion: We conclude that covering a Gore-Tex Dualmesh patch with a latissimus dorsi rotational flap is a convenient technique for complex chest wall reconstruction in paediatric oncology patients, that may be easily combined with further chest wall stabilizing surgery. Multidisciplinary collaboration is advised to design the best surgical strategy.

Date: 19/06/2008
Time: 17:45–19:00

CHI 2-7

«…, dass überall Kinderärzte und Chirurgen zusammen arbeiten, dann kann der Erfolg nicht ausbleiben…», Conrad Ramstedt (1867–1962), pioneer in early pediatric surgery

R. Weil. Chirurgische Klinik, Kinderspital St.Gallen

Pylorusresection, intraluminal dilatation of the pylorus, gastroenterostomy, pyloroplasty or temporary jejunostomy were the surgical therapies in the early 20th century in children with infantile hypertrophic pyloric stenosis, a disease first described 1888 by Harald Hirschsprung (1830–1916). Surgery however was not considered as first option due to a high post operative mortality (>60%). Mortality of 10–46% was observed in patients with a conservative regimen. The surgical breaktrough occurred, when Conrad Ramstedt (1867–1962) introduced his method of pyloromyotomy in 1912 (June 18th, 1912). Mortality rate of 7% published in a large study in 1930 prooved the efficacy of his technique. He himself by this time lost only 2 children out of 60.
We present the evolution of the therapy in infantile pyloric stenosis and honour the achievement of Conrad Ramstedt, known also as a former Generaloberarzt.
By this way, we fullfill a prediction of Professor Sauerbruch, made in the early 1920th, when he presented Ramstedt personally to his students with the following words:
«Von diesem Mann werden Sie einmal mehr hören, als die heutigen Pädiater und Chirurgen ahnen.» ("Future Pediatricians and surgeons will learn more about this man, more than we can possibly imagine at present.")

Date: 20/06/2008
Time: 11:00–12:00

PED 3-1

Efficacy of immunotherapy in children with ige-mediated anaphylaxis to hymenoptera venom: how many are protected at re-stings?

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Background: Hymenoptera venom allergies in children are of great concern because young patients tend to be more prone to re-stings than adults.
Objective: This study aims to determine the protective effect of venom immunotherapy (VIT) with and without use of emergency rescue medication (ERM, i.e. oral antihistamines and steroids) to re-stings in childhood.
Methods: 83 children (mean age 9.2 years; range 3.7–15.5) with grade III or IV allergy to bee (n = 49), wasp (n = 29) or both hymenoptera venoms (n = 5) were retrospectively followed-up via file and telephone survey between June 2006 and June 2007. Mean follow-up period was 7.7 years (range: 0.3–14.3 years) after commencement of VIT and mean duration of venom therapy was 3.6 years, with 50% of patients still undergoing VIT at time of survey. Primary endpoints included number of re-stings and percentage of systemic reactions to re-stings. As secondary endpoint we evaluated how many patients were carrying and taking ERM at re-sting. Furthermore, we investigated clinical efficacy of ERM in addition to VIT.
Results: 49 children (59%) had been re-stung 108 times by the insect they were allergic to. The rate of re-stings was 0.23 per patient per year of follow-up with no difference between bee and wasp stings. 16% of children re-stung by bees developed immediate systemic reactions as compared to 6% of patients re-stung by wasps (p = 0.25). The majority of patients (87%) carried ERM, but only 75% took it after re-sting. None used the adrenaline auto-injector. Children not taking ERM experienced significantly more frequent systemic allergic reactions to re-stings compared to those taking ERM (19% versus 4%, p <0,01). Furthermore, grades of systemic reactions were milder in children taking ERM after re-sting with 3 grade I reactions in comparison to the reactions in children not taking ERM after being re-stung: 2 grade III reactions with need of emergency medical attention, 1 grade II, and 2 grade I reactions.
Conclusions: A majority of children are being re-stung after commencement of VIT with no difference in prevalence of bee and wasp re-stings. VIT in children has been proven to be a successful therapy providing complete protection in 94% of children with allergy to wasp-venom and in 84% with allergy to bee-venom. ERM provides an effective adjunct therapy with reduced incidence and severity of systemic allergic reactions to re-stings.

Date: 20/06/2008
Time: 11:00–12:00

PED 3-2

Use of complementary and alternative medicine by patients presenting to a paediatric emergency department

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1 University Children's Hospital of Zurich;
2 Paracelsus Hospital Richterswil

Objective: Although the popularity of complementary and alternative medicine (CAM) has risen in the last decade, information about its use in paediatric patients presenting to an emergency department (ED) is still sparse.
Design and setting: A cross-sectional survey was performed of paediatric patients presenting to an urban, tertiary paediatric ED. 1143 questionnaires (68% of those distributed) were available for analysis.
Results: Overall 58% (n = 665) of all respondents admitted some forms of CAM therapy to their child, 25% (n = 291) at present illness. 31% of the respondents (n = 354) used CAM prescribed by a physician, 50% (n = 575) as self-medication. Patients presented to the ED mostly because of an infection (42% of total; 29% of these used CAM) or a trauma (38% of total; 19% of these used CAM). Parents of CAM-users were significantly older, more often born in Switzerland and had significantly higher school education than those of the non-users. Nearly two third of the administered CAM therapies were not prescribed by a physician and half of the families using CAM did not discuss this with their general practitioner. Parental requirements implied that medical professionals on a paediatric emergency department should know effects and side-effects of CAM therapies and even be able to recommend them.
Conclusion: The use of CAM by the study population is very frequent, even in trauma patients. It is characterised by a high rate of self-medication and exclusion of the physicians from the decision process. The parents demand the consideration of CAM and wish an open discussion with the medical professionals about CAM.

Date: 20/06/2008
Time: 11:00–12:00

PED 3-3

Acute alcohol intoxication in children – large increase of cases in the last decade

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Background and aim: In recent years several indicators in the media suggested rising numbers of children with alcohol abuse. To validate this suggestion we collected further data on alcohol intoxications. The aim of the study is to analyse data of inpatients of our hospital with regard to patient numbers, severity of intoxication, medical complications und surrounding socio-economic factors.
Methods: The present study is a retrospective analysis of 160 records of patients admitted to the university children's hospital Zurich due to alcohol intoxication during the period 1998–2007.
Results: Out of 160 patients the gender is nearly balanced; 81 males/ 79 females. The age range is between 1 and 16 years(y) (median 14.63 y; SD 1.44y). In 27% (n = 43) of cases the children were monitored in the intensive care unit because of an impaired state of consciousness. 52% (n = 83) of our patients had a mild hypothermia with body temperature below 36C°. (median 35.75C°). At admission blood alcohol concentration (BAC) was measured in 76% (n = 122) of cases. The median BAC was 1.78‰. During hospitalisation the children had normal blood sugar levels and no physical complications. High percentage alcohol was consumed in >65% of cases; mainly mixed with energy drinks or orange juice. Only 4 girls drunk alcopops. In 14.5% (n = 23) the patients showed additionally cannabis abuse. 21% (n = 34) had a physical injury. Two girls had signs of sexual violation. 43% (n = 69) lived in single parent families. One third had a history indicating psychosocial problems.
Conclusions: Compared to the years of 1971–97 we found a significant increase of alcohol intoxicated children, particularly in females. Although a high percentage of cases showed a severe intoxication we had no complications and the therapy was only basic supportive medical care. Because of the high number of psychosocial problems the focus during hospitalisation should be on the evaluation of the patients psychological status, social background and family interactions to avoid further substance abuse. The event of alcohol intoxication is mostly harmless but is often an indicator for significant underlying psychosocial problems.

Date: 20/06/2008
Time: 11:00–12:00

PED 3-4

Neural tube defects in switzerland from 2001 to 2007: are periconceptional folic acid recommendations followed?

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Background: Neural tube defects (NTD) are common congenital anomalies. The etiology of NTD is complex, with both genetic and environmental factors implicated. The present study was performed to analyze the birth-prevalence of NTD in Switzerland from 2001 to 2007 and to identify possible risk factors.
Methods: Diagnosed cases of NTD in all pediatric units in Switzerland and four prenatal ultrasound centers were reported to the Swiss Pediatric Surveillance Unit from January 2001 to December 2007. Patient, mother, and NTD characteristics were assessed prospectively with a questionnaire.
Results: Data of 140 newborns and fetuses with NTD were studied. The major group suffered from myelomeningocele (70%), followed by anencephaly (16%) and encephalocele (14%). The prevalence of NTD life born children between 2001 and 2007 was 0.12‰, corresponding to 9–10 affected newborns each year. About the same number of pregnancies was annually interrupted because of a fetus with NTD. Correct periconceptional folic acid supplementation was taken by 5% of the women. Remarkably, 39% of the women with an affected pregnancy were not Swiss citizens – almost twice the rate of foreigners living in Switzerland.
Conclusions: NTD remain a frequent problem in Switzerland. Although a correct periconceptional folic acid supplementation is effective in reducing the prevalence of NTD, women still do not follow these recommendations. Possible reasons are lack of awareness and communication problems. Consequently, only a public health policy including folic acid fortification of food is likely to result in significant prevention of NTD.

Date: 20/06/2008
Time: 11:00–12:00

PED 3-5

Sexual games at school: the importance of the network

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2 Groupe Protection de l'Enfant, Département de l'enfant et de l'adolescent, Hôpital Universitaire Genève; 3 Service Santé de la Jeunesse, Office de la Jeunesse, Genève

Aims: Children regularly exhibit sexual behaviour and participate in sexual game at school. It is therefore important for the paediatrician to assess what can be considered normal for a child in relation to his developmental age and to be able to recognise the pathological attitude called sexual behaviour problems (SBP). SBP must be managed with the network put in place in order to reduce parental and teachers' anxiety.
Method: SBP relates to children aged between 2 and 12 years old. SBP are defined as children initiating coercive behaviour involving sexual body parts, in a manner that is developmentally inappropriate and potentially harmful to themselves or others. The first step will be to assess if the child's behaviour is problematic or not. The reference schools is the Child Health Service ("Service Santé de la Jeunesse" or SSJ) represented by nurses and paediatricians as well as the "Service médico-pédagogique» (SMP), itself composed of child psychologists and psychiatrists. Sometimes, paediatricians from a private practice or a hospital can be the first interlocutor for the parents. In case of confirmed SBP, the second step will be the psychological and somatic investigation to help the children involved, as well as their parents and teachers. It is very important at this stage to properly coordinate the network. The third step is to organise for the child's care: therapeutic, legal, social, etc.
Results: In 2007, 50 cases of sexual problems were addressed to the SSJ. The majority of them concerned sexual games and were satisfactorily managed by the intervention of the SSJ, which consisted of discussions with the children, parents and teachers. When necessary, sexual educators intervened in the classroom. About 20% of the cases required the intervention of the Emergency Unit of the SMP (EU); these cases were serious or generated a lot of emotions. The EU offered the psychological help to the persons involved as well as a class intervention when needed. Among the situations evaluated by the EU, 30% were sexual games, 20% SBP and 50% in between. The small number of SBP was not correlated with the importance of adults' emotional reactions.
Conclusion: Parents and teachers need rapid answers to their questions. A correct assessment of the children's sexual behaviour and a good communication between professionals can therefore help to reduce the emotional responses and improve the outcome.

Date: 20/06/2008
Time: 11:00–12:00

PED 4-1

Management of cryptorchidism in children: guidelines from the swiss paediatric urology group

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Aim: To develop clinical guidelines on the management of cryptorchidism in pre-pubertal boys, from timely diagnosis through therapy to long-term follow-up and prognosis.
Method: These guidelines are based on a systematic review of the literature conducted by a group from the Centre Hospitalier Universitaire of the Canton de Vaud (CHUV), which is currently submitted for publication. The method used in this review will be available upon publication, but can be requested from the corresponding author. This work was proposed for discussion to the newly created Swiss pediatric urology group (Swiss PU) during their first meeting in Berne in January 2008. At the end of the day, the attending members of Swiss PU endorsed the following statements and recommendations.
Results: Five definitions, twenty recommendations and eight statements were drafted. The degree of the underlying evidence and the strength of the recommendations are indicated. As is true for most clinical guidelines, our statements and recommendations have no coercive value; they should rather be a decisional help for the physician and provide a basis for argumentation, supported by the best scientific evidence available to date. This work represents the opinion of Swiss PU in the management of cryptorchidism in non-syndromic children.
Conclusions: Cryptorchidism is best diagnosed clinically, and treated by surgical orchidopexy, without a routine biopsy. The surgical evaluation should be performed between 12 and 18 months of age, or upon diagnosis if this comes later. If no testis is palpable, or if other signs of hypovirilization such as hypospadias are present, chromosomal sex and hormonal status must be assessed. We do not advise hormone therapy. Laparoscopy is the best way to diagnose and manage intra-abdominal testes.

Date: 20/06/2008
Time: 11:00–12:00

PED 4-2

Children with disorders of sexual differentiation (dsd) and their parents need an early multiprofessional care to cope with the particular sex development

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The presentation of ambiguous genitalia in newborns is considered as a diagnostic and psychosocial emergency. Early mistakes in management may result in long lasting psychosocial disturbances and unfavorable medical outcome. To examine the problems and needs of the patients and their families, a multicentre inquiry (Netzwerk Intersex) was performed recently, results of the Eastern Swiss patients being presented here.
Methods: 9 families of patients with DSD were assessed for satisfaction with medical and psychological care, knowledge on diagnosis, gender role and quality of life by questionnaires and had a visit and counseling by a trained psychologist. 15 parents, 10 of their children and 2 adult patients were examined, while 2 adult patients, in whom sexual ambiguosity was minimal and diagnosis made during puberty, declined to participate.
Results: Diagnoses encompassed androgen insensitivity (4 female (f) sex of rearing, 1 male (m)), congenital adrenal hyperplasia (CAH, 4 f), mixed gonadal dysgenesis (MGD, 3f, 1m), 46XY-DSD with adrenal insufficiency (3 f) and oviduct persistence (1m). In 2 patients (CAH 4 y, and MGD, 7 y), gender was reassigned from male to female after the 3rd month of life, with psychological counseling, so far without adverse effects. The most frequent complaints of parents concerned the newborn period, including incompetent and confusing medical advice, disparaging care, time pressure to decide on sex of rearing and lack of psychological and emotional support. The endeavour of our recently established multiprofessional team of endocrinologists, surgeons, social workers and psychologists specialized in pediatrics and DSD, is to reduce these problems to those of acceptance of chronic disease needing continuous medical care and surgical interventions. So far results are very encouraging.
Conclusion: Counseling parents of newborns with DSD in the presence of a psychologist is important, because medical personnel may be unaware of psychic distress. To ensure a straightforward diagnostic process during the diagnostic window of postnatal "puberté en miniature", specialists with expertise in DSD are to be involved immediately after birth. However, surgical interventions and gender assignment should be delayed until a precise diagnosis and therapeutic concept have been set up with experts and parents or patients to guarantee optimal health and well-being of the patient.

Date: 20/06/2008
Time: 11:00–12:00

PED 4-3

Exercise training increase bone mineral density in children with type 1 diabetes

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Aims: Osteoporosis is a common long-term complication of type 1 diabetes (T1DM). This study aimed to assess the effects of a 9-month impact exercise training program on bone mineral density (BMD) in children with T1DM.
Methods: This was a clinical controlled trial including 27 children with T1DM (age 10.5 ± 2.4) divided in 2 groups: Exercise (n = 15) and Control (n = 12). We assessed whole body, lumbar, spine (L2-4) and femoral neck BMD; fat and lean tissue mass (LTM) by dual-energy x-ray absorptiometry (DXA Lunar Prodigy™); anthropometric measures, disease duration, glycemic control (HbA1c), physical activity by accelerometer before and after the 9-month training program. The intervention consisted of two 90-minute sessions per week of impact physical activities (rope skipping, jumping, ball games and gymnastics).
Results: There were no significant differences among groups at baseline. Three patients had whole body BMD z-score below –1 (osteopenia) and none of them had osteoporosis. The intervention resulted in significant changes in height (1.27 ± 0.53 cm, 95%CI: 0.17-2.36; p = 0.02), weight (1.27 ± 0.59 kg, 95%CI: 0.04-2.50; p = 0.04), and whole body BMD (0.017 ± 0.007 g/cm², 95%CI: 0.001-0.033; p = 0.03). Bone mineral density did not increase significantly at other sites. LTM increased in the training group (p = 0.001), whereas fat mass increased in the control group (p = 0.04). However, intervention effects were not significant among groups.
Conclusions: We demonstrated that regular physical activity with impacts on the skeleton (jumping, running) improves whole body bone mineral accretion in children and adolescents with T1DM. They should therefore be encouraged to practice regular activities to enhance peak bone mass and prevent osteoporosis later in life.

Date: 20/06/2008
Time: 11:00–12:00

PED 4-4

Role of the antenatal and postnatal ultrasound in the diagnosis of vesicoureteral reflux

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Antenatal hydronephrosis (ANH) is a frequent anomaly found on foetal echography. Actually there is no consensus recommendations for the postnatal follow-up and the necessity to practice a voiding cystouretrography (VCUG) for the diagnosis of the vesicoureteral reflux (VUR), thus leading to a number of unnecessary and irradiating examinations. The goal of this study was to evaluate the role of the antenatal and postnatal ultrasound in the diagnosis of VUR in neonates presenting ANH.
Methods: we prospectively followed 121 patients with the diagnosis of ANH defined as a foetal renal pelvis 05 mm after 20th week of gestation. All infants had two successive ultrasound examinations of the urinary tract at 5 days and 1 month after birth. Only children with persistence of dilatation with an anterior posterior diameter (APD) 05 mm or presenting an ureteral dilatation on one or both postnatal ultrasound had a VCUG at 6 weeks after birth.
Results: 89 children (74%) had a VCUG and a VUR was detected in 10 patients (11%). Among those, 5 had a high grade reflux (> grade II) with a maximal postnatal APD of 14 mm ± 7. We found a positive correlation between the severity of reflux and the degree of APD on the antenatal and postnatal ultrasound (Mann-Whitney test, $p < 0.05$). To determine the sensibility and specificity of the antenatal and postnatal ultrasound for the VUR, we used the ROC curve and found with a cut off level of 7–9 mm a sensibility of 90% and specificity of 12.6% for the antenatal ultrasound and a sensibility of 90% and specificity of 44.3% for the postnatal ultrasound. Interestingly all children with a severe VUR had an APD 010 mm on the antenatal and prenatal ultrasound.
Conclusion: Although we observed that the ultrasound had a poor specificity for the diagnosis of the VUR, this study demonstrated the utility of the antenatal and postnatal ultrasound in selecting patient at risk for a severe VUR. We recommend that all the newborns with an antenatal hydronephrosis 07 mm have an ultrasound at 5 days and 1 month after birth; the VCUG should be done at 6 weeks if one of the ultrasound showed the persistence of an APD 010 mm or the presence of a ureteral dilatation.

Date: 20/06/2008
Time: 11:00–12:00

PED 4-5

Abdominal ultrasound in the first three days of pyelonephritis has no immediate impact on its management

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Background: Diagnosis of pyelonephritis in children implies a subsequent complex management including imaging studies. Recently, we demonstrated oral antimicrobial treatment to be safe, effective and feasible in children >6 months of age, allowing for outpatient management. Next, in a prospective study we asked if routinely performed abdominal ultrasound within three days from diagnosis of pyelonephritis is needed in an attempt to further simplify its management in children.
Patients and methods: Children presenting in the emergency department with suspected diagnosis of pyelonephritis based on clinical signs, symptoms and leukocyturia between 10/ 2007 and 02/2008 were enrolled. Diagnosis of pyelonephritis was confirmed by positive urine culture. Children >6 months were treated with ceftibuten for 14 days; children 2–5 months with ceftriaxon for 3 days, followed by cefixim for 11 days, children <2 months with parenteral amoxicillin and gentamicin for 14 days. Abdominal ultrasound was carried out within 3 days from diagnosis to rule out abscess, pyonephrosis, or other abnormalities predisposing to complications.
Results: 49 children (29 girls; mean age 2.4, median 0.5, range 0.02–19.1 years) with urine culture confirmed pyelonephritis and abdominal ultrasound within 3 days from diagnosis were included. Ultrasound in 3 children indicated suspicion for a yet unknown anomaly (one pelvic kidney, one hydroureteronephrosis + primary obstructive megaureter, one duplex kidney with slightly dilated upper-pole pyelon) and confirmed known anomalies in 4 other children (vesicoureteric reflux in two, megaureter and multiple renal cysts in one each). In addition, ultrasound supected infravesical obstruction by swelling of bladderwall in one and of duplex kidney in another child. With respect to the management of pyelonephritis none of 9 children with an anomaly or obstruction visualized by ultrasound led to change of medical treatment or urged for surgical intervention or hospital admission.
Conclusions: In this series in children from all ages abdominal ultrasound within 3 days from diagnosis of pyelonephritis had no immediate impact on its management. Nevertheless, before abstaining from performing ultrasound at diagnosis of pyelonephritis can be recommended, it remains to be seen if initial ultrasound findings in comparison with follow-up studies provide information on upper urinary tract involvement potentially influencing further management.

Date: 20/06/2008
Time: 11:00–12:00

PSY 2-1

A child psychiatry unit functioning within a paediatric ward

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Following an evaluation of the needs of children and adolescents aged from 0 to 18 years, the Vaud Canton in Switzerland encouraged the creation of specialised psychiatric care within open setting general paediatric wards in certain general hospitals.
In 2007 such a unit, with a 4 bed capacity, was inaugurated in the paediatric ward of Aigle general hospital. We are currently assessing interdisciplinary collaboration and cohabitation of our unit within this setting.
We intend to present this particular functional concept in relation to interdisciplinary and networking practices from both child psychiatry and paediatric perspectives. Our aim is to discuss difficulties encountered in setting up such a unit, conceptual differences between professional backgrounds, and mutual gains resulting from close collaboration between these two initially diverse professional fields.

Date: 20/06/2008
Time: 11:00–12:00

PSY 2-2

Connecting to the network: the role of cpe in ticino in the care chain of psychological treatment in childhood

G. G. Magnolfi. Servizio medico-psicologico di Lugano, Lugano-Viganello

The CPE, health care semi-residential structures being part of the Territorial Medical-Psychological Services in Ticino exist from more than 40 years. The methods of care and, in part, the cases relating to these services, have been changed in recent years, trying to respond with greater attention to the demands and needs of the territory. Their organization, formerly quite "closed" and protective towards children suffering from very serious mental illnesses, now increasingly favours the exploitation of resources of the little patient and his family, the alliance with their healthy side "not to play the game of psychosis" (Green), working on possible children integration in social groups they belong to.
The possibility to apply a part-time care, realized for almost all of our children, then, if favourable conditions are built to allow it, leads to the development of an individualized therapeutic project in an integrated and multidisciplinary work within the network of care, still capable of improvement in his future

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Date: 20/06/2008
Time: 11:00–12:00

PSY 2-3

One year of "maternology"

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Since April 2007, there is an agreement between obstetricians, child psychiatrists and midwives of the birth ward of the regional Ticino Hospital Beata Vergine in Mendrisio, which aims to offer the possibility of consulting with a child psychiatrist to pregnant women or to mothers after childbirth who feel uneasy with their new situation or show some risk of future psychicological disturbance.
The consulting is being offered by the presence of a child psychiatrist during at least one weekly staff meeting and the possibility of direct announcement of "uneasy" clinical situations by the obstetrician to the child psychiatrist. Equally, the nursing staff has the regular opportunity of discussing with the psychiatrist about apparently difficult mother-child situations, and there is the possibility of answering to a mother's spontaneous demand for psychological support.
In the aim of further sensibilization to the problems of motherhood and very early infancy, a specific training course has been started with introduction to more specific chapters of the argument, in the shape of short moments of discussion between the staff and specialists of different categories (pediatrician, obstetrician, child psychiatrist). The presence of the child psychiatrist is moreover found to be appreciated when young families are to be accompanied in their grief for intrauterine or postpartum loss of their child.
The present contribution starts from direct statements by members of the obstetric staff and aims to enumerate several situations of uneasiness that classically come to their observation and create, at least in the mind of the staff, the request for specialist aid. Starting from discussions between staff and child psychiatrist, we try to reassume some of the main obstacles to effective access to therapy despite adequate perception of psychological problems. Inside the offer of child psychiatric consulting, particular attention has been paid to the possibility of preventing postpartum depression and it's serious consequences for mother-baby relationship. We try to confront the named aim (preventing PPD) to one year's clinical findings, and to formulate other possible or necessary indications for the opportunity of child psychiatric consultation during pregnancy. We specially propose to examine under this point of view general situations of excessive anxiety or panic attacks during pregnancy.

Date: 20/06/2008
Time: 11:00–12:00

PSY 2-4

Recommendations on the treatment of adhd in children & adolescents

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ADHD is the most commonly diagnosed childhood psychiatric disorder with a prevalence of 3–5% (6–12 yrs). Treatment of ADHD in children and adolescents has previously been controversially discussed in the public. In the meanwhile, very similar recommendations if not a broad consensus amongst the associations of pediatricians and child & adolescent psychologists in the US and Europe concerning the guidelines on treatment of ADHD have been established (1–4).
A variety of specialists dealing with ADHD in clinic or in private practice therefore sought to summarize the existing guidelines and to present recommendations, according to their long standing personal experience and in the tradition of the SGP and SGKJPP (5). The purpose of this paper was to improve the quality of medical care and to facilitate decision making for practioners working at a distance from academic centers.
Individual and parent education and training, counselling of teachers in preschool and in school plays an important role in understanding and providing basic information on ADHD and gives advice for the challenges of everyday life those families are facing. The use of stimulants and other drugs in the treatment algorithms has to be decided on an individual basis. Correct use of drug treatment won't cure the disease, but usually has a considerable beneficial effect on the individuals' quality of life and on reducing oppositional-defiant and aggressive behaviour.
The various options of psychotherapy and alternative treatment procedures will be discussed.

Literature

[1] Taylor et al. (2004): European clinical guidelines for hyperkinetic disorder – first upgrade. Eur Child Adolesc Psychiatry (Suppl.1) 13:I/7-I/13.
[2] American Academy of Pediatrics (2001). Clinical Practice Guideline: Treatment of the School-Aged Child with Attention-Deficit/Hyperactivity Disorder. Pediatrics 108 (4) : 1033-1044

Date: 20/06/2008
Time: 11:00–12:00

CHI 3-1

Is there an ideal age for hypospadias surgery?

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Purpose: Hypospadias is the most common malformation of the penis. The assumption that hypospadias may affect children's psychosexual development adversely is the major rational for recommending surgery early in life. We performed a comprehensive, cross-sectional investigation of the psychosexual development of boys operated on for hypospadias and compared it to a healthy control group in order to add evidence to this assumption.
Patients and methods: Sixty-eight children and adolescents (7–17 years) who had surgery for hypospadias were assessed by a standardized interview adressing penile self-perception, gender-role behaviour, sexual experiences, and sexual attitude. Scores were compared to an age-matched control group consisting of 68 boys after hernia repair. In addition charts were analyzed for patients and medical datails. Patients' knowledge of hypospadias was assessed. The Pediatric Penile Perception Score, the Gender-Role Questionnaire by Ijntema and Cohen-Kettenis, and a self-developed questionnaire on first sexual experiences and sexual attitude comprised the standardized assessment instruments.
Results: Boys with hypospadias did not significantly differ from the control subjects with regard to penile self-perception, gender-role behaviour, first sexual experiences, and sexual attitude. Younger age and the patient's knowledge of hypospadias predicted a more positive penile self-perception, while a more pronounced masculine gender-role behaviour was best predicted by younger age at final surgery.
Conclusions: The psychosexual development after hypospadias repair is similar to that of healthy children. But the later corrective surgery is completed, the more likely the patients become insecure with regard to gender-role behaviour. Puberty seems to be a critical time for all patients, during which they require regular urological follow-ups and benefit from age-appropriate information about their penile condition.

Date: 20/06/2008
Time: 11:00–12:00

CHI 3-2

Traumatic retrograde degloving of the penile skin in a child

S. Tercier, C. Gapany, P. Frey. CHUV Lausanne

Aims: Traumatic injuries to the penis are rarely reported in the paediatric literature. Iatrogenic trauma during circumcision seems to account for the larger part of injuries. While penis amputation requires complex reconstructive surgery, degloving injuries and other skin lesions with or without tissue loss most often needs to be treated by partial- or full-thickness skin grafting, or by local flaps with possible subsequent skin sensitivity disorder, chronic lymphedema and even loss of the erectile function.
Methods: We report the case of an eleven year-old boy, who sustained a blunt trauma of the groin and the penis, caused by a bicycle handlebar, which resulted in total degloving of the penile skin. At clinical examination under general anaesthesia the cavernous bodies, urethra and scrotum were found to be intact. For postoperative urine drainage and balloon catheter was inserted. After extensive irrigation with an antiseptic iodine containing solution, the almost entire shaft skin was primarily sutured to the remaining proximal skin of the penile basis. No wound drainage was applied, however topic antibiotic ointment was administered. The non-compressive dressing was changed every second day.
Results: The patient could be discharged with satisfactory wound healing, however, with signs of partial skin necrosis after 21 days. At day 40, the sutured skin showed focal superficial necrosis, which was handled conservatively. At the 3-month follow-up, necrotic areas had dried off and were removed. The underlying tissue showed full epithelium coverage, scaring was minor and cosmetically adequate. The boy described full sensitivity to touch and normal erectile function.
Conclusions: In penile skin degloving injury primary suturing proved to be an efficient therapy.

Date: 20/06/2008
Time: 11:00–12:00

CHI 3-3

Urethro-fistular calculus after exstrophy-epispadias surgery

S. Tercier, C. Gapany, P. Frey. CHUV Lausanne

Aims: Distal urethral calculi, although fairly common in children from developing countries, are rarely larger than 10 mm. Such calculi are mostly composed of struvite and uric acid.
Methods: We report the case of an eight-year-old Senegalese boy, who underwent primary bladder closure for exstrophy at the age of 16 months, followed by epispadias surgery 6 months later. The patient was lost for follow-up after exstrophy-epispadias surgery for 6 years. At 8 years of age, he presented with a fistulized distal urethra forming a diverticulum-like structure containing a round stone with a diameter of 28 mm extending from the urethra into the fistula.
Results: The fistula was opened, the stone removed, the fistula resected and urethroplasty was performed.
Conclusions: Such urethral calculus has, to our knowledge, not been previously reported in exstrophy-epispadias complexes surgery. This case shows the necessity and importance of careful long-term follow-up, as such a severe complication may develop well after primary surgery.

Date: 20/06/2008
Time: 11:00–12:00

CHI 3-4

No retro-look: retroperitoneoscopy

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Purpose: Minimal invasive techniques in pediatric urology are becoming more widely established. Retroperitoneoscopic surgery is still controversially discussed because of the technical challenge. We report our experience with retroperitoneoscopy.
Patients and method: From May 2002 to October 2007 retroperitoneoscopy was performed in 27 patients, aged 4 months to 13 years (mean 29 months) including 15 nephrectomies (3 bilateral), 11 partial nephrectomies (8 upper, 3 lower pole) and 1 resection of an abdomino-scrotal hydrocele. Follow up included regular clinical visits and ultrasound.
Results: Retroperitoneoscopy was completed successfully in 25 patients; conversion was required in 2 cases due to unclear anatomy. Mean operative time was 200 minutes. Mean hospital stay was 4.25 days. Postoperatively one patient developed fever of unknown origin an other patient showed urinary secretion from the incision. In the long term follow up 2 children developed febrile UTI, in one case due to reflux into the remaining ureteric stump. All remaining moieties are growing well and only in two patients after lower pole heminephrectomy small cystic lesions were detected. Cosmetic outcome is very satisfactory with hardly visible scars.
Conclusion: For nephrectomy and heminephrectomy retroperitoneoscopy is an effective and a safe approach. With growing experience we hope to expand the indication for reconstructive surgery as well.

Date: 20/06/2008
Time: 11:00–12:00

CHI 3-5

Compressed collagen gel matrix in urinary tract tissue engineering

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Aims: Collagen is a structural protein, which can act as cell carrier matrix, allowing cell adherence and multiplication and which is therefore extensively used for tissue regeneration, however, its mechanical properties are limited. Plastic collagen compression, a novel matrix manufacturing procedure, significantly increases its mechanical properties to neo-tissue level. This controlled, cell-independent process allows engineering biomimetic matrices.
Methods: Rat-tail collagen type I solution in acetic acid is mixed with Eagle minimum essential medium, the solution is neutralized with NaOH, prior to addition of alpha-MEM medium and cast into rectangular moulds. Once gel formation by polymerization at room temperature is completed, plastic compression of the collagen gels is achieved by loading with 120 g. Compression induces expulsion of over 95% of the water to result in a 30–40 micrometer thick matrix with increased mechanical properties. This matrix is then seeded with human bladder smooth muscle and urothelial cells. Cell growth and differentiation are analyzed, using scanning electron microscopy, conventional histology and immunohistochemistry. Cell viability and proliferation are quantified for 14 days in vitro.
Results: Both cell types adhere and proliferate on and the matrix surface forming dense cell layers after two weeks in culture. Furthermore urothelial cells show spontaneous stratification. However, smooth muscle cells seeded within the construct, assessed with the Alamar Blue assay, show no proliferation but no signs of senescence are observed. Cellular distribution within the matrix is also evaluated using conventional histology and confocal microscopy. During the in vitro culture lasting 14 days, smooth muscle cells show a tendency to migrate to the matrix surface.
Conclusions: Plastic compressed collagen matrix show significant potential for bladder tissue regeneration, as not only it offers adequate mechanical strength but also allows efficient cell seeding inside and on the matrix resulting in adequate cell proliferation and differentiation.

Date: 20/06/2008
Time: 16:30–17:30

PED 5-1

National screening program for biliary atresia: a pilot project

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2 Gastroentérologie et hépatologie pédiatrique, Hôpital des Enfants, Université de Genève

Introduction: Biliary Atresia (BA) is the most frequent etiology of neonatal cholestasis. Its incidence in Switzerland is 1/17800 births, close to other diseases, for which a neonatal screening is performed. BA is suspected in case of neonatal jaundice, discolored stools and hepatomegaly. The treatment of BA is sequential: in the neonatal period the Kasai operation must be performed rapidly. If the Kasai fails and biliary cirrhosis progresses liver transplantation is needed. Increased age at Kasai operation significantly decreases the chances of the child to survive with his own liver. In Switzerland the median age at Kasai is 68 days, late as compared to international standards. We present a national screening project aiming at earlier diagnosis and treatment of BA.
Methods: A stool color card (SCC), showing different stool colors, to be compared with the baby's stool color, can help to detect neonatal cholestasis. Such SCC are used in Taiwan and Japan, and proved to be a simple, non-invasive, efficient, low-cost mass screening method for early diagnosis and management of BA. The SCC and an informative brochure are distributed and explained to the parents after birth. Parents discuss the SCC with their pediatrician one month later. If the stool color is abnormal, the baby is sent urgently to a pediatric gastroenterologist for further investigations. Data on the SCC is communicated (by fax or a password protected website) to the study centre, where an anonymous record is hold. The original
of the SCC remains in the baby's file. The study coordinator contacts the physician of all babies with abnormal stool color, to know the final diagnosis. Consent of parents for these measures is given by signing the SCC. Feasibility of this screening program is assessed by the number of SCC received at the study centre, as compared to the national annual birth number. Efficacy is assessed by 1) the number of BA patients identified annually via the screening, 2) the reduction of age at Kasai operation, as compared to the pre-screening era, 3) the ratio of BA patients whose jaundice cleared after the Kasai operation and the ratio of patients alive with their native liver at the age of 5 years.
Conclusion: This simple screening using a SCC is expected to reduce the age at BA diagnosis, the age at Kasai operation, and the need for pediatric liver transplantation. Thus, the expected benefit is for the patient, as well as for the collectivity.

Date: 20/06/2008
Time: 16:30–17:30

PED 5-2

Performance of a whole-blood t cell interferon-gamma release assay in the diagnosis of childhood tuberculosis infection – the bern experience

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Background: BCG vaccination or exposure to environmental mycobacteria may interfere with the tuberculin skin test (TST), thus limiting the usefulness of the TST for the diagnosis of Mycobacterium tuberculosis (Mtb) infection. Novel T cell interferon-gamma release assays (TIGRA) are very specific in detecting Mtb infection, but so far the experience in pediatric populations is only limited.
Aims: We prospectively studied the agreement of TST and the TIGRA in the pediatric population of a Swiss university hospital.
Methods: Between March 2005 and January 2008 all children who had a diagnostic workup for Mtb infection were prospectively enrolled and simultaneously tested by TST and the whole-blood TIGRA Quantiferon®-TB Gold In-Tube (Cellestis Inc., Carnegie, Australia). The risk of Mtb exposure was categorized for each patient.
Results: A total of 55 patients were included (mean age, 7.25 years). Of these, 15 had active tuberculosis (TB), 28 had latent infection, 10 had no exposure, and 2 had been exposed perinatally. The BCG vaccination rate was 46%. The positivity rates of TST and TIGRA were 27.8% (22/79) and 20.3% (15/74), respectively. Four (5.4%) TIGRA tests gave indeterminate results. The agreement between TST and TIGRA was as follows: 48x TST-/TIGRA-; 12x TST+/TIGRA+; 5x TST+/TIGRA- (3 of these patients were BCG vaccinated); 3x TST-/TIGRA+. Concordance for all observations resulted in κ-value of 67% and was unaffected by the BCG vaccination status (κ = 72%). TIGRA results were significantly correlated with the degree of TB exposure (P = 0.002), while TST results were not (P = 0.054). Low mitogen responses significantly correlated with low age (P = 0.014).
Conclusions: TST and TIGRA performed similarly in this typical Swiss setting. The low rate of indeterminate TIGRA results underscores its usefulness in clinical practice. The result of the mitogen response strengthens the validity the test result and indicates that negative antigen responses in young children must be interpreted with caution. The significant correlation of the TIGRA test results with the risk of TB exposure indicates a higher specificity of this test in comparison with the TST.

Date: 20/06/2008
Time: 16:30–17:30

PED 5-3

E-asthma – web based clinical decision support for pediatric asthma patients

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2 Klinik für Kinder und Jugendliche, Stadtspital Triemli

In the past, the benefit of "Clinical Decision Support Systems" (CDSS) has been demonstrated in several studies and for various illnesses. CDSS are electronical systems providing interactive support for clinical decision making and are considered an innovative method to improve the implementation rate of evidence-based guidelines. In light of the federal «E-Health Initiative» and the expected transposition towards electonical medical record systems in most Swiss private practices within the next few years, the integration of CDSS tools in such record sytems suggests itself. With "E-Asthma" we present such a newly developed CDSS for the diagnosis and treatment of asthma within the ambulatory setting. "E-Asthma" allows quality-oriented documentation and provides interactive evidence based treatment recommendations within an electronic medical record system. In our oral presentation, we will demonstrate layout, functionality and practical applications of this module. Besides delivering improved quality management measures, «E-Asthma» may also provide an important contribution for improved asthma diagnosis and treatment leading to better care for patients with asthma.

