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Ped-O 1

Natural history of recurrent cough in children

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Aims: Recurrent cough (RC) is common in childhood and an important cause of primary care visits. Despite that, data on the natural history of recurrent cough in unselected children are scarce.
Methods: In a population-based cohort in Leicestershire (UK) we measured recurrent cough (defined as chronic night cough + cough apart from colds + GP visits for cough) with repeated questionnaires. For non-wheezers, we computed frequency of recurrent cough at different ages, and determined predictors of RC in multivariable logistic regressions.
Results: We had data on 1247 children aged 1 year, and data on 1127, 1267, 1410, 825 aged 2, 4, 6 and 9 years respectively. Prevalence of RC at these ages was 17%, 19%, 21%, 16% and 12% respectively. Of all children with RC at age 1 year, 62% continued to report RC at age 2 years, and 46%, 35%, 28% at age 4, 6 and 9 years. Factors associated with RC at all ages were: south Asian ethnicity, chronic rhinitis and frequent snoring. Other factors associated with RC during the first 2 years of life were nursery care and posseting. In contrast, attendance to nursery care in infancy protected from RC at age 6 and 9 years. Associations with family history of atopy were marginal.
Conclusions: Recurrent cough is common and tracks strongly during childhood. At all ages, there are strong associations with upper respiratory symptoms. This might be explained by an increased susceptibility to upper respiratory infections.

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Ped-O 2

Changes in Health Related Quality of Life (HRQL) after Spinal Fusion and Scoliosis Correction in Patients with Cerebral Palsy

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Background: There is little literature on the impact of spinal fusion for scoliosis in patients with cerebral palsy (CP) regarding the health related quality of life (HRQL). The purpose of this study was to evaluate the outcome of spinal fusion and scoliosis correction measured by the subjective change in the HRQL as well as the objective radiological parameters. Various factors that could influence the subjective outcome were examined to investigate their correlation to the results of HRQL.
Methods: A retrospective review of 50 consecutive patients with CP, who had spinal fusion for scoliosis with minimal 2 years of follow-up. Radiographic data were obtained from preoperative, postoperative and last follow-up examinations. The assessment of the HRQL was done through a modified version of the "Caregiver Priorities and Child Health Index of Life with Disabilities" (CPCHILD) questionnaire, assessed by the caregivers of the patients.
Results: A significant improvement ($p = 0.001$) of HRQL was found between the preoperative and the postoperative status. The satisfaction rate of the patients (respectively their caregivers) with the outcome of the operation was 91.6%. There was average scoliosis correction of 64.3% and pelvic tilt correction of 57.7%. At the last follow-up the average scoliosis angle was 32.0°, pelvic tilt was 8.8°. No significant correlation between the amount of scoliosis angle correction and the subjective change in the HRQL could be established ($R^2 = 0.321$, $p = 0.078$). No correlation between the occurrence of complications and changes in the HRQL ($p = 0.122$) or the satisfaction rate with the outcome of the operation ($p = 0.764$) was found. Extension of spinal fusion to sacropelvis has no influence on the occurrence of complications ($p = 0.42$) or on the changes in HRQL ($p = 0.71$)
Conclusion: Life quality improves after spinal fusion and scoliosis correction in patients with CP. There is high satisfaction rate of the patients and their caregivers.

Ped-O 3

Predicting bacteremia in children with cancer and fever in chemotherapy induced neutropenia. Results of the prospective multicenter SPOG 2003 FN Study

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Study Aim: To develop a score predicting the risk of bacteremia in pediatric patients with cancer and fever in neutropenia (FN), and to evaluate its performance.
Methods: Pediatric patients with cancer presenting with FN induced by non-myeloablative chemotherapy were observed in a prospective multicenter study. A score predicting the risk of bacteremia was developed from a multivariate mixed logistic regression model. Its cross-validated predictive performance was compared to that of published risk prediction rules.
Results: Bacteremia was reported in 67 (16%) of 423 FN episodes. In 34 (8%) episodes bacteremia was not known at reassessment after 8–24 hours of hospitalization. Predicting bacteremia at reassessment was better than prediction at presentation with FN. The reassessment score predicting future bacteremia in 390 episodes without known bacteremia used: hemoglobin ≥90 g/L at presentation, platelet count <50 G/L, shaking chills ever observed, or other need for hospitalization according to the treating physician. Presence of anyone item predicted bacteremia with 100% sensitivity, with 54 (13%) episodes classified as low-risk, and a specificity of 15%.
Conclusions: This reassessment score, simplified into a low-risk checklist, identifies pediatric patients with FN at risk for bacteremia not yet known. It has the potential to contribute to reduction of antimicrobials, and to shortening of hospitalizations in pediatric patients with cancer and FN.

