Annual Joint Meeting of the Swiss Societies for Paediatrics, Child and Adolescent Psychiatry, Paediatric Surgery

Lugano, June 19–21, 2008
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Physical activity patterns in children with cerebral palsy measured by accelerometers: a pilot study
K. Sonderegger1, F. Fugsteg-Büsch2,3, E.D. De Bruin1, R. Möller1, N. Ruch1, A. Meyer-Heim1
1 Institute of Human Movement Sciences and Sport, ETH Zurich; 2 Institute of Sport Sciences, Swiss Federal Office of Sport (BASPO), Mägglingen; 3 Rehabilitation Centre, University Children’s Hospital Zurich, Afdeltum am Albis
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) were 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: 72.6 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 = 0.39, 36.0 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.
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% 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 than those of “late or no T.S” group (spleen size within 1st year of P.S: 92% compared to 54% respectively or 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.

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

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1 Institute of Social and Preventive Medicine, University of Bern; 2 Paediatric Hematology-Oncology Unit, CHUV Lausanne; 3 Dept of Psychology, University of Sheffield, England; 4 Paediatric Hematology-Oncology Unit, University Children’s Hospital Zurich

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 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.

Conclusion: Contrary to recommendations, only a minority of adult survivors of childhood cancer in Switzerland go to regular follow-up visits, 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.
Enteral tube-weaning in infants in Switzerland – a successful interdisciplinary approach of the children's hospital lucerne

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

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 lose their ability to drink and eat. Programs of tube weaning are 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 feeding 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 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

Alcohol intoxication among adolescents: Evaluation of a new Ambulatory consultation

M. Duran1, D. Aladjem1, M. Suchef2, L. Lauer3, M. Alvarez4, M. Callbach1
1 Consultation pour adolescents, Département de l’enfant et l’adolescent, Hôpitaux Universitaires de Genève
2 Service d’abus de substances, Département de psychiatrie, Hôpitaux Universitaires de Genève
3 Service d’endoscopy, Département d’endoscopie, 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 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

<table>
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<th>Item</th>
<th>% of patients</th>
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<td>Learnt something during the consultation</td>
<td>72%</td>
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<tr>
<td>The consultation was useful</td>
<td>60%</td>
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<tr>
<td>The consultation improved the communication</td>
<td>32%</td>
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</table>

Table 2: summary of items about alcohol consumption

<table>
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<th>Item</th>
<th>% of patients</th>
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<tbody>
<tr>
<td>No new alcohol intoxication after the consultation</td>
<td>68%</td>
</tr>
<tr>
<td>• once or twice</td>
<td>16%</td>
</tr>
<tr>
<td>• more than twice</td>
<td>16%</td>
</tr>
</tbody>
</table>

When they answered the questionnaire, none of them think to have an alcohol problem. 40% were affected by social difficulties concerning 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 from it, 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.
Effects of late cord-clamping on cerebral and systemic circulation in premature infants (VLBW)

M. Nelle1, O. Linderkamp2, C. Höcker2.

Effects of late cord-clamping on cerebral and systemic circulation in premature infants (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, ml/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/m²) were estimated. Statistic: unp. t-test.

Results (4 h after birth)

<table>
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<th>Early cord clamping</th>
<th>Late cord clamping</th>
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<td>MBP</td>
<td>36 ± 4</td>
<td>44 ± 7</td>
<td>0.03</td>
</tr>
<tr>
<td>LVO</td>
<td>241 ± 38</td>
<td>245 ± 25</td>
<td>NS</td>
</tr>
<tr>
<td>ACA</td>
<td>0.16 ± 0.03</td>
<td>0.20 ± 0.05</td>
<td>NS</td>
</tr>
<tr>
<td>SVR</td>
<td>123 ± 40</td>
<td>145 ± 30</td>
<td>0.05</td>
</tr>
<tr>
<td>Hct</td>
<td>0.46 ± 0.4</td>
<td>0.55 ± 0.5</td>
<td>0.002</td>
</tr>
<tr>
<td>cerebral HbT</td>
<td>7.4 ± 1.8</td>
<td>11.1 ± 4.1</td>
<td>0.04</td>
</tr>
<tr>
<td>systemic HbT</td>
<td>154 ± 27</td>
<td>181 ± 24</td>
<td>0.05</td>
</tr>
</tbody>
</table>

Conclusions: Late cord-clamping improves MBP, systemic vascular resistance, hemoglobin, systemic and cerebral hemoglobin transport in premature <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).

Lighting conditions differed significantly between the two groups (intervention vs control group: 511.8 lux ± 45.3 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 ± 45.3 vs 31.2 days ± 45.3).

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, wake and content. At age 5 weeks sleep was clustered between midnight and 4 am, 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 (02.06 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.

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


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.
Oral presentations – PSY 1

Date: 19/06/2008
Time: 16:15–17:30
PSY 1-1

Childhood autism: an interdisciplinary approach
P. Weber1, E. Ister1, K. Schnecke2
1 Department of Neuropediatrics, University Children’s Hospital Basel; 2 Office of Child and Adolescents Psychiatry Basel-Land; 3 Clinic of Child and Adolescents Psychiatry, University Psychiatric Clinic, Basel

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 conclusion 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 present a multisteped child with 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 distinctly 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.


Network therapy: a systemic model
R. Trélat1, J. Jouvent2, M. Ostier3
1 Sozialpädiatrisches Zentrum, Children’s Clinic, Kantonsspital Lugano-Viganetto; 2 Department of Social Work, Zurich University of Applied Sciences; 3 Integrierte Psychiatrie Winterthur

Definition: The network therapy includes professionals, family and child; it presents all third parts, gives time to share the experiences and thoughts, and looks for consensus, in respective mediation, coaching, as well framing for multiproblematic and underorganised situations. It limits overreaction and overinvolvement, isolation and helpless, gossip and power positions.

Illustrations: Are illustrated two long term multidisciplinary psychopedagogical guidance networks, one of an anorexic infant, another of a preschooler batteries in the case of a boy, who was born preterm at 28 weeks. In the neurodevelopmental follow up autism was disclosed. Intensive early education and treatment distinctly 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.


Troubles anorexiques en milieu psychiatrique intrahospitalier. Quel sens et quels soins donner aux symptômes dans l’articulation du parcours patient de l’enfant à l’âge adulte?
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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 (N = 455), sent to general practitioners, psychiatrists, psychotherapists, psychologists in private practices, child psychiatrists, 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 a greater area of 220’000 people was 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 teams 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 extramilitar interactions and in daily life activities.
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 bowel 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 preformed spring-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.

**Method:** 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 gradual reintegration of the abdominal content, obviating the need for 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:** 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.
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.

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 OsthesslichKinderklinik we created a multiprofessional management team taking care of the follow up for patients withoesophageal disorders, mainly for the treatment of anastomotic leaks, anastomotic strictures, gastrooesophageal reflux, tracheomalacia, dysmotility, respiratory function, scoliosis and nutritional problems. We would like to present this multidisciplinary team and its management methods to pediatricians and pediatric surgeons 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 life of the patients and their parents.

Methods: 19 patients after oesophageal atresia surgery and 11 with peptic stenosis of the oesophagus 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 specialists from the very beginning of the diagnosis and treatment. The outcome and the quality of life of these patients and their parents depends on the structured organization of such a team. A questionnaire is being created to evaluate the satisfaction of the patients and their parents. Preliminary results should be available within a few weeks.