Date: 20/06/2008
Time: 16:30–17:30

PED 5-4

Efficacy of high dose inhaled dscg on asthma control in young children

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Background: In current therapeutic guidelines disodium cromoglycate (DSCG) is only regarded as an alternative controller treatment for children with mild persistent asthma and its use in childhood asthma management is in steep decline. The overall clinical efficacy assessment of DSCG is hampered by a major contribution of early clinical trial conduction with limited sample size and suboptimal design and unknown inhaler device performance.
Aims: The aim of this ongoing randomized open labelled study is to investigate the efficacy of high dose inhaled DSCG on asthma control in children aged 6 to 16 years using an optimised perforated vibrating membrane nebulizer (e-Flow®).
Methods: Children with stable atopic asthma are included in the study. Children are randomized to either inhaled DSCG at a dose of 80mg per day, administered in three inhalations or inhaled corticosteroids (ICS) without dose adjustments, over a period of six months. DSCG was inhaled using the eFlow® device, ICS were given by PMDI. FeNO and Symptom scores were assessed monthly, whereas clinical examination, lung function, blood eosinophil counts and serum cortisol level were measured at baseline, 3 and 6 months.
Results: Preliminary Data is available for 28 children (aged 11.9 + 2.9 yrs; 12 girls). Baseline characteristics were similar in both groups. Symptom scores, lung function and serum cortisol levels remained unchanged in both groups. FeNO levels were reduced in the ICS group but not in the DNCG group. Blood eosinophil counts were lower in the DSCG group at 3 and 6 month compared to baseline.
Discussion: Given the preliminary results of the ongoing study, treatment with high doses of DSCG has similar effects on symptoms and lung function as compared to ICS in children with allergic asthma, whereas FeNO decreased only in ICS treated children. Asthma control was comparable in both treatment groups.

Date: 20/06/2008
Time: 16:30–17:30

PED 5-5

Management of acute bronchiolitis: can evidence-based guidelines alter clinical practice?

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Background: Acute bronchiolitis is the most common lower respiratory tract infection in infants and there is no evidence that drug treatment alters its natural course. Despite this, most Swiss paediatricians reported in 2001 to prescribe bronchodilators and inhaled corticosteroids (ICS). This situation led to the creation of national guidelines followed by a tailored implementation program. The aim of this study was to examine if treatment practices changed after the implementation of the new guidelines.
Methods: We sent a questionnaire on treatment of bronchiolitis to all Swiss paediatricians before (2001) and after (2006) creation and implementation of national guidelines (2003–2005). Guidelines were created in collaboration between all paediatric pulmonologists and implemented carefully using a multifaceted approach.
Results: Questionnaires were returned by 541 paediatricians (58%) in 2001, and 639 (54%) in 2006. While both surveys showed a wide variation in the treatment of bronchiolitis between physicians, reported drug prescription decreased significantly between the two surveys. For outpatients, general use (for all patients) of bronchodilators dropped from 63% to 23%, and general use of ICS from 37% to 7%. For inpatients, general use of bronchodilators and ICS dropped from 56% to 32% and from 32% to 7% respectively (all p <0.0001). The decrease was evident in all regions, among hospital and primary care physicians, and among general paediatricians and paediatric pulmonologists.
Conclusions: This is one of the first studies showing that national guidelines together with a tailored implementation program can have a major impact on medical management practices in a country.

Date: 20/06/2008
Time: 16:30–17:30

PED 6-1

Physical activity and cardiopulmonary fitness in children with chronic diseases

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Aims: This study aimed to compare daily physical activity and cardiopulmonary fitness in children with chronic diseases, such as juvenile idiopathic arthritis (JIA), Type 1 diabetes (T1DM), and obesity (OB), with healthy subjects (HC).
Methods: This was a cross-sectional study including 136 children (JIA: n = 24, mean age: 10.7 ± 3.0; T1DM: n = 16, mean age: 11.2 ± 2.8; OB: n = 38, mean age: 9.0 ± 1.5; and HC: n = 58, mean age: 9.5 ± 2.6). There was no sex difference between groups. Daily physical activity (PA) was measured for at least 4 days (mean: 6.3 ± 1.1) with an accelerometer worn at the waist during waking hours. We assessed cardiopulmonary fitness by measuring the peak oxygen consumption (VO2max) during a maximal treadmill test. PA data were compared using ANCOVA with age as-covariate.
Results: Total daily physical activity was significantly lower in JIA (504 ± 162 vs. 740 ± 535 count/minute, p = 0.021) and in OB (606 ± 274, p = 0.047) compared to HC. Physical activity was not different in T1DM compared to JIA or OB, but it was significantly higher in OB than JIA (p = 0.008). We showed that HC spent less time in light PA (68.6 %) than other groups (OB: 71.3 %, p <0.001; JIA: 73.9%, p <0.001 and T1DM: 77.9%, p <0.001); and more time in moderate to vigorous PA (31.4%) compared to T1DM (22%; p <0.001), JIA (26%, p <0.001) and OB (28.7 %, p <0.001). The differences in physical activity level between the three chronic diseases groups were also significant (p <0.001 for all). Cardiopulmonary fitness was similar between HC and T1DM. In addition, OB (36.38 ml/kg/min, p <0.001) and JIA (38.2, p <0.001) children had a significantly lower VO_2 max than HC (46.18). Obese children had also lower VO_2 max than T1DM (43.8, p = 0.002).
Conclusions: Obese and JIA children are globally less active than healthy children, JIA being even less active than obese subjects, probably due to articular pain and deconditioning. Physical activity in OB children may be reduced due to exercise intolerance, social exclusion or low self-esteem. Moreover, both groups spend less time in moderate to vigorous activities compared to healthy children. We show that this sedentary behavior is associated with low cardiopulmonary fitness. Diabetic children have similar cardiopulmonary fitness and daily physical activity compared to healthy subjects; however they do not spend as much time in moderate to vigorous physical activities. The fear of hypoglycemia during exercise may explain these findings. We conclude that the impact of childhood chronic diseases on physical activity level and cardiopulmonary fitness should not be underestimated. Physical activity prescription should be adapted to the child's capacity and fear.

Date: 20/06/2008
Time: 16:30–17:30

PED 6-2

Health-related quality of life and physical activity level are reduced in pre-pubertal obese children

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Objective: The main purpose of this study was to evaluate the associations between health-related quality of life (HRQOL), BMI, physical activity level and socioeconomic status (SES) in pre-pubertal children.
Patients and methods: This was a cross-sectional study including 94 children aged 6 to 11 years (mean 9.2 ± 1.6 y), divided in 2 groups: 47 obese and 47 lean subjects. Health-related QOL by the Swiss French version of the Child Health Questionnaire (PF50), 7-day physical activity count by Actigraph accelerometer, past 12-month physical activity, anthropometrics and SES by questionnaires.
Results: Obese children had significantly lower total HRQOL (76 ± 15 vs 85 ± 6%, p <.001) as well as physical and mental health, social, family activity and cohesion subscales, except emotional behavior and bodily pain, compared to non-obese children. Obese children had lower 7-day physical activity count (324.9 ± 67.7 vs 378.6 ± 111.5, p = 0.02) and past 12-month physical activity (0.8 ± 1.3 vs 3.9 ± 3.8, p <.001) than lean controls. Univariate regression analysis showed that HRQOL sub-scales were inversely associated with BMI and positively related to 7-day physical activity count and past 12-month physical activity. Physical activity variables were inversely associated with BMI (r = –0.35, p = 0.002). Multivariate regression analysis including BMI, gender, age, physical activity and SES showed that being obese was the only significant predictor of HRQOL (r = 0.24, p <.001). Among SES data, only a higher non university education of the father was associated with some HRQOL subscales.
Conclusion: We demonstrate that HRQOL as well as physical activity level are reduced in pre-pubertal obese children, compared to lean subjects. Body mass index but not physical activity neither SES is an independent predictor of HRQOL. We conclude that the impact of obesity on the child's physical and mental health and functioning, as well as parent's emotions should not be underestimated, even in this very young population. It is urgent to identify effective intervention to increase both HRQOL and physical activity.

Date: 20/06/2008
Time: 16:30–17:30

PED 6-3

Association of anthropometric measures and their changes to cardiovascular risk factors in children

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In adults, central obesity is a better predictor of cardiovascular risk than BMI, but data in children are scarce. Cross-sectional studies regarding the association between cardiovascular risk factors (CVRF) and BMI as compared to central obesity or direct anthropometric measures of fat mass in children are controversial and longitudinal data are lacking.
Methods: We examined a randomly selected population-based sample of 503 1st and 5th grade Swiss schoolchildren in a cross-sectional design and then followed 185 children (randomly selected control group) for 10 months as part of the KISS study (Kinder-Sportstudie). Outcome measures included highly sensitive C-reactive protein (hs-CRP), sex hormone binding globulin (SHBG), fasting insulin concentrations, the homeostasis model assessment of insulin resistance (HOMA) and blood pressure (BPmean) at baseline as well as changes over 10 months. Exposure variables included BMI, waist circumference, waist to hip ratio, waist-to-height ratio, single skinfold measurements as well as the sum of 4 skinfolds (biceps, triceps, scapula, suprailiac).
Results: After adjusting for age and sex, BMI, waist circumference and the sum of 4 skinfolds were the 3 measurements that were best associated with CVRF in children cross-sectionally and longitudinally. In cross-sectional analyses, BMI, waist circumference and sum of 4 skinfolds were significantly correlated to hs-CRP, Insulin, HOMA, SHBG and BPmean (0.28 <0.42, all p <0.001) and no clinically significant differences between those measures were observed. In longitudinal analyses, correlations between deltas of outcome and exposure variables were less correlated, but still significant for insulin, HOMA and SHBG (0.16 <0.29, all p <0.05). Stratifying by sex, pubertal status (prepubertal vs. pubertal) or weight status (normal weight vs overweight or obese) did not significantly alter these results. The additional variance in CVRF explained by other measures of obesity in addition to the base model (age, sex, BMI) was minimal for most CVRFs.
Conclusion: Compared to other anthropometric measures of obesity, we did not find any clinically relevant disadvantage of BMI used as a continuous variable in predicting traditional and novel cardiovascular risk factors both in cross-sectional as well as in longitudinal analyses.
Supported by the Federal council of Sports, Magglingen, Switzerland.

Date: 20/06/2008
Time: 16:30–17:30

PED 6-4

Physical activity levels and patterns of 7- and 11-year-old swiss schoolchildren

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It is well documented that a certain amount of physical activity (PA) has an impact on health benefits in children as well as in adults (WHO). Little is known about the activity pattern of Swiss primary school children.
Aims: The objective of the study was therefore to determine (a) the extent and intensity of PA in Swiss schoolchildren (also in comparison to children of other countries), (b) the pattern of PA within the week, (c) the factors, which are associated with low PA, and (d) participation in sport clubs.
Methods: 540 children from 26 classes were randomized and stratified by age (1st and 5th grade), geographic region (urban, rural) and by ethnicity (10–30% migrants). 391 children (73%) wore an accelerometer (MTI Actigraph) slightly superior to the right iliac crest for 7 consecutive days. Total PA (total counts/daily hours awake), as well as the time spent in moderate and vigorous PA were assessed. PA was also determined using questionnaires, which included information on the socioeconomic status. Skinfold thickness was measured with skinfold calipers.
Results: Boys were significantly more active than girls in both age groups, 5th grade children were less active than 1st grade children, even though the participation in sport clubs increased from 43 to 68% in boys and from 35 to 58% in girls. Swiss children were significantly more active compared to Portuguese, Danish and Northern American children. Total PA of Swiss children was significantly higher on weekdays than on weekends (773 ± 21 vs. 676 ± 28 cpm, $p < 0.01$). Age, female sex and overweight were significantly associated with low total PA (681 ± 38 vs. 750 ± 25 cpm, $p < 0.05$, for overweight vs. normal weight children). Media use, ethnic origin, educational status of parents, parental degree of PA, overweight and sports club participation of parents did not correlate with total PA.
Conclusion: Despite the increased participation of 5th grade children in sport clubs, total PA decreased from 1st grade to 5th grade children. Since PA was mainly reduced on weekends, activity based intervention programs should include the whole family. Special attention should be given to families with overweight children.
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Date: 20/06/2008
Time: 16:30–17:30

PED 6-5

Aspirin resistance in children after interventional cardiac catheterisation

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Background: During recent years, several large trials have demonstrated the benefit of aspirin (AS) for the prevention of arterial thrombotic events in adults. AS has also been increasingly used as an anti-platelet agent in children. However, no well-designed clinical trials assessing the clinical efficacy and dosing of AS in children have been performed. In children, AS is usually administrated at a dose between 1 and 10 mg/kg/day. The term "aspirin resistance" (ASR) describes the clinical inability of aspirin to protect individuals from arterial thrombotic events (clinical ASR) or the failure of aspirin to produce an expected response on one or more laboratory measures of platelet activation and aggregation (laboratory ASR). In adults, a prevalence of 5–51% of ASR had been reported.
Methods: The frequency of ASR in children with single congenital heart disease after interventional cardiac catheterization was studied. Diagnosis of ASR is based on results from platelet aggregometry (defined as arachidonate-aggregation amplitude 020%) and is compared to results from the platelet-function-analyser (PFA). In addition side effects were monitored.
Results: Of 71 children (median age 10y, range 10 mts-20y) that had given consent, 15 had been excluded (reasons: lost to follow up, no/incomplete blood samples, withdraw of consent, switch to coumarin postoperatively, use of EMLA local anaesthesia). Of 56 children included to date, 39 have completed the study. ASR was detected in 6 (15%) of the 39 children. Of these 6 children, 3 had a normal response to AS after doubling the dose (from 3–5 to 6–10 mg/kg), 1 child did not respond to a dose increase, and 2 were unavailable for further studies. 5 (9%) children showed minor bleedings while no thrombotic events were recorded during AS-therapy. While PFA results where abnormal in 29% of children without a bleeding history prior to inclusion, only 2 of 6 children with ASR according to aggregation-studies had normal PFA results (results will be updated at the meeting).
Conclusion: SR is detected in 15% of children after interventional cardiac catheterization. In contrast to adults, ASR can be overcome in some of these children by increasing the dose of AS. While aggregation studies are still the gold-standard to define therapy response, PFA results have a low specificity for the diagnosis of ASR in children.

Date: 20/06/2008
Time: 16:30–17:30

CHI 4-1

How is life with bladder exstrophy? A long-term follow-up study (38–69 years)

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Purpose: To described the urological, nephrological, orthopaedic and psychosocial long-term outcome of patients born with classical bladder exstrophy and treated with bilateral ureterosigmoidostomies according to Mathisen in early childhood.
Patients and methods: Out of a consecutive list of 42 patients born in Switzerland and treated at the University Children's Hospital in Zurich between 1937 and 1968, patients were located and invited to participate in this follow up study. It includes pediatric and adult chart analysis, personal history with a detailed sexual and social history including a preset range of questions about living with this malformation, assessment of renal function and morphology by MRI and orthopaedic follow-up.
Results: 25 patients (21 males and 4 females) were assessed, mean age 50 years (37–69). 13 patients (52%) had their ureterosigmoidostomie still in place, they all were completely continent. 15 (60%) patient have normal renal function or mild chronic kidney disease as assessed by serum Crea and glomerular filtration rate (MDRD). 10 out of 26 kidneys were judged normal in the MRI (n = 16) study. 2 patients had renal transplants and 1 was on hemodialysis. One patient suffered from adenocarcinoma of the colon. 76% of patients are married or live in a stable relationship, 24% have children after normal conception, and an additional 16% are fertile with technical assistance. About 30% reported no sexual activity at all or inability for sexual intercourse. All but 2 patients are professionally and socially successful. Patients have no or only mild clinical signs of hip osteoarthritis, also radiologically no or only mild degrees of osteoarthritis are detected.
Conclusion: Most patients born with bladder exstrophy and treated with ureterosigmoidostomies are able to lead very successful lives both socially and sexually, but at least 30% are severely handicapped in their sexual lives. The (small) majority of our patients have a normal renal function and a well functioning ureterosigmoidostomy. This is quite astonishing, given the fact that ureterosigmoidostomies according to Mathisen are nowadays considered to be refluxing high pressure reservoirs also at high risks for tumor formation. The orthopaedic outcome suggests that the altered pelvic geometry in BE does not pose a high risk for development of hip osteoarthritis.

Date: 20/06/2008
Time: 16:30–17:30

CHI 4-2

Anterior osteotomy of the superior and inferior ramus pubis in exstrophy closure

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Aims: To present our surgical approach for pelvic closure by pubic ramotomy in exstrophy patients.
Methods: We present the technical details of the anterior osteotomy of the superior and inferior ramus of the pubic bones and report the results of ten patients having undergone this procedure to facilitate symphysial approximation and abdominal wall closure.
Results: Primary tension-free abdominal wall closure and symphysial approximation could be achieved in all cases. Wound healing was never compromised and secondary dehiscence was never observed.
Conclusions: Considering the dysmorphic pelvis in the patient with exstrophy-epispadias complex, anterior osteotomy of the superior and inferior ramus of the pubic bones seems to be an easily performed efficient method to facilitate bladder exstrophy repair and it can be performed by the pediatric urologist. No further skin incisions or turning of the patient are necessary.

Date: 20/06/2008
Time: 16:30–17:30

CHI 4-3

Subureteral endoscopic injection using stabilized non-animal hyaluronic acid/dextranomer gel (Deflux®) as first line treatment of vesicoureteral reflux (VUR) in children

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Purpose: Endoscopic correction of vesicoureteral reflux (VUR) has become an established alternative to long-term antibiotic prophylaxis and ureteral reimplantation. At our institution, the procedure is used since 1988. A number of tissue augmenting substances have been used, and since 2002 we are using hyaluronic acid/dextranomer (Deflux®) as first line treatment. The aim of the study was to assess the data of patients treated with Deflux® and compare them to the results of a former study, including 82 patients treated with collagen injection.
Methods: Retrospective study of the charts of 79 children with VUR grade II-V, also including children presenting with additional malformations such as duplex ureter, posterior urethral valves (PUV), diverticulum or neurogenic bladder. All patients underwent subureteral Deflux®-injection. Outcome was verified with voiding cysto-urethrography (VCU) 3 months (all patients) and 12 months (22 patients) after operation.
Results: From January 2002 to August 2007, 79 children (56 girls, 23 boys) with a total of 125 ureters underwent subureteral Deflux®-injection. 59 patients presented bilateral VUR. Additional malformations were duplex ureters in 16 patients, PUV in 4 patients, diverticulum in 3 patients, neurogenic bladder in 2 patients and ectopic orifice in 1 patient. VUR grade was II in 34 ureters, III in 52 ureters, IV in 32 ureters and V in 7 ureters, respectively. VCU 3 months after infection therapy demonstrated no VUR in 56 patients (71%) and diminished VUR grade in additional 13 patients (total 87.3%). After a second injection therapy (21 patients), the postoperative VCU was negative in 16 children. Due to persistent VUR, 2 children underwent a third Deflux®-injection with success. Serious complications were not noted.
Conclusion: Subureteral Deflux®-injection is a safe method for treatment of VUR, even for high-grade reflux. It demonstrates minimal morbidity and low complication rate. The success rate is higher compared to the treatment using collagen injection. Using a differentiated injection technique (HIT), the results are even similar to these of ureteral reimplantation.

Date: 20/06/2008
Time: 16:30–17:30

CHI 4-4

Poly(acrylonitrile)-based hydrogel (Hypan®) as a therapeutic bulking agent in urology

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Aims: Vesicoureteral reflux and urinary incontinence can be treated by endoscopic injection. Injectables such as collagen and hyaluronic acid-dextranomer are commonly used, however are not offering optimal therapeutic effect yet.
Methods: In the present study a novel application for poly(acrylonitrile)-based hydrogel (Hypan®), already tested in other clinical indications, has been investigated for its aptitude as an injectable bulking agent. This non-degradable, extremely hydrophilic hydrogel has the capacity of a 6-fold swelling once implanted into the body, assuring the bulking effect. Deposits of this bulking agent have been implanted into the submucosal space of the bladder of 6 mini-pigs and the histological reaction, as well as the physical implant behavior have been studied over a time period of 6 months.
Results: The implants have developed the desired bulking effect by the above described hydrophilic swelling. They have been enclosed into a fine fibrous tissue capsule and provoked a very limited inflammatory reaction.
Conclusions: This study has showed that poly(acrylonitrile)-based hydrogel implants injected into the bladder are well tolerated and, due to their defined bulking ability, feasible for the treatment of vesico-ureteral reflux and urinary incontinence. To determine the therapeutic efficiency of this treatment the implants will be studied in a vesicoureteral reflux and urinary incontinence animal model.

Date: 20/06/2008
Time: 16:30–17:30

CHI 4-5

Endoscopic and surgical treatment of vesico-ureteral reflux in children: comparative long-term follow-up

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Aims: This retrospective study analyzes the long-term results of endoscopic and surgical treatment of vesico-ureteral reflux in children.
Methods: A cohort of 130 patients, 67 girls and 63 boys with a mean age of 30 months were either treated by endoscopic subureteral collagen injection (SCIN) for 92 and by Cohen's reimplantation surgery for 123 refluxing ureteral units. Mean follow-up was of 4.2 years varying from 1 to 8.7 years. Reflux recurrence, urinary tract infection (UTI) and kidney function were evaluated.
Results: After SCIN reflux was absent in 64 at 6 months. 20% of the initially 92 refluxing ureters were injected twice. After one or two injections reflux was absent in 71%. In 21% recurrent reflux was of grade I or II, not requiring further treatment. UTI was observed in 27%. After Cohen ureteral reimplantation reflux was absent in 96% at 6 months. UTI was observed in 23%. Renal function at diagnosis and follow-up was compared in children with bilateral grade III only. In patients treated with SCIN it was normal in 77% preoperatively and in 90% at follow-up. In patients treated by open surgery it was normal in 47% preoperatively and in 76% at follow-up.
Conclusions: For high-grade vesico-ureteral reflux re-implantation surgery remains golden standard. SCIN is indicated for low and medium grade reflux. Recurrent bacteriuria was observed more often after SCIN and pyelonephritis after open surgery. The renal function seems to be preserved with both techniques.

Date: 21/06/2008
Time: 09:15–10:30
CHI 5-1

Fast recovery following endoscopic stenting for pancreatic duct injury

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Aim: The management of paediatric pancreatic trauma remains controversial. In patients with ductal disruption some authors recommend early surgery in order to decrease duration of parenteral nutrition and prevent pseudocyst formation. We followed a minimally invasive approach for the same goals but aiming also for a shorter postoperative course and hospital stay.
Methods: We report two cases (6 and 7 years respectively) presenting to our hospital with increasing abdominal pain following a handlebar injury. Further investigations with ultrasonography and CT scan showed signs of pancreatic duct lesions with beginning pseudocyst formation. Patients were immediately started on TPN and Octreotide. As soon as the overseas ordered Pigtail stents (5 Fr diameter, 7 cm length) had arrived, ERCP was performed and placement of the pigtail stent beyond the site of duct lesion could be achieved.
Results: One day after stent placement the patients were asymptomatic. They started on clear fluids shortly after the procedure followed by build-up of oral nutrition the following day. Patients were discharged 7 and 2 days respectively following stenting and controlled with regular ultrasounds. In the first patient, the initial pseudocyst was disappeared after 3 weeks. The second patient had no pseudocyst detectable on ultrasonography on day 2. In patient 1 the stent was removed as day surgery after 4 weeks. On follow-up 4 months after stent removal abdominal ultrasound showed normal pancreas with no signs of fluid collection. In patient 2 stent removal is still pending.
Conclusion: In our cases, minimal invasive stenting of the pancreatic duct was feasible, abbreviated the natural course of complicated pancreatic duct injury and shortened the hospital stay as well as the duration on parenteral nutrition.

Date: 21/06/2008
Time: 09:15–10:30
CHI 5-2

Modified whipple's procedure for large solid and papillary neoplasm of pancreatic head

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Solid and papillary epithelial neoplasm (SPEN) of the pancreas is a rare tumour occurring mainly in adolescent girls and young women. Local growth is slow, and metastasis (mainly hepatic) are rare. The prognosis is usually good, provided complete surgical excision is achieved. We report on a 13 years old girl who discovered an abdominal mass by auto-palpation. Initial imaging work-up showed a 7 cm diameter partially cystic mass in the pancreatic head, displacing the duodenum, with normal tumour markers, and no extra-pancreatic extension. Duodenal duplication was suspected, but SPEN of the pancreas was eventually diagnosed by operative findings and surgical biopsy. The child underwent secondary total excision of the lesion by pylorus-preserving pancreaticoduodenectomy. Intra-operative frozen sections of the pancreas cut surface were free of tumour. Prior to digestive reconstruction, in order to avoid any intestinal traction or twist, the distal duodenum and jejunum were transposed behind the superior mesenteric vessels, small bowel was placed on the right side and colon on the left side of the abdomen (like after cure of midgut malrotation), with appendectomy. Bilio- and pancreatico-digestive anastomoses were performed on two separate Roux en Y loops, placed behind the jejunal loop anastomosed to the first duodenum. Post operative course was uneventful. Histology confirmed complete tumour excision, and the girl is doing well 10 months after surgery. This technique allowed a safe reconstruction of separate alimentary, pancreatic and biliary conduits after removal of a large SPEN of the pancreatic head, avoiding any traction or twist of the intestinal loops with minimal additional intestinal dissection as compared to classic techniques.

Date: 21/06/2008
Time: 09:15–10:30
CHI 5-3

Colon-Carzinom bei einem 15-jährigen Mädchen mit Turner-Syndrom

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A 15-year-old female with turner syndrome first presented to her GP with unspecific abdominal pain. A blood examination revealed slightly elevated inflammatory values. The diagnosis of a gastroenteritis was made.A few days later the pain increased dramatically and she also passed fresh blood in her stools. Further examinations, including ultrasound, colonoscopy and CT-scan, revealed a colonic cancer in the distal recto-sigmoid. The cancer had perforated the bowel wall and built an abscess.We performed a colostomy and treated her with antibiotics intravenously. After 2 blocks of Chemotherapy (Oxaliplatin/Capecitabin) we performed a tumor resection (RO) with an end-to-end anastomosis. The tumor was staged as a pT3pN0 (0/24) G2 adenocarcinoma of the mucinous subtype. She went through another 6 blocks of chemotherapy, 4 with Oxaliplatin/Capecitabin. Due to hepatotoxicity, a monotherapy with Capecitabin (2x) followed. The colostomy could be closed after 11 months. Now, 15 months after being diagnosed with cancer our patient is doing well with no signs of tumor recurrence. Colonic cancer is a very rare condition in the paediatric age group. A review of the current literature didn't show many cases. There is a case report of a patient with Turner Syndrome from Japan (2001), a report of 7 cases from England (2001), a retrospective multicenter study from Israel with 7 cases (2006), and a few others. The mortality rate is very high as the cancer is often diagnosed at a very late stage of the disease, often with metastasis. The aim of presenting this case is to alert us all and emphasise how important an early diagnosis is, and that chemotherapy combined with radical surgery and in selected cases with radiotherapy can increase the survival rate. Although colorectal carcinoma is extremely rare in children and adolescents, we have to think of it as part of the differential diagnosis of abdominal pain and anemia of unknown origin, and a late diagnosis is associated with a very poor prognosis, often being fatal.

Date: 21/06/2008
Time: 09:15–10:30
CHI 5-4

Laparoscopic gastropexy in children with gastric volvulus

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Aim: The aim of this study was to evaluate the results of our technique of laparoscopic gastropexy in children with gastric volvulus.
Methods: The files of all children with gastric volvulus operated with a laparoscopic gastropexy in 2 Swiss medical institutions were analyzed.
Results: Fifteen children were included in our study. Range of age was between 1 and 11 months. Main symptoms included sudden postprandial crying, probably related to abdominal pain, vomiting and irritability. In 3 patients apneic episodes associated with cyanosis, pallor and hypotonia were recorded. In all cases the diagnosis was established with upper GI series. Organo-axial gastric volvulus was found in all cases. The laparoscopic gastropexy was performed with an 8-mm Hg CO_2-pneumoperitoneum using 3-mm instruments and a 4 or 5 mm telescope. Our technique include 3 steps: 1) esophagofundopexy, 2) phrenofundopexy and 3) anterior gastropexy. Average time of surgery was 60 minutes. One conversion was performed. Follow-up ranged from 1 month to 7 years. To date, all patients are free of symptoms.
Conclusion: Our technique of laparoscopic gastropexy is a good option in children with gastric volvulus

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Date: 21/06/2008
Time: 09:15–10:30
CHI 5-5

Combined endoscopic and laparoscopic procedures in children

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Aims: Several combined laparoscopic and endoscopic procedures have been described in adults but not in children. The aim of this study is to report our experience in children and to assess the usefulness of these procedures.
Patients and results: Since 1999, 13 patients ranging from 2.4 to 14.2 years of age were treated by combined endoluminal and endocavitary approaches. They included: 2 endoluminal removals of large jejunal polyps far below the ligament of Treitz and thus inaccessible for conventional endoscopy; 1 localization of bleeding site and treatment of ileal vascular dysplasia; 3 assisted gastrostomies, the laparoscopy helping the PEG procedure to avoid a colonic perforation; 3 redo laparoscopic Heller procedures to assess by endoscopy the level of the myotomies; and by colonoscopy: 1 combined laparoscopic coagulation and endoscopic injection of a very severe rectal vascular Klippel-Trenaunay malformation; by combining laparoscopic mobilization of the bowel with colonoscopic polypectomy, previously inaccessible large polyps could be snared in 2 patients. All the combined surgical procedures were performed successfully without conversion. A precise timing of the procedure and the use of long atraumatic clamps is mandatory to avoid bowel distension due to endoluminal insufflation. If a perforation sould occur, diagnosis could be immediately done and subsequent suture or resection achieved by laparoscopy.
Conclusion: Combined endoluminal and endocavitary approaches to bowel lesions are feasible and appear to have several advantages in children. The described procedure increases the safety of the otherwise difficult polypectomy and also avoids laparotomy with enterotomy or bowel resection as the alternative.

Date: 21/06/2008
Time: 09:15–10:30
CHI 5-6

Percutaneous endoscopic gastrostomy in children at the Ostschweizer Kinderspital: a safe technique with limited medical complications when managed by a multiprofessional team

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Aim: Percutaneous endoscopic gastrostomy (PEG) is widely used for establishing enteral feeding in children with special nutritional needs. To identify possible risk factors for complications after PEG insertion, is one of the major aims of our multidisciplinary team, involving a pediatric gastroenterologist (1), a pediatric surgeon (2), two nurses (3) and a nutritionist (4). We consider itessential to work in such a team, meet regularly, and present ourselves as a permanent professionalteam to optimize recognition of complications such as misplacement, early dislodgement, infections,pneumoperitoneum and others as early as possible. We would like to emphasize the need of workingtogether with the Home Care service of the providers of enteral nutrition and act as a teaching center for Spitex services.
Methods: The PEG Team of the Ostschweizer Kinderspital was introduced 4 years ago due to the lack of medical support for patients with PEG. The team meets regularly, establishes guidelines for indication and postoperative follow up of the patients and their parents, is very active in the team work with the Home Care services and emphasizes in teaching the nurses of the Spitex in the care of the PEG. 46 patients (18m, 28f) are currently treated and managed in this center.
Results: Out of the 46 patients 1 patient had an interposition of the colon, 15 had local wound infections, 6 had hypergranulation and 4 had a gastroeeosophageal reflux that was not present before the gastrostomy. 2 patients died of other causes not relatated to the PEG.
Conclusion: We would like to present our team to pediatricians, pediatric surgeons and nurses as a model of good clinical practice in the management of patients with PEG. Quality of life of patients with PEG was improved significantly since the existence of the team, postoperative complications could significantly be decreased. We would like to encourage other pediatric hospitals to establish such team and hopefully work out a swiss multicenter data bank with pediatric patients with PEG.

P1

The role of medical genetics in paediatric primary care

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Aims: Birth defects still are the leading cause of death during the first year of life. Prevalence of serious congenital and genetic disorders diagnosed before 1 year of age is 2–2.5%. Long-term studies that include conditions diagnosed later in childhood give an overall estimate of 4% (2800/ year in Switzerland). Medical Genetics is changing the face of Paediatrics by identifying an increasing number of causes of human biologic variation. For the paediatricians in the Primary Care the question is important: At which point in the course of evaluation should the patient be referred to Medical Geneticists? Methods: We have tried to pinpoint some indicators that should prompt the paediatrician to consider a genetic cause or contribution to a patient's condition and act accordingly. These indicators are not 100% sensitive or specific, but they should raise the awareness of genetic influences on the patient.
Results: Developmental delay (DD) is frequent and carries a high possibility for genetic disorders. We recommend a referral to the Medical Geneticist in the forthcoming situations: when there is a severe or progressive DD, a positive family history, a child with dysmorphic features, an increase in symptoms or when the parents consider further children. Single anatomic variations are common, but two or more minor anomalies are much more likely to indicate the presence of a syndrome with genetic implications. Unusual early onset of adult diseases, abnormally severe reactions to infections or metabolic stress, unexplained laboratory values, exceptional histopathology or recurrent miscarriages of the mother are also suggestive for a strong genetic influence. For a child in whom a cause of disorder was still not recognized, a periodic reconsideration of aetiology is appropriate because of progressively increasing medical and genetic knowledge.
Conclusions: The genetic evaluation of birth defects or DD has not only medical, but also ethical, social and legal issues, as Primary Care physicians have a duty to provide information about available options. Despite evidence about clinical utility and treatment options for many genetic conditions is limited, a genetic test result is the gold standard to confirm a diagnosis and provide genetic risk information to family members. Furthermore, we are obliged to increase our understanding of the pathology of genetic diseases and the correlation between a person's genotype and phenotype.

P2

Juvenile bullous pemphigoid masquerading as physical abuse

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Introduction: Bullous Pemphigoid (BP) is a subepidermal blistering autoimmune disease that most commonly affects adults of 60 years and older, but rarely also occurs in children.
Case report: A 6-year-old boy presented with a 2-year history of recurrent skin lesions, apparently occurring after falls during playtime. Clinical examination showed multiple erosions and scabs on hands, feet, knees and elbows. He had no history of allergy or eczema, and there was no known skin disorder in his family. His wounds were surgically treated. After Discussion with the paediatrician concerning the possibility of child abuse, the child was referred for dermatologic evaluation. Antigen mapping of a skin biopsy excluded hereditary epidermolysis bullosa, while direct immunofluorescence studies (IF) revealed linear C3- and IgG-deposits along the epidermal basement membrane zone (BMZ), consistent with BP. A split skin examination demonstrated localization of the target antigen at the base of the blister, and Western Blot analysis confirmed the diagnosis of anti-p200 pemphigoid, a recently described subentity of BP. The patient was treated with oral Dapsone and topical corticosteroids.
Discussion: BP is an autoimmune subepidermal blistering disorder associated with tissue-bound and circulating autoantibodies reactive with glycoproteins of the BMZ. More than 50 cases of childhood BP have been published, with evidence of autoantibodies targeted against the human BP antigens. So-far, this is the first report of childhood BP targeting the 200-kD antigen. The disease is not life threatening and usually self-limiting, but requires long-term use of immunomodulatory and immunosuppressive agents. In the situation of otherwise unexplainable recurrent skin injuries of the extremities, the differential diagnosis should be considered, especially when discussing the possibility of child abuse or neglect.
Conclusions: Children are particularly at risk for recurrent skin injuries. In unclear situations, the possibility of physical abuse or neglect should be taken into consideration, while at the same time the physician in charge should keep in mind the differential diagnosis of rare hereditary or autoimmune dermatological diseases. These can be easily established by a skin biopsy incl. IF performed in an experienced paediatric dermatology unit.

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P3

Virtual reality (vr) based paediatric interactive therapy system (pits) for enhancement of motor learning in children with central motor impairment – a feasibility trial

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Aims: Restoration of upper-limb sensori-motor function in children with congenital or acquired motor dysfunctions is primarily based on repetitive and task-oriented training. Active participation is crucial for successful neurorehabilitation. A virtual reality based paediatric interactive therapy system (PITS) that allows children to practice specific movements of the upper limbs with immediate feedback about their motor performance has been developed by the Institute of Neuroinformatics (UZH/ETH). The objective of this clinical pilot study was to investigate its feasibility in paediatric patients.
Methods: PITS consists of bimanual cybergloves, a high-adjustable table, a flatscreen and a PC workstation and offers 3 scenarios which allow further variation and shaping of tasks. Participants were recruited from an in-patient rehabilitation setting (Rehabilitaton centre Affoltern, University Children's Hospital Zurich) and followed up over a three-week period in which PITS prototypes were integrated into the 3 times/weekly occupational therapy sessions. The outcome measures during the 3 week trial were the Melbourne Test (MT), Box and Block Test (BBT), Nine Hole Peg Test (NHPT) and a motivational questionnaire measured in pre- and post design.
Results: PITS has been tested on 4 children (mean age 13.5 y range 11–15 y, 4 boys, 0 girls). Diagnoses were traumatic brain injury, Guillain-Barré-syndrome, plexusparesis and meningomyelocele. Improvements in hand function corresponding to improved test scores in the pre- and postassessments were: MT + 11,22% ($p = 0.109$, ns), BBT + 7,8 items ($p < 0.05$, s), NHPT – 3,76 sec ($p = 0.279$, ns). During exercise patient motivation was found to be high and could be maintained over 3 weeks by using 3 different VR scenarios and variations of difficulty.
Conclusions: PITS has been tested a feasible, applicable and motivating VR-system which can be included in a rehabilitation training program for children with congenital or acquired central motor dysfunctions. Preliminary Results are promising. PITS may be an effective additive to a conventional rehabilitation program. It additionally offers the potential for use as a group therapy tool, which allows increased therapy frequency while maintaining personnel costs, as well as a tool for telerehabilitation. Further research with a greater number of participants and transfer studies are needed to investigate if the system is effective compared to real-world occupational therapy.