Ped-O 4

Normative Oscillometric Blood Pressure Values for Pre-School Children

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Objectives and study: Commercially available oscillometric blood pressure (BP) devices are increasingly used in children. Due to technical differences and device specific algorithms validated for measurements in adults, oscillometric BP readings in children often deviate from auscultatory measured BP values. Thus, normative auscultatory BP standards cannot simply be conferred and device specific normative data sets for children may be required.
Methods: Standardized BP measurements were performed in German pre-school children aged 5 to 7 years by 2 different oscillometric devices (Boso medicus prestige™ and Omron M5 professional™). In a subgroup also auscultatory BP measurements were performed. The median of 3 consecutive measurements was used for analysis and calculation of BP percentile curves.
Results: BP measurements were performed in 7417 children with a mean age of 5.8 ± 0.5 years. BP was correlated to height, weight and BMI (all $p < 0.0001$). The 5th, 50th, 90th and 95th systolic blood pressure percentiles were 89, 102, 113, 117 mm Hg (for both oscillometric devices). The 5th, 50th, 90th and 95th diastolic oscillometric BP percentiles were 54, 65, 74, and 77 mm Hg for the Boso and 49, 60, 69, and 73 mm Hg for the Omron device, respectively. Oscillometric measurements were 2.1 mm Hg higher or systolic (both devices, $p < 0.0001$), and 1.6 mm Hg higher (Boso, $p < 0.0001$) or 1.9 mm Hg lower (Omron, $p < 0.0001$) for diastolic BP when compared to auscultatory BP.
Conclusion: Oscillometric devices facilitate BP measurements in children, however oscillometric BP assessment requires device specific reference values or correction factors.

Ped-O 5

Impact of food allergy on the quality of life in Swiss children

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Background: Food allergy in children significantly affects the quality of life. Its impact can be analyzed by the validated French versions of the Food Allergy Quality of Life Questionnaires, parent's form and children's form (FAQLQ-PF and FAQLQ-CF).
Objectives: The aim of our study was to evaluate the quality of life in a sample of Swiss children with IgE-mediated food allergy, confirmed

by a standardized food challenge, or a convincing history of severe systemic reaction and positive allergy tests.
Methods: Information were collected with the FAQLQ-PF questionnaires (parents of children 0–12 years old) and the FAQLQ-CF questionnaire (children of 8-12 years old) during a scheduled allergy clinic visit, and analysed in term of emotional impact, food anxiety and social and food limitations. Patients were divided according to the FAQLQ-PF questionnaire in three age groups: group 1 from 0 to 3 years, group 2 from 4 to 6 years and group 3 from 7 up to 12 years.
Results: 62 food allergic patients were included, with a girl/boy ratio of 1:1.38. Median age was 7.8 years. 58% were known to be allergic to one food, 18% to two foods, 24% to 3 or more foods. Peanuts (71% of children) as well as hazelnuts (29%), walnuts (23%) and eggs (22%) were the major allergens. 61% of children suffered from a mild systemic reaction due to food allergy, 39% from a severe systemic reaction. Young children (group 1) showed significantly better quality of life than older children (group 2 and 3), (Global score (GS) = 0.66 versus 2.13; p = 0.02). Children suffering from severe systemic reactions showed worse scores than the ones with mild reactions (GS = 2.32 versus 1.47; p = 0.039). Children with an allergic mother showed worse scores (GS: 2.22 versus GS = 1.29; p = 0.002), as the ones with allergic siblings (emotional impact score = 2.12 versus 1.38 p = 0.03). Multiple regression calculations could show that these variables were independent. The type of food allergens has no significant impact on quality of life.
Conclusion: Food allergy affects the quality of life of Swiss children. Older children, children with severe systemic reactions and children with mothers or siblings also affected by allergies show worse quality of life scores.

