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Free communications

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FM01

Susceptibility of Escherichia coli strains in children with community-acquired urinary tract infection in Tessin

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Background: Treatment with a third-generation cephalosporin is currently advised for Swiss children with urinary tract infection. On the contrary, co-amoxyclav is currently recommended in Northern Italy.
Objective: To prospectively assess the susceptibility of Escherichia coli strains isolated from children with a symptomatic community-acquired urinary tract infection.
Methods: Susceptibility of 100 Escherichia coli strains causing symptomatic community-acquired urinary tract infections was assessed in children (73 girls and 27 boys aged between 5 weeks and 17 years, median 33 months) attending our Emergency Unit. Strains from children receiving antimicrobial prophylaxis or prescribed antimicrobials in the previous month were excluded.
Results: High rates of ampicillin (39 strains) and co-trimoxazole resistance (21 strains) and low rates of nitrofurantoin resistance (4 strains) were identified. No resistance was identifid for co-amoxiclav and third generation cephalosporins.
Conclusions: Among children of Tessin with symptomatic community-acquired urinary tract infection uropathogenic Escherichia coli strains resistant to nitrofurantoin, co-amoxiclav and third generation cephalosporins are uncommon. It is tempting to assume a more severe resistance pattern in children with hospital acquired urinary tract infection and in children receiving antimicrobial prophylaxis or recently prescribed antimicrobials (Borsari AG, et al. Clin Ther 30:2090-5,2008).

FM02

Hemolytic-uremic syndrome in Switzerland: a nationwide surveillance from 1997 to 2003

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Background: Hemolytic-uremic syndrome – characterized by microangiopathic anemia, thrombocytopenia, and acute renal impairment – is a leading cause of acute renal failure in childhood. In its typical presentation, it is preceded by an episode of diarrhea mostly due to Shiga-like toxin-producing *Escherichia coli*. Multiple studies indicate important geographical variation in regard of clinical, epidemiological, and microbiological features. Beside some single center studies, nationwide data on childhood hemolytic-uremic syndrome in Switzerland are not available.
Methods: A prospective, national surveillance through the Swiss Pediatric Surveillance Unit from April 1997 through March 2003.
Results: One hundred-fourteen cases (median age 20.5, range 0–161 months; 50% boys) were reported by 38 pediatric units (annual incidence 1.42 per 10^5 children aged 16 years or less). Shiga-like toxin-producing E. coli were isolated in 60% of tested stool specimens, serotype O157:H7 in eight of them. Sixteen children presented with only minimal renal involvement, including three with the primary site of infection within the urinary tract. Twelve patients presented with atypical hemolytic-uremic syndrome, six of them due to infection with *Streptococcus pneumoniae*. Overall mortality was 5.3% (6/114), including two children with atypical hemolytic-uremic syndrome due to *Streptococcus pneumoniae*. The severity of thrombocytopenia significantly correlated with mortality.
Conclusions: Hemolytic-uremic syndrome is not rare in Switzerland, predominantly affecting children less than five years of age. Contrasting other countries, E. coli O157:H7 play only a minor role in the etiology. Incomplete manifestation with minor renal involvement is not uncommon and in some cases the primary site of the infection is located in the urinary tract. Severe thrombocytopenia seems to be associated with a bad outcome. The highest mortality (33%) is observed in *Streptococcus pneumoniae*-associated atypical hemolytic-uremic syndrome. Robust clinical, epidemiological and bacteriological data have been established through this prospective cross-sectional study using a national surveillance unit.

FM03

Passive Smoking Increases Blood Pressure in Pre-School Children

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Introduction: Passive smoking is associated with early arterial damage, arterial stiffening and increased arterial pressure in adults. Parents smoking at home are mostly responsible for children's exposure to tobacco smoke. The influence of passive smoking on blood pressure in childhood has not been so far studied. We aimed to analyze the influence of passive smoking and other preventable risk factors on blood pressure in a large cohort of pre-school children.
Methods: As part of a screening project, blood pressure, BMI and fat mass (by skinfold thickness) were determined in 4231 pre-school children (5.8 ± 0.6 years) and potential preventable risk factors (parents' smoking habits) were documented.
Results: Smoking was reported by 28.6% of the fathers and 20.7% of the mothers. The prevalence of parental smoking decreased with increasing educational level. Significantly higher BP values were observed in children of smoking parents (systolic BP +1.1, 95% confidence interval +0.6 to +1.6 mm Hg, $p < 0.0001$; diastolic BP + 0.5, 95% confidence interval +0.1 to + 0.9 mm Hg, $p < 0.02$). Moreover, BP showed a linear correlation with BMI (systolic $r = 0.29$, diastolic $r = 0.21$; both $p < 0.0001$) and fat mass (systolic BP $r = 0.13$, $p < 0.0001$, diastolic $r = 0.11$, $p < 0.0001$).
Conclusion: Already in pre-school children, environmental factors like passive smoking or dietary habits play a crucial role in determining BP level. Because blood pressure increase related to passive smoking may be only partially reversible after cessation of the exposure, the present data strongly emphasize the importance of implementing smoke-free environments for children at home and in public places. Prevention with healthful lifestyle training already in childhood, together with prevention programs addressed to the entire family seems to play a crucial role in preventing cardiovascular diseases.

FM04

Education and Employment in young adults with congenital heart disease

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Introduction: Because of improved survival of children with congenital heart disease the focus shifts toward quality of life. This study therefore assessed school education and employment of these young adults.
Methods: A questionnaire was sent to all young adults between 16 and 18 years with congenital heart disease treated at our institution (N = 326). We asked for educational level, employment status and socio-economic status. These data were compared with the official statistics of our region, which was published by the government
Results: The questionnaire was returned by 207 of 326 patients (63%). 42 (21%) finished school in the highest degree. 61 (30%) in second, 77 (37%) in third and 24 (12%) in the lowest degree or in a special school for disabled children. The distribution in the general population was 16%, 41%, 33% and 10% respectively. After the compulsory school of 9 years, 44 (21%) of those with congenital heart disease attended higher level schools towards a university degree. This is more than the 17% of the general population. 140 (68%) learned a profession on the regular level, where 6 (3%) learned it on an adapted level. 12 (6%) couldn't find a job. Of those 5 thought it was because of their heart defect, whereas 7 thought it was because of other reasons. There was no significant difference regarding the severity of the defect. 30 of those learning a profession answered, that this was not their desired profession.
Conclusion: School education in children with congenital heart disease is good in general, but there is a slightly higher percentage of children in the lower grades. Employment status is surprisingly high compared to other publications. One reason might be the relative low unemployment rate of our region. But despite this pleasant result, approximately a fifth of all couldn't learn their desired profession.

FM05

Sudden cardiac death: clinical evaluation of paediatric family members

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Introduction: The annual incidence of sudden cardiac death (SCD) in the general population is estimated as 1 in a 1000. A number of SCDs are caused by inherited cardiac diseases, such as cardiomyopathies, channelopathies or conduction system disorders.
Patients and methods: A total of 36 consecutive families referred after a sudden cardiac death (n = 29) or life-threatening event (n = 7) of a family member, were analyzed. Mean age of the 58 paediatric family members was 8.3 ± 6.1 years. Children were first (n = 19) or second-degree (n = 39) relatives. Stepwise evaluation included history, 12-lead and 24-hour electrocardiogram (ECG) as well as 2D echocardiography in all. Additional investigations were signal average ECG, cardiac magnetic resonance imaging (cMRI), event recorder and genetic testing when indicated.
Results: Inherited heart disease was identified due to autopsy, family or post-event evaluation in 23/36 (64%) families: cardiomyopathies (n = 12), channelopathies (n = 9), and sinus node dysfunction (n = 2). A total of 18 (31%) of the evaluated paediatric relatives were diagnosed with likely inherited heart disease: cardiomyopathies (n = 6), channelopathies (n = 8), and sinus node dysfunction (n = 4). The proportion of affected first-degree (n = 6, 32%) and second-degree relatives (n = 12, 31%) did not differ. The following evaluations contributed either separately or combined to the diagnosis: 12-lead ECG (n = 12), 24-hour ECG (n = 10), 2D echocardiography (n = 8), signal average ECG (n = 1) and cMRI (n = 1). Channelopathies were genetically proven in 3 children. None of the affected patients suffered from cardiac symptoms before evaluation although previous syncope was reported in two. Prevention of SCD or of a cardiac event was initiated in 16/58 (28%) by implantation of an antibradycardia device (n = 3), an implantable cardioverter defibrillator (ICD, n = 6) and/or medication with antiarrhythmic drugs (n = 8). Subsequently, no cardiac event was noted in any patient. Appropriate and successful ICD discharges occurred in 4 (22%) of the affected patients.
Conclusion: A stepwise, comprehensive clinical investigation of SCD or life-threatening event families identifies a substantial number of paediatric relatives at risk for sudden cardiac death. This allows for targeted prevention by effective treatments and evaluation of further relatives.

Free communications – Ambulant Paediatrics

FM06

«walk-out» Patienten in der interdisziplinären Notfallstation der Universitäts-Kinderkliniken Zürich

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Introduction: The aim of our study is to analyse the reasons why patients and their parents leave our emergency department (ED) before they have be seen by an emergency doctor. They were all triaged by a nurse (Cat 1 "to be seen immediately" to cat 5 "can wait 2 hours"). Additionally we studied the grad of illness of these children, the waiting time in the ED, the further progress of the illness, the parents satisfaction and their socioeconomic background.
Methods: We conducted a phone survey of all parents who left our emergency department with their children without a medical consultation in December 2007 and April 2008.
Results: In this two months 137 (2.6%) out of a total of 5205 children left our ED without a medical consultation. The weekly distribution of the cases is even. 132 (96%) received a triage category 4 or 5. 81 (59%) didn't visit another doctor in the progress. One child had a pneumonia and one a pyelonephritis. The mean waiting time before they left was 76 minutes. The parents overestimated this time (+32 min). The reasons why they left our ED were: 73 (70 %) problem solved by itself or by a nurse, 58 (55%) waiting time too long. 56 (53%) left the ED satisfied, 45 (33%) unsatisfied. There was no difference regarding age, domicile and socioeconomic status.
Conclusions: The 2.6% walk out rate and the mean waiting time of 76 min. before leaving is low compared to other studies. The children are not critically ill and in the progress were no bad outcomes. A high percentage of the parents leave the ED satisfied because of the received advice at the triage. The walk out rate could be reduced through a pleasant waiting area and a good catering.

FM07

Intensive insulin treatment in children and adolescents with newly diagnosed type 1 diabetes: multiple daily injections vs. continuous subcutaneous insulin infusion

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Background: Intensive insulin treatment in children and adolescents with newly diagnosed type 1 diabetes proved to be safe and feasible. The aim of our study was to compare intensive insulin treatment by means of multiple daily injections (MDI) vs. continuous subcutaneous insulin infusion (CSII) in newly diagnosed type 1 diabetes with respect to HbA1c, insulin dose, weight, frequency of ketoacidosis, and hypoglycaemia.
Methods: Prospective cohort study of newly diagnosed type 1 diabetes in children and adolescents admitted to the children's hospital Luzern, from 1/2003 to 12/2007. Follow-up with HbA1c (DCCT/IFCC), weight, insulin dose, incidence of hypoglycemias. Data are mean [SD], differences between groups are calculated by student's t-test and Mann Whitney rank sum test.
Results: 76 children were included, 44 male/32 female, mean age 9.2 [4.2] years, mean duration of hospital stay 11.7 [3] days, mean duration of follow-up 2.5 [1.4] years. 62 children (81%) were treated with MDI, 14 children (19%) with CSII. The mean age in the MDI vs. CSII group was 9.5 [4.2] vs. 8.0 [4.1], (p = 0.19); the mean HbA1c 8.1 [1.4] vs. 7.9 [0.4]% (p = 0.98); the mean insulin dose 35.6 [20.9] vs. 24.6 [8.4] U/d (p <0.001), corresponding to 0.75 vs. 0.58 U/kg BW, and the mean BMI was 19.7 [4.3] vs. 18.7 [3.8] (p = 0.44). There were 7 patients admitted with ketoacidosis in the MDI group (4.7/100 PY) vs. 0 in the CSII group, and there were 3 (2.1/100 PY) vs. 1 (2.6/100 PY) severe hypoglycaemias.
Conclusions: Intensive insulin treatment with MDI and CSII is safe with low rates of ketoacidosis and hypoglycemias. Treatment results with MDI and CSII are comparable, however, insulin need and the rate of ketoacidosis are significantly lower in patients treated with CSII.

FM08

Adolescents with Essential or White Coat Hypertension Display an Altered Circadian and Ultradian Blood Pressure Rhythmicity

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Introduction: Adults with primary or white coat hypertension display blunted cardiovascular rhythms. We studied cardiovascular rhythms among children with primary hypertension or white coat hypertension.
Methods: Cardiovascular rhythms were analyzed by Fourier analysis of 24h ambulatory blood pressure profiles in 129 children with primary hypertension (age 14.7 ± 2.6 years) and in 54 children with white coat hypertension (age 14.5 ± 3.0 years). 146 age- and height-matched healthy subjects (14.6 ± 2 years) represented the control group.
Results: The day/night mean arterial pressure ratio was lower in children with primary hypertension and white coat hypertension compared to controls (1.13 vs. 1.16 vs. 1.21, respectively; p <0.0001). The prevalence of 24 h blood pressure rhythms was similar among the groups but prevalence of 12h blood pressure rhythms was increased in primary hypertension (67%) and white coat hypertension (72%) compared to controls (51%, p <0.0001). The amplitudes of the 24 h, 8 h, and 6 h blood pressure rhythms were reduced in primary hypertension and white coat hypertension compared to controls (p <0.05). Primary hypertension and white coat hypertension patients displayed delayed 24 h, 12 h, 8 h, 6 h acrophases (the time from midnight to the highest value during the rhythm) in comparison with controls (p <0.05).
Conclusion: Children with primary hypertension or white coat hypertension exhibit abnormal circadian and ultradian cardiovascular rhythms with higher prevalence of non-dipping compared to normotensive children. Children with white coat hypertension display intermediate patterns of cardiovascular rhythms in comparison to both normotensive and primary hypertensive patients.

FM09

A school-based physical activity program increases fitness and decreases adiposity in primary school children (KISS): a cluster-randomized trial

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Background: Childhood obesity and physical inactivity are increasing dramatically worldwide with detrimental effects on fitness, cardiovascular health and quality of life. Schools provide an ideal setting for preventive interventions. We therefore conducted a randomised controlled trial to determine whether a school-based physical activity (PA) program during a full school-year improves body composition, fitness, cardiovascular health, PA and quality of life in primary school children.
Methods: Twenty-eight classes from two of 26 provinces in Switzerland were cluster randomized to the intervention (16 classes, n = 297) and control (12 classes, n = 205) groups. The intervention consisted of a multi-component PA intervention including daily physical education. Primary outcomes included body fat (skinfold thickness), aerobic fitness (shuttle run test), PA (accelerometry), and quality of life (questionnaires). Children and parents in the control group were not aware of the existence of an intervention group. Analyses were done according to intention to treat.
Results: 498/540 completed the pre- and postintervention assessment, mean age was 6.9 ± 0.3 years for 1st grade and 11.1 ± 0.5 years for 5th grade. After adjustment for grade, gender and cluster, children in the intervention arm compared with controls showed statistically significant additional average changes in body mass index by –0.22 (95% CI –0.38 to –0.05, p = 0.009), in sum of four skinfolds by –1.78 mm (–3.38 to –0.19, p = 0.02), in aerobic fitness z-scores by 0.22 (0.01 to 0.42, p = 0.04), in moderate-vigorous physical activity in school by 14 min (5 to 23, p = 0.008) and in the cardiovascular risk score by -0.18 (–0.30 to –0.05, p = 0.005). Overall daily physical activity and quality of life did not change differently.
Conclusions: A school-based multicomponent physical activity intervention including compulsory elements can improve physical fitness and reduce adiposity and a composite cardiovascular risk factor score in children. Implementation of such a program may help to improve health and fitness of our children, and also improve health later in life by reducing cardiovascular and other diseases.
Supported by the Swiss Federal Office of Sports and the Swiss National Foundation (PMPDB-114401)

FM10

The success of a general school-based physical activity intervention on bone mineral content depends on pubertal stage but not on gender

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Background: It is not clear whether sensitivity of bone to physical loading varies for different pubertal stages and among genders. We therefore performed a randomised controlled trial in to determine whether a school-based physical activity (PA) program during a full school-year influences bone mineral content (BMC) and whether there are differences in response for boys and girls before and during puberty.
Methods: Twenty-eight 1st and 5th grade classes were cluster randomised to an intervention (INT, 16 classes, n = 297) and control (CON; 12 classes, n = 205) group. The intervention consisted of a multi-component PA intervention including daily physical education during a school year. Each lesson was predetermined, included 10 min of jumping or strength training exercises and was the same for all children. Measurements included anthropometry, tanner stage, and BMC for total body, femoral neck, total hip and lumbar spine using dual-energy X-ray absorptiometry (DXA). PA was assessed by accelerometers. Bone parameters were normalized for gender and tanner stage. Analyses were performed by an adjusted regression model.
Results: 275 of 380 children who initially agreed to have DXA measurements had also post-intervention DXA and PA data. Mean age of prepubertal and pubertal children at baseline was 8.74 ± 2.07 and 11.13 ± 0.60 years, respectively. 64/144 girls and 86/131 boys were prepubertal at the end of the intervention. Compared to CON, children in INT showed statistically significant increases in BMC of total body (adjusted z-score differences: 0.110; 95%-CI 0.042 to 0.178), femoral neck (0.136; 95%-CI 0.014 to 0.257), and lumbar spine (0.110; 95%-CI 0.028 to 0.191). There was no gender*group, but a tanner*group interaction favoring prepubertal children.
Conclusion: This study shows that a general, but stringent school-based PA intervention can improve femoral neck, lumbar spine and total body BMC in elementary school children. Pubertal stage, but not gender seems to determine bone sensitivity to physical loading.
Supported by the Swiss Federal Office of Sports and the Swiss National Foundation (PMPDB-114401)

Free communications – Chronic diseases and long-term effects

FM11

Efficacy of rhDNase on lung function in patients with cystic fibrosis: a long-term retrospective observational study

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Introduction: Recombinant human deoxyribonuclease (rhDNase) has been shown to improve lung function in the short-term and to reduce the number of pulmonary exacerbations in patients with cystic fibrosis (CF), but its long-term effect on airway inflammation and function has not been assessed.
Methods: In a retrospective observational study we evaluated the long-term effect of rhDNase on bacterial acquisition and forced expiratory indices (FEV_1, FEF_{50}), functional residual capacity (FRC_{pleth}), lung clearance index (LCI), trapped gas (V_{TG}), airway resistance (sR_{eff}), and PaO_2 in 170 children (85 males; 85 females) with CF receiving or not rhDNase (2.5 mg/day) over an observation period ranging from 5 to 18 years of age. Linear mixed model analysis and binary logistic regression analysis were used to assess drug efficacy on lung function.
Results: A significant improvement on lung function, comparing a 10 yrs period prior rhDNase with a 10 yrs period after initiation of rhDNase treatment could be demonstrated by LCI ($p = 0.002$). This effect on ventilation inhomogeneities was better shown in CF patients with age <10 yrs. We did not find any long-term effect of rhDNase treatment on any of the other examined lung function parameters.
Conclusion: This observational study showing only modest long-term efficacy of rhDNase in CF patients raises concerns about cost effectiveness and, therefore, prospective long-term studies on rhDNase efficacy in CF patients are warranted.

FM12

Use of pegylated Interferon in children with chronic hepatitis B and C: the Geneva and Lausanne experience

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Aims: Hepatitis B and C cause morbidity and mortality. Vertical infection is the most important route of transmission in children and long-term spontaneous clearance is known to be low. Children with active disease who are at high risk to develop cirrhosis and hepatocarcinoma were treated in our divisions.
Methods: HBV and HCV groups received for 48 weeks subcutaneous recombinant peginterferon alfa-2a (Pegasys®) at a dosage of 100 µg/m² once per week in combination with oral ribavirin (15 mg/kg x day in 2 doses) for the HCV group. Physical examinations, viral load, ALT levels and blood count were determined during the treatment and the follow-up (3 and 6 months after the end of the treatment).
Results: HBV group: 15 patients, median age 12,5 y (4–17 y), 4 horizontal, 10 unknown and 1 sexual transmissions, all HBsAg/HBeAg positive. 5/9 had HBeAg seroconversion with one concomitant HBsAg seroconversion, 4 did not have seroconversion, 2 discontinued therapy after 12 weeks because of elevated transaminase or no response to treatment, 4 are still on therapy. HCV group: 10 patients, median age 7y (3–15 y), 8 vertical, 2 unknown transmissions. 5 genotypes 1A, 2 genotypes 1B, 3 genotypes 3A, 7/10 had negative viremia after 24 weeks of treatment (early viral response), 2 are still on therapy, 1 stopped the therapy after 36 weeks because of no response (genotype 1A). At present, no negative viremia at 24 weeks relapsed in the follow-up period.
Conclusion: Our patients tolerated well the therapy with minor side effects. Weekly peginterferon was well accepted even in very young

children. We had 5/15 HBeAg seroconversion with one HBsAg seroconversion and 7/10 early viral response with hepatitis C. In the literature, 1/3 of children with hepatitis B have a sustained response to therapy and 1/10 will become both HBeAg and HBsAg negative. In hepatitis C, between 44 and 75% will have sustained viral response according to the genotype. Therefore, in our hands, both HBV and HCV therapies seem to be more effective than reported in the literature, certainly due to precocious intervention.

FM13

Management of infantile feeding disorders: A multidisciplinary approach

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Aim: Feeding disorders in infants and young children differ in their etiology, accompanying symptoms and investigative steps required. Guideline-orientated, multimodal therapeutic approaches have been successful for the treatment of these disorders. At the Ostschweizer Kinderspital we created a multiprofessional management team taking care of the follow up of patients with infantile – feeding disorders, taking the whole family setting into consideration.
Methods: 24 patients (11 F, 13 M) with infantile feeding disorders from Eastern Switzerland and Zürich have been diagnosed and treated by the multiprofessional team consisting of a child psychiatrist, a pediatric gastroenterologist, a pediatric neurologist, a developmental neurologist, a nutritionist and a speech therapist. All patients and their families are seen in a clinic which is coordinated by a nurse as a case manager.
Results: The feeding disorders of all 24 patients could be classified into one of the six known different types. They all had a full work up by the specialists involved in the team. Out of 15 patients with enteral tube feeding, 11 could be weaned successfully in a limited period of time, 4 are still partially fed by a gastrostomy tube. 4 out of 11 patients were hospitalised for three weeks for the weaning process. 7 could be weaned on an outpatient basis. 12 out of all 24 patients finished their treatment without complications, the rest are still undergoing therapy.
Conclusion: We would like to show that it is of major importance for the management of infants with feeding disorders to investigate and treat them in a multidisciplinary setting with a permanent case manager. The therapeutic outcome, the weaning process and the quality of life of these patients and their families depend on the structured organisation of such a team. We have established such a program for the last two years and were able to treat most patients on an outpatient basis. Our aim is to create a door to door clinic involving all specialists at the same time.

FM14

Neuropsychological sequelae after pediatric traumatic brain injury – prognostic factors during the acute phase

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Introduction: Traumatic brain injury is the most common cause of mortality in childhood and has a high morbidity in survivors. During the recent years, it became more and more evident that neuropsychological problems are frequent, even after minor or moderate traumatic brain injury. Clinical symptoms during the acute phase are prognostic for later neurological outcome. However, their significance for neuropsychological outcome is less well documented.
Methods: 131 patients aged 2 months to 16 years at the time of traumatic brain injury during the years 2001 to 2008 were included into this retrospective analysis. Children with non-accidental trauma or a preexisting neuropsychological diagnosis were excluded. Clinical parameters as Glasgow coma scale [GCS], duration of coma and intubation, duration of impaired consciousness, intracranial pressure, vegetative symptoms, focal neurological symptoms and seizures were recorded. These data were analysed in correlation to detailed neuropsychological assessments (depending age by SON-R, K-ABC and WISC) at follow up 2-12 months after traumatic brain injury.
Results: Initial GCS was 13–15 in 61 patients, 9–12 in 30 patients and 3–8 in 40 patients. Mean duration of coma (GCS ≤8) was 48.9 hours (range 1–560). Focal neurological signs were present in 23 children. Mean IQ at follow up assessment was 100.6 (45–125). Initial low GCS (≤8), duration of coma and intubation were negatively correlated with neuropsy-chological outcome, especially IQ scores. A score of focal neurological symptoms, consisting of cranial nerve dysfunction, decerebration symptoms, ataxia and paresis of limbs correlated most significantly with later adverse neuropsychological performance.
Conclusion: Clinical parameters provide a prognostic factor for later neuropsychological outcome, helping to identify children at risk for neurodevelopmental long-term sequelae and to offer optimal therapy and follow up.

FM15

Bacterial meningitis in the neonate and neurological outcome – experience from the Children's Hospital St. Gallen

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Introduction: The incidence of neonatal bacterial meningitis is up to 0.5/1000 births with a high mortality (5–10%) and a high risk for neurological sequelae. Early onset meningitis is defined as illness before 7 days of age, late onset meningitis occurs after 7 days of age. The predominant pathogens are Group B streptococci and Escherichia coli but other gram-negative bacteria such as Klebsiella spp., Citrobacter spp. and Enterobacter spp. are also, although less commonly implicated.
Methods: We reviewed cases of neonatal bacterial meningitis in the last 5 years at our hospital with special regard to the neurological outcome.
Results: In the last 5 years we had 5 cases of severe neonatal bacterial meningitis: *Early-onset meningitis:* 1 child with pseudomonas meningitis and 1 child with E. coli meningitis, both children required a shunt because of hydrocephalus and both have developmental delay. In addition the child with E. coli meningitis has sensorineural hearing loss requiring a cochlear implant. *Late-onset meningitis:* 1 child with H. influenzae type B who developed cerebral palsy and visual impairment. 2 children with Citrobacter koseri meningitis, both developed ventriculomegaly requiring shunting, developmental delay and in one case, severe epilepsy.
Conclusion: We present 5 cases of severe neonatal bacterial meningitis seen in the last 5 years at the Children's Hospital St. Gallen. All of them developed severe neurological sequelae presenting with developmental delay, cerebral palsy, epilepsy, hearing impairment, visual disorder or hydrocephalus requiring shunting. Early and adequate antimicrobial therapy is essential but often does not prevent neurological sequelae. Neuroimaging is helpful in determining some of the potential complications. Long-term follow up should include monitoring of developmental status, epilepsy, hearing and visual acuity.

Free communications – Haematology/Oncology

FM16

Bleeding risk after tonsillectomy: To screen or not to screen and how?

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Introduction: Bleeding after tonsillectomy remains a feared complication. Most ENT surgeons therefore ask for a preoperative coagulation screening, although a series of recent publications show the poor predictive value of such tests. The value of a detailed bleeding history has been less accurately studied. Aims of this study were: i/ to determine the rate of bleeding after or during tonsillectomy in a wide non-selected pediatric population; ii / to determine the sensitivity, specificity, positive and negative predictive values of a comprehensive bleeding history versus those of the laboratory screening; iii / to suggest future guidelines.
Methods: Prospective study of 420 children successively admitted for tonsillectomy over a period of 30 months in a pediatric university hospital. A detailed questionnaire about bleeding symptoms was filled up preoperatively and each child had usual laboratory screening tests. Surgeons used two different techniques: cold versus diathermic resection of the tonsils.
Results: Per-operative bleedings were relatively frequent (15%) but always moderate (15–70 ml) and expected, being related to the cold dissection technique. No severe per-operative bleed (>70 ml) was observed. Relevant postoperative bleedings were rare (1%). Preoperative coagulation screening and bleeding history were both unable to predict bleeding risk. The detailed questionnaire about bleeding symptoms used in our study showed a very high NPV (98%), similar to the one of the coagulation screening.
Conclusion: As reported by others, bleeding history and coagulation

tests are poor screening instruments and unable to predict which patient will bleed. Our study shows that postoperative bleeds are exquisitely rare in the setting of a completely negative bleeding history, allowing to spare lab testing in the majority of the children. Lab screening remains useful in the context of either positive or incomplete history; if it's negative, the child can proceed safely to tonsillectomy; if not the whole screening should be repeated and if positive again, the operation should be postponed until a specialized haemostasis consultation has been completed.

FM17

Langerhans cell histiocytosis in all its variety. A retrospective single centre evaluation

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Introduction: Langerhans cell histiocytosis (LCH) is a rare disease that can occur at any age, affecting any organ of the body. LCH is a clonal disease, however, until today a clear distinction between a reactive or malignant disease has not been ascertained. We retrospectively analysed the data of all children diagnosed with LCH in our clinic since 1980.
Methods: From January 1980 to December 2008, 46 children were diagnosed with LCH. In 44 pts diagnosis was confirmed by histological examination. According to disease extension at diagnosis, 2 groups were defined: single system and multi system disease.
Results: Median age at diagnosis was 3.5 y (0–15.5 y), 56% male. 32 pts (70%) presented with single system disease with involvement of bone in 90%, two pts showed skin and eye involvement, respectively. 14 pts (30%) presented with multi system disease. These children were younger (median age 1.3 y), and all were treated with chemotherapy. Six of them showed recurrent or progressive disease and were retreated with chemotherapy. One pt received allogenic stem cell transplantation due to multiple relapses. Two children died; a 1.1 year-old-girl prior to diagnosis and a 1.5 year-old-boy with progressive disease under treatment 7 months after diagnosis. Compared to pts with multi system disease, only 10 pts (31%) with single system disease received chemotherapy. The follow-up (FUP) of all living children ranges from 0.1 to 14.3 y. Two are under treatment, 2 and 6 months after diagnosis, and 10 children are still on periodical FUP 0.7–11.2 y after diagnosis.
Conclusion: The presentation of LCH is multifaceted and the course of the disease is often unpredictable. Knowledge of the disease is required for early recognition and intuition for the therapy of the individual child is necessary to avoid over or under treatment. In our series we observed a substantial number of benign courses of disease after biopsy only in single system diseases, and changeful, unpredictable and threatening courses in particular in very young children, who, except in two cases, survived their disease.