P4

Safety and efficacy of paediatric outpatient radiofrequency catheter ablations

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Aim: Radiofrequency catheter ablations (RFA) are frequently performed as treatment for supraventricular tachycardia in children older than 4 years of age. Aim of this study was to evaluate safety and efficacy of paediatric outpatient RFA.
Patients and methods: Between 06/2002 and 03/2007, 201 RFA were prospectively analyzed. Exclusion criteria for outpatient procedures were complex RFA in congenital heart disease, arterial access or distance to home more than 1 hour. All RFA were performed under general anaesthesia. In case of transseptal puncture, patients received a single-shot dose heparin. All patients underwent postprocedural echocardiography and electrocardiogram and were discharged within 6 hours after Conclusions of RFA. To identify potential complications after discharge, parental follow-up phone calls the day after outpatient RFA procedure were performed.
Results: A total of 65/ 201 (32%) patients aged 13.6 ± 3.8 years qualified for outpatient RFA. Accessory pathway ablations (n = 33) and atrioventricular node modifications (n = 28) were the most common RFA. A transseptal approach was performed in 24 RFA. Median procedure time was 1.5 hours (range: 1.1 ± 4.3), with a median fluoroscopy time of 10 minutes (range: 5–86). RFA was

successful in 63/65 (97%) patients. Postprocedural echocardiography with special attention for intracardiac thrombi, pleural effusion and inflow patterns from systemic veins or the coronary sinus were normal in all patients. Anaesthetic adverse events, predominantly post-interventional nausea and vomiting, were observed in 9 (10%) patients. Hospital discharge within 6 hours after Conclusions of RFA was practicable in all but one patient due to ongoing nausea. Follow-up phone calls did not reveal further complications. Recurrence of tachycardia after RFA was observed in 4 of 65 (6%) patients.
Conclusions: Outpatient RFA are feasible and safe in selected paediatric patients. No RFA related complication was observed. Anaesthetic adverse events were nausea and vomiting due to general anaesthesia. Success rate and recurrence rate of tachycardia was favourable after outpatient RFA.

P5

1-year effect of an inpatient intervention on aerobic fitness in 43 girls and boys with juvenile obesity

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Background: In most patients with juvenile obesity limitations in aerobic fitness occur, especially if the overweight exceeds the age related 98th percentile. Such decondition-ing leads to restrictions in the career prospect or even to physical disability. Specific therapeutic exercise is needed to treat those patients. The present study evaluates a 1-year follow-up of an inpatient intervention on aerobic fitness.
Methods: 43 obese girls (n = 14) and boys (n = 29) were tested at the beginning (IN), at the end of an inpatient program lasting 8 weeks (8W), after 6 month post-hospitalization program (6MFU = 6-month follow-up) and again 6 month later (12MFU = 12-month follow-up). They were 13.7 ± 2.4 years old with a body weight of 90.5 ± 19.6 kg and a body mass index of 33.9 ± 4.8 kg/m^2 (for all subjects age related percentile >98). The inpatient program was based on a multicomponent treatment and education focusing on nutritional changes, behavioral modifications and physical activity (at least 45 min/day; 50–75% of maximal heart rate; in swimming, walking, hiking, jogging, skiing, and ball games), in which the participants took part in daily organized activities. A major effort was done in planning for, and monitoring each patient's post-hospitalization program including the identification of a team of health professionals located close to the patient's home. Maximal aerobic power tests were performed on a cycle ergometer. The study was analyzed, using a two-way ANOVA with repeated measurements.
Results: VO_2max percent predicted increased throughout the inpatient program from 52.8 ± 11.1 to 69.1 ± 9.2% (p_{IN-8W}<0.001). This increase lasted for the whole outpatient period: at 6MFU 69.3 ± 11.2 and at 12MFU 67.1 ± 13.6%; $p_{IN-6MFU}$<0.001 and $p_{IN-12MFU}$<0.001; $p_{8W-6MFU}$= ns(1.00), $p_{8W-12MFU}$= ns(0.50); $p_{6MFU-12MFU}$=ns(0.45). A similar pattern occurred in both genders: boys IN 50.0 ± 9.5, 8W 66.8 ± 9.1, 6MFU 67.3 ± 10.4, 12MFU 64.6 ± 14.1%; p_{IN-8W}<0.001, $p_{IN-6MFU}$<0.001 and $p_{IN-12MFU}$<0.001; $p_{8W-6MFU}$=ns(1.00), $p_{8W-12MFU}$= ns(0.93); $p_{6MFU-12MFU}$=ns(0.81); girls IN 58.6 ± 12.3, 8W 74.1 ± 7.3, 6MFU 73.4 ± 12.0, 12MFU 72.3 ± 11.1%; p_{IN-8W}<0.001, $p_{IN-6MFU}$<0.001 and $p_{IN-12MFU}$<0.001; $p_{8W-6MFU}$= ns(1.00), $p_{8W-12MFU}$= ns(1.00); $p_{6MFU-12MFU}$= ns(1.00).
Conclusions: A specific therapeutic exercise as a component of a multidisciplinary inpatient program and a comprehensive follow-up outpatient program is a successful method to improve a clinically relevant aerobic deconditioning to an almost normal fitness in both genders.

P6

Assessing motivation, ability as well as psychosocial situation predicts efficiency of pediatric practitioners in children with obesity

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Background: Treatments of obesity by single persons are of little success. The present study Aimed to evaluate, if subgroups of obese children qualify for an intervention by a family pediatrician only.
A secondary goal was to investigate if ambulatory parameters like nutritional behaviour, physical activity and psychosocial situation co-determine the success of an intervention on obese children.
Methods: 71 obese children (BMI >97 BMI-percentile), aged 12 ± 4.5 years (28 girls, 43 boys) were divided in groups concerning nutritional behaviour and physical activity respectively personal and family resources. Motivation and ability were scored from 1 (bad) to 5 (very good). At the beginning and end of a therapeutic intervention at a paediatric practitioner (18.3 ± 11.9 months) BMI was verified and compared within the groups.
Results: All patients combined showed a moderate decrease in BMI during intervention, which was neither medically nor statistically of relevance (p = ns (0.14). Nutritional behaviour and physical activity showed significant group differences (p = 0.001 resp. 0.002) as follows: the poorer the motivation and ability concerning nutrition and physical activity, the higher the BMI above the 90. percentile. Moreover, there was a significant effect within the different groups (p = 0.006 resp. 0.001). The BMI of those patients with the lowest rating further worsened, whereas the better scored patients improved. A clinically relevant deterioration occurred especially in the groups scored 3 and 4. In line with the nutritional and physical activity assessments the personal and family resources showed significant differences between the groups as well (p <0.001, the worse the resources, the higher the BMI above the 90. percentile). But there was only a similar trend among the groups in the course of the intervention (p = ns (0.13)).
Conclusions: The possibility to predict whether patients are responders or non-responders is of particular importance for medical practitioners in terms of determining the appropriate medical intervention. Our study supports the implementation of a simple tool assessing nutritional, physical activity and psychosocial situation by medical practitioners to predict the therapeutic effect before an intervention in children and juveniles with obesity. This may improve and strengthen the efficiency of medical practitioners and may help to select those patients potentially successful with only a single intervention.

P7

Iron deficiency syndrome – an important differential diagnosis and a possible comorbid reason for an inefficient medicinal methylphenidate-therapy in children diagnosed with an attention deficit hyperactivity disorder (adhd)

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Objectives: The Iron Deficiency Syndrome (IDS) as a preliminary stage of the Iron Deficiency Anemia (IDA) often remains unrecognised. The cardinal symptoms are for instance tiredness, depressive mood, concentration deficit, interferences in attention and retentiveness. In children these symptoms can be misinterpreted as symptoms of ADHD. The beginning of an IDS pathology is postulated when Ferritin reaches a value below 50 ng/ml (range 300–30 ng/ml).
Methods: In 2007 almost all of the newly allocated children with symptoms of ADHD and children who received a Methylphenidate-therapy which could not be adjusted optimally were tested for IDS. When Ferritin values were 50ng/ml or lower, iron was substituted perorally over 3 months at least. The course was analysed both clinically and by means of standardised ADHD-questionnaires.
Results: 1) In 37 out of 65 (57%) children who were differential-diagnostically tested for IDS, a Ferritin value of 50 ng/ml or lower was detected. 2) In 22 out of 52 (42%) children diagnosed with ADHD who could not be optimally adjusted to Methylphenidate and/or suffered from disturbing symptoms such as sleep disorders, decline in appetite and depressive mood, respectively, Ferritin values of 50 ng/ml or lower were detected as well. All children were treated with iron perorally over 3 months at least. In the course, 9 out of 37 (24%) children within the first group no longer displayed any ADHD pathology. Within the second group, Methylphenidate was much more effective and negative symptoms less pronounced.
Conclusions: The Iron Deficiency Syndrome constitutes an important differential diagnosis in children with symptoms of ADHD. Children and adolescents diagnosed with ADHD and comorbid IDS displayed a better medicinal adjustment with Methylphenidate and showed significantly less often negative symptoms after an effective iron-therapy. The Iron Deficiency Syndrome decreases the activity of the iron-dependent enzymes and thus influences the metabolism of neurotransmitters.

P8

Heterozygous hypomorphic stat3 mutations in 4 swiss patients of 3 unrelated families with classic autosomal dominant hyper ige syndrome

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Aims: Hyper-immunoglobulin E syndrome (HIES) is an autosomal dominant disorder characterized by a highly elevated serum IgE, eczema, recurrent staphylococcal skin abscesses and cyst-forming pneumonia, with disproportionately milder inflammatory responses,

referred to as cold abscesses, and skeletal abnormalities. Therapy involves lifelong antibiotic and antimycotic prophylaxis, and occasionally surgical abscess drainage. The molecular defect, heterozygous mutations of STAT3 has recently been identified
Methods: We enrolled 4 patients from 3 unrelated families with the classic symptoms of HIES and looked for mutations in the gene encoding human signal transducer and activator of transcription 3 (STAT3) by sequence analysis.
Results: We found hypomorphic heterozygous STAT3 mutations in all four patients with HIES suggesting a dominant negative effect. We identified a novel R335W mutation within the DNA-binding domain in two related patients from Sri Lanka, and a V637M mutation in the other two non-related Swiss patients. The V637M mutation is one of the four known STAT3 hotspot mutations and is located within the SH2 domain. There was no difference in the clinical phenotype between patients with mutations in the DNA binding or SH2 domain.
Conclusions: For four decades the molecular basis of HIES has remained elusive. Here we show that dominant-negative mutations in the STAT3 gene result in classic multisystem HIES in four patients followed in Switzerland. The identification of STAT3 as the major causative gene of HIES will facilitate early and definitive diagnosis as well as treatment, hopefully leading to the prevention of serious infectious complications and sequelae.

P9

A 16.7 mb deletion on 18q21.2-q22.2 in a patient with dysmyelination, muscular hypotonia and mental retardation

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Partial deletions of the long arm of chromosome 18 produce a highly variable phenotype. Mental retardation, short stature, foot deformities, midface hypoplasia, hypotonia, hearing impairment, strabism, atretic or stenotic external auditory canals and genito-urinary malformations are features the most commonly reported. We report on a 14 months old boy with moderate delay of psychomotor development, muscular hypotonia, tapering fingers, short feet, brachycephaly and a dysmorphic facial phenotype (frontal bossing, broad nasal bridge). Cranial MRI showed a marked global cerebral atrophy affecting gray and white matter with dysmyelination. Extensive neuro-metabolic investigations showed normal Results. Cytogenetic analysis revealed an unbalanced de novo translocation t(16;18)(q22;q21.2) with a deletion of 16.7 Mb of the long arm of chromosome 18q21.2-q22.2 as determined by array-CGH. The interstitial 18q deletions are rare and vary in size, but proximal breakpoints are most commonly within bands 18q21.2 to q22.2. Investigations of the individuals with interstitial 18q deletions have further narrowed the critical areas for certain features. Abnormal cerebral myelination was reported to be associated with a small deleted region at 18q23 [Linnankivi et al., 2006]. In our patient the typical white matter abnormalities are present without haploinsufficiency of the assumed region. In individuals presenting with developmental delay, brain MRI findings with abnormal myelination may lead to the suspicion of a neuro-metabolic cause. In these cases, if also some of the other clinical features are fulfilled, the diagnosis of 18q deletion may be established using first 18q subtelomeric FISH and, secondly, molecular analysis.

P10

An infant with failure to thrive: insidious presentation of a group B streptococcal ventriculitis

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Aim: To illustrate a rare cause of failure to thrive.
Methods: 1. case report of a breastfed infant first presenting with a failure to thrive, later followed by symptoms of intracranial hypertension; 2. literature review.
Results: The most common cause of failure to thrive in breastfed infants is insufficiency of breast milk in an otherwise healthy child. However, progressive feeding difficulties and failure to thrive can be the very first signs of a primary group B Streptoccocal ventriculitis. This rare disease does not present with acute symptoms of sepsis or meningitis, but evolves in an insidious and apyretic way. The diagnosis is therefore often delayed and suspected only after intracranial hypertension appears. CSF-culture confirming group B Streptoccocal infection and MRI are the necessary investigations for the diagnosis. Therapy requires a ventricular shunt and long-course antibiotics.
Conclusions: Insufficiency of breast milk as a cause of failure to thrive should be made only after exclusion of several other diagnosis, among which group B Streptoccocal ventriculitis is an insidious and rare cause.

P11

Coronary artery fistula in a newborn with down syndrome

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Background: Congenital heart defects (CHD) occur in 30–60% of children with Down syndrome with ventricular septal defects and endocardial cushion defects being the commonest lesions. As children with Down syndrome may develop early pulmonary hypertension with reduced left to right shunting, a CHD may be present even in the absence of a cardiac murmur.
Patient: We report on an 8 days old, male newborn (gestation age: 39 5/7, birth weight: 3210 g), fourth child of non-consanguine parents. Pregnancy, birth and postpartal adaptation was uneventful. The newborn showed typical dysmorphic signs of a Down syndrome. Chromosomes analysis confirmed the diagnosis of a free translocation of chromosome 21. Pulse oxygen, femoral pulses and blood pressure were normal. The clinical examination revealed no signs of congestive heart failure nor a cardiac murmur. Electrocardiography was inconspicuous. Routinely performed transthoracic echocardiography displayed a dilatation of the left proximal coronary artery with a moderate coronary artery fistula from the ramus interventricularis to the right ventricular apex. Echocardiographic evaluation and follow up at the age of 3 months revealed stable haemodynamics as well as an asymptomatic patient. No treatment had to be initiated, further follow up was scheduled for semianual intervals.
Conclusions: In children with Down syndrome, an echocardiographic evaluation even in the absence of a cardiac murmur remains essential to rule out a CHD, preferably in the first weeks of life. Coronary artery fistulas may be clinically silent and remain asymptomatic during childhood. However, life-threatening complications like infective endocarditis, myocardial infarction, ventricular arrhythmias, and heart failure can develop. An early diagnosis as well as close echocardiographic follow up is mandatory to define the optimum timing of a surgical or interventional closure of haemodynamically active fistula.

P12

Path from thrombocytopenia to wiskott-aldrich syndrome

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Introduction: Thrombocytopenia in a newborn can have many different aetiologies. After exclusion of infection, asphyxia, placental insufficiency and systemic illnesses, antibody-mediated thrombocytopenia should be considered. The latter condition shows a spontaneous improvement within the first months of life. We present the case of a newborn with persisting thrombocytopenia and the path that led to diagnosis.
Case-report: Our patient was born by elective caesarean section because of a breech presentation following an otherwise uncomplicated pregnancy. On the first day of life he was hospitalized because of multiple haematomas and petechiae. An infection could be excluded. The genotyping of the maternal, paternal and the patients platelets showed an incompatibility (mother: HPA-1bb, father: HPA-1aa, child: HPA-1ab) but no antibodies could be detected. In the first few weeks of life, the platelet count increased slightly to a maximum of 106 $x10^9$/L (normal range: 150–450 $x10^9$/L). The infant developed normally but often had blood in the faeces and showed petechiae. At the age of three months the platelet count was at 20 x 10^9/L, splenomegaly was detected and anaemia had developed (haemoglobin of 66 g/L). PCR for Cytomegalovirus was positive in serum and bone marrow samples. After two weeks of intravenous ganciclovir, the serum PCR became negative but the infection recurred within a month. Additionally, the boy developed a chronic rash on face and body. With the triad of thrombocytopenia, recurrent infection and eczema we suspected a Wiskott-Aldrich Syndrome, which was subsequently confirmed by genetic testing, revealing an Arg86His mutation (c.257G>A) in the WAS gene. Haematopoietic stem cell transplantation is planned.
Conclusions: Persisting neonatal thrombocytopenia in a child with skin rash and "uncommon infection" may be early symptoms of a Wiskott-Aldrich syndrome.

P13

Human donor milk banking in Switzerland

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Background: More and more human milk banks are being set up around the world to supply preterm infants, sick newborn infants and also older recipients (eg. those under treatment for cancer) with milk

from donors. Preterm infants profit from donor milk when their own mother's milk is not available. Human milk has many advantages, such as decreasing the risk of necrotising enterocolitis that occurs frequently in preterm infants. Quality control according to evidence-based guidelines is extremely important for the operation of a human milk bank.
Objectives: The Swiss Human Milk Bank Committee was founded in early 2006 by more than 30 delegates from different hospitals that operate donor milk banks. The poster lists the benefits and rationale of human donor milk and outlines the current situation of the six official milk banks in Switzerland.
The strategies for the future, to optimise quality control of human donor milk in Switzerland for the fragile neonate, including national standardised guidelines are listed. There is a list of scientific references and a graphic display of when each milk bank was established, and whether clinical guidelines are used or not. Further information includes, donor screening, bacteriological testing of milk, quantity of human milk processed, number of donors and recipients for the year 2007.

P14

Case report: intra-arterial lysis of an acute thrombotic occlusion of the right medial cerebral artery in a girl with autoimmune cytopenia

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Introduction: We discuss the intra-arterial lysis of an acute thrombotic occlusion of the right medial cerebral artery (MCA) in a patient with acute haemolysis and partial renal failure in the context of autoimmune haemolytic anaemia and thrombocytopenia (Evans Syndrome, ES). We aim to review the risk factors and the management of acute stroke in childhood.
Case report: An 11-year-old girl who had been diagnosed with ES 8 years earlier presented with severe haemolytic crisis. High dose steroids, immunoglobulins and erythrocyte transfusions improved her condition within 72 hours. A transient decrease in urine output and an increase in creatinine and blood urea occured despite adequate intravenous hydration. After defaecation the patient developed acute left hemiplegia on the 4th day of admission. Diffusion-weighted MRI showed ischaemia of the right MCA territory due to an intravascular occlusion in segment 1. Intra-arterial thrombolysis using recombinant tissue type plasminogen activator (rt-PA) was successfully performed within three hours after the onset of neurological symptoms. The patient was able to move her left arm and leg again after 12 hours. The follow-up angio-CT and the follow-up angio-MRI after 8 days were both normal. The further diagnostic work-up remained normal.
Discussion: The incidence of stroke in childhood is 1.8 per 100 000 children per year with 8% mortality and 80% morbidity. Hemiplegia is the most common clinical presentation. Large outcome studies comparing intra-arterial lysis with conservative therapy and anticoagulation or no treatment are not available. The success of intra-arterial thrombolysis critically depends on time to treatment and the availability of adequate imaging facilities. "Weak" risk factors for thrombotic events were present in our patient: autoimmune disease, steroids, haemolysis and partial kidney failure. Vascular events have not been reported in patients with ES so far.
Conclusions: Early diffusion-weighted MRI and immediate intra-arterial thrombolysis were the cornerstones of the successful treatment of acute stroke in our patient. Outcome studies for therapeutic management of stroke in childhood are needed.

P15

The health status of children without residence permit consulting the childhood hospital of lausanne (chl)

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Aims: To integrate social, economic and medical data concerning children without a residence permit taken into care by the Childhood Hospital of Lausanne.
Methods: This is a prospective study by a question paper including the socio demographic, medical and education data of 103 children without residence permit, consulting the CHL for the first time between August 2003 and March 2006. These children were then convoked for a second check-up after 1 year, in order to allow a regular monitoring.
Results: Among the children that are native of Latin America (87%), 36% are less than 2 years old and 64% are between 2 and 16 years old. This population of children lives in precarious conditions with a family income lower than the poverty level (89% of the families live with less than 3000.– CHF/month) and an accommodation hosting more than two persons per room (72%). Despite these conditions, 45% of the children got a medical insurance during the year after their first consultation. The main reasons for consultation are mainly infectious symptoms, a check-up requested by the school or a check-up concerning new-born children. The majority of them are in a good health status and are affected by pathologies that are similar to the ones of other children of the same age (otitis, eczema, diarrhoea of nurseling, etc). At least 14% of the children are obese and 28% are overweight. Anaemia needing an iron treatment, concerns the 8% of the children whose haemoglobin was taken (43%). All children in age of education went to school during the year after the first check-up.
Conclusions: These children, mainly native of Latin America, live in very precarious conditions. At least, their general health status was good and the majority of them could benefit of regular check-ups. Prevention, focused on healthier life, is particularely important among this population characterized by a high incidence of overweight and obesity.

P16

Wolff-parkinson-white syndrome in a child with melas syndrome

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Introduction: MELAS (mitochondrial myopathy, encephalopathy, lactic acidosis, and stroke-like episodes) is a multisystemic disorder caused by different point mutations of the mitochondrial DNA.
Case report: A 12-years old boy presented with migraine-like headache, vomiting, impaired visual acuity, and recurrent focal seizures. At the age of 7, a catheter ablation because of a WPW syndrome was performed. Lactate was increased in blood (5.6 mmol/l) as well as in cerebrospinal fluid (3.7 mmol/l) and remained increased. EEG showed regular epileptic discharges on the left occipital region, MRI revealed a localized T2-hyperintensity of the left occipital cortex and a cerebellar atrophy, and echocardiography showed a hypertrophic cardiomyopathy. Molecular analysis of mitochondrial DNA in blood lymphocytes confirmed the mutation A3243G in tRNA with a mutation load of 57%. An antiepileptic therapy with levetiracetam was started, but the seizures recurred. Furthermore, a therapy with L-arginine was installed both at acute episodes and as oral daily supplementation. Seven months after diagnosis the boy shows a truncal and limb ataxia and recurrent seizures as left-sided cloni with a new epileptic focus on the right central side. The hypertrophic cardiomyopathy is not of hemodynamic significance.
Conclusions: MELAS represents a multisystemic disease. Cardiac involvement includes hypertrophic or dilated cardiomyopathy and WPW syndrome. The prevalence of WPW syndrome among patients with MELAS syndrome is much higher than in the normal population. Therefore, patients with WPW syndrome and neurologic abnormalities consistent with MELAS syndrome should be screened for the A3243G mutation.

P17

A rare cause for failure to thrive: Imerslund-Gräsbeck syndrome

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Introduction: Imerslund-Gräsbeck Syndrome (IGS) is a rare autosomal recessive disorder characterized by megaloblastic anemia with asymptomatic proteinuria. Further symptoms include failure to thrive (FTT), neurological deficits and recurrent infections. Mutations in two different genes (cubilin and amnionless gene) are known to cause a defect of the ileal vitamin B_{12} receptor, leading to selective vitamin B_{12} malabsorption, requiring lifelong vitamin B_{12} therapy.
Case report: A 25 month old boy was referred with fatigue, diarrhoea, vomiting, aphthous stomatitis and failure to thrive. The patient eats a balanced diet appropriate for age including meat. Periodic vomiting and diarrhoea were interpreted as recurrent gastroenteritis. The past medical history is otherwise unremarkable, with normal psychomotor development. His parents are first cousins of Turkish descent. Physical examination showed an alert child with pallor and obvious oral aphthous ulcers; weight was 11.9 kg (10th percentile), height 83.5 cm (10th percentile), and head circumference 50.5 cm (75th percentile). Lab Results revealed pancytopenia with severe megaloblastic anemia (hemoglobin 74g/l. MCV 113fl, MCH 41 pg), neutropenia and thrombocytopenia (83 G/l). Proteinuria was present. Vitamin B_{12} levels were not measurable (<33 pmol/l). Tests for

cystic fibrosis, celiac disease, folic acid, vitamin deficiency, metabolic disorders and infectious gastroenteritis were normal. Genetic analysis for IGS was done in light of typical clinical and laboratory findings and parental consanguinity. Treatment with intramuscular vitamin B_{12} was initiated upon clinical suspicion. One week later, gastrointestinal symptoms as well as the thrombocytopenia subsided. After one month, hemoglobin, MCV and MCH were all in normal range. Accelerated physical growth and normal development followed thereafter. Proteinuria persisted, as described in the literature.
Conclusions: IGS represents a rare etiology for a common pediatric problem. Megaloblastic anemia with characteristic clinical signs warrant further investigations of this uncommon disorder. Urinalysis for proteinuria provides further evidence, yet the final diagnosis can only be made through confirmation of specific gene mutations. Despite its rare occurrence, IGS remains an important differential diagnosis in chronic FTT, especially in the light of potential subsequent psychomotor retardation and the availability of simple and effective treatment.

P18

Neonatal hypoglycemia: dont forget the hormones!

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Background: We report an unusual but potentially life threatening cause of neonatal hypoglycemia due to the combination of two rare congenital endocrine disorders.
Case report: A 3-day old male newborn was admitted to our neonatal intensive care unit with hypoglycemia requiring i.v. glucose treatment. The child presented with dysmorphic facial features without midline defects, syndactyly of the toes, muscular hypotonia, micropenis and left inguinal testis. Laboratory investigations revealed hyponatremia and hyperkalemia. Congenital adrenal hyperplasia (CAH) was suspected and treatment started with mineralocorticoids and glucocorticoids. Further he developed direct hyperbilirubinemia. However, in contrast to the expected Results, the neonatal screening test was normal, and we found very low cortisol, 17-OH-progesteron, DHEA levels in combination with a very low ACTH. ACTH-test confirmed severe adrenal insufficiency and abdominal MRI imaging revealed hypoplastic adrenal glands. Besides the pathologic pituitary-adrenal axis, the child showed unmeasurable IGF-1 levels, low gonadotropines, and compensated hypothyroidism, strongly suggestive of multiple pituitary hormone deficiencies. On cerebral MRI the pituitary was hypoplastic with a small remnant of the adenohypophysis on the sellar floor, aplasia of pituitary stalk and ectopic neurohypophysis.
Discussion: For the neonatologist, the combination of severe hypoglycemia and micropenis should always rise suspicion of isolated GH deficiency or combined pituitary hormone deficiency. Hyponatremia can be a first sign of salt wasting in the context of adrenal insufficiency further supported by hyperbilirubinemia. Congenital adrenal insufficiency is a potentially lethal disease and is not detected by neonatal screening, unless due to congenital adrenal hyperplasia. Although molecular mechanisms of pituitary development have been in part elucidated in the past 15 years, the features in our patient are not typical of any described defect in known transcription factors.

P19

Use of complementary medicine in type 1 diabetes mellitus in Switzerland – a general survey

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Background: Type 1 Diabetes mellitus (T1DM) is caused by destruction of the beta cell leading to lack of insulin secretion. The only effective treatment consists in exogenous insulin replacement. Nevertheless, many complementary treatments have been tried in T1DM. The purpose of this study was to assess overall prevalence of use and specific modalities of complementary medicine (CM) used by our population of patients with T1DM.
Methods: A questionnaire was distributed among patients with T1DM at three different settings during three consecutive months. The questionnaire consisted of items on personal data, duration of diabetes, insulin treatment method and questions on the use of CM including personal goals and achievements.
Results: A total of 342 subjects with T1DM participated in the study. 246 were adults(119 females) and 96 were children (40 females). The adult cohort had a mean age of 41.7 years and a mean diabetes duration of 19.1 years, the paediatric subjects were 11.2 years and had a mean diabetes duration of 4.9 years. 96.7% of the adults and 51% of the children were treated with intensive insulin therapy (MDI/CSII). A total of 48 subjects (14%; 13.4% adult, 18.5% paediatric; 20 male, 28 female) clAimed to ever have used CM to treat their diabetes. The treatments were cinnamon (14), zinc (5), chromium (2), magnesium (11), vitamins (3), special beverages (10), fish oil (1), special diet (1), ginseng (3), Bach flower (1), homeopathy (12), bioresonance (5), acupuncture (2), massages (2), craniosacral therapy (1), Qi Gong (1) and Chinese quantum method (1). The primary goal as well as the primary achievement were improvement of general well-being, followed by amelioration of glucose stability, reduction of blood glucose and insulin dose, reduction of frequency of hypoglycaemia, improvement of physical fitness and reduction of appetite. The mean duration of use was 16.1 months. Mostly, the CM was recommended by friends/family members (21), family doctors (7) or complementary health professionals (7).
Conclusions: Even though T1DM is known to be a chronic condition requiring regular application of subcutaneous insulin, 14% of the subject in our study cohort used CM to treat their disease. Thus, to achieve an optimal patient care, a collaboration between diabetologist / primary care physician and professionals in CM needs to be established. Therefore, the issue of CM should be raised at some point when caring for diabetic patients.

P20

Experiment of nature: do girls need estrogens before puberty?

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Background: Cytrochrome P450arom (AROMATASE) catalyses the conversion of androgens to estrogens. P450arom is encoded by the CYP19 gene on chromosome 15p21.1. The enzyme is located in the endoplasmatic reticulum of steroidogenic tissues such as the ovary/testis and placenta, as well as other tissues including the brain, adipose tissue, liver, muscle, and vascular endothelium. In hereditary aromatase deficiency, lack of estrogens and subsequent androgen excess Results in virilisation of 46,XX fetuses as well as retarded fusion of metaphyses with consecutive gigantism, osteoporosis and metabolic syndrome in both sexes.
Case report: The family history of this Swiss patient is unremarkable; there is no consanguinity. The patient is a 3.5-kg product of a full-term pregnancy. No medications were taken during pregnancy. About 28 weeks into pregnancy, the mother began to suffer from severe acne, lowering of the voice and hirsutism. At birth, the infant had virilised external genitalia (Prader V). No gonads were palpable. Presenting with sonographically normal female internal genitalia and a 46, XX karyotype, the child was raised as a girl. Classic CAH due to 21-hydroxylase deficiency was ruled out by normal 17-a-OH-Progesterone on ACTH-testing. Androgen levels were elevated, estrogens very low. At the age of 3 years, large ovarian follicles were found as a result of dramatically increased plasma FSH, bone age was retarded. The diagnosis of aromatase deficiency was suspected and confirmed genetically. Oral estrogen substitution was started, resulting in a decrease of ovarian size, advance of bone age, improvement of bone mineral density and lipid profile, and normalisation of FSH. During childhood, oestrogen dosage was regularly adapted, depending on bone age, presence or absence of ovarian follicles and level of plasma FSH.
Conclusions: Loss of placental aromatase activity leads to virilisation of both pregnant mother and 46,XX fetus. During childhood, substitution with small doses of estrogen is necessary in girls with severe aromatase deficiency for normal skeletal maturation and mineralization. Estrogen supplementation prevents follicule maturation in the ovaries by inhibition of FSH secretion. Regular controls of FSH, ovarian size, bone age and bone mineral density are recommended to establish the exact estrogen dose. Increasing doses of estrogen are required during puberty for normal female sexual development.

P21

Features of resistant supraventricular tachycardia in infancy

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Aim: Most episodes of supraventricular tachycardia (SVT) in children occur in the first few months of life. Although the prognosis is generally very good, there are cases with very resistant initial course requiring multiple antiarrhythmic drug (AA) therapies to control SVT. It was the Aim of this study to analyze the features associated with resistant SVT in that specific age group.

Methods: A retrospective study was undertaken covering the period from 1997 to 2007 with evaluation of all patients suffering from SVT with initial manifestation in the first year of life. The course was defined as complicated in case more than one AA was required for oral longterm treatment or in case further SVT-attacks occurred during AA treatment.
Results: 46 neonates / infants had SVT in the first year of life, 77% had the first episode in the neonatal period. 67% were male. 13 patients or 28% already had SVT during fetal life. Complicated course with resistant SVT was observed in 15 patients (33%), all being neonates. Preexcitation syndrome on the resting ECG was even underrepresented in patients with resistant course (2 of 15 or 13%) compared to 26% in the overall population. Patients with already intrauterine SVT significantly more frequently had complicated courses (8 of 15 or 53%) compared to 23% in patients with first manifestation only after birth (p <0.01). Outcome did not differ between the two groups of patients with complicated or uncomplicated course of SVT, with comparable lenght of AA prophylaxis (median of 11 and 10 months respectively) and 95% of all patients being free of recurrencies at last observation. There was no mortality associated with SVT in the first year of life but occurred in one patient with hydrops due to intractable SVT during fetal life.
Conclusions: One third of patients with SVT occurring in the first year of life exhibited a complicated initial course . Factors associated with resistant SVT were a history of already intrauterine SVT and neonatal age. Preexcitation syndrome was not a risk factor. Outcome in patients with complicated course did not differ from the entire population and was very good.

P22

Hyperprolactinemia in adolescents: be aware of it!

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Background: Complaints about unspecific disturbances, such as headache, vertigo and palpitations are not uncommon in adolescents and are often ascribed to psychosocial stress. The pediatrician is challenged to detect those patients with an underlying disease. We report a case where amenorrhea led to the diagnosis of a pituitary prolactinoma.
Case: A 14 year old girl presented with recurrent episodes of vertigo, shivering, heart palpitation, sensation of heat and anxiety. She had been seen at the emergency unit for similar symptoms as well as adynamia and headache at least four times during the past 12 months. Her parents were divorced and she reported to be under considerable strain because of poor performance at school. She was overweight (BMI + 1.52 SD), blood pressure, heart and respiration rate were normal, pubertal stage was adult. Except for 2 café au lait spots on the right arm and left leg, clinical examination was unremarkable, as well as ECG and an orthostase test. Consultation of a pediatric psychiatrist was recommended. The symptoms did not resolve, and on the following visit, the patient mentioned to have amenorrhea for 3 months. Pregnancy could be excluded, and pelvic ultrasound was inconspicuous. Hormonal analysis showed normal values for estradiol, LH, FSH, thyroid parameters and growth factors, but a markedly elevated prolactin (3836 µIU/ml, n <425), which was confirmed by 2 further measurements (3720 µIU/ml, 3977 µIU/ml). MRI of the pituitary revealed an adenoma of 1.5 x 0.8 cm. The patient was referred to neurosurgery for transphenoidal resection.
Discussion: Even if prolactinomas are very rare in childhood and menstrual irregularities are common the first years after menarche, hyperprolactinemia has to be ruled out in every patient with amenorrhea. As anxiety and depression can be found in hyperprolactinemic patients, prolactin measurement should be considered, when such symptoms are described.

P23

Smoking behaviour among Swiss childhood cancer survivors

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Purpose: Cigarette smoking places childhood cancer survivors at risk for various adverse health effects. Nonetheless, representative data on smoking in survivors are scarce. We determined prevalence and predictors of current smoking among adult childhood cancer survivors in Switzerland in comparison with a representative population survey.
Methods: A detailed questionnaire was sent to all former childhood cancer patients registered in the Swiss Childhood Cancer Registry (SCCR) who had survived at least five years, were aged ≥20 years at the time of the survey and had a valid address (N = 832). Information on current smoking from this questionnaire was combined with prospectively collected clinical information from the SCCR database. Results were compared with population-based data from the Swiss Health Survey (SHS) 2002 (N = 6517).
Results: By December 1st 2007, the response rate was 58% (N = 484), but the study is ongoing. Among males 26% (95% CI 18–32%) of those aged 20–29 years and 21% (13–29%) of those aged 30-40 were current smokers. Among females 18% (12–24%) of 20–29 year olds and 14% (6–23%) of those aged 30-40 smoked. In the representative SHS, the proportion of current smokers was considerably higher: 46% and 41% in males and 34% and 32% in females aged 20–29 and 30–39 respectively. In a multivariable logistic regression, females, survivors diagnosed before age 5 and those with late effects were less likely to smoke. Socioeconomic determinants such as education, profession and parental education were strong determinants of smoking in the SHS but not in survivors.
Conclusions: The prevalence of current smoking was only half in childhood cancer survivors compared to the general population in Switzerland, but still far too high. Risk factors for smoking differ between cancer survivors and the general population. Further efforts are needed to develop prevention and smoking cessation programs in this high risk group.

P24

Spinal epidural haematoma in an 11-year-old boy with haemophilia A

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Background: Spinal epidural haematoma is a rare complication in haemophiliac patients.
Case report: We report on an 11-year-old boy with severe haemophilia A, who presented with acute onset of intense thoracolumbal back pain. A trauma was initially denied. Subsequently, the patient mentioned that one week before he had back pain after two children had jumped on his back. The pain had lasted following a single FVIII substitution of 50 units/kg. The clinical examination on admission showed meningeal irritation without neurological deficits. A factor replacement therapy was immediately started before imaging was obtained. Subsequently, magnetic resonance images (MRI) revealed an extensive spinal epidural haematoma ranging from C3 to L1 with spinal compression. A conservative therapy with factor replacement (50 units/kg every 8 hours during the first 36 hours, 50 units/kg and 25 units/kg alternating every 12 hours from day 2 to day 7, and 50 units/kg every 24 hours from day 8 on), analgetic therapy and bed rest led to a complete recovery with resolution of the epidural haematoma on the follow up MRI at day 7 after initial presentation.
Conclusions: In haemophilic patients with acute onset of intense back pain, a spinal epidural haematoma should be considered and a factor replacement therapy should be started immediately. Magnetic resonance imaging allows establishing the diagnosis. In the absence of neurological deficits, intensive factor replacement therapy and bed rest result in a complete recovery with no need for surgical decompression.

P25

Staphylococcal scalded skin syndrome: a case report

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Introduction: Staphylococcal Scalded Skin Syndrome (SSSS) is a Staphylococcal toxin-mediated disease (STMD), caused by staphylococcal exotoxines. Children in the first year of life are most commonly affected. We report the case of a 6-week-old infant who presented with this syndrome.
Case report: Our patient was born after an uncomplicated full term pregnancy; there were no peri- or postnatal complications. At the age of five weeks he presented conjunctivitis which was treated with anti-septic eye-drops for two days. At the age of six weeks he presented nasal congestion that was attributed to a viral infection and treated with nasal decongestants. The mother noted a foul odour around the left ear. Two days later he presented with an erythemous macular skin rash beginning behind the left ear, which spread rapidly to the face and became generalised. Mucous membranes were not affected and there was no fever. Within 24 hours, the macules evolved into flaccid bullae and skin desquamation was observed. Nikolsky sign was positive. The child remained systemically well, although in obvious pain. A full blood count and the C-reactive protein were within the

normal range, blood culture was sterile. Swabs of the ears and nose grew Exfoliatine B producing Staphylococcus aureus. Skin biopsies showed intraepidermal, sub-corneal clivage, with no inflammatory cells. The patient received analgesics and intravenous Flucloxacillin and Clindamycin. No new blisters occurred after 48 hours. He was discharged after 5 days. Intra-nasal mupirocin was prescribed for staphylococcal eradication in household contacts. Conclusions: Our case shows the potential of exfoliatine producing Staphylococcus aureus to cause a severe blistering disease in young children. The differential diagnosis includes toxic epidermal necrolysis and epidermolysis bullosa. Timely diagnosis, usually requiring a skin biopsy, is important for optimal management.