Ped-O 6

Neonatal pulse oximetry screening for congenital heart defects in Switzerland: Analysis of positive test results

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Background: Timely recognition of critical congenital heart defects in neonates is eminent to decrease associated morbidity and mortality. Clinical symptoms are often missing within the first hours of life and 30% of the patients with critical congenital heart defects leave the maternity unit without being diagnosed. Early detection can be improved by the measurement of the transcutaneous oxygen saturation within the first hours of life. Recent studies of the pulse oximetry (POX) screening report a specificity of 99.8% and a sensitivity of 39–83% to detect congenital heart defects in neonates with a postductal transcutaneous saturation lower than 95% within the first hours of life. POX screening is recommended in Switzerland since 2005 by the Swiss Associations of Neonatology and Paediatric Cardiology. The aim of this study was to describe patient characteristics in all screening-positive individuals.
Methods and results: From January 2009 to January 2011 all Swiss Paediatric Cardiologists were asked to fill out a standardized questionnaire for each patient they have seen for an assessment after a screening – positive test result. A total number of 48 questionnaires were returned. There were 27/48 (56%) neonates with a structural heart defect (true positive POX screening test results) including 16 neonates with potentially critical heart defects: Hypoplastic left heart syndrome (n = 1), d – transposition of the great arteries (n = 7), total anomalous pulmonary venous drainage (n = 4) and coarctation of the aorta (n = 4). In 21/48 (44%) of the patients, intracardiac anatomy was normal (false positive POX screening test results). The low saturation was explained in 10/21 patients with normal intracardiac anatomy by evidence of pulmonary hypertension on echocardiography and a right to left shunt through patency of the foramen ovale or arterial duct. Due to the study design with voluntary return of the questionnaire, this analysis is limited by an underreporting and the true number of patients detected with POX screening in Switzerland may even be higher.
Conclusion: The current practice of POX screening in Switzerland yielded a timely diagnosis of potentially critical heart defects in 21 patients within the last 2 years. This is further evidence for the benefit of the POX screening and encourages us to continue with POX screening in all Swiss birth clinics.

Ped-O 7

Early diagnosis of Autism Spectrum Disorders with the use of the M-CHAT: Results from canton Ticino

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Introduction: Although early diagnosis and treatment of children with Autism Spectrum Disorders (ASD) are considered crucial for improving the level of functioning and quality of life, ASD is not always accurately diagnosed during the preschool years. In fact, previous studies show that ASD is often not recognized until entry into public school, especially for higher functioning individuals. For these reasons, we introduced the *Modified Autism Checklist for Toddlers* (M-CHAT) during the annual check-up for 2-year-olds. The aim of this study was to determine the impact of this procedure in the precocity of ASD diagnosis in the southern part of Switzerland.
Methods: We introduced the screening instrument M-CHAT at the annual check-up for 2-year-olds. The M-CHAT is an autism questionnaire for parents consisting of 23 questions about early developmental issues with particular regard to social interaction. Children who were suspected of having ASD were referred to the Paediatric Neurology Unit in Bellinzona, for specific assessment.
Results: Actually 70% of the paediatricians in Ticino used the M-CHAT questionnaire during the annual check-up for 2-year-olds. In 2009, 14 children were further evaluated: 9 showed autistic spectrum disorders (8 M, 1 F); the mean age of the diagnosis was 3.8 years. In 2010, 11 children were referred for specific assessment. Ten children had infantile autism (7 M and 3 F). The mean age for ADS diagnosis was 2.9 years.
Conclusion: After the introduction of the M-CHAT during the annual check-up for 2-year-olds, we observed an increased number of children that underwent assessment for suspicion of ASD in our department comparing the previous years. In fact, we are not far from the incidence estimated at the world-wide level (0.6%). Importantly in 2010, we were able to diagnose ASD at a mean age lower than 3 years. This approach shows the importance of paediatricians in the early detection of ASD. Paediatricians should already begin to recognize children with ASD at the preschool age in order to begin therapy as soon as possible. Early diagnosis and intervention may have a decisive impact on a child's development and subsequently on integration into kindergarten and public schools.