Hemato logic and subjective changes after subtotal splenectomy in children with hereditary spherocytosis
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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, clinical 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 successful 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. Refilucyte 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 (OPS) were not observed. All children interviewed reported an improvement in their general condition. In the majority of the cases, these statements were affirmed 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.
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 familial 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 computer tomography (CT) of the head at the age of 3 11/12 years was performed to investigate cutan findings on the scalp. It showed completely normal intracranial 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 ventriculoperitoneal shunt was inserted. Postoperative CT-scan of the head showed a decrease in ventricular size and again no further abnormalities. Otherwise neurological 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.

Kohler's disease of the tarsal navicular:

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Case report: A healthy 6 year old boy presented for ataxic gait and limp of the left foot. There was no swelling or bruising, and weight bearing, although painful, was possible. 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 ataxic gait 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 Sudeck’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 further 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 case 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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Modified technique of mesenterico-rex shunt with insufficient length of the jugular vein graft


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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 endoscopy, pre-operative check-up, including retrograde trans-jugal 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-clavicular part to its confluence with subclavian vein. Nevertheless, 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.

Subtrochanteric rotational osteotomy for the treatment of reduced femoral anteversion

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Background: Reduced femoral anteversion, acetabular retroversion and irregularities of the femoral head-neck-offset cause reduced internal rotation of the hip which may lead to femoro-acetabular impingement (FAI) which is nowadays regarded to be a prearthrotic condition of the hip. If the femoral anteversion 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 anteversion.

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
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 (5.8–18.0).

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.

Lattisimus 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. Neoadjuvant 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 cases with large thoracic wall defects needing complex reconstruction, taking the somatic growth potential into account.

Conclusion: We conclude that covering a Gore-Tex Dualmesh patch with a lattisimus 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.
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. Patients were divided in two groups: 56% of patients were taking ERM at re-sting. Furthermore, we investigated clinical efficacy of ERM in addition to VIT.

Results: 43 children (52%) 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 = 0.32). 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 re-sting: 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 who carry and take ERM at re-sting will be an effective adjunct therapy with reduced incidence and severity of systemic allergic reactions to re-stings.

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 and multifactorial. In Switzerland, since 2001 it is advised to women of child-bearing age to take periconceptional folic acid supplementation to reduce the risk of NTD. The aim of this study was to analyse the prevalence of NTD in Switzerland from 2001 to 2007 and to identify possible risk factors.

Methods: Diagnosed cases of spina bifida and anencephaly were identified from the Swiss Pediatric Surveillance Unit from January 2001 to December 2007. Patients were assigned to one of four groups: those with no periconceptional folic acid supplementation, those with periconceptional folic acid supplementation, those with other supplements, and those with no supplement. Additional data such as age, parity, smoking, alcohol consumption, and socio-economic factors were collected. The prevalence of NTD birth defects was determined for each group. Patients were then further subdivided into women with and without at least one nontoxic prenatal ultrasound scan.

Results: Data of 140 newborns and fetuses with NTD were studied. The majority of patients had a mild hypothermia with body temperature below 36.5°C. 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 drank alcohol and 10 boys were found to be alcohol users. 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. A. 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 psychosocial 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.

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 conducted further data on acute alcohol intoxication in children. The aim of the study is to analyse data of inpatients of our hospital with regard to patient numbers, severity of intoxication, medical complications and surrounding socio-economic factors.

Methods: The present study is a retrospective analysis of 160 records of patients admitted to the University Children's Hospital Zürich due to alcohol intoxication during the period 1998–2007.

Results: The gender ratio is nearly balanced: 81 males/ 79 females. The age range is between 1 and 16 years (median 14.63 y; SD 1.44y). In 27% (n = 43) of the children the mental care 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 36.5°C. 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 drank alcohol and 10 boys were found to be alcohol users. 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. A. 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 psychosocial 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.
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 it is necessary to develop a validated and cost-effective cystourethrography (VCUG) for the diagnosis of the vesicoureteral reflux (VUR), thus leading to a number of unnecessary and misleading examinations. The aim 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 fetal renal pelvis diameter ≥ 5 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 dilation with an anterior posterior diameter (APD) ≥ 5 mm or presenting an urateral dilatation of postnatal "puberté en miniature", specialists with expertise in DSD are to be involved immediately after birth. However, surgical expertise in DSD are to be involved immediately after birth. Nevertheless, before abstaining from performing ultrasound at 5 days and 1 month after birth, it remains to be seen if initial diagnostic ultrasound and diagnosis made during puberty, declined to participate.

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 I) with a maximal antepapillary APD of 14 mm or 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). The sensitivity and specificity of the antenatal and postnatal ultrasound for the VUR, we used the ROC curve and found with a cut off level of > 10 a sensitivity of 76.2% and specificity of 92.6% for the antenatal ultrasound and a sensitivity of 90% and specificity of 44.3% for the postnatal ultrasound. Interestingly all children with a severe VUR had an APD 0.10 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 presence of an APD 010 mm or the presence of a ureteral dilation.

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 leucocytocyna between 10/ 2007 and 02/2008 were enrolled. Diagnosis of pyelonephritis was confirmed by positive urine culture. 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.

Results: Children presenting in the emergency department with suspected diagnosis of pyelonephritis based on clinical signs, symptoms and leucocytocyna between 10/ 2007 and 02/2008 were enrolled. Diagnosis of pyelonephritis was confirmed by positive urine culture. 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.

Conclusions: In this series in children from all ages abdominal ultrasound within 3 days from diagnosis of pyelonephritis has no 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.

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 ± 0.53 kg, 95%CI: 0.04-0.50; p = 0.04), and whole body BMD (0.017 ± 0.007 g/cm²; 95%CI: 0.01-0.03; p = 0.03). Bone mineral density did not increase significantly at other sites. LIM increased in the training group (p = 0.001), whereas fat mass increased in the control group (p = 0.04). However, both changes did not reach statistical differences.

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. Improvement in muscle mass and bone mineral density indicates that this intervention allowed to practice regular activities to enhance peak bone mass and prevent osteoporosis later in life.
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.

One year of “maternology”

S. Staub-Ghielmetti, G. Caccia, F. Bianchi, M. Pagliarani, T. Chiaravalloti. Ospedale Beata Vergine, Mendrisio

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 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 reassure 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 its 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.

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 to facilitate decision making for practitioners 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


Is there an ideal age for hypospadias surgery?

D. Weber, M. Landolt, V. Schoenbucher, R. Gobet. Division of Pediatric Urology, University Children’s Hospital Zurich

Purpose: Hypospadias is the most common malformation of the penis. The assumption that hypospadias may affect children’s psychosexual development adversely is the major rationale 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 addressing 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 details. Patients’ histories of hypospadias was assessed. The Pediatric Penile Perception Score, the Gender-Role Questionnaire by Jöntema 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 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.

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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.

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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 (6 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 and one 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 nephrectomy 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.

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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 micron 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 immunohistochromesthesia. 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 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 shows adequate mechanical strength but also allows efficient cell seeding inside and on the matrix resulting in adequate cell proliferation and differentiation.
been demonstrated in several studies and for various illnesses. CDSS

In the past, the benefit of “Clinical Decision Support Systems” (CDSS) has

Y. Nordmann1, R. Guggenheim2.

Mtb infection, but so far the experience in pediatric populations is only limited.