P26

Cardiovascular findings in turner syndrome: it is not only the aortic arch

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Background: Turner Syndrome (TS) occurs with a prevalence of 1 per 2500 live-born females. The cardiovascular system is affected in 17–44% of the patients. Most common findings are bicuspid aortic valve, valvular aortic stenosis, aortic coarctation and aortic aneurysm. A rare but certainly underestimated abnormality is partial anomalous pulmonary veinous connection (PAPVC), consisting of one ore more pulmonary veins draining into the right atrium, leading to a volume overload of the right ventricle and if untreated to pulmonary hypertension during the second or third decade of life. We report a case series of 2 patients with TS and PAPVC.
Case report: *Patient A:* An asymptomatic 9 years-old girl, was diagnosed of TS because of failure to growth. Screening echocardiography detected dilated right-sided cardiac structures and vena anonyma, leading to suspicion for PAPVC. PAPVC of the left upper pulmonary vein was then confirmed at cardiovascular magnetic resonance (CMR). *Patient B:* A 26 years-old young lady with TS underwent surgical repair for aortic coarctation in early childhood. She presented now with increasing exercise-related dyspnea. Echocardiography showed dilated right-sided cardiac structures and CMR confirmed PAPVC of the right upper pulmonary vein. PAPVC was surgically repaired and the patient is doing well.
Discussion: The high prevalence of cardiovascular abnormalities in patients with TS warrants a screening echocardiography by a pediatric cardiologist. Besides to the known malformations of the left ventricular outflow tract and the aortic arch, particular attention should be payed to the pulmonary veins. If echocardiography is not conclusive, due to a limited echo window, further evaluation with CMR is recommended. Early detection of PAPVC is important, as this lesion has a good prognosis, if corrected before pulmonary hypertension develops. Finally ECG registration is mandatory in the cardiovascular evaluation of these patients, as TS is often associated to a prolonged QT-interval.

P27

Temporomandibular joint arthritis in patients with juvenile idiopathic arthritis: efficacy of intraarticular corticosteroid injection as measured by mri and clinical examination

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Background: Temporomandibular joint (TMJ) arthritis in children with juvenile idiopathic arthritis (JIA) can lead to disturbed growth of the mandible and may result in an asymmetric lower jaw or in general retrognathia. Because TMJ arthritis is often asymptomatic the efficacy of intraarticular steroid injections (IAS) is difficult to assess clinically.
Objective: To assess the efficacy of IAS into the TMJ by magnetic resonance imaging (MRI) and clinical examination.
Patients and methods: JIA-patients with active TMJ arthritis on MRI were included into this study. An injection of 5 mg triamcinolone hexacetonide and 0.3 ml lidocaine 1% into affected TMJs was performed. Clinical examination at baseline and after IAS included pain, tenderess and maximal mouth opening. After a minimum interval of 3 weeks after IAS control MRI was performed. The amount of joint fluid and extent of joint enhancement following intravenous Gadolinium administration were used to assess the level of inflammation on MRI. A cohort of patients without TMJ involvement on MRI served as control group for the clinical symptoms.
Results: The study group consisted of 21 patients (15 female/ 6 male), the control group of 17 patients (7 female/ 10 male). The baseline mean maximal mouth opening was significantly different with 41 mm (range 34–54) in patients with affected TMJs as compared to 46mm (range 41–57) in patients without TMJ involvement ($p = 0.005$). After a median time of 42 days (range 7–164) the mean maximal mouth opening increased by 1.8 mm in the study group ($p < 0.003$) as compared to 0.5 mm in the control group ($p = 0.15$). However, the difference between study group and controls did not reach statistical significance ($p = 0.16$). Pain on chewing or yawning had completely resolved in all 5 patients (7 TMJs) and tenderness had resolved in 7/11 TMJs with the respective symptoms. Follow up MRI was performed after a mean interval of 52 days (range 21–125). 23/36 affected joints showed improvement and 6/36 complete resolution of inflammation on MRI.
Conclusions: In our cohort of JIA patients with MRI proven TMJ IAS led to resolution of clinical symptoms and significantly improved mouth opening in the majority of patients. However, MRI examination showed improvement but not complete resolution of inflammation in most of the affected joints. Longer follow up is warranted to assess the significance of persistent MRI changes for the mandibular growth in our patients.

P28

Salt sensitivity of low birth weight children

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Aims: Compromised intrauterine fetal growth leading to low birth weight (<2500 g) is associated with adulthood renal and cardiovascular disease. The Aim of this study was to assess the effect of salt on blood pressure (salt sensitivity) in children with low birth weight.
Methods: Caucasian children (n = 35, 43% female, mean age 11.4 ± 2.1 years) born with low birth weight (mean 1852 g, range 690–2499 g) were prospectively investigated. Of these, 17, 8, and 10 children were born at term small for gestational age, preterm small for gestational age and preterm with appropriate weight for gestational age, respectively. A group of 15 Caucasian healthy, age- and weight-matched children, born at term with appropriate weight for gestational age after uneventful pregnancies (weight range 2730 to 4890 g) served as control. The glomerular filtration rate was calculated using the Schwartz formula, and renal size was measured by ultrasound. Salt sensitivity was assigned, if mean 24-hour blood pressure increased by >3 mm Hg on a high as compared to a controlled salt diet.
Results: Baseline office blood pressure was increased and glomerular filtration rate was reduced in children born with low birth weight as compared to children born term with appropriate weight for gestational age ($p < 0.05$). Twenty-four out of the 35 children investigated complied with the dietary adjustments. Salt sensitivity was present in 37% and 47% of all low birth weight and small for gestational age children, respectively. This exceeds observations of salt sensitivity in young adults from the same geographical region (n = 41, 46% female, mean age 25.9 years) presenting with a prevalence of salt sensitivity of 26%. Kidney length and volume were reduced compared to normal values adjusted for height and weight. Salt sensitivity inversely correlated to kidney length ($r^2 = 0.307$, $p = 0.005$) and volume ($r^2 = 0.171$, $p = 0.044$).
Conclusions: A reduced renal mass in growth restricted children coincides with prevalent salt sensitivity. Presumably a consequence of "fetal programming", directed prophylactic antihypertensive interventions such as salt restriction are enabled.

P29

Acute renal failure in a neonate after acyclovir overdose. Complete recovery in the long term follow-up

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Background: Acyclovir has been used widely for treatment of documented or suspected herpes simplex infection in newborns. It is generally well tolerated, although neurotoxicity and renal impairment has been described. In newborns no adverse side effects has been reported so far. We report the largest dose of acyclovir inadvertently administered to a neonate with the highest measured acyclovir blood concentration in a neonate.
Case report: A two day old, 3170 g female newborn with vesiculo-bullous skin lesions was transferred to the neonatology unit

suspecting a herpes simplex infection. An intravenous treatment with acyclovir was started immediately. On day four sleeplessness and continuous crying was noted, on day five an unusual increase in weight to 3660 g, and on day six a rise in plasma creatinine and urea were noted (211 μmol/l and 9,6 mmol/l, respectively). At this time a dosing error of acyclovir was discovered. The patient had been given 3 times 100 mg/kg instead of 3 times 10 mg/kg per day intravenously over 4 days. Acyclovir was immediately discontinued. Creatinine and urea decreased to normal values by day 9. On day 6 after birth a renal ultrasound showed marked enlargement of the kidneys with hyperechogenicity. Renal ultrasound was normal six weeks after birth. Follow-up examination showed normal renal function and ultrasound. The acyclovir blood levels were measured retrospectively in a serum sample from day 4 and in several samples from day 6 to day 8. The highest level was 277 μg/ml on day 6 followed by a rapid decrease over two days. All cultures taken from the child remained negative for herpes simplex virus and final diagnosis was incontinentia pigmenti Bloch-Sulzberger syndrome.
Conclusions: Renal failure following acyclovir therapy can occur in newborns after administration of very high doses only. Acute impairment of renal function is rapidly and fully reversible after termination of acyclovir administration. Although acute renal failure occurred in our patient, follow-up over the time of 7 years revealed no renal sequelae, underlining a full reversibility of toxic renal side effects.

P30

Agreement rates between actigraphy, diary, and questionnaire for children's sleep patterns: recommendations for clinical and research practice

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Aims: The evaluation of children's sleep-wake patterns is essential for the identification and management of sleep problems. Sleep-wake patterns of children can be assessed by different Methods. However, none of previous reports provide the clinician or sleep researcher with information about the interchangeable use of the most common used Methods in clinical practice (actigraphy, diary, and questionnaire). Do parents accurately report on their child's sleep? How well do actigraphy, dairy and questionnaire data agree? Can these Methods interchangeably be used? These questions can only be answered by the statistical approach proposed by Bland and Altman (1986, 1999). The Aims of this study were [1] to describe sleep-wake patterns in a non-clinical sample of healthy children by measures derived from questionnaire, diary, and actigraphy and [2] to report rates of agreement between Methods according to Bland and Altman (1986, 1999).
Methods: Cross-sectional study of 50 kindergarten children, age 4 to 7 years. Sleep-scheduled times (sleep start, sleep end, assumed sleep, actual sleep time, and nocturnal wake time) were assessed by different Methods. The study included data from 7 nights of actigraph recordings and sleep diary over the same time period, and from a questionnaire, asking about children's normal sleep scheduled times.
Results: Differences between actigraphy and diary were ± 28 minutes for sleep start, ± 24 minutes for sleep end, and ± 32 minutes for assumed sleep indicating satisfactory agreement between Methods, while for actual sleep time and nocturnal wake time agreement rates were not sufficient (± 72 minutes, ± 55 minutes, respectively). Agreement rates between actigraphy and questionnaire as well as between diary and questionnaire were insufficient for any investigated variable. Sex and age of children, and SES did not influence the differences between Methods.
Conclusions: Actigraphy and diary may interchangeably be used for the assessment of sleep start, sleep end, and assumed sleep, but not for nocturnal wake times. The diary is a cost-effective and valid source of information, while actigraphy may provide useful information about nocturnal wake times. It is insufficient to collect information by a questionnaire asking about children's normal sleep patterns. Therefore, we recommend that the diary should be a standard tool in the assessment of children sleep-wake patterns.

P31

Isotretinoin-induced transient ileitis mimicking crohn's disease

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Background: The use of systemic isotretinoin, a vitamin A acid derivative, has improved outcomes in patients with acne vulgaris and is recommended if first line therapy is not effective. However, significant adverse effects may occur. Only a few cases with temporal association with the use of isotretinoin and inflammatory bowel disease (IBD) in patients without a history of intestinal problems are reported. Although IBD and transient ileitis are described as a possible adverse drug reaction in the product information, little attention has been given in the literature and even in current recommendations to this association. In Switzerland 5 cases of suspected association between isotretinoin use and IBD were registered by Swissmedic in the last 10 years (1997–2007). We report a patient with probable isotretinoin-induced transient ileitis that demonstrates the potential significant morbidity associated with isotretinoin use.
Case report: A 15-year-old male adolescent presented with occult bloody diarrhea, vomiting and abdominal pain emerging over the last two days. His past medical history was unremarkable except for severe acne vulgaris. The patient had followed a two-month course of isotretinoin for acne prior to the onset of symptoms, shortly after increasing the dose from 30 to 60 mg daily. His family history was negative for IBD. A diagnosis of ileitis was suspected by abdominal ultrasound showing long-segment swelling of the terminal ileum predominantly of the submucosa with enlarged lymph nodes surrounding this area and inflammatory participation of the vermiform appendix without typical signs for acute appendicitis. After discontinuing isotretinoin the patient never had abdominal pain or blood in the stool again with subsequent normal ultrasound. Leucocyte count, C-reactive protein and ESR were normal. Infectious causes were ruled out and calprotectin in stool was normal.
Conclusions: This case of probable isotretinoin-induced transient ileitis suggests that patients presenting with acute abdomen or symptoms of IBD should be asked about current or past use of isotretinoin. It is conceivable that isotretinoin is acting as a trigger for IBD in already predisposed individuals, or unmasking symptoms in patients with preexisting but subclinical disease. Physicians and patients should be made aware of this possible association and this should be included in the consent process required before isotretinoin is prescribed.

P32

Unusual late life threatening event as first presentation of factor X deficiency (fxd)

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Introduction: We present a 2 year-old boy with acute intracerebral haemorrhage (ICH) as first manifestation of a FXD. This is the first reported case of ICH in FXD past the neonate period. The literature of this rare disorder is reviewed.
Case report: A 2 year-old boy presented with meningismus without fever. CRP and leukocytes were elevated. The cerebrospinal fluid was hemorrhagic, and antibiotics were started. A computed tomography due to rapid clinical deterioration showed intracranial bleeding. Due to abnormal clotting tests (Quick 0%, aPTT>180 sec.) an operation was not performed. Until a severe FXD was proven (FX activity <1%) the patient was managed with fresh frozen plasma (FFP) and later with Prothromplex concentrate (PCC), before he was switched to a weekly prophylactic provision of factor IX Behring. He was discharged without apparent neurological deficit.
Discussion: XD is an autosomal recessive disorder (incidence of homozygosity 1:1 million). Factor X is the first protease in the common pathway of thrombin formation. The gene for FX on chromosome 13 is adjacent to the FVII gene. Most carriers are asymptomatic. In our case the parents of the patient are consanguine (FX- activity of both reduced to 50%). The mother and her sisters have a positive bleeding history. 42 of the 102 FXD patients in the largest published FXD cohort (Greifswald Registry) are symptomatic. In childhood, a wide spectrum of bleeding symptoms ranging from easy bruising up to severe intraarticular hemorrhages are the first signs of FXD. ICH has only been reported in 7 patients, all of which presented their disorder in the perinatal period. Severe residual neurological impairment remained in 6. Treatment options consist of FFP, PCC or pasteurized FIX Behring, containing high amounts of FX. Only 7 patients of the Registry are on regular prophylaxis. In our patient a prophylaxis with FIX Behring once a week was started. He suffered no major bleeding and no treatment induced complication within the last 3 years. This observation supports previous reports that FIX Behring allows a low risk and successful treatment and prophylaxis in FXD patients.
Conclusions: This is the first report of a non-neonatal ICH in FXD. Even in previously asymptomatic individuals, the late occurrence of life threatening bleeding events may occur. In these patients a regular prophylaxis should be discussed. Prophylaxis with FIX Behring has so far been successful, with no bleeding or adverse events.

P33

Effect of physical activity on overnight hypoglycaemia in a population of diabetic adolescents: a study based on continuous glucose monitoring.

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Background: Physical activity (PA) has a positive influence on the cardiovascular system and the well-being in patients with type 1 diabetes mellitus (T1DM). On the other hand it may cause post-exercise hypoglycaemias, which is an important issue for both health care practitioners and diabetic patients. Continuous glucose monitoring (CGMS) has been increasingly used to understand the interaction between PA and glycaemia.
Aim: To examine the effect of physical exercise on overnight glycaemia in adolescents using a CGMS.
Methods: Ten subjects with T1DM (8 males; age 13 to 19 years) underwent two blocks of CGMS recording blood glucose levels between 30 and 72 hours under standardized conditions concerning diet and insulin regimen. In an interval of 4 months, a PA block followed a sedentary block (SB). The PA block consisted of an exercise of 30 minutes at 60% of the determined Vmax on a cycle ergometer at mid-afternoon.
Results: Baseline glucose levels were similar in both blocks (SB: 8.9 mmol/L ± 1.65; PA: 9.4 mmol/L ± 2.40). 55% of the subjects presented with more hypoglycaemic events during PA compared to SB (mean 34.2 vs. 46.4). Overnight hypoglycaemia was observed after exercise in 2/10 subjects (20%). Risk for nocturnal hypoglycaemia was significantly increased ($p < 0.006$) in patients suffering from hypoglycaemic events during PA. In contrast, PA and hypoglycaemic events did not correlate with HbA1c and any other data (BMI, age, sex, duration of diabetes).
Conclusions: 1) A diabetic patient regularly suffering from hypoglycaemia is more prone to experience hypoglycaemic events following PA. 2) These data implicate that patients with T1DM have to be even more careful about their blood sugar, insulin doses as well as caloric intake following PA.

P34

Macrocepahly after cavopulmonary anastomosis

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Introduction: Patients with complex congenital heart malformations, in whom a biventricular repair is impossible, currently undergo the Fontan procedure with one ventricle maintaining the systemic arterial circulation and passive systemic venous blood return directly into the lungs. The first step is the creation of a bidirectional cavopulmonary anastomosis (BDCPA) performed during the first year of life. The prerequisites for the passive blood flow through the lungs are a normal pulmonary vascular pressure and resistance. In the following patient the pathologic elevated central venous pressure was leading to an excessive growth of the head.
Case report: The female patient underwent a one and a half chamber repair with a BDCPA for a severe Ebstein's anomaly with hypoplastic right ventricle at the age of six months. Head circumference increased from birth 36 cm (P75) to 43 cm (P50–75) at the time of the operation up to 46.6 cm (P90–97) at 9 months and 48.5 cm (>P97) at 12 months, reaching 49.8 cm (>P97, +3SD) at the age of 16 months. Haedmodynamic evaluation at that time identified an elevated pressure in the superior caval vein and pulmonary arteries of 20 mm Hg mean (normal <15 mm Hg) due to an unrestricted forward flow through the pulmonary valve as well as a stenosis of the left upper pulmonary vein. Ultrasound of the head showed a slight hydrocephalus. After reducing the venous pressure by interventional disconnection of the right pulmonary artery from the pulmonary bifurcation by implantation of an Amplatzer duct occluder and surgical relief of the pulmonary vein stenosis, the head growth slowly normalized. During postoperative follow up the head circumference measured 50.5 cm (P97) at the age of 24 months.
Conclusions: The reversible macrocephaly is generated by transient elevation of the venous pressure in the cerebral venous sinuses. This clinical phenomenon may be explained by a communicating hydrocephalus because of decreased reabsorption of cerebrospinal fluid (CSF) as a result of decreased driving pressure between the external CSF space and the draining skull veins. Repeated measurements of the head circumference in patients after complex cardiac surgery with BDCPA is easy to perform and may identify patients with unfavorable haemodynamics.

P35

Cinca syndrome – a diverse clinical spectrum

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Background: CINCA (chronic infantile neurologic cutaneous articular) syndrome is an autoin-flammatory disease which begins in the neonatal period or early infancy. Typical symptoms include urticarial rash, episodes of fever, chronic meningitis, painful arthropathy, inflammation of eyes, and hearing loss. Some children show characteristic facial features consisting of frontal bossing and saddle nose, and blond hair and finger clubbing. Diagnosis is confirmed by identifying mutations in the CIAS 1 gene.
Method: We report 3 patients (14 y. old male, 15 y. and 20 y. old females) each with distinct disease onset, time of diagnosis, evolution, outcome and CIAS 1 gene mutation.
Results: ⅗ patients showed the typical physiognomy and chronic urticarial rash beginning in early infancy. ⅔ had febrile episodes. ⅔ developped painful arthropathy with overgrowth and disorganisation of the epiphyseal plates. 1 girl had arthralgias without persistent arthritis. Eye involvement with chronic severe keratitis (boy), short episodes of conjunctivitis and acute uveitis (20 y.o. girl) or papilloedema (15 y.o. girl) was characteristic. ⅔ had growth (<3P) and developmental retardation. Two patients (20 y.o. girl, boy) had chronic meningitis, sensorineural and conductive hearing loss, the 15 y.o. girl had high frequency hearing loss. ⅔ suffered from episodes of severe headaches, the 15 y.o. girl with papilloedema from headaches and vomiting. The laboratory findings revealed anemia, thrombocytosis, leucocytosis, high ESR and CRP in all patients. We found 3 different mutations in the CIAS 1 gene . On subcutaneous Anakinra therapy (IL-1 receptor antagonist) the urticarial rash quickly disappeared in all, the arthropathy improved in ⅔, the hearing loss in ⅓, the headache in ⅗, eye involvement in all. The definite genetic diagnosis was made at the age of 9 y. in the boy and 5 y. in the 20 y.o. girl. The 15 y.o. girl showed only mild symptoms (except papilloedema) explaining why the diagnosis was not made until 14 y. of age after having had pneumonia with persistently high laboratory inflammatory markers.
Conclusions: CINCA syndrome is important to consider in the differential diagnosis of urticarial rash, arthropathy, and periodic fever starting in infancy. Early initiation of therapy (Anakinra) improves the outcome of arthropathy significantly and may prevent mental retardation, cerebral and chronic eye involvement. The drug is well tolerated.

P36

An adolescent with a vasculitis, it is not always Henoch-Schönlein purpura

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Systemic vasculitides diagnosed in children are mainly Henoch-Schönlein purpura (HSP), Kawasaki Disease and post-infectious vasculitis. The more severe forms of vasculitides are very rare during childhood and adolescence, with an estimated prevalence of about 1 for 100'000 children. We present the case of a 13 years old boy who presented with pain and swelling in the left ankle and with purpura on the adjacent skin. The laboratory showed proteinuria and microhematuria. The patient was diagnosed as HSP and received symptomatic treatment. Few days later, he developed bloody diarrhoea and abdominal pain. The abdominal ultrasound was normal, and he was treated with prednisone for a few days. Three weeks later, he presented diffused arthralgia, a painful and swollen calf, persistent abdominal pain, one episode of hematemesis, and persistent hematuria and proteinuria. A Doppler ultrasound did not show thrombosis. He was then sent to the hospital for further investigations. The systematic anamnesis revealed asthenia and weight loss since a few months. He was also complaining about aphtous stomatitis, bloody nasal discharge, conjunctival hyperaemia, rash on the face and the elbows, headache and cervical pain. The laboratory showed systemic inflammation (ESR 47), normal blood count, normal liver, kidney and muscles values, elevated complement (C3 = 1.66 g/l), normal antistreptolysin-O, negative antinuclear antibodies. C-ANCA were positive with elevated anti-proteinase 3 (147 kU/l). The diagnosis of Wegener's Granulomatosis was suspected on the clinical and laboratory findings. It was confirmed by a CT scan showing maxillary and ethmoidal mucosal thickening and multiple small opacities on both lungs, and by the histopathology of kidney, intestinal and nasal mucosa. Our patient was treated with high dose steroids to induce the remission and azathioprine to allow steroid weaning. The evolution was favourable with complete inactivity of the disease and disappearance of the c-ANCA. Vasculitis

is a less common disease in childhood and adolescence. The most usual presentation is Henoch-Schönlein purpura which is often self-limited and has a good prognostic depending on the renal condition. This case shows us than even during this period of life we do not have to forget other vasculitis like Wegener's granulomatosis.

P37

Haemolytic-uraemic syndrome (HUS) causing temporary renal failure, transient exocrine but persistent endocrine pancreatic insufficiency

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Background: HUS, the triad of microangiopathic hemolytic anaemia, thrombocytopenia and renal insufficiency, is the leading cause of renal failure in previously healthy children.
Case report: A previously healthy 2 year old girl presented with a 3 day history of bloody diarrhea, vomiting, loss of appetite, and progressive weakness. She was afebrile. 2 weeks ago, while on holiday in France, she had had an episode of diarrhea for 4 days. There were no epidemiological risk factors for Shiga toxin producing E. coli (STEC). At admission haemolytic anaemia, thromboytopenia, and impaired renal function were present. Stool examination demonstrated STEC. Sonography showed heightened echogenicity of the kidneys and mural edema of the colon. Erythrocyte and platelet transfusions were required. Renal function deteriorated with development of anuria and edema. The patient was transferred on day 3 to a university hospital for peritoneal dialysis (PD).Peritonitis occurred and was treated with intravenous antibiotic therapy. Arterial hypertension developed. Insulin-dependent diabetes mellitus (IDDM) and exocrine pancreatic dysfunction with voluminous stools and weight loss developed. After 17 days of dialysis, renal output and function were restored to normal. After cessation of PD, hypertension and IDDM persisted. Treatment at discharge after 42 days of hospitalisation consisted of an angiotensin-converting enzyme inhibitor, subcutaneous insulin, and pancreatic enzyme substitution. Four months after diagnosis the antihypertensive treatment could be stopped whereas treatment with insulin was still necessary. Exocrine pancreatic function normalized with catch-up weight gain and regular stool pattern without enzyme susbstitution.
Conclusions: In addition to causing renal failure in children, HUS can also rarely cause endocrine and exocrine pancreatic deficiency. Monitoring in hospital should include not only renal function, fluids/electrolytes and blood pressure, but also pancreatic endocrine (glucose) and exocrine (weight gain, stool pattern) function.

P38

Severe encephalopathy in an infant due to subclinical maternal pernicious anemia

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Introduction: Developmental regression in infants can have various causes including severe neurometabolic disorders. Vitamin B_{12} deficiency is a treatable cause and typically occurs in breastfed infants of vegetarian mothers.
Case report: A 7 month old, exclusively breastfed boy presented with developmental regression, generalized muscular hypotonia, irritability, and failure to thrive since the age of four months. Brain MRI showed delayed myelination, EEG revealed generalized slow activity. Laboratory evaluation presented a megalobastic anemia (Hb 90 g/l, MCV 94 fl, MCH 33 pg) and serum vitamin B12 was markedly decreased (60 pg/ml). Methylmalonic aciduria was found and total homocysteine in plasma was elevated. Therefore, the diagnosis of vitamin B_{12} deficiency was made. Cobalamin substitution was initiated and within few days the child became more awake, his development markedly improved, and EEG showed a normal Background activity. Exact history of the mother did not disclose special dietary habits and vegetarian diet was explicitly negated. Laboratory evaluation of the mother did not show anemia, but her vitamin B_{12} level was also decreased (117 pg/ml). Schilling test disclosed a lack of intrinsic factor in the mother and confirmed the diagnosis of a subclinical pernicious anemia.
Conclusions: Acquired Vitamin B_{12} deficiency can cause irreversible neurologic symptoms in infancy including encephalopathy, seizures, and developmental regression. Vitamin B_{12} deficiency is a treatable disorder and has to be searched. Mostly, the mothers are vegetarian, but pernicious anemia can cause the same symptoms. Therefore, mother's normal dietary habit does not rule out this diagnosis.

P39

Prenatal mass in the heart and neonatal nodules in the brain as typical findings in tuberous sclerosis

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Case report: A large cardiac mass surrounding the great arteries and a second small mass in the anterior lateral papillary muscle were detected at echocardiography in a fetus at 30 weeks of gestation. Neonatal echocardiogram demonstrated additional multiple tumors. Cardiac MRI showed the full extent of the tumor with infiltration of the interventricular septum and of the left and right anterior ventricular wall. Another small tumor was detected in the anterior lateral papillary muscle. Brain MRI was performed suspecting tuberous sclerosis (TS). This showed multiple subependymal nodules. At the age of 5 months the patient is seizure free, development age-appropriate. The rhabdomyomas did not cause hemodynamic complications or rhythm disturbances and remain stable in size.
Discussion: TS is an autosomal dominant multisystem disorder mainly affecting skin, brain, and heart. Clinical signs suspicious for TS appear at distinct age. Rhabdomyomas are multiple, present prenatally, and are of hemodynamic significance only in few patients. Other neonatal findings are cutaneous white spots and subependymal nodules and/or cortical tubers. However, angiofibroma, epilepsy, and mental retardation appear only later.
Conclusions: Multiple cardiac tumors detected prenatally are most suspicious for TS. The diagnosis can be confirmed if brain MRI shows subependymal nodules, cortical tubers, or cerebral white matter radial migration lines. Neuroradiological findings correlate with neurologic and cognitive outcome, whereas cardiac tumors are mainly harmless and regress spontaneously.

P40

Drug management of chronic systemic hypertension in childhood: a systematic review of the literature

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Aims: Appropriate drug management of chronic systemic hypertension is crucial. Unfortunately, there have been few drug trials for antihypertensive treatment in childhood and recommendations have been extrapolated from data obtained in adult patients. The purpose of this study was to systematically review the studies published between 1995 and 2007 that deal with the effect of antihypertensive drugs on childhood hypertension or proteinuria.
Methods: Medline, Current Contents, personal files and reference lists were used as data sources.
Results: 52 out of 79 initially found reports were excluded. Consequently 27 articles were retained for the final analysis. The blood pressure reduction was similar with converting enzyme inhibitors (10.7/8.1 mm Hg), angiotensin II receptor antagonists (10.5/6.9 mm Hg) and calcium channel blockers (9.3/7.2 mm Hg). In addition converting enzyme inhibitors (by 49 percent) and angiotensin II receptor antagonists (by 59 percent) significantly reduced pathological proteinuria, which was not influenced by calcium channel blockers.
Conclusions: In children, like in adults, the blood pressure reduction of converting enzyme inhibitors, angiotensin II receptor antagonists and calcium channel blockers is identical (and likely identical to that obtained with more traditional agents like diuretics and Beta-blockers). In children with pathological proteinuria angiotensin converting enzyme inhibitors or angiotensin II antagonists are superior to calcium channel blockers.
Simonetti GD, Rizzi M, Donadini R, Bianchetti MG. Effects of antihypertensive drugs on blood pressure and proteinuria in childhood. J Hypertens 2007;25:2370-6.

P41

Consider the diagnosis of acute hemorrhagic edema of young children in a non-toxic appearing child who has acute onset of two unusual skin lesions: cockade and edema

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Background: Acute hemorrhagic edema is a cutaneous, small vessel leukocytoclastic vasculitis of young children. The disease has been rarely reported (no more than 100 cases in a review published in 2004). Believing that the disease is not as rare as assumed but is

likely overlooked and underreported we report our experience and the Results of an extensive review of the literature.
Methods: We report six new cases. A thorough computer-based search of the literature in English, French, German, Italian, Portuguese or Spanish language revealed 287 published cases.
Results: The 293 children (boys: 67 percent) ranged in age between 2 and 60, median 11 months. In 193 children the disease developed after a simple acute respiratory illness, an acute diarrheal disease, or a urinary tract infection. At presentation the children were nontoxic in appearance. The dramatically presenting skin lesions included a) large, round, red to purpuric plaques predominantly over the cheeks, ears, and extremities and b) mostly tender edema of the distal extremities, ears and face. An involvement of body systems other than skin occurred in 25 children. The presumptive diagnosis of Waterhouse-Friderichsen sepsis, severe infection of the skin or non-accidental skin bruising was initially made in some children. The children spontaneously recovered without sequelae within 2 to 60, median 10 days.
Conclusions: Acute hemorrhagic edema of young children is a rather rare but very benign vasculitis whose diagnosis is mostly straightforward. Pediatricians often lack the educational Background to appreciate the condition since reputed textbooks do not mention the condition or mention it only in passing.

P42

Overweight and eating disorders in primary care: a sentinella reporting in 2–20 years old patients

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Aims: To describe overweight or eating disorders in primary care consultations of Swiss children or adolescents and analyze responses by physicians.
Methods: 150 to 200 primary care physicians participating in the Swiss Sentinel Surveillance Network in collaboration with the Swiss Federal Office of Public Health register their consultations over one year for selected health conditions. We describe reports of consultations where overweight or eating disorders were identified in subjects aged 2-20 years by physicians, patients or their relatives, or referring professionals, between 29.12.2007 and 15.2.2008.
Results: 189 consultations were registered in the first 7 weeks of declaration. A short majority concerned female (58%) and 12–20 years old (53%) patients. Half were reported by pediatricians, one third by general practitioners and the remaining minority by internists. The sample included two thirds of Swiss-German and one third of Swiss-French cases.
In the male subgroup aged 2–20 and in female children aged 2–11, almost all reported consultations were characterized by overweight. Among female teenagers, underweight was reported in 29% whilst overweight was recorded in 60%. Anorexia was noted in 68% of reported consultations of underweight female teenagers.
In underweight patients, advice given by physicians frequently covered both nutrition and physical activity (38%) or nutrition only (29%), while no specific recommendations were recorded for the remaining third. In case of overweight, for one half of consultations patients received both nutritional and physical activity recommendations, for 12% nutritional only, and for one quarter patients were not advised in these domains. No specific treatment was usually proposed to overweight patients (65%), except when bulimia was diagnosed; in such case, one third of patients were proposed a psychological/psychiatric treatment, whereas both psychological and pharmacological treatments were frequently offered for underweight teenagers. Therapy was most often motivated by physicians (50%) or by relatives (44%), more rarely by patients themselves (7%).
Conclusions: These preliminary data indicate that in some primary care consultations of young patients with overweight or eating disorders, advice was not given on nutrition and physical activity. This observation needs to be later confirmed with the totality of the consultations registered in 2008 and reasons will be further investigated.

P43

Vaginal voiding: a common cause of daytime urinary leakage in the school-age girl

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Background: Vaginal voiding secondary to urethro-vaginal reflux is an under-recognized cause of daytime urinary leakage in girls. The history of these girls is very distinctive: they are dry when going to the toilet but frequently wet their panties in moderate amounts immediately after the voiding. The purpose of the present retrospective analysis was to estimate the frequency of vaginal voiding as the cause of daytime urinary leakage in school-age girls, and to study the effect of simple instructions intended to amend the problem.
Methods: Girls with vaginal voiding were identified in a group of 39 girls referred to one of us because of daytime urinary leakage. They were evaluated by a noninvasive screening protocol. Girls with vaginal voiding were instructed on how to achieve better toilet habits.
Results: Vaginal voiding was found in 12 pre-pubertal girls with daytime urinary leakage aged between 8.5 and 14, median 11 years. They all had history of small leakage immediately after voiding. A body mass index greater than the corresponding 85th centile was noted in 5 and labial fusion in 2 further girls. The remaining 5 girls were noted to adopt a hairpin posture while sitting on the toilet. Instructions Aimed at improving toilet habits amended the disturbance in the 12 girls.
Conclusions: Vaginal voiding is a common cause of daytime urinary leakage in school-age girls. It occurs a) in heavy-set girls, b) in girls with labial fusion, and c) in lender girls who adopt a hairpin posture while sitting on the toilet. The condition is easily diagnosed by an adequate history and amended by instructions Aimed at improving toilet habits. With no need for specialized urologic investigations, the outpatient pediatrician can properly handle the condition.

P44

Unclear swelling of the popliteal fossa due to a giant pseudoaneurysm associated with osteochondroma

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Background: Osteochondromas are the most frequent benign bone tumours in up to 2% of the population. They develop in adolescence and may rarely present with vascular complications. Before ossification the tumour is a cartilaginous exostosis which becomes an osteochondroma. We report a vascular complication associated with osteochondroma in a 13 year-old boy.
Case report: A 13 year-old boy developed mild swelling and pain in the left popliteal fossa after a bicycle tour without obvious trauma. The pain and swelling progressed over the next 2 weeks until the patient could no longer perform his daily activities. On physical examination an indurated mass with a 10 x 7 cm diameter was found in the left popliteal fossa. Neither sensory nor motor deficits were found and peripheral pulses were present. In his history the boy suffered from multiple tibial and fibular osteochondromas. Radiography revealed an osteochondroma arising from the lower femur just above the left knee. Arthroscopy showed normal anatomy of the menisci. An MRI scan demonstrated a popliteal artery mass of 5.4 x 5 x 10 cm, and an arteriography showed a contrast medium jet into a pseudo-aneurysm arising from the anterior surface of the popliteal artery with distal patency. During surgery, the popliteal artery was exposed through a medial, longitudinal incision and the pseudoaneurysm was resected. The defect in the popliteal artery was closed with a xenopericard patch. The postoperative course was uneventful and the patient recovered completely.
Conclusions: Unclear swelling of the popliteal fossa may rarely be caused by a vascular complication of an osteochondroma leading to a giant pseudo-aneurysm. The popliteal artery is fixed in Hunter's canal and in its trifurcation, therefore distal portions have little mobility. Local compression of the artery by an osteochondroma can stretch the vessel and lead to rupture of the artery by continuous rubbing.
MRI and/or angiography are needed to plan surgical repair.

P45

Post-natal diarrhoea due to intestinal hypoperfusion as first sign of interrupted aortic arch in a newborn

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Background: Main causes of death due to undiagnosed congenital heart disease (CHD) in newborns are leftsided outflow tract abnormality like hypoplastic left heart, interruption or coarctation of the aortic arch. Patients usually present with signs of cardiovascular collapse after spontaneous closure of the ductus arteriosus during the first days of life. Intestinal hypoperfusion may result in necrotizing enterocolitis with hematochezia or diarrhoea as warning signs.

Case report: 18hours after initial uneventful adaptation a male term newborn (weight 3580 g) presented with nausea, loose abnormal smelling stools and mottled skin. Mild tachypnoea occurred, postductal oxygen saturation was 87%. At admission in the neonatology unit blood gas value was normal besides elevated lactate (5.9 mmol/L). Sepsis was suspected due to clinical presentation. Blood pressure was above normal limits for the left arm and failed for the right arm and legs, misattributed to unsettling. 4hours later irritability increased, poor peripheral perfusion, nausea and again abnormal smelling stools occured. Blood gas demonstrated further lactate elevation (6.5 mmol/L). Thorough physical reevaluation revealed grade 3 pansystolic murmur at left sternal edge and absent femoral pulses. Blood pressure was unchanged for the left but significantly lower for the right arm, measurement failed for both legs. Transthoracic echocardiography revealed discontinuity between ascending and descending aorta, a ductus-dependent perfusion of the descending aorta and a large ventricular septal defect (VSD) with bidirectional shunting. After fluid resuscitation and reestablishment of ductal patency with intravenous prostaglandin the newborn was hemodynamically stable and transferred to the University Children's Hospital. Cardiac MRI confirmed Type A interrupted aortic arch with an arteria lusoria arising from the descending aorta. Surgical reconstruction of the aortic arch and VSD patch closure was done without intra/postoperative complications.
Conclusions: Postnatal diarrhoea may be an alarming sign for deficient bowel perfusion. Besides arterial hypotension due to sepsis an important differential diagnosis of intestinal hypoperfusion is ductus dependent leftsided outflow tract abnormality such as interrupted aortic arch. Measurement of four-limb blood pressure in critical ill newborns is mandatory. Postductal pulse-oxymetric screening within the first 12 hours of life may be effective for early detection of CHD.