Ped-O 8

Abdominal ultrasound in the first three days of febrile urinary tract infection has no direct impact on its treatment

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Background: Current practice of management of febrile urinary tract infection (UTI) in children includes imaging at acute infection and after 4–6 weeks to detect children at risk for complications or recurrence. We asked whether abdominal ultrasound in the first 3 days after diagnosis of febrile UTI impacts on clinical management.
Methods: Children presenting at the emergency department with suspected diagnosis of febrile UTI (clinical signs and leukocyturia) were prospectively followed. Diagnosis was confirmed by urine culture. Antimicrobial treatment was installed along the Swiss recommendations. Abdominal ultrasound was performed within 3 days after diagnosis of UTI to rule out complications (e.g. abscess, pyonephrosis) that might immediately influence treatment, and subsequent uroradiologic evaluation including abdominal ultrasound and voiding cystourethrogram (VCUG) was performed 6–8 weeks later.
Results: 116 children with febrile UTI and a median age of 7.4 months (mean, 15.4; range, 0.2–88 months) were included. 89/99 children (58% girls) with first febrile UTI and 16/17 children (71% girls) with ≥1 previous febrile UTI were subjected to uroradiologic evaluation 4–6 weeks after the initial ultrasound. None of the initial ultrasounds showed obstruction or complication of UTI with immediate implications for treatment. Thus, the initial ultrasound did not impact on any change of management of febrile UTI in 99 children with first or 17 with at least one previous febrile UTI. Initial ultrasound from children with first febrile UTI recognized 8/9 and 1/3 anatomic anomalies of the kidney and the ureter, respectively, eventually diagnosed in the subsequent uroradiologic evaluation.
Conclusion: The immediate impact on clinical management of abdominal ultrasound performed in the first 3 days after diagnosis of febrile UTI was lower than 1 in 116 children. This justifies revisiting the imaging work-up in children with febrile UTI.

Ped-O 9

Iron-deficiency among children 6–12 months: is an immigrant more at risk?

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Introduction: Iron-deficiency with or without anaemia is the most common deficiency in the world. Its prevalence is increased in developing countries and in low socio-economic populations. The aims of the study were to determine prevalence of iron-deficiency among Swiss and immigrant infants and identified risks factors.

Method: We conducted a prospective study and included every child notified for routine check-up at age 6-months. We followed them at 12 months old. Children were divided into two groups: immigrants' and non-immigrants' children, according to length of stay of parents in Switzerland. We measured haemoglobin, ferritin, anthropometric data, familial and nutritional status at 6 and 12 months.
Results: 509 infants were included, divided into two groups: immigrants' children (N = 297) and non-immigrants' children (N = 212). Prevalence of iron-deficiency was 8 cases (3.1%) in the non-immigrant's children and 17 (9.3%) in immigrant's children. A significant difference was noticed between groups for iron-deficiency at 12 months (p = 0.005). Logistic regression identified immigration in the last 5 years, low income or unemployment, breastfeeding after 6 months, early cow's milk introduction (before 9 months) and baby cereals after 9 months as risks factors for iron-deficiency.
Conclusion: We conclude that prevalence of iron-deficiency in immigrant population is higher than in non-immigrant. Immigration and category of employment are risk factors for iron-deficiency, as starting baby cereals before 9 months is a protective factor. Prevalence obtained was far under those mentioned in literature in similar populations. Good socio-economic conditions in Switzerland, quality of food for pregnant women and young infants may be the explanation.