FM18

Physical activity levels among Swiss Childhood Cancer Survivors

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Background: There is evidence that physical activity may mitigate or prevent some late effects of childhood cancer.
Aim: To describe physical activity levels in Swiss childhood cancer survivors and compare them to the general population.
Methods: The Swiss Childhood Cancer Survivor Study (SCCSS) sent a detailed questionnaire to all children registered in the Swiss Childhood Cancer Registry between 1976–2003, who had survived ≥5 years, were aged <16 yrs at diagnosis and ≥20 years at the survey and had a valid address (N = 1218). Results were compared with population-based data from the Swiss Health Survey 2002 (SHS) (n = 6517). An index created by the Federal Office of Sport (FOSPO) was used to classify physical activity levels in 3 groups (inactive, partly active, active). Effects of gender, age (20–30 years, 31–40 years) and other potential risk factors on FOSPO were assessed with multinomial logistic regressions in both populations.
Results: Response rates were 73% (886/1218) in the SCCSS (survey ongoing) and 64% in the SHS. 13 % of survivors were limited in activity due to a handicap. This proportion did not differ by age or gender, but was higher in survivors of CNS and other miscellaneous intracranial and intraspinal neoplasms (24%), neuroblastoma and other peripheral nervous cell tumors (20%), and malignant bone tumors (48%) compared to other diagnostic groups (all p <0.01). Among cancer survivors, 48% (95% CI 32–65%) were active, 46% (33–58%) partially active and 6% (2–10%) inactive, compared to 46% (37–55%), 48% (41–54%) and 6% (4–9%) in the SHS. In the multivariate regression, female gender (p <0.01) malignant bone tumor (p = 0.05) and age >30 years (p <0.01) was associated with inactivity or reduced activity.
Conclusion: In spite of their severe illness experience and treatment history, physical activity levels among childhood cancer survivors are similar to the general population. In knowledge of their increased risk for developing cardiovascular disease and obesity, physical activity should be promoted in childhood cancer survivors.

FM19

Protein-energy malnutrition in pediatric solid tumors: clinical approach and algorithm for nutritional support

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Introduction: Protein-energy malnutrition (PEM) occurs up to 50% in pediatric cancer patients treated in developing countries, whereas its prevalence in industrialized countries varies and has been mainly studied in acute leukemia. We report on our approach in evaluating and following pediatric cancer patients since 2 years.
Methods: Patients were considered at low or high risk for PEM according to disease type and stage. At diagnosis and regular intervals during treatment, clinical parameters were evaluated such as height/weight (W/H), body mass index, anthropometric measures, blood chemistry and hormones for growth and bone metabolism were analyzed and bone density assessed by Dual-energy X-ray absorptiometry (DEXA). In case of weight loss >15% or W/H <85%, an algorithm indicated the corresponding nutritional support extending from oral formulas up to percutaneous gastrostomy (PEG).
Results: Forty-three patients were assessed, 17 treated for acute leukemia and 26 for solid tumors. Nine were low-risk and 34 high-risk patients. Two out of 9 low-risk patients needed oral supplements; 12 of 34 high-risk patients received nutritional support, either as oral supplements (3), nasogastric tube (3), oral supplements and tube (2), PEG (2), parenteral nutrition (2). High-risk patients had a lower W/H ratio than low-risk patients, but both increased the ratio during treatment. The mean value of pre-albumin as parameter for subclinical malnutrition was under the lower normal limit at diagnosis and moved into the normal range during treatment, and so was IGF1. Bone metabolism increased during treatment, but without significant effect on bone density.
Conclusion: The described approach to the detection and care of PEM in pediatric cancer patients revealed subclinical malnutrition at diagnosis in the majority of patients and improvement throughout treatment. Thus, PEM is a reality in our setting and nutritional support must be integrated from the very beginning into the therapeutic program.

FM20

Palliative care for children and adolescents in German-speaking Switzerland: A needs analysis across three diagnostic groups

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Introduction: Parents caring for a child with a life-limiting illness are confronted with a multitude of demands and have tremendous needs for support from health care professionals. Research on paediatric palliative care (PPC) in Switzerland is limited. To better develop concepts for PPC services, perceptions of affected families need to be systematically examined. International studies on PPC often include only one group of diagnosis, e.g., mainly cancer. The purpose of this study was to identify the needs of parents caring for a child with a life-limiting illness depending on the diagnostic group.
Methods: Fifteen parents from three equally sized groups of diagnoses (cancer, neurological disorders, and non-cancer/non-neurological conditions) were interviewed. Interview questions regarded various aspects of PPC such as communication with professionals, needs for support in the care of the child, and bereavement support. Eligibility criteria were: active PPC at the time of data collection and child's age between 1–18 years. Families whose child had died no more than two years ago were also included.
Results: Parents of children with neurological and metabolic disorders expressed their needs and concerns primarily in terms of an interdisciplinary care team. In Switzerland, psychologists and social workers in hospitals support children with cancer and their families, whereas children with neurological or metabolic diseases lack this support. Parents of children with cancer emphasized their need for an easy accessible and continuous bereavement care.
Conclusion: Acknowledging the needs of parents caring for a child with a life-limiting illness may lead to a better understanding of the different challenges for families and promote PPC services.
The study was funded by the Swiss foundation Pro Pallium.

FM21

Brief intervention addressing excessive cannabis use in young people consulting family doctors: a pilot study

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Introduction: Switzerland has one of the highest prevalence of young cannabis users in the world. Most these young people are in contact with a family doctor at least once a year. These doctors can create opportunities for discussing health compromising cannabis use. We developed a brief primary care intervention addressing excessive cannabis use (defined as weekly or more frequent use) and piloted its use by family doctors in consultations with young people.
Methods: Ten family doctors and five young people collaborated with the research team in developing the brief intervention and the pilot study. The intervention was based on the guide known as the 5As (assess, advise, agree, assist, and arrange). The family doctors trained the use of the intervention with adolescent actors whose feed-back informed further refinements the study. Young people consulting for any health problem were recruited in the practices before the consultation. They were asked to provide their e-mail or mobile phone contact details for follow-up. Cannabis use and associated psychosocial factors were assessed using a confidential questionnaire administered before the consultation and one month later.
Results: Of 81 young people invited to participate, 78 (70% female) agreed (96% participation). One in seven (13.2%, 95%CI: 7.5–18.9%) used cannabis at least once a week. Data at one month was available for 42% of those who had provided e-mail contact details and 91% of those who had provided their mobile phone number (63% overall). There was a 15% reduction in the number of excessive cannabis users at follow-up. There was also 18% reduction in the number of excessive alcohol users at follow-up. In most cases, the intervention lasted no more than 5 minutes.
Conclusions: This brief intervention shows promise in reducing excessive cannabis use in young people consulting in primary care. Its effects may extend to other substance use. This pilot study has informed the design of a randomised trial to assess the effectiveness of the brief intervention in reducing excessive cannabis and other substance use in young people attending primary care.

FM22

What barriers do boys encounter to talk about sexuality-related problems with health providers?

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Purpose: Although young males also have sexually-related concerns, they are mostly absent from specialized services. Our objective is to assess what barriers prevent young males from talking about sexuality-related problems with health providers.
Methods: In the context of a qualitative study, we conducted 2 focus groups gathering 12 boys aged 17–20. Discussions were triggered through the presentation of 4 vignettes corresponding to questions posted by 17–20 year old boys and girls on www.ciao.ch concerning various sexual dysfunction situations. In order to avoid having to talk about their own experience, participants were asked what they would do in those cases.
Results: Participants declared that they often did not know where to consult, whom to address for help, and where to go anonymously for this type of problem. The main barriers to talk about these issues were shame and embarrassment and the implications for masculinity and pride. Two criteria for consulting a health professional were raised: (1) a problem which is long-lasting and (2) considered as physical (vs. psychological). Many boys do not imagine talking to their GP about sexual concerns because it is not the doctor's role, it lacks anonymity (towards parents in particular), and it is embarrassing. Nonetheless, the neutral status and the nonjudgmental character of a doctor were raised as positive points. Theoretically, a family planning center was presented as the most appropriate place to consult because it gathers professionals who are used to these types of problems, it is free and anonymous. However, in practice, they consider it as mainly intended for girls. Participants strongly valued the Internet as a positive tool to break down barriers for boys to consult in face-to-face settings: a consultation on the Internet was presented as a first step which can help define the seriousness of a problem, whether it is worth consulting for, and where to consult.
Conclusions: Health professionals must communicate a clear message that these issues can be discussed to make it more socially acceptable for boys to disclose. As some participants suggested, a mandatory visit to a health professional for all teenage boys might be necessary as an incentive for future consultations and a way for it to become eventually part of the norm. The Internet and new technologies could be an efficient way to reach male adolescents concerning sexually-related problems.

FM23

Adolescent victims of child abuse and their perpetrators

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Objective: The aim of this study is to describe the adolescent victims referred to the child protection team at the University Children's Hospital Zurich with a special focus on their perpetrators.
Method: Since 2003, the child protection team has registered standardized data on each case. Characteristics are analyzed for the 1'484 referrals between 2003 and 2006 comparing adolescent victims (n = 290) with victims under the age of 12 (n = 1181).
Results: Adolescents were mainly referred for sexual (50%), physical (27%) and psychological maltreatment (14%). In all these types of maltreatment the majority of victims was female (sexual maltreatment: 92%; physical maltreatment: 70%; psychological maltreatment: 67%). Overall, female victims accounted for 81% of the adolescent sample compared to 56% in the sample of children under the age of 12. Twenty percent of the sexually and 14% of the physically maltreated adolescent cases are victimized by peers. The juvenile perpetrators are predominantly male (91%). Juvenile perpetrators also account for 11% of the victims under the age of 12.
Conclusions: The dominance of female victims in adolescent referrals is overwhelming, special prevention efforts are needed. As research on child protection cases at hospitals is still scarce – especially on perpetrators –, we also encourage hospital child protection teams to a more thorough documentation for the support of future analyses.

FM24

Suicide attempt in adolescent in central Valais: need for prevention

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Introduction: Suicide is an important problem in Switzerland, regarding the whole population, starting already at a very young age.
Methods: We collected data about all the children and adolescents hospitalised between 1. 1. 2006 and 31. 12. 2008 for suicide attempt. We also collected data from the network "Réseau Entraide Valais", which aim is early detection of distress, in order to prevent suicide at all age. A report about health of pupils of age 11 to 15 made by the "Observatoire valaisan de la santé (OVS)" from 1994 to 2006 was used to understand the possible causes of distress among children.
Results: 30 adolescents aged from 12 to 15 where admitted because of a suicide attempt: 25 girls and 5 boys, 16 /30 being 15 years old. The younger were 12 years old (4/30). In 15/30, it appeared clearly to be an act of rebellion or a call for help, rather than a real suicide attempt. In 24 cases, the method used was the ingestion of medicines. Most of the cases only required medical monitoring, but in one case the patient had to be admitted to the ICU for respiratory assistance. When asked about the trigger leading to a suicidal act, 22 patients incriminate a problematic relationship with their parents. In 12 cases, we clearly identified a difficult family and social situation (parents affected by psychiatric illnesses, parental conflicts). In the majority of cases, the parents were separated. OVS report shows that about 8% of all pupils feel lonely, about 50% of them find it difficult to speak about their worries to their father, 25% to their mother.
Conclusion: The issue of suicide is a real problem among children in Valais. It is not exclusively a medical issue, but involves the whole society. The "Réseau Entraide Valais" http://www.reseau-entraide-vs.ch/involves the direct environment of persons in crisis, as well as institutions and public associations. The access to these resources and the communication in the network itself should be a priority in the aim of a better prevention of suicide.

FM25

Adolescent friendly health services in detention

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Introduction: The health of young offenders is known to be poor. They are in much need of appropriate primary care and mental health services. Dedicated health services in juvenile detention centres are recommended by the World Health Organisation, yet few such structures exist in European youth detention centres. The objective of this paper is to describe the medical service of a youth detention centre in Geneva, highlighting the role this service has in lessening the barriers to care for adolescents during and following their detention.
Method: Qualitative and descriptive analysis of the service.
Results: The service employs a nurse, a general practitioner, a psychologist and two psychiatrists and is part of a network of youth services within Geneva University Hospitals. It provides 3000 consultations a year to approximately 400 adolescents aged 11 to 18

years (20% female). In accordance with the rights of detainees, a confidential health consultation is offered to all detainees within 48 hours of detention (90% attend). The nurse does the triage to organise rapid referral for a general medical or psychiatric consultation if needed. For those who are only detained a few days, the focus of the nurse, medical and psychiatric consultations are on the identification of health problems and the provision of information on appropriate health services for these health problems in the community. The young offenders who stay for more than two weeks have a more detailed medical and mental health assessment. They also attend health education sessions. In addition to the care offered for the identified health problems emphasis is placed on explaining the right to confidential health care and orienting the adolescents towards adolescent friendly health services in the community following their detention.
Conclusion: For many young offenders contact with the medical health service during detention is the first regular health care contact they have had in years. Providing adolescent friendly health services and guidance in detention has the potential to improve these adolescents' access to care beyond detention as they return into the community.

Posters – Neonatology

P001

Incidence and Severity of Nasal Trauma due to Continuous Positive Airway Pressure in Neonates: a Prospective Study

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Introduction: Nasal Continuous Positive Airway Pressure (nCPAP) has become of increasing importance in the management of neonatal respiratory distress over past decades. Nasal injury is a complication commonly observed in practice, associated with discomfort, infection and functional or esthetical sequelae.
Methods: The objective of this prospective observational study was to describe incidence, severity and evolution of nasal trauma due to nCPAP in neonates. All neonates admitted in our Neonatal Intensive Care Unit between January 2002 and December 2007 who were supported by nCPAP (Infant Flow Driver) were included. The nose of the patients was regularly inspected and lesions were reported. We pre-defined 3 stages of nasal injury following the general standardized classification of the decubitus lesions from the National Pressure Ulcer Advisory Panel (NPUAP): I persistent erythema not fading, II superficial ulceration, III necrosis involving entire thickness of skin.
Results: 989 neonates were enrolled. Mean gestational age was 33.6 weeks (SD 3.7), mean birth weight 2142 grams (SD 840), with a 1.4 male/female ratio. Nasal injury was reported in 420 (42.5%) patients, corresponding to a general incidence of 30.6 lesions/1000 days of nCPAP treatment. Lesions were respectively of stage I, II and III in 371 (88.3%), 46 (10.9%) and 3 (0.7%) of cases. Incidence and severity of lesions were inversely correlated with gestational age and birth weight. Most of the lesions (>90%) appeared during the first 5 days of nCPAP. Progression of lesions into more severe stages was observed. Persistent visible scars were present in two cases.
Summary: Nasal trauma secondary to nCPAP application is a very frequent iatrogenic complication in neonates, especially in immature infants. Intervention studies are needed for development of strategies of prevention and management.

P002

Evidence for a role for M-ficolin in the neonatal period: association with need for mechanical ventilation and mortality in premature infants with necrotising enterocolitis

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Objective: Necrotising enterocolitis (NEC) causes significant mortality in premature infants. We have recently reported elevated MASP-2 levels in infants with NEC, suggesting that the lectin pathway of complement activation may be involved in the pathogenesis of NEC. M-, L- and H-ficolins recognize microorganisms and activate MASP-2, but their role in host defense is largely unknown. This study investigated whether ficolin concentrations are associated with NEC.
Study design: Case-control study including 30 premature infants with NEC and 60 matched controls. M-, L- and H-ficolin were measured in cord blood using TRIMFA. Logistic regression analysis adjusted for gestational age and birth weight was performed.
Results: Of the 30 NEC cases (median gestational age, 29.5 weeks), 12 (40%) were operated and 4 (13%) died. No difference regarding ficolin concentrations was found when comparing NEC cases versus controls ($p > 0.05$). However, infants who died of NEC had significantly lower M-ficolin cord blood concentrations than NEC survivors (for M-ficolin <300 ng/ml; multivariate OR 12.35, CI 1.03–148.59, $p = 0.048$). M-, L- and H-ficolin concentrations were positively correlated with gestational age ($p < 0.001$) and birth weight ($p < 0.001$). Infants with low M-ficolin required significantly more often mechanical ventilation after birth (multivariate OR 10.55, CI 2.01–55.34, $p = 0.005$).
Conclusions: M-, L- and H-ficolins are already present in cord blood and increase with gestational age. Low concentration of M-ficolin was associated with higher NEC-associated fatality and with increased need for mechanical ventilation. Future studies should assess whether M-ficolin is involved in neonatal disease.

P003

Magnesium Chloride in Infants with Persistent Pulmonary Hypertension: The Berne Experience 1997–2007

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Introduction: Persistent pulmonary hypertension of the newborn (PPHN) occurs in 0.43 to 6.8 per 1000 live births. It can occur as a primary condition or be secondary to meconium aspiration, respiratory distress syndrome, infection, congenital heart disease or diaphragmatic hernia. Magnesium chloride (MgCl) is a potent vasodilator and has been used to reduce the high pulmonary arterial pressure for years despite of the current lack of published evidence.
Methods: Tertiary single centre retrospective study including all neonates treated with MgCl for PPHN in the 12-bed Paediatric Intensive Care Unit (PICU) between 1997–2007.
Findings: 89 patients with a mean gestational age of 38 3/8 weeks (29 3/7–42 1/7 weeks) were treated for PPHN between 1997 and 2007. Main diagnoses were congenital heart disease (37%), meconium aspiration (27%) and congenital diaphragmatic hernia (11%), only 7% were diagnosed with idiopathic PPHN. After a mean loading dose of 0.97 mmol/kg/h for 20 minutes, a continuous infusion of 0.27 mmmol/kg/h was administered for a mean time of 30.1 hours (3–74). Mean Magnesium blood concentration after 12 hours was 3.3 mmol/l. 19.1% of all patients received additional inhaled nitric oxide to treat PPHN. Mean PaO2 increased from 64.2 mm Hg before treatment to 82.4 mm Hg after 12 hours. Mean airway pressure remained stable with 14.5 cmH_2O. Inotropic score was 9.7 and 8.9 respectively. Oxygen index (OI) decreased from 16.3 to 9.7. 70.8% of all patients treated with MgCl survived to hospital discharge.
Discussion: This study describes the largest group of neonates treated with MgCl for PPHN in medical literature. The preliminary data suggest that MgCl is a feasible and save treatment for PPHN without important adverse effects; there were especially no major hypotensive events. To augment the evidence of MgCl's benefits, randomized controlled studies should be performed.
Conclusion: In our experience MgCl is a relatively safe and low-cost medication that should make part of the armamentarium of the modern treatment of PPHN.

P004

Fitness to fly in ex-premature babies with bronchopulmonary dysplasia: a "home made" ability test

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Introduction: Commercial long distance flights expose travelers to hypoxia. This hypoxia is even more challenging in patients with pre-existing lung disease. This question is of particular importance for ex-premature babies with bronchopulmonary dysplasia (BPD) and children with cystic fibrosis. Aircraft cabins are pressurized to an altitude of 1500–2400 m above sea level, making the oxygen partial pressure being 15 kPa, or equivalent to a FiO_2 of 15–16% at sea level. Standardized tests for fitness to flight are either unreliable (normobaric hypoxia) or technically challenging (hypobaric hypoxia).
Patient, method and result: 5-month-old girl, ex-premature (30 weeks gestational age) with mild bronchopulmonary dysplasia without

need for supplemental oxygen at hospital discharge. Because of the need for a trans-atlantic flight, we decided to test her tolerance to hypobaric hypoxia. We did this by monitoring her transcutaneous oxygen saturation during a car trip of 120 minutes duration from Sion (512 m), the goal being to reach the Grande Dixence dam (2365 m). A mobile oxygen source was provided for security reason. The Grande Dixence dam could not be reached, because of progressivly lowering of saturation levels (<92%) between 1800 and 2000 m. A moderate (87%) but persistent (>5 min) desaturation appeared when reaching 2000 m, prompting for oxygen administration and return to the Rhône valley level. Because of this result, we decided to monitor her oxygen saturation during the flight, and provide supplemental oxygen if needed. Our patient received oxygen from arrival at cruising altitude for the whole flight, which she completed without problem.
Conclusion: In countries with high mountains reachable by car, like Switzerland, a trip to sufficient altitude while monitoring the transcutaneous oxygen saturation allows to perform a non expensive hypobaric hypoxic fitness to flight test. A "false" positive test due to the position in the car safety seat (as described for preterm and term newborns), however has to be excluded by a journey in the same condition and of same duration without change of altitude. A larger study is needed to define more accurately the protocol and to measure positive and negative predictive values.

P005

Current State of the Neonatal Pulse Oximetry Screening for Congenital Heart Defects in Switzerland

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Introduction: Timely recognition and treatment of congenital heart defects (CHD) is crucial for a good outcome, but can be challenging in neonates because of an initial lack of clinical signs. Pulse oximetry screening can improve the detection rate of CHD. It was recommended in 2005 by the Swiss Societies of Neonatology and Paediatric Cardiology as follows: Newborns in whom postductal transcutaneous pulse oximetry is below 90% or persists for several hours below 95% need to undergo an echocardiogram to rule out CHD. It is not known whether this screening is actually practised, and if there are methodological difficulties that prevent nationwide screening or that have made changes to the recommendations inevitable.
Methods: A questionnaire about the adopted in-house policy of pulse oximetry screening was sent to all Swiss maternity units in summer 2008. Results were set in relation to the total number of newborns in Switzerland in 2007.
Results: Response rate was 100% within 10 weeks. 103 of the 136 maternity units (76%) actually perform the screening as recommended, and 85% of all neonates in Switzerland are screened. Maternity units linked to a paediatric unit showed significantly higher screening rates ($p = 0.019$). The screening rate in "birthing units" (Geburtshäuser) was significantly lower than that in hospitals (3/20 versus 100/116; $p < 0.001$). Reasons for not accomplishing the screening are: Financial factors (14), the confidence that clinical signs are sufficient to detect CHD (11) and missing in-house guidelines (4). No adaptations needed to be made to the screening protocol.
Conclusion: The recommendations regarding the pulse oximetry screening were easily transferred into clinical routine by most of the Swiss maternity units within the last 3 years. The negative outlook of maternity units which are not performing the screening is not based on evidence but on structural and financial considerations. Further efforts are needed to convince all maternity units in Switzerland of the need for the screening.

P006

Systematic hearing screening test with the evoked oto-acoustic *emissions* (TEOAEs) of the newborns in 2008

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Introduction: Significant bilateral hearing loss is present in 1 to 3 per 1000 newborns. Screening by high-risk is less expensive but can identify a maximum of 40 to 50% of newborns with significant congenital hearing loss. Systematic screening of all newborns can detect a minimum of 80% of newborns with significant hearing loss. The method used has a false-positive rate between 2 to 3%.
Objective: As a significant, undetected bilateral hearing loss will impede speech, language, and normal cognitive development, the goal of the screening is early detection with appropriate intervention no later than 6 months after birth.
Methods: Implemented testing methods consisted by measuring evoked otoacoustic emissions (TEOAEs), using Echoscreen®, performed in all newborns in their first 2 to 3 days of life in the nursery. If a first and second test failed, a third test was done. If the third test failed, the child was referred for audiologic assessment 4–6 weeks later. Other clinical examinations like transient evoked otoacoustic emissions, auditory brainstem responses, tympanometry and behavioural audiometry were done to confirm the hearing loss.
Results: Of 1600 newborns born in our hospital in 2008, 1597 were tested for TEOAE's (99.8%). 1589 newborns (94.5%) had a normal TEOAE's test, 1 showed bilateral permanent hearing impairment (0.06%), 3 (0.18%) are still studied. 43 newborns were submitted to a second test, and 17 to a third test, while 4 (0.25%) had not done the subsequent test. The false-positive rate was 2.2%.
Conclusions: Bilateral permanent hearing impairment was proved in 0.06 % of newborns. Efforts should be made to avoid losses of follow-up and reimbursement of the costs of screening by the basic insurance.

P007

Prevalence of exclusive breastfeeding at 6 months in preterm infants (≤35 0/7 weeks gestational age) at the Neonatal Clinic, University Hospital Zurich, Switzerland

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The World Health Organization (WHO) recommends exclusive breastfeeding for 6 months. A national Study of 2003 has shown that 20% of Swiss babies had reached this goal. There are no comparable data for preterm infants. In a retrospective study in 2002 and 2005, we examined 125 preterm infants (≤35 0/7weeks gestational age) regarding prevalence of exclusive breastfeeding at 6 months non-corrected age. Seventeen percent of the infants were exclusively breastfed at 6 months. In the cohort of 2002, we compared the two extremes: exclusive breastfeeding for 6 months (14 infants) and weaning during the first 1–2 weeks at home (18 infants). There was no significant effect on breastfeeding duration regarding the following factors: infant's gestational age and birth weight, mother's age, tube feeding, oxygenation, CPAP-therapy, time of first kangaroo session and time of infant's first feeding (breast or bottle). The only parameter which might predict a longer duration of breastfeeding is 100% breast milk feeding at discharge from the hospital.

P008

Unusual presentation of group B streptococcal (GBS) infection in neonates and infants

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Introduction: Group B streptococcus (GBS) is the most frequent bacterial pathogen in neonates. GBS infection is classified by the age at onset: early onset (0–6 days), late onset (7–89 days) and late, late onset (>90 days). The incidence of early onset GBS disease has declined with maternal intrapartum chemoprophylaxis, while the incidence of late onset GBS has not changed. Late onset disease presents mainly as bacteraemia, meningitis, pneumonia or arthritis/osteomyelitis.
Methods: Presentation of 3 unusual cases of late onset or late, late onset GBS infection.
Case 1: A 3-week-old girl presented with fever (39.2 °C), rhinitis and mildly floppy tone. Initial blood tests showed WBC of 2.5 G/l and CRP of 7 mg/l (24 h later 11.4 G/l, 239 mg/l). Amoxicillin and gentamycine was started. 6 hours later she developed right third cranial nerve palsy. CSF analysis revealed pleocytosis and GBS consistent with meningitis. Blood culture grew GBS. Intravenous immunoglobulin (IVIG) was added. The girl recovered completely. Development was normal (EQ 102) at 6 months of age.
Case 2: A 10-week-old girl suffered from uncomplicated pyelonephritis. As urine culture grew GBS, ceftriaxone was switched to amoxicillin with rapid recovery.
Case 3: A 5-month-old boy with pyelonephritis presented with low plasma sodium (116 mmol/l), high potassium (5.8 mmol/l) and normal creatinine. Urine culture grew GBS. Ultrasound showed normal kidneys and dilated ureters. Cystography revealed a left grade V vesico-ureteric reflux. Ceftriaxone was switched to amoxicillin. Within 5 days, all findings normalised, suggesting transient pseudohyopoaldosteronism due to GBS.
Conclusion: Early onset GBS infection is a well known disease in neonates whereas late onset and late, late onset GBS infection are less common and present with a broad clinical spectrum as demonstrated in this 3 cases. Up to 20–30% of neonates with late onset GBS meningitis have a preceding infection of the upper respiratory tract as in our case. Focal neurologic deficits are rare complications of meningitis and known to be associated with adverse neurologic outcome. IVIG has been suggested as a adjuvant therapy in early onset GBS sepsis; whether it had any impact on the course in our patient with meningitis remains speculative.

P009

Peripartal acquired Campylobacter Jejuni infection: an unusual cause of colitis in the newborn

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Background: The presence of bloody stool in the newborn is a rare and serious sign, and can be secondary to infections, necrotizing enterocolitis, volvulus, invagination or cow milk allergy. Campylobacter Jejuni (CJ) has rarely been reported as a cause of infectious colitis in the neonatal period. We here present such a case.
Case presentation: The patient was born at term after an uneventful pregnancy. Because of prolonged rupture of the membrane, the mother received one dose of antibiotics. Post-natal adaptation was excellent (Apgar 9-10-10). On day 2 of life, the newborn was found to be markedly lethargic, febrile (38.1), with copious bloody diarrhea. Physical exam showed a febrile lethargic jaundiced newborn. BP was 68/48 mm Hg. After volume resuscitation she was immediately started on clamoxyl-garamycine. Blood/CSF cultures were negative, but stool cultures grew CJ, sensitive to gentamicine and erythromycin. On further questioning, the mother had an episode of febrile gastroenteritis with watery diarrhea less than 2 weeks before delivery, but she then did not seek medical advice. After delivery, the mother had normal stools, and cultures were negative. On clarythromycin, the infant fully recovered with no relapse.
Discussion: CJ infection in the neonatal period is rare. It has been associated with maternal and fetal death, premature delivery, and infection in the fetus or newborn. Although typically sensitive to macrolide antibiotics, CJ is often sensitive to aminoglycosides, as in our patient and in reported cases. If recognized early, this uncommon neonatal infection should respond to a neonatal sepsis treatment, and carries a good prognosis. Outbreaks of CJ infection in the NICU have been described, and such infection should be reported to the infection control authorities.