P46

"You don't deserve a new heart!" psychosociocultural contraindications for paediatric heart transplantation

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Introduction: Heart transplantation is a therapeutic option for children with end stage heart disease due to myocardial dysfunction or congenital heart disease. Frequently the progression of the illness is rapid and difficult to predict. The decision pro transplantation arises often in an advanced state of the disease, mostly in an ICU setting and under time pressure. The following cases illustrate the challenge in appropriate transplantation decisions especially in children with difficult and different psychosociocultural Background.
Patient 1: Two years old foreign girl with severely symptomatic restrictive cardiomyopathy. Her father was well integrated in Switzerland; her mother did not speak German yet. The girl's clinical condition initially improved with anticongestive medication. But feeding problems occurred: she refused to eat, did not accept to swallow medication, but vomited instead. Her situation deteriorated and recurrent haemodynamically significant arrhythmias were noted. We decided against transplantation because of the extremely high risk of non-compliance of the patient with regards to the postoperative immunosuppressive therapy. Finally, she died from progressive low cardiac output in the context of persistent atrial flutter.
Patient 2: 12 years old male patient with dilated cardiomyopathy requiring inotropic support. The patient and his family were foreigners, isolated for their language, culture and religion. His condition worsened and he was listed for cardiac transplantation after having obtained parental consent. A left ventricular assist device was implanted as a bridge to transplantation. Four months later, the parents suddenly decided against transplantation and the device was finally removed. The boy is now, two years later, on stable outpatient medication and goes to school for two days a week.
Conclusions: These two patients are representatives of a small group of paediatric patients with end stage heart disease in whom a heart transplantation is contraindicated because of psychosociocultural reasons. An ethical dilemma is created for the caring team: a known beneficial therapy is withheld from the child. It is our task and responsibility to develop strategies for an early recognition of psychosociocultural contraindications for heart transplantation with the aim for an optimal and holistic patient management including palliative care and child protection.

P47

Hypertonic saline inhibits in vitro growth of pseudomonas aeruginosa (pa) isolates from patients with cystic fibrosis (cf)

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Aims: Inhalation of 7% hypertonic saline (~1 M NaCl) is one of the therapeutic modalities Aimed at improving mucus clearance from the CF lung. While inhalation of hypertonic saline BID or QID over prolonged periods of time has been shown to reduce the frequency of pulmonary exacerbations, its direct effect on growth of Pa has not been investigated. We studied the in vitro growth characteristics of Pa exposed to various salt concentrations.
Methods: Clinical Pa isolates cultured from sputum samples of patients with CF were incubated in Luria-Bertani broth supplemented with various concentrations of sodium chloride. Growth curves and time-kill assays, respectively, were performed with or without continuous or intermittent exposure to 0.15–1 M saline.
Results: Growth of Pa was consistently inhibited by 1 M saline, but not by concentrations equal or less than 0.5 M. Two of 10 isolates investigated showed weak growth after >6 hours of incubation, the remaining 8 isolates were still fully inhibited after 24 hours. A mucoid phenotype did not affect salt susceptibility. In time-kill analyses, 1 M saline exerted a bacteriostatic, but not bactericidal effect. Short-term exposure to 1 M saline for 15 to 120 minutes induced a "posthypertonic" inhibitory effect lasting approximately 120 minutes following removal of salt. This effect occurred irrespective of the duration of preceding saline exposure. Repetitive short term exposure to 1 M saline (1 hour) followed by incubation in medium without saline (3 hours) – simulating in vivo exposure of Pa in CF patients inhaling hypertonic saline – resulted in persistent inhibition of bacterial growth. Repetitive subculturing of Pa in 1 M saline did not result in adaptive resistance after 5 passages.
Conclusions: Hypertonic saline used for inhalation in patients with CF inhibits growth of Pa in vitro. If also occurring in vivo, this effect may contribute to the observed prevention of acute pulmonary exacerbations in clinical trials using 7% saline. Alternatively, bacterial stress responses induced by exposure to hypertonic saline may affect bacterial virulence factor expression and antimicrobial susceptibility.

P48

Dynamics of colonization with stenotrophomonas maltophilia (SM) in patients with cystic fibrosis (CF)

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Background: The pathogenic role, need for treatment, and implications for nosocomial spread of respiratory tract colonization with Sm in patients with CF are controversial.
Methods: We conducted a retrospective study in CF patients <16 years of age, who were colonized at least once with Sm. Individual episodes of colonization were defined as being separated by at least 12 months and 3 Sm negative cultures.
Results: Fifty-two patients were included. The mean age at first recovery of Sm and the duration of follow-up were 6.2 ± 4.3 and 5.0 ± 4.5 years, respectively. In a Kaplan-Meier analysis, the proportion of Sm-free patients decreased linearly between birth and 16 years of age. Seventeen patients (33%) were colonized only once. These patients had a similar age (5.7 vs. 6.1 years, p = 0.83) compared to patients with repetitive Sm positive cultures (3.9 vs. 6.7 years, p = 0.08). Once-only colonization was associated with less frequent use of antibiotics during the 6 months preceding the first recovery of Sm (5/17 vs. 24/35 patients; risk ratio 0.46; 95% CI 0.25–0.83). The frequency of resistance to cotrimoxazole (TMP-SMX) (0 vs. 4 patients, NS) and the use of TMP-SMX following the first recovery of Sm (7 vs. 14 patients, NS) were similar. Nineteen patients (37%) had one episode of Sm colonization with >1 consecutive positive cultures (mean duration, 0.9 years), 16 patients had multiple episodes (31%). TMP-SMX resistance occurred in 4 patients (8%).
Conclusions: Acquisition of Sm occurred at a constant rate over the first 16 years of life. One third of patients were Sm positive only once. The use of TMP-SMX did not appear to influence the frequency of Sm recovery despite that fact that most isolates were sensitive to this agent in vitro.

P49

Premature loss of primary teeth, a cardinal clinical feature of childhood hypophosphatasia

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Introduction: Exfoliation of primary teeth in the absence of trauma in early childhood is very rare. However, the clinician should know the most common conditions of this symptom such as early-onset periodontitis or hypophosphatasia.
Methods: The case of a 4 9/12 y old girl with premature loss of primary teeth is reported. Results At the age of 1 y the patient got her first deciduous teeth; already 9 months later she lost one of them, followed by continuous spontaneous loss of a total of 5 incisors. Because of pain during mastication, an orthodontist was consulted who did not detect conspicuous findings. A pediatric dentist suspected hypophosphatasia which was proven by low plasma alkaline phosphatase (54 U/l; age-matched controls: 96–297), high plasma pyridoxal-5-phosphate (194 nmol/l; controls 35–100), and elevated urinary phosphoethanolamine (59 mmol/mol creatinine; age-matched controls <20). Hand and knee radiographs showed no signs of rickets. Molecular analysis of the TNSALP gene disclosed that the girl carried the mutation c.211C>T (R54C) on the maternal allele as did her 6.5 y old healthy brother; the 2nd mutation on the paternal allele was not found (courtesy of Dr. E. Mornet, Paris). This constellation is compatible with a dominant mild form with variable clinical expression or with compound heterozygosity comprising one undetected mutation. Clinical follow-up of both children showed no additional symptoms until age 5 6/12 and 6 10/12 y, respectively.
Conclusions: Hypophosphatasia is an uncommon disorder with a highly variable clinical expression; in contrast to the severe forms, milder variants can easily be overlooked. Premature loss of teeth should raise the suspicion of early onset periodontitis, syndromal disorders such as Jeune-Tommasi-Freycon-Nivelon syndrome [MIM 208750], lacrimoauriculodental digital syndrome [MIM 149730] and Papillon-Lefèvre syndrome [MIM 245000], and child abuse. In the clinical context the most likely diagnosis is hypophosphatasia, a connective tissue disorder which can easily be diagnosed by simple laboratory tests. As in our case dental involvement generally is limited to the primary frontal teeth, but rarely may result in secondary teeth loss and involvement of alveolar bone. A positive effect of medical treatment (Vitamin D, zinc, magnesium, or enzyme substitution) has not been proven effective. However, early diagnosis is important to start timely orthodontic measures and to provide genetic counseling.

P50

Association of sports club participation with fitness and fatness in children

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Decreased fitness and increased fatness are relevant factors for decreased cardiovascular and bone health in children. One way to increase physical activity, and hence fitness, and to reduce the risk for overweight might be regular sports club participation.
Aim: The purpose of this study was to investigate the association of sports club participation with fatness and fitness in children in general, and in those with increased risk for overweight and/or low fitness. Methods: A cross-sectional study was conducted in a random sample of 502 1st and 5th grade primary school children. Fitness components were determined by ten motor tests and body fatness by the sum of four skinfolds and waist circumference. All measures were normalized for gender and grade. Regular sports club participation was defined as participation of at least once a week.
Results: Two thirds of all children were participating in a sports club. Girls' and boys' participation rate, as well as those of overweight children and of children with overweight parents were comparable to their respective normal weight peers. In contrast, children from migrant families (OR 0.31; 95% confidence interval, 0.20–0.48) and those from inactive parents (OR 0.16; 0.05–0.45) participated significantly less (all P <0.001). Sports club participation was generally associated with aerobic fitness (0.55 > beta > 0.41, all P <0.01) and partly with speed, strength, and coordination (0.37 > beta >0.21, all P <0.05). In overweight children and in children from overweight parents and migrant families, this association was not found. There was no association between sports club participation and measures of fatness in any of the groups.
Conclusions: Regular sports club participation rates were high, and were in general associated with higher levels of most fitness components in school-age children. Participation rates were lower for children of migrant families and children from inactive parents. In addition, the association between sports club participation and fitness-components was not found in overweight children, and in children from overweight parents and migrant families.
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P51

Choroid plexus papilloma: a rare cause of nonobstructive hydrocephalus

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Introduction: Hydrocephalus Results from an imbalance between production and resorption of cerebrospinal fluid (CSF). It is known as obstructive (anatomic or functional obstruction to CSF flow), or nonobstructive (impaired resorption or increased production of CSF). We report a case of a child with choroid plexus papilloma, a rare cause of nonobstructive hydrocephalus.
Case report: A 3 months old girl was admitted with a three days history of irritability, lethargy, poor appetite and vomiting. She was born at term by vaginal delivery after an uneventful pregnancy with growth parameters (birth weight, length and head circumference) in average values. Physical examination on admission revealed a wide open and bulging anterior fontanel, open skull sutures, dilated scalp veins, setting-sun eye sign and axial hypotonia with head lag. The head circumference was 7.5 SD above average, with body weight and length being +0.5 SD only. Ultrasound and MR imaging studies revealed an important quadriventricular hydrocephalus without obstruction to CSF flow and a voluminous vascularized left lateral ventricle mass arising from the choroid plexus, compatible with a papilloma. Complete surgical resection of the mass was achieved, without residual tumor at the post-operative MRI. Histological studies confirmed the choroid plexus papilloma. Despite removal of the papilloma, persistent hydrocephalus was treated with a VP shunt.
Discussion: Choroid plexus tumors are rare tumors of neuro-ectodermal origin, accounting for less than 1% of intracranial tumors and separated into benign papillomas (80%) and malignant carcinomas (20%). They are mostly diagnosed before age 2 and usually grow in lateral ventricles. They usually present with signs of increased intracranial pressure due to excessive production of cerebrospinal fluid, as shown in our case. As most cases occur in infants, a congenital origin is questionable. The prognosis for the benign tumor is excellent after surgical resection. The use of radiotherapy and chemotherapy is restricted to malignant tumors.
Conclusions: This case shows a rare cause of hydrocephalus due to hypersecretion of CSF by a choroid plexus papilloma and emphasizes the need to recognize the signs and symptoms of increased intracranial pressure in infants, as well as the importance of careful follow-up of head circumference growth.

P52

"Rota" seizures: not only in asia

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Background: Most convulsions during infections in otherwise healthy children are simple febrile seizures. Prognosis is good and routine performance of extensive laboratory investigations is not warranted. Children with rotavirus gastroenteritis can present with clinically worrisome afebrile cluster convulsions. We present two such cases.
Cases: Two afebrile boys (17 and 19 months old) were admitted to our hospital because of convulsions. Both had 4 recurrent seizures within 5–10 hours, each lasting less than 15 minutes. One patient had 2 generalized tonic-clonic and 2 generalized tonic convulsions, while the other had 2 lateralized convulsions with right upper limb tonic movements and loss of consciousness followed by two generalized toncio-.clonic seizures. Both patients experienced gastrointestinal symptoms prior to the convulsions: one had vomiting and diarrhea 8 hours before the first convulsion; the other had developed acute mild gastroenteritis 2 days before admission. Both patients had rotavirus found in stools. None of them was dehydrated neither had electrolyte or other laboratory abnormalities. They were given iv phenytoin after the 3rd convulsion. Both patients left hospital within 3 days without anti-epileptic treatment. Follow-up has been uneventful.
Discussion: Rotavirus associated convulsions in children are known, although mostly described in Asia. They are usually described as short-lived, tonic-clonic or partial seizures, occurring in cluster within 24 hours, at any time during the rotavirus gastroenteritis, sometimes even before its onset. Digestive symptoms may be mild, preventing clinicians to link up the convulsions to an afebrile benign

gastroenteritis. The relationship between the virus and the neurological symptoms is not clear. Prognosis is generally good. Rotavirus infections are common; «rota» seizures apparently not. Clinicians should be aware of "rota" seizures to refrain from wrong epilepsy diagnosis, overinvestigation and useless long term treatment.

P53

Cerebral sino-veinous thrombosis (SVT) and ent infection: report on 2 cases

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Introduction: Ear, nose and throat (ENT) infections are common in pediatrics, usually treated by oral antibiotics. Such infections are considered mostly benign, and complications rarely occur. We present two cases of SVT in relation to ENT infections.
Cases presentation: Case 1: this 7-year-old male presented a mild orbital tumefaction and good general condition. He was treated by oral Amoxi-Clav (50 mg/kg/day tid) for periorbital cellulitis. Despite an initial favorable evolution, he had relapsed fever five days later and developed diplopia. Head CT revealed pansinusitis, a frontal epidural collection and thrombosis of the right superior ophthalmic vein extending into superior sagital vein. He was treated with IV amikacine and ceftriaxone for 12 days followed with another 2 weeks of oral amoxi-clav, and anticoagulation with low molecular weight heparin (LMWH).
Case 2: this 19-month-old female initially exhibited a bilateral purulent otorrhea and upper eyelid tumefaction, treated with oral Amoxicillin (60 mg/kg/day tid) and Ibuprofen. Within 24 hours, she developed vomiting and an altered mental status. Head MRI showed a pansinusitis. Lumbar puncture revealed mild inflammation. Although initial outcome was favorable under i.v ceftriaxone, fever relapsed after the fourth day. A repeat head MRI revealed persistent pansinusitis and a left transverse sinus thrombosis extending into the left jugular vein (Lemierre disease). She required a left mastoidectomy and right paracentesis. Cultures of pus revealed a fusobacterium. Treatment was pursued with IV ceftriaxone and clindamycin for a total of 6 weeks along with LMWH. In both patients, an MRI was repeated within 6 weeks of therapy revealing partial resolution of the SVT. Both are being treated with LMWH for 3 months. Thrombophilia work-up is being pursued.
Discussion: SVT are rarely observed. Canadian ischemic stroke registry estimated its incidence at 0.7 per 100'000 children per year, and found ENT infections as one of the commonest cause. The recent occurrence of two cases in our local hospital is likely to be coincidental. Epidemiological studies are desirable to evaluate if current treatment attitude may be associated with an increased rate of severe complications.

P54

Cerebral folate deficiency in a child with alpers-syndrom: a case report

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Introduction: Cerebral folate deficiency has, amongst other neurometabolic disorders also been described in Alpers disease. The detection of folate antibodies in some of the affected patients has given rise to the idea of an underlying autoimmune process. We report on a girl with Alpers-Syndrom and cerebral folate deficiency who possibly derived clinical benefit from folinic acid supplementation.
Patient and methods: Pregnancy and birth were uneventful. The early development of the child was characterized by psychomotor retardation, especially in motor skills and speech. At the age of 3 10/12 years the girl presented for the first time at our hospital with status epilepticus. A cerebral infection was excluded, amino-acids in plasma and organic acids in urine were normal. Pyruvate was not increased, lactate and ammonia were slightly elevated. A congenital disorder of glycosylation could be excluded. Cranial CT revealed a "cerebral swelling". In the EEG typical changes as previously described for Alpers-Syndrom were seen. Because of repeated mainly myoclonic convulsions the girl received anticonvulsant therapy. Lafora disease was excluded by skin biopsy. Following repeated episodes of status epilepticus the suspected diagnosis of Alpers-Syndrom could be proven by molecular analysis (compound heterozygote mutation in nuclear-encoded DNA polymerase). Due to a deficiency of folate (22,9 nmol/l, n: 63–111 nmol/l) in cerebrospinal fluid and folate antibodies in serum a supplementation therapy with folinic acid was initiated. After normalization of folate in CSF the anticonvulsant therapy could be considerably reduced.
Results and discussions: The analysis of folate metabolites in cerebrospinal fluid is part of the basic investigations when searching for neurotransmitter disorders. We found low folates in a girl diagnosed with Alpers-Syndrom. Substitution with folinic acid allowed reduction of anticonvulsant therapy leading to considerable clinical improvement. Deficiency of folate in cerebrospinal fluid in Alpers-Syndrom and consecutive benefit from supplementation with folinic acid has not yet been reported. The reason for the reduced cerebral folate concentration is still a matter of investigation.

P55

Mycoplasma pneumoniae associated opsoclonus-myoclonus-ataxia syndrome – a case series

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Introduction: Opsoclonus-myoclonus-ataxia syndrome is a rare acquired movement disorder occurring in all age groups, predominantly in infants. Although the exact pathogenesis is still undefined, there is strong evidence for a paraneoplastic or parainfectious immune process resulting in central nervous system dysfunction. Mycoplasma pneumoniae has been implicated in a number of immune-mediated neurologic diseases. However, the association of Mycoplasma pneumoniae and opsoclonus-myoclonus-ataxia syndrome is not well established so far.
Methods: Case series report and review of literature.
Findings: We present three cases with opsoclonus-myoclonus-ataxia syndrome in adolescents following an infection with Mycoplasma pneumoniae. After ruling out a neoplastic etiology of the symptoms, all three patients were treated with oral steroids. They showed a full recovery without any cognitive impairment and, within a year, no relapses were reported.
Discussion: OMAS seems to be a previously undescribed central nervous system complication of M. pneumoniae infection. Compared to paraneoplastic forms of OMAS in infants, M. pneumoniae associated cases of OMAS in adolescents show a rather benign course without neurodevelopmental long term sequelae. OMAS should be added to the spectrum of M. pneumoniae associated neurologic complications; nevertheless neoplasia has to be ruled out in all cases of OMAS.

P56

Extensive aplasia cutis congenita (acc) of the scalp – successful conservative management

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Aim: ACC is characterized by focal absence of the epidermis, subcutaneous tissue, galea and rarely cranial bone. Patients with large scalp or skull defects are at risk for lethal hemorrhage or meningitis, especially if the dura is exposed. It is not clear if patients with large defects require surgical treatment. We present a patient in whom conservative therapy produced excellent Results.
Case report: A full-term newborn was transferred to us on the first day of life for evaluation of a large cranial skin defect noted at birth. Family history was unremarkable. The 38-y-old mother was a non-smoker, with no history of substance abuse or infections during pregnancy. Caesarean section was performed because of a pathologic cardiotocogram. Physical examination revealed a full thickness defect of the dermis and bone in the midline area of the posterior fontanelle extending to both parietal regions of the cranial vertex with exposure of the dura (6x4cm). There was no cerebrospinal fluid leakage. No other malformation was detected. Intravenous antibiotics were initiated for 5 days. In addition, antiseptic dressings were applied until the defect of the scalp epithelialised and healed at 6 weeks. The bone defect closed by 6 months. A large scar remained over the vertex of the head; treatment with osmotic scalp expander is planned.
Conclusions: ACC is a rare disorder. Most reported cases are sporadic but an autosomal dominant pattern of inheritance also occurs. Infection with varicella or herpes simplex virus, or consumption of drugs such as alcohol, cocain and heroin, during pregnancy have been proposed as etiologies.
Conservative treatment with moist, antimicrobial-impregnated dressings is the therapy of choice for small defects. Optimal treatment of extensive defects overlying the sagittal sinus is controversial, Most authors recommend urgent surgical intervention

to prevent fatal hemorrhage, or infection, and to prevent cerebral injury. Our experience demonstrates that first line conservative, non-operative management should be considered even in cases with large defects. This may prevent infants from untimely operative procedures with elevated operative risk.

P57

Qt-interval prolongation caused by healthfood products?

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Aim: To describe a patient with QT-interval prolongation on electrocardiogram (ECG) as well as other symptoms/signs possibly attributable to intake of "life style and wellness" dietary supplements (DS).
Methods and results: A 15 yr old female competitive sports swimmer presented with a history of having had a vasovagal syncope one month earlier. During the past few weeks she had suffered from unexplained sweating and was unable to perform her usual physical exercise. Four months earlier she had had a normal exercise ECG. Past medical history was unremarkable. Family history revealed no incidents of sudden death or recurrent syncope. Although she did not take medication on a regular basis, over the preceding 5 months she had supplemented her diet with 3 DS products containing guarana and green tea extracts. Physical examination was unremarkable. Her ECG showed sinus rhythm with a heart rate of 50 beats per minute. The maximum corrected QT-interval (QTc) was 500 ms and there were notched T-waves in several leads. A 24h-ECG revealed a QTc of 430–580 ms and episodes of sinus tachycardia up to155 beats per minute for several hours after taking the DS. An echocardiogram was normal. Hemoglobin, serum electrolytes, creatine-kinase, C-reactive protein and renal, liver, and thyroid function tests were normal. Serologic testing for Epstein Barr virus showed no evidence of recent infection. After discontinuation of DS, the ECG abnormalities completely resolved and symptoms disappeared. Five months later the patient remained free of symptoms and resumed competitive swimming.
Discussion: Resolution of symptoms and normalization of the ECG after discontinuation of DS intake suggest a causative role of the DS. Green tea extracts and guarana contain caffeine, which usually does not prolong the QTc when one adheres to the recommended daily allowance. At high doses it acts as an ergolytic. Whether other components of the compound contributed to the ECG abnormalities remains speculation. There may be a genetic susceptibility to drug induced changes of repolarisation patterns in some individuals. Life style DS with "adrenalizing" components are becoming increasingly popular among adolescents and are easily purchased over the counter. At least one fatality has been reported in association with guarana. As DS are not considered to be medication, the heart of the matter may be missed when taking the standard medical history.

P58

Ulcus vulvae acutum – painful genital ulcers are not always due to herpes simplex virus (HSV)

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Background: Genital ulcers are uncommon in children. The broad differential diagnosis includes mainly infectious causes and raises the question of sexual abuse.
Case report: A previously healthy 12 y old girl presented with a history of sudden onset of fever, dysuria and pain in the genital region. Clinical examination revealed three ulcers in the labia majora and minora measuring up to 2 cm in diameter. Physical examination was otherwise unremarkeble. HSV infection was suspected and aciclovir was initiated. Sexual contact was denied. Laboratory tests for sexually transmitted diseases were negative. Despite antiviral therapy the painful ulcers persisted and an abscess developed. Amoxicillin/clavulanic acid was added. Sterile pus discharged spontaneously from the abscess. A biopsy was taken, but no specific histopathology was found. Cultures, PCR (blood and swabs from the ulcers) and serology remained negative for bacterial, viral and fungal infectious agents. The patient developed cough, left-sided pulmonary crepitations, and findings consistent with atypical pneumonia on chest x-ray. Complement-fixation test for Mycoplasma pneumoniae rose from 1:10 to 1:640 within 12 days. A diagnosis of Ulcus vulvae acutum Lipschütz associated with M. pneumoniae was made. Oral clarithromycin was started and the vulvar ulcerations rapidly disappeared.
Conclusions: Our Results strongly suggest that M. pneumoniae was the cause of both atypical pneumonia and the ulcers. Ulcus vulvae acutum is a rare clinical entity characterized by acute painful genital ulcers. It was first described in 1913 by Lipschütz in an adolescent girl; it was attributed to an non-venereal infectious agent. The multiple ulcers are usually painful and accompanied by local lymphadenopathy and fever. Healing can be spontaneous but may take several weeks and leave scarring. Since the initial report several aetiologies have been proposed, among others HSV, syphilis, chlamydia, EBV and Beçets disease. Althoug the disease is rare, it is important to include in the differential diagnosis of genital ulcers. To our knowledge there is only one other report in the literature of genital ucers attributable to M. pneumoniae infection.

P59

Early blood exchange transfusion in malignant pertussis

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Introduction: Bordetella pertussis infection is a potentially serious illness especially in very young children and can be lethal in 1% of those younger than 2 months old. B. pertussis is the leading cause of death due to bacterial infection in children aged less than 2 months, and the 3rd cause in children up to 18 years of age. Malignant pertussis is a rapidly evolving combination of pneumonia, respiratory failure, severe leucocytosis, neurologic involvement and finally, severe pulmonary hypertension leading to death in 75% of cases.
We report a case of malignant pertussis in an 8-week-old baby with a favourable outcome due to aggressive management and early blood exchange transfusion.
Case report: D0: 8-week-old girl born at 38 weeks, hospitalized for bronchiolitis with oxygen requirements after an 8-day history of rhinorrhea and cough. Chest X-ray (CXR) showed peribronchial thickening.
D2:Paroxystic cough with desaturations and fatigue. Blood exams showed hyperleucocytosis (91 G/L), elevated CRP (83 mg/L) and hyponatremia (Na 123 mmol/L). CXR showed a left sided infiltrate for which empirical treatment with IV cefuroxime and oral clarithromycine was started.
D3: PCR positive for *B. Pertussis.* Cefuroxime stopped and clarythromycine pursued.
D4-6: Progressive respiratory failure requiring intubation and high frequency oscillatory (HFO) ventilation. CXR worsened with bilateral upper infiltrates and atelectasis.
D7:Peak WBC count at 119,4 G/L and mild pulmonary hypertension confirmed by echography. A one-volume blood exchange transfusion was performed with resultant decrease in WBC count to 34 G/L. Continuous IV heparin treatment was started for thromboembolic prophylaxis.
D9: Withdrawal of heparin after 48h. due to thrombocytopenia.
D12:Seizures due to hypoxic spells successfully treated with midazolam. Brain MRI normal.
D17: Persistence of a mild pulmonary hypertension responding well to inhaled NO for 24 h.
D22: Extubation.
D30: Discharge from the Intensive Care Unit.
D36: Pulmonary CT-scan showing multiples residual condensations.
D37: Discharge from hospital.
Conclusions: According to the literature, risk factors for severe fatal B. pertussis infection are leucocytosis, seizures or encephalopathy, lack of vaccination and pneumonia. So far, HFO ventilation, vasodilatators such as NO or sildenafil and ECMO have not shown effectiveness in preventing malignant pertussis related deaths.
The recent use of double-volume blood exchange transfusion (Romano M.J., Pediatr 2004) and leucopheresis (Marek J., Pediatr Crit Care Med 2006) has proved successful in 2 infants with malignant pertussis by preventing leukocyte microthrombi formation in the pulmonary veins and subsequent pulmonary hypertension.
The early use of exchange transfusion also seems appropriate for the management of malignant pertussis and should be initiated before severe pulmonary hypertension develops.

P60

Unexplained severe periventricular leukomalacia in an otherwise healthy premature infant: role of maternal morganella morganii chorioamnionitis?

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Introduction: Several risk factors have been linked to the development of periventricular leukomalacia, including chorioamnionitis. Morganella morganii has been reported as a rare pathogen for chorioamnionitis. We present a case where both neonate's leukomalacia and maternal Morganella morganii chorioamnionitis occur, and question a possible causality.

Case report: A 36 years-old healthy woman presented at 24 weeks pregnancy with premature contractions and was given oral betamethasone. At 28 weeks, she showed another episode of spontaneous onset of labour with low-grade fever and moderately elevated CRP (28 mg/L), clinically consistent with chorioamnionitis. Membranes were intact. Four hours after admission, a girl was delivered vaginally with good Apgar scores. She showed a moderate metabolic acidosis (pH 7.14) and hypoglycaemia (0.9 mmol/l), which both resolved within the two first hours of life. Apart from a transient leucopenia (4.8 G/L) on day one, the clinical evolution was uneventful. The infant did not present any clinical or biological signs of infection and was not treated with antibiotics. Nevertheless, ultrasound studies on day one showed diffuse periventricular hyperechogenicity and cerebral MRI on day five showed a massive bilateral periventricular leukomalacia. Meanwhile, placental histology showed severe chorioamnionitis and cultures were positive for Morganella morganii.
Discussion: Morganella morganii is a gram-negative enterobacteria, which has most commonly been described as a nosocomial pathogen in urinary tract infections, postoperative wounds and immunocompromised adults. It is a rare cause of early-onset systemic neonatal infection through maternal chorioamnionitis. Late-onset neonatal infections, rarely associated to meningitis or brain abscesses, have also been reported. The clinical presentation of the mother and child in this case is compatible with a subclinical infection causing preterm delivery and in utero brain injury, with lesions objected at birth. Indeed, the association between chorioamnionitis, preterm delivery and periventricular leukomalacia is well established, but Morganella morganii has not yet been associated to this constellation.
Conclusions: Morganella morganii is a rare cause of neonatal infection. The case reported showed a poor neurological outcome without any risk factor other than Morganella morganii maternal chorioamnionitis. This causality is debatable, but would represent the first case described.

P61

Influenza-associated encephalopathy

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Background: Pediatricians are familiar with influenza virus infection. As a complication, febrile seizures are seen in 3% of young children affected. Influenza-associated encephalopathy, in contrast, is very rare in Europe and not well appreciated as a clinical entity. It has been repeatedly reported in Japanese children, exposing them to high morbidity and mortality. We report on an unusual case of a child, who presented with febrile seizures due to a "trivial" viral infection, but who finally died of acute influenza-associated encephalopathy.
Case: A 3 year old girl was well until the day before admission, when she developed a high fever. The following day she experienced a prolonged febrile, generalized, tonic-clonic convulsion and was admitted to hospital. The convulsion lasted for 60 minutes, until it could be disrupted with lorazepam and phenytoin. Lumbar puncture revealed a normal cell count. Empirical therapy with acyclovir and ceftriaxone was started. Despite cessation of convulsions, she failed to regain consciousness. A MRI of the brain was performed thereafter, which did not show pathology. Influenza A virus was detected by culture from nasopharyngeal swab, but not in the cerebral-spinal fluid (CSF). An intermittent non-convulsive status epilepticus persisted for the next 3 days despite maximal anticonvulsive therapy under continuous EEG-monitoring. High intracranial pressure was treated unsuccessfully. The girl showed clinical signs of progressive brain herniation with massive brain edema on CT and loss of cortical signals from somato-sensory evoked potentials. "Comfort care" was initiated and the girl died on day 8. Autopsy showed massive brain edema, herniation and histologically hypoxic-ischemic lesions, unfortunately PCR on brain tissue was not examined.
Conclusions: Encephalopathy due to influenza virus is associated with sudden onset of high fever, severe generalized seizures, rapidly progressive coma, and death within some days. Little is known about the incidence of this fatal disease in Europe. In 90% of the reported cases from Japan, the cell count and the protein concentration of the CSF were within normal limits. Only in few cases, the influenza virus could be detected in the CSF by PCR or culture. Therapy remains symptomatic. Although many issues regarding pathogenesis remain to be clarified, influenza virus infection plays a role in triggering the development of encephalopathy.

P62

Assessement of the interobserver-variability of the apgar-score using video sequences

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Purpose: The Apgar-Score is used to assess the immediate neonatal adaptation to extra-uterine life in newborn infants for over 50 years. Moreover, it allows to document the effectiveness of neonatal resuscitation. Despite that an increasing number of extremely preterm and sick newborn infants are being resuscitated due to advances in neonatal medicine, the Apgar-Score has not been adjusted. For example, there are no consistent data on the use and significance in very preterm infants. Likewise, no accepted standard for reporting a score in newborn infants undergoing resuscitation after birth exists, where many score parameters are altered by resuscitation measures. Moreover the scoring definitions in textbooks and guidelines vary substantially. The purposes of our study were to assess the interobserver variability of the Apgar-Score and to gather more information on how the infant's maturity, the breathing pattern or respiratory support and the experience of the observer may affect the score.
Methods: 31 neonatologists, 36 obstetricians and 37 midwives looked over 20 video sequences showing delivery room resuscitations of 20 newborn infants (gestational age 29–31 wks) in order to assign the Apgar-Score. No audio sound was performed. The heart rate was shown optically by finger tapping. Each sequence took 20 seconds. In between the sequences little breaks were interposed in order to write the scores down.
Results: Overall, this study revealed a high interobserver variability. In 15 out of 20 sequences the median value of the total Apgar-Score was the same for neonatologists and midwives. The scores given by obstetricians were tendentially lower. That may be due to the less frequent assignment of Apgar-Scores by obstetricians in general, and more particularly in preterm infants. For all observers, in the majority of sequences, heart rate showed the lowest and skin colour the highest standard deviation.
Conclusions: Given the high interobserver variability, the significance of the Apgar-Score may rightly be questioned. This is of importance as the Apgar-Score is widely used in research when it comes to compare newborn infants. Therefore, there is an urgent need for perinatal health care professionals to be consistent in assigning an Apgar-Score, both on timely assessment at the set time points and with regard to the definition of each parameter. Therefore, better guidelines need to take into account prematurity and resuscitation measures.

P63

Educational level and employment of swiss childhood cancer survivors

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Purpose: We assessed the educational level and employment of adult childhood cancer survivors in Switzerland.
Methods: A detailed questionnaire was sent to all former childhood cancer patients registered in the Swiss Childhood Cancer Registry (SCCR) who had survived at least five years, were aged ≥20 years at the time of the survey and had a valid address (N = 832). Information on schooling, education and work situation from this questionnaire was combined with prospectively collected clinical information on tumour type and therapy from the SCCR database. Results were compared with population-based data from the Swiss Health Survey (SHS) 2002 (N = 6517).
Results: By December 1st 2007, 484 questionnaires (58%) have been returned, however the study is still ongoing. Former diseases included leukaemias (188), lymphomas (98), CNS tumours (51), embryonal tumours (68), bone tumours or soft tissue sarcomas (52) and other tumours (27). 152 survivors (31%) had received remedial teaching during their school years and 133 (27%) had had to repeat a year. Among males aged ≥30 years (N = 104), 41% (95% CI 32–51%) had a university degree, compared to 30% in the SHS. Among females (N = 70) this proportion was 27% (17–38%), compared to 14% in the SHS. 84% (76–91%) of males aged ≥30 years had a full-time job and 6% (2–12%) were part-time employees. Corresponding figures for females were 36% (24–47%) and 43% (31–55%) respectively. Respective numbers in the SHS were 88% and 11% for males, and 24% and 47% for females. In total, 4% of all survivors received a disability benefit. This proportion arose to 24% in the survivors with a CNS tumour.

Conclusions: Despite transient problems with schooling, adult survivors of childhood cancer in Switzerland seem to have a normal or even better educational level and current work situation compared to the general population.

P64

Spinal cord emergencies: clinical presentation and work-up of acute myelopathies in childhood

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Introduction: Acute myelopathies are rare in childhood, but they are associated with a substantial risk of long-term neurological deficits. Based on four cases we discuss clinical presentation, differential diagnosis, management issues, and outcome of acute myelopathies in childhood.
Case reports: All four children were previously healthy and presented with non-traumatic acute paresis and sensory loss of the lower limbs. Patient 1 is a 12 years old girl presenting with a history of subfebrile temperature and sore throat for several days, followed by acute and severe back pain, progressive paresis of the legs, sensory deficits with a sensory level, and urinary retention. Investigations showed myelitis caused by EBV. She was treated with steroids and the outcome was favorable. Patient 2 is an 11 years old boy with lower back pain radiating to the legs followed by sudden paraparesis and bladder dysfunction. MRI showed spinal cavernoma with acute bleeding. Treatment with steroids was initiated and the cavernoma was surgically removed with full recovery. Patient 3 is a 10 year old boy with sudden onset of paraesthesia in the thighs and a rapidly progressive paresis of the legs. Within two hours he developed complete paraplegia and sensory dysfunction in the lower limbs. Treatment with steroids and aspirin was started. Spinal MRI showed T2-hyperintensity with contrast enhancement. The etiology of the transverse myelopathy was most probably spinal ischemia. Unfortunately he remained paraplegic and has neurogenic bladder and bowel dysfunction. Patient 4 is a 5 years old girl with paraparesis and pain in both legs occurring one day after she fell while dismounting from her bike. Spinal MRI showed localized symmetric contrast enhancement in the anterior horns in the thoracolumbar myelon. Treatment with steroids was started and CSF was normal. Symptoms improved over weeks and the etiology remained unknown.
Conclusions: In acute myelopathy spinal imaging (preferably MRI) is urgent to rule out spinal cord compression and to look for evidence of inflammation or ischemia, and represents the first and most important diagnostic procedure. Early transfer to a tertiary care centre and treatment with steroids has to be considered. In most cases, history and clinical examination allow differentiation from Guillain-Barré syndrome (no motor and sensory level) or spinal tumor (rarely presenting as acute myelopathy).

P65

Serum concentration of H-ficolin and the risk of fever and neutropenia in pediatric cancer patients

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Background: The impact of H-ficolin (Hakata-antigen, Ficolin-3), an important component of the lectin pathway of complement activation, on susceptibility to infection is largely unknown. This study Aimed to determine if H-ficolin serum concentration is associated with fever in severe neutropenia (FN) in children with cancer.
Methods: H-ficolin was measured by time resolved immunofluorometric assay (TRIFMA), in serum taken at time of diagnosis from 94 children treated with chemotherapy for cancer. Association of FN episodes with H-ficolin concentration was analyzed by exact multivariate Poisson regression accounting for chemotherapy intensity and duration.
Results: Median H-ficolin serum concentration was 26 mg/L (range, 6 to 83; IQR, 20 to 39) with 7 (7%) children having low H-ficolin (<14 mg/L). H-ficolin concentration was higher in children with fever at diagnosis than in those without (median, 37 vs. 24; p = 0.036) and was correlated with C-reactive protein (p = 0.014). During a cumulative chemotherapy time of 82 years, 177 FN episodes were recorded, 35 (20%) of them with bacteremia. Children with low H-ficolin had a significantly increased risk to develop FN (multivariate RR, 2.20; 95%CI, 1.28 to 3.59; p = 0.005), resulting in prolonged duration of hospitalization and of intravenous antimicrobial therapy. They showed a tendency towards more frequent episodes of FN with bacteremia (RR, 2.89; 95%CI, 0.93 to 8.18; p = 0.068). Conclusions: In this study, low serum concentration of H-ficolin was associated with an increased risk of FN, in particular of FN with bacteremia, in children treated with chemotherapy for cancer. Low H-ficolin thus represents a novel risk factor for chemotherapy-related infections.

P66

When staphylococcus aureus infection needs rescue therapy: successful treatment of septic shock and purpura fulminans with vasopressin and protein c.