Ped-O 10

Usefulness of chest x-rays in guiding treatment of children with lower respiratory tract infections

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Background: The usefulness of chest x-rays (CXR) to guide treatment of lower respiratory tract infections (LRTI) is controversial. We investigated the diagnostic value of CXR in children with LRTI compared to clinical assessment. This as a subanalysis of the ProPAED study investigating whether PCT guided AB treatment of children with LRTI reduces AB exposure.
Methods: The ProPAED study investigated previously healthy children, 1 mo to 18 yrs, presenting with LRTI to emergency units of two pediatric tertiary care hospitals in Switzerland between 01/2009 and 02/2010, randomized to AB treatment according to either internationally recognized guidelines or PCT guided treatment to assess the usefulness of PCT guided AB treatment. In this subanalysis, we compared CXR diagnosis with clinical diagnosis of pneumonia, PCT values, and AB use.
Results: 293 of 337 children with LRTI underwent CXR, mean age 3.9 yrs (range 0.1–17.5), 61.2% were hospitalized. 203 patients were clinically assessed as CAP, 90 as NonCAP. 67% (n = 136) of clinical CAP were confirmed by CXR, 34% (n = 31) of clinical Non-CAP had a positive CXR nevertheless. In the PCT group (N = 135) 56% (N = 76) had CXR positive, in 24 of them (31%) PCT was <0.25 and were thus not treated with AB despite positive CXR. In the GL group (N = 146) this compares to 79 (54%) patients with positive CXR, in which PCT was <0.25 in 29 (37%) patients, 9% (N = 7) of them were not treated with AB. Of patients with CXR positive CAP AB treatment was given to 68% (N = 52) and 85% (N = 67) of patients in the PCT and GL group, respectively.
Conclusions: CXR guided AB treatment lead to overtreatment of children with LRTI in our study population. PCT should be considered as an alternative biomarker guiding prudent antibiotic treatment in children.

Ped-O 11

Usefulness of biomarkers to restrict antibiotic prescribing for lower respiratory tract infections in children

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Background and aims: The identification of lower respiratory tract infections (LRTI) in need of antibiotic treatment (AB) remains challenging. This study investigated procalcitonin (PCT) and c-reactive protein (CRP) as predictive biomarkers of bacterial infection not in need of antibiotic treatment.
Methods: Data were obtained from a randomized controlled trial (ProPAED) comparing pediatric febrile LRTI treated with AB either according to international guidelines or to PCT cut offs accepted for AB of LRTI in adults. Of 337 children enrolled in the original trial, all previously healthy children with complete diagnostic and outcome data were analyzed. Test performance parameters of PCT and CRP in predicting outcome with restricted AB were calculated.
Results: 125 patients were eligible for analysis of PCT and CRP dependent treatment outcome. The PCT and CRP values were higher in patients with community acquired pneumonia as compared to those with other LRTI (p <0.05). PCT (cut-off 0.25 mg/dl) combined with CRP (cut-off 40 mg/dl) showed a sensitivity of 81% (CI 95 67-96) and a negative predictive value of 88% (CI 95 77.3–97.7)) to rule out non-favorable outcome of children with LRTI without AB.
Conclusion: The combination of low PCT and CRP values may predict favorable outcome of restricted AB for LRTI in children. Larger prospective intervention studies with higher PCT cut offs are needed to further crystallize outcome driven paediatric cut offs for biomarker guided AB of LRTI in children.

Ped-O 12

Group therapy of overweight children in Switzerland – medical results after one year*