P011

Opsismodysplasia in a newborn: Case report of a spondyloepimetaphyseal dysplasia

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Introduction: Opsismodysplasia is a spondyloepimetaphyseal dysplasia caused by a defect of chondroosseous transformation, with only about 40 cases previously published. The inheritance is thought to be autosomal recessive, no causative gene has yet been identified. It is characterized by consistent clinical signs including craniofacial abnormalities, rhizomelic micromelia and muscular hypotonia. The diagnosis is mainly based on specific radiological findings. The clinical outcome is highly variable, depending on the severity of the phenotype; however, neonatal lethality is common.
Case report: We report on a term boy, first-born to a healthy, non-consanguine somalian couple. Prenatal ultrasound examination at 30 weeks of gestation demonstrated a shortening of both femora. At birth, the boy presented with markedly reduced response to stimulation, irregular breathing and reduced muscle tone. Weight was 2710 g (<3rd percentile), length 45 cm (<3rd percentile). Clinically, he showed dysmorphic features such as frontal bossing, broad and depressed nasal bridge, short nose with anteverted nostrils, long philtrum, hypertelorism, low set ears, and large fontanelles. Limbs were short, in particular hands and feet. Chest was narrowed and abdomen prominent. Skeletal survey revealed the characteristic findings for Opsismodysplasia: a major delay in epiphyseal ossification, shortening of tubular bones, especially those of metacarpals, metatarsals and phalanges, platyspondyly, square shaped iliac bones with lateral and medial spurs. Further investigations such as echocardiography, ultrasound of brain and abdomen as well as ophtalmological examination were all normal. Severe muscular hypotonia caused feeding problems and necessitated nasogastral tube feeding. Although the boy showed impressive thoracic narrowness, he was eupnoeic. The further clinical course was so far uneventful. Investigations on chromosomal and microarray analysis are not completed yet.
Conclusion: Opsismodysplasia is characterized by a distinct phenotype with specific radiological findings. Awareness of this disorder and clinical follow-up documentation is of utmost importance to understand the natural course of this rare skeletal dysplasia. More data are needed to counsel parents regarding recurrence risk, prenatal diagnosis and prognosis.

P012

Valsartan fetopathy: a case report

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Background: ACE inhibitors and angiotensin II receptor type 1 antagonists (sartans) are contraindicated during pregnancy due to impaired renal development resulting in tubular dysgenesis. The wide spectrum of manifestations ranges from death as a result of severe complications of renal failure to reversible effects on amniotic fluid after drug withdrawl as well as late preterm and term gestations with only minor problems during neonatal period and favorable outcome concerning renal function.
Case report: At 35 weeks of gestation a 34-year old woman who received valsartan during whole pregnancy for essential hypertension presented with preterm labour and complete anhydramnion. The spontaneously delivered boy showed typical signs of sartan fetotoxicity including neonatal anuria, enlarged hyperechogenic kidneys, arterial hypotension, limb contractures as well as a narrow chest and skull bone hypoplasia. There were no signs of respiratory distress and lung hypoplasia could be excluded by chest X-ray. Volume expansion, dopamine infusion and furosemide administration resulted in a normalization of urine output within 48 hours. Peritoneal dialysis could be avoided. Maximum serum creatinine was 245 µmol/l and decreased to age-related normal values within 14 days. On follow-up controls renal function remained stable.
Conclusion: Intake of angiotensin II receptor antagonists during pregnancy results in an interdisciplinary challenge for perinatal medicine. Women who become pregnant need to be changed to a different antihypertensive drug class. Due to an unpredictable course patients need to be treated and followed in neonatal units with adequate pediatric nephrological expertise.

P013

Fatale cerebral hemorrhage caused by Vitamin K deficiency

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Introduction: Bleeding due Vitamin K defiency (VKDB) in the neonate has been largely but not completely been eliminated by recommendations for the application of Vitamin K after birth. More recently, in 2003 the recommendations were adapted to the current use of 2 mg Konakion orally at the age of 4 h, 4 days and 4 weeks after birth. The Swiss Pediatric Surveillance Unit (SPSU) registers all cases of VKDB including as far as ascertainable the age of onset and possible factors contributing to the hemorrhage. We report a case of a deliberate non-use of Vitamin K by the parents resulting in a profound intracebral hemorrhage followed by a severe neurological outcome.
Case report: A 7 week old boy presented acutely on the emergency departement with hematemesis and pallor. On admission, he was in a reduced state, the fontanelle was bulging and the left pupil was enlarged and not reactive to light. Ultrasound examination and CT scan revealed a large subdural bleed with midline shift and partial herniation of the temporal lobes. Coagulation studies revealed a very low quick value and a prolonged PTT time. There were no signs for hepatic insufficiency and the coagulation defect resolved rapidly following substitution with Vitamin K and fresh frozen plasma. On further enquiry, the parents reported that they had refrained from giving Vitamin K after birth to protect their child from "unnatural chemicals". The child underwent emergency operative decompression of the subdural hematema and could be extubated 3 days later. He developed severe neurological sequela with seizures, muscular hypertonia and pathologic electrophysiological results.
Conclusion: Although it is quite rare, hemorrhage due to Vitamin K deficiency may have serious complications, especially if cerebral bleeding occurs. This reinforces the importance of informing the parents and encouraging the application of Vitamin K after birth. It ist to be expected that the SPSU registry will throw more light on the incidence and on the circumstances surrounding Vitamin K deficiency bleeding in Switzerland.

P014

Know it and see it – disruption of the brachial plexus root

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Introduction: Palsy of the brachial plexus is a common congenital injury (Incidence: 1 per 1000 births per year). In nearly 90% of cases spontaneous recovery can be observed. It still is difficult to predict to which degree the function of the arm will be restored. Therefore, invasive diagnostic procedures – like contrast agent imaging – might be necessary. In our case magnetic resonance imaging could reveal the extent of the lesion, whereas ultrasonography was of no diagnostic value.

Case: Female newborn, spontaneous birth in 42nd week of gestation. Difficult delivery because of macrosomia (birth weight 4540 g), APGAR score 0/4/5, arterial umbilical cord pH 7,27. Cardiopulmonary reanimation because of neonatal asphyxia. Therapeutic hypothermia and urgent referral to the NICU. Even though the baby did recover quickly from asphyxia, she seemed to neglect her right arm completely. Additionally, on neurologic examination there were no spontaneous arm movements and no Moro reflex or palmar grasp on the right side. Therefore the MRI – planned to detect possible cerebral sequelae of asphyxia – was extended to the cervical spine. The scan revealed an intraspinal, extramedullar bleeding on level C4-Th2 which had not been identified by ultrasound. The bleeding was interpreted as indirect sign of a disruption of the upper and lower brachial plexus. Although MRI showed marked regression of the findings within one week, the clinical course still was sobering: despite immobilization and daily physiotherapy, the baby did not move her right arm at all. Additional electrophysiological studies (electromyography and nerve-conduction) three weeks afterwards showed no response to external stimuli. This confirmed the presumptive diagnosis of disruption of the brachial plexus. Therefore, neurosurgical options had to be considered.
Conclusion: The degree of brachial plexus lesions cannot be evaluated by ultrasound. MRI is a valuable non-invasive method to gain detailed information about the underlying lesion and should be considered if there is no recovery of the palsy within one week.

P015

Waving Hand & Broken Head

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Introduction: According to Levy et al. (2005) approximately 1.5% of caesarean sections performed involve a deeply impacted head of the newborn. Often they follow a complicated 1st stage of delivery, including high doses of oxytocine.
Case: After an uneventful pregnancy at 40 weeks gestational age an infant girl was born as the first child of a healthy 27-year G1P1. Since spontaneous delivery became difficult and the CTG showed repeated fetal decelerations, the decision for urgent secondary caesarean section was made. Obstetricians reported that the fetal head had become wedged in the pelvis and was thus markedly complicating the labour. Initially, the newborn was depressed but adapted well in the postnatal course. APGAR score was 6/9/10, umbilical artery pH 7.24, birth weight 3505 g. However, after three uneventful days the girl showed intermittent myocloni of the left hand aggravated by distress, i.e. hunger. The baby finally was referred to our neonatal Unit for evaluation of these possible seizures. On physical examination, the girl appeared to be in a good condition – apart from two impressive cephalic haematomas and intermittent myocloni of the left hand. Initial cerebral ultrasound revealed a impression fracture of the skull and a large epidural haematoma resulting in a slight midline shift. Cerebral perfusion still was normal in the ACA. In the further course, a MRI-scan revealed several lesions in the brain parenchyma and the decision for neurosurgical intervention was made to reduce the apparently still increasing pressure affecting brain tissue. On operation, the skull fracture was found to be dislocated towards the brain, consisting of multiple fractured pieces. After decompression of the haematoma and remodelling of the skull, the further clinical course after operation was uneventful and the girl was discharged home soon after. The patient has been seen on a regular basis since then.
Four months after neurosurgical intervention skull X-Ray showed appropriate ossification and no evidence of a possible osteogenesis imperfecta was detected. The girl reached her neuromotor milestones and had no further focal seizures.
Conclusion: Even caesarean section might be traumatic. Neonatal skull fractures are rare and represent an interdisciplinary challenge. In our case, the *"waving hand"* fortunately gave us the hint to look for a *"broken head"…*

P016

Vagal bradycardia at term

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We discuss the case of a newborn boy presenting well into term with severe bradycardic events sinus pauses up to 5.4 seconds. Sinus bradycardia below 80 bpm and sinus pauses or asystole of more than 2 seconds are considered pathologic at term. After exclusion of specific causes, the diagnosis of vagal hyper-reflectivity (VHR) was retained, a state caused by unbalance between sympathetic and parasympathetic activity, the latter overriding the former. It is thought to be a functional and transitional anomaly of the sympathetic and vagal tone during the first month of life and may lead to prolonged monitoring and delayed hospital discharge. This form of rare bradycardia can be treated with atropine and allowed in our case immediate resolution of events and safe discharge from hospital. When VHR is diagnosed, atropine is the treatment permitting resolution of symptomatic episodes of bradycardia and early and safe discharge from hospital

P017

Risk for failure of oral treatment in infants and children with febrile urinary tract infection

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Background: Febrile urinary tract infection is one of the most common serious bacterial infections in pediatric patients. It is associated with relevant morbidity during the acute phase and risk of long-term renal sequelae. Therefore, a rapidly efficacious treatment is mandatory and until recently, at least initial parenteral antibiotic treatment has been advised. However, based on newer studies, Swiss recommendations consider an exclusively oral treatment using a third-generation cephalosporin in patients older than six month. To date, no data are available estimating the potential rate of patients necessitating reevaluation or hospital admission because of delayed success or failure of such oral outpatient treatment.
Methods: Retrospective chart-review of patients older than six months hospitalized (January 2003 – December 2007) with suspicion of febrile urinary tract infection.
Results: One hundred-seventy-four patients (median age 17, range 6.5–189 months; 82% girls) were included. Temperature at admission was 38.8 ± 1.2 °C, leucocytes and C-reactive protein were 20.0 ± 6.9 G/l and 128 ± 87 mg/l, respectively. *E.coli* were found in 144, *Enterococcus* spp in 8 (4/8 as single pathogen); *Klebsiella* spp in 6, and *Proteus* spp. in 2 patients. Antimicrobial resistance to third-generation cephalosporins was found in nine (*Enterococcus* spp, n = 8 and *Klebsiella pneumoniae*, n = 1). Patients remained afebrile after 37.5 ± 28.1 hours with 45 patients (26%) still presenting fever on third day. Persistence of fever was significantly associated with higher CRP and leucocytes at admission ($p < 0.01$). Fifteen (9%) and 3 (2%) patients suffered from repetitive vomiting (≥ twice/day) during the first and second day, respectively. Patients with vomiting also had a tendency (not significant) towards higher CRP at admission.
Discussion: The present data support the potential for safe and efficacious oral antibiotic treatment in an outpatient setting in the majority of children with febrile urinary tract infection aged six months and older. However, in the present cohort, a substantial number of patients would have needed reevaluation because of persistent fever and some might have risked inappropriate treatment because of vomiting. Patients with higher inflammatory signs at admission seem to be at particular risk for delayed response to treatment. Additionally, a small number of patients were at risk of treatment failure because of antimicrobial resistance. Therefore, urine culture prior to initiation of empiric treatment is still strongly recommended.

P018

Transient secondary pseudohypoaldosteronism in four infants with obstructive uropathy

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Introduction: Pseudohypoaldosteronism (PHA) is characterized by renal tubular resistance to aldosterone resulting in hyponatremia, hyperkalemia and metabolic acidosis. There are primary and secondary forms of PHA. The primary form can be divided in two different forms: type I and type II. Secondary PHA has rarely been described in infants and children with acute urinary tract infection and/or obstructive uropathy.
Case reports: We report 4 patients seen in our hospital between 2000 and 2009 with secondary PHA due to urinary tract infection and/or obstructive uropathy:

P.	Age	Sex	Symptoms	Na	K	Urine culture	Type of uropathy
1	2 mo	m	poor feeding, vomiting	115	6.8	Beta-häm. streptococcus	uroteropelvic junction (UPJ) obstruction
2	2 mo	f	vomiting, dehydratation	114	6.5	Staph. aureus	left duplex kidney; upper pole hydronephrosis/-ureter w/o anatomical obstruction
3	6 mo	m	vomiting, poor feeding	107	6.9	Hafnia alvei	UPJ obstruction, urinoma after pyeloplasty
4	5 mo	m	fever, poor feeding	119	5.6	E. coli	ureterovesical junction (UVJ) obstruction

All patients presented with hyponatremia, hyperkalemia (see table, results in mmol/l) and metabolic acidosis. After treatment and sodium substitution hyponatremia, hyperkalemia and metabolic acidosis resolved in all cases.
Conclusion: Hyponatremia and hyperkalemia due to various causes, including acute renal failure and secondary PHA, may be associated with obstructive uropathy with or without infection. Secondary PHA mimicking acute Addison crisis arrises from partial lack of responsiveness of the renale tubule to endogenous mineralocorticoids. Given our experience with 4 patients in 9 years this entity seems to occur more frequently than expected from literature.

P019

Acute renale failure in a child with pneumonia, volume depletion and (ab)use of Ibuprofen

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Background: Non-steroidal anti-inflammatory drugs (NSAID) are increasingly popular in hospital medicine and general practice. The vast majority of healthy children who ingest therapeutic doses of NSAID for a limited duration tolerate them without any significant adverse effects. However, the risk of renal toxicity is potentially increased in situations where there is stimulation of the renin-angiotensin system such as with volume depletion or in preexisting chronic renal disease.
Case: We present the case of a previously healthy 9 year old boy with a history of a febrile respiratory illness over 10 days with reduced food and fluid intake. Because of fever and pain, he was treated with Ibuprofen at a therapeutical dosing. At admission, he was in reduced general condition and in respiratory distress. A pleuropneumonia was diagnosed with a chest X-ray. Together with oliguria, he showed a reduced kidney function with an increased creatinine of 220 mmol/l and an urea of 38 mmol/l without *electrolyte dysbalance* or acidosis. Urinanalysis revealed proteinuria and red blood cell casts compatible with tubulointerstitial nephritis. Renal ultrasound was normal, ANA and ANCA were negative, immunoglobulins and complement proteins C3, C4 were normal and the direct coombs test was negative. After intravenous rehydration, antibiotic therapy and discontinuation of NSAID, renal function recovered completely within 36 hours.
Conclusion: In an era of widespread popularity of NSAID use in children, awareness of the possible adverse renal effects of NSAID is needed, especially in children with complex medical problems or with volume depletion. The number of episodes with renal dysfunction is likely to be underestimated and underreported because the kidney dysfunction is often mild and resolves spontaneously. In situation of dehydration, pre- existing renal impairment, concomitant therapy with nephrotoxic drugs, paracetamol should be used as first choice anti-pyretic treatment and the use of NSAID should be avoided.

P020

Nephrotic syndrome and headache: think about thrombosis of cerebral vessels

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Background: Nephrotic Syndrome (NS) induces a hypercoagulable state with a consequently increased predisposition of thrombosis. The prothrombotic tendencies are due to imbalance between thrombotic and antithrombotic factors. Low molecular weight proteins as antithrombin III, protein C and S are excreted in urine. Due to compensatory protein synthesis in liver high molecular weight proteins like fibrinogen are increased. Other contributing factors include volume depletion, platelet hyperaggregation, immobilization and steroid therapy. The incidence of thrombosis in children with NS is estimated to be less than 10%. Commonly affected vessels for thrombosis include deep leg veins, caval and hepatic veins and cerebral sinuses.
Case: We report a 4,5 year old girl suffering from a first episode of a steroid-sensitive NS under treatment with prednisolon (60 mg/kg/m^2 daily). 7 days after beginning of steroids, she presented to the emergency department because of severe frontal headache. There was no impairment of consciousness, no seizures or cranial nerve deficits. Bilateral papillary edema was observed. Proteinuria had already improved, demonstrating a sensitivity to steroids. However plasma albumin was still very low (7 g/l). Computed tomography showed a partial thrombosis of the superior sagittal sinus reaching the right transverse sinus. Immediately, intravenous heparinisation followed by low molecular weight heparin treatment was started. The girl recovered clinically within a few days without any obvious neurologic sequelae.
Conclusion: Thromboembolic complications in children with NS are rare but potentially life threatening with significant morbidity in case of delayed diagnosis. In children with NS and new onset of headache, sinus venous thrombosis should always be considered and rapidly looked for by neuroimaging studies. Immediate anticoagulation might help to avoid neurologic sequelae.

P021

Xanthogranulomatous Pyelonephritis – a reason for anemia, hypertension and anorexia in childhood

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Introduction: Xanthogranulomatous Pyelonephritis is a rare, severe and atypical form of chronic renal parenchymal inflammation in childhood. The manifestation is non-specific and by mimicking neoplastic renal diseases the preoperative diagnosis can be difficult.
Case report: We report a 16-month-old boy who presented with paleness, anorexia, fever, arterial hypertension and an unilateral tender palpable abdominal mass. Laboratory investigations showed anemia, leukocytosis with left shift, elevated CRP and pyuria with growth of Proteus mirabilis in the urine culture. Ultrasound revealed an enlarged right kidney with abnormal parenchymal structure, hypoechogenic areas and calcifications. CT-scan demonstrated multiple low-density fluid-filled calices, hydronephrosis and calculi in a massive enlarged kidney. Functional examination in late phase of CT showed absent excretion of contrast medium. We diagnosed Xanthogranulomatous Pyelonephritis preoperatively, based on clinical and radiological features. The child was first treated by antibiotic therapy, blood-transfusion and antihypertensive therapy, followed by transperitoneal complete nephrectomy. The histopathological findings confirmed the diagnosis of Xanthogranulomatous Pyelonephritis.
Conclusion: Xanthogranulomatous Pyelonephritis is a rare and uncommon disease in childhood, but should be considered in children presenting with chronic infection, anemia, anorexia or hypertension. Radiological examination may allow a preoperative diagnosis. Total nephrectomy is the treatment of choice, leading to a good prognosis in affected child.

P022

Treatment of accidental digital injection of adrenaline from an auto-injector: case report and literature review

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Introduction: Adrenaline (epinephrine) auto-injectors are used for treatment or prevention of severe allergic reactions. A potential side effect is necrosis induced by severe local vasospasm.
Case report: We describe the case of a five year old boy who accidentally injected 0.3 mg adrenaline in his left thumb. The distal phalanx of the thumb was pale and cool; no pain was reported. The use of local warmth and nitroglycerine plaster (5mg) had no effect. Consequently 0.25 mg phentolamine (Regitin®, 10 mg dissolved in 10 ml NaCl) was injected around the puncture area, resulting in full recovery.
Literature review: Seven case reports and one review showed findings consistent with our report: un-successful administration of local warmth or topical nitroglycerine and fast recovery after local injection of phentolamine. Phentolamine is an alpha-receptor-blocker antagonizing the effect of adrenaline. A compartment syndrome is possible because of the limited space. Phentolamine is effective up to twelve hours after the adrenaline injection. In contrast, another study (a case series of 28 unintentional adrenaline auto-injections) reported that 24/28 patients needed minimal, if any therapy. Vasodilatation was achieved by the application of physical stimulation, heat or transdermal nitroglycerine. Only three patients required infiltration (2 phentolamine, 1 terbutaline).
Conclusion: Injection of adrenaline in acral parts of the body can have potentially disastrous side effects. In case of a telephones request patients are advised to submerge the affected body part in

warm water and provide massage. If symptoms do not completely resolve within thirty minutes, they should seek medical help. Transcutaneous application of nitroglycerine can be tried. In case of persistent symptoms, phentolamine must be injected locally. If the auto-injector has been previously used by another person then the rules of accidental needle injury must be followed. Instruction on how to use an auto-injector is important for prevention of severe allergic reaction and accidental misuse.

P023

Transplant coronary artery disease is diagnosed later and is more severe in pediatric heart recipients transplanted in infancy

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Introduction: Transplant coronary artery disease (TCAD) represents the largest cause of late graft loss and the most frequent indication for retransplantation in pediatric recipients. Intravascular ultrasound (IVUS) and coronary flow reserve (CFR) allow early detection and follow-up of TCAD.
Methods: We retrospectively reviewed pediatric heart recipients with TCAD. TCAD was diagnosed by angiography or IVUS. Patients transplanted at <6 months of age (infants) were compared to those transplanted at >6 months (children). Stable TCAD was defined as no change in severity after serial evaluation and progressive TCAD as worsening disease or death as a result of TCAD.
Results: Of 310 transplanted children, 52 (17%) were diagnosed with TCAD. Of those, 20 (38%) were infants and 32 (62%) were children at the time of transplant. Age at TCAD diagnosis was significantly lower in the infant group (8.3 ± 3.4 vs 14 ± 5.8, p <0.001). The number of prior rejection episodes was significantly lower in the infant group (3.3 ± 1.7 vs 5.8 ± 3.6, p <0.01). Nevertheless, infants with TCAD had significantly more episodes of rejection compared to those without TCAD (0.83 ± 1.14 vs 3.35 ± 1.73, p <0.0001). More patients in the children group had stable TCAD (76% vs 55%, p <0.01). Compared to children, infants had a longer time from transplant to diagnosis of TCAD (8.0 ± 3.4 vs 5.4 ± 3 years, p <0.01) but their angiographic score was higher at diagnosis (3 ± 1 vs 1.7 ± 1.3, p <0.01). Coronary flow reserve in the infant group was significantly lower (2.3 ± 0.5 vs 3 ± 0.3, p <0.05) at the time of TCAD diagnosis and showed a progressive decline compared to the children group (CFR 2 years post-TCAD diagnosis 2.1 ± 0.6 vs 2.9 ± 0.7, p <0.01). There was a trend towards shorter time from diagnosis of TCAD to death or retransplant in the infant group (0.9 ± 0.9 vs 2.8 ± 3.4 years, p = 0.06).
Conclusion: TCAD in infant recipients is diagnosed later, appears worse by angiography and is associated with a lower CFR at the time of diagnosis which tends to decline over time. Despite fewer rejections in infant recipients with TCAD, there is a trend towards shorter time to death or retransplantation, reflecting a more aggressive nature of TCAD.

P024

Estimation of the risk for sudden cardiac death and implantation of an ICD in a patient with dilated cardiomyopathy due to Naxos Disease

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Introduction: Palmoplantar hyperkeratosis, woolly hair and cardiomyopathy is the characteristic triad of Naxos Disease. This inherited syndrome was first identified among individuals from Naxos, Greece. Mutations in constituents of cardiac and skin cell adhesion cause the particular phenotype. We report on a boy with Naxos disease and dilated cardiomyopathy and discuss the indication and reflections considering the implantation of an implantable cardioverter-defibrillator (ICD).
Case report: A ten years old boy with Naxos disease with pronounced palmoplantar hyperkeratosis and light, woolly hair presented with decreasing exercise tolerance. Echocardiography demonstrated biventricular dilatation with an impaired systolic and diastolic function (ejection fraction 30%, norm >55%). 24 hour Holter-monitoring showed multiple polyformic premature ventricular complexes; the late potentials were pathologic. Cardiac catheterisation revealed elevated enddiastolic pressures in both ventricles and a moderate stenosis of the aortic valve, which was successfully dilated by balloon valvuloplasty. Cardiac Magnetic Resonance Imaging showed several areas of myocardial late-enhancement as a sign of fibrotic replacement of the myocardium. These findings were indicative for increased risk of sudden cardiac death (SCD) and the implantation of an ICD was decided. Further follow up (two years) was uneventful. The patient is not restricted in his daily activities and enjoys an excellent quality of life.
Discussion: The identification of patients at high risk for sudden cardiac death is challenging in children because of the heterogeneity of the underlying myocardial diseases. In syndromatic cardiomyopathies such assessment should include definition of the syndrome and advanced morphologic, electrophysiologic and haemodynamic evaluation including cardiac catheterisation and magnetic resonance imaging. Improvement of the impaired haemodynamic status by an appropriate medication, interventional catheterisation or cardiac surgery represents the first step toward SCD prevention. Furthermore ICD implantation needs to be considered in selected patients as a safe and effective prevention of SCD. Finally heart transplantation may be discussed in patients with severely impaired function.

P025

Two unusual cases of pericardial effusion after cardiac surgery

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Introduction: Pericardial effusion can occur as part of almost any pericardial disorder. It can develop in acute pericarditis or may be seen as an incidental and silent finding in a variety of systemic disorders, after trauma or after cardiothoracic surgery.
Case report: We report two patients with persistent pericardial effusion after cardiac surgery. Case 1: A 25 year old young man with Down's syndrome underwent surgical patch closure of a ventricular septal defect (VSD). One month after the procedure he developed a haemodynamic compromising circular perdicardial effusion of 14 mm on echocardiography. Despite treatment of postcardiotomy syndrome with diuretics, non-steroidal anti-inflammatory drugs and prednisone the patient developed tachycardia, tachypnea and fever six weeks later. Pericardial drainage revealed chylopericard with 700 ml of milky-fatty fluid. After six weeks of medium chain triglycerides (MCT) diet regression of the effusion could be noted and the patient showed complete recovery. Case 2: A 1½ year old boy was operated for subaortic VSD and subaortic membranous stenosis. During postoperative course chylopericardial effusion and serous pleural effusion stopped spontaneously after 48 hours. On postoperative day 12 a percutaneous pericardial drainage was inserted due to an increasing circular pericardial effusion on echo and revealed 165 ml chylopericard. The chylopericard persisted for five weeks and required a combined conservative-surgical management including surgical creation of pericardial window, treatment with somatostatin, total parenteral nutrition followed by MCT diet for two months with a good outcome.
Conclusion: Isolated chylopericard after cardiac surgery is a rare complication. Chylopericard should be suspected, if anti-inflammatory treatment of postcardiotomy syndrome is not effective. Diagnosis is made by puncture and labarotory analysis. Treatment of chylopericard can be difficult and requires a combined an interdisciplinary approach.

P026

First degree atrioventricular (AV) block caused by Lyme borreliosis

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Introduction: Lyme borreliosis is the most common tick borne infection in Europe. It is caused by the spirochete *Borrelia burgdorferi* and is transmitted by the tick *ixodes ricinus*, which is endemic throughout Europe. The sundry clinical manifestastions can be categorized as early localized (erythema migrans), early disseminated (lymphadenitis cutis benigna, facial palsy, lymphocytic meningitis, carditis) or late (arthritis, encephalomyelitis). While dermatological or neurological manifestations are common, cardiac involvement in childhood is rare (<0.5%).
Case report: A 13 years old girl presented with exercise-induced shortness of breath. She frequently went for walks in the forest and reported having had several tick bites in recent months. Physical exam (PE) revealed physiological heart sounds without cardiac murmurs. The electrocardiography (ECG) revealed first-degree atrioventricular (AV) block with a PR interval of 280 ms. Minimal holosystolic mitral insufficiency was evident on echocardiography. Blood serology demonstrated antibodies to *B. burgdorferi*: ELISA (VIDAS) IgG / IgM positive (10.17, positive cut off index: 1.0) and Western Blot IgG + IgM positive. Four days after initiating per os antibiotic therapy with amoxicillin (12.5 mg/kg/dose 4 x/day for 21 days) ECG and echocardiography returned to normal. Within 5 weeks exercise tolerance returned to normal.
Discussion: The main manifestations of Lyme carditis are high degree AV block, often associated with myocarditis or pancarditis. According to international guidelines Lyme carditis without AV block III° in children may be treated with per os antibiotic therapy (amoxicillin or doxycyclin [only if >8 years]).

Conclusion: The clinical findings, serology, and rapid response to antibiotics are highly suggestive of mild carditis due to *B. Burgdorferi*. Exercise-induced shortness of breath may represent a mild symptom of carditis. When carditis is suspected, an ECG should be performed.