T cell interferon-gamma release assays (TIGRA) are very specific in detecting

may interfere with the tuberculin skin test (TST), thus limiting the usefulness of

BCG vaccination or exposure to environmental mycobacteria

Performance of a whole-blood t cell interferon-gamma

and the ratio of patients alive with their native liver at the age of 5 years.

2) the reduction of age at Kasai operation, as compared to the pre-screening

of this screening program is assessed by the number of SCC received at the

centre, where an anonymous record is hold. The original

SCC is communicated (by fax or a password protected website) to the study

centre, which are very specific in detecting

Mtb infection

are electrical 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 economical

medical record systems in most Swiss private practices within the next few

years, the integration of CDSS tools in such record systems seems itself.

With “E-Asthma” we present such a newly developed CDSS for the diagnosis and
treatment of asthma within the ambulatory setting. “E-Asthma” is an asthma

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 measures,

“E-Asthma” may also provide an important contribution for

improved asthma diagnosis and treatment leading to better care for patients

with asthma.

Introduction:

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 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 were acquired by fax or a password protected website to the study centre, where an anonymous record is held. The original 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 per age and population group received at the study centre, as compared to the national annual birth number. Efficacy is assessed by 1) the ratio 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.

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

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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 in a multifaceted approach.

Results: Questionnaires were returned by 541 paediatricians (58%) in 2001, and 639 (64%) 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 all public and private 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.
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 = 58, 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 co-variate.

**Results:** Total daily physical activity was significantly lower in JIA (SO4 ± SD2 vs. 740 ± 835 count/minute, p = 0.012) and in OB (606 ± 274, p = 0.047) compared to HC. Physical activity was not different in T1DM compared to JA 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 %; JIA: 73.9 %; 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 VO2 max than HC (46.18). Obese children had also lower VO2 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 children; 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.

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

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In adults, 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 <0.001). 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.
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 administered 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.
How is life with bladder exstrophy? A long-term follow-up study (38–69 years)

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Purpose: To describe 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 16% are sterile. Patients have no or only mild clinical 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 (still) majority of our patients have a normal renal function and a well functioning ureterosigmoidostomy. This is quite astonishing, given the fact that the (small) majority of our patients have a normal renal function and a well functioning ureterosigmoidostomy. This is quite astonishing, given the fact that

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

P. Ramseyer, M. Duboli, B.J. Meyrat, P. Frey. CHUV Lausanne

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 sympathectomy and approximation of the pelvic arch that is impossible to achieve. After surgical exploration of the pelvis in the patient with exstrophy-epispadias complex, anterior osteotomy of the superior and inferior rami of the pubis 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.

Results: 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 16.3 years (38–69 years)

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

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Aims: This retrospective study analyses 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 ranging 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 77% preoperatively and in 76% at follow-up.

Conclusions: For high-grade vesico-ureteral reflux re-implantation surgery remains gociculated. Several studies showed that 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.
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 initial radiographic evidence of ductal disruption, we 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 on two cases (5 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 lesion with beginning pseudocyst formation. Patients were immediately started on TPN and Octreotide. As soon as the overseas ordered Pigtails 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 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 9. In patient 1 the stent was removed as day surgery after 4 weeks. On follow-up 4 months after 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 complications and shortened hospital stay as well as the duration on parenteral nutrition.

Laparoscopic gastropathy 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 gastropathy in children with gastric volvulus.

Methods: We present the cases of all children with gastric volvulus operated on by the senior author (JL) and his team at the University hospital of Lausanne between 1998 and 2007. A consecutive sample of 12 patients (11 boys and 1 girl) were identified.

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 gastropathy was performed with an 8 mm H2 CO3-pompy to avoid bowel distension, 2 5 mm ports and a 4 or 5 mm telescope. Our technique include 3 steps: 1) esophagogastropathy, 2) pneufundopaxy and 3) anterior gastropathy. Average time of surgery was 60 minutes. One conversion was performed. Follow-up ranged from 1 month to 7 months. To date, all patients are free of symptoms.

Conclusion: Our technique of laparoscopic gastropathy is a good option in children with gastric volvulus
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 disorders 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 genetic causes of common and rare diseases. 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 Genetics?

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 following situations: when there is a severe or progressive DD, a positive family history, a child with dysmorphic features, increases 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, abnormal severe reactions to infections or metabolic stress, unexplained laboratory values, exceptional history of miscarriages or recurrent miscarriages of the mother are also possible causes of human biologic variation. For the paediatricians in the changing face of Paediatrics by identifying an increasing number of causes of human biologic variation. For the paediatricians in the forthcoming situations: when there is a possibility for genetic disorders. We recommend a referral to the Medical Geneticist in the following situations: when there is a severe or progressive DD, a positive family history, a child with dysmorphic features, increases 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, abnormal severe reactions to infections or metabolic stress, unexplained laboratory values, exceptional history of miscarriages or recurrent miscarriages of the mother are also possible causes of human biologic variation. For the paediatricians in the changing face of Paediatrics by identifying an increasing number of causes of human biologic variation. For the paediatricians in the

Discussion: The Personal Identification of an individual is accurate 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 implications. 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 for 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.

References

Virtual reality (vr) based paediatric interactive therapy system (pits) for enhancement of motor learning in children with congenital 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 (Rehabilitation 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-Barre-syndrome, peripheral neuropathy, 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 tele-rehabilitation. 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.

Safety and efficacy of paediatric outpatient radiofrequency catheter ablation

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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 immunosuppressive and immunomodulatory agents. 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.

References

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

D. Wilke1, K. Eng2, L. Holper1, E. Chevrier1, D. Kipfer1, A. Meyer-Heim1, 1 Rehabilitation Centre, Afinity am Albis, University Children’s Hospital Zurich; 2 Institute of Neuroinformatics, University ETH Zurich

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 (Rehabilitation 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-Barre-syndrome, peripheral neuropathy, 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 tele-rehabilitation. 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.
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. 0.009), 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 resp. the worse 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.

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


Objectives: The Iron Deficiency Syndrome (IDS) as a preliminary stage of the Iron Deficiency Anemia (IDA) can be difficult to recognise. The cardinal symptoms are for instance tiredness, depressive mood, concentration deficit, interferences in attention and retardation. 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. In Ferritin values were 50 ng/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 differentially-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 symptoms of ADHD and children who received a Methylphenidate-therapy as a preliminary stage of the iron deficiency syndrome (IDS) as a possible comorbidity after an inefficient medicinal methylphenidate-therapy in children diagnosed with an attention deficit hyperactivity disorder (adhd)
among which group B Streptococcal ventriculitis is an insidious and Conclusions: For four decades the molecular basis of HIES has remained elusive. Here we show that dominant-negative mutations in the gene of HIES will facilitate early and definitive diagnosis as well as treatment, hopefully leading to the prevention of serious infectious complications and sequelae.

A 16.7 mb deletion on 18q21.2-q22.2 in a patient with dysmalfication, 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, strabismus, atrial or stenotic external auditory canals and genito-urinary malformations are features that are most commonly reported. We report on a 14 months old boy with moderate delay of psychomotor development, muscular hypotonia, tapering fingers, short feet, broad thumbs 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 neurometabolic investigations showed normal results. Cyto genetic analysis revealed an unbalanced de novo translocation t(18;16)(q22.1;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 18q12.1 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 the future, 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.

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 insufficiently 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 Streptococcal 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 Streptococcal 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 diagnoses, among which group B Streptococcal ventriculitis is an insidious and rare cause.

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-consanguineous 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.
Electrocardiographic evaluation was inconclusive. 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 conditions 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.