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The present interim analysis of a Swiss national multi-centre study funded by the FOPH examines the implementation of multiprofessionel group programmes to treat overweight children and their effects on medical parameter before (T0) and after 1 year (T2).
Methods: In an on-going clinical cohort study, data between 03/2009 and 12/2010 from 22 active out of 27 certified Swiss programmes were analysed. Programmes including children and parents were standardised according to paediatric guidelines to guarantee the reimbursement by health insurances. Co-morbidity, family origin, body mass index standard deviation score (BMI-SDS), waist circumference, blood pressure (BP) and metabolic markers were assessed at T0 and T2 (means ± SD).
Results: 1292 overweight children were registered for therapy. After motivational analysis, 23% of them did not pursue admission procedure. After psychological and medical evaluation, 529 children started group therapy (41%). Group size was 11.6 ± 3.5 children and scantily cost-effective. At baseline, 342 children were assessed so far: age 12.2 ± 2.2 years, 53% female, 45% migrants, BMI-SDS 2.7 ± 0.6 and waist/height ratio 0.59 ± 0.7. Overweight appeared at 6.1 ± 3.7 years; physical, nutritional or psychological therapy had been individually started before T0 in 3, 11 or 15% of cases, resp. Children were extremely obese in 59%, obese (BMI >P97& <P99.5) in 34% and overweight in 7% of cases. The prevalence of co-morbidities was high: 75% of orthopaedic disorders, 21% of asthma, 29% of attention deficit, 16% of hypertension and 7% of sleep apnoea. Moreover, 68% of mothers and 77% of fathers were overweight or obese. At T2, weight gain in fathers was significant. Longitudinal data of 113 children were analysed (dropout rate 8.1%) showing significant reductions in BMI-SDS (–0.19 SD ± 0.36, p <.001), waist/ height ratio, systolic BP and fasting glucose. Follow-up of elevated levels of glucose or ALAT (at T0: 14 and 26% increased, resp.) was done by the physician in less than 50% at T2.
Conclusion: Standardised multiprofessionel group therapy significantly improves adiposity and health of obese children, though demands on health care professionals and families are high. The local availability of programmes remains insufficient and the group setting, time and effort required exceed by far the resources of most families with obese children. Nevertheless, the individual medical care for obese children must be improved by diagnosing and treating co-morbidity early and adequately.

Ped-O 13

High serum leptin and soluble cellular adhesion molecules levels are associated with impaired endothelial function in pre-pubertal obese children

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Background: Soluble inflammatory mediators, such as leptin and cellular adhesion molecules (CAMs), have been shown to act as potential biomarkers in cardiovascular diseases. The aim of this study was to assess their levels and determine their associations with arterial vascular function in pre-pubertal obese children.
Methods: Forty-four obese (age 8.3 ± 1.5 yrs) and 23 non-obese pre-pubertal children (age 9.0 ± 1.6 yrs) were studied. The flow-mediated dilation (FMD) of the brachial artery was assessed by high resolution ultrasound. Circulating levels of adipocytokines (leptin, adiponectin, resistin), soluble adhesion molecules (intercellular adhesion molecule-1 [ICAM-1]), vascular cell adhesion molecule-1

[VCAM-1], sE selectin), and other inflammatory biomarkers (sP selectin, high-sensitivity C-reactive protein [hs-CRP], IL-1α and TNF-α) were measured in both groups.
Results: Obese subjects had significantly increased levels of leptin (mean 41.3 ng/ml, CI95% 31.9–50.8 vs 14.6, 9.4–19.8, p = .0002), resistin (2.6, 2.2-3.1 vs 1.7, 1.4–2.0, p = .003), VCAM-1 (415.0, 325.1–995.6 vs 371.3, 291.2-470.2, p = .03), sE-selectin (37.3, 30.4-51.0 vs 20.2,16.1–32.4, p = .0008), sP-selectin and hs-CRP compared to non-obese subjects. Obese children with low FMD (<5.5%) had significantly higher leptin and VCAM1 levels than subjects with normal FMD. The BMI-SDS (t = 3.4, p = .003) and leptin (t = 2.6, p = .017) were independent predictors of FMD.
Conclusions: Early endothelial cells activation and pro-inflammatory state appear before puberty in obese children, in association with high leptin level. Leptin is an independent determinant of endothelial dysfunction of the peripheral circulation.

Ped-O 14

Effects of group therapy on physical fitness and functional capacities of overweight children and adolescents in Switzerland: the KIDSSTEP study*