P027

«incidental finding» severe subaortic stenosis

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Introduction: Obstruction of the left ventricular outflow tract may be found at subvalvular, valvular and supravalvular level. All forms of aortic stenosis account for about 3–5% of all congenital heart defects, subaortic stenosis being the second most frequent form, but rarely seen in newborns.
Case: We report a 5½ year old girl, who was referred because of a pneumonia and a newly diagnosed 4/6 systolic heart murmur. At the age of 6 months the girl had already had an echocardiography because of a heart murmur with normal results. In the meantime she was fine and physical workout was performed without cardiac symptoms. On admission there was a 4/6- systolic ejection murmur with p.m. at the left sternal border and radiating to the neck. In addition, an impressive jugular thrill was palpable, whereas the praecordial region was not hyperactive. Transthoracic echocardiography revealed a fibromuscular subaortic stenosis narrowing the left ventricular outflow tract by half and a severe myocardial hypertrophy. CW-dopplerechocardiography showed a maximum systolic/mean pressure of 190/100 mm Hg. The patient was referred to Zurich for a surgical resection of the subvalvular stenosis. The procedure could be performed without any complications and the girl was dismissed 2 weeks after surgery. The follow-up echocardiographic study (7 weeks after the operation) showed a mild residual obstruction of the left ventricular outflow tract with a maximal pressure gradient of 18 mm Hg. The remodelling of the left ventricle was supported by propanolol.
Conclusion: Subaortic stenosis is one of the congenital heart defects which show a progression over time. It should be considered in all cases of newly audible heart murmur or if an innocent heart murmurs changes its characteristics. Normal echocardiography in the past medical history does not excuse from further investigation.

P028

Improvement of Diastolic Dysfunction by Levosimendan assessed by Tissue Doppler in a Child on Extracorporeal Membrane Oxygenation (ECMO) Support after Surgical Repair of Tetralogy of Fallot

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Introduction: Levosimendan is a calcium-sensitizing agent with inotropic properties which has been shown to improve cardiac function and hemodynamic performance in adults with severe heart failure. Experience in children is limited and promising, but there is no report of the use of levosimendan in children with diastolic dysfunction.
Case report: An 8 year-old girl was referred for late repair of Tetralogy of Fallot (TOF). Pre-operative echocardiography confirmed TOF with severe pulmonary stenosis (Z-score –5) and a small but apex-forming left ventricle (mitral valve Z-score –1) with normal systolic function. She underwent surgical repair with VSD patch closure and interposition of a RV-to-PA conduit and was unable to be weaned of cardio-pulmonary bypass despite high doses of inotropic support, so she was started on ECMO support. Post-operative echocardiography revealed no residual anatomic defects, normal systolic function but biventricular diastolic dysfunction [mitral Doppler E/A = 2.7 (N <1.5), tricuspid E/A = 1.7 (N <1.5), pulmonary vein S/D = 0.3 (N >0.5), mitral and tricuspid Tissue Doppler Imaging (TDI) Ea 4 and 5cm/s (N >8), mitral E/Ea = 16 (N ≤8)], confirmed by elevated filling pressures (CVP 20–25 mm Hg and LAP 20–22 mm Hg), and levosimendan was administered during 24 h. On post-operative day (POD) 3, her clinical condition improved and she was successfully weaned off ECMO on inotropic support (milrinone, adrenalin and noradrenalin). Because of persistent elevated filling pressures and diastolic dysfunction by echocardiography, she received a second infusion of levosimendan on POD 8 with improvement of filing pressures and weaning off inotropic support on POD 11. Echocardiography at 3 weeks post-surgery showed normalization of mitral and tricuspid TDI Ea (14 and 8cm/s) and mitral E/Ea ratio (E/Ea = 8), suggestive of improvement of diastolic function.
Conclusion: In diastolic dysfunction requiring ECMO support, levosimendan contributed to ECMO weaning and improvement of diastolic function, as documented by normalization of invasive filling pressures and TDI parameters. Prospective studies in larger populations are needed to confirm this potential beneficial effect of levosimendan.

P029

Asymptomatic right encircling aortic arch associated with ventricular septal defect and biscuspid aortic valve

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Introduction: Encircling aortic arch is a rare form of vascular ring usually associated with severe tracheo-bronchial compression presenting with symptoms of upper airways obstruction. The treatment consists of surgical ligation of the ligamentum arteriosum and/or uncrossing of the aorta. Previous descriptions of asymptomatic patients have never been reported to our knowledge.
Case report: A 19 month-old boy was referred for ventricular septal defect closure. Echocardiography showed a large perimembranous ventricular septal defect (VSD) with high flow pulmonary hypertension and left heart dilation. The aortic valve was bicuspid with neither stenosis nor regurgitation. The aortic arch anatomy was difficult to delineate by echocardiography but the aortic arch appeared right-sided with suspicion of a left aberrant subclavian artery. By cardiac magnetic resonance imaging (MRI), the ascending aorta was right-sided, bifurcating to the left behind the oesophagus and trachea just above the tracheal bifurcation, with a left-sided descending aorta, a left ligamentum arteriosum and a retro-oesophageal left sub-clavian artery, realizing an encircling aortic arch. The child underwent VSD surgical closure with a bovine pericardial patch and surgical ligation of the left ligamentum arteriosum. His post-operative course has been uneventful with no respiratoty symptoms allowing for early extubation.
Conclusion: Symptoms of tracheal obstruction have been described in some patients only after extubation in the post-operative period of cardiac surgery, probably in relation to change in the mediastinal geometry after sternotomy. For this reason and despite the absence of respiratory symptoms, surgical ligation of the ligamentum arteriosum should be performed in patients undergoing cardiac surgery.

P030

Large ascending aorta aneurysm with severe aortic regurgitation in a 7 year-old girl with Marfan syndrome

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Introduction: Dilation of the ascending aorta in patient with Marfan syndrome is rare in early childhood. Medical management with angiotensin converting enzymes inhibitors (ACEI) or beta-blockers (BB) is recommended until severe dilation is present that requires surgery.
Case report: A 7 year-old girl was referred to our institution with physical features of Marfan syndrome associated with dyspnea on exertion and palpitations. The chest X-ray revealed marked cardiomegaly with widened mediastinum. Echocardiography showed aneurysmal dilation of the ascending aorta (aortic valve annulus 3.7 cm, sinus of Valvalva 8.5 cm) with severe aortic regurgitation and left ventricular dilation (8 cm) with mildly depressed left ventricular function (FS 28%). Cardiac magnetic resonance imaging (MRI) confirmed the dissecting aneurysmal dilation of the ascending aorta with severe aortic regurgitation (RF 44%) and left ventricular dilation. After an initial unsuccessful aortic valve preserving operation consisting of aortic valvuloplasty with interposition of a 28 mm Valsalva conduit, the child underwent a Bentall procedure with reimplantation of the coronary arteries on a biological valve conduit. Unfortunately, because of severe left ventricular dysfunction, the child could not be weaned off cardio-pulmonary bypass and was placed on extracorporeal life support with a centrifugal left ventricular assist device (LVAD). Unfortunately, she did not recover sufficient function to be weaned off the LVAD and died on post-operative day 5 from low cardiac output.
Conclusion: Cardio-vascular complications of Marfan syndrome include aortic root dilation, mitral valve prolapse, mitral and aortic valve regurgitation, LV dilation and pulmonary artery dilation. Early medical management and close follow-up of pediatric patients with Marfan syndrome is important to detect progression of aortic root dilation and aortic regurgitation, as surgical mortality in patients with advanced disease and compromised left ventricular function is high.

P031

Tetralogy of Fallot with absent pulmonary valve, total anomalous pulmonary venous connection and major systemic to pulmonary collateral artery

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Introduction: Absent pulmonary valve syndrome is a relatively rare variant of tetralogy of Fallot (TOF), and has been reported in 4 patients in association with totally anomalous pulmonary venous return (TAPVR). Although up to 25% of patients with TOF/absent pulmonary valve syndrome have systemic-to-pulmonary collaterals, this is the first time it is reported in association with TAPVR to our knowledge.

Methods: A 2-year-old boy was referred to our institution for cyanosis and mild signs and symptoms of heart failure. Transthoracic echocardiography showed TOF with features typical of absent pulmonary valve syndrome. There was severe pulmonary valve stenosis with a peak systolic gradient of 85 mm Hg and free pulmonary regurgitation, associated with aneurysmal dilatation of the branch pulmonary arteries. There was also a secundum atrial septal defect and anomalous pulmonary venous connection to a non-obstructed confluent leading to the right atrium at the junction of superior vena cava. A tubular structure was visualized between the abdominal aorta and the inferior vena cava, with continuous systolo-diastolic accelerated flow. Cardiac MRI confirmed that this structure was a major aorto-pulmonary collateral artery.
Results: We performed a staged approach, starting with interventional embolization of the collateral artery followed by successful surgical transatrial patent foramen ovale enlargement and redirection of the anomalous pulmonary venous connection to the left atrium using a pericardial patch, patch closure of the ventricular septal defect and right ventricular outflow tract reconstruction with a Labcor® valved conduit.
Conclusions: This rare association of TOF/absent pulmonary valve, TAPVR and MAPCA illustrates the importance of a complete preoperative diagnosis, for successful staged hybrid management.

P032

Pulmonary hypertension and macrocephaly in a newborn: does it ring a bell?

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Background: Persistent Pulmonary Hypertension of the Newborn (PPHN) occurs in about 1:500 term and pre-term infants. Its aetiology often remains unclear, but common causes include structural lung disease (e.g. congenital diaphragmatic hernia), lung pathology (e.g. meconium aspiration pneumonia, hyaline membrane disease) and group B streptococcal infection.
Case report: A term-born boy presented with postnatal respiratory distress and need for supplemental oxygen. Besides tachypnea (respiratory rate 110/min) and macrocephaly which was interpreted as familial, clinical examination was normal. Echocardiography revealed severe PPHN. Underlying structural lung or cardiac disease, significant lung pathology (only radiological signs of slight aspiration of amniotic fluid) and neonatal infection were excluded. Bone morphogenetic type II receptor (BMPR-II) gene analysis to rule out familial pulmonary hypertension was normal. Tachypnea and supplemental oxygen need persisted, and the cause of the PPHN remained unknown. At the age of 6 weeks, cardiac catheterization was performed. It showed cardiac high-output-failure and distended veins coming from the brain. Subsequently, vena galeni malformation was diagnosed on angio-MRI examination of the brain.
Conclusion: In this neonate, vena galeni malformation resulted in high-output cardiac failure with PPHN due to pulmonary hypercirculation. The combination of PPHN with macrocephaly should prompt search for arteriovenous cerebral malformations.

Posters – Adolescent Medicine

P033

Youth-friendly primary-care services: do they make a difference?

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Introduction: Primary care has a key role to play in addressing young people's health needs. Yet young people face barriers in accessing this care. Recommendations to remove these barriers have been proposed under the concept of youth-friendly services. The aim of this study was to review the evidence on the effectiveness of youth-friendly services in improving young people's health.
Methods: MEDLINE, PsycLit, Embase and the Cochrane databases were searched between 2000 and 2005 for studies in which youth-friendly health care provision had been evaluated. The reference lists of the resulting abstracts were also searched manually for other relevant studies. Papers in which primary-care services for young people were only described but not assessed were excluded.
Results: Twenty seven articles were included for review. Most came from countries with established economies, but there were also studies from Africa, South America, China and Bangladesh. Most had methodological limitations in that they were uncontrolled observational studies. The evidence indicated that youth-friendly initiatives can improve access to care and provider performance in addressing young people's health concerns. There was no reliable evidence, however, for the effect of youth-friendly primary-care services on young people's health outcomes.
Conclusions: More evidence of the benefits of youth-friendly primary-care services on young people's health is needed. Promoting the socio-economic and political conditions in which innovative, well assessed youth-friendly primary-care services can be implemented is also a priority.

P034

Health Networking and Adolescent Friendly Health Services in Geneva

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Introduction: Adolescent Friendly Health Services are services that are available, accessible, acceptable, appropriate and equitable for adolescents. The Adolescent and Young Adult program at Geneva University Hospitals is a multidisciplinary team including primary care physicians (general practitioners, internists and paediatricians), nurses, gynaecologists and psychiatrists. Its aim is to improve the quality of care provided to young people. In this paper we describe the activities of the program and the role of the network around this service in Geneva's community to improve health care access and adherence to care for adolescent patients.
Method: The Adolescent and Young Adult program has developed strategies in 3 directions: health care, training and research. The multidisciplinary team, trained in adolescent health and recognized as expert team, used these strategies to make connexions with the different services in the city. A qualitative study involved the partners of the network and the adolescent clients to identify the key points of this network and the positive and negative outcomes after 4 years of existence of this program.
Results: In addition to general adolescent health care the activities of the program include: networking especially with schools, social services and juvenile justice services; support and advice for family doctors in the city; multidisciplinary health care in complicated situations (i.e. violence, multiple risk behaviours, pregnancy or sexual aggression); assessment and specialized care for diagnoses such as eating disorders, substance use or chronic illness. A training program has been tailored to enhance primary care physicians' competences in this field: appropriate communication with young people, the role of families, the need for confidentiality, etc. Different actors in the network (e.g. family doctors) collaborate in the research activities.
Conclusion: Such a reference centre is a support and useful specialized program. Networking with community services and family doctors can help to address the needs of adolescent and young adult through adolescent friendly health services.

P035

Cannabis use trajectories among Swiss adolescents

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Introduction: Swiss data indicate that one fifth of current 16–20 year-old cannabis users do not use tobacco and seem to do better than those smoking both substances. The aim of this research is to assess the substance use trajectories of cannabis users who do not use tobacco and those who use both substances from age 17 to age 23.
Methods: Using data from the TREE longitudinal data base, 328 out of 1796 youth 18.3%; 45% females) who smoked cannabis only (Group CAN; N = 46; 36% females) or concurrently with tobacco (Group CANTAB; N = 284; 46% females) at T1 (2001; age 17) were followed at T4 (2004; age 20) and T7 (2007; age 23). Two additional outcome groups were included at T4 and T7: those using only tobacco (Group TOB) and those not using any of these substances (Group NONE). Data were analyzed separately by gender.
Results: Females in group CAN at T1 were as likely to be in group TOB (35%) or NONE (35%) at T4 and the percentages increased to 41% and 47%, respectively, at T7. Males in group CAN at T1 were more likely to be in group TOB at T4 (33%) and T7 (61%) than in group NONE (23% and 15%, respectively). Females in group CANTOB at T1 were mainly in group TOB at T4 (52%) and T7 (61%), while males in CANTOB at T1 remained mainly in the same group at T4 (75%) and T7 (61%). Only 10% of females and 5% of males in group CANTOB at T1 were in group NONE at T4 and 15% and 12%, respectively, at T7.
Conclusions: Adolescents using only cannabis are globally less likely to continue using cannabis in young adulthood than those using both substances, although a fair percentage (specially males) switch to tobacco use. This result confirms previous research indicating that nicotine dependence and persistent cigarette smoking may be the main public health consequences of cannabis use. A gender difference arises among those using tobacco and cannabis at age 17: while females become mainly tobacco smokers, the majority of males continue to use both substances. Although these results could be explained by a substitution effect, teenagers using both substances seem to have gone beyond the experimentation phase and should be a motive for concern.

P036

Are adolescent high internet users at greater risk for their health

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Introduction: The impact of intensive internet use on global health is poorly known. This study examines the relationship between internet use and the health status of adolescents. We hypothesized that high internet users have a poorer health state.
Methods: Data were drawn from the 2002 SMASH database, a survey of Swiss adolescents aged 16 to 20 years. Adolescents using internet at least 2 hours daily in the last month constituted the High Internet Users group (HIUs; n = 208; 2.8%) and all others (n = 7340; 97.2%) the control group. Analyzed health factors were: self-perceived health level, depressive tendencies, overweight, self-estimated sleep quantity, as well as frequent occurrence of headache and back pain. Health factors significant at the bivariate level ($p < 0.05$), plus possible confounders (age, gender, academic track (student/apprentice), residence (rural/urban), parental level of education, nationality (Swiss born/not) and presence of a chronic illness), were included in a multivariate analysis (results shown as adjusted odds ratios (AOR) with [95% confidence interval]).
Results: Bivariate analysis demonstrated that HIUs were significantly more likely to be males (82% vs. 51%), younger (17.6 ± 1.1 vs. 17.9 ± 1.2 years), living in a rural area (59% vs. 44%), overweight (16% vs. 10%) and reporting more insufficient sleep (48% vs. 38%). After controlling for the significant confounding factors, the multivariate analysis showed that overweight (AOR = 1.66 [1.12–2.46]) and reported insufficient sleep (AOR = 1.50 [1.12–2.01]) remained significantly linked to HIUs.
Conclusions: Although adolescent intensive internet users were a minority in 2002, the current trends seem to indicate an increase of this behavior in that age group. At greater risk for overweight and insufficient sleep, this growing population shows a poorer health state. Therefore, pediatricians should inquire about internet use intensity and convey preventive messages about the possible consequences of its excessive use as part of their consultation.

P037

Influence of parenting skills on obese children's outcome after individual or group therapy

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Introduction: Childhood obesity results from a large spectrum of genetic and psychosocial influences, making therapy complex and difficult. As family-based multiprofessional interventions prove to be most successful, the influence of parenting skills – either demanding or supportive – on changes in BMI, burden by difficulties or quality of life (QoL) in European obese children was investigated.
Methods: In the longitudinal "Euregio-KIG" cohort study of the German, Austrian and Swiss Lake Constance region, 312 obese children (means ± SD: 12 ± 2.2 years, BMI-SDS 2.26 ± 0.50) were included in a multiprofessional obesity therapy either in an individual n = 84 or group n = 228 setting. In 184 families out of these, the influence of the parenting dimensions of demandingness and supportiveness (n = 28 or n = 156, resp.) on children's weight loss (delta BMI-SDS), on strengths & difficulties score (SDQ) and QoL (KINDL Ob.) before the end of one year programs could be examined by validated questionnaires (BzGA study, Germany 2005) and by univariate analysis of variance, controlling for age and gender as well as for age and BMI of parents.
Results: BMI-SDS decreased by 0.38 after individual therapy, $p < 0.05$, and by 0.09 after group intervention, $p > 0.05$. In both groups QoL improved significantly by 10.5 and by 4.1, resp., both $p < 0.05$. Baseline SDQ was similar in both groups, but significantly reduced after group intervention only (delta = –2.65, $p < 0.05$, versus –1.03, n.s., after individual therapy). Parental behaviour had no effect on these changes, but a high supportiveness resulted in a significant reduction of BMI-SDS by –0.22, $p < 0.05$, after group therapy only. Unexpectedly, low supportive parental behaviour was associated with a decrease in BMI-SDS after indivi-dual therapy. So far no effects were observed following demanding parental behaviour.
Conclusion: Therapy improves well-being in obese children. Supportive parental qualities of upbringing seem to promote a favourable outcome of obese children after group therapy. An individualized therapy may compensate for parental deficits and thus reduce obesity. However, results are biased by the higher drop out rate during individual intervention as well as by regional differences in therapeutic settings and should be corroborated by randomised controlled trials.

P038

Waist-to-height ratio, a good predictor of metabolic syndrome in children for the paediatrician

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Introduction: The actual prevalence of childhood obesity is increasing. Cardio-vascular and metabolic complications require an early and systematic screening at the paediatrician office. Waist circumference (WC) and BMI are widely used. However, the normal range of theses predictors are age-dependent, and it implies referring to tables. Recently, a waist-to-height ratio (WHtR) above 0.5 has been shown to be a good predictor of metabolic syndrome (MS) in adults, but studies in the paediatric population are scarce on this topic.
Methods: This is a cross-sectional study, including 20 obese children (age in years 11.35 ± 0.41). Measurements of body weight, height, and waist circumference were taken. We assessed fasting glucose, fasting insulin, triglycerides, high-density lipoprotein cholesterol (HDL-C), and blood pressure.

Results:	WC		BMI z-score		WHtR	
	Pearson	p	Pearson	p	Pearson	p
Fasting glucose	0.349	.221	0.086	.727	0.344	.229
Insulin	0.789	.001*	0.279	.247	0.581	.029*
TG	0.639	.014*	0.263	.292	0.566	.035*
HDL-c	–0.415	.141	–0.376	.125	–0.230	.428
Systolic BP	0.783	.001*	0.621	.003*	0.688	.005*
Diastolic BP	0.255	.359	0.409	.073	0.326	.236

Conclusions: We could demonstrate that waist-to-height ratio correlates with metabolic syndrome as well as waist circumference, and better than BMI z-score (which doesn't reflect abdominal fat). Moreover, it has the advantage to be performed quickly at the paediatrician office, and doesn't require any reference table. In fact, we think that every child with a value of waist-to-height ratio above 0.5 should be proposed a metabolic workup.

P039

Does metformin improve adiposity and components of the metabolic syndrome in youngsters? Randomized placebo controlled trial in 70 obese adolescents

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Objective: As results on the efficacy of metformin to reduce obesity and components of the metabolic syndrome in youngsters at risk for diabetes are conflicting, this was studied in European obese adolescents in addition to so far unsuccessful lifestyle intervention.
Methods: We included obese adolescents (13.8 years, 65% female, 33% immigrants, BMI 33.1 kg/m^2), with insulin resistance (HOMA-IR >P.97, 5.0) and/or impaired glucose tolerance (total n = 243). After 6 months of multiprofessional individualized lifestyle intervention, 70 out of 86 adolescents without improvement in BMI and HOMA-IR were enrolled in a randomized double blind controlled study with placebo (n = 34) or metformin (2x500 mg/d, n = 36) in addition to ongoing lifestyle intervention for another 6 months in 2 centers. Anthropometric (BMI-SDS, waist circumference) and metabolic parameters (OGTT, HOMA-IR, Insulin sensitivity index (ISI), lipids) were measuredevery 6 months.
Results: During lifestyle intervention alone, BMI and HOMA-IR deteriorated significantly (to 35 kg/m^2 and 6.1, resp.). In the subsequent medication period, HOMA-IR improved by 1 point in both groups (in 73% vs 54% of metformin vs. placebo patients) as did fasting insulin, but BMI remained unchanged. ISI, however, improved in the metformin group only (p <0.01). Other components of the metabolic syndrome remained unchanged. High fasting insulin predicted a subsequent BMI increase. irrespective of medication. Drop outs rate for side effects was 7% (1 metformin, 4 placebo patients).
Conclusion: In pubertal obese European adolescents unresponsive to behavioral therapy, hyperinsulinaemia predicted weight gain, but insulin resistance slightly improved without weight change both during metformin and placebo plus lifestyle intervention, probably due to enhanced compliance with healthy lifestyle as a pure placebo effect. Metformin dose may have been too low to achieve significant additional effects.

P040

Associations between physical activity level, cardiorespiratory fitness and cardiovascular diseases risk factors among obese children and their mother

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Introduction: The aim of this study was to compare physical activity, cardiorespiratory fitness and cardiovascular diseases (CVD) risk factors among obese children and their mother.
Methods: This was a prospective study including 18 pre-pubertal obese children (female 39%, age 9.6 ± 1.2 yr) and their mother (age 38.5 ± 5.4 yr). Measures included: seven-day physical activity count by accelerometer; cardiorespiratory fitness (VO_2max); body mass index (BMI); waist-to-hip ratio, body fat by DXA; blood pressure (BP); fasting blood lipids, glucose, insulin, HOMA-IR and high-sensitive C-reactive protein (hs-CRP) levels.
Results: Thirty-four percent of mothers were overweight and 3 of them were obese. Children had higher waist-to-hip ratio (0.90 ± 0.06 vs 0.83 ± 0.06, p = .002), insulin (11.6 ± 4.1 vs 8.0 ± 5.2 mU.L^{-1}, p = .03) and HOMA-IR levels (2.4 ± 0.9 vs 1.7 ± 1.1, p = .048), and moderate-to-vigorous physical activity (51.8 ± 22.8 vs 29.3 ± 21.8 min.day^{-1}, p = .017) compared to mothers, whereas the later had higher total cholesterol (5.2 ± 0.8 vs 4.6 ± 0.8, p = .02) and diastolic BP (73.5 ± 10.1 vs 67.1 ± 7.5 mm Hg, p = .011) than children. There were no differences among groups for total physical activity, VO_2max, BMI, total body fat, systolic BP, blood lipids and hs-CRP levels.
Conclusion: Pre-pubertal obese children have increased risk of central adiposity and insulin resistance indices compared to their mother, but moderate-to-vigorous physical activity remains higher in children. Total physical activity, cardiorespiratory fitness, whole body fat, systolic BP, and blood lipids are similar among children and their mother. Therapeutic interventions focusing on mother's lifestyle changes might be interesting approaches to prevent the spread of CVD in obese children.
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P041

Feasibility and effects of whole body vibration training in children

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Introduction: Whole body vibration (WBV) has spread within the last years and is now a common exercise mode in fitness centres. There are several studies about WBV with contradictory conclusions. In particular, there is lack of scientific information about WBV training in children. Therefore, the aim of this study was to analyse the feasibility of WBV and its effect on motor performance in healthy children.
Methods: WBV using a side alternating Galileo platform (Novotec/GER) was applied with 12–15 Hz to 13 children and with 25–27 Hz to 12 children. 12 children without training served as control group. The children were 11–12 years old and randomly assigned to the different groups. WBV was performed in slight squat position twice a week for 3 minutes over a training period of 4 weeks. Motor performance was tested pre and post training assessing coordination (sideward jumping for 15 sec), power (standing long jump) and dynamic stability (stability index of MFT S3-Check).
Results: Although some subjects showed reddening of the legs during training or itching at the shanks for some minutes after training, WBV produced no permanent negative secondary effects and was well tolerated by the children. All three groups showed a significantly increased performance in sideward jumping and a small non-significant increased performance in long jump. No interaction between the different WBV training methods or between WBV groups and control group could be found.
Conclusion: WBV of this intensity seems to be feasible in children of this age and may be transferred in rehabilitation setting studies. The improved performance in all children is possibly caused by learning or developmental effects. The lack of a WBV training effect may be explained by the fact that children of this age cannot be regarded as untrained, which seems to diminish the effect of WBV as also seen in adult studies.

P042

Physical activity intervention for obese children: what are the mid terms benefits on cardiovascular risk factors?

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Introduction: Childhood obesity is a major public health issue as it is associated with premature development of cardiovascular diseases (CVD). We report blood pressure and endothelial function evolutions, 2 year after a 3 or 6 months physical activity program including 38 pre-pubertal obese children.
Methods: This is a follow-up study (mean time: 1.96 ± 0.35 years) including 20 obese children (age in years 11.35 ± 0.41). We assessed blood pressure (BP) by ambulatory monitoring; endothelial function by echocardiography; arterial stiffness using applanation tonometry; body mass index (BMI); body composition by dual-energy x-ray absorptiometry (DXA); physical activity by accelerometer and biologic markers.
Results: Mean BMI z-scores (3.7 ± 0.2 vs. 3.6 ± 0.2 kg.cm^-2, p = .32), total body fat (41.9 ± 1.6 vs. 42.8 ± 1.5%; p = .39) and physical activity (703.4 ± 85.2 vs. 574.4 ± 99.5 cpm, p = .297) were stable over time. Daytime and 24h diastolic BP z-scores significantly declined (day: 2.0 ± 0.4 vs. –0.01 ± 0.4, p = .006; 24 h: 1.4 ± 0.4 vs. 0.3 ± 0.4, p = .043), while systolic BP z-scores were at the limit of significance (day: 2.7 ± 0.6 vs. 1.3 ± 0.5, p = .055; 24 h: 2.4 ± 0.4 vs. 1.4 ± 0.45, p = .067). Diastolic 24 h BP decreased more in children who had reduced BMI z-scores compared to the one who did not (p = .028). Systolic hypertension rate dropped from 50 to 27.8% and diastolic hypertension from 41.7 to 5.6%. Arterial flow-mediated dilation decreased (6.4 ± 0.7 vs. 4.8 ± 0.6 %, p = .039), and was associated with high body fat (p = .020) and 24h systolic BP (p = .049). There was no difference between the children who performed 3 vs. 6 months of physical activity.
Conclusions: We investigated the mid term impact of a physical activity program on CVD risk factors in obese children. The beneficial effects on blood pressure observed at the end of the intervention were still present, and even further improved. However, endothelial function was still impaired in these patients. Our results highlight the importance to perform regular physical activity for the prevention of hypertension.

P043

Virtual Reality Soccer Scenario to Enhance Active Participation of Robotic-Assisted Gait Training in Children

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Introduction: Virtual environments can make repetitive motor rehabilitation exercises more motivating and thereby more effective. Children in particular show insufficient motivation towards the training process, as the regular training might be too monotonous and lacks stimulation entertainment. Therefore, we developed a virtual reality (VR) based soccer scenario that provided interactive elements to engage patients during robot-assisted treadmill training. The aim of the study was to compare the immediate effect on motor output during different supportive conditions in patients and healthy control children while training with the Lokomat driven gait orthosis (DGO).
Methods: Ten patients (four males, six females, mean age 14.6 years, std. 3 years) with different neurological gait disorders and eight healthy children (two males, 6 females, mean age 12.6 years, std. 3.3 years) were instructed to walk on the Lokomat in four different randomly presented conditions: (1) walk normally, (2) therapists' instruction to promote active walking, (3) VR as motivating tool to walk actively, and (4) VR tool combined with therapists' instruction. The measured motor output is expressed by force exertion provided by DGO.
Results: Active participation in patients and control children increased significantly when supported and motivated either by therapists' instruction or by VR scenario compared with reference measurement (normal walking) ($p < 0.0001$).
Discussion: In conclusion, the used VR scenario induces an immediate effect on motor output that is of similar magnitude as the effect resulting from verbal instructions by therapist. Therefore, VR represents a valuable tool for patients and healthy subjects to keep motivation high during DGO training. Further development needs to focus on the implementation of interactive design elements that keep the motivation high across and beyond Lokomat training sessions especially in pediatric rehabilitation.