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 particular haematomata and petechiae. An infection could be excluded. The genotypy 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 x109/L (normal range: 150-450 x109/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 20 x 109/L. Splenomegaly was detected and anaemia had shown 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.

Human donor milk banking in Switzerland
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. Qualify 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 donating 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.

Case report: intra-arterial lysis of an acute thrombotic occlusion of the right middle 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 MCA territory (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 increased serum creatinine were noted. Elevated angiotensin II levels confirmed the diagnosis of intravascular hypertension. 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 intracavalral occlusion in segment 1. Intra-arterial thrombolysis using recombinant tissue type plasminogen activator (t-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 also normal. The further diagnostic work-up remained normal.

Discussion: The incidence of stroke in childhood is 1.8 per 100000 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 angioplasty 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.

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

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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 CHIL 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: This is 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 particularly important among this population characterized by a high incidence of overweight and obesity.

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 headaches, 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 large right temporal atrophy. A 12-year old boy presented with seizures (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 large right temporal atrophy. A 12-year old boy presented with seizures.

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.

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 anaemia with asymptomatic proteinuria. Further symptoms include failure to thrive (FTT), neurological deficits and recurrent infections. Mutations in two different genes (cubulin and amnionless gene) are known to cause a defect of the ileal vitamin B12 receptor, leading to selective vitamin B12 malabsorption, requiring lifelong vitamin B12 therapy.

Case report: A 25 months old boy was referred with fatigue, diarrhoea, vomiting, aphtous 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 anaemia (hemoglobin 74g/l, MCV 113fl, MCH 41 pg), neutropenia and thrombocytopenia (83 G/l). Protein C was present. Vitamin B12 levels were not measurable (<33 pmol/l). Tests for
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 paediatric patients with T1DM.

Methods: A questionnaire was distributed among patients with T1DM at three different settings during three consecutive months. The questionnaire consisted of 20 items asking about intake 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) c/o .Mied to ever have used CM to treat their diabetes. The treatments were cinnamon (14; zinc (7), and (2), DHEA (2), craniosacral therapy (1), Pong and (1) and Chinese quantum method (1). The primary goal as well as the primary achievement were improvement of general health—being followed by alleviation of symptoms. 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.

Neonatal hypoglycemic – don’t forget the hormones!

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Background: We report an unusual but potentially life threatening case 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, microopenia and left inguinal testis. Laboratory investigations revealed hyperkalemia, hyperuricemia and hyperbilirubinemia. Congenital hyperaldosteronism (CAH) was suspected and treatment started with mineralocorticoids and glucocorticoids. Further he developed direct hypobulinemia. 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. Subsequent MRI imaging revealed hypoplastic adrenal glands. Besides the pathologic pituitary- adenal axis, the child showed unmeasurable IGFI 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 fusion of metaphyses with consequent gigantism, osteoporosis and metabolic syndrome in both sexes.

Discussion: For the neonatologist, the combination of severe congenital endocrine disorders and hypoglycemia is a diagnostic challenge. The results of neonatal screening tests are often misleading, especially in syndromes with adrenal insufficiency. Radiological investigations are helpful but not sufficient. Hypoplastic adrenal glands are suggestive of multiple pituitary hormone deficiencies. Moreover, the diagnosis of congenital hyperinsulinism should be considered in neonates with hypoglycemia and other symptoms such as weight loss, failure to thrive and hypoglycemia. 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 pituitary development. Hypoplastic adrenal glands were reported in patients with congenital adrenal hyperplasia, 21-hydroxylase deficiency, 3ß-hydroxysteroid dehydrogenase deficiency and mitochondrial defects. In our patient, other causes for hypoglycemia could be excluded. The patient’s history suggested a diagnosis of multiple pituitary hormone deficiencies. In hereditary disorders of hGH, prolactin, ADH, ACTH, cortisol, gonadotropin and insulin synthesis, the pituitary gland is composed of several cell types. In hereditary hypoplasia of the pituitary, the anterior pituitary becomes underdeveloped, resulting in severe dwarfism.

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 good, there are controversies with regard to the initial course and subsequent management. In particular, the role of antiarrhythmic therapy is controversial and may be helpful in selected cases. The primary goal of this study was to analyze the features associated with resistant SVT in that specific age group.

Background: Most episodes of supraventricular tachycardia (SVT) in children occur in the first few months of life. Although the prognosis is generally good, there are controversies with regard to the initial course and subsequent management. In particular, the role of antiarrhythmic therapy is controversial and may be helpful in selected cases. The primary goal of this study was 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 treatment. 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.

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 on a case where hyperprolactinemia led to the diagnosis of a pituitary tumour.

Case: A 14 year old girl presented with recurrent episodes of vertigo, headache, nausea and vomiting, 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 orthostatic test. Consultation of a pediatric psychiatry 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 examination revealed a medium sized tender cystic mass in the left ovary. Ultrasound was inconspicuous. Hormonal analysis showed normal values for estradiol, LH, FSH, thyroid parameters and growth factors, but a markedly elevated prolactin (3063 mIU/ml, n < 225), which was confirmed by 2 further measurements (5720 mIU/ml, 3977 mIU/ml). MRI of the pituitary revealed an adenoma of 1.5 x 0.8 mm. The patient was referred to neurosurgery for transphenoidal resection.

Discussion: Even if prolactinomas are very rare in childhood and adolescence they are often ascribed to psychosocial stress. The pediatrician is challenged to detect those patients with an underlying disease.

P23

Smoking behaviour among Swiss childhood cancer survivors

Purpose: To describe smoking behaviour among Swiss 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 22% 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 haemophilic patients.

Case report: We report on an 11-year-old boy with severe haemophilia A, who presented with acute onset of intense thoracolumbar back pain. A trauma was initially denied. Subsequently, the patient reported a fall 10 days before where 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 revealed a slight left sided weakness without neurological deficits. A factor replacement therapy was immediately started before imaging was obtained. Subsequently, magnetic resonance imaging (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 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 diagnosing 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 erythematous 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. Nikolay 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 intradermal, sub-coneal crivage, with no inflammatory cells. The patient received antibiotics and intravenous Fluocxacillin and Clindamycin. No new blisters occurred after 48 hours. He was discharged after 5 days. Intra-nasal mupirocin was prescribed for stomatitis. Prophylaxis of household contacts. Conclusions: Our case shows the potential of exfoliatine producing Staphylococcus aureus to cause a severe blistering disease in young children. The different provocation includes topical or systemic necrosis and epidermolysis bullosa. Timely diagnosis, usually requiring a skin biopsy, is important for optimal management.

Cardiovascular findings in turner syndrome: II. Not only the aortic arch
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Background: Turner Syndrome (TS) occurs with a prevalence of 1 per 2500 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 venous connection (PAPVC), consisting of one or more pulmonary veins draining into the right atrium, leading to a volume overload of the right ventricle and right heart failure, 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 3-year-old girl, was diagnosed of TS because of failure to growth. Screening echocardiography detected dilated right-sided cardiac structures and venous anomaly, 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.

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 the mandible and may result in an asymmetric lower jaw or in general.

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 examination and after IAS included pain, tenderness and maximal mouth opening. After a minimum interval of 3 weeks after IAS control MRi was performed. The amount of joint fluid was quantified and correlation of intravenous Fluocxacillin and Clindamycin was 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 significant (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 inflammatory changes 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.