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Introduction: Obese children spend usually less time in physical activities and have lower cardiorespiratory fitness that their non-obese counterparts. The purpose of this study was to assess changes during family-based group therapy in physical fitness and functional capacities in overweight children and adolescents.
Methods: We present data of 77 girls and 68 boys (mean age 12.2 ± 2.2 years) of the ongoing national multi-centric study of multiprofessional group therapy for childhood obesity, collected between 03/2009 and 12/2010 in 22 certified programs in Switzerland. The therapy includes adapted physical activity sessions 1-2 times per week or once every 2 weeks. The following outcome measures (Eurofit test battery) were performed at baseline and after the intensive phase of therapy (at 6–9 months): Single Leg Balance Test, Plate Tapping Test (coordination), Long Jump Test (legs explosive strength), Sit Up Test (abdominal strength and endurance), Shuttle Run (10 x 5 meter, speed), and Leger Fitness Test (20m-shuttle run, aerobic fitness).
Results: At baseline, obese children had significantly reduced balance, coordination, abdominal and legs muscle strength, speed and Leger test running distance, compared to Swiss normative references. From baseline to the end of the intensive phase of therapy, changes in BMI-SDS were -0.15 ± 0.34 (p <0.001) and –0.29 ± 0.39 (p <.0001) in girls and boys, respectively. Significant improvements in single leg Balance (girls 10.0 ± 99.8 and boys 29.1 ± 47.3%), plate tapping time (10.8 ± 13.7 and 10.8 ± 14.9%, resp.), Sit ups (35.9 ± 70.8 and 15.4 ± 51.1%, resp.), and Leger test distance (19.2 ± 62.1 and 23.1 ± 44.8%, resp.) were observed. Long jump and shuttle run results did not change, probably due to biomechanical limitations.
Conclusion: Physical fitness and functional capacities are reduced in overweight children and adolescents in Switzerland. Regular exercise training during 6 to 9 months, combined with family-based group therapy, significantly improves their balance, coordination, abdominal muscle strength and aerobic fitness. These changes are associated with reductions of the degree of adiposity.

Ped-O 15

Juvenile Dermatomyositis in Western Switzerland

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Introduction: Juvenile dermatomyositis (JDM) is a rare chronic, autoimmune disease affecting mainly the muscles and skin and which lead to impairment of physical function and endurance. The incidence described in USA is 2.5–4.1 cases per million children. Recent studies describe that two third of the patients followed a chronic disease course, whereas only one third showed monocyclic disease course.
Objectives: Assess disease characteristics and outcome of JDM in Western Switzerland.
Methods: Retrospective pediatric rheumatology chart review in Lausanne and Geneva.
Results: We describe a population of 13 JDM patients followed in western Switzerland from 1997–2010. The incidence of JDM was similar to other studies (3.5 cases per million children). The mean age of onset was 6 years (3–10 years), the mean follow-up 6 years (3 months – 13 years). All patients presented a rash (Gottron's rash, heliotrope rash or extensor surface rash, in one patient only during follow-up), muscle weakness and evidence of myositis (biopsy, elevated muscle enzymes, magnetic resonance imaging or by electromyography). The median delay of diagnosis was 2 months, 3 patients had a longer delay because of misleading symptoms. Follow up of >1 year is available for 12 patients. In comparison with other studies, evolution was predominantly monocyclic (50% of all cases showed remission within 2 years after diagnosis). All patients were treated by corticosteroids and Methotrexate. If skin manifestation predominated, Hydroxychloroquin was added (40% of all cases). In the 6 other patients with chronic disease course, multiple treatment changes were required: Ciclosporine, Azathioprin, immunoglobulines, Infliximab, Adalimumab, Rituximab. Only one patient had pulmonary complications (interstitial pneumonia) and one patient presents functional impairment. Calcinosis was present in 3 patients (23%). All patients showed at one point impregnation of corticosteroid treatment. One patient presented pancreatitis and fungal infection related to treatment. Growth impairment was seen in 3 patients (23%).
Conclusion: In our study we show a better outcome compared to other recent reports. This could be due to early diagnosis and rapid treatment onset or less severe presentation of the disease in our patients. Early referral to a reference center and rapid treatment with corticosteroids combined with Methotrexate may improve the outcome.