P044

Sport and inflammatory Diseases in Childhood: what is the current practice in the region Bern?

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Introduction: In the multidisciplinary outpatient clinic for rheumatology the team is often asked by the parents if sports are forbidden, allowed or recommended. Several studies demonstrated that children with juvenile idiopathic arthritis (JIA) have a lower physical condition compared to healthy children of the same age and sex. For those reasons we decided to investigate the practice of sports among our patients.
Method: During a 6 month period we asked all the children aged 7 years and older attending our outpatient clinic to fill out a questionnaire. The questionnaire focused on the kind of sport practised, the frequency of the training, and the participation in competition. All the 47 patients who filled out the questionnaire have an inflammatory rheumatic disease (39/47 with JIA). The children were aged 7–17 years old (mean 12,5 +/ 3.03 SD). 15 of the 47 patients were not on medication. 13 patients were under NSAIDs (nonsteroidal antiinflammatory drugs) alone. 12 patients had Methotrexate and 6 patients anti-TNF. Steroids alone or in combination were used for 4 children.
Results: Only 4 patients (9%) do not have an extracurricular sport activities. All other patients are attending an extracurricular sport at least once a week, and 27 patients (58%) are doing an extracurricular sport activity twice a week or more. All kinds of sports are represented: swimming (14 pat.) and biking (13 pat.), which are joint sparing activities are often practised. But soccer (6pat), jogging (6 pat.) and skiing (7 pat.) are also very popular. Even demanding activities on the joints such as artistic skating are practised. Rarer sport kinds as sailing, golf, or hornus playing are reported. Half of the patients regularly take part in competition but only 4 of them have sport ambitions. 40% of the patients are members of a sports club.
Conclusion: Spontaneously our patients seem to have realised that a sport activity is important even they have an inflammatory rheumatic disease. Most of them seem to chose an activity which is joint sparing. They also understand our message that sport activities are a role not only for the physical condition but also have positive psychosocial aspects.

P045

Fatal pulmonary embolism in an adolescent with ulcerative colitis

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Introduction: Patients with inflammatory bowel disease (IBD) have a higher risk for thromboembolic complications, such as deep venous thrombosis and pulmonary embolism (PE), that is explained due to the chronic inflammation. Prolonged immobilisation, dehydratation, surgery and side effects of medications (e.g. corticosteroids) are additional risk factors.
Case report: We report the case of a boy, who was diagnosed with ulcerative pancolitis (UC) at the age of 6y. At the age of 7 years, he developed a subacute cerebral sinovenous thrombosis while treated with high doses of corticosteroids and azathioprin for an acute relapse. He showed complete resolution of the cerebral sinovenous thrombosis after 4 months treatment with heparin. The family history was unremarkable for thromboembolism. Additional congenital and acquired causes for thrombophilia, such as factor V Leiden, prothrombin mutations and Anticardiolipin antibodies were excluded. A hypercoagulable state was noted with increased platelet counts, increased d-Dimer and thrombin antithrombin complex and slightly decreased Protein S and C levels. Thus, prophylactic antithrombotic therapy was recommended in situations with a greater risk for thrombosis. In the following years, the patient showed a difficult course of the UC with frequent relapses and steroid dependence despite treatment with methotrexate. At the age of 15 y, during a subacute relapse of UC with worsening diarrhea and tiredness, the boy experienced a sudden collapse in his home and died from an acute PE. He didn't have any signs of deep venous thrombosis. The autopsy revealed a massive central pulmonary embolus with superficial calcifications adherent to the main pulmonary artery. These calcifications indicate previous PE, although the patient never complained about symptoms suggestive for PE.
Discussion: The risk for thromboembolic complications is well established in patients with active IBD with a 3–4 fold higher risk compared to the general population. To our knowledge, this is the first case report of an adolescent with UC who died after a fatal PE, in whom no risk factors were identified in addition to the chronic inflammation and prothrombotic side effects of the medications. In conclusion, thromboembolic complications should be considered also in children with IBD and prompt prophylactic antithrombotic therapy in situations with a greater risk for thrombosis.

P046

When teenager's nail polish hides a dermatological condition: a case of twenty nail dystrophy

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Introduction: Severe nail changes are relatively rare in children. They can appear as isolated events or associated with other symptoms. Rarely they are a sign of nutritional deficiency. Even in the absence of a serious medical condition they often lead to uncertainty and psychological burden, especially in affected adolescents. Wanting diagnosis may lead to medical odyssey and potentially damaging therapeutic polypragmasia.
Methods: Illustrated case report and review of literature.
Findings: This twelve years-old girl was seen in her paediatrician's office for a regular preventive medical check-up. The otherwise healthy teenager had a striking nail polish of both her fingers and her toes. After a short joking about the latest fashion trends the girl's mother explained, that the nail varnish was to hide a strange and ugly change of her daughter's hand and feet nails that appeared some months ago. After removing the varnish the nails showed a longitudinal ridging, multiple pits, a brittleness and a sandpaper like appearance. There were no signs of other dermatological conditions. The girl was referred to a dermatologic nail clinic and was diagnosed with idiopathic trachyonychia.
Discussion: Trachyonychia, the so called "twenty nail dystrophy", is a visual diagnosis rarely demanding nail biopsy. The nail changes appears typically in childhood and early adolescence and consist of a typical sandpapered appearance with a rough, lustreless nail plate of grayish-white colour, a longitudinal ridging and superficial pits and usually affect all twenty nails symmetrically. The condition can both occur as an isolated nail abnormality as well as a common manifestation of alopecia areata, lichen planus, psoriasis, eczematous changes, vitiligo, onychomycosis but also IgA deficiency or primary biliary cirrhosis. The isolated, idiopathic form is proposed to have an autosomal dominant inheritance and is known to be a benign condition that, especially in children, undergoes spontaneous remission.
Conclusion: Nail disorders in childhood should be carefully investigated and concomitant pathologies ruled out. Reassurance and at most topical medication are the cornerstones of care in patients with the relative benign but cosmetically distracting diagnosis of twenty nail dystrophy.

P047

Missed genital inspection as part of routine exami-nation in prepubertal girls: Be prepared for a surprise

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Introduction: Main causes for a growing abdominal mass in adolescent girls with primary amenorrhoea include tumour, bladder dilatation secondary to urine retention and haematocolpos due to imperforate hymen, horizontal vaginal septum or vaginal atresia. Patients may present with cyclical lower abdominal pain, urinary and rectal symptoms.
Case report: A 12 year old girl was referred with a history of increasing abdominal pain and progressive abdominal distension for the last 4 weeks. Former medical history was uneventful. The abdominal pain was described as dull and extended into the lumbar region. It was felt permanently rather than in a cyclic pattern. Bowel or bladder disturbances, lack of appetite, weight loss and night sweats were negated. The patient did not have menarche. The abdomen was uniformly distended and a well defined smooth mass reaching from the pelvis to the umbilicus was palpated. Breast and pubic hair development had reached Tanner stage 3, there was no evidence of developmental anomaly. Suspecting a cystic or malignant abdominal mass genital examination was not performed at the time. Haematological values were normal. A transabdominal ultrasound of the abdomen revealed the presence of a large haematocolpos and haematometra. The following gynaecological examination showed a bulging and slightly livid imperforated hymen. Incision and surgical repair of the hymenal membrane with slow release of 1.6 litres of menstrual blood was performed by the gynaecologist.
Conclusion: Haematocolpos and haematometra in female adolescents may remain silent for a long period of time. Alarming signs emphasizing the diagnosis are cyclical abdominal pain in pubertal girls without menarche. On clinical examination a palpable abdominal mass urges immediate genital inspection. The diagnosis is still escaping paediatric routine examination until the time of menarche, especially in absence of symptoms. If treatment is delayed there may be serious impairment of fertility secondary to haematometra, haematosalpinx, salpingitis and endometriosis. Therefore inspection of the external genitalia is imperative in all girls before puberty and mandatory for early detection of imperforate hymen.

Posters – Oncology/General Paediatrics/Neuropaediatrics

P048

Ethical issues in the care of paediatric patients: development and evaluation of practical workshops for medical and nursing staff

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Introduction: Medical-nursing teams in many paediatric fields are confronted with difficult and complex situations. In paediatric oncology, a interdisciplinary team composed of physicians, nurses, psychologists, social workers, teachers, pharmacists, representatives of religious worship, take care of the child and its family. Within the framework of a Master in philosophy of medicine, under the direction of "Culture and Health" (University of Lyon, France), we intend to study the usefulness of practical application of ethics as a tool in decision-making for the best care of children.
Methods: Objectives: To present, apply and evaluate an ethics workup in daily pediatric practice to help to improve clinical decisions, to help physicians and nurses of the oncology unit to get familiar with medical ethical conflicts and diemma through teaching ethics workshops, and to support discussion and exchange. All staff members of the oncology unit will be invited to an ethical workshop once a month for 5 months, each lasting 1H30. A health care professional involved in the care of the patient will present a problematic situation that has either occurred in the past or is still on-going and raises ethical questions related to the child's medical history. The meeting will be guided by a doctor of medicine and philosophy who is head of the ethics unit of the University hospital (CHUV). This workshop will be evaluated by a written questionnaire at the end of the meeting.
Results: During these 5 workshops, the type of up-coming topics, recurrent problems, participation and the possible impact on present and future practice will be evaluated. We will analyze the way the workup is conducted, its meaning for the professionals and the outcomes, and compare with the literature existing in this field.
Conclusion: Depending on the impact of these first workshop series, extension to all paediatric units and the integration of an "Espace Ethique" within the DMCP will be considered.

P049

Long-term childhood ALL survivors: Health-related quality of life after a relapse

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Background: During the last decades, treatment of acute lymphoblastic leukemia (ALL) in children has improved, with current 5-year survival rates reaching 85%. About 20% of children with ALL suffer a relapse and 1/3 of these can be retreated successfully. Little is known on long-term health-related quality of life (HRQOL) in this group.
Objective: To assess whether quality of life in adult long-term survivors of childhood ALL differs between those with and without a relapse.
Methods: In the Swiss Childhood Cancer Survivor Study (SCCSS), we sent a questionnaire to all former childhood cancer patients who had been diagnosed with cancer before age 16 and survived for more than 5 years. HRQOL was assessed with the SF-36, a self-assessment tool with 36 questions. It measures eight dimensions (scales) of health status: physical functioning, role limitations due to physical health, bodily pain, general health perceptions, vitality, social functioning, role limitations due to emotional problems, and mental health. Scores on each scale range from 0–100, with a score of 100 indicating the highest rating of health. Additionally, a physical and a mental component summary scale can be derived from these eight scales by factor analysis.
Results: 381 ALL survivors aged ≥20 years were eligible for the survey. By January 2009, 283 (74%) participated, but recruitment is still ongoing. Of these, 44 (16%) had had a relapse. Mean values for the domain of general health and for the physical component summary were 55.7 and 55.6 respectively in survivors without relapse and 52.1 and 54.1 respectively in survivors with relapse ($p = 0.023$ and 0.055). The other seven domains and the other summary scale did not differ between those with and without relapse.
Conclusions: In general, health-related quality of life in long-term ALL survivors is comparable to the general population, and similar between those with and without relapse. However, two sub scores of SF-36 are slightly lower. Further analyses will determine risk factors for a reduced HRQOL in long-term ALL survivors.

P050

Sinus Histiocytosis with massive lymphadenopathy (Rosai-Dorfman-disease) treated with Cladribine (2-CdA)

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Background: Sinus Histiocytosis with massive lymphadenopathy or Rosai-Dorfman-disease is a rare but well defined histiocytic proliferative disorder of unknown origin that usually presents with cervical lymphadenopathy, fever and leucocytosis in an otherwise healthy child. Elevation of cytokines such as Il-6 represents the activity of the disease. Although many patients undergo spontaneous remission, a subset of patients has a more serious course. For these patients steroids and chemotherapeutic agents such as etoposide or 6-mercaptopurine with low dose methothrexate have been used. Because treatment response with these drugs is often insufficient, new approaches come to clinical application. Successful therapies have been reported occasionally with Cladribine (2-CdA), a well known purine analogue extremely toxic to mature, non dividing lymphocytes, generally used in the treatment of hairy cell leukaemia. Cladribine impairs the function of monocytes including inhibition of Il-6 production.
Case report: We report on a 5 years old boy who was admitted for massive bilateral cervical lymph nodes after exclusion of a known infectious origin by the local paediatrician. The child was febrile, but in good general condition, the lymph nodes were tender. To rule out

bacterial lymphadenitis colli antibiotic treatment was given without any improvement. Finally lymph node histology showed characteristic features of Sinushistiocytosis type Rosai-Dorfman. Treatment with steroids, 6-mercaptopurin and methothrexate failed. Because of progressive enlargement of the cervical nodes and further dissemination to the jugular and mediastinal lymph nodes we were forced to look for alternative treatment modalities. Therapy with Cladribine (2-CdA) was begun. Shortly after the first cycle dramatic shrinkage of the lymphadenopathy was evident with further regression occurring after subsequent cycles. Biological inflammatory parameters such as Il-6 decreased. There was no apparent toxicity to observe. After 12 months follow-up off therapy, complete clinical and biological response is sustained without any recurrence.
Conclusions: Our findings confirm the promising efficacy of cladribine and suggest that therapies targeting specifically cytokines might be useful in some cases of otherwise therapy refractory patients with Sinus Histiocytosis with massive lymphadenopathy (Rosai-Dorfman-disease).

P051

Thrombocytopenia, disseminated intravascular coagulopathy, microangiopathic haemolytic anaemia and a vascular tumour: The Kasbach-Merritt phenomenon

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Introduction: A vascular tumour combined with disseminated intravascular coagulation (DIC), thrombocytopenia and microangiopathic haemolytic anaemia are the characteristics defining the Kasabach-Merritt phenomenon (KMP). Kaposiform haemangioendotheliomata (KHE) are vascular tumours, along with haemangiomata and tufted angiomata, and although KHE is rare, it is frequently associated with KMP. Treatment options include surgery, radiation, embolisation and medication. However, no large scale or prospective studies are available which have established a standard of care for KHE.
Case: A 20 month old boy presented with multiple haematomata of different sizes and stages, an enlargement of the liver and spleen, and swollen testicles. He was otherwise in good general condition. Laboratory findings showed haemoglobin 88 g/L, platelets $12x10^9$/L, white blood cells $15.4x10^9$/L, PT 46%, aPTT 40 sec, TT 20 sec, fibrinogen 0.57 g/L and D-dimers >20 ug/mL. All other parameters were normal. Abdominal CT/MRI revealed an inoperable mass extending from the retroperitoneum into the intraabdominal and mediastinal space and infiltrating several organs. A tumour biopsy proved a KHE. On the day after biopsy, the DIC was further complicated with acute intra-abdominal bleeding. This necessitated an emergency omentectomy and repeated administrations of thrombocytes, fresh-frozen-plasma, tranexam acid and activated factors VII and XIII. After stabilisation, a systemic therapy with prednisone, vincristin and cyclophosphamide was initiated. After six months of therapy, the tumour has not gone into remission, however it has shrunk and the patient shows a good general condition.
Discussion: In our case an extended KHE caused a life-threatening KMP with a severe DIC and acute bleeding following biopsy surgery. In conclusion, physicians should be aware of the potentially life-threatening KMP as a complication of vascular tumours in childhood, and carefully plan diagnostic and therapeutic strategies.

P052

Pernio (chilblains) in 2 children: A rare cause of painful or discoloured toes

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Kinderklinik Aarau

Background: Pernio (chilblains) is a rare cause of painful or blue fingers or toes in childhood. Low temperatures cause a localized inflammatory lesion of the skin.
2 case reports: Patient A: A 14 yr. old girl presented with pain and multiple, purplish, edematous lesions of the toes of 14 days duration. All toes were warm and the foot pulses were strong. Mechanical pressure or injury was denied. Blood tests showed no signs of inflammation. Serological tests and anti-DNA-Ab, ANCA, ENA 1, ENA 2, Antiphospholipid-Ab were normal. X–ray of the feet, sonography of metatarso-phalangeal-joints and duplex sonography of all vessels of the lower extremities were unremarkable. 19 days after presentation the toes were painless, and the skin had normal colouring. Some small dry and hyperkeratotic lesions were present on the dorsal side of the toes. Patient B: A 10 yr. old girl presented with a 3 month history of multiple, pruritic, painless, purplish-red, hyperkeratotic skin lesions of the toes of 3 months duration. Laboratory tests were normal.
Discussion: Pernio occurs most commonly among young women between the ages of 15 and 30 years, but may occur among older individuals or children as well. Acute pernio (A) may develop 12–24 h after exposure to the cold. Chronic pernio (B) occurs with repeated exposure. Low body weight seems to be a predisposing factor. Lesions usually begin in fall or winter. Incidence among children is unknown. Frequently described symptoms are: intense pain, itching, and burning of reddened skin which may also blister. Cold induced trauma is thought to result in vascular damage from tissue hypoxemia and secondary inflammatory reaction. In some patients cryoproteins or cold agglutinins are detectable. Therapy consists of preventing low temperatures. Nifedipine has been demonstrated to be effective.
Conclusion: Pernio is a diagnosis of exclusion (in particular systemic lupus erythematodes should be excluded). Laboratory evaluation is unremarkable. The most important "therapy" is prevention.

P053

Simplifying Paediatric Immunization With a Fully Liquid Combination Vaccine: Evidence From A Time-Motion Study In India

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Background and aims: Immunization is essential to achieve the Millennium Development Goal (MDG) of reducing child mortality. Fully liquid combination vaccines have been developed to rationalize vaccine delivery and to simplify supply and administration of vaccines. A study was carried out to understand implications of a fully liquid pentavalent DTP-HepB-Hib vaccine in terms of resource requirements, efficiency and impact on vaccination programs.
Methods: A time-motion study was conducted at the Institute of Child Health (ICH) in Calcutta, India. The study compared a single fully liquid DTP-HepB-Hib vaccine vs. a lyophilized vaccine with two vials requiring reconstitution. 312 children were vaccinated over 6 weeks in 2006. An analysis was done to estimate potential time and programmatic impact.
Results: Study results indicated significant time savings for vaccine preparation and total vaccine consultation for the fully liquid vaccine of 52 % and 23% ($p < 0.05$) as compared to the lyophilized vaccine. At current vaccine load time savings at ICH would be between 15 and 25 working days per year. Extrapolated to India, these delivery time savings could be around 100,000 working days per year.
Conclusions: The single vial fully liquid DPT-HepB-Hib combination vaccine offers important time gains for vaccine delivery as compared to a vaccine requiring reconstitution. Combination vaccines simplify logistics and delivery, critical in countries with health staff shortage. Fully liquid combination vaccines might contribute to better resource management and ultimately improve efficiency of immunization programs.
Funding: Novartis Vaccines and Berna Biotech

P054

"Oh, Baby, please don't cry!" Osteopathic treatment reduces crying intensity and duration in infants with colic, compared to treatment as usual

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Background: Infantile colic (IC) affects 8–40% of infants, and causes distress both to children and parents. IC is defined following Wessel's "Rule of Threes": Crying at least or more than 3 hours per day, on more than 3 days per week, for more than 3 weeks. Generally, a lack of etiology and consensus of treatment is observed. Treatment as usual (TAU) consists of thorough medical check to exclude possible concomitant illness, and of behavioral advice such as rocking, walking, etc.. Osteopathic treatment (OT) is a complementary therapy and consists of diagnosing musculo-skeletal strain patterns in the body, followed by techniques to release these strains.
Aim: To compare TAU and OT with regard to IC.
Method: 16 infants (mean age: 8 weeks; range: 2–12 weeks) were prospectively enrolled in the study. All infants underwent thorough medical check, and IC was diagnosed. They were randomly assigned either to TAU or OT. 10 infants entered the OT and 6 the TAU branch. OT consisted of four sessions within 4 weeks; infants in the TAU branch received no further treatments. After 4 weeks, all infants were thoroughly examined and assessed again by the same Pediatrician. Parents completed questionnaires with regard to family strain, crying intensity and duration, and sleep, before and after OT or TAU. Parents registered crying and sleep patterns with a daily sleep log and sleep was objectively assessed with actigraphs.
Results: Infants of the two groups did not differ with respect to gender, age, or birth weight. Likewise, mothers did not differ with respect to age, vocational career, education, quality of pregnancy, or any perinatal conditions. Com-pared to infants of the TAU group, crying

intensity (F(1, 14) = 4.58, p <.05) and duration (F(1, 14) = 4.78, p < .05) decreased significantly in infants of the OT group. Overall family strain decreased for all families irrespective of group (F(1, 14) = 4.40, p < .05), though with significant strain reduction for families with infants of OT (F(1, 14) = 4.96, p < .05). No significant results were observed with respect to sleep.
Conclusions: IC causes distress both on the infant and the family system. OT may successfully reduce crying intensity and duration in the infant, and therefore significantly relieve family strain.

P055

Acute Life-Threatening Event (ALTE) in an infant: think about star anise intoxication

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Introduction: In the past years a series of cases was published about neurologic and gastrointestinal symptoms after the ingestion of star anise *(Illicium verum)*. Mostly, the cause of the intoxication was the treatment of infant colic pain by herbal teas containing Chinese star anise contaminated with Japanese star anise (*Illicium anisatum)*.
Observation: We report the case of a two-month-old infant who developed symptoms of poisoning some hours after receiving star anise tea. Hyperexcitability, hypertonia of the extensor muscles, horizontal intermittent nystagmus, episodes of cyanosis and important regurgitations were observed. All the complementary exams were normal and the symptoms disappeared spontaneously after 48 h. We could prove the contamination of the Chinese star anise preparation with Japanese star anise. The product was then prohibited from selling.
Conclusion: The diagnostic evaluation for ALTE should include the search for ingestion of star anise tea, which can cause various neurologic or gastrointestinal symptoms by overdose of Chinese star anise or by contamination with Japanese star anise. Indeed, Japanese star anise contains sesquiterpenic lactones that are potent neurotoxins. Actually, star anise products are in free sale in Switzerland and the potential adverse effects linked to its consumption are not well known of the parents, care personal and pharmacists. New cases should be prevented by more ample information.
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P056

Acute Respiratory Insufficiency as a challenging Presentation in Guillan-Barré-Syndrom

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Background: Guillain-Barré-Syndrom (GBS) is an acutely evolving, immune mediated, inflammatory disorder of the peripheral nervous system leading to demyelination and axonal loss. Clinical hallmarks are a rapidly progressive, symmetric weakness and areflexia. Sudden repiratory insufficiency is a rather rare complication in GBS.
Case report: A 3 3/12 year old boy presented to our emergency department with acute onset of respiratory insufficiency and loss of consciousness. His recent history revealed a non-febril infection of the upper respiratory tract 3 weeks earlier. In addition the patient had complained of unilateral leg pain 3 days prior to admission. He had been treated for assumed coxitis fugax with an unremarkable ultrasound examination of the hips. On admission the child required intubation and mechanical ventilation. Due to the depressed consciousness and respiratory insufficiency neurologic evaluation could not be completed. Initially diagnosis was unclear and the cranial CT showed no pathologic findings. A few hours later when the effects of medication for intubation could be excluded, a flaccid tetraplegic paralysis with areflexia and autonomic dysfunction with elevated blood pressure and tachycardia became obvious. Clinical findings and results of cerebrospinal fluid examination with an increased level of protein and normal cell count lead to the diagnosis of GBS. A spinal MRI showed characteristic enhancement of the cauda equina nerve roots. For completion of diagnosis electrophysiologic investigations as somato-sensible evoked potentials and nerve conduction velocity showed typical signs of demyelination. Under standard therapy with intravenous immunoglobulin there was a slow but favourable return of sensibility and motor activity. Initial attempt at extubation failed due to the presence of a postintubation subglottic granulom but was successful after laser therapy to remove the granuloma.
Conclusion: GBS is a well known but not very frequent disease in childhood. Diagnosis of GBS can be challenging especially if respiratory insufficiency is one of the first symptoms. In our case, unilateral leg pain three days prior to hospital admission had been the only and unusual preceding symptom. Neurological examination, cerebrospinal fluid findings, spinal MRI and electrophysiologic investigations are characteristic for the diagnosis of GBS.

P057

Skin rash, fever and hearing loss should evoke Muckle Wells syndrome

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Background: The Muckle Wells syndrome, belonging to the cryopyrin-associated periodic syndromes (CAPS) is generally characterized by progressive sensorineural hearing loss, fever and cold-induced urticaria. It is commonly linked to mutations in the gene NLRP3 on chromosome 1.
Case-report: A 2-years-old boy known since his first year of life for a severe bilateral sensorineural hearing loss, fever, urticaria-like rash twice a month and occasional ankle arthralgia, showed a high elevated sedimentation rate. The laboratory work-up could exclude a chronic infection, immunodeficiency, auto-immunity and allergies. Hearing loss-linked syndromes like Alport, Pendred and connexine 26-linked syndromes could be excluded. Q703K mutation on gene NLRP3 was found and based on the symptoms and the genetic analysis, a diagnosis of Muckle Wells syndrome was confirmed. The patient started a daily subcutaneous treatment of Anakinra, a recombinant selective interleukin-1 receptor antagonist, which induced significant improvement on fever, rash and arthralgia within 3 months of treatment.
Conclusion: In the presence of hearing loss, urticaria and fever in a child, it is important to look for CAPS. Mutation Q703K on the gene NLRP3 is found in 3% of the general population and has been reported to be associated with atypical neurological cryopyrin-associated periodic syndromes. We report here the first case of classical Muckle Wells syndrome with this mutation, associated with early-onset hearing loss. Since some years, a very efficient treatment, Anakinra, is available for this condition.

P058

A Rare Cause of Macrocephaly: Megalencephalic Leukoencephalopathy with Subcortical Cysts

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Introduction: Megalencephalic leukoencephalopathy with subcortical cysts (MLC) is a rare white-matter disease, first described by M. van der Knaap in 1995. Diagnostic features include macrocephaly in infancy with mostly mild clinical signs of white matter affection at time of diagnosis and typical MRI. Clinical signs in early infancy include mild spasticity, ataxia, mild cognitive decline and sometimes epilepsy. The course of the disease can vary, about 50% of the patients show slow progression. MRI shows extensive bilateral white-matter changes with cysts in the temporal regions. Based on the clinical and MRI features, MLC can be distinguished from other conditions with macrocephaly (Alexander's disease, Canavan's disease, Glutaric aciduria type I). Inheritance is autosomal recessive, in 80% pathogenic mutations in the MLC1 (22q13.33) gene are found.
Case report: We report a female patient born to non-consanguineous parents who was referred for excessive macrocephaly by the age of 6 months. Head circumference had been normal at birth. At first presentation, she showed extreme macrocephaly (+8 SD); her motor and cognitive development was appropriate to her age and she showed a normal neurological examination. Cerebral ultrasound revealed megalencephaly with hyperechogenic white matter with multiple small and single wider cysts in the subcortical region. MRI confirmed megalencephaly with normal CSF spaces. White matter of both hemispheres seamed swollen, T_2 white matter signals were isointense to CSF. Corpus callosum, capsula interna and cerebellum were not affected. Genetic testing confirmed a compound heterozygosity for two of the (more than 50 known) pathogenic mutations in the MLC1-Gene. Now 18 months old, the girls motor and cognitive development is still appropriate to her age. She shows no neurological pathologies, did not experience any seizures and her EEG is normal.
Conclusions: MLC is a rare cause of progressive macrocephaly in infancy to be considered in differential diagnosis. Ultrasound can presume, MRI confirm the diagnosis. No curative therapies are known until now. Prenatal diagnosis is possible.

P059

Bowel-perforation of ventriculo-peritoneal shunts: Still an unknown complication?

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Introduction: Implantation of a ventriculo-peritoneal (VP) shunt is an established therapy for hydrocephalus. Perforation of VP-Shunts into

abdominal structures, such as bowel, scrotum, vagina or bladder is a rare but potentially deadly complication.
Case report: In June 2008, a 2 year old boy suffered a severe accidental brain injury with intracerebral haemorrhage and subsequent hydrocephalus. A VP-shunt was placed. In October 2008 a revision was required because of an abscess just above the valve. In November 2008 during a weekend at home, the boy presented with recurrent vomiting and irritability. Parents detected an anal protrusion of a white tube when changing nappies. Diagnostic work-up in Hospital neuchatelois did not revealed a protrusion of the tube, but a mild tenderness in the left lower abdomen. CBC revealed leucocytosis (14.1 10^9/l) and elevated CRP (23 mg/ml). In abdominal X-ray the tip of the VP-shunt was located in the peri-rectal area. Treatment with Meronem and Vancomycin was started. Rectoscopy in the Inselspital Berne confirmed the assumed perforation. Intra-operative taken cultures revealed Staphylococcus aureus and Klebsiella ornithinolytica, liquor cultures Staphylococcus aureus. The VP-Shunt was replaced.
Discussion: Complications from VP-shunt malfunction due to obstruction or infection are well-known. Perforations of abdominal structures such as rectum, vagina or scrotum have been described in ca.100 cases. In younger children, longer VP-shunts are usually placed to allow an adaptation with growth. Younger children have been found at a greater risk for perforations. Early diagnosis of such complications might be difficult, as clinical signs of peritoneal irritation, and signs of meningeal irritation secondary to the VP-shunt infection are usually lacking.
Conclusion: Given the frequent use of VP-shunt, pediatricians should be aware of potential complications, including intestinal perforations; in awareness of the high mortality of such complications, a high index of suspicion is required.