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Background: Purpura fulminans (PF) and catecholamine refractory septic shock are associated with a high mortality. There is a growing body of data supporting the use of vasopressin and protein C (PC) as a rescue therapy but there are only few reports on this treatment in staphylococcal sepsis and PF in children.
Methods: Case report and review of literature.
Findings: We report the case of a 12 year old boy who presented with PF and septic shock secondary to community acquired, methicillin-sensitive Panton Valentine leukocidin (PVL) producing S. aureus. Despite adequate fluid and catecholamine resuscitation and adequate antimicrobial therapy, blood pressure remained low and the boy showed persistent lactic acidosis, rhabdomyolysis, disseminated intravascular coagulation (DIC), and organ failure. Therefore vasopressin with a starting doseof 0.001 U/kg/min was administered 12 hours after admission and a PC-concentrate-infusion with an initial bolus of 100 U/kg followed by a maintenance dose of 10 U/kg/h was started on day 2. Under this rescue regimen a substantial improvement could be observed within 48 hours. Despite a SAPS II score predicting a mortality of 70%, the boy survived without impairment of central nervous functions. No amputations and skin grafts were necessary. Renal failure required hemodialysis for three months. Five months after diagnosis, the boy suffers from improving critical illness polyneuropathy and functional deficits caused by extensive muscle necrosis making him partially wheel-chair dependant.
Discussion: PVL is a potent exotoxin encoded about 5% of S.aureus isolates, many of which are methicillin-resistant. It is mainly associated with severe necrotizing pneumonia; its association with PF is less well established. Vasopressin as a rescue therapy of catecholamine refractory shock in children is still under debate. The experience of non activated PC-concentrate as an anti-coagulatory, pro-fibrinolytic and anti-inflammatory therapy is only limited.
Conclusions: Our experience demonstrates that the use of vasopressin and PC-concentrate can be successfully used as a rescue therapy in septic shock and PF secondary to PVL producing S. aureus.

P67

The role of reagent strip urinalysis in infants with congenital cataracts

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Background: Congenital cataracts are often isolated (dominant inheritance) but are sometimes associated with congenital infections (including rubella, toxoplasmosis, herpes simplex virus, and cytomegalovirus) and with various systemic, genetic, or metabolic conditions (including Turner's Syndrome, Down Syndrome, galactosemia, and peroxisomal disorders). In addition, cataracts occur in the oculo-cerebro-renal syndrome of Lowe, a rare X-linked multisystemic disorder whose features include ocular congenital cataracts, developmental delay (with cognitive impairment), and proximal renal tubular dysfunction. Unfortunately Lowe Syndrome is often difficult to identify. We report on a new case of Lowe Syndrome and suggest that urinalysis may aid in early disclosure of renal impairment.
Case report: In a floppy infant the diagnosis of congenital cataracts was made at the age of 1½ month. The final diagnosis of Lowe Syndrome was suspected biochemically and subsequently confirmed using molecular biology techniques at the age of 3 months.
Conclusions: Urinalysis for proteinuria and glucosuria, using a simple and cheap reagent strip tool, may disclose proximal tubular

dysfunction. In infants with congenital cataracts, urinalysis is a valuable aid in early clinical suspicion of Lowe Syndrome (before the onset of massive renal involvement and developmental delay).

P68

Characteristics of ADHD subtypes in a group of children from canton Ticino

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Aim: Attention-deficit/hyperactivity disorder (ADHD) is a common childhood-onset disorder with a significant impact on public health. The Aim of this study was to study ADHD symptoms and DSM-IV subtypes in children and adolescents in Ticino, along with their features, and therapy and follow-up.
Methods: A total of 97 children with a diagnosis of ADHD, recognized between 2000 and 2006, were examined and their clinical characteristics, school Results and therapy were studied.
Results: We considered 97 children (89 male/8 female) aged 3 to 14 years (mean age 8.1) who were followed by our department of child neurology. We divided the children into the three following groups: pre-school (11), primary school (67) and secondary school (18). The subtype most represented in the pre-school group was Hyperactive-impulsive at 50%. In the primary school group we found the following divisions: Inattentiveness: 37%; Hyperactive-impulsive: 18%; and Combined: 45%. For the adolescents in the secondary school group, the most-represented subtype was inattentiveness at 67%. The therapies were distributed in the three groups in the following way: ergotherapy: 45% pre-school, 28% primary school and 27% secondary school; psychomotor: 54% pre-school, 34% primary school and 16% secondary school; pedagogical support: 18% pre-school, 70% primary school and 83% in secondary school. Drug therapy with methylfenidate was followed by 50% of the children and varied between 9% in pre-school children and 89% in the secondary school.
Conclusions: The distribution of the subtype of ADHD is remarkably different between the three groups: the pre-school aged children showed more Hyperactive-impulsive problems, while later problems of inattentiveness are present. Behavioural therapies are available for the children of pre-school age, while the boys of secondary school mainly benefit from therapy with methylfenidate. As the most-represented group is that of the primary school age, in the future paediatricians should be able to recognize children with ADHD already at the preschool age in order to begin therapy as soon as possible.

P69

A population-based study of epilepsy in children from canton Ticino

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Introduction: Epidemiological studies of childhood epilepsy are of importance in order to compare incidence and prevalence rates, age distribution, inheritance, seizure types, epilepsy syndromes and treatment strategies.
Aim: To analyse epilepsy characteristics, outcome and antiepileptic drug (AED) use in a children's neurological department located in the southern part of Switzerland.
Methods: All the children referred to and analysed by our department were aged from birth to 16 years with the epilepsy diagnosis recognized between January 2000 and December 2007.
Results: 183 children (99 male/84 female) were included in the study. Mean age at first attack (range) was 5,2 (0-16) years. A majority of the patients, 58%, had focal or focal plus secondarily generalized seizures. Epilepsy was classified in 45% of patients. The most common syndrome of childhood absence epilepsy occurred in 20%, and rolandic epilepsy in 14%. 34% showed different disorders associated with epilepsy: The most common associated disorders are cortical malformation (8%) and chromosomal abnormality (7%). 72% were seizure-free at the last follow-up visit after at least 6 months. Antiepileptic drug (AED) treatment was used in 89% of patients (most common: valproate 38%, carbamazepine 11%, topiramate 10 %, sultiam 8%). 88% were on monotherapy, 9% on two drugs and 3% on three or more.
Conclusions: This study provided valuable information on epilepsy characteristics, outcome and AEDs in a child population in the southern part of Switzerland. Epilepsy was classified in 45% of the patients, more than two-thirds were seizure-free and the majority were on monotherapy.

P70

Rupture of brain arteriovenous malformation presenting as acute, isolated, non-traumatic headache

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Background: In childhood, tension-type headaches and migraines account for more than 95 percent of non-traumatic headache cases. Hence, obtaining a neuroimaging study is not indicated in children with acute non-traumatic headache and a normal neurological examination. On the contrary, urgent neuroimaging is warranted if headache is associated with neurological dysfunction. We report on three children with ruptured brain arteriovenous malformations who presented with acute headache but without any associated neurological dysfunction.
Reports: In our emergency unit between 2002 and 2006, at least 60–70 children per year presented with a history of acute, non-traumatic headache. Three boys, aged 6, 7 and 9 years, presented with sudden onset of an intense headache, which was not accompanied by neck stiffness, altered mental status, or focal neurological signs. Neuroimaging studies were performed because the headache was persistent, intense and refractory to medical management. Two of the children had a contrast-enhanced CT scan and the third an MRI. Test Results demonstrated an intracranial hemorrhage temporo-parietal right in all three patients. The patients recovered without sequelae following surgery (N = 2) and surgery plus endovascular embolization (N = 1).
Conclusions: In childhood, intracranial parenchymatous hemorrhage is the most typical clinical presentation of brain arteriovenous malformations. The clinical features of children with either tension-type headache or migraine and those of children with a ruptured arteriovenous malformation may be initially identical. In children with acute non-traumatic headache, the following "red flags" have been recently recommended: aged 3 years or less, morning or nocturnal headache (or vomiting), headache increased by Valsava or straining, explosive onset, arterial hypertension, meningeal signs, papilledema, motor asymmetry, ataxia, gait disturbance, and abnormal deep tendon reflexes.

P71

Dangerous complications of retropharyngeal infections through the "danger space"

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Background: Retropharyngeal deep neck infections are typically associated with an infection of the nasopharyngeal structures. The retropharyngeal lymph structures undergo atrophy at puberty explaining the predominence of local infections in school children. We discuss a typical but dangerous complication of this infection.
Case report: A 7-year-old boy was brought to the emergency department for evaluation of a prolonged upper respiratory infection with fever, headaches and diminished neck movement. On examination he had a sore throat, difficulty in swallowing and a positive Bolte-sign. Computed tomography showed a diffuse swelling in the retropharyngeal space, cervical bilateral lymphadenitis and pansinusitis. I.v.-antibiotic treatment was started but 2 days later he developed oliguria and a polyserositis with ascites, pleural and pericardial effusions. A follow up computed tomography identified a bilateral abscess of 14 cm length extending from the level of C2 until the level of the T8 vertebral body. The abscess had to be drained by cervical incision and later on by thoracotomy.
Discussion: Complications of retropharyngeal infections constitute true surgical and medical emergencies. An abscess in the "danger" space between the alar and prevertebral fasciae may drain into the posterior mediastinum and result in a widespread necrotizing process with pericardial or pleural abscess and often sepsis. Early diagnosis and timely debridement are the mainstays of successful treatment. Mediastinal drainage may be attained by the cervico-mediastinal approach, which can be effective in early mediastinitis, but thoracotomy is generally indicated once the necrotizing process has entered the "danger" space.
Conclusions: It is important to remember, that the only route for direct infection from the neck into the mediastinum is the retropharyngeal fascia. Therefore children with a continuous fever and pain upon treatment of retropharyngeal infections have to be evaluated early for complications including acute necrotizing mediastinitis.

P72

Violence incident reporting system in geneva children's hospital

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Aim: The violence against medical and paramedical staff in paediatric wards, including emergencies, is a real problem. We wanted to know if it has increased recently.
Method: We used the incident reporting system introduced in our ward in 2001, to evaluate how often the staff had to deal with violence from patients or parents. The incident reporting system is based on a volontary and non obligatory declaration, not including serious events like lethal or severe damage to a person.
Results

Year	Violence from patient	Violence from parents	Total violence indicents (% of total incidents reported TIR)	Nb of hospitalisations	violence incidents / nb hosp. ‰	Problem with medication (% of TIR) of TIR)
2002	1	0	1 (2)	2174	0.45	15 (37)
2003	34	7	41 (41)	2377	17.25	28 (28)
2004	23	8	31 (29)	2713	11.43	22 (21)
2005	16	17	33 (24)	2885	11.44	37 (27)
2006	41	18	59 (34)	2324	25.39	27 (16)

Between 2002 and 2006 we noticed an increase of the incidents due to violence from patients or parents (p <0.0002; 2002–04 vs 2005–06). On the contrary, the usual incidents mainly reported in the litterature (related to medication)decreased during the same period. The violence incidents happened in 3 situations: agressive parents against staff in the emergency room, agressive patients in both paediatric and pedopsychiatric wards.
Conclusions: Although incident reports are based on a volontary declaration, our data demonstrate an increase of the perception of violence by hospital staff.
In Conclusions, the staff must be aware of the violence problem and have a good training about dealing with it in order to correctly manage these patients and/or agressive parents. Due to our «incident culture» we were able to reduce usually reported incidents such as medication problems. However, the evolution of the society led to an increase of incidents related to agressive behaviour.

P73

Use of pegylated interferon in children with chronic hepatitis B and C: the geneva experience

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Aims: Hepatitis B and C cause morbidity and mortality from liver disease. Vertical infection is the most important route of HBV and HCV transmission in children in CH and long-term spontaneous clearance is known to be low. Children with biochemically and histologically active disease who are at higher risk to develop cirrhosis and hepatocarcinoma were treated in our division.
Methods: Both B and C hepatitis patients had a liver biopsy. Patients with chronic hepatitis B received subcutaneous recombinant peginterferon alfa-2a (Pegasys®) at a dosage of 100 μg/m² once per week for 48 weeks. Patients with chronic hepatitis C received the same therapy in combination with oral ribavirin (15 mg/kg x day in 2 doses) for 48 weeks. Viral load, ALT levels, blood count and physical examinations were determined during the treatment and follow-up.
Results: HBV group: 9 patients, median aged 10,5 ± 6,5, 2 horizontal transmissions, 7 unknown (adopted), all were HBsAg/HBeAg positive. 1 discontinued therapy after 12 weeks because of elevated transaminase despite reduced doses, 1 is still on therapy, 2/8 had HBeAg seroconversion and 5 did not have seroconversion. HBsAg remained positive in all. 4/8 experienced leukopenia or elevated transaminase as side effects. HCV group: 4 patients, aged between 3 and 15 years, 3 vertical transmissions, 1 unknown, 2 genotype 1A, 1 genotype 1B and 1 genotype 3A. 3/4 had negative viremia after 24 weeks of treatment, 1 started therapy 10 weeks ago. In 12/13 children, transient flu-like symptoms of variable intensity were observed during the first weeks of treatment.
Conclusions: Our patients tolerated well the therapy with minor side effects. Weekly peginterferon was well accepted even in very young child. We had 2/8 HBeAg seroconversion and 3/4 sustained viral response (SVR) with hepatitis C. In the literature, 1/3 of children with hepatitis B have a sustained response to therapy and 1/10 will become both HBeAg and HBsAg negative. Between 44 and 75% will have SVR according to the genotype in hepatitis C. Histological abnormalities in asymptomatic children with both HBV and HCV show a bad correlation with liver test.
As cirrhosis and hepatocellular carcinoma are common complications in adulthood, we should consider peginterferon treatment already in younger age, even with modest responses.

P74

Which childhood cancer patients fail to access specialised paediatric oncology care?

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Objective: Specialised paediatric oncology care is essential to obtain maximal cure rates for childhood and adolescent cancer patients. The Swiss Childhood Cancer Registry (SCCR) registers all paediatric cancer patients in Switzerland. We determined the proportion and characteristics of paediatric patients, who were not registered in the SCCR and not treated in one of the 9 specialised paediatric oncology departments (SPOG centres).
Patients and methods: We matched children registered in the SCCR against records in cantonal cancer registries (ASRT) for regions where the latter exist (58% of Switzerland), using a specialised software for record linkage. All children aged <16 years at diagnosis with malignant primary site tumours diagnosed between 1990 and 2004 were included in the analysis.
Results: About 16% childhood cancer patients (169 of 1077) had never visited a specialised paediatric oncology centre. An additional 6% (69 of 1077) had been treated in a SPOG centre but not reported to the SCCR. Adolescents (14–15 years) were with a proportion of 34.7% at highest risk to be treated outside a SPOG centre (51 of 147, p <0.0001). Children suffering from malignant bone tumours and soft tissue sarcomas were also at increased risk to be treated in other clinics (p = 0.061). Of all patients not treated in SPOG centres, 28.9% (33 of 114) were treated in a general paediatric hospital and 38.6% (44 of 114) in an adult clinic (missing information for 32.5%). The proportion treated by adult specialists remained stable over time, while the contribution of general paediatric hospitals decreased.
Conclusions: A relevant proportion of childhood cancer patients in Switzerland were not treated in a specialised paediatric oncology centre.

P75

Lung function, exhaled markers of airway inflammation and atopy status in paediatric patients with clinically stabile inflammatory bowel disease

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Background: The inflammatory bowel diseases (IBD), e.g., Crohn's disease and ulcerative colitis are chronic inflammatory disorders of the intestine and/or colon associated with a variety of systemic manifestations. Different bronchopulmonary involvement has been reported both in adult and paediatric patients. Exhaled nitric oxide (FeNO) is recognised as marker of airway inflammation in patients with many chronic respiratory diseases. There are only few data about lung function and no data about FeNO in paediatric patients with IBD.
Aim: The primary Aim of the presented study was to measure lung function, FeNO and nitrite (NO2) concentration in EBC in paediatric patients with clinical stabile IBD.
Patients and methods: 20 children with clinical stabile IBD were enrolled in this prospective study. 19 healthy children and 20 children with mild persistent asthma served as a control groups. The mean age in IBD patients was 13,9 years (range 10,1–18 years), and the mean duration of disease in IBD patients was 20 months (range 7–60 months). After the clinical examination and completion of questionnaire, the patient underwent the following tests in order: skin-prick testing (SPT), measurements of FeNO, spirometry, body plethysmography, and TLCO and collection of exhaled breath condensates.
Results: The median FeNO in IBD group was 14 ppb (range 3,6–78,3 ppb) and herewith significantly higher than median FeNO in the group of healthy children (6,2 ppb, range 3,4–17,1 ppb). 6 children (32%) showed at least one pathological pulmonary function test. In 5/6 was the type of pulmonary dysfunction described as obstructive, whereas lung function was normal in all healthy controls. The concentration of NO2 EBC did not differ significantly between the group of IBD and healthy control subjects. 9/20 children with IBD showed a minimally one positive reaction in SPT compared to 3/19 healthy controls (p = 0.08). After adjustment for positive skin prick testing, there was still significantly difference in FeNO between IBD patients and healthy control subjects.
Conclusions: In our study, children with clinical stabile IBD shown elevated incidence of lung function abnormalities and compared with

healthy controls higher FeNO levels. As FeNO levels were not significantly explained by the elevated incidence of atopy in IBD patients, airway inflammation may be present in children with clinical stable IBD.

P76

Emergency service utilization by adolescents

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Aims: Adolescents are described in literature as frequent users of emergency services. In Switzerland, 40 % of 16–20 years declare not to have a primary care physician, particularly boys. The Aims of this study were to review adolescents' utilization of the paediatric emergency service (ES) of Geneva University Hospital, and to identify their characteristics.
Methods: A retrospective survey of 8499 children (0–11 years) and adolescent (12–16 years) visits to the emergency service in 2006. The following items were used in the analyses: age, sex, schedule of visits, degree of emergency and reasons for visits. Reasons for visits were categorized as follow: medical (including cardio-vascular, endocrine, gastro intestinal, genitor-urinary, haematological, infectious and neurological problems), trauma, wound, psychiatric, poisoning (substances and medication), gynaecologic, ill treatment and polytraumatized.
Results: Adolescents accounted for 14.2% of the ES visits. There were no difference in sex-ratio and schedule of visits between children and adolescent. A lower degree of emergency (adolescent: 67.5%; children: 62.9%, $p < 0.05$) was found in adolescent. Trauma (adolescent: 39.8%; children: 14.4%) and psychiatrics' problems (adolescent: 4.3%; children: 0.3%) were more prevalent in adolescents than in children. For the adolescent cohort, boys were overrepresented (boys: 55%; girls: 44.8%). No difference for schedule of visits and degree of emergency were observed between boys and girls. The main reason for visits was trauma for boys (47.7%) and medical problems for girls (42.8%). Psychiatric and poisoning were also more frequent in girls than in boys.
Conclusions: Most of the adolescents' visits to ES are not urgent, which may be explained by the fact that many adolescents do not have a primary care physician and have to consult a hospital ES when they have a medical or surgical problem. Adolescents' boys visited ED more frequently than girls. This disparity is also found in children.

P77

Acute lower abdominal pain in girls – the challenge of early recognizing, diagnosing and treating ovarian torsion

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Introduction: In girls with acute lower abdominal pain ovarian torsion must be taken into consideration. Prompt diagnosis of ovarian torsion is important to preserve ovarian function and prevent complications. However the diagnosis can be challenging because the symptoms are relatively non-specific and ultrasound can be misleading.
Case reports: Between March 2006 and November 2007 we examined six girls aged 6 3/12 to 14 8/12 years presenting with lower abdominal pain and nausea at our emergency department. All of them were referred by the physician with suspected acute appendicitis or acute constipation. Clinical reevaluation and diagnostic work-up resulted in the diagnosis of ovarian torsion. All patients had a symptom of acute onset of lower abdominal pain (100%). Four (66%) reported nausea and vomiting. The most common sonographic finding was enlarged ovary with heterogeneous appearance and numerous small peripheral follicles in the ovarian parenchyma. Intraovarian blood flow was diminished or absent in 4 (66%) cases. Torsion involved the right ovary in 3 patients (50%) same as the left (50%). Diagnostic laparoscopy and detorsion was performed in all patients. 4 patients had a solitary cyst as an underlying cause for ovarian torsion.
Conclusions: Ovarian torsion should be considered in any girl with acute onset of lower abdominal pain. Typical aspect in the ultrasound such as heterogeneously enlarged ovary and diminished or absent blood flow in the ovarian vessels are not always present. Early laparoscopy (even as a diagnostic tool) and detorsion should be performed to preserve ovarian function and avoid complications.

P78

Atopy and pfapa syndrome: a protective or a risk factor?

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Background: PFAPA syndrome is a recurrent febrile disease first described in 1987 by Marshall et al, and characterized by periodic fever, aphtous stomatitis, pharyngitis and cervical adenitis. In a cohort of patients investigated in 2006 by Tasher et al, a high frequency of atopy was reported in PFAPA patients.
Aim: To investigate the frequency of atopy in PFAPA patients, compared with the general population.
Patients and methods: In 2 of the participating centers of the European registry for PFAPA (Lausanne-Geneva, n = 45 and Bordeaux, n = 39), we questioned all patients or their parents during a phone call interview to complete the personal and family history of atopy. We used the same questionnaire for a control group taken from a general pediatric consultation.
Results: Atopy was present in 16/45 (35,6%) and 15/39 (38,5%) PFAPA patients from Lausanne-Geneva and from Bordeaux, respectively, and in 17/47 (36,2%) of the control group. The family risk of atopy was the same in PFAPA patients and in the control group. We have compared clinical characteristics of atopic and non-atopic PFAPA patients. Frequency of some symptoms were different between atopic and non-atopic patients, but the differences were not statistically significant (pharyngitis 97% versus 89%, headache 39% vs 15%, abdominal pain 68% vs 45%, neurological symptoms 10% vs 0%).
Conclusions: The frequency of atopy is not different between PFAPA patients and general population. Atopic PFAPA patients do not seem to have a separate clinical profile. These Results do not suggest a relationship between atopy and PFAPA nor that a Th2 cytokine profile is implicated in the pathogenesis of PFAPA syndrome.

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Familial aspect in pfapa syndrome

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Background: PFAPA syndrome is a recurrent febrile disease first described in 1987 by Marshall and characterized by periodic fever, aphtous stomatitis, pharyngitis and cervical adenitis. Since this first description no clear etiology has been found. In opposite to other auto-inflammatory diseases, no genetic origin was underlined and no familial tendency was reported until now. To better understand this disease, we created a European web-based multicentric registry with the participation of 8 countries and 14 rheumatologic centers.
Aim: To investigate the eventual familial tendency to present PFAPA or an other chronic inflammatory disease.
Patients and methods: In 2 of the centers participating to the registry (Lausanne-Geneva, Switzerland and Bordeaux, France), we questioned all patients or their parents during a phone call interview to complete the family history. We used the same questionnaire for a control group taken from a general pediatric consultation. In the questionnaire, we asked for positive family history of recurrent fevers, PFAPA, and rheumatologic diseases (chronic inflammatory).
Results: Eighty-four patients with PFAPA were recruited: 45 in Lausanne-Geneva and 39 in Bordeaux and were compared to 47 control children. Family history for recurrent fever was positive in 19/45 (42%, CI95: 28–56) and 18/39 (46%, CI95: 30–62) PFAPA patients from Lausanne-Geneva and Bordeaux respectively, and always negative in the control group. 6/45 (13%, CI95: 3–23) and 3/39 (8%, CI95: 0–17) PFAPA patients had a family member with PFAPA, but none in the control group. The differences between both PFAPA group and the controls are statistically significant. The family history for rheumatologic diseases (chronic inflammatory) seemed to be more frequently positive in the Swiss PFAPA group (14/45 = 31%) than the French PFAPA group (5/39 = 18%) and the controls (7/47 = 20%), but the differences are not significant.
Conclusions: These data show that history of recurrent fever and PFAPA is found more often in patients with PFAPA than in the general pediatric population. They suggest a familial susceptibility and a potential genetic origin for the PFAPA syndrome. This opens a wider spectrum for future research.

P80

Infliximab in pediatric inflammatory bowel diseases: the geneva experience

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Introduction: Infliximab (IFX, Remicade®), a monoclonal antibody against tumor necrosis factor-alpha, has been shown to be effective for the treatment of refractory Crohn's disease (CD), Ulcerative Colitis (UC) or indeterminate colitis (IC). This retrospective study evaluates the efficacy and safety of IFX in patients with inflammatory bowel disease (IBD) in our tertiary pediatric gastroenterology service.
Methods: 12 pediatric patients (age 11.6y ± 0.98 y) with IBD who received at least one dose of IFX were reviewed. The administration protocol was IFX 5 mg/kg at week 0, 2 and 6 with a maintenance dose every 8 weeks. The indication for IFX, the disease activity index at week 0, 6 and 52 as well as side effects were evaluated.
Results: 8 patients with CD and 4 patients with UC or IC were treated with IFX between 2004 and 2008. Indications for treatment with IFX were active disease which was refractory to steroids (n = 10) and/or severe perianal disease in 2. At time of first infusion 10 patients were on Prednisone (0.4–2 mg/kg×d) and 6 on Azathioprine. A total of 109 doses of IFX were administered, ranging from 2 to 21 infusions per patient. Usually no premedication was given. At 6 weeks (after 3 doses) 7/8 (87%) children with CD were in remission, mean Harvey-Bradshaw disease activity index (HBI) decreased from 7.1 to 2 ($p < 0.01$). All patients with UC were in remission, Pediatric ulcerative colitis activity index (PUCAI) decreased from 38.75 to 3.75 after 3 doses of IFX ($p < 0.01$). On the long-term follow-up 3 patients are still in their first year of therapy and 4 patients are off IFX after 1 year of treatment. 7/9 were free of steroids after one IFX-year. Two patients could not be weaned off IFX and are on a second year. During IFX therapy 2 patients lost response to IFX, one was switched to Adalimumab and the other was on Azathioprine and Prednisone. 5 significant side effects were observed (4.5% of infusions): 2 skin rashes, fever in one patient and 2 anaphylactic reactions.
Discussion: Treatment with IFX is effective in most children with IBD which is refractory to standard immunosuppressive therapy. Nevertheless at long-term 40% of our population could not be weaned off IFX or had to be discontinued due to loss of response (Antibody to IFX, ATI) or severe adverse reactions.

P81

Exhaled nitric oxide is elevated in young preschool children with persistent asthma at school age

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Background: Respiratory symptoms are common in early childhood. The clinical characterization of disease presentation and hence, its likely disease progression has so far been proven difficult. We have recently shown that wheezy young children less than 4 years of age with a stringent index for the prediction of asthma (API) at school age have elevated levels of FeNO compared to children with recurrent wheeze and a loose index for the prediction of asthma at school age or children with recurrent cough.
Objective: To follow up children from the previous study in order to assess persistent asthma at school-age and to evaluate the predictive value of FeNO measured in early preschool-age for the persistence of asthma at school age.
Methods: All children from the original study now being at the age of 6-10 years were followed up by a phone call and interviewed using a modified ISAAC questionnaire. They were asked to take part in an additional visit in order to perform skin prick testing (SPT), lung function and FeNO measurements. FeNO was assessed by a single breath on-line method according to ATS/ERS recommendations. SPT was performed for five common allergens including birch and hazel, 6-grass mix, animal dander of cat and dog, and dust mite. Spirometry was measured according to current recommendations.
Results: From the original 391 children 123 were not followed up because they were younger than 6 years. From the remaining 268 children ≥6 years a total of 167 children were followed up (mean ± SD; 7.5 ± 0.84 years), whereas 101 children were lost for follow up. From 94 children a whole dataset was achieved including FeNO, lung function and SPT at school age, whereas from 73 children only questionnaire data was available. FeNO measured at age of 5 to 47 month in children with current asthma at school age was 9.65 (5.8–17.1) median (IQR), compared to 6.5 (4–9.8) in those without persistent asthma at school age ($p < 0.001$).
Conclusions: Wheezy young children less than 4 years of age and persistent asthma at school age have elevated FeNO levels. Further analysis will specifically look for effects of inhaled steroids at the time of FeNO measurement and various other influences, the calculations of sensitivity, specificity and predictive values. In addition the value of FeNO will be compared to the asthma predictive index, and whether the measurement of FeNO in addition to the API enhances the detection rate of future asthmatics at school-age.

P82

From initial common back pain in children to final diffuse metastatic rhabdomyosarcoma

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Introduction: Many children complain once in their childhood of back pain. Unlike adults, back pain in children is often associated with serious disease. We report on a 7 year old child who presented back pain, weakness and hypercalcemia.
Case report: Four weeks before diagnosis, the child experienced two traumas, one falling down the stairs without pain and one week later during a trampoline session with acute and persistant pain. In spite of appropriate analgesia with paracetamol and ibuprofen, back pain worsened with appearance of weakness, night pain and finally impossibility to move. At this moment, the child was addressed for further investigation. On physical examination, the child looked tired, pale, was slightly dehydrated and suffered intense back pain. No neurological deficit was found. Standard X-ray and CT of the spine showed compressive fractures of D4-D6-D7-D8-D11-L1-L4. Laboratory analyses revealed hypercalcemia, hyperuricemia, elevated LDH, and discrete thrombocytopenia. At this stage, a differential diagnosis between histiocytosis and leukaemia was considered. For diagnostic reasons, a bone marrow aspiration and biopsy were performed revealing surprisingly medullary infiltration by small blue round cells (95%), negative for histiocytic or leukemic markers, but positive for rhabdomyosarcoma. The alveolar type was confirmed by cytogenetics (fusion complex PAX3-FKHR with translocation T2;13). A small nodule on the postero-lateral face of the left arm proved to be the primary tumor by MRI and biopsy. Hypecalcemia was secondary to the diffuse bone metastasis and needed treatment with hyperhydratation, furosemide and pamidronate for several days.
Conclusions: Back pain in children is rarely common, especially in young ones. Persistence of pain, progression, occurrence at night and resistance to analgesia need active search for an underlying disease.

P83

Food poisoning leading to fulminant hepatitis

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Objective: Case report of a child with hepatic failure due to food poisoning by the emetic toxin of *Bacillus Cereus*.
Methods: Case report, literature review, and Discussion about the etiologic agent.
Results: A previously healthy 9 year old girl was brought to the emergency room by her parents in respiratory arrest, after several episodes of vomiting and a tonic-clonic seizure. After respiratory, haemodynamic, neurologic and metabolic stabilization, severe liver failure with clotting disorders, severe metabolic acidosis and hypoglycemia, was diagnosed. Extensive investigations failed to reveal an obvious cause for this acute hepatic failure (no evidence for viral, drug, toxic, autoimmune, metabolic and hematologic causes). Further questioning revealed that the patient had ingested a reheated meal of spaghetti carbonara about 12 hours before she was brought to the hospital. Microbiological investigations of the reheated spaghettis revealed the presence of the emetic toxin of *Bacillus Cereus*. After 2 days of intensive supportive care, the outcome was favorable and a liver transplantation could be avoided.
Conclusions: Although usually benign, food poisoning can, in rare cases, cause serious complications. Our Results show that the causal agent *Bacillus Cereus* and his toxin should be included in the differential diagnosis of fulminant hepatitis.

P84

Case report: acute adrenal insufficiency (addison crisis) under low-dose inhaled fluticasone proprionate

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Safety of inhaled fluticasone proprionate (IFP) has been established in children at doses of 200 mcg daily; however, doses up to

500 mcg IFP are generally considered as safe. We present a patient where under low dose IFP (250 mcg daily) acute adrenal insufficiency (AI) developed and made ICU treatment necessary. This seven year-old boy was referred from a regional allergy clinic with a 36 hour history of fatigue, lethargy, anorexia and vomiting. Fluticasone proprionate 125 mcg had been inhaled twice daily because of asthma over the past 19 months. Before arrival at the paediatric emergency unit, intravenuous fluids had been administered by a primary care team under assumption of developing septic shock. With this pre-treatment, our patient on admission had evidence of compensated metabolic acidosis (pH 7.34, pCO_2 27 mm Hg, BE –10.2), hyponatremia (125 mmol/l), mild hyperkalemia (5.2 mmol/l) and normoglycemia. A morning serum cortisol concentration measured eight days prior to admission had been extremely low (6.9 nmol/l, normative values 68–473); a work-up of this intriguing finding was not performed in due time. Serum cortisol concentration in our lab was only inadequately elevated given the clinical presentation and compatible with AI. Intravenuous high-dose glucocorticoid replacement and volume expansion brought about rapid clinical improvement. As a complication of AI rhabdomyolysis developed and made vigorous hydration with isotonic crystalloid liquids necessary. Follow-up 3 months later under reduced IFP doses showed normal random serum cortisol concentration and a normal ACTH test, adding to the evidence that in our patient AI had been triggered iatrogenically by the formerly used 250 mcg IFP. A further finding in our patient was markedly reduced growth velocity while on additional inhalative budenoside therapy until two months before onset of AI. We summarize that in our patient AI developed under low-dose inhaled corticosteroids; further, that the clinical presentation of AI in our patient was initially mistaken for septic shock, and that previous high dose inhalative steroid therapy had a systemic impact on our patient by means of impaired growth. We may conclude from our patient that AI may develop even under a low dose of inhalative corticosteroids, which is generally accepted as safe. Therefor, AI should also be considered in children with a history of low dose inhalative corticosteroids when presenting with respective signs and symptoms.

P85

The individual plan for school integration: results after three years of utilization in schools and preschool institutions in geneva

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Objectives: In September 2004 we introduced the Individual plan for school integration (projet d'accueil individualisé, PAI) in all schools and preschool institutions in Geneva. This document promotes quality of life through school and social integration of children and adolescents with chronic diseases, in defining the child's specific health needs, the treatments and measures necessary in the school, and the roles and responsibilities of the partners involved. We presented the first Results during the annual meeting of the Swiss Pediatrics Society (SSP) in Montreux (June 2005). Its utilization has now been generalized and we present here the Results for 2006–2007 after three years of experience.
Population and methods: The PAI is used for every child and adolescent who attends school, apprenticeship or preschool institution, with a chronic disease having an impact on his schooling. We now use two different types of PAI: the complete PAI which has been used since the beginning and the simplified PAI which was developed during year 2006–2007 for children with conditions having only few specific health needs at school (i.e. asthma). A monitoring sheet was filled by the school nurse or the school doctor for each child and adolescent with chronic conditions.
Results: From September 2006 to June 2007, 427 children and adolescents out of 75'232 benefited from a PAI (192 girls, 235 boys) representing a prevalence of 5.7 of 1'000. The median age was 9.1 years (1–21 years). The most frequent diagnoses were allergies (161), asthma (62), epilepsy (59) and diabetes (48). Most health needs regarded treatments (278), mobility (223), special diet (137), and sportive activities (120). For allergies, asthma and epilepsy, there were usually less than three different health needs, whereas for diabetes there were usually more than three. We filled 383 emergency protocols and 32 school evacuation protocols. Comparison with the previous year will also be provided.
Conclusions: The PAI is now well-known by all partners, and its utility acknowledged by the schools and the child/adolescent's doctors. It clarifies in lay language the problems that could happen in schools and provides solutions, while taking into account everybody's view. It reassures parents and teachers and develops a better knowledge of the children and adolescents with chronic diseases at school. The simplified PAI is especially adequate for more simple pathologies.

P86

Standardized exercise challenge test for preschool children using the forced oscillation technique

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Background: Asthma diagnosis and documentation of disease control in preschool children is difficult. Exercise induced bronchoconstriction (EIB) is a common feature of childhood asthma and tests to evaluated EIB are established for school-children. As young preschool-children are not able to cooperate to perform Spirometry, the gold standard of EIB-testing, no standardized test is available for children younger than 6 years.
Aims: To evaluate a standardized EIB-test for children between 3 and 6 years.
Methods: 42 children aged 3 to 6 years were included in a cross-sectional study. 19 with a doctor diagnosed asthma and 23 healthy controls. Children were running together with the investigator in order to achieve submaximal heart rate (>195 b/min, Polar®) within 2 minutes and to keep this heart rate for 4 minutes. Lung function was assessed by forced oscillation technique (FOT) and was performed at baseline and at 3, 6, 10, 15 minutes after the run. A positive EIA was defined as an increase in resistance of ≥30% in Rrs4-24Hz or 32% in Rrs8Hz to baseline.
Results: Demographical characteristics were similar except for allergic sensitisation ($p < 0.001$) and cough without cold ($P < 0.005$). Baseline Rrs8Hz and Rrs4-24Hz were similar in both groups (7.38 ± 1.3 kPa / [L/s] and 7.95 ± 1.9 kPa / [L/s], respectively; $p = 0.27$). A positive EIB test in was present in 10/19 children with asthma compared to 2/23 healthy controls ($p = 0.005$). The sensitivity of the test was 0.52, specificity 0.92, positive and negative predictive values were 0.83 and 0.72, respectively. If only symptoms (coughing, wheezing or desaturation) were taken into consideration the difference between the two groups was less significant ($p = 0.03$)
Conclusions: Regarding sensitivity and specificity of the test compared to the standardisized exercise challenge test in school children. In the current study normal values are to be established. Effects of current asthma-treatment are to be investigated.

P87

Exhaled nitric oxide and its role in monitoring asthma control in children in comparison to spirometry and bronchial responsiveness

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Objective: Despite highly effective therapy options being available, total asthma control is only achieved in a very small portion of asthmatic children. It could be expected that the measurement of objective inflammatory or physiological parameters would be helpful to monitor asthma.
Methods: In a double-blind, randomised controlled study, we recruited 30 mild poly-allergic, asthmatic patients on a low dose therapy (200 mcg BDP equivalent) with inhaled corticosteroids. We measured exhaled nitric oxide (eNO) with the Niox mino®, bronchial responsiveness to Mannitol (BR) and spirometry at visit 1 and visit 2 (three months later). The patients were randomly assigned to three groups according to the Results of the above mentioned measurements (group 1 = eNO, group 2 = BR and group 3 = spirometry, respectively). Anti-inflammatory therapy was doubled in group 1, when eNO at visit 1 was >22 ppb, in group 2, when bronchial hyperreactivity was present and in group 3, when FEV_1 was <80% of predicted, respectively, by a respiratory Physician, blinded to the remaining Results. In addition, the patients filled in an asthma control test (score) and a life quality questionnaire (QoL) at visit 1 and visit 2.
Results: 24 patients successfully completed the study (8 in each group). In group 1(eNO group) the mean (SD) eNO levels were 24ppb (±18ppb) at visit 1 and 11ppb (±9ppb) at visit 2 and improved significantly from visit 1 to visit 2 ($p = 0.02$). In addition, score and QoL improved from 22 to 24 ($p = 0.02$) and from 146 to 153 ($p = 0.05$), respectively. However, BR as well as FEV1 did not improve significantly ($p = 0.08$; $p = 0.2$). In group 2 and 3 there was no difference between visit 1 and visit 2 for all objective parameters (eNO, BR and spirometry) as well as for the score and QoL.
Conclusions: The guidance of anti-inflammatory therapy according to the objective measurement of eNO improves asthma control in mild poly-allergic, asthmatic patients and should therefore be recommended as a standard measure in the follow-up of these patients.