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 adult cardiovascular system problems 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 matched children, born at term with appropriate weight for gestational age were used (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 weight (p < 0.01). Twenty-four out of the 35 children (69%) were salt sensitive. Salt sensitivity was present in 37% and 47% of all low birth weight and small for gestational age children, respectively. Salt sensitivity was significantly correlated with fasting and insulin concentrations (r2 = 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.

Acute renal failure in a neonate after acyclovir overdose.
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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 neurological 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 temperature of 38.6 °C and an increase of six 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.

Agreement 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 (actigraphy, diary, and questionnaire). Do parents accurately report on their child's sleep? How well do actigraphy, diary and questionnaire data agree? Can these Methods interchanged? 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-selected group of healthy children. 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. The study was performed in 1999.

Methods: Cross-sectional study of 50 kindergarten children, age 4 to 7 years, including data from sleep schedules (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.

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 to the need for recommendations to this association. In Switzerland 5 cases of suspected association between isotretinoin use and IBD were registered by Swissmedic (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 diarrhoea, 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 vermiiform 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.

Conclusions: This case of probable isotretinoin-induced transient ileitis suggests that patients presenting with acute abdomen or symptoms of IBD should be asked about the 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.

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 meninengis 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 (PTT > 180 sec) an operation was not performed. Until a severe FXD was proven (FXD activity only 1%) the patient was managed with fresh frozen plasma (FFP) and later with Prolonged complex (PCC) and FIX Behring concentrates. The patient was switched to a weekly prophylactic provision of factor IX Behring. He was discharged without apparent neurological deficit.

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

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Introduction: Continuous glucose monitoring (CGMS) has been increasingly used to understand the interaction between PA and glycemia.

Aim: To examine the effect of physical exercise on overnight glycemia 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 in all patients suffering from hypoglycaemic events during PA. In contrast, PA and hypoglycaemic events did not correlate with Hba1C and any other data of peak, duration of hypoglycaemia.

Conclusions: 1) A diabetic patient regularly suffering from hypoglycaemia is more prone to experience hypoglycaemic events following PA. 2) These data imply the patients with T1DM have to be even more careful about their blood sugar, insulin doses as well as caloric intake following PA.

Macrolecephaly 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 reversible macrocephaly is generated by transient normal pulmonary vascular pressure and resistance. In the following years, the pulmonary vein stenosis, the head growth slowly increased from birth 36 cm (P75) to 43 cm (P50–75) at the time of the surgical relief of the pulmonary vein stenosis, the head circumference measured 50.5 cm (P97) at the age of 24 months.

Haematodynamic evaluation at that time identified an elevated pulmonary vascular resistance and normal pulmonary vascular pressure. In the following months, the pulmonary vascular resistance slowly normalized, and the head circumference reached 49.8 cm (>P97, +3SD) at the age of 16 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 resistance to the cerebral venous 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 with T1DM have to be easy to perform and may identify patients with unfavorable haemodynamics.
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 that even during this period of life we do not have to forget other vasculitis like Wegener’s granulomatosis.

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

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Background: HUS, the triad of microangiopathic hemolytic anemia, 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 diarrheas for 4 days. There were no epidemiological risk factors for Shiga toxin producing E. coli (STEC). At admission haemolytic anemia, thrombocytopenia, and impaired renal function were present. Stool examination demonstrated STEC. Sonography showed heightened echogenicity of the kidneys and renal edema of the colon. 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). PD was stopped whereas treatment 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. For 6 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 stools without enzyme substitution.

Conclusions: In addition to causing renal failure in children, HUS can 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.

Severeencephalopathy in an infant due to subclinical pernicious anemia

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Introduction: Developmental regression in infants can have various causes including severe neurometabolic disorders. Vitamin B12 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 megaloblastic anemia (Hb 90 g/l, MCV 94 fl, MCH 33 pg) and serum vitamin B12 was markedly 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 B12 deficiency can cause irreversible neurologic symptoms in infancy including encephalopathy, seizures, and developmental regression. Vitamin B12 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.

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 not present at birth, are multiple, and present perinatally, and are of hemodynamic significance only in few patients. Other neonatal findings are cutaneous white spots and subependymal nodules and/or cortical tubers. However, angiolipobroma, epilepsy, and mental retardation appear only later.

Conclusions: Multiple cardiac tumors detected perinatally 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.

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, Blanchetti MG. Effects of antihypertensive drugs on blood pressure and proteinuria in childhood. J Hypertens 2007;25:2370-6.

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

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1 Institute of Social and Preventive Medicine, University of Lausanne; 2 Swiss Federal Office of Public Health

Aims: To describe overweight or eating disorders in primary care consultations of Swiss children and 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: There were 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 presented 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 the non-accidental skin bruising was initially made in some children. The children spontaneously recovered without sequelae within 2 to 60, median 10 days. Conclucy: 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.

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 excision of 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 fibular 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-anerysm 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.

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

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Background: Vaginal voiding secondary to urethral-vaginal reflux is an under-recognized cause of daytime urinary leakage in girls. The history of these girls is very distinct: 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 selected according to 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 6.5 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 tender 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.
"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 to proceed with 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 severe 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 he was listed for cardiac transplantation after 12 years old male patient with dilated cardiomyopathy (SM) in patients with cystic fibrosis (CF)

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Aims: Inhibition of 7% hypertonic saline (~1 M NaCl) is one of the therapeutic modalities Aimed at improving mucus clearance from the CF lung. While inhibition 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 intermittent or continuous 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 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 at least 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.
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 TNALP 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 8 10/12 y, respectively.

Conclusions: Hypophosphatasia is an uncommon disorder with a highly variable clinical expression comprising one of the most severe forms, milder variants can easily be overlooked. Premature loss of teeth should raise the suspicion of early onset periodontitis, syndromic disorders such as Jeune-Tommasello-Richet syndrome (MIM 208750), lacrimoauriculodental 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 tooth 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.

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 measures were normalized for gender and grade. Regular sports club participation was defined as participation of at least once a week.

Results: There was no association between sports club participation and fitness-components was not found in overweight children, and in children from overweight parents and migrant families. This study was supported by the Federal council of Sports, Magglingen, Switzerland.

Discussion: Exclusion 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 TNALP 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 8 10/12 y, respectively.

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Cerebral folate deficiency in a child with alpers-syndrom: F. Hoegger1, E. Proietti1, P. Schlaeppy1,2, B. Perret1,3, D. Logchamps4, 1 Département de pédiatrie, Hôpital Neuchâtelois, Neuchâtel; 2 Pédiatre, Peseux, Neuchâtel; 3 Pédiatre, Marin, Neuchâtel; 4 Département de Pédiatrie, CHUV, Lausanne

Introduction: Ear, nose and throat (ENT) infections are common in children with Alpers-Syndrom. Despite an initial favorable evolution, he had subsequent episodes of pan-sinusitis and a left transverse sinus thrombosis extending into the superior sagittal vein. He was treated with IV amikacine and ceftriaxone for 12 days followed with another 2 weeks of oral amoxi-clone, and anti-coagulation with low molecular weight heparin (LMWH).

Case 2: this 19-month-old female initially exhibited a bilateral purulent otitis media and upper eyelid tumefaction, treated with oral Amoxicillin (50 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 (Leniere 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 3 weeks. A repeat head MRI showed no progression.

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.

Cerebral folate deficiency 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 1/2 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 manic-myoclonic convulsions the girl received anticonvulsant therapy could be considerably reduced.