P060

A not-uncommon form of non-epileptic seizures: benign paroxysmal tonic upgaze of infancy

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Introduction: Benign paroxysmal tonic upgaze of infancy is a transient movement disorder characterized by episodes of sustained upward deviation of the eyes.
Reports: Between January and December of 2007 we made the diagnosis of benign paroxysmal tonic upgaze in 6 infants (4 boys and 2 girls) referred with suspected epilepsy. The infants, who had been born at term or near-term without perinatal or neonatal disease alarm indicators after an uneventful pregnancy, were clinically healthy. In the 6 infants episodes of sustained, conjugate, upward deviation of the eyes (often with incomplete downward saccades on attempted downgaze) lasting 3 to 5 seconds were first observed between the age of 2 and 7 months. The episodes were associated with preserved horizontal eye movements and a normal level of consciousness. In the 6 infants the episodes were documented by video-recording and interictal electroencephalography was normal. The tendency to recurrent episodes of upward deviation of the eyes remitted 2 to 6 months after onset without any drug management.
Conclusion: The present experience confirms that paroxysmal tonic upgaze of infancy is a benign, transient and likely not-uncommon movement disorder best managed with parental reassurance. Misdiagnosing the condition with epilepsy might result in redundant investigations and management, which can cause apprehension in the parents. Unfortunately existing textbooks do not refer to the condition or mention it only in passing.

P061

Detection of posture in children by an electronic-garment – a pilot study

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Background: Postural management plays an important role in children with central motor impairment. Several therapeutic interventions aim to improve postural control. However, to date there exist no satisfactory measurements which allow to evaluate therapeutical approaches. This pilot study introduces an electronical garment for posture recognition and aims to investigate its reliability and feasibility in the community setting.
Method: A comfortable, loose fitting shirt with integrated acceleration sensors developed at the Wearable Computing Lab of ETH Zurich was used to assess several standardised postures in healthy children. For comparison of the tool, skin attached acceleration sensors and a video based analysis system (Dartfish®) were used.
Results: Data of healthy children (n = 22) suggest that 6 different sagittal postures can be differentiated by the lose-fitting electronical shirt. The wireless data transfer (Bluetooth) has proven to be reliable and allows the tool's use without restrictions in activities of daily living. According to the questionnaires, acceptance of the children to wear a lose electronic garment was high.
Conclusion and outlook: The loose fitting electronical shirt has been proven as a feasible and reliable option to detect posture in healthy children. Our data justify further studies applying this system to investigate posture also in children with motor impairment. This innovative tool may offer – in the long run – the possibility to log specific movements of the trunk and upper extremities to assess outcome of rehabilitative interventions.

P062

Outcome of acute peripheral facial palsy in children – a catamnestic study of 84 patients

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Introduction: Causes, therapy and prognosis of acute peripheral facial palsy differ among children and adults, but there are few studies regarding the pediatric population only. This survey gives an overview of the etiology and outcome in children.
Patients and methods: 84 children aged 10 months to 16 $^3/_{12}$ years, who were seen at the children's hospital of Eastern Switzerland during the years 1998–2007 because of a facial palsy, except of congenital and traumatic causes. Data about etiology, symptoms, diagnostics and therapy were gathered retrospectively from the medical histories, duration and outcome by questionnaires. 6 of 9 patients with residual symptoms were seen for control and recovery was graded by the House-Brackmann scale.
Results: There were 83 cases of unilateral and 1 bilateral case with no relevant differences in gender or side. The etiology was in 26 cases (31.0%) neuroborreliosis (NB), in 6 (7.1%) other infections (3 otitis media (OM), 2 varizella, 1 herpes encephalitis) and in 1 case (1.2%) a toxicity of Methotrexat was suspected; 51 cases (60.7%) were classified as idiopathic (IPFP). Between June and November, the number of cases with NB rises to 53.3%. 4 patients with IPFP received steroids. 75 (89.3%) recovered completely (HB-Grade I). Full recovery was reached between 2 and 365 days (median 21 days). 9 patients (10.7%) showed slight residual symptoms (HB-Grade II). Of these, 6 were classified as IPFP, 1 was caused by NB and 1 by OM.
Discussion: In this study almost a third of the palsies were caused by NB, during the summer and fall even more than half. These findings are similar to results of previous studies and confirm the importance of a lumbar punction. Patients with palsy due to borreliosis show a higher recovery rate than those with OM or IPFP. Since all patients with NB had received antibiotic treatment, no conclusions can be drawn regarding its impact on the natural history of the disease. The palsy recovered in almost 90% completely even without treatment with steroids and residual symptoms were slight.

P063

Central Nervous System Tumors in 75 Pediatric Patients with Neurofibromatosis Type I in Bern

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Introduction: Neurofibromatosis type 1 (NF1) is a relatively common disorder with predisposition to central nervous system (CNS) tumors.
Methods: From a series of 75 pediatric patients with NF1 in Bern, we performed a retrospective assessment regarding the prevalence and distribution of CNS tumors in these patients.
Results: Of 75 patients with NF1 (mean age at last follow-up: 10.2 years, range 1–18 years), 39/75 (52.0%) had a neuroimgaging study during the course of follow up. In 17/75 patients (22.7%) one or more CNS tumors could be detected (8 female and 9 male patients). Imaging studies showed 13/17 (76.5%) optic pathway gliomas with 4 (23.5%) involving only one optic nerve, 4 (23.5%) involving both optic nerves, 7 (41.2%) involving the chiasma and 1 (5.9%) involving the optic tract. One patient showed thalamic pilocytic astrocytoma, 8 patients multiple hamartomas and 2 patients cerebral gliomas. Clinical problems related to these tumors were visual impairment in 6 patients, high intracranial pressure in 2 patients, symptomatic epilepsy in 2 patients and central pareses of facial nerve in 1 patient. Seven patients were asymptomatic. Treatment was started in 3 patients due to progressive visual impairment: first choice was carboplatin for 10–16 months. Due to recurrent progression in 2 patients a second course with etoposide or etoposide/temozolomide was initiated. All three children showed severe visual handicap, but stable disease over the last few years.
Conclusion: CNS tumors are affecting nearly one quarter of pediatric patients with NF1, fortunately progressive functional deficits are rare. Careful neurological and ophthalmologic follow-up during childhood and adolescence is essential. Due to the nature of these tumors, treatment should only be started in case of proven progressive symptoms.

P064

When a goiter is just the tip of the iceberg

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A 14-year-old boy presented with goiter in the emergency department. Dysphagia and dyspnoe had been present since one week. Three days before hospital visit his mother detected a pretracheal swelling that visibly increased in size. No fever or weight loss occurred in the previous weeks. On physical examination, the boy showed a large symmetric goiter. The thyroid tissue was homogeneously enlarged, painless, and of hard consistency. No locoregional or distant lymph nodes were enlarged. Clinically the boy was in a euthyroid state. Laboratory tests revealed normal erythrocyte, leucocyte, and platelet counts without blasts. Blood chemistry showed no signs of increased cell lysis. TSH and T4 were in the normal range. The thyroid ultrasound showed a hypoechogenic and inhomogeneously enlarged thyroid tissue with small calcifications. The lower pole of the left thyroid lobe showed retrosternal extension. Further, one single enlarged supraclavicular lymph node with inhomogeneous tissue pattern was found. On chest radiograph, multiple mediastinal enlarged lymph nodes with deviation of the trachea were seen. Relevant tracheal deviation and compression from an enlarged thyroid mass were confirmed on MRI. The histological analysis of the excised supraclavicular lymph node revealed the diagnosis of a precursor T-lymphoblastic lymphoma. Chemotherapy according to the EURO-LB 02 study was initiated. The boy reached complete remission at day 33. The most frequent etiologies of goiter during puberty are Hashimoto thyroiditis or M. Basedow. A goiter is an unusual primary clinical sign of a lymphoma. However, a rapidly growing goiter in association with symptoms of dysphagia and dyspnoe/stridor must always be considered as a true emergency and warrants immediate radiological investigations to visualize the mediastinum and the trachea.

P065

Case Presentation: Autoimmune Disease, Not Iodine Deficiency, as Underlying Disease of Goiter in a 9 Year-Old Girl from the Grison Alps

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Background: The association between goiter and iodine deficiency is particularly well known in areas historically burdened with iodine deficiency such as the Swiss Alps. We report a child from the Grison Alps with goiter initially suspected to be caused by iodine deficiency.
Case report: A 9 year old girl with goiter was referred by the paediatrician, who had suspected iodine deficiency as underlying cause, based on the information that the family had temporarily switched from iodized to non-iodized salt. Iodine substitution (150 ug/day) had been initiated without laboratory evaluation and maintained for 1 year, without normalisation of thyroid size. History taking gave no hard evidence for increased risk of iodine deficiency. Physical examination revealed goiter stage II-III in an otherwise healthy girl. Laboratory evaluation revealed a euthyroid state, normal iodine urine secretion and, as expected, positive thyroid-antibodies (TPO-Ab 112 U/ml; Tg-Ab 1100 U/ml) leading to the diagnosis of euthyroid autoimmune thyroiditis Hashimoto. Iodine supplementation was stopped and thyroid function regularly followed-up.
Discussion: In the Swiss Alps iodine deficiency and associated diseases including goiter historically were endemic. However, government-guided nation-wide iodine supplementation has been ensured by iodization of table salt and animal food for over 50 years. Today, normal consumption of dairy products and meat contribute to roughly 60% of the daily recommended intake. Therefore, iodine deficiency can almost certainly be excluded in healthy individuals in Switzerland and other causes must be ruled out before making the diagnosis of iodine-deficiency-induced goiter and before starting iodine therapy.
Conclusions: Iodine deficiency, though once endemic, today is an extreme rarity in Switzerland. Autoimmune disease, not iodine deficiency, is the leading cause of goiter in Switzerland. Finally, as always in medicine, the making of the diagnosis should precede the therapy.

P066

Neonatal diabetes mellitus and exocrine pancreas insufficiency due to pancreas hypoplasia: Case report

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Neonatal Diabetes Mellitus (NDM) is a rare condition, occurring in 1:300'000–400'000 live births. Only a few cases of pancreatic hypoplasia or agenesis have been reported as cause of NDM. Mutations in transcription factors involved in pancreas development were found in some of these cases. We present the clinical course of a girl born at term to non-consanguine parents with pancreatic hypoplasia. She was born at 40+2 gestational weeks with marked intrauterine growth retardation (1600 g, P<3, z-Score: –3.84) after an uneventful pregnancy. On the first day of life she presented with hypoglycaemia and was admitted to our NICU. After administration of i.v. glucose high blood glucose values of between 18–23 mmol/L were noted together with glucosuria +++ but without ketonuria beginning at 19 hours of life without normalisation after stopping glucose infusion. Since NDM was suspected, i.v. insulin therapy was established and replaced by an s.c. insulin pump therapy after definitive diagnosis. The daily insulin requirements were low ranging from 0.1–0.4 U/kg/d in the first two months. The diagnosis of NDM was established on the basis of hyperglycemia with glucosuria and undetectable levels of C-peptide, endogenous insulin and absence of autoimmune diabetes mellitus specific antibodies. Additional exocrine pancreas insufficiency was suspected because of impaired weight gain and undetectable pancreatic elastase (<15 ug/g; Ref >200) in the faeces after 3 weeks, which was treated by pancreatic enzyme substitution. MRI of the abdomen revealed only rudimentary pancreatic tissue confirming severe pancreas hypoplasia. Permananet neonatal hyperglycemia demands measurement of insulin, C-peptide, beta-cell specific antibodies, search for exocrine pancreas insufficiency and radiologic evaluation of pancreas morphology. This rare condition appears to be treatable by high caloric intake under insulin and pancreatic enzyme substitution, although little data is available on the long-term outcome.

P067

Endothelial biochemical markers are increased in children with type 1 diabetes and are associated with diastolic blood pressure

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Introduction: Type 1 diabetes (T1DM) is associated with premature atherosclerosis. Biochemical markers of vascular function are related to retinopathy in children with T1DM but there is no information about their relationships with high blood pressure.
Methods: This is a cross-sectional study including 29 children with T1DM, without known renal complications, and 39 healthy controls (age 10.5 ± 0.5 vs. 9.8 ± 0.4 years). We measured 24-hour blood pressure (BP) by ambulatory BP monitoring; circulating levels of soluble intercellular adhesion molecule-1 (ICAM-1), vascular cell adhesion molecule-1 (VCAM-1), and sE-selectin, as indices of vascular endothelial cell activation; concentrations of sP-selectin and IL1- , as markers of platelet activation and inflammation, respectively. We also assessed glycemic control and lipid profile.
Results: Groups had similar physical characteristics and lipid profile, except body mass index (BMI) which was higher in T1DM children than healthy controls (18.6 ± 0.5 vs. 16.7 ± 0.4 $kg.m^{-2}$, p = .003). Children with T1DM had increased diastolic BP (24 h DBP z-score 0.61 ± 0.18 vs. –0.65 ± 0.16, p <.001), even after adjustment for BMI, as well as higher VCAM-1 concentrations (492.4 ± 66.6 vs. 340.2 ± 37.5 $ng.ml^{-1}$, p = .039) compared to healthy children. There was also a trend towards increased sE-selectin levels in T1DM subjects (33.5 ± 3.4 vs. 25.6 ± 2.7 $ng.ml^{-1}$, p = .069). Diastolic BP z-scores were associated with sE-selectin (t = 2.57, p <.05), disease duration (t = 2.15, p = .042), and triglycerides levels (t = 2.58, p = .017) in the T1DM group. Concentrations of sE-selectin were also related to triglycerides levels (t = 3.2, p = .004).
Conclusions: In our study, children with T1DM present the first hint of cardiovascular diseases. They have increased diastolic blood pressure and some biochemical markers of endothelial dysfunction, compared to healthy subjects. Concentrations of sE-selectin are well correlated with diastolic BP and could be used as a screening tool to detect patients at risk of vascular complications.

P068

Diabetes and immune thrombocytopenic purpura – a new association?

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Introduction: Type 1 diabetes is often associated with autoimmune diseases such as thyroiditis and celiac disease. In rare cases it can be part of manifestations of autoimmune polyglandular syndromes (APS), characterized by at least two endocrine autoimmune diseases associated to non-endocrine autoimmune disease. The case reported includes type 1 diabetes, thyroiditis and immune thrombocytopenic purpura (ITP), unusual for both, diabetes and APS.
Case report: A 13-year-old healthy Caucasian boy, known for allergic rhinitis treated by preseasoneal immunotherapy, presented with multiple haematoma and developed a goitre. ITP (Coombs test and ANA AB+) and Hashimoto's thyroiditis (anti-TPO-AB+) were diagnosed. L-T4 therapy was started. Severe ITP was resistant to

iv immuno-globulins and dependent on high dose steroids. Under such therapy, the patient developed polyuria and polydipsia. Type 1 diabetes was diagnosed (anti-GAD and anti-IA2 AB+) and insulin treatment initiated. He also presented a thoracic zona. Severe thrombocytopenia persisted despite cyclosporine therapy. Rituximab therapy was started. Further work-up revealed a slight elevation of anti transglutaminase AB and T- lymphopenia. Anti 21-hydroxylase AB were negative. Parathyroid function was normal. Serology for viral hepatitis C was negative.
Conclusion: This case is a challenge for establishing a causative diagnosis. APS 1-3, FAS gene mutations (ALPS), primary immune deficiency, IPEX-syndrome or extra-hepatic autoimmune manifestations in hepatitis C need to be considered. FAS gene mutations (ALPS) are unlikely in the absence of lymphocytosis. Chronic ITP resistant to therapy has already been reported in association to thyroid and celiac disease but it has been described neither with diabetes nor with APS. APS-1 appears unlikely in the absence of candidosis, hyperparathyroidism and adrenal insufficiency. But mutations of the APS-1 gene (AIRE) have to be excluded due to APS-1 broad clinical variability and to its potentially lethal complications. Symptoms seem to correspond mostly to APS-3 with an exception for age of onset. A predisposing genotype implicated in central control of the immune system has to be the cause and needs further investigations.

P069

Hydrometrocolpos, hypothalamic hamartoma and polydactyly- pitfalls and differential diagnosis in neonatal presentation of Pallister-Hall syndrome.

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Background: Pallister Hall Syndrome (PHS) is a rare disorder that may affect the development of several organ systems. The most common features are: polydactyly of the fingers or toes and hypothalamic hamartoma, which can lead to seizures or endocrine abnormalities. Other features include malformation of the airways such as bifid epiglottis, imperforate anus, and kidney abnormalities. Although signs and symptoms vary in severity, only a small percentage of affected children develop serious complications.
Method: case report
Results: Birth at 36 wks of gestation after a normal pregnancy, BW 2700 g, APGAR 3/6/7. She was intubated from days 1–4 of life due to respiratory distress caused by an intra-abdominal cystic mass (already detected on ultrasound during gestation) which impaired lung function. Postnatal abdominal ultrasound showed bilateral hydronephrosis and vaginal atresia with hydrometrocolpos. After renal drainage creatinin levels normalised and hydronephrosis disappeared. Echocardiography was normal. In addition to the intra-abdominal cystic mass (hydrometrocolpos) and dysplastic kidneys, other dysmorphic findings were present: postaxial polydactyly, hypoplastic fingernails, microcephaly, small nose and hypothalamic hamartoma (seen in cranial ultrasound and MRI). The phenotype was suggestive of Pallister-Hall Syndrome (DD Mc Kusick-Kaufman, Bardet-Biedl), which was later confirmed genetically by detection of a GLI3 mutation. The parents were not consanguineous. Currently the girl suffers so far only from isolated growth hormone deficiency, with no signs of hypoglycaemia. She had retarded motoric function and decreased growth velocity, both of which improved after initiating s.c. growth hormone substitution.
Conclusion: The clinical picture of Pallister-Hall syndrome can be similar to Bardet-Biedl and Mc Kusick Kaufman syndromes during the neonatal period. The importance of detailed genetic counselling and molecular genetic approach can optimize clinical management and improve long-term follow up.

P070

Identification of a novel *SRY*-mutation causing 46,XY complete gonadal dysgenesis in two half-sisters by paternal gonadal mosaicism

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Introduction: Mutations and deletions in the *SRY*-gene have been identified in about 15% of individuals with 46,XY complete gonadal dysgenesis (CGD). The *SRY* gene, located at Yp11.3, encodes the sex-determining region Y protein and is the testis-determining factor (TDF), which initiates male sex determination. Mutation of *SRY* prevents testes differentiation from occurring, resulting in CGD.
Methods: We describe the clinical, endocrinological and molecular characteristics of two half-sisters with CGD, related by their deceased father.Genetic investigations were performed after one sister was diagnosed for primary amenorrhea and infantilism of secondary sexual development, hypergonadotropic hypogonadism, infantile uterus and hypoplastic bilateral gonads. The karyotype revealed 46,XY. *SRY* deletion was excluded by FISH analysis. Direct *SRY* gene sequencing was performed according to standard methods.
Results: Sequencing of the *SRY* coding region revealed a novel, hemizygous missense mutation c.347T>C resulting in a replacement of Leucin to Serin at position 116 (p.Leu116Ser). The mutation is located in the HMG-domain, an evolutionary conserved region, with *in silico* analysis (PolyPhen software) predicting a pathogenic impact. Therefore, p.Leu116Ser is likely to adversely affect *SRY*'s DNA-binding capacity. Mutation analysis of the sister as well as of 50 male control individuals is ongoing.
Conclusions: We describe a novel mutation in the *SRY* gene, p.Leu116Ser causing Swyer syndrome phenotype in two half-sisters related by their common father. The recurrent phenotype in this family is most likely due to paternal gonadal mosaicism.

P071

Methylation profiling in cases with uniparental disomy identifies novel imprinted genes on chromosome 15

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One of the major features associated with imprinting is the presence of parent-of-origin specific Differentially Methylated Regions (DMRs). Thus, the maternal and paternal genomes possess distinct epigenetic marks which distinguish them at imprinted loci. In order to identify novel imprinted genes on chromosome 15, we have profiled DNA methylation in cases with uniparental disomy of chromosome 15 (UPD15). Methylated DNA was immunoprecipitated using antibodies for 5-methyl cytidine, and hybridized to high-density oligonucleotide arrays with complete coverage of chromosome 15, generating profiles of the paternal and maternal methylomes. Comparison of six individuals with maternal versus paternal UPD15 reveals more than twenty DMRs on chromosome 15. Putative DMRs were validated by bisulfite sequencing, confirming the presence of parent-of-origin specific methylation marks in multiple samples. Many are associated with known imprinted genes within the Prader-Willi/Angelman syndrome region, such as *SNRPN* and *MAGEL2*, validating this as a method of detecting imprinted loci. However, more than half of the novel DMRs identified are located outside of 15q11-q13, and are associated with genes not previously thought to be imprinted. These include *IGF1R* at 15q26.3, which plays a fundamental role in growth regulation, and *GABRG3*, a gene which has previously been shown to be abnormally expressed in autism. Many DMRs occur at CpG islands or overlap conserved non-coding regions, suggesting a role in regulating gene expression.
These data provide an imprinting map of chromosome 15, demonstrate that the number of imprinted loci in humans is much higher than previously thought, and identify novel candidates for human disease.

P072

Are children and adolescents with rheumatologic disorders withdrawn from steroid therapy at risk for adrenal crisi?

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Background: Long term supraphysiological glucocorticoid (GC) therapy blocks the endogenous hypothalamus-pituitary-adrenal axis. Every GC therapy for more than 2–4 weeks harbors a risk for a steroid withdrawl syndrome (Addison's crisis). Therefore tapering steroids after long term treatment is mandatory.
Aim: To assess adrenal function after termination of GC therapy in pediatric patients treated in the ambulatory care centre of rheumatology at Universitiy of Berne.
Method: 19 patient (9 f/10 m) aged 2–15 years were withdrawn from steroid therapy (mean 1.4 mg/kg/d prednisone equivalent) with a standard protocol. Two to four weeks after termination of therapy adrenal function was assessed by determination of basal serum ACTH and cortisol as well as stimulated cortisol by standardized low-dose ACTH test. Evidence of adrenal insufficiency in low-dose ACTH test resulted in repetition of the assessment 6 months later.
Results: Follow-up by history revealed that none of the patients had symptoms or events suggestive of adrenal insufficiency following >12 months after steroid therapy. Only 3 patients used hydrocortisone replacement once as recommended during self-managed minor illness. Two to four weeks after steroid withdrawl 6 patients had

abnormal low serum cortisol levels at baseline and 10 patients failed the low-dose ACTH stimulation test. Importantly, only 4 patients had both low basal cortisol and failure in adrenal stimulation test. Six months later, in reassessment of the patients who failed in adrenal stimulation test 4 subjects had low serum cortisol at baseline and 7 failed adrenal stimulation testing again. Detailed analysis of the characteristics between the groups will be presented.
Conclusion: Patients with rheumatologic disorders who have been treated with high GC doses but have been tapered from their steroids according to current recommendations, seem to have no increased risk for adrenal crisis. However, when looking biochemically, adrenal function tests reveal insufficiency in 53% of the patients immediately after steroid withdrawl which persisted in 37% 6 months later.

P073

An uncommon cause of hypocalcemia in a newborn

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Introduction: Hypocalcemia is a common and frequent metabolic problem in newborns. It is usually detected by routine blood examinations or screening in trembling neonates. The varied causes include transient forms due to maternal disorders of the calcium homeostasis or diabetes mellitus and anticonvulsive treatment of the mother, but also persistent forms due to neonatal disease, such as Di George Syndrome, hypopara-thyreoidism or secondary forms due to neonatal hypomagnesemia.
Case report: We present a mature male neonate of a diabetic mother (weight 3640 g; P: >90%), delivered at 37 0/7 weeks of gestation with an uncomplicated adaptation. In the first hours of life, he developed symptomatic hypoglycaemia treated with intravenous glucose therapy. As irritability persisted despite correction of hypoglycaemia additional investigations lead to the diagnosis of a hyerphosphatemic (max 2.9 mmol/l; normal 1.5–2.2 mmol/l)) hypocalcemia (ionized Ca: min 0.78 mmol/l, normal 1.13–1.32 mmol/l; Ca total: min 1.4 mmol/l; normal ≥1.8 mmol/l) and hypomagnesemia (min 0.6 mmol/l; normal-Newborn: 0.8–1.1 mmol/l). Substitution with calcium and magnesium was initiated immediately followed by rapid clinical improvement and normalisation of serum calcium and magnesium. As the need for substitution with calcium and magnesium continued to persist beyond four weeks of life, we measured the parathormone level in serum which was in the low normal range (minimal 2.6 pmol/l = 24.7 pg/ml; normal 1.3–6.8 pmol/l) despite simultaneous low normal serum calcium concentration. Maternal causes for transient hypocalcemia were excluded and after six weeks we performed further investigations due to a suspected persistant endocrine abnormality. However at that time, PTH started to rise and substitution of calcium and magnesium could be reduced and stopped without any further need. Vitamin D metabolites, lymphocyte subsets and fluorescence in situ hybridisation of the diGeorge region on chromosome 22q gave normal results.
Conclusion: As demonstrated by this case history, duration of transient neonatal hypocalcemia due to maternal diabetes mellitus which usually lasts for a few days to a few weeks may persist longer than expected in some neonates.

P074

Extraadrenal Phaeochromocytoma as a Rare Cause of Arterial Hypertension in an Adolescent

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Introduction: In contrast to adults, phaeochromocytoma is rarely found in children. Two types of phaeochromozytoma can occur, an intra- and an extraadrenal one, the later called paraganglioma. The clinical symptoms are caused by high levels of plasma catecholamines.
Case report: A 16-year-old female adolescent was referred to our hospital because of arterial hypertension. Clinical history was characterised by episodes of hot flashes with sweating, blushing, and obstipation for about 2 years. She got an acute vision reduction three days before admission to our hospital. Blood pressure was elevated with 200/135 mm Hg. Echocardiography showed a severe concentric hypertrophy of the left ventricle (LV) without obstruction of the leftventricular outflow tract. Ocular fundus showed an edema of the macula and the papilla due to the arterial hypertension. High levels of urine and plasma catecholamines led to the diagnosis of a neuroendocrine tumor. MIBG-scintigraphy was negative, but DOPA-PET-CT demonstrated 2 tumours as a manifestation of 2 extraadrenal phaeochromocytomas, one intraabdominal, the other intrathoracical (40x24x29 mm and 8x14 mm respectivly). Diagnosis of bifocal extraadrenal phaeochromozytoma was made, and macroscopically total resection of the abdominal mass was done after initiating perioperative antihypertensive treatment with Doxazosine. Histological work-up confirmed the diagnosis and blood pressure normalised without any medication. The thoracic mass will be resected in a second operation. Due to possible syndromic features a genetic work-up was initiated.
Conclusion: Acute impairment of the vision due to arterial hypertension inducing severe concentric LV hypertrophy may be caused by phaeochromocytoma/paraganglioma even in adolescents. Diagnostic and preoperative work-up are complex and need interdisciplinary cooperation.

P075

Management of neonatal primary hyperparathyroidism with Cinacalcet after surgery failure

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Introduction: Neonatal primary hyperparathyroidism (NPHPT) can lead to rapid and severe complications: nephrocalcinosis, bone damage, neurological disabilities and death. Surgery is the treatment of choice and is effective in most cases. When surgery fails, trials with Biphosphonates were done with relative efficacy. We report a case treated successfully with Cinacalcet, a calcimimetic. To our knowledge, it's the first use of Cinacalcet in this indication in children.
Case report: A newborn girl presented with poor feeding, lethargy and apnoea. The laboratory values revealed: hypercalcemia, and hyperparathyroidism. The parathyroid gland was removed and the histological exam confirmed parathyroid chief cell hyperplasia. Postoperatively, PTH stayed high at 24 pmol/l and calcium at 5 mmol/l. The patient underwent six interventions (neck and arms) which were unable to detect the residual parathyroid tissue. The renal ultrasound showed nephrocalcinosis. The creatinine clearance was normal. The mineralometry showed a Z score at –2.2. The patient received a treatment with Biphosphonates, PTH decreased at 9 pmol/l and calcium at 3 mmol/l. In 2006, a trial with Cinacalcet was performed: PTH lowered to the normal range and calcium lowered to 2.7–2.9 mmol/l. No secondary effect of this treatment was observed.
Discussion: NPHPT is different from PHPT of adolescence witch is mostly secondary to parathyroid adenoma or MEN. NPHPT is usually associated with homozygote mutation of the Calcium sensing receptor protein (CasR) on the chief cell of the parathyroid gland. CasR plays a central role in calcium homeostasis through its ability to regulate PTH and renal calcium absorbtion. In normal subject, hypercalcemia leads to activation of the CasR, and inhibition of PTH secretion and calcium reabsorbtion. Cinacalcet, a calcimimetic agent of class II acts by enhancing the CasR sensitivity to calcium leading to PTH suppression calcium decrease.
Conclusion: Surgery remains the treatment of choice for severe NPHPT, our case illustrates that Cinacalcet can be a safe and efficient alternative to surgery.