P88

Case report: it looks like an osteomyelitis but... what else could it be?

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Walking disability, cutaneous redness and tenderness near to a joint are not so rarely seen in general practice. Osteomyelitis or septic arthritis must be diagnosed very quickly in order to avoid serious complications. However, these symptoms can also be present in other illnesses that should not be forgotten because treatment and outcome are different. We present the case of a 3 years old boy who had pain in the left foot and was progressively wobbling. He had fever, redness and swelling of the left tarsus. No other joint was involved. An osteomyelitis was suspected and the child was hospitalized. The blood analysis revealed a maximal CRP at 19, ESR maximal at 62, WBC of 14 with left deviation and negative procalcitonin. Blood cultures were sterile. ASLO, FR and ANA were negative. As the child had diarrhea 2 weeks before the onset of the symptoms, serology for Lyme, Salmonella, Shigellosis and Yesinia were also done and were positive only for Salmonella. A foot X-ray showed nothing pathological. A scintigraphy showed hypercaptation in the left tarsus without clear localization, and no liquid collection seen by ultrasonography. No treatment was started. As the local status worsened, he was transferred to CHUV for investigations. An MRI showed an inflammation of the muscles around the tarsus and of the subcutaneous tissue compatible with a cellulitis without sign of osteomyelitis or osteoarthritis. An infectious disease was diagnosed and an antibiotherapy was started. The evolution slowly improved but there was no resolution. He was then hospitalized again and was seen by our orthopedist who diagnosed a Köhler disease because of the local status, the absence of infectious signs on the blood tests and the X-ray that showed an osteonecrosis of the navicular bone. The antibiotherapy was then stopped and a synthetic plaster was put for 6 weeks. After 6 weeks of plaster immobilisation, clinical examination was normal. X rays confirmed the diagnosis of Kohler disease. Köhler disease is an osteochondrosis of the tarsal navicular. It is seen in children between 2 and 9 years old without sex ratio. The main symptom is foot pain that is augmented by acupressure. The X-ray shows a small tarsal navicular with high density. The resolution is spontaneous and without repercussion.
Köhler disease should then be part of the differential diagnosis of each suspected osteomyelitis or cellulites that has an atypical clinical presentation or follow-up.

P89

Case report: recurrent fever is not always a pfapa

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Fever is a very frequent symptom in children. Recurrent fever is less frequent but it is a problem that every pediatrician can see. The more common cause is recurrent viral infection but autoinflammatory diseases should not be forgotten. We present the case of a 5 years old girl who had recurrent fever episodes of 38.5°–40°C, lasting 3 or 4 days every 2 weeks since the age of 1 year. Fever was associated with pharyngitis (streptococcus negative, except twice), aphtous stomatitis, vomiting and abdominal pain, cervical and inguinal adenopathies. There were neither cutaneous eruption nor arthralgia. Between the episodes she was doing well and her physical examination was normal. The laboratory showed systemic inflammation (ESR 30, CRP 117), normal liver and kidney values, slightly elevated IgG and IgA values, normal IgM and IgD. The clinical presentation suggested a PFAPA syndrome and we completed the investigations to exclude other autoinflammatory diseases. A blood cells count, done the first day of the fever, allowed to exclude a cyclic neutropenia and a genetic analysis excluded a TRAPS (TNF receptor associated periodic fever). She had a good antibody response to vaccines except for Pneumococcus. She was then vaccinated with Prevenar® and the response was good. While waiting for the Results our patient was given oral prednisone at the beginning of each fever episodes and had a good response to that treatment. The urinary analysis that was done during an episode of fever revealed a high concentration of mevalonate (20 mmol/ mol creatinin, N <2) that was confirmed by a second analysis. The diagnosis of HIDS (hyper IgD syndrome) was confirmed by a genetic analysis showing 2 MVK mutations : p. Ile268Thr/ p.Val337Ile.Because she was heterozygous, a spontaneous mutation was unlikely and we recommended a genetic analysis of the parents.
Our case shows that PFAPA (periodic fever, aphtous stomatitis, pharyngitis and cervical adenitis) and HIDS may have a very similar clinical presentation. Except for the very short periodicity of the fever episodes, our patient presents as a typical PFAPA . Moreover, our patient had normal IgD values which did not exclude HIDS, as previously described in the literature.
In patients where a PFAPA is suspected, other autoinflammatory disease should be evoked and investigated. In particular, urinary mevalonic acid at the beginning of the fever episode should be measured.

P90

Arterial hypertension in childhood – diagnosis and differentialdiagnosis remain a challenge

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Aims: Arterial hypertension in childhood remains a special diagnostic and differential diagnostic challenge for the paediatrician. Owing to its low incidence and uncommon occurrence, a delay in diagnosis or even a misdiagnosis are regularly observed. We describe a case where the correct diagnosis was first established in early adolescence and where the delay had important health implications for the patient.
Case report: A 12 year old previously apparently healthy boy presented to his paediatrician with new onset of exercise induced precordial chest pain. The physical examination revealed arterial hypertension with a systolic pressure of 160 mm Hg as well as a systolic murmur. Referral to an adult cardiologist resulted in the diagnosis of assumed familial idiopathic arterial hypertension. As part of the work-up to exclude secondary renal involvement, the patient was referred to a paediatric nephrologist who in turn suggested a consultation by a pediatric cardiologist. Finally the diagnosis of a severe arterial hypertension (systolic BP right arm 200 mm Hg) caused by a coarctation of the isthmus of the aorta (gradient 110/47 mm Hg) was established clinically and echocardiographically. In addition a mild stenosis of the aortic valve and a severe left ventricular hypertrophy were established. More than one year after diagnosis and consequent successful implantation of dilatative stents at the coarctation the patient requires drug therapy for a persisting arterial hypertension.
Conclusions: Standardised monitoring of blood pressure as well as bilateral palpation of peripheral and inguinal pulses should remain part of the regular paediatric assessment.

P91

Single origin of the right pulmonary artery from the ascending aorta –- rare differential diagnosis of failure to thrive in children

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Objective: Anomalous origin of one pulmonary artery from the ascending aorta is a rare anomaly, which usually presents with congestive heart failure at an early age.
Method/result: We report about a 3 months old boy, who was sent to our outpatient clinic for evaluation of failure to thrive. The patient presented in congestive heart failure with respiratory distress and tachypnea. Physical examination showed a hyperactive precordial impulse and a 3/6 medium to high pitched murmur at the left sternal border. Echocardiography revealed an anomalous origin of the right pulmonary artery from the ascending aorta with high blood flow. The left pulmonary artery had its normal origin. The pressure in the right ventricle was systemic. The little boy underwent surgical correction with separation of the right pulmonary artery from the aorta and reimplantation in the common pulmonary artery. He had an uneventful recovery although the pulmonary hypertension required selective vasodilatators to normalise more rapidly.
Conclusions: Congenital heart disease is a well known differential diagnosis in evaluating children with failure to thrive. Although frequent lesions as VSD or AV canal are more likely, rare reasons as anomalous origin of a single pulmonary artery has to be kept in mind.

P92

Maternal luteoma, an unusual cause of virilization of a female newborn

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Background: Virilization in female newborns most frequently Results from congenital adrenal hyperplasia secondary to 21 hydroxylase deficiency. Intersex genitalia in the newborn require urgent management as they could be the first manifestation of a life-

threatening adrenal insufficiency. If the mother presents signs of virilization during pregnancy, the differential diagnosis must include a maternal androgen-secreting tumor, a fetal aromatase deficiency in which fetal androgens cannot be converted into estrogens by the placenta resulting in virilization of the fetus and the mother, or a P450 oxidoreductase deficiency, a congenital adrenal hyperplasia with combined deficiency of 21 hydroxylase and 17 -hydroxylase associated with various degrees of adrenal insufficiency, ambiguous genitalia, polycystic ovaries and skeletal malformations
Case report: A full term newborn from non-consanguineous parents was admitted for a disorder of sex development with marked clitoromegaly without palpable gonads in the genital folds. Pregnancy was marked by virilization of the mother with deepening of her voice and excess hair growth on her face and abdomen. Karyotype of the newborn revealed 46XX and 17OH-Progesterone was normal, ruling out classical adrenal hyperplasia. The other adrenal and ovarian hormones were within the normal range on day 3 and MRI excluded polycystic ovaries. High levels of total testosterone were detected in maternal serum five days after delivery (>50 mcg/l [0.05 mcg/l – 0.95 mcg/l] and pelvic ultrasound showed an ovarian mass which was confirmed by the CT-scan and compatible with a luteoma or a primary malignancy of the ovary. The spontaneous regression over two weeks, with normalization of the testosterone level, led to the Conclusions of a maternal luteoma. Without treatment all signs regressed in the mother, but clitoromegaly persisted in the newborn. Surgical intervention will be planned later.
Discussion: The first step in the approach of a newborn with ambiguous genitalia is to exclude a life-threatening congenital adrenal hyperplasia. The 17OH-Progesterone must be measured initially. Maternal luteoma is a rare benign tumor of pregnancy that must be considered in case of maternal virilization. It is of spontaneous resolution and fetal virilization Results from tumoral hypersecretion of testosterone with passage through the placenta. This case emphasizes the importance of a detailed maternal history, which can rapidly lead to the diagnosis.

P93

Short-term outcomes in a group of children with ASD following with a developmentally based model

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The present work shows the rehabilitation process and the evolution of a little group of 3 children with ASD. All the children presented severe ASD symptoms (absence of language, lack of interaction, auto and hetero-aggressive behaviors, stereotypes and perseveration). One of the children had also West Syndrome. They were all diagnosed by clinical and standardized test by NPI of Italy. The rehabilitation process took place during six months based on DIR principles (Stanley Greenspan & Serena Wieder) and CIDT (clinical intervention developmental team): an intensive comprehensive developmental based program, working in a permanent alliance with family and school. The Program took into account the functional emotional developmental level of each child, his sensory and motor profile and the kind of relationship established with caregivers and team. Family, interdisciplinary team and school worked together taking into account the strengths and weakness of each child and designing an individualized approach to each child. After 6 months of intervention important clinical Results were seen: all the children were more attentive and engaged and have developed grades of intentionality and communication, the auto and hetero-aggressive behaviors have totally diminished and children were more adapted at school and at home. The Aim of this work is to demonstrate that even though the initial late age of the intervention (seven years) and the severity of the symptoms, a good evolution could be expected with the implementation of an intensive developmental based program. All work and evolution is accompanied by videos.

P94

An unusual case of severe anemia and thrombocytopenia in a 10 month old infant in switzerland

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We report the case of a 10 month old infant with severe malaria, who presented with a high parasitemic load. Following diagnosis, he was treated in our clinic in collaboration with the Swiss Tropical Institute. Despite advanced hemolysis and the rarity of this disease in infants in Western Europe, prompt recognition and appropriate treatment led to a favourable course.
Case: A 10 month old infant was sent to our clinic due to recurrent fever with anemia and thrombocytopenia. The otherwise healthy boy had visited his family in Guinea and the malaria prophylaxis with Mefloquin had been stopped early due to application problems. On the return voyage he developed a recurrent, gradually climbing fever, which was controlled by acetaminophen, so medical attention was not sought for 6 days. On hospital admission physical exam showed an infant in a reduced general condition, with multiple petechiae. His temperature was 37.8 °C. Lung examination revealed generalised rales and tachypnea, the liver and spleen were palpable 4 cm below the costal arch. Laboratory studies showed anemia, thrombocytopenia, elevated liver and inflammatory parameters and signs of hemolysis. Glucose, INR, TT, kidney and urine analysis were normal. In the thick blood smear malaria was confirmed, the thin smear revealed an 8% infestation with Plasmodium falciparum. After consulting the Swiss Tropical Institute he was started on an iv antimalarial regimen with quinine and clindamycin (each 10 mg/kg tid) for 7 days. Over the following 3 days the parasite load gradually decreased, but he developed severe hemolysis. The renal function was maintained. Due to symptomatic anemia he required two blood transfusions. The thrombocyte nadir was 50 G/l. Ten days after hospital admission the patient had fully recovered.
Discussion: In Western Europe malaria is a rare disease usually related to travelling to endemic areas. Children of citizens with a migration Background, the “visiting friends and relatives”, are at particularly high risk. Between 2003 and 2005 666 cases of malaria were reported in Switzerland. Only 20% of infections occur in patients younger than 19 [1]. No such case of severe pediatric malaria with poor prognostic factors (low age, high parasite load, respiratory distress) in Switzerland has been previously published.
Our case highlights the importance of close collaboration between institutions (referring pediatrician, pediatric hospital and tropical institute) to establish the optimal therapeutic care. This management resulted in a favourable course and a good outcome despite the fact of relatively delayed referral and poor prognostic findings at initiation of therapy.
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P95

Hypercalcemia in children associated with congenital toxoplasmosis and cytomegalovirus infection

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Introduction: Hypercalcemia in children is a rare condition, explained in the majority of cases by hyperparathyroidism, hypervitaminosis D, alterations of vitamin D metabolism and malignancy. Only a few infectious diseases have been reported to cause hypercalcemia: tuberculosis, leprosy and some fungal infections (coccydioidomycosis, histoplasmosis, candidiasis, Pneumocystis carinii).
Case presentation: We present a 13 months old boy, born in Peru at 36 weeks of GA, who developed after a few months growth retardation with microcephaly, anorexia, fatigue, crying, polyuria-polidipsia, hepato-splenomegaly, iron deficiency anaemia, dyserythropoiesis and leucopoenia. A congenital toxoplasmosis (mother also infected, with positive IgM), a cytomegalovirus infection and a severe hypercalcemia (3.90 mmol/L) with hypercalciuria (8.4 mmol/mmol creatinin) and hypophosphatemia (1.0 mmol/L) were diagnosed. In several measurements, 25 OH-D3, 1,25-OH2-D3 were normal, PTH is very low and PTHrP is normal. We did not find evidence of malignancy, of granulomatous or genetic diseases and conditions to explain hypercalcemia. The only diseases diagnosed were a congenital toxoplasmosis and a probably acquired cytomegalovirus infection, never reported before in association with hypercalcemia. Considering the impairment of this hypercalcemia with specific symptoms, after two months of observation, we treated the boy with pamidronate 7 mg i.v. Due to the relapse of hypercalcemia (3.21 mmol/L) with related symptoms, a second dose of palmidronate was given 5 weeks laters. Thereafter, the serum calcium values became normal, the hypercalciuria resolved and the symptoms disappeared, with a catch-up growth in stature and weight. Simulatenuously, the child was treated for toxoplasmosis with pyrimethamin and sulfadiazin.
Conclusions: Infections and granulomatous diseases are suspected to cause macrophages activation and the production of metabolites that may induce hypercalcemia. The presentation of similar cases may help to confirm this hypothesis or to develop new ones regarding calcium homeostasis of the organism. Biphosphonate therapy induced a rapid and complete resolution of the hypercalcemia and related symptoms.

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Retinopathy, progressive intracerebral calcification and epilepsy in a premature new-born: case report of a new entity

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Background: The most common cause of cerebral calcifications and retinopathy in infancy are intrauterine infections by toxoplasmosis, rubeola, CMV and herpes (TORCH). Calcifications can also be seen in tumoral, endocrine, metabolic and degenerative conditions (Aicardi-Gouttière, Cockayne syndromes). We present an infant with a severe retinopathy, first attributed to prematurity, and progressive cerebral calcifications who was finally diagnosed with a recently identified disease, cerebral retinal microangiopathy with calcifications and cysts (CRMCC,Crow et al, 2008)
Case report: This prematurely born (28 weeks) 18 month-old boy had a history of intrauterine growth retardation, intraventricular hemorrage and severe bilateral retinopathy. Cerebral echography at 3 weeks revealed a few periventricular echogenicities. At 9 months, he was hypotonic with rowing eye movements and nystagmus. Head circumference was normal. Development was mildly delayed. At 11 months he presented partial seizures, lethargy and irritability. Multiple intracerebral calcifications and hydrocephalus were seen on CT scan and a VP shunt was inserted. Cerebral MRI showed extensive bilateral cystic thalamic lesions with calcifications, and diffuse white matter abnormalities. CSF showed pleiocytois, elevated proteins and lactate. PCR in CSF and serologies for TORCH were negative Serum lactate, calcium, phosphate, alpha-interfron (also in CSF) were normal.
Conclusions: CRMCC is a new multisystemic disease entity of probably genetic origin which includes Coats retinopathy and leucoencephalopathy with calcifications and cysts (Labrune et al, 1996). Very early onset has been described in only one case and represents a diagnostic challenge.

P97

Lung function and respiratory symptoms at school age in children after premature birth due to amniotic infection or pre-eclampsia

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Background: Preterm infants exposed to amniotic infection (AI) have better postnatal pulmonary function than those exposed to pre-eclampsia (PE) despite accelerated morphologic lung maturation in the latter. Most likely, this is due to a relative lack of surfactant, ascribable to low stimulant cytokine and high TNF-alpha levels. Preterm after AI may benefit from increased activity of the Bronchus-associated lymphoid-tissue (BALT). Various lines of evidence suggest that perinatal factors are important in determining susceptibility to atopy and asthma. In a follow-up study we tested the hypothesis that preterm infants exposed to AI have a higher risk to develop asthma and allergies than children exposed to PE.
Methods: A total of 65 ex-premature born children were followed up at an age between 7 and 13 years. All children have been treated at the neonatology department of the university hospital Zurich and extensive data on perinatal outcomes was available for all children. The assessment included measurements of fractional exhaled nitric oxide (FeNO), pulmonary function (spirometry, whole body plethysmography and forced oscillation [FOT]), bronchial responsiveness to methacholine, skin prick test. The history of respiratory symptoms was assessed using a modified ISAAC-questionnaire.
Results: A full data set was obtained in 37 of the children; partial assessment was obtained in the remainders. Children after PE showed significantly increased airway resistance (FOTavr4-24Hz; $p = 0.03$) and a lower ratio FEV_1/FCV ($p = 0.013$), indicating smaller airway caliber. For all other parameters no difference was found between the two groups. The vast majority of children (83.8%) showed severe (52%) to moderate (31.8%) bronchial hyper-responsiveness.
Conclusions: Bronchial hyper-responsiveness persist to school age in most the children after premature birth. There is no evidence that preterm infants exposed to AI have a higher risk to develop asthma or allergies later in life compared to preterm infants exposed to PE. In contrast children after PE show a tendency for increased airway obstruction.

P98

Invasive infections by group a beta-haemolytic streptococcus cause similar morbidity compared to invasive pneumococcal and meningococcal infections

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Introduction: Invasive infections are known for serious complications such as septic shock, multiorgan failure and have significant mortality. *Streptococcus pneumonia***e** and *Neisseria meningitidis* are the most common causative pathogens beyond the newborn period. Group A beta-haemolytic streptococcus (GAS) causes a variety of infectious diseases ranging from acute bacterial pharyngitis and pyoderma to invasive and toxin-mediated diseases.
Patients: Between January 2007 and 2008 nine children aged 2 to 13 years presented at our emergency department with septic shock due to GAS. Streptococcal septic shock followed skin or upper respiratory tract infection in all nine cases, including a 2-year-old boy with chicken pox, a 2-year-old boy with paronychia, a 7-year-old-girl with appendectomy, a 5-year-old boy with Influenza A infection, a 2-year-old girl with otitis media with perforation, a 6-year-old boy with mastoiditis, and two boys and one girl, aged 5 to 13 years with acute pharyngitis. After preceeding infection lasting one to seven days, all children deteriorated within less than one day, proceeded to sepsis, to septic shock and consecutive multiorgan failure. 8 of 9 patients needed full cardiovascular support at the intensive care unit. Invasive GAS infection was confirmed by positive culture from blood or pleural fluid in eight patients. Two patients had antibiotic treatment initiated before presentation. All nine patients survived, however one patient developed severe necrotising fasciitis and fully recovered but another following septic ischemic necrosis partially lost his arms and legs. In the same period of time we diagnosed meningococcal sepsis in six children, aged 1 to 15 years and pneumococcal sepsis in eleven patients, aged 6 months to 15 years.
Conclusions: Invasive GAS infections cause significant morbidity. While varicella and influenza infections are well-known risk factors predisposing for severe GAS infection in our case most children had presumably preceeding common viral infections. Not only the frequency of invasive GAS infection in our paediatric emergency patient group was comparable to invasive pneumococcal and meningococcal infections but also its course was as fulminant and severe. Clinical observation and perception of rapid deterioriation are the only clues to early diagnosis.

P99

Post-Kawasaki disease arthritis

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We report the case of a patient who developed symptoms of polyarthritis after being treated with immunoglobulin (IVIG) for Kawasaki disease (KD). A three year old previously healthy girl presented with the criteria for KD. She was treated with IVIG and high dose aspirin. Due to a persistence of fever, she was subsequently treated with a second dose of IVIG with an excellent clinical response. En days after discharge from the hospital, the patient was rehospitalized for reapparition of the lymphadenopathy, and polyarticular arthralgia with a refusal to walk primarily in the morning. Lab studies showed anemia, elevated CRP, WSR, leukocytosis and thrombocytosis. In view of the clinical picture, the differential diagnosis of juvenile idiopathic arthritis systemic form, an infectious process, a relapse of Kawasaki disease, or a tumoral etiology was evoked. Viral serologies and the Mantoux test were negative. A tumoral etiology was excluded due to the normalization of the lymphadenopathy without treatment, normal imagery, and lab studies. Rheumatoid factor and ANA were also negative. A review of the literature, revealed five other cases of arthritis post-Kawasaki disease. These patients responded to treatment with high dose aspirin, ibuprofen, or steroids. Our patient was treated with ibuprofen at 40 mg/kg. This treatment led to an excellent clinical evolution. Arthritis may be seen in the acute or subacute stage of KD in 15–45% of patients. According to the literature, there are two clinical scenarios in which arthritis is a symptom. In the first, the arthritis is polyarticular and appears at the same time as the other classical symptoms of KD involving the small or large joints. In the second, the arthritis appears ten days after the illness and implies mostly the larger joints. The incidence of post-Kawasaki arthritis is reported at 2%. These cases are reported to have elevated inflammatory markers. This form of polyarthritis usually cedes after treatment of 10 to 60 days. The etiology of this complication is not yet clear, but there are suspicions that this may be a reactive arthritis due to an unknown pathogen. The diagnosis of polyarthritis post-Kawasaki is a difficult one to make and should always be considered when there is a persistence of arthritis and elevated inflammatory markers after the diagnosis of Kawasaki disease.

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The pediatrician before the psychological consultation

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The Aim of this work is to emphasize the priviledged place the pediatrician has as the first contact with the child's psychological symptomatology and the clinical implications it determines whether is is its early detection or its correct treatment. The pediatric consultation characteristics in relation to the adult are analized, the different psychological symptom modalitites are detailed, and the most frequent behaviours before such symptoms are described. Furthermore, the manifested and latent content present in the consultation are conceptualized. Finally, a parallel is drawn between the somatic and psychological symtomatology and the pediatrician's conceptualization of both. Potential risks of wrong interpretation are described and an analysis of the different treatment options is presented together with clinical examples.

P101

Pityriasis lichenoides et varioliformis acuta and typhoid fever

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Introduction: Pityriasis lichenoides (PL) is a rare cutaneous disorder of unknown aetiology. There are two types: Pityriasis lichenoides et varioliformis acuta (PLEVA or Mucha-Habermann) generally presents as an acute-to-subacute skin eruption of multiple, small, red papules that develops into polymorphic lesions and vacillates with periods of varying remissions as well as possible sequelae of hyper/hypopigmentation and varicella-like scars. Pityriasis lichenoides chronica (PLC) has a more gradual manifestation of very small red-to-brown flat maculopapules with mica-like scale; it also follows a relapsing course but with long periods of remission. They are believed to be different presentations of the same process, with the process being more rapidly accelerated in PLEVA. There is currently no standard of therapy.
Case report: We present a 9 10/12 year old originally Afghan boy, living in Switzerland for five years without any contact to foreign people, in slightly reduced general condition with existing fever since 2 weeks up to 39.5 °C. There was a cutan rash with papules and vesicles, partly necrotic eruptions and scars, which has a history of 14 months with fluctuating localization. The biopsy in May 2007 was histopathologically compatible with PLEVA. The serologies for HIV, toxoplasmosis, EBV, adenoviruses, VZV and CMV were all negative. After a positive test for salmonella in the stools and the diagnosis of Typhoid fever we started antibiotic therapy wich resulted in a rapid decrease of the fever and complete remission from the skin lesions.
Conclusions: A number of acute exanthems (eg, PL, pityriasis rosea, acute lichen planus, guttate psoriasis, erythema multiforme) are believed to be caused by a hypersensitivity reaction to infectious agents. Familial outbreaks, clustering of cases, and co-morbid symptoms have been used to support these relationships in PL, although clear causality is lacking. Elevations of pathogen-specific antibody titers also have been offered as proof of causality, but such immunologic responses may represent epiphenomena caused by non-specific immune responses to unknown pathogens. The most commonly reported associated pathogens are EBV, Toxoplasmosis and HIV. In our case we assume a relation with Salmonella typhi.

P102

Long term outcome of catheter-related arterial thrombosis in infants

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Background: Catheter-related arterial thrombosis is an increasingly recognized complication in infants with congenital heart disease undergoing cardiac surgery or catheterization. No information is available on long term outcome of these thrombotic complications.
Methods: Children with catheter-related thrombosis of the inguinal arteries occurring at our institution between 12/01 and 5/07 were investigated. Follow-up consisted of a Doppler-ultrasonographic examination and a clinical examination including measurements of blood pressure and both leg length and circumference.
Results: Data of 50 patients with a total of 54 catheter-related arterial thrombotic complications are presented. The median age of patients at the time of arterial thrombosis was 45 days (range: 1 day – 7 months). Arterial thrombosis was related to indwelling arterial catheters in 34% and cardiac catheterization in 66%. Forty-six of the 50 patients were treated initially with heparin for a median of 20 days, followed by acetylsalicylic acid in 29 patients for a median of 5.5 months or oral anticoagulation in 5 patients for a median of 3 months. Three patients received initial therapy with acetylsalicylic acid only and one patient was not treated. Follow-up was performed at a median of 19 months (range: 2 months – 5.5 years) after diagnosis. Doppler ultrasound examinations in 44 of the 50 patients showed complete resolution in 32 (73%) and no resolution in 12 (27%). Differences in blood pressure of more than 15 mm Hg was present in 7 (14%) of the 50 patients, in 6 of them Doppler ultrasound revealed no resolution of thrombosis. Leg length difference of more than 1cm was present in 4 (8%) of 50 patients, 2 of them showing no resolution by Doppler ultrasound. Difference in leg circumference of more than 1cm was present in 7 (14%) of the 50 patients. Of these 7, only 1 showed no resolution by Doppler ultrasound. In 1 patient claudicatio was present.
Conclusions: Catheter-related arterial thrombosis in infants with congenital heart disease is associated with long term complications including persistent vascular occlusion, leg length differences and loss of important vascular access for future catheterizations. Further studies are required to assess optimal prophylaxis and treatment of these thrombotic complications.

P103

Psychosomatic versus somatopsychic disease: a tricky question

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We present four cases treated in our children's clinic:
1) A 14 year old boy was admitted to our hospital for clarification of headaches and visual changes, present for over 2 years in combination with a depressive mood. All investigations to date had been negative and the referring colleagues presumed the headaches to be in connection with his mental state.
2) An 11 year old boy presented with severe pain in his gluteal region. He showed alterations in psychological state and a hysterical attitude towards all investigations.
3) A 12 year old girl was hospitalised after a bicycle accident with cerebral concussion. Apart from a persisting headache, neurological examination showed no pathological findings. Her Background and behaviour were highly suggestive for a psychological cause of the headache.
4) The final case is that of a 10 year old girl previously hospitalised with abdominal pain, coincident with colonisation of the bowel with Blastocystis hominis and, due to a difficult social situation, thought to be psychosomatic in nature. Three months later, the girl complained of leg pain.
All four cases presented with severe stress factors and behaviours suggestive of psychological origin, leading to initial suspicion of psychosomatic illness. Contrary to the first impressions, further investigations revealed the presence of considerable somatic disease (pseudotumor cerebri (1), chronic recurrent multifocal osteomyelitis (2), septic sacroileitis (3), skull fracture with intracerebral hematoma with sinus vein thrombosis (4)).
Our examples show the importance of regarding each case objectively and without presumption, even in the presence of a very likely psychological etiology (influenced for example, by circumstances of referral, suggestive behaviour, psychosocial stress factors etc.). Although physical symptoms in all four cases could easily be explained by underlying psychoemotional stress factors, and in each case a substantial need for psychotherapeutic treatment was ascertained, careful investigation revealed treatable somatic disorders. Therefore, these cases confirm that precise evaluation for somatic disease is an absolute necessity, even in presumably apparently clear cases of psychosomatic cause.

P104

Effects of leboyer delivery on blood viscosity, hemorheologic and circulatory parameters in term neonates

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Objective: This study was done to compare postnatal alterations in blood viscosity, hematocrit value, plasma viscosity, red blood cell aggregation, and red blood cell deformability in term neonates undergoing both early umbilical cord clamping and delivery according to the Leboyer method.
Study design: The umbilical cords of 15 healthy, term infants were clamped within 10 seconds of birth (early cord clamping), and 15 infants delivered according to the Leboyer method were placed on the mother's abdomen, and the umbilical cords were clamped

3 minutes after birth. Hemorheologic parameters were studied in umbilical cord blood at 2 hours, 24 hours, and 5 days from the time of delivery.
Results: The residual fetal placental blood volume decreased from 45 ± 8 ml/kg (x ± SD) after early cord clamping to 25 ± 5 ml/kg after delivery by the Leboyer method. After Leboyer-method delivery, the hematocrit value rose from 48% ± 5% at birth to 58% ± 6% 2 hours after delivery, 56% ± 7% at 24 hours, and 54% ± 8% after 5 days. Blood viscosity in the Leboyer-method group increased by 32% within the first 2 hours but did not change significantly during the following 5 days. Plasma viscosity, red blood cell aggregation, and red blood cell deformability were not affected by the mode of cord clamping.
Conclusions: Delivery by the Leboyer method leads to a significant increase in blood viscosity as a result of increasing hematocrit value, whereas other hemorheologic parameters are similar to those of infants with early cord clamping.

P105

Transient erythroblastopenia of childhood (TEC) and colitis both associated with human herpes virus 6 (HHV- 6): a case report

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Introduction: We describe the case of a 19 months old boy suffering from transient erythroblastopenia of childhood (TEC) and, timely associated, bloody diarrhea. Both anemia and diarrhea were associated with human herpes virus (HHV-6) infection. To our best knowledge HHV-6-associated colitis in an immuncompetent host has not been described to date.
Methods: TEC is characterized by temporary anemia caused by a decrease of red blood cell precursors. Among other viral infections HHV-6 has been implicated in pathogenesis of TEC. The boy was admitted to our hospital because of anemia (Hb 57g/l) of unknown origin. He had no other symptoms. Upon a complete diagnostic investigation the diagnosis of TEC was made and HHV-6 as etiological agent was detected in the bone marrow aspirate by PCR. Shortly after the patient presented with persistent partly bloody thin stools. Colonoscopy showed an unspecific colitis with a predominance of eosinophils. Biopsy of the colon was positive for HHV-6 (PCR) as well. Both anemia and colitis disappeared or resolved spontaneously, and the boy has fully recovered.
Results: HHV-6 was detected in the bone marrow aspirate by PCR as cause for the described episode with TEC. Furthermore, HHV-6 has also been isolated in the biopsy specimen from the colon epithelia by PCR in the context of the accompanying colitis.
Conclusions: We describe an episode of TEC caused by HHV-6 infection accompanied by an HHV-6 colitis. Taken into account the timely association and the clinical presentation it is very likely that both symptoms were associated with the same pathogen, HHV- 6.

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Paediatric registries in switzerland – overview and call to join forces

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Aims: Patients registries are essential tools for surveillance of rare diseases and contribute to improve diagnostic and therapeutic approaches. Our Aim was to gain an overview over paediatric registries in Switzerland, to exchange information and to provide mutual support.
Methods: Via a mailing addressed to the president of the SSP, to all heads of tertiary care centres, to all presidents of paediatric subspecialties and to other personal contacts we searched for existing and planned paediatric registries in Switzerland and organised a first meeting.
Results: The meeting was attended by the coordinator of Orphanet Switzerland (www.orpha-net.ch) and by representatives of five existing paediatric registries: the Swiss Childhood Cancer Registry (www.kinderkrebsregister.ch), the Swiss Neonatal Network and Follow-up Group (www.neonet.unibe.ch), the Swiss Neuropaediatric Stroke Registry (www.neuropaediatrie.ch\snpsr) the Swiss neuromuscular network (www. treat-nmd.eu) and the Swiss Paediatric Renal Registry. Registries being planned: the Lysosomal Registry, the Swiss Cleft Palate Registry and the Paediatric Rheumatology Registry. These registries Aim to describe incidence, prognosis and time trends of rare diseases in Switzerland, contribute to aetiological research, evaluate current diagnostic work up, treatment and follow up procedures and assist in joining international clinical studies. Registries are also important to facilitate provision treatments for rare diseases. All registries share common problems: completeness of case ascertainment and follow-up, ethics approval (23 different ethics committees), creation and support of databases, legal support, and problems with funding, because usual funding bodies such as the SNF do not support the daily running of registries.
Conclusions: Paediatric registries are not only good research instruments, but also essential tools to improve and standardise diagnostic work-up and care for children with rare diseases. Problems with case ascertainment and follow-up, methodological problems, legal barriers and difficulties to obtain funding are shared by all of them. Joining forces and collaborating within the Swiss Society of Paediatrics might help to maintain and further develop these essential tools. People who are interested to join the group contact Maja Steinlin (maja.steinlin@insel.ch) or Claudia Kuehni (kuehni@ispm.unibe.ch) or one of the other co-authors.

P107

Oropharyngeal trauma in children: don't brush it off too easily

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Background: Oropharyngeal trauma in children is a common event, typically caused when the child trips over holding an elongate object (like a toothbrush) in his mouth. Fortunately most of these accidents are benign, but potentially life threatening injuries do occur.
Methods: Between January 2004 and March 2008 four children with upper airway obstruction following oropharyngeal trauma were diagnosed at our emergency department. Signs and symptoms at presentation, diagnostic examinations and outcome of these children were reviewed.
Results: Two children were girls, two were boys. The mean age at diagnosis was 5 years. In three cases the penetrating object was a toothbrush and in one case a drumstick. In one patient the toothbrush was still lodged in the oropharynx and had to be removed surgically. In the other cases the objects had already been removed either by the child or his caretakers. Three accidents were caused by falls, in one case the girl reported that her mother had intentionally stuck the toothbrush into her throat. All patients initially remained asymptomatic (6 to 48 hours) and then progressively developed a painful neck swelling with signs of upper airway obstruction. One boy presented with serious respiratory distress. Although visual inspection of the oral cavity was unremarkable in all patients, conventional X-ray and CT scan identified para- and retropharyngeal swelling and emphysema. All children received antibiotics and anti-inflammatory drugs. Three children could be treated conservatively. In one case a surgical intervention was necessary (to remove the toothbrush), this girl developed a deep abscess above the parotis one month later and had to be treated surgically again. All children recovered without sequelae.
Conclusions: Complications following oropharyngeal trauma typically occur after a symptom-free interval of several hours. A thorough history of the accident is necessary to identify potential child abuse. Due to the mechanism of injury the oral impact zone normally can't be seen directly. Careful instruction of the parents when to return is mandatory. A follow up visit should be scheduled and in-hospital-observation as well as treatment with antibiotics and anti-inflammatory drugs should be considered early. In case of upper airway obstruction a lateral cervical X-ray is helpful to quickly assess the extent of the injury.

P108

Foreign material associated early postoperative infective endocarditis in complex congenital heart malformation

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Introduction: Foreign material is frequently used in cardiac surgery for the operative treatment of complex congenital heart defects (CHD). Nevertheless, foreign material is a well known risk factor for postoperative infective endocarditis (IE).
Methods: Case report of an early onset postoperative IE in an infant operated for a complex CHD.

Case report: A 4-day-old boy was operated for Taussig Bing anomaly with hypoplasia of transverse aortic arch and coarctation by arterial switch, extended resection of coarctation, combined with enlargement of the aortic arch and closure of a ventricular septal defect (VSD) using a xenopericard patch. On postoperative day 7 he developed fever, tachypnea and showed reduced microcirculation. C-reactive protein (CRP) was elevated (250 mg/l), blood cultures grew Staphylococcus aureus and echocardiography demonstrated a vegetation of 8x5 mm attached at the right side of the patch. Imminent pacing failure required replacement of epicardial pacing leds which were colonized with coagulase negative staphylococci. With antibiotic treatment with flucloxacillin, teicoplanin and gentamicin the patient merely stabilized, blood cultures were negative only after 7 days but a new VSD within the infected patch material developed on day 16. This early onset postoperative IE required re-operation to control the overwhelming endocarditis. The xenopericard patch was resected and the residual VSD could be closed directly without further use of foreign material. Cultures from resected patch and close endocardial samples grew Staphylococcus aureus. The patient showed a rapid postoperative recovery. After antibiotic treatment for 4 weeks the child could leave the hospital in good clinical condition.
Conclusions: Early onset foreign material associated IE after pediatric cardiac surgery has to be taken into account as a potential life-threatening complication. Early diagnosis by clinical symptoms, blood culture and echocardiography may implicate early surgical redo, because infectious control may not be achieved with antibiotics alone.