Discussion: The analysis of folate metabolites in cerebrospinal fluid is part of 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.
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 defect size. This may prevent infants from untimely operative procedures with elevated operative risk.

**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 to 155 beats per minute for several hours after taking the DS. An echocardiogram was normal. Hemoglobin, serum electrolytes, creatinine, 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 if one adheres to the recommended daily allowance. At high doses it acts as an ergotropic. 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.

**Ulcus vulvae acutum – painful genital ulcers are not always caused by HSV**

C.M.B Luhmann1, M. Gittermann1, H. Yurtsever2, H.P. Gnehm1.

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 recent 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 unremarkable. 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 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 spontaneously 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 a non-venerial 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. Although 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 ulcers attributable to M. pneumoniae infection.

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

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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: Paroxysmic 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 clarithromycin was started.

D3: PCR positive for B. Pertussis. Cefuroxime stopped and clarithromycin 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 microthrombosis 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.

**Early blood exchange transfusion in malignant pertussis?**

M. Martinez1, I. Rochat1, R. Corbelli1, P. Tissiere2, C. Barazzone-Arigofto1. 1 Pediatric Pulmonology, Children’s University Hospital, Geneva; 2 Pediatric Intensive Care Unit, Children’s University Hospital, Geneva

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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 GL) 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 enterobacteriaceae, which has most commonly been described as a nosocomial pathogen, but can cause 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 objecting 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.

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 presented with 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 lorpazepam 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 could be detected in the CSF by PCR or culture. Therapy remained symptomatic. Although many issues regarding pathogenesis remain to be clarified, influenza virus infection plays a role in triggering the development of encephalopathy.

Assessment 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 effects of resuscitation, such as late neurological sequelae. 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 viewed 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 seen 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 time at points and with regard to the definition of each parameter. Therefore, better guidelines need to take into account prematurity and resuscitation measures.

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, 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 SCCT 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 (88), CNS tumours (51), embryonal tumours (68), bone tumours or soft tissue sarcomas (93) 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 230 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% (70–91%) of males aged 230 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% for males, and 24% and 47% for females. In total, 4% of all survivors received a disability benefit. This proportion rose 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 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 paraesthesia 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 of the lumbar 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 stepwise 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 = 0.0036) and was correlated with C-reactive protein (r = 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 = 0.069). 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 protamine 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 dose of 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 wheelchair dependent.

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 reagent strip, may disclose proximal tubular function.
were on monotherapy.

patients, more than two-thirds were seizure-free and the majority southern part of Switzerland. Epilepsy was classified in 45% of the This study provided valuable information on epilepsy on three or more. 

Antiepileptic drug (AED) treatment was used in 89% of patients (most 
cortical malformation (8%) and chromosomal abnormality (7%). 72% 
common syndrome of childhood absence epilepsy occurred in 20%, seizures. Epilepsy was classified in 45% of patients. The most 
183 children (99 male/84 female) were included in the study.

Results: We considered 97 children with a diagnosis of ADHD, recognized 
between 2000 and 2006, were examined and their clinical 
characteristics, school Results and therapy were studied.

Methods: A total of 97 children with ADHD, recognized between 2000 and 2006, who were followed by our department of child 
we divided the children into the three following groups: pre-school (11), primary school (67) and secondary school (18). The 
neural imaging study was not indicated in children with acute non-traumatic headache and a normal neurological 
On the contrary, urgent neuroimaging is warranted if headache is associated with neurological dysfunction. We report on three children with disrupted 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 8 years, presented with sudden onset of an intense headache, which was not 
accompanied by neck stiffness, altered mental status, or focal 
nonal imaging 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 tempo-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 case of 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 byValsava or straining, explosive onset, arterial hypertension, meningal signs, papilledema, 
motor asymmetry, ataxia, gait disturbance, and abnormal deep tendon reflexes.

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

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 ADHD, recognized between 2000 and 2006, who were followed by our department of child 
we divided the children into the three following groups: pre-school (11), primary school (67) and secondary school (18). The 
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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 analyze 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 36%, carbamazepine 11%, topiramate 10 %, 
sulthiam 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.
P72

Violence incident reporting system in geneva children’s hospital

A. Giroud Rivier1, P. Mueller1, A.-L. Rougemont2, L. Rubbia-Brandt2, and C: the geneva experience

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

Aims: 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.

Methods: We reviewed 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 voluntary and non obligatory declaration, not including serious events like lethal or severe damage to a person.

Results:

<table>
<thead>
<tr>
<th>Year</th>
<th>Violence from patients</th>
<th>Violence from parents</th>
<th>Total violence incidents (% of total workers reported TIR)</th>
<th>Nb of hospitalisations</th>
<th>Violence incidents / 100 hosp. yr</th>
<th>Problem with medication (% of TIR)</th>
</tr>
</thead>
<tbody>
<tr>
<td>2002</td>
<td>0</td>
<td>0</td>
<td>(23)</td>
<td>2174</td>
<td>0.45</td>
<td>15 (27)</td>
</tr>
<tr>
<td>2003</td>
<td>1</td>
<td>0</td>
<td>(24)</td>
<td>2377</td>
<td>17.25</td>
<td>26 (50)</td>
</tr>
<tr>
<td>2004</td>
<td>2</td>
<td>0</td>
<td>(27)</td>
<td>2713</td>
<td>11.43</td>
<td>22 (42)</td>
</tr>
<tr>
<td>2005</td>
<td>1</td>
<td>0</td>
<td>(26)</td>
<td>2885</td>
<td>11.44</td>
<td>27 (57)</td>
</tr>
<tr>
<td>2006</td>
<td>1</td>
<td>0</td>
<td>(25)</td>
<td>2324</td>
<td>25.39</td>
<td>27 (56)</td>
</tr>
</tbody>
</table>

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 literature (related to medication) remained during the same period. The violence incidents happened in 3 situations: aggressive parents against staff in the emergency room, aggressive patients in both paediatric and pedopsychiatric wards.

Conclusions: Although incident reports are based on a voluntary 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 aggressive patients. 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 aggressive behaviour.

P73

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


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2 Division of clinical pathology, HUG, Geneva

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 biochemical and histological evidence of chronic liver disease who are at higher risk to develop cirrhosis and hepatocarcinoma were treated in our division.

Methods: We included hepatitis B and C hepatitis patients with a liver biopsy. Patients with chronic hepatitis B received subcutaneous recombinant peginterferon alfa-2a (Pegasy®) 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 ± 6.2, 2 horizontal transmission (1 ungenital), 7 vertical transmission (1 ungenital, 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 chronic hepatitis C had a liver biopsy. 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.

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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 (ASIFT) 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 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 department. 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 (p = 0.001). Children suffering from malignant bone tumours and soft tissue sarcomas were also at increased risk to be treated in other clinics (p = 0.001). 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 stable 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 broncho pulmonary 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 clinically stable IBD.

Patients and methods: 20 children with clinical stable 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 (2.5 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 stable IBD showed 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.

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, gastrointestinal, genitourinary, 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: 38.9%; children: 14.4%) and psychiatric 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.

Acute lower abdominal pain in girls – the challenge of early recognizing, diagnosing and treating ovarian torsion
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Introduction: Ovarian torsion should be considered in any girl with acute lower abdominal pain. The clinical presentation is often non-specific and ultrasound can be misleading. Prompt diagnosis of ovarian torsion is important to preserve ovarian function and avoid complications.

Case report: 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 pelvic inflammatory disease. The diagnosis was established by intraoperative findings. 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 an enlarged ovary with heterogeneous appearance and numerous small peripheral follicles in the ovarian parenchyma. Intraoperative 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.