P076

Hyperlipidemia management in children: a retrsopective study of 90 children

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There is convincing evidence that atherosclerotic process begins in childhood and is enhanced in case of associated high lipid levels. Inherited disorders of lipid metabolism are the major cause of lipid abnormalities in children. However, due to the epidemic of obesity, prevalence of secondary hyperlipidemia is increasing, leading to a threatening outburst of cardiovascular disease in the future. Prevention by lifestyle modifications (diet and exercise) and drug therapy in high-risk children is now widely recommended. We retrospectively studied 90 children referred to the Lipid Clinic in the Children Hospital of Geneva from 2002 to 2008. Age at referral, origin, aetiology, co-existence of additional cardiovascular risk factors, work-up and management were reviewed. Of the 90 children collected, 43% had a monogenic hypercholesterolemia, 28% a polygenic hypercholesterolemia, 14% a secondary hypercholesterolemia and 15% other dyslipidemia. Molecular analysis of the LDL receptor gene was performed in 70% of the children with monogenic disease with discovery of a mutation in 50%. Half of our patients had 2 or more cardiovascular risk factors : 33% had high Lp(a) levels, 27% had low HDL-C levels and 25% were overweight. High-risk group was defined as LDL-C levels >4.9 mmol/l or LDL-C levels >4.1 mmol/l in addition with severe familial history of atherosclerosis and/or other risk factor. These criteria were reached in 42% of our collective. In the high-risk group, children older than 6-year-old undergo a vascular exploration by measurement of intima-media-thickness (IMT) and research of endothelial dysfunction. Anomalies in the IMT (>0.5 mm) were found in 17/23 children (74%), with evidence of endothelial dysfunction in 40%. Drug therapy (ezetimibe and/or statins) was introduced in 21/38 children (55%), around the age of ten. No secondary effects, except

slight elevation of plasmatic creatine kinase level, were observed. Stabilisation or even regression of irregularities of the carotid wall was documented during the cardiac follow-up. These data showed that targeted tracking is mandatory already in childhood. More awareness of paediatric predictors of atherosclerosis is needed among physicians. Drug therapy is to be reserved to high risk children with careful follow-up, as criteria for risk stratification upon lipid levels are still lacking in childhood.

P077

Hypoglycemia and darkening of the skin – when hormone meets metabolism

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Introduction: Hypoglycemia is one of the most frequent metabolic disorders in children. Its rapid diagnosis and treatment is essential to prevent neurological complications. Multiple causes can lead to hypoglycemia, counterregulatory hormones may play a crucial role. Adrenal insufficiency is a potentially life-threatening emergency. Immediate clinical suspicion and treatment is mandatory.
Case-report: We report the case of a 5-year old boy from Erytrea who presented with severe hypoglycemia of 1.6 mmol/l and loss of conscience. Sodium was within normal range and patient history revealed high consumption of salt. Insulin levels were normal while cortisol was undetectable and ACTH markedly elevated >1250 ng/l. Diagnosis and treatment of primery adrenal insufficiency was established. Different causes of adrenal insufficiency such as solid tumors, Tuberculosis and HIV infection were excluded by ultrasound and laboratory tests. Even though an autoimmune disease is frequently associated with adrenal insufficiency, so far no antibodies could be found. MRI of the adrenals and photographs of the skin will be presented. We started substitutional treatment with hydrocortisone and fludrocortisone. Clinical follow-up was favorable.
Conclusion: Acute adrenal insufficiency is an uncommon cause of hypoglycemia and a wide range of exams is warranted. Once the diagnosis is made, lifelong treatment is standardized, including stressadaption of hydrocortisone dosages. Late diagnosis, especially delayed substitution, may have deleterious consequences.
Take home message: In case of hypoglycemia, adrenal insufficiency must be considered and treated with high priority.

P078

Administration of "tooth friendly" homeopathic globuli resulting in severe enteral bicarbonate loss in an infant

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Background: Homeopathy is frequently used in children as a first line treatment or additive to classical medicine. To make these globules "tooth friendly" and to lower the risk for caries, they are increasingly coated with sugar substitutes like Xylitol instead of saccharose. Non-digestible and/or non-absorbable sugar substitutes are known for their laxative effect.
Case report: We report on a 2-month old girl who was referred to our department because of a failure to thrive. She was breastfed in the first month of life. Thereafter, she was converted to formula milk. Simultaneously, she developed large amounts of daily watery stools and began loosing weight. The initially suspected infectious colitis was ruled out by inconspicuous stool cultures. Physical examination at admission revealed persistent diarrhoea and weight loss of 300 g in 3 weeks. There were no signs of dehydration except for dry lips. The girl was tachypnoeic with a deep and laboured breathing pattern. Blood gas analysis revealed a metabolic acidosis (pH 7.11) with severe bicarbonate loss (HCO_3 5 mmol/l, BE –22.7). There was no improvement after a resuscitation bolus of normal saline solution. Laboratory analysis showed normal anion gap, severe hyperchloraemia and low potassium level. Further laboratory investigations with regard to a metabolic disorder (ammonia, glucose, lactate, urine sample for aminoacids and organic acids) were normal. During hospitalisation, only slow compensation was achieved by permanent Na-bicarbonate infusion and potassium substitution. However, the girl regularly gained weight. When further questioning the mother, she reported an administration of Similasan®-globules, widely used as tooth friendly homeopathic globules against sleeping disorders. For the past few weeks she had given a dosage of up to 50 globules per day, accounting for a total daily dose of 100 mg/kg/d Xylitol.
Conclusion: Our case shows that Xylitol consumption does not only have laxative effects but may cause considerable weight loss and life threatening enteral bicarbonate loss when given in an overdose in infants. Paediatricians should be aware of this potential risk and inform parents.

Posters – Pneumology/Gastroenterology

P079

Chest CT in bronchopulmonary dysplasia: clinical and radiological correlations

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Introduction: Chest CT is very sensitive in assessing pulmonary damage in bronchopulmonary dysplasia (BPD). We established a CT score and investigated whether there was a correlation between radiological pulmonary lesions and relevant BPD clinical data such as gestational age, type and duration of mechanical ventilation and severity of BPD. We also assessed the score's usefulness in predicting complications of BPD.
Method: Retrospective review of 19 premature infants with BPD born between 1998 and 2007 who underwent at least one chest CT during their first year of life. A total of 29 CT were blindly evaluated by two radiologists for the presence or absence in 6 lobes (lingula included) of pulmonary parenchymal abnormalities described in BPD (areas of decreased attenuation, presence of bullae/emphysema, bronchial wall thickening, bronchiectasis, linear and subpleural opacities). This score was then compared with the most relevant clinical data and the long term evolution of these patients.
Results: All CT showed abnormalities. The most frequent lesion was bronchial wall thickening observed in all patients, followed by linear (89.5%) and subpleural (89.5%) opacities. Areas of decreased attenuation were found in 68.4%. Bullae/emphysema and bronchiectasis were the less frequent item described (26.3% and 21.1% respectively). The presence of areas of decreased attenuation correlated significantly with BPD severity ($P = 0.03$). Moreover, combining the presence of bronchiectasis with attenuation enables a more precise prediction of severity ($p = 0.02$). However, there was no significant correlation between the CT score and clinical data or middle term clinical evolution.
Conclusion: This study demonstrates the feasibility of a CT score in BPD and its usefulness in assessing the severity of this disorder. Areas of decreased attenuation seem the most sensitive to predict BPD severity. Though more patients are needed to validate this approach, these results could be helpful for further prospective studies.

P080

Comparison of three different methods of FeNO measurement in children

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Background: Measurement of fractional exhaled nitric oxide (FeNO) as a marker of airway inflammation may be useful in the diagnosis and management of asthma in children. Practicability is crucial for successful implementation of FeNO into asthma management.
Objective: The study aimed at comparing a conventional chemiluminescence NO analyser (EcoMedics®) with a hand-held device (NIOX MINO®) and offline FeNO measurement using a commercially available system in an unselected cohort of children aged 6–16 years. A secondary objective was to confirm FeNO stability over time in 15 samples from adult volunteers obtained using the offline system.
Methods: Sixty-six children (mean ± SD age 11.8 ± 3.0 years) underwent single breath FeNO measurement in triplets with each device. Offline collected FeNO was measured after offline breath collection into a Mylar balloon and subsequent analysis using the chemiluminescence NO analyser. Variability and between – method agreement were assessed, and stability over time within the Mylar balloons was tested by repeated hourly measurements.

Results: FeNO levels ranged from 2 to 113ppb. Intra-class correlation was excellent (r = 0.98, p <0.001 for each pair). Bland-Altman plots and back-transformation of logarithmic mean differences revealed fair agreement between methods. Stability over time was confirmed over 10 hours both at room temperature and when stored under cooling conditions.
Conclusions: FeNO values obtained using the chemiluminescence NO analyser, the portable NIOX MINO® system and the offline collection technique show between–method agreement lying within a clinically acceptable range.

P081

Fatal Asthma: Could it be prevented?

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Introduction: Asthma is the most common chronic disorder in childhood and adolescence. Over the last two decades, the prevalence seems to have increased, especially in industrialized countries. In contrast, the incidence of asthma-related deaths has decreased and fatal asthma in childhood is now a rare event.
Methods: An 11 year old boy with a sudden onset of dyspnoe at the playground alarmed his father by phone. On the father's arrival a few minutes later, the boy was cyanotic and subsequently soon lost consciousness. The father immediately brought him to the next hospital in his own car. At the hospital, the boy was in an asystolie and immediate resuscitation was started. After one hour of cardiopulmonary resuscitation circulation was reestablished. But the child had developed a severe hypoxic encephalopathy. Taking a careful history revealed symptoms attributed to asthma since the age of two years. There was only one hospitalisation for an asthma attack at the age of two years. Follow-up was irregular by few appointments with the paediatrician and the general practitioner. The boy had only short-acting beta2-agonists as needed, and during viral infections he used inhaled steroids for 1–2 weeks. There had never been a consultation with a paediatric pulmonologist.
Conclusion: It is debatable if this life threatening event could have been prevented under a consequent preventive therapy with inhaled steroids. Retrospective studies show that 39% of cases with severe asthma attacks have potentially preventable factors. Most of these children have mild or moderate asthma, and they often do not have their rescue medication with them. This case underlines the importance of an optimal therapy management and follow-up for all children with asthma to reduce mortality. There is an urgent need for good instructions for unexpected situations, and a written management plan for emergencies should always be given.

P082

Recurrent croup linked to IgA deficiency?

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Background: Recurrent croup is a well-recognized clinical entity characterized by repeated episodes of inspiratory stridor and barking cough. The causes of these episodes are thought to be viral infections, occurring especially before the age of 5 years.
Case report: We present a 5-year old boy with recurrent episodes of croup, 17 of which occurred during the last year without any seasonal pattern. Eight of these episodes required hospitalization. The patient's history was otherwise uneventful. The family history only revealed mild recurrent episodes of croup until the age of 5 in a second-grade cousin. Investigations including chest x-rays, fluoroscopy of the upper gastrointestinal tract, blood tests with differential blood counts, total IgE and measurement of C1-esterase-inhibitor activity and concentration were normal. Lung function testing showed moderate reduction of peripheral airway flows with good reversibility upon inhalation with beta-2 agonists. Nasal and oral exhaled nitric oxide was normal. Bronchoscopy revealed unspecific inflammation of the whole tracheobronchial tree, consistent with recurrent episodes of gastric fluid regurgitation. Streptococcus pneumoniae was the only organism isolated from bronchoalveolar lavage fluid. Immunological studies revealed total IgA deficiency as well as the absence of secretory IgA. The patient was treated with inhalative steroids, oral omeprazole and a two-week-course of amoxicillin to eradicate Streptococcus pneumoniae. During that time, the croup episodes did not re-occur, upon cessation of omeprazole symptoms resumed with two new croup episodes.
Discussion: This patient presents with an unusual frequency and severity of recurrent croup episodes. We speculate that several factors may contribute: (1) gastrooesophageal reflux, (2) airway hyperreactivity involving both upper and lower respiratory tract and (3) IgA deficiency. The exact contribution and a possible interaction of these entities in relation to recurrent viral infections will be discussed.

P083

Cutaneous manifestations as a rare initial presentation of cystic fibrosis

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Background: Cystic fibrosis (CF) is the most common life-threatening autosomal recessive disease among Caucasians, but is rare in the African population. It usually presents with pulmonary (e.g. recurrent infections) or gastrointestinal symptoms (e.g. failure to thrive). Over 1600 mutations located on chromosome 7 have been described with great variability in clinical phenotype particularly in regard to onset, progression, and severity of the disease.
Case report: We report the case of a 1 month old girl from a black African mother and a Caucasian father who presented with a rash on the cheeks and perineum. The rash consisted of exfoliative dermatitis with erosive lesions and was resistant to local treatment. It then progressed over the entire body. She developed diffuse edema and hepatomegaly with poor weight gain. Laboratory investigations demonstrated a normochromic normocytic anemia, hypoalbuminemia and low zinc levels. Skin biopsy suggested dermatitis consistent with a nutritional deficiency such as acrodermatitis enteropathica. Fecal elastase was markedly decreased but unfortunately sweat testing was not possible at this time due to the widespread dermatitis. Gastro-intestinal symptoms disappeared after introduction of pancreatic enzymes, zinc and vitamin supplementation with rapid resolution of the skin lesions. Sweat test performed later was clearly positive (Chloride = 90 & 86 mmol/l). CFTR gene sequencing revealed a compound-heterozygous genotype with F508del / T501I mutations; the latter a new missense mutation in exon 10 not previously described in the literature.
Conclusion: Cutaneous manifestations as the initial presentation of CF are rare. They are almost always secondary to malabsorption, therefore diminished fecal elastase as sign of exocrine pancreatic insufficiency, is the investigation of choice. This is particularly pertinent if the sweat test is not possible initially due to the condition of the skin. Furthermore, we recommend gene sequencing analysis to confirm the diagnosis where extended panel mutations have not identified two common mutations. In our case we found a new mutation for CF.

P084

Abdominal pain in cystic fibrosis: it is not always distal intestinal obstruction syndrome!

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Background: Distal Intestinal Obstructive Syndrome (DIOS) is a common complication in Cystic Fibrosis (CF), which presents with symptoms of bowel obstruction, ranging from chronic recurring cramping abdominal pain to acute obstruction with vomiting and abdominal distension.
Case report: A 17 year old male CF patient presented with a history of recurrent pain in the right lower abdominal quadrant. He was afebrile and showed no other gastrointestinal symptoms such as vomiting or diarrhea. His current therapeutic regimen consisted of daily chest physiotherapy, inhalations with salbutamol, ipratropium bromide, sodium chromoglycate, rhDNAse and tobramycin, pancreatic enzymes, vitamin and calcium substitution, as well as oral azithromycin. Clinical examination showed a localized tender pain without palpable mass in the right lower abdominal quadrant without any other pathological findings. Laboratory investigations (differential blood counts, blood sedimentation rate, C-reactive protein, pancreas-amylase, liver enzymes, creatinine and urinanalysis) were normal. Abdominal X-ray and abdominal ultrasound showed signs of stool impaction present within the whole colon. The patient was diagnosed as having DIOS and was treated with rectal enema of saline and oral acetylcysteine, after which he passed great amounts of stool with relief of the pain. Within the following two years however, the patient reported having recurrent pain during urination as well as recurrent abdominal pain as described above. Urinalysis revealed recurrent sterile leucocyturia and later bacteriuria with *E. Coli*. Uroflow measurements and ultrasound of the kidneys and of the urinary tract were normal. Urethritis was postulated and several courses of oral antibiotics given without significant relief of the pain. After two years, cystoscopy was performed and revealed a fistula between the intestine and the bladder. On abdominal MRI exam and later surgical intervention, chronic purulent appendicitis was diagnosed.
Conclusion: Appendicitis always needs to be considered in the differential diagnosis of abdominal pain in CF patients, especially as regular antibiotic treatment in these patients may conceal clinical symptoms of appendicitis.

P085

Beware of the nanny – recent cases of infant tuberculosis in Geneva

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Introduction: There has been a recent outbreak of tuberculosis in young children at the University Children's Hospital in Geneva. We report on 7 children diagnosed with active pulmonary or miliary tuberculosis in the past 12 months.
Methods: We reviewed children diagnosed with active tuberculosis at our hospital in the past 12 months. We identified 7 children aged 3 months–6 years (mean: 2 years 9 months), including a pair of twins.
Results: Diagnosis was based on clinical and radiographic findings including tuberculin skin tests and interferon gamma assays, and close contact with an adult index case. Surprisingly, 4/7 children did not travel outside Switzerland and contracted the disease through their nanny, while only 3/7 had a history of travel to a high-risk country.

Age	Symptoms	Chest X-ray and CT scan	BK
9 mths	Fever, cough	Consolidation, adenopathies, miliary infiltrates	positive
9 mths	None	Consolidation, adenopathies	negative
4 yrs	Fever, cough	Consolidation	negative
6 yrs	Cough	Consolidation, adenopathies	negative
4 yrs	Cough	Hilar adenopathy	negative
3 mths	Cough	Consolidation	negative
3 yrs	Cough	Hilar adenopathy	negative

Conclusion: We were impressed by the high number of infants recently diagnosed with active tuberculosis at our hospital. Risk stratification of tuberculosis patients usually focuses on age, immune status and country of origin. However, except for their young age, most of our patients did not belong to a group particularly at risk, but contracted the disease in Geneva, with the nanny being the source of infection.

P086

Chronic cough – not always harmless

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Background: Cough is probably the single most common complaint in children presenting to a physician. Most of these coughs are caused by viral infections and no investigations are necessary. In children with chronic cough (>3–4 weeks), the physician has to weigh the possible causes (e.g. congenital anomalies, foreign body, cystic fibrosis) in view of their prevalence at different ages. Rarely there are serious causes for chronic cough which demand immediate treatment.
Case presentation: A previously healthy 14 years old girl presented with a history of persistent cough for nearly one year, aggravating with additional shortness of breath and enlarging infraclavicular lymph nodes in the days prior to admission. CT scan of the chest showed a massive mediastinal mass and a pulmonary infiltration of the right lobe. A lymph node extirpation and a percutaneous lung biopsy revealed a Hodgkin's lymphoma of nodular sclerosing subtype, including lung involvement. Because of dramatic deterioration with respiratory insufficiency the girl was started on steroids before enrolling her on the European Hodgkin's lymphoma protocol (EuroNet-PHL-C1). After two days the child was free of pulmonary symptoms and we could start regular chemotherapy. Four months earlier a pulmonary examination with exercise testing was performed because of her persistent cough and exercise limitation. The physician postulated diminished physical fitness because of insufficient exercise. At that time a chest x-ray was not performed.
Conclusions: Hodgkin's lymphoma can develop over several months, and in case of pulmonary involvement, the only symptom can be chronic cough. A child with persistent cough should always be investigated carefully. After excluding common diseases like asthma and chronic sinusitis, a pulmonary examination should always include a chest x-ray to exclude rare diseases.

P087

A rare case of pulmonary hypertension in childhood

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Background: Pulmonary hypertension (PAH) is characterized by pulmonary vascular obstructive disease and right-sided heart failure. Here, we describe a rare cause of PAH in childhood.
Case report: A 15-month old hypotrophic female child was referred to our emergency department with unclear episodes of apnoea and cyanosis, resembling seizures or blue spells. Except for prematurity the patient's history was uneventful. The family history revealed that the parents were cousins of first degree and that a sister with known hypertrophic cardiomyopathy of unknown origin had died at the age of 17 months after similar episodes, without post-mortem examination having been performed because of ethnic reasons. In our patent, echocardiography revealed severe PAH. Right heart catheterisation (RHC) confirmed the diagnosis and showed good vasoreactivity to 100% oxygen. No structural cardiac defects were found. Bone morphogenetic type II receptor (BMPR-II) gene analysis to rule out familial pulmonary hypertension was normal. The patient remained stable under treatment with continuous home oxygen, diuretics and oral bosentan. Further evaluation by high-resolution CT of the chest showed bilateral diffuse ground-glass opacities. Subsequent thoracoscopic lung biopsy revealed multiple areas with thickening of the alveolar septae due to abnormal proliferating capillaries, and the diagnosis of pulmonary capillary hemangiomatosis was made. The patient unfortunately died at the age of 29 months of a pulmonary hypertensive crisis during a family visit in Tunisia. No autopsy was performed.
Comment: Pulmonary capillary hemangiomatosis (PCH) is an uncommon cause for pulmonary arterial hypertension. So far, only 16 paediatric cases have been reported in the literature. PCH has to be diffentiated from idiopathic pulmonary arterial hypertension to avoid life-threatening vasodilator-induced pulmonary edema. The differentiation is difficult and can only be done by lung biopsy. Suspicion should increase when CT scan shows signs not compatible with idiopathic pulmonary hypertension alone.

P088

Spontaneous, idiopathic chylothorax in 6-month old

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Background: Spontaneous chylothorax is a rare but life-threatening condition in children. Onset is mostly in the neonatal period and occurs seldom in older infants. For the clinician a major challenge lies in the decision between conservative and surgical management.
Case presentation: We report a 6-month old infant admitted in the pediatric emergency ward for progressive respiratory distress with subsequent finding of a left pleural effusion. Pleural drainage revealed a chylothorax which was treated successfully by chest tube placement and a combined conservative therapy with medium-chain triglyceride (MCT) diet, total parental nutrition (TPN) and 22 days of high-dose somatostatin treatment. The infant was discharged home on a MCT diet after 12 weeks without any acute sequelae. A normal diet was introduced at home after a delay of 6 weeks with no symptoms of reoccurrence of chylothorax.
Conclusions: Management of congenital chylothorax requires a multidisciplinary approach. Treatment options include pleural drainage, cessation of enteral feeding and initiation of TPN and may also compromise surgical treatment. Experience with somatostatin treatment is limited. Combined conservative therapy including high-dose somatostatin may stabilize idiopathic chylothorax in children.

P089

Eosinophilic esophagitis: an increasing entity in children

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Introduction: Eosinophilic esophagitis (EoE) is characterized by an isolated eosinophilic infiltration of the esophagus unresponsive to acid blockage. EoE is a growing clinical entity among both the paediatric and adult population with a reported prevalence of about 4/10'000 population. In the past, symptoms of EoE were often confused with gastroesophageal reflux disease (GERD). Thus, many children were unsuccessfully treated with medications used for GERD. As the incidence of EoE is increasing it is important for physicians to both identify patients with EoE and understand the diagnostic work-up and the available treatment options.
Methods: Patients diagnosed with EoE between 2004 and 2009 at the Children`s hospitals of Lucerne and Geneva were retrospectively enrolled. Demographical and clinical characteristics, symptoms at diagnosis, treatment and follow-up were studied. EoE was defined as 20 or more eosinophils per high-powered field on mucosal biopsies from the esophagus.
Results: 9 children (5 male/4 female) aged 1.5 to 14 years (median 7 years) were studied. They presented with nonspecific symptoms, including vomiting (n = 5), abdominal pain (n = 2), failure to thrive (n = 3), dysphagia (n = 3), food impaction (n = 4) and torticollis (n = 1). In 7 children allergic work-up revealed a sensitisation to allergens (3 perennial, 3 saisonal, 1 food). The time between the onset of symptoms and the diagnosis was 3 months to 8 years. Treatment of EoE included topical corticosteroids (n = 8) and/or dietary (n = 3), i.e. elimination or elemental diet. In one case esophageal dilatation was necessary because of secondary esophageal narrowing. In all cases the symptoms improved under therapy.

Conclusion: The clinical presentation of EoE varies widely and the symptoms are quite unspecific. It is important to recognize EoE as differential diagnosis to GERD, as correct treatment alleviates symptoms, decreases the risk of food impaction and induces histological remission. However, more has to be learned about the natural history of this "new" disease.

P090

Severe eosinophilic enterocolitis in newborn with abnormal fœtal echography

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Introduction: Eosinophilic enterocolitis is relatively rare in newborn and infants. At our knowledge, there is no previous describing a subject with antenatal abnormal bowel echography, fast and severe developpement of symptomatology after birth.
Case: C. was born at 35 SA 3/7, birth weight is less than percentile 10. Routine foetal echography at 30SA, reveal important bowel dilatation without systemic impact on the fœtus. First meconium was emited in delivery room, it was accompagned by macroscopically red blood. C. didn't need any support and start maternal milk feeding. In the first 24 h, apparition of frequent vomiting and repeated bloody stools. Blood eosinophilic count at peak was 4990 G/l. We first maked a barium contrast, with suspicion of gastric malposition. The first signs of intestinal obstruction, associated with bad general statement and increased capillary refill were noted at day 8. Sepsis, necrozising entercolitis and bacterial and parasitic gastroenteritis were exclueded. Antibiotics and total parenteral nutrition were started withdrawal of enteral feeding. When C. recovered at day, we started with a hydrolysat milk formula. Because of symptomatology reappearance at day 18, an endoscopy with bopsies was performed. Results revealed the presence of eosinophilic infiltration in the mucosa of the small and large bowel as well as in the rectum. Specific and total IgE were normal and skin prick tests were negatives. 16 days after introduction of a amino-acid milk, C. recovered completely. Discharge of hospital ad day. Now, C.is 7 month old, growth at percentile 25 and developpement is normal. There is no atopic history in the relatives.
Discussion: Bowel enlargement in foetal echography was prior associated with surgical gastro-intestinal abnomalies, but however neonatal eosinophilic enterocolitis must be part of the differential diagnosis, even if it was a fortuitous discovery in our case. The immunological mechanisms were not well understood, however the antigenic presentation in utero could be the same as seen in cow's milk allergy. The recognition of this pathology and important and the fast dietary change is important. Challenge to reintroduction at 12 month is possible with precaution.

P091

A rare cause of chronic diarrhea in infancy: a case report

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Introduction: Neuroblastoma and ganglioneuromas can be associated with secretory diarrhea, hypokalemia, achlorhydria and metabolic acidosis due to secretion of Vasoactive Intestinal Peptide (VIP) by the tumour. In children the VIP syndrome is often confused with infections or malabsorption syndromes.
Case: We present a 14-month-old girl suffering from progressive diarrhea since the age of 6 months, abdominal distention and failure to thrive since the age of 12 month. At first consultation in our hospital she presented with four watery stools per day without signs of dehydration, but hypokalemia (2.5 mmol/l) and metabolic acidosis (pH 7.27, Bic 11 mmol/l, base excess –14 mmol/l). In a meticulous diagnostic workup, malabsorption syndromes, maldigestion, infectious, metabolic and immunological causes explaining her diarrhoea could be excluded. Endoscopic evaluation of the upper and lower gastrointestinal tract including mucosal biopsy were not diagnostic. Finally, both an increased VIP serum level and increased urine catecholamines were detected. After a vigorous search, a computer tomography-assisted somatostatin-receptor scintigraphy revealed a pelvic, prevertebral tumour located in the sacral region. Retrospectively, this tumour was present in both examinations, pelvic ultrasound and MRI scan, however, was interpreted as faecal masses. After gross tumour resection gastrointestinal symptoms resolved completely. Histopathology revealed a well differentiated neuroblastoma with favourable histology and without amplification of the oncogene MYCN. According to the postoperative staging and the favourable biological criteria of this neuroblastoma, the patient was categorised in the low risk group, and accordingly, a "wait and see" strategy was chosen with no further therapy. Four months after tumour resection the patient remains clinically, radiologically and metabolically (normal serum VIP, normal urine catecholamines) in remission.
Conclusion: Although neuroendocrine tumours are rare causes for secretory diarrhea in children, after exclusion of other common causes, clinicians should consider the differential diagnosis of a VIP syndrome in the context of an endocrine active neoplasia, especially if hypokalemia and metabolic acidosis are present.

P092

Unexpected recovery from fulminant liver failure caused by *amanita* poisoning in a young infant

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Introduction: *Amanita phalloides* is responsible for the majority of fatalities caused by mushroom poisoning, leading to fulminant liver failure with a mortality rate of 40 to 80%. There are no well designed randomized trials addressing the efficacy of standard treatment strategies. The only proven therapy for patients unlikely to recover from acute liver failure is liver transplantation. Survival rate without transplantation is 10 to 30%. Several sets of criteria for emergency transplantation for patients with acute liver failure have been proposed, however they have not been clearly assessed in the context of *amanita phalloides* poisoning. Fatal prognostic factors are: short interval between ingestion and diarrhoea, young age (<10 y) and severity of quick and prothrombin perturbation. Here we report a case of an infant with *amanita phalloides* intoxication and fulminant liver failure, with several fatal prognostic factors and transplantation criteria, who underwent spontaneous recovery in the hours prior to the programmed living donor liver transplantation, by signs of factor V increase.
Methods: Case report of a 17-month old boy with *amanita phalloides* poisoning.
Results: Mild symptoms (vomiting and diarrhoea) began 6 hours after poisoning (ingestion of pasta soaked with a sauce of *amanita phalloides*). Diagnosis was confirmed by positive urine alpha-amanitin. A conservative treatment with silibine and active charcoal was immediately introduced. 36 hours after poisoning the boy presented with liver failure and a stage I encephalopathy; prothrombin time was >100 ms, factor V 10%. Hepatic cytolysis peaked at 60 hours after ingestion (ASAT 14250 U/l, ALAT 14118 U/l), with factor V <10%. The infant was programmed for very urgent living-donor liver transplantation. Yet, 72 hours after ingestion spontaneous increase of factor V (30%) was observed and the transplantation postponed. Liver function resumed after 14 days and the boy is in perfect general condition at 6 months after poisoning.
Conclusion: Liver transplantation should rapidly be considered in children with *amanita phalloides* poisoning. Nevertheless liver function should be closely monitored until the intervention, because of possible spontaneous recovery even in fatal appearing cases, and even in very young infants; factor V monitoring seems a good assessment parameter.