P109

Premature closure of the ductus arteriosus in utero: treatment and evolution

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Introduction: The ductus arteriosus (DA) plays a major role in fetal circulation. Antenatal ductus closure is uncommon and can lead to pressure overload of the right ventricle (RV) and consecutive RV hypertrophy. We present 5 newborns with in utero restrictive DA.
Methods: Restrospective study of newborns with in utero restrictive DA. Charts and echocardiography were reviewed. 3 patients were identified as having a restrictive DA in utero and 2 others were identified because of post natal severe RV hypertrophy with low transcutaneous saturation and absent DA on echocardiography in the first hours after birth.
Results: Pre-natal diagnosis was made between 1 and 6 weeks before birth, all babies were born by ceasarian section because of severe RV hypertrophy and decreased RV function. Post-natal diagnosis was made at time of echocardiography between 1 and 10 hours of life. Clinically all patients had abnormally low transcutaneous saturation (84% [range: 80–88%]) not responding to supplemental O_2. All patients developed respiratory distress needing non invasive or mechanical ventilation. Echocardiography demonstrated in all patients a severe RV hypertrophy (RV free wall 7.3 mm (5.8–8.5 mm), free wall of left ventricle (LV) 3.0 mm [2.5–3.6 mm]), 4 patients presented a small RV chamber with mean end diastolic RV volume of 9 ml/m^2 (5–14 ml/m^2) and one a dilated RV. All patients demonstrated RV diastolic dysfunction with consecutively bidirectional shunt at the foramen ovale level. In 3 patients there was high systolic RV pressure with increased tricuspid regurgitation. In those 3 patients, initial RV systolic function was decreased. DA was either absent or smallish. Treatment ranged from clinical observation to intensive treatment with O_2, NO and sildenafil. Catecholamine infusion was given to sustain RV systolic and diastolic function in 3 patients. All patients survived with regression of RV hypertrophy and of RV dysfunction within months.
Conclusions: Premature closure of the DA can be diagnosed in utero and close monitoring of RV function is mandatory. Depending on the time between DA restriction and birth, post natal evolution could be complicated with need of RV afterload reduction and catecholamines, but once initial adaptation is overpassed RV hypertrophy regression occurs in all patients.

P110

Linear morphea follows blaschko's lines

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Aims: The aetiology of morphea (or localized scleroderma) remains unknown. It has previously been suggested that lesions of linear morphea may follow Blaschko's lines and thus reflect an embryological development. However, the distribution of linear morphea has never been accurately evaluated. We Aimed to identify common patterns of clinical presentation in paediatric patients with linear morphea and to establish whether linear morphea follows the lines of Blaschko.
Methods: A retrospective chart review of 65 children with linear morphea was performed. According to clinical photographs the skin lesions of these patients were plotted onto standardized head and body charts. With the aid of Adobe Illustrator a final figure was produced including an overlay of all individual lesions which was used for comparison with the published lines of Blaschko.
Results: Thirty-four (53%) patients had the en coup de sabre subtype, 27 (41%) presented with linear morphea on the trunk and/or limbs and 4 (6%) children had a combination of the two. In fifty-five (85%) children the skin lesion were confined to one side of the body, showing no preference for either left or right side. By comparing the overlays of all body and head lesions with the original lines of Blaschko there was an excellent correlation.
Conclusions: Our data indicates that linear morphea follows the lines of Blaschko. We hypothesize that in patients with linear morphea susceptible cells are present in a mosaic state and that exposure to some trigger factor may result in the development of this condition.

P111

Triplets' long-term developmental outcome at early school age: severe birth weight discordance displays a major predisposing factor for motor and cognitive impairment.

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Background: The incidence of multiple deliveries has increased since the 80's in western countries, which has contributed to the rise of preterm births and associated perinatal and developmental risks. Nevertheless, only few studies have focused on the long term neurodevelopmental outcome of this population group, despite its importance for perinatal practical advising. It is hypothesized that multiple gestation offspring are at increased risk for adverse long term outcome compared to singletons.
Aim: To assess, at school age, motor and cognitive development of children born from a triplet pregnancy, to compare the outcome to population norms and to investigate the predictive value of specific neonatal risk factors for adverse developmental outcome.
Methods: 65 live-birth newborns from 22 triplet pregnancies were prospectively enrolled since birth. At age 6 years, the Zurich Neuromotor Assessment (ZNA) was administered to test neuromotor skills and cognitive competences were assessed with Kaufmann ABC.
Results: 44% of the subjects were female and 11% small for gestational age (SGA = BW <10th percentile); Mean birth weight (BW) 1661g, mean gestational age (GA) 32.7 weeks, mean socioeconomic status (SES) 6. 83% were conceived by assisted reproductive technologies (ART). Triplets scored significantly lower on the ZNA in all timed components: pure motor ($p < 0.001$), adaptive fine- ($p = 0.001$) and gross-motor ($p < 0.001$) and balance ($p < 0.01$) tasks compared to test reference. The mean Mental Processing Composite of the Kaufman-ABC (94.8, ±10.4 SD, $p < 0.001$), with all subtests of the Sequential Mental Subscale (short term memory, $p < 0.001$), and the Achievement Scale (arithmetic and riddles, $p < 0.01$) was affected. Most of the cognitive and motor outcomes were positively correlated with SES, while sex, GA, BW, SGA status, ART, and birth order, had no significant influence on cognitive achievement and only minor impact on some motor subscales. Weight discordance of >25% at birth (23% of cohort) was associated with significantly decreased cognitive and, to a lesser extent, motor performance.
Conclusions: Triplets have a higher risk of cognitive and motor impairment at early school age compared to the normal population. Importantly, severe weight discordance at birth, rather than birth weight, seems to play a major predisposing factor. Larger study samples and matched control population are needed to better define other independent risk factors in the triplets' development.

P112

Basilar thrombosis in children: a rare but potentially life-threatening problem

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Background: Basilar thrombosis is a rare, but distinct entity of paediatric stroke, causing potentially life-threatening infarctions.

Knowledge of its natural history, best diagnostic and treatment approaches and also outcome is still very limited.
Methods: Data from the Swiss Neuropaediatric Stroke Registry (SNPSR) was scanned for basilar thrombosis diagnosed by brain imaging. The SNPSR is a prospective population-based study, including all children from birth to 16 years living in Switzerland and suffering from stroke.
Results: Since the start of the SNPSR in 2000, only 4 cases of basilar thrombosis (out of 94 reported childhood strokes) were recorded. This corresponds to an incidence of 0.5/1 million children/year. 3 were boys (aged 5;8, 7;6 and 9;7 years, respectively) and one was a teenage girl (aged 13;9 years). At initial presentation, all patients suffered from impaired consciousness, from syncope-like short moments, to sustained somnolence and coma. Other symptoms included headache (in ¾), gait disturbance (¾), vomiting (¾), slurred speech (¾), and neuropsychological symptoms such as mood swings (¼) or agitation (¼). Possible triggering factors were preceding viral infections in two cases. Identified potential risk factors were protein S deficiencies in two cases, and protein C reduction of unknown clinical significance in the other two cases. Delay from onset of symptoms to diagnosis was 12-48 hours. Treatment varied from one patient to the other and consisted in (systemic or intraarterial) thrombolysis and/or heparinisation and/or aspirin. Data on outcome are still preliminary: Despite the life-threatening initial presentation, substantial recovery can be observed. Clinical outcome showed mild to moderate sequelae with residual motor, cerebellar and/or neuropsychological problems.
Conclusions: Basilar thrombosis in children is a rare but serious condition. Because of its largely non-specific clinical features, reaching diagnosis can be very challenging. Moreover, delay in treatment initiation might be prejudicial for the neurological and neuropsychological outcome. Better knowledge of this condition should help us reach the diagnosis faster, thus reduce the still considerable "door-to-table"-time and potentially improve the outcome both in terms of revascularization after thrombolysis and functional recovery.

P113

Neonatal listeriosis, still a burden

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Introduction: Listeriosis is relatively rare and occurs primarily in newborn infants, elderly patients, and patients who are immunocompromised. In neonates, infection with Listeria monocytogenes can present as a life-threatening illness. As in group B streptococcal infections, listeriosis can manifest as early-onset disease with sepsis and/or meningitis (mortality rate of 20–30%, significantly higher in preterm infants) or late-onset disease manifesting primarily as meningitis.
Aim: To present the case of a preterm infant with early-onset neonatal listeriosis and unusual pulmonary complications.
Case report: This male infant was delivered by CS at 26 5/7 weeks of gestation because of suspected amnion infection syndrome following an otherwise unremarkable pregnancy. The patient required respiratory support with HFOV and iNO and circulatory support with inotropes and vasoactive agents. There was a marked maculopapular exanthema on the trunk. Gram stain of a tracheal aspirate revealed gram positive rods, later identified as Listeria monocytogenes which also grew from blood cultures and both sides of the placenta. Amoxicillin/gentamycin was administered for three weeks. The hospital course was complicated by the development of pneumothoraces and extensive bilateral pneumatoceles as well as PIVH grade III. The patient was discharged at 41 3/7 weeks corrected gestational age. On his first developmental follow-up at a corrected age of three months, only subtle neurologic abnormalites were noted.
Conclusions: Neonatal listeriosis is still a non-negligible cause of infection in preterm and term newborns. A disseminated rash with small, pale, granulomatous nodules is characteristic and should alert the clinician of the possibility of neonatal listeriosis. Pneumonia is commonly present (50%) and, as demonstrated by our case, can lead to significant complications. Ampicillin in combination with an aminoglycoside such as gentamicin is the therapy of choice. Listeria is not susceptible to cephalosporins of any generation.

P114

Invasive fungal infections in the picu

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Introduction: Pediatric invasive fungal infections (IFI) yield high mortality. The Aim of the study was to identify risk factors, review diagnostic Methods of fungus identification and antifungal treatment.
Methods: A retrospective study over an 87 month period reviewed all pediatric intensive care unit (PICU) patients with IFI, defined as cultural or histological agent identification in a sterile body tissue. Success pertaining to antifungal treatment was analyzed respective to underlying disease and identified fungus.
Results: Eighteen children were identified yielding an incidence of 22 / 10'000 patients accounting for 0.23% of our PICU population. Half showed positive fungal blood cultures, whereas 39% were diagnosed histologically (all aspergillus species). Invasive infections with aspergillus (IA) and invasive candida (IC) were equally distributed. Two thirds of patients were immunocompromised, the majority of iatrogenic origin. The IFI attributable mortality rate was 50%, with dying patients surviving an average of 5 days after IFI diagnosis. Survival was 42% in immunocompromised patients and 67% among those without known or iatrogenically induced immunodeficiency. Fifty three percent of the treated patients survived IFI. Preemptive treatment and modification of antifungal therapy were based on clinical response and did not improve outcome. Immunosuppresion and IA were the most relevant risk factors.
Conclusions: IA and IC show comparable incidence which is a function of diagnostic enforcement. Immunosuppression and IA pose the most relevant risk with respect to morbidity and mortality. Immunocompromised patients are more likely to contract IA and less likely to be diagnosed early. IA is likely to be missed if not investigated histologically. If IFI is presumed in an immunocompromised patient, a biopsy of the suspected organ should be considered.

P115

Management of an animal bite of unknown origin: a case report

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Aim: The majority of animal bites are inflicted by mammalians such as dogs (80%), cats (10%), rodents and humans (3% each). Other animals are rarely involved or not identified at all. Here we discuss the management of a patient presenting with a bite of unverified origin, considering that medical literature does not provide official guidelines on this issue.
Case description: A teenager in rural north-western Switzerland presented with two bites in his left lower leg, possibly snake bites. One hour after the incident, he stated having stepped on a tail-like object in the bushes of his home garden without being able to identify the animal. On examination, no systemic or local symptoms were found except two deep, paired slit-formed puncture wounds on the left lateral sura and a superficial scratch mark. Following local desinfection, intravenous amoxicillin/clavulanate was given; due to the unclear vaccination status, active and passive tetanus inoculation was administered. Mainly since rabies is extremely rare in that area, specific vaccination was not recommended by experts. The patient was clinically monitored for uneventful 24 hours and discharged from hospital care with antibiotic treatment to be continued for an additional 6 days. So far, veterinarians and zoologists could not identify any animal that would match the bite pattern observed.
Conclusions: Bites of unidentified origin should always be treated as those of a possibly venomous snake or of a mammalian carrying rabies, even in instances deemed unlikely, since imported and escaped, possibly illegal animals can never be excluded. The bite pattern is rarely helpful for identifying the biting species, and the history of the accident is notoriously unreliable for their identification. Therefore, retrospectively, we would vaccinate the patient against rabies. In keeping with published recommendations and our findings, we endorse the following steps: 1st evaluation of cardiovascular state and concomitant injuries (vessels, nerves, tendons, bones, joints); 2nd meticulous desinfection of the wound and appropriate débridement; 3rd primary closure except high risk wounds and treatment delay >24 h; 4th prophylactic antibiotics, usually amoxicillin/clavulanate; 5th immobilization/elevation of extremities; 6th tetanus vaccination, if indicated, and 7th, compulsory rabies vaccination; 8th observation for 24 hours in case envenomation leads to severe systemic or local symptoms.

P116

Absence status epilepticus as a first manifestation of an absence seizure disorder

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Introduction: Absence status epilepticus (ASE) is a form of nonconvulsive status epilepticus occurring in all age groups. Precise epidemiological data are scarce, and ASE is likely to be underdiagnosed. In childhood, it is observed almost exclusively in individuals with a known idiopathic generalized epilepsy disorder

(Korff CM, 2007). We report on a boy presenting with a mildly decreased state of consciousness, revealing as an ASE, as a first manifestation of absence epilepsy.
Case: A 13 year old young man, treated with Ritalin for an ADHD, otherwise healthy, presented with a mildly reduced state of consciousness, mnestic disturbances and repeated vomiting. After having ruled out intracranial mass lesion, metabolic disturbances, systemic illnesses, toxins and infection, an EEG performed 24 hours after the onset of symptoms showed a continuous discharge of generalised 3 Hz spike waves, revealing the diagnosis of an absence status epilepticus. Successful treatment was started with i.v. lorazepam, and continued with valproic acid. The next day, our patient could be discharged in an unremarkable clinical state, showing only scarce epileptic activity on his EEG.
Discussion: Nonconvulsive status epilepticus is one of the aetiologies to be considered in the differential diagnosis of impaired consciousness, even if mild. Even though unusual, it does occur in children with no history of an idiopathic generalised epilepsy disorder. Diagnosis is readily established using a standard EEG analysis. ASE usually responds well to anticonvulsive treatment with benzodiazepines or valproic acid.

P117

Cyclic electrographic seizures in childhood

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Introduction: Cyclic electrographic seizures are a form of status epilepticus with seizures recurring at relatively constant intervals from seconds or minutes (Friedman et al. 2007). Little is known about the occurrence of this phenomenon in children. We worked up our experience with cyclic seizures gained in our institution over a period of 12 years.
Methods: We retrospectively reviewed the database of all digitised EEG recordings performed in our tertiary paediatric care centre from January 1995 until December 2007. All recordings showing at least 5 seizures were analysed using compressed spectral array (CSA). Those showing seizures which recurred at relatively constant intervals were included in the study. We searched the patients' files for age, sex, underlying aetiology, whether an epilepsy disorder had been known, and outcome. Antiepileptic medication before and during the EEG was noted, and whether it showed an effect. The seizure discharges were analysed regarding duration, localisation, and intervals between discharges.
Results: 29 patients were included. Median age was 10 months (range 1 day to 16.9 years). 55% were female. The most common aetiologies were acute neonatal hypoxic ischaemic brain injury, acute inflammatory diseases of the central nervous system and intracranial haemorrhages. In a considerable part of patients the aetiology remained unknown. Outcome was normal in 1 patient. 4 patients died. The remaining suffered from epilepsy, neurological and/or cognitive deficits.
Conclusions: Cyclic electrographic seizures are not uncommon in childhood and are easily detected by CSA. In our patients, they occurred predominantly in neonates and/or in the context of an acute disease. Outcome remained unfavourable in most patients. The underlying pathophysiology is unknown. Further studies are needed in order to better understand the phenomenon of cyclic seizures in children.

P118

Lemierre syndrome: a prompt diagnosis and a favourable outcome. Case report

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Introduction: We report a case of classical Lemierre syndrome, the second treated in our Pediatric Unit in 2 years, in which a prompt diagnosis based on the previous experience is associated to a very good outcome.
Discussion: An otherwise healthy 17 years old girl presented to our emergency room with one week-persistent fever, pharingitis, headache. Since a few days she developped pain in the left cervical region and in the left hypochondrium. Phlogistic blood values were elevated. A thoracic radiography was still normal and an abdominal ultrasound scan revealed a hepatosplenomegaly. Considering the septic condition an empirical antibiotic therapy with Ceftriazone was started. Nonetheless clinical conditions of our patient worsened after 2 days with dyspnea, tachypnea and persistent fever. A thoracic X-ray investigation showed then clearly bilateral pulmonary infiltrates and an important left pleural effusion. The presumptive diagnosis of a Lemierre Syndrome was confirmed the third day of hospitalisation with a Doppler Scan of the left cervical region wich revealed a left jugular vein thrombosis. A MRI study showed the thrombus extending in left jugular vein until the base of the brain. We added Clindamycin to the antibiotic therapy in the Aim to cover the classical etiologic agent (Fusobacterium Necrophorum – a strict anaerobic bacterium); in the following few days a thoracotomy was necessary to void a massive effusion from the left lung. Thereafter the clinical conditions of our patient improved: the fever subsided and the respiratory distress disappeared. After 3 weeks of endovenous antibiotical treatment , the girl was discharged with oral antibiotics for further 3 weeks. Blood culture remained negative.
Conclusions. Early diagnosis and prolonged antibiotic therapy can further a positive outcome in Lemierre syndrome with no necessity of anticoagulation. Even with negative blood culture, the clinical presentation and imaging findings play a main role in the early recognition of this syndrome.

P119

Autologous human bite

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Background: Human and animal bite wounds can cause infections with otherwise unusual bacteria. Effective management requires rapid medical evaluation, may necessitate surgical intervention and antibiotic treatment. Close clinical follow-up is recommended to minimize the risk of late complications.
Case report: A 10-year-old previously healthy boy bite with his teeth in his right knee while jumping on a trampoline-like air-inflated mattress. He presented in the emergency room with a superficial lesion on his right knee. After inspection and disinfection, he was empirically treated with oral amoxicillin-clavulanic acid, his leg was immobilised. Two days later the boy showed a painful knee with joint effusion and redness. Puncture and rigorous joint irrigation were performed. Opaque joint aspirate revealed >70'000 cells/ l including 83% polynuclear cells. After parenteral administration of amoxicillin-clavulanic acid for 14 days the wound was healed and the boy discharged. The microbiologic work-up of the joint aspirate revealed no microorganisms in the Gram stain, and by culture no growth of bacteria within 72 hours. Eubacterial PCR from joint aspirate detected streptococcal DNA. Original cultures of the joint fluid grew Actimomyces species. 7 days after hospitals discharge the boys' right knee was swollen and painful. Joint puncture revealed clear fluid with 1250 cells/µl and the boy was treated with oral amoxicillin-clavulanic acid for 7 days. Bacterial cultures and eubacterial PCR from second joint aspirate remained sterile. Because of persistent swelling of the knee for two months and a hyperintense lesion in the vastus lateralis muscle by sonography, MRI was completed and was normal. The boy recovered completely. Actimomyces species grown and identified in culture from joint fluid were subjected to PCR and sequencing. The sequences identified Actinomyces odontolyticus. Actinomyces are bacteria of commensal mouth flora, best known to cause cervicofacial, pulmonal or abdominal abscesses. To our knowledge actinomyces arthritis in children is not reported.
Conclusions: Exploration of bite wounds is essential in order to prevent underestimation of their extension. Close clinical follow up allows early detection of complications. Although Actimomyces arthritis in a child is a rarity, following bite injury, and human bites in particular, infections by otherwise unusual microbial agents including contaminating commensal mouth flora have to be considered.

P120

Double intussusception in a 5,5 years old child

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Introduction: The intussusception in the pediatric age is an acute condition, wich can often be resolved conservatively (enema) but can become a surgical emergency needing intervention. The classical triad are acute recurrent abdominal pain, vomiting and blood in the stool. The abdominal pain is colicky with the child becoming quite and confortable between episodes. The idiopatic intussusception (95% of all cases) is the most common cause of intestinal obstruction among children between the age of 3 months to 3 years. In older children as well an intussusception must be considered in the presence of the classical symptoms: in these cases she Results usually from a lead point that is pulled distally by peristaltic activity. The most common lead point are Meckel diverticulum, lymphoma, polyps and other intraluminal patologies.
Case report: An otherwise helthly 5,5 years old child came to the emergency room because of severe abdominal pains wich suddently appeared 2 hours before consultation. The child was in reduced general condition, very suffering, inconsolable and banding forward during the pain attacks. The abdominal pains were of short duration (few minutes) with short pain free intervalls and were accompanied by vomiting and voiding of stool with fresh blood. The clinical suspicion of intussusception was confirmed by abdominal unltrasonography. A gastrografin / air enema evidentiated the ileo – coecal intussusception, wich could be only partially resolved. Therefore an urgent surgical approach was decided. The laparotomy revealed a

double intussusception: an ileo – ileal invaginated intestinal tract was further invagineted through the ileo-coecal valve. The manual devagination revealed an enormous ulcerated Meckel diverticulum as causative lead point. The gut was vital and no resection was needed. The postoperative course was favourable.
Conclusions: Intussusception beyond the typical age (infants and toddlers) present with the same symptomatology but Results usually from a lead point, most common a Meckel diverticulum as in our case. To our knowledge a double intussusception is a very rare event.

P121

Profound hyperbilirubinemia – a rare manifestation of acute epstein barr virus infection in adolescence

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Epstein-Barr virus (EBV) infection is frequently associated with mild hepatitis and hematologic abnormalities. Severe hyperbilirubinemia has been described in association with cholestatic hepatitis and haemolytic anemia in patients with acute EBV infections.
Case report: A 15-year-old previously healthy girl was referred to our hospital with a 10-day history of malaise, fatigue, headache, low grade fever and new onset of jaundice. On physical examination, she was moderately jaundiced with mild tonsillitis, unilateral cervical lymphadenopathy and mild hepatosplenomegaly. Laboratory values revealed a cholestatic jaundice with a total bilirubin of 286 mol/l (direct 133 µmol/l), moderately elevated aminotransferases (ASAT 258 U/l, ALAT 117U/L) and a Coombs positive hemolytic anemia (Hb 78 g/l). Direct antiglobulin test was positive for cold-reactive IgM autoantibodies (Anti-C3d positive, 30°C). Acute EBV infection was confirmed by highly positive EBV early antigen. Other causes of liver disease (infectious, metabolic, toxic, autoimmune) were excluded. In the following days, she became profoundly jaundiced with a maximal total bilirubin of 787 mol/l (direct 471 µmol/l) on day 4 following admission. The molar ratio of unconjugated bilirubin to albumin, was 0.78 (measure for free unconjugated bilirubin). Treatment with corticosteroids and albumin transfusion was commenced and she showed a prompt clinical response with a complete resolution of disease within a few weeks.
Discussion: In rare cases, EBV infection can be complicated by cold agglutinin hemolytic disease and cholestatic hepatitis resulting in extreme hyperbilirubinemia with the potential risk of kernicterus. Although kernicterus has never been reported in acute EBV infection, it has been described in two patients with severe viral hepatitis as well as in patients with inherited glucuronyltransferase deficiency (Crigler-Najjar syndrome). In Crigler-Najjar syndrome, a molar ratio of unconjugated bilirubin to albumin >1 is associated with an increased risk for kernicterus. Treatment consists of supportive measures including corticosteroids, albumin transfusions and, if not sufficient, plasmapheresis.

P122

Orphanet website: a new version

L. D'Amato-Sizonenko

Aims: To better serve its 25,000 daily users, Orphanet, the European information server on rare diseases and orphan drugs, has launched a new version of its website.
Methods: Orphanet (www.orpha.net) provides access to an encyclopaedia of rare diseases and orphan drugs and to a directory of services in 37 countries, including Switzerland (www.orpha-net.ch). It provides information on specialized clinics, clinical laboratories, research activities and patients' organisations.
Results: The new website is a customised portal providing a multitude of information and services all available from the site's homepage, accessible in 5 languages: English, French, German, Italian and Spanish. The portal is fully accessible to visually and physically impaired users. The database of rare diseases and orphan drugs has been enhanced with new epidemiological informations, including mode of inheritance and genetics. Links to related information sources, (Swiss-Prot, GenAtlas or EuroGenTest) are just a click away. A major feature of the update is the addition of a classification scheme that categorises and cross-references any given disease by scientific, medical and genetic criteria. This new service is unique in its kind and will allow access to information from a generic category. Searching for a rare disease by clinical signs has been improved. The orphan drug section has been enriched to provide information on the stage of development for any particular molecule from the moment it receives "European Medicines Agency" orphan designation until its market authorisation in Europe. The website also now provides access to the list of on-going clinical trials by molecule and to all orphan indications of a designated molecule – a service strongly requested by patients. Finally, navigation of the new site has been simplified to adequately guide first-time users – a growing part of the website's visitors as the word of Orphanet spreads. Orphanet Switzerland (www.orpha-net.ch), active since 2001, is in charge of collecting information about the national resources in the field of rare diseases.
Conclusions: The new version of the Orphanet website, the 4th one in its 11 years of existence, positions Orphanet as a real portal, adapted to the diversity of its audience – one-third patients and their relatives and two-thirds health professionals, scientists, teachers and students. Orphanet is one of many organisations throughout Europe dedicated to improving conditions for rare disease patients and their families.

P123

Review of clinical, neurophysiological and magnetic resonance imaging manifestation of adem in 8 pediatric patients

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Aim: Acute disseminated encephalomyelitis (ADEM) is a rare, immune-mediated inflammatory disease in childhood. We describe its clinical characteristics and review them in context of neurophysiological and neuroimaging features.
Methods: A retrospective (2001–2007), observational, single center multi-case report study was conducted. Clinical and neuroimaging features as well as biological markers of 8 patients (aged 5 to 15 years), diagnosed with ADEM were evaluated.
Results: Eight Patients (median age: 10y 3/12m) with ADEM were seen between 2001–2007. There was a male predominance (6/8). Four patients had associated infections and fever, four presented only neurological symptoms. Neurological presentation varied between ataxia (4 patients), unilateral sensory deficit (4 p.), altered level of consciousness (3 p.), dysarthria (3 p.), hemi- or monolimb paresis (1 p.), seizures (2 p.) and diplopia. An infectious aetiology could not be established in the CSF of any of the patients, whereas the serology was positive in five patients: Mycoplasma IgM (2 p.), Enterovirus (3 p.), Parainfluenza III (2 p.), HSV IgM (2 p.), Influenza A (1 p.). Increased CPR levels and oligoclonal banding (CSF) could be each observed in a single patient. Five patients showed pathological EEG findings, marked by a reduced Background rhythm frequency, only one showed focal discharges. On MRI scans two patients presented 2–3 lesions, five patients presented more than three lesions. Lesions were localised in both hemispheres, corpus callosum, thalamus, the basal ganglia, pons, cerebellum and myelon. Four patients received parenteral high-dose steroids over 3–5 days followed by oral steroids. Complete Remission was found in five patients. Three patients showed discrete neurological sequelae. The number and localisation of MRI lesions did not correlate with the severity of acute clinical manifestation. There was no distinct effect of Steroid therapy on neurological outcome.
Conclusions: ADEM is a potentially severe demyelinating disorder that shows a wide variety in clinical manifestation. In our population no correlation between the number and localization of MRI scan lesions and severity of acute clinical manifestation was recognized. There was no difference in recovery (time and outcome) in patients with or without steroid treatment. Total remission seemed to be more likely if lesions were not located in the pons, thalamus or basal ganglia.

P124

Der unabdingbare Blick in die Windel – eine aussergewöhnliche Vulvaveränderung bei einem termingeborenen Mädchen

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Wir berichten über ein 3 Tage altes, am Termin geborenes Mädchen, welches anlässlich der ersten Vorsorgeuntersuchung eine interlabiale, zystische Raumforderung zeigte. Die weissliche Läsion lag in der anterioren Vulvahälfte und hatte eine glatte Oberfäche mit feiner Vaskularisation. Die medial verschobene Urethra und das medio-posteriore Hymen konnten identifiziert werden. Die Miktion war nicht beeinträchtigt. Die Entbindung nach unauffälliger Schangerschaft erfolgte über eine primäre Sectio wegen Beckenendlage. Apgar war 9/10/10 bei 1, 5 und 10 Minuten, Nabelarterien-pH 7.31. Geburtsgewicht war 2790 g (P15), Länge 46 cm (P3), Kopfumfang 35,5 (P80). Der weitere postnatale Verlauf war ebenfalls unauffällig. Wir beschreiben anhand von Illustrationen, die Differentialdiagnose und die entsprechende Diagnostik einer zystischen Raumforderung der Vulva im Neugeborenenalter:
– prolabierte Urethra
– prolabierte ektopische Ureterocele
– Hydro (metro) -colpos
– vaginales (botryoides) Rhabdomyosarkom
– Paraurethralzyste
Eine sorgfältige Körperuntersuchung gehört zum Standart der pädiatrischen Untersuchungen im Säuglingsalter. Bei eindeutigem Befund, können unnötige Zusatzuntersuchungen unterlassen werden und die Eltern beruhigt werden.

P125

A functional constipation with unusual clinical manifestation

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Clinical case: An 8 year and 3 month old girl with known chronic coprostasis and encopresis from the age of 2–3 years. She wears diapers where she leaves the faeces or she dirties her underwear. She does not use the toilet, being scared to get sucked in by it. Status after being hospitalized for constipation and encopresis at the age of 4 years, when a dolichomegasigma had been found. In the following the paediatrician had not been consulted for this problem. The girl has been followed for a long time by a psychiatrist. She presents at our emergency room with worsening abdominal pain and repeated episodes of vomiting with beginning in the preceding night. She isn't able to drink, since 4 days she has not eaten anything. She has not evacuated stools in 7 days, since 3 days she takes a contact laxative based on senna extract, and as well an osmotic laxative. She reaches our emergency room in very diminished general conditions, much afflicted; she has no fever, the abdomen has an impressive aspect, being extremely globose and painful. We suspect a toxic megacolon caused by cronic constipation. An emergency operation is carried out with manual evacuation of the faeces in the rectum and placement of a caecostomy. An x-ray confirms the megacolon. The following course is favourable. The girl progressively begins to feed again. Osmotic laxative treatment and daily lavages of the colon through the stoma. Toilet-training three times per day after the meals to accustom the girl to evacuate the bowels normally. The contacts between the girl and her parents are being limited to some hours a day, to train her to obtain a certain independence and self-responsibility. Slow improvement. An anorectal manometry is carried out which has shown the absence of the rectoanal inhibitory reflex. To exclude an aganglionosis of the colon, biopsies are taken of the distal colon and the caecostomy, with normal results. Following the stoma is being closed. After hospitalization for three months the girl is being discharged from the hospital in good general conditions. The ampulla of the rectum still appears dilated, but less than in the preceding controls. The girl has learned to use the toilet three times a day and evacuates faeces daily. She still takes an osmotic laxative.
Conclusions: The chronic functional constipation in childhood is a frequent problem which, when non treated correctly, can evolve into a dramatic clinical manifestation. The chronic functional constipation is a demanding problem which the paediatrician has to address with constancy.

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Menstrual staphylococcal toxic shock syndrome in a 15-year-old girl: a case report

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Introduction: Menstrual staphylococcal toxic shock syndrome (TSS) is a toxin-mediated systemic disease of acute onset characterized by fever, hypotension, myalgias, rash, multiple-organ failure and late desquamation of hands and feet. It is associated with vaginal colonization with toxin-producing Staphylococcus aureus during menstruation and leads to high morbidity and mortality. Although rare since the chemical composition of tampons has been optimized, menstrual TSS is still observed, primarily in healthy young women. According to the Swiss Federal Statistical Office, between 1995 and 2005 only five women with TSS were recorded.
Case description: A 15-year-old girl presented acutely ill with fever (41 °C), dizziness, confusion, headache, flank pain and vomiting. On clinical examination, she showed a facial rash, ubiquitous maculopapular exanthema, diffuse abdominal pain and myalgias. As 100/µl leukocytes were found in the urine, we suspected pyelonephritis and ceftriaxone (2 g/d i.v.) was started, after which
her clinical condition improved rapidly. Unexpectedly, urine cultures revealed only insignificant growth of S. aureus. Reevaluating the history and symptoms, we found that the patient had recently menstruated and last used and removed a tampon on the day of admission. Therefore we strongly suspected menstrual TSS and subsequently detected toxic shock syndrome toxin (TSST-1) in the girl's urine isolate of S. aureus, which is known to be responsible for most menstrual toxic shock syndromes. Typically, blood cultures remained negative. After four days' treatment with ceftriaxone, the patient was discharged in good condition and was treated with oral clindamycin for ten more days. Two weeks later she showed skin desquamation of hands and feet.
Conclusions: In septic appearing young women, menstrual TSS has to be considered in the differential diagnosis. Quick tampon removal, cardiovascular stabilization and adequate antibiotic therapy are crucial in the management of this life-threatening disease.

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Use, outcome and risk of central venous or arterial lines in neonatology

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Aims: Reported healthcare-associated infection (HAI) rates in neonatal care range up to 9 per 1000 patient days. In particular, preterm and extremely low birth weight (ELBW) infants are at high risk as they require longer care and are more likely to be exposed to invasive devices. Bloodstream infection is the leading HAI among neonates. We observed the incidence of BSI and their association with the type of central venous or arterial line in use.
Methods: A prospective cohort study conducted from 2001 to 2007. Inclusion criteria were all neonates with a birth weight below 1500 g, or any medical device in place, or having undergone any surgical intervention, or in need of parenteral nutrition. All neonates in the study were followed until discharge. BSI and catheter surveillance was carried out by a dedicated infection control nurse using adapted CDC HAI definitions. The Cox hazard proportional model was used for adjusted risk analysis.
Results: A total of 99 BSI (bacteraemia or clinical sepsis) were detected among 1552 neonates with 1928 central lines and a total of 10, 655 catheter days. The median days of catheter use [interquartile range] were 7 [2–10] for silastic catheters (SVC), 3 [1–4] for umbilical artery catheters (UAC), 3 [1–5] for umbilical venous catheters (UVC) and 13 [6–48] for tunnelled venous catheters (TVC). The incidence density (ID) was highest among UAC (13.9 episodes/1000 catheter days), followed by UVC (12.9/1000), SVC (8.6/1000) and TVC (2/1000). The total ID was 9.3/1000 catheter days. Adjusted risk factor analysis showed significant Results for the use of silastic catheters (HR [95% CI]: 2.4 [1.2–4.7], p = 0.01) and ELBW infants (HR [95% CI]: 3.1 [1.7–5.7], p <0.001).
Conclusions: Central venous and arterial lines are of benefit but also a risk, especially for ELBW children. Umbilical catheters show the highest ID, but because catheter duration is highest among SVC, they are the most likely to infect.

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Associations among obesity, blood pressure, and left ventricular mass

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Aims: To measure resting and ambulatory systemic blood pressure (BP) and left ventricular mass (LVM) in pre-pubertal obese and lean children, and to determine their relationships.
Methods: Cross-sectional study including 44 obese and 22 lean pre-pubertal children (mean age 8.8 ± 1.5 years). We measured: casual and 24-hour ambulatory BP; LVM and LVM index (LVMI) by echocardiography; whole body lean tissue and fat mass by DEXA.
Results: Mean 24-hour systolic BP (124.8 ± 14.2 vs. 105.5 ± 8.8 mm Hg), diastolic BP (72.8 ± 7.3 vs. 62.7 ± 3.8 mmHg) and LVMI (36.1 ± 5.8 vs. 30.9 ± 5.7, g/m-2.7) were significantly higher in obese than in lean subjects. Systolic ambulatory hypertension was present in 47.6% of obese children, casual BP missing the diagnosis in 55% of cases. Body fatness, lean tissue mass and 24-hour BP correlated positively with LVMI. When adjusted for body fatness, LVMI was associated with 24-hour systolic BP only (adjusted R2 = 15.9%; p = 0.001).
Conclusions: Ambulatory systemic hypertension and increased LVM appear before puberty in obese children. Left ventricular mass is partially determined by systemic BP; however increased LVM may occur before the development of hypertension. We conclude that prevention and treatment of childhood obesity should be initiated as early as possible to prevent the premature development of hypertension and end-organ damage.

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The adolescent in hospital protagonist of his disease

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Aim: In our hospital we tried to improve the communication between the adolescents and the paediatrician giving to the teen agers admitted into the clinic the possibility of having a physician expert in their problems. Our study wants to examine the efficacy of the counselling to the teen agers during the illness.
Methods: We identified one physician as reference to the teen agers. This physician offered himself to help them giving a service of counselling. The objet of the counselling was to help the adolescent becoming more conscious about himself so that he can lives his problems with more tranqullity. This happened with a direct relationship between the adolescent and his counsellor, without the presence of the parents. The counselling happend during the hospitalization or in the first aid station and then continued with periodic visit after the dismissal. The counselling is composed by few parts: the welcome, the observation, the empathic listening, the personalization of the feelings, the personalization of the sense and the restitution to the patient and then to his family.
Results: The adolescents admitted to the counselling showed a better compliance and recovered sooner than others. They valueted this possibility very important as documented by their firm presence at the following visits after the dismissal.
Conclusions: The counselling let us to understand that every adolescent is unique, everyone is the protagonist of his own story, even during his disease.

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Swiss pediatrician's training needs in promoting and supporting breastfeeding

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Pediatricians play an important role in promoting and supporting breastfeeding.Many actions have been realised in Switzerland leading to an increase in the number of breastfeeding women and duration of breastfeeding. The current obesity epidemic justifies continuing these efforts.Are Swiss pediatricians sufficiently prepared to support breastfeeding? Do they need information or training in this field?
Objectives: To clarify the pediatrician's position in supporting breastfeeding. To identify their needs and the type of information or training desired.
Method: A questionnaire was proposed in 2007 to all Swiss pediatricians (1200, of whom 800 primary care pediatricians) through the website and the journal of the society.
The survey concerned frequency of breastfeeding questions during consultations, the possibility of referring to a lactation consultant, the observed breastfeeding duration, possible difficulties in supporting breastfeeding, information and/or training needs.
Results: 285 pediatricians answered (35,6% primary care pediatricians) with a proportional representation of the 3 linguistic regions, 70% consulting in urban areas, 27% in rural areas. The majority of the pediatricians are often consulted about breastfeeding (›2/3) and meet no difficulties in this field (‹2/3). Among those (›1/3) who sometimes encounter difficulties, 1/4 attribute them to a lack of training. Lactation consultants are available in all areas. The average breastfeeding duration is estimated at 2–4 months. 3/4 of the pediatricians desire information for the promotion of breastfeeding and 2/3 for practical advice, in priority through the journal and the website of the society.
Conclusion: The participation rate of the pediatricians indicate their interest in breastfeeding promotion. Although 2/3 of the Swiss pediatricians often consulted about breastfeeding encounter no difficulties in this field, 3/4 wish to receive further information. This survey confirms the pediatricians' needs for information on breastfeeding and gives indications on the type and the means by which this information could be developed.

The numbers refer to the pages of this supplement.