Conclusions: PFAPA syndrome is a recurrent febrile disease first described in 1987 by Marshall et al, and characterized by periodic fever, aphthous 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% vs 89%, headache 39% vs 15%, abdominal pain 69% 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.

Familial aspect in pfapa syndrome
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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. 6/14 (35%, CI95: 19–51) 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 these 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.
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 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 low index for the prediction of asthma at school age or children without recurrent coughs.

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 measured 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 ± 47 month in children with current asthma at school age was 9.65 (5.8–11.1) median (IQR), compared to 6.5 (5–9.8) 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 predicitive values. In addition the value of FeNO will be compared to the asthma predicitive index, and whether the measurement of FeNO in addition, the API enhances the detection rate of future asthmatics at school-age.
500 mcg IFP are generally considered as safe. Therefor, AI should also be considered in children with a history of low dose corticosteroids, which is generally accepted as safe. Therefor, AI developed under low-dose 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 present 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 and the roles and responsibilities of the partners involved. We recruited 30 mild poly-allergic, asthmatic patients on a low dose treatment (200 mcg BDP equivalent) with inhaled corticosteroids. We measured exhaled nitric oxide (eNO) with the Niox mini®, 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 FEV1 was significantly (p = 0.03) lower than baseline. Results: Demographical characteristics were similar except for allergic sensitisation (p < 0.01) and cough without cold (p < 0.005). Baseline RrsHz and Rrs-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 standardized 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.

**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 as bronchocstriction (EIB) is a common feature of childhood asthma and tests to evaluate 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 c 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 Rrs-24Hz or ≥32% in RrsHz to baseline.

**Results:** Demographical characteristics were similar except for allergic sensitisation (p < 0.01) and cough without cold (p < 0.005). Baseline RrsHz and Rrs-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 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 standardized 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.
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, Shigelllosis and Yesinia were also done and were positive only for Salmonella. A foot X-ray showed nothing pathological. A scintigraphy showed hypercapacitation 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 osteon 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 blood analysis 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 Köhler disease. Köhler disease is an osteonecrosis of the navicular bone. 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 naviculare 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 cellulitis that has an atypical clinical presentation or follow-up.

Case report: recurrent fever is not always a psapfa

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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 most common cause of recurrent viral infection is recurrent adenovirus infection. The differential diagnosis of recurrent viral infections and of autoinflammatory diseases should not be forgotten. We present the case of a 5 years old girl with etiologically recurrent fever episodes of 38.5°C to 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 PSAPA 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 episode 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.Ile626Thr/ p.Val337Ile. Because she was heterozygous, a spontaneous mutation was unlikely and we recommend a genetic analysis of the parents.

Our case shows that PFAPA (periodic fever, aphthous 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, urin ary mevalonic acid at the beginning of the fever episode should be measured.
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 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 [0.05–0.95 mcg/l]) and pelvic ultrasound showed an ovarian mass which was confirmed by the CT-scan and compatible with a uroateria 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.

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 (coccidioidomycosis, 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, anaemia, fatique, 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 (8.4 mmol/mmol creatinin) and hypophosphatemia (1.0 mmol/L) 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 antimarial regimen with quinine and clindamycin (each 10 mg/kg tid) for 7 days. Over the following 5 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 thombocyte 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 Western Europe has been previously reported to our knowledge.

This case highlights the importance of close collaboration between institutions (referring pediatrician, pediatric hospital and tropical institute) to establish the optimal management of these patients. Our case 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.

References:

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 parasite 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. A full blood test revealed anemia and severe platelet reduction and an 8% infestation with Plasmodium falciparum. No such case of severe pediatric malaria with poor prognostic factors (low age, high parasite load, respiratory distress) in Western Europe has been previously reported to our knowledge.

This case highlights the importance of close collaboration between institutions (referring pediatrician, pediatric hospital and tropical institute) to establish the optimal management of these patients. Our case 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.

References:

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


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. They described a developmentally 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 emotionally engaged and had 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.
Retinopathy, progressive intracerebral calcification and epilepsy in a premature new-born: case report of a new entity

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Poster presentations

Background: The most common cause of cerebral calcifications and retinopathy in infancy are intrauterine infections by toxoplasmosis, rubella, CMV and herpes (TORCH). Calcifications can also be seen in tumoral, endocrine, metabolic and degenerative conditions (Aicardi-Goutière, Cockayne syndromes). We present an infant with a severe retinopathy, first attributed to prematurity, and progressive cerebral calcifications which 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 hemorrhage and severe bilateral retinopathy. Cerebral echography at 3 weeks revealed a few periventricular echogenecities. 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 pleocytosis, elevated proteins and lactate. PCR in CSF and serologies for TORCH were negative Serum lactate, calcium, phosphate, alpha-interferon (also in CSF) were normal.

Conclusions: CRMCC is a new multisystemic disease entity of probable genetic origin which includes Coats retinopathy and leukoencephalopathy with calcifications and cysts (Labruné et al., 1996). Very early onset has been described in only one case and represents a diagnostic challenge.

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

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Introduction: 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 this organ is important in the induction of 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 85 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. This population included measurements of fractional exhaled nitric oxide (FeNO), pulmonary function (spirometry, whole body plethysmography and forced oscillation [FGO]), 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 (FOTs4-24Hz; p = 0.03) and a lower ratio FEV1/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 airway hyper responsiveness.

Conclusions: Bronchial hyper-responsiveness persist to school age in most 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.

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 being serious such as septic shock, multorgan failure and have significant mortality. Streptococcus pneumoniae 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 pyodermas 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 preceding infection lasting one to seven days, all children deteriorated within less than one day, proceeded to sepsis, to septic shock and consecutive multorgan 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 children survived, however one patient developed severe necrotising fascitis 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 preceding 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 deterioration are the only clues to early diagnosis.

Post-Kawasaki disease arthritis

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We report the case of a patient who developed symptoms of polyarthritus after being treated with immunoglobulin (IVIG) for Kawasaki disease (KD). A three year old previously healthy girl, was 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 diagnosis she suffered from the hospital. The patient was rehospitalized for reapprarition of the lymphadenopathy, and polyarticular arthritis 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 implicates mostly the large joints. The incidence of post-Kawasaki arthritis is reported at 2%. These cases are reported to have elevated inflammatory markers. This form of polyarthritus usually cedes without 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 polyarthritus 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.
M. Albisetti1, S. Kroiss-Benninger1, O. Kretschmar2, I. Forster3, M. Schmugge1,
in infants and HIV. In our case we assume a relation with Salmonella typhi.
non-specific immune responses to unknown pathogens. The most immunologic responses may represent epiphenomena caused by antibody titers also have been offered as proof of causality, but such although clear causality is lacking. Elevations of pathogen-specific symptoms have been used to support these relationships in PL, acute lichen planus, guttate psoriasis, erythema multiforme) are A number of acute exanthems (eg, PL, pityriasis rosea, decrease of the fever and complete remission from the skin lesions. After a positive test for salmonella in the stools and the diagnosis of histopathologically compatible with PLEVA. The serologies for HIV, vesicles, partly necrotic eruptions and scars, which has a history of living in Switzerland for five years without any contact to foreign We present a 9 10/12 year old originally Afghan boy, standard of therapy.
to be different presentations of the same process, with the process being more rapidly accelerated in PLEVA. There is currently no to be different presentations of the same process, with the process being more rapidly accelerated in PLEVA. There is currently no
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 which 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.