P093

Portal hypertension due to portal thrombosis as primary manifestation of Alagille syndrome

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Introduction: Syndromic paucity of bile ducts or "Alagille syndrome" is characterized by peculiar facies, chronic cholestasis, posterior embryotoxon, butterfly-like vertebral arch defects and vascular malformations, e.g. peripheral pulmonary artery hypoplasia or stenosis. Almost 90% of Alagille syndrome patients carry a mutation in JAG1 gene. We present a 3-year-old female child with "partial" or "incomplete" Alagille syndrome. She had no significant liver pathology, but other major syndromic features were present.
Case report: A 3-year-old girl was admitted for severe upper gastrointestinal bleeding. Clinical examination revealed a characteristic facial appearance, splenomegaly and heart murmur. There was no evidence of cholestasis, coagulopathy or liver dysfunction. Liver ultrasound showed no irregularities, but revealed significant portal hypertension with vascular oesophageal derivation. CT scan indicated portal thrombosis. Endoscopy showed esophageal varices grade II-III as the cause of bleeding. Further work-up revealed cardiac malformation (peripheral pulmonary arterial stenosis), butterfly vertebra and posterior embryotoxon. Liver biopsy demonstrated mild peri-sinusoidal fibrosis, but neither paucity of intrahepatic ducts, nor cholestasis were noted. The suspected diagnosis of Alagille-Syndrome was confirmed by genetic findings of a mutation in the JAG 1 gene (p.Cys196Gly). A mesenteric-portal vein-bypass (Rex-Shunt) was performed to restore physiologic blood flow to the liver with favourable outcome.
Discussion: We present a patient with upper gastrointestinal bleeding (GI) as primary manifestation of Alagille syndrome. GI bleeding occurred secondary to portal hypertension, probably caused by portal thrombosis due to vascular malformation. This case illustrates the phenotypic variability of Alagille syndrome and emphasizes that absence of cholestasis, jaundice or liver dysfunction does not exclude Alagille syndrome. Genetic testing (JAG 1 gene) allows to confirm the diagnosis.

P094

Respiratory Picornaviruses as Major Cause of Morbidity in Children – a Retrospective Study

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Introduction: Picornaviruses have emerged as major causes of lower respiratory tract infections in children. However, children often suffer from co-infections with other respiratory viruses, questioning the relative contribution of picornaviruses to clinical manifestations.
Methods: In 344 patients admitted between November 2006 and October 2007 because of an acute respiratory tract infection, we performed respiratory virus testing from nasopharyngeal secretions using in parallel both a direct fluorescent antibody panel (DFA) and a Luminex® multiplex polymerase chain reaction (PCR) testing for influenza A and B, hMPV, adenovirus, parainfluenza 1-4, picornaviruses, RSV A, B, and coronaviruses. All children with comorbidities and known respiratory coinfections were excluded resulting in a homogenous group (n = 111) of previously healthy children with picornavirus infection. Patients with positive PCR and DFA (group A) and those with positive PCR, but negative DFA (group B) were compared with respect to symptoms, duration of hospitalization and complications. Three DFA positive, but PCR negative cases were excluded.
Results: 108 patients were enrolled, with 84 patients in group A (78%) and 24 patients in group B (22%). There was no significant difference between group A and B regarding median age (18 months), duration of hospitalisation (3.0 vs. 3.5 days) or duration of oxygen requirement (1.0 day). Group A was more commonly given the diagnosis of obstructive bronchitis (71% vs. 46%, p = 0.04), was more commonly described to wheeze (79% vs. 58%, p = 0.05), and to have retractions (60% vs. 38%, p = 0.07). Group B more often presented with fever (46% vs. 25%, p = 0.05).
Conclusion: Picornaviruses cause substantial in-patient morbidity in otherwise healthy children. Using PCR as the gold standard, there were no substantial clinical differences between patients with positive or negative DFA regarding the severity of infection. Thus, DFA may be a cost-saving technique to diagnose picornavirus infections.

P095

Human Metapneumovirus Infections – Biannual Epidemics and Clinical Findings in Children in the Region of Basel, Switzerland

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Human metapneumovirus (hMPV) epidemics vary in time and severity. We report findings for PCR for hMPV and respiratory syncytial virus (RSV) performed on nasopharyngeal aspirates (NPA) of hospitalized and outpatient children with respiratory tract infections between October 2004 and April 2008. A total of 3934 NPAs were tested for hMPV and 3859 for RSV. Of these 198 (5%) were hMPV positive and 869 (23%) were RSV positive. Median age was 17 months and 9 monthsfor hMPV and RSV, respectively. 59% of hMPV and 58% of RSV patients were hospitalized. Proportions of hMPV positive samples for the four winter seasons were 0.4%, 11%, 0.2% and 14%.
For RSV, they were 28%, 15%, 28% and 28%.
Conclusions: HMPV epidemics follow a biannual variation in our area. Major epidemics were observed in winter seasons staring in odd years (2005/06 and 2007/08), minor epidemics in those starting in even years (2004/05 and 2006/07). RSV epidemics usually follow a reciprocal biannual pattern, leading to annually alternating major RSV and hMPV epidemics.

P096

Immune response to influenza vaccination in children treated with methotrexate or/and tumor necrosis factor-alpha inhibitors

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Introduction: In children treated with immunsuppressive medication such as methotrexate and tumor necrosis factor-alpha (TNF-α) inhibitors, additional immunization is recommended due to elevated susceptibility to infections. However, it is unclear if adequate antibody response to vaccinations can be established in children receiving methotrexate and/or TNF-α inhibitors.
Methods: In a prospective open label study, we assessed seroconversion and seroprotection upon influenza vaccination in 31 children with autoimmune disease treated either with methotrexate (n = 16), TNF-α inhibitors (n = 7) oder both (n = 8) and a control group of 10 immunocompetent children. Influenza antibody titres were determined by haemagglutinin inhibition assay, before and 4–8 weeks after vaccination.
Results: Pre-vaccination seroprotection (titre ≥1:40 of ≥2 of 3 influenza strains) was present in 42% of the treatment group and 30% of the control group. After vaccination, a protective titre was achieved in 87% of the treatment group and 90% of the control group. Seroconversion was defined as the change from a negative titre (<1:40) to a protective titre (>1:40) with at least a 4-fold titer increase. This was documented in 57% resp. 50% (B strain), 46% resp 75% (A/H3N2 strain) and 58% resp. 80% (A/H1N1) in the treatment group resp. control group. Safety evaluation of vaccination showed no serious adverse events.
Conclusion: Children under methotrexate and/or tumor necrosis factor-alpha (TNF-α) inhibitors can be safely and effectively immunized against influenza, with a seroprotection after vaccination comparable to immunocompetent children.

P097

Interpretation of primary care physicians' attitude regarding routine rotavirus immunization using diffusion of innovation theories

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Objective: To evaluate primary care physicians' attitude towards implementation of rotavirus (RV) immunization into the Swiss national immunization schedule.
Design and settings: An eight-question internet-based questionnaire was sent to the 3'799 subscribers of InfoVac, a nationwide web-based expert network on immunization related issues which reaches >95% of pediatricians and smaller proportions of other primary care physicians. Five demographic variables (gender, specialty, year of final examination, work environment, area of residence) were also inquired. Descriptive statistics and multivariate analyses for the main outcome variable "acceptance of routine RV immunization" and other variables were performed. Dissemination of innovation theory was used for final assessment of all data.
Results: 977 questionnaires were returned (26%). Fifty percent of participants were pediatricians. Based on the current burden of disease, routine RV immunization was supported by 146 participants (15%) – so called early adopters –, dismissed by 620 (64%), leaving 211 (21%) undecided. However, when asked whether they would recommend RV vaccination to parents if it were officially recommended by the federal authorities and reimbursed, 467 (48.5%; so called early majority) agreed to recommend RV immunization. Multivariate analysis revealed that physicians who would immunize their own child (OR 5.1; 95% CI 4.1–6.3), hospital-based physicians (OR 1.6; 1.1–2.3) and physicians from the French speaking area (OR 1.6; 1.2–2.3) were significantly more likely to support RV immunization.
Conclusion: Diffusion of innovation theory predicts successful implementation if 50% of a given population (early majority) support an innovation. Thus, introduction of RV immunization in Switzerland is likely to be successful, if the (i) federal authorities issue an official recommendation and (ii) costs are covered by basic health care insurance.

P098

Complicated Sinusitis: Case Series

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Introduction: Children experience frequent infections of the upper respiratory tract per year. Of those, 0.5–10% develop a bacterial sinus infection. Intracranial spread of this infection is a rare but serious complication. Imaging studies usually are not necessary to confirm the clinical diagnosis and guide treatment in uncomplicated cases. However, high-resolution CT and/or MRI-imaging should be considered in cases with atypical presentation or suspicion of intracranial complication.
Methods/results: In a population-based retrospective case series, 4 children with complicated sinusitis seen in our institution between December of 2007 and July of 2008 were enrolled. Age ranged from 8.5 to 13.1 years. All children presented with leading symptoms of headache and fever. Symptoms for central nervous system involvement included signs of raised intracranial pressure (vomiting), of meningeal inflammation (headache, meningism) and of focal neurological deficit (seizures, urinary retention/incontinence, hearing loss, paresis of the abducens and trigeminus nerves). Focal meningoencephalitis was found in two patients, meningitis in one patient. CT scan demonstrated pansinusitis in all cases; in addition, one patient each had intracranial-intraorbital empyema and thrombosis of the sinus cavernosus/sagittalis. Pathogens included S. pneumoniae (n = 2, isolated from blood and CSF culture) and group A streptococcus (n = 2, isolated from blood and nasopharyngeal swab culture). All children were initially treated with ceftriaxon or amoxicillin + clavulanic acid (1 case); antibiotic regimen was specified according to

microbiological results. All patients underwent endoscopic sinus surgery (drainage, puncture or incision of the sinus) and recovered completely.
Conclusion: Complicated sinusitis is a rare event but can lead to serious cerebral consequences. Immediate intravenous antibiotic treatment is justified in case of clinical suspicion of intracranial complication.

P099

A Bump to the head: Major Complications after Minor Head Injury

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Introduction: We present an unusual clinical course following a banal head injury.
Case report: After trivial banging the head at the "Maiensäss" (alp) a 7 year old boy developed a painful swelling measuring 3x4 cm. One week later he knocked his head again and putrid discharge was noticed. The lesion did not respond to a treatment consisting of several surgical revisions and oral antibiotic therapy with amoxicillin / clavulanat. Swab cultures remained negative for bacteria and fungi. On first admission in our hospital three weeks later, the scalp lesion had further increased with a purulent centre and a scaly border. Debridement was performed and smears as well as a biopsy from the margin of the lesion were taken. Assuming a deep fungal infection a therapy with systemic Itraconazol and topical silver-sulfadiazin was commenced. The antibiotic therapy was continued for another week. After a three months course of systemic Itraconazol and regular wound dressings, epithelialization was completed with a remaining extensive scar formation and alopecia. The causative pathogen identified was Trichophyton verrucosum found only in the biopsy but not in swab cultures. The extended history revealed intensive contact to cattle at the "Maiensäss" carrying the same germ.
Discussion: Kerion celsi is a deep localized tumor-like trichophytia mainly on the scalp. It was named by the ancient Roman physician Celsus and means 'the honeycomb'. Animal transmitted fungal skin infection can present with severe inflammatory reaction and may be mistaken for bacterial abscess, leading to incorrect treatment with antibiotics. In case of unsuccessful antibiotic therapy, fungal infection must be actively investigated and often a biopsy is needed to elicit the pathogenic agent. Typical carriers are cattle and pets as guinea pigs and hamsters. Systemic steroids are controversial whereas topical steroids are disapproved in general.
Conclusion: Deep suppurative skin lesions unresponsive to treatment should prompt early attention to fungal infection. A biopsy of the margin of the lesion should be performed. Smears are often misleading and may delay appropriate therapy, resulting in more extensive scaring alopecia. In general, systemic antifungal treatment is required for several weeks to months to achieve infection control.

P100

Tick-borne encephalitis presenting with fever without localising signs

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Introduction: Fever accounts for up to 17% of all paediatric emergency department visits. Twenty percent of febrile children have fever without localising signs (FWLS). Routine investigations include urinalysis and chest radiography. In older children performing a lumbar puncture is optional when clinical signs of meningoencephalitis (ME) are lacking. Tick-borne encephalitis (TBE) is diagnosed easily in patients with fever and signs of ME. Diagnosis of TBE in patients without neurological signs will only be established if lumbar puncture and serological blood testing is considered.
Methods: Between 2000 and 2008 all cases of TBE occurring in children were assessed on the basis of clinical records. In all these cases those with fever as the leading manifestation were further analyzed.
Results: The case definition of FWLS (in particular without neurological signs), CSF pleocytosis and positive TBE serology in blood was fulfilled in 4 children (3 male, 1 female). The median age was 5.8 years with a range of 4 to 9 years. All patients presented with fever (38.5–40.0 °C). This was the second febrile episode with an symptom-free interval. Neurological examination was normal, there were no signs of ME. All cases were associated with a reduced general condition and headache. 3 cases presented with additional vomiting. Urinalysis (all patients) and chest radiography (2 patients) were normal. Tick exposure was remembered in one case. Lumbar puncture was performed in all patients: CSF examination showed a pleocytosis (total cell count 67–164/µl, mononuclear 27–57), values for protein were 304–577 mg/l and glucose 2.9–4.1 mmol/l. Bacterial cultures were all negative. There were raised inflammatory parameters: leucocytosis (11.9–34.5 G/l) and/or elevated C-reactive protein (3–69.7 mg/l). The detection of specific IgG and IgM established the diagnosis of TBE.
Conclusion: The aim of this study was to evaluate cases of TBE virus infection without signs of ME. TBE virus has to be considered as a causative agent of flu-like symptoms in children. In these cases lumbar puncture and blood serology are essential for establishing the diagnosis of TBE. This approach alleviates the need for ancillary testing and antibiotic treatment. TBE is a mandatory reportable disease in Switzerland. Our strategy to perform a lumbar puncture for case inclusion of TBE in patients with FWLS most likely results in an increase in the incidence for TBE in Switzerland.

P101

Liver abscess complicating a perforated appendicitis

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Background: Complicated inflammatory disease of the appendix is commmon in children. Peritonitis and pelvic/abdominal abscesses are the most frequent sequellae. On the other hand, pylophlebitis (portal thrombosis and infection) and liver abscesses (LA) are rarely reported nowadays, but were much more common in the preantibiotic era. Because of its potential severity, early recognition of a LA as a complication of a (ruptured) appendicitis is of the utmost importance.
Case presentation: A previously healthy 3-year-old girl presented with initial typical signs and symptoms of acute appendicitis, confirmed to be perforated at operation. IV antibiotherapy with co-amoxycillin was started, for 7 days, with a good clinical answer (no fever or abdominal pain). Three days after finishing antibiotics, she presented high fever again, with abdominal pain. On physical exam, there was abdominal tenderness and guarding, but no hepatosplenomegaly. An abdominal ultrasound disclosed a right liver lobe abscess. Intravenous Augmentin® therapy was immediately started, with a rapid and complete clinical recovery, obviating the need for percutaneous drainage. Cultures of peritoneal fluid grew E.coli and Enterococcus faecium, both sensitive to Augmentin®. Six months later, she is doing well, with no clinical signs of portal hypertension, and with a normal abdominal ultrasound-doppler.
Discussion: Intrahepatic abscess is, nowadays, an extremely rare complication of (ruptured) appendicitis. This case report calls for the following concluding remarks: 1) in case of prolonged fever in a child with appendicitis, a complete abdominal echographic survey should be obtained to rule out an abscess formation, either in the liver or peritoneal cavity; 2) portal hypertension is a known complication of pylephlebitis. Children with liver abscess should be followed then accordingly (clinical and ultrasonographic followup).

P102

A rare cause of abdominal lymphoma and intussusception

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Introduction: Intussusception are mostly caused by viral infection or idiopathic at infancy.
Case presentation: A girl presented with a history of nausea, emesis, diarrhea, weight loss and inappetence after a four week vacation in India. Abdominal ultrasound showed a small bowel intussusception. Upon further examinations (abdominal CT with combined sellink techniques to show the small bowel, Meckel's scintigraphy, *bone scintigraphy* and MRI) an intra- and retroperitonial mass suspicious for abdominal lymphoma was discovered. Broad blood and urine testing, microbiological and parasitological stool tests were performed. In two different stool cultures gardia lamblia was found. Treatment for seven days with metronidazol was applied. Finally a laparoscopy with biopsy revealed reactive changes in structural normal lymph nodes without malignity. Up to now the patient has shown complete healing and normal weight gain.
Discussion: In all imaging methods there was always suspicion of lymphoma. At the end infection with gardia lamblia was the reason for the abdominal lymph node reaction and distribution. Though unusual for our region lambliasis was described before causing lympho-proliferative diseases of benign character.
Conclusion: An intussusception can be caused by a benign lymphoma induced by the infection with gardia lamblia. Furthermore this case could be a request for broadening of our diagnostic views in a global migrating society.

P103

Brodie's abscess of the left tibia caused by *Brucella*: an unusual presentation of an uncommon pathogen

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Clinical history: An 11-year-old girl of Portuguese origin presented to our outpatient service with a one-week complaint of left lower limb pain, without fever or other systemic symptoms. On physical examination, palpation of the proximal tibia elicited maximal pain, whereas knee movement was not limited, but the degree of the pain restrained walking. Complete blood count was normal and biological makers of inflammation were low (CRP 18, ESR 18 mm/h). CT scan and MRI showed a 6 x 4 mm elliptic lesion in the growth plate of the proximal metaphysic of the left tibia, with minimal surrounding edema, compatible with a bone abscess. Due to localization and size of the abscess, surgery was excluded to avoid definitive sequelae on the growth cartilage. Three blood cultures were drawn at the time of admission and treatment of flucloxacilline 150 mg/kg/j i.v was initiated. After seven days, blood cultures were positive for an initially unidentified Gram-negative coccobacillus. A positive identification of *Brucella* sp was obtained by molecular biology. The patient had traveled to a rural area in Portugal every year, but had no recollection of exposure to unpasteurized milk products. Treatment was switched to rifampicine and doxicycline for a total duration of 6 weeks leading to rapid resolution of signs and symptoms.
Discussion: Brucellosis is the most common zoonosis worldwide, but is present in Switzerland only as a disease imported by travelers and migrants. It is caused by the ingestion of unpasteurized milk products. The protean nature of the clinical manifestations contributes to the diagnostic challenge in non-endemic countries. Brucellosis should be kept in mind as a possible cause of osteoarticular disease, prolonged fever or abdominal symptoms in children who have traveled to countries where the pathogen is endemic.
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P104

Therapeutic options in chronic recurrent multifocal osteomyelitis (CRMO): from non-steroidal anti-inflammatory drugs (NSAIDs) to TNF-α inhibitors

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Background: CRMO is characterized by a prolonged, fluctuating course with recurrent episodes of pain occurring over several years. CRMO is multifocal and mostly seen in long bones as tibia and in the clavicle, less frequently the spine and pelvic bones. The radiographic appearance suggests subacute or chronic osteomyelitis. Histopathological and laboratory findings are nonspecific. Bacterial culture is negative. CRMO is diagnosed by exclusion of the two main differential diagnoses – bacterial infections and tumor.
Methods: we report 6 patients (9,11,15,16 and 17 year old females, 15 year old male), each with distinct disease onset, time of diagnosis, evolution, therapy and outcome
Results: In our patients symptoms began between the age of 4 to 13, definitive diagnosis was made after 2 month up to 2 years after first symptoms. 4/6 showed typical multifocal location for pain two girls had just one fokus. In all patients laboratory findings were unspecific with elevated ESR. A histology was done in 5/6 Patients, which showed unspecific chronic inflammation, bacterias could not be found. HLA-B 27 was negative in all patients, ANA in 3/6 positive. 4/6 patients got at the beginning of symptoms a therapy with antibiotics. 4/6 patients had a good improvement with NSAIDs. 2 girls had a prolonged course of disease with several relapses. High doses of analgetics, steroids, calcitonin and bisphosphonates were applied without success. Only therapy with TNF-α inhibitors was able to control disease.
Conclusion: CRMO is important to consider in the differential diagnosis of septic osteomyelitis, bone tumors, juvenile idiopathic arthritis and histiocytosis. Most cases can be successfully treated with NSAIDs. In therapy resistant CRMO cases TNF-α inhibitors is a promising therapeutical option.

P105

PFAPA syndrome: A new consensus on classification criteria

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Introduction: PFAPA syndrome is diagnosed according to criteria proposed initially and modified by different authors. They are based on clinical features and the exclusion of other periodic fever syndromes. An analysis of a large cohort of patients has shown weaknesses for these criteria. We aimed to propose revised classification criteria based on a consensus among international experts in the field.
Methods: We used the Delphi method followed by a consensus conference. For a specific question, consensus was considered to be reached if at least 80% of the participants agreed. 20/21 contacted experts accepted to participate to the Delphi survey. 11 experts participated to the consensus conference on November 2008 in Morges.
Results: With the Delphi survey, the experts proposed first a list of criteria they would consider for diagnosis. In a second step, they selected the most relevant criteria for diagnosis of PFAPA syndrome. The following criteria were accepted during the consensus conference: 1. Periodic Fever for at least 6 months (a. Daily fever of at least 38.5 °C (axillar) for 2 to 7 days, b. At least 5 regularly recurring fever episodes with maximum of 2 months interval between them.); 2. Pharyngitis, cervical adenitis, oral aphtae: at least one in every episode and at least 2 out of 3 in the majority of episodes; 3. Exclusion of other causes of recurrent fever (clinical or by laboratory depending on history); 4. Exclusion of infections, immunodeficiency and cyclic neutropenia; 5. Disease onset before the age of 6 years; 6. Full recovery between episodes; 7. Normal linear growth; Supportive criterion: Prompt response to steroid treatment. The exact definition and descriptors for these criteria were also reviewed by consensus.
Conclusion: Based on the consensus of a group of international experts in the field of periodic fever syndromes, a new set of classification criteria has been proposed. This classification needs now to be tested prospectively on a cohort of patients with periodic fever to assess the sensitivity and specificity of these criteria.

P106

Age-dependency of sting recurrence in children with hymenoptera venom immunotherapy: higher prevalence of stings but fewer systemic reactions in younger children

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Introduction: Hymenoptera venom allergies in young patients are of great concern because children tend to be more prone to re-stings than adults.
Methods: 83 children 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. Mean follow-up period was 7.7 years after commencement of VIT. Group 1 included preschool children (<6 years, n = 17), group 2 school children (6–10 years, n = 39) and group 3 young adolescents (>10–16 years, n = 32). Endpoints were rate of re-stings and percentage of SR to re-stings in relation to age.
Results: 49 children (59%) had been re-stung 108 times by the insect they were allergic to. Younger children were being stung significantly more often than older children: the rate of re-stings per year was 0.41 in group 1 (preschool age), 0.21 in group 2 (school age) and 0.15 in group 3 (adolescents) respectively. Regression analysis showed a significant age dependency in prevalence of re-stings ($p = 0.001$). SR upon re-sting were fewer in younger children with 1/29 stings (3%) in group 1, 2/47 stings (4%) in group 2, and 5/32 stings (15.6%) in group 3. Differences in prevalence of SR were significant ($p < 0.05$) between preschool and school children (group 1 und 2) as compared to adolescents (group 3). Overall, there was a trend for higher prevalence of SR upon re-stings with increasing age ($p = 0.079$).
Conclusion: A majority of children with hymenoptera venom allergies are being re-sting. The younger the patients, the higher the prevalence of re-stings. In younger children however, recurrence of systemic reactions upon re-stings is lower as compared to adolescents.

P107

Successful treatment of macrophage activation syndrome as initial presentation of systemic onset juvenile idiopathic arthritis with interleukin-1-antagonist anakinra in a 3 years old girl

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Introduction: Macrophage activation syndrom (MAS) is a potentially life-threatening complication of systemic childhood inflammatory disorders, such as systemic onset juvenile idiopathic arthritis. The immunopathogenesis of MAS is characterized by excessive activation and proliferation of T lymphocytes and macrophages resulting in hemophagocytosis. Immunosuppressive medication such as corticosteroids and ciclosporin are currently used, but can fail in severe cases. This is the first report of successful treatment of MAS with anakinra, an interleukin-1-receptor antagonist.
Methods/results: A 3 years old girl presented with a 3 days history of refusal to walk with fever up to 40°C. Due to remarkably elevated inflammation markers the girl was started on antibiotic treatment for suspected osteomyelitis. However, bone scintigraphy could not reveal any osseous focus. Infectious causes (cultures in blood, urine, cerebrospinal fluid and extensive serologies) could be excluded. The girl developed macular exanthema, hepatosplenomegaly, generalized lymphadenopathy, tenosynovitis on hands and feets, anemia, hyperferritinemia and finally presented encephalopathy with status epilepticus. Presence of phagocytosis by macrophages in cerebrospinal fluid and bone marrow confirmed our suspicion of macrophage activation syndrome. High dose corticosteroids, ciclosporin and intravenous immunoglobulins were uneffective. Anakinra 50 mg (3.5 mg/kg/dose) s.c. daily, later increased to 75 mg (5.4 mg/kg/dose) per day over a week's period, led to prompt response with resolution of fever and clinical improvement.
Conclusion: Anakinra was an effective treatment in this severe course of MAS, indicating that IL-1 is a major mediator in the underlying inflammatory disease. Further studies have to be established to confirm this first report.

P108

Uveitis, CNS involvement, sensorineural hearing loss and cutaneous findings: the Vogt-Koyanagi-Harada syndrome

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A 13 years old African black patient was hospitalized with asthenia, abdominal pain, weight loss, headache, photophobia, hearing loss and subfebrile temperature. Clinical examination revealed a nucal rigidity and a bilateral anterior uveitis. The audiogram revealed a bilateral high frequency deficit. CBC count was normal, CRP 198 mg/l, SR 82 mm/h. CSF showed a pleiocytosis (487/µl) with hyperproteinorachia (980 mg/l). Blood and CSF cultures were sterile. Search for viral infection and for auto-antibodies was negative. Cerebral MRI showed an encephalitis with inflammatory lesions in the white matter and grey nuclei. Vogt-Koyanagi-Harada (VKH) syndrom was diagnosed. High dose systemic and topic ocular cortisone therapy was started with a rapid decrease of symptoms and of the inflammatory syndrome and disappearance of the cerebral lesions within two weeks. Methotrexate treatment was then added. VKH syndrom is an idiopathic multisystem disorder, which typically affects Hispanic, Japanese and pigmented individuals. After a prodromal phase of neurological (meningitis, encephalopathy) and auditory manifestations (tinnitus, vertigo, hearing loss), an acute uveitic phase follows, characterized by bilateral anterior granulomatous and/or multifocal posterior uveitis with exsudative retinal detachments. A convalescent phase follows several weeks later, characterized by localised alopecia, vitiligo and poliosis. A chronic-recurrent phase can occur with exacerbations of the anterior uveitis. The prognosis depends mainly on the ocular features, and the early recognition and aggressive treatment of the disease. In the case of posterior ocular segment involvement, of late diagnosis or incorrect initial therapy, prognosis is guarded with only 50% of patients having a final visual acuity better than 5/10. Although rare in the paediatric age, VKH syndrome must be considered when facing an association of symptoms affecting eye, hearing, CNS, the skin and phanera so as to initiate the local and systemic cortisone therapy quickly, as the timing of the treatment is essential for ophthalmologic prognosis.

P109

A 2-month- old girl with a rash: neonatal lupus syndrome

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A 7 weeks-old girl was seen for a rash. She was born at term from a 27-year old mother with a history of chronic urticaria after an uncomplicated pregnancy. No skin lesions were noted at birth. Growth and development were normal. No family history of other skin conditions or inflammatory disease. Skin: several macules with central atrophy and raised margins, 5 to 15 mm, on the scalp, the face, the chest and abdomen as well as the lower sacrum, buttocks and upper legs. Conjunctivae and oropharynx were clear. No lymphadenopathy or hepatosplenomegaly. The rash grew slowly over 1–2 weeks. ECG: no atrioventricular heart block. Echocardiography: small patent foramen ovale. Skin biopsy: compatible with a lupus.
Laboratory (child): mild anemia, neutropenia and thrombocytosis. ESR: 17 mm/h, CRP <1 mg/l, ALAT 66 U/l, ASAT 84 U/l, Gamma-GT: 52 U/l, Ca 2.77 mmol/l; HIV, Parvovirus B19 and CMV infections were excluded. Anti-nuclear antibodies >1/1280, anti-SSA antibodies 134 kU/l. Anti-SSB, anti-Sm and anti-phospholipid antibodies, C3, C4 complement and RF: normal. No proteinuria.
Laboratory (mother): anti-nuclear, anti-SSA + SSB, anti-U1RNP antibodies positive.
Differential diagnosis: based on skin lesions is large: urticaria, erythema marginatum, tinea corporis, seborrheic dermatitis, ichthyosiform genodermatosis, erythema annulare centrifugum, familial annular erythema, erythema multiforme, infantile epidermodysplastic erythema, fungal infections, congenital Lyme disease, annular erythema of infancy, erythema gyratum atrophicans, congenital rubella, CMV infection and congenital syphilis.
Conclusion: This case report emphasizes the need to perform immunological studies to any infant up to 8 months of age with annular or polycyclic rash and/or any degree of heart block as well as to its mother, in order to exclude a neonatal (and maternal) lupus.

The numbers refer to the pages of this supplement.