Psychosomatic versus somatopsychic disease: a tricky question S. Pradet1, S. Neté, A. Jágy1, A. Somverville1, U. Hunziker1, 1 Kantonsspital Winterthur, 2 Kinderklinik Zürich 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 at 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 psychosomatic 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 psychosomatic 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 osteonecrosis (2), septic saccroilitis (3), skin 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 psychosomatic 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 psychosomatic 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.


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.

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**P105**

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

G. Oesch¹, U. Cafisch⁴, G.E. Pfyffer¹, H. Haechler³, J. Spalinger¹.

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 immunocompetent 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 persistently 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 a 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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**P106**

**Paediatric registries in switzerland – overview and call to join forces**

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Introduction: Paediatric 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.org) by representatives of five existing paediatric registries: the Swiss Childhood Cancer Registry (www.kinderkrebsregister.ch), the Swiss Neonatal Network and Foundation (www.neone.net), the Swiss Paediatric Stroke Registry (www.neuropaediatric.ch) and the Swiss Paediatric 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 gold standard 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.

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**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 chews on a hard object (e.g. 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 a foreign 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 object could be 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.

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**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 forms an important 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.
hours of life. Clinically all patients had abnormally low transcutaneous saturation and absent DA on echocardiography in 3 patients. All patients survived O2, NO and sildenafil. Catecholamine infusion was given to sustain RV systolic and diastolic function in 3 patients. All patients survived O2, NO and sildenafil. Catecholamine infusion was given to sustain RV systolic and diastolic function in 3 patients. All patients survived. RV systolic and diastolic function in 3 patients. All patients survived. RV systolic and diastolic function in 3 patients. All patients survived.

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.

Premature closure of the ductus arteriosus in utero: treatment and evolution
T. Boulos-Ksontini1, N. Sekarski1, J.F. Tolsa2, E.J. Meiboom3, S. Di Bernardo4. 1 Pediatric Cardiology Unit, CHUJ/ Lausanne; 2 Neonatology Unit, CHUJ/ Lausanne
Introduction: The ductus arteriosus (DA) plays a major role in fetal circulation. Antenatal ductus closure is uncommon and can lead to preterm birth and need of RV afterload reduction and catecholamines, but once initial adaptation is overpassed RV hypertrophy regression is rare, but distinct entity of paediatric patients with 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 Photoshop the true length of each line 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.

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 severe long-term outcome compared to singletons.

Aim: To assess, at school age, motor and cognitive development of children born from a triplet pregnancy. The aim of this study was to compare the outcome of these children with that of the corresponding birth weight discordance pairs and the population norms and to investigate the predictive value of specific neonatal risk factors for adverse developmental outcome.

Methods: 65 live-born children 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 Kaufman-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). Triplet 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 (84.8, ±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 <0.05) 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: Triplet's 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.

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 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 Photoshop the true length of each line 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.

Basilar thrombosis in children: a rare but potentially life-threatening problem
M. Gautschi1, E. Bolthausen2, T. Schmitt-Mecheke3, M. Weissert4, E. Weh1, M. Steinitz1. 1 University Children’s Hospital Bern; 2 University Children’s Hospital Zurich; 3 Children’s Hospital Lucerne; 4 Children’s Hospital St.Gallen
Background: Basilar thrombosis is a rare event, but distinct entity of paediatric patients with 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 Photoshop the true length of each line was produced including an overlay of all individual lesions which was used for comparison with the published lines of Blaschko.

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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 disease and manifest primarily as meningitis.

Aim: To present the case of a preterm infant with early-onset neonatal listeriosis and to discuss the natural history, best diagnostic and treatment approaches, and also outcome is still very limited.

Case report: This male infant was delivered by CS at 26 5/7 weeks of gestation because of suspected amniotic fluid infection syndrome following an otherwise unremarkable pregnancy. The patient required respiratory support with HFNO and INO and circulatory support with inotropes and vasoactive agents. There was a marked maculopapular exanethema 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 patient required 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 a bite. Even in instances deemed unlikely, since imported and those of a possibly venomous snake or of a mammalian carrying a bite, considering that medical literature does not provide official guidelines on this issue, 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.

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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 presented in an immunocompromised patient, a biopsy of the suspected organ should be considered.

Absence status epilepticus as a 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.
to the antibiotic therapy in the Aim to cover the classical etiologic
in left jugular vein until the base of the brain. We added Clindamycin
with a Doppler Scan of the left cervical region which revealed a left
an important left pleural effusion. The presumptive diagnosis of a
investigation showed then clearly bilateral pulmonary infiltrates and
2 days with dyspnea, tachypnea and persistent fever. A thoracic X-ray
started. Nonetheless clinical conditions of our patient worsened after
headache. Since a few days she developed pain in the left cervical
emergency room with one week-persistent fever, pharyngitis,
An otherwise healthy 17 years old girl presented to our
Introduction: Cyclic electrographic seizures are a form of status
in children. In order to better understand the phenomenon of cyclic seizures in
occurred predominantly in neonates and/or in the context of an acute
1 patient. 4 patients died. The remaining suffered from epilepsy,
were included in the study. We searched the patients’ files for age,
sex, underlying aetiology, whether an epilepsy disorder had been
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Spreads. Orphanet Switzerland (www.orpha-net.ch), active since 2001, is in charge of collecting information about the national resources and 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.

**Case report:**

**Profound hyperbilirubinemia – a rare manifestation of acute Epstein-Barr virus infection in adolescence**

S. Yammine, A. Duppenthaler, S. Lüer, S. Schibli.
University Children’s Hospital, Inselspital Bern

**Aim:**
Orphanet website: a new version

**Methods:**

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.

**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 EuroGeneTest) are just a click away. A new 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 enhance the 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 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 kept clinical trials information accessible in a well-structured manner.

**Conclusions:**

Intussusception beyond the typical age (infants and toddlers) presents with the same symptomatology but Results usually from a point lead, most common a Meckel diverticulum as in our case. To our knowledge a double intussusception is a very rare event.

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


**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 study 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: 10 y 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.), 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.

Four 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.
Extremely low birth weight (ELBW) infants are at high risk as they require care ranging up to 9 per 1000 patient days. In particular, preterm and report healthcare-associated infection (HAI) rates in neonatal infants.


The counseling let us to understand that every adolescent is unique, everyone is the protagonist of his own story, even during his disease.

Results: A total of 98 BSI (bacteremia or clinical sepsis) were detected among 1552 neonates with 1928 central lines and a total of 10,655 catheter days. The median day of catheter use (interquartile range) was 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 [8–48] for tunneled 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 (2.6/1000) 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 high risk, especially for ELBW children. Umbilical catheters show the highest risk, but because catheter duration is highest among SVC, they are the most likely to infect.

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

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 in their problems. Our study wants to examine the efficacy of the counselling to the teen ages during the illness.

Methods: We identified one physician as reference to the teen ages. This physician offered himself to help them giving a service of counselling. The object of the counselling was to help the adolescent becoming less conscious about himself so that he can lives his problems with more tranquility. This happened with a direct relationship between the adolescent and his counsellor, without the presence of the parents. The counselling happened during the hospitalization or in the first aid station and then continued with periodic visit after the discharge. 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 valued 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.
Swiss pediatrician's training needs in promoting and supporting breastfeeding

N. Pellaud, E. Jeannot. Société suisse de pédiatrie

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.

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