PedChir-O 1

Adherence to perioperative antimicrobial prophylaxis in a children's hospital

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Introduction: Perioperative antimicrobial prophylaxis (POAP) is effective in preventing postoperative infections. Institutional guidelines for POAP are a tool to rationalize antimicrobial use in the perioperative setting.
Methods: Application of POAP was investigated among consecutive children undergoing any invasive intervention under anaesthesia at a tertiary paediatric surgical centre during six weeks in 2010. Perioperative documentation was checked for indication, choice, dose and duration of POAP and compared to a local guideline.
Results: 710 children with a median age of 4.6 years (mean 6.0, range 0.01–23 years, 18% <1 month) fulfilled the inclusion criteria. POAP was administered to 225 of 710 (32%) children with the highest rates in cardiothoracic (36/38) procedures, followed by craniofacial/neurosurgical (26/29), trauma/orthopaedic (73/141) and urologic operations (27/68). Overall, the indication for POAP was appropriate in 215 of 225 prescriptions (96%), however, correctly applied regarding antimicrobial choice, dose and duration in only 41% (89/215). The choice of antimicrobial was as advised in 76% (171/225) with the recommended dose administered in 130 of those 171 patients (76%). POAP was correctly given as a single shot in 30% (51/171) and during 24 hours in 52% (89/171). Recommended duration of POAP was exceeded in >50% of laparotomies and neurosurgical procedures and in >25% in urological, trauma/orthopaedic and neurosurgical operations. Dosing errors were seen most frequently with amoxi-clav (>50% of the prescriptions).
Conclusion: Indication of POAP usually accorded with the local guideline. However, several targets for optimization of POAP were identified, in particular duration of POAP, choice of antimicrobial and its dosing. Adherence varied by the type of procedure, potentially allowing for targeted interventions. In order to further optimize POAP a multidisciplinary discussion and review of the local guideline commonly involving surgeons, infectious diseases specialists and pharmacists may be successful.

PedChir-O 2

When three make one – a triple magnet ingestion case report

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Introduction: Young children are known to swallow small objects, usually without danger for their health. On the other hand, magnet ingestion in children, even if a rare entity, may cause dangerous gastrointestinal complications. Multiple magnets, if ingested, may attract each other maintaining the intestinal tissue between them, and lead to bowel obstruction or perforation.
Case report: A 2-years-old boy presented a partial bowel obstruction due to consecutive ingestion of 3 magnet pieces on 2 following days. Symptomatology was acute including continuous crying, vomiting, abdominal pain and surgical abdomen on physical examination. After one night observation, the control X-rays did not show any movement of magnetic pieces and a surgical intervention was performed. Hopefully, no bowel necrosis or perforation was found in our patient, no post-operative complication occurs and the child could be discharged after 5 days.
Discussion: Between 2003 and 2006 twenty cases of intestinal perforations and one death due to ingestion of several magnet pieces were reported [1]. A recent literature review [2] describes more than 100 cases of magnet ingestion. Our case is particular because of atypical magnet location: one piece in the stomach and two another in transverse colon, attracting each other and causing bowel obstruction. In that situation, double intervention was necessary: laparotomy as well as intraluminal introduction of metallic mandrill to proceed to dispatching and removal of gastric magnet.
Conclusion: It is important to make aware parents about the risk of magnet ingestion and remind to physicians that these cases could be easily misdiagnosed without careful history taking. Early intervention is important to avoid complications.

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PedChir-O 3

Neuroendocrine tumors in children

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Introduction: Neuroendocrine tumors are extremely rare in children. We report on our series of three different neuroendocrine tumors.
Patients: A 13 year old girl had an appendectomy for appendicits. The histological section showed inflammatory changes of the appendix and additionally a neuroendocrine tumor which had already invaded the serosa. Therefore as a curative approach a right hemicolectomy was performed. A 13 year old boy was admitted with hematemesis. The gastroduodenoscopy showed a bleeding duodenal ulcer. Gastrin levels were markedly elevated. Therefore a CTscan was performed and a gastrin producing neuroendocrine tumor in the pancreas was found. As a curative approach a partial pancreatectomy was performed. The histological section showed a R1 resection and peripancreatic lymph node involvement. Therefore a re-resection of a part of the pancreas with radical pancreatic lymph node resection and consecutive splenectomy was performed. A 12 year old boy was admitted with headache due to high blood pressure. The diagnostic workup identified a tumor in the right adrenal gland. The diagnosis of a phaechromocytoma was suspected by MRI and highly elevated catecholamine levels in blood and urine. A tumor adrenalectomy confirmed this diagnosis.**Conclusion:** Neuroendocrine tumors are exclusively rare and require a high index of suspicion. However tumors are often diagnosed by chance. An evaluation of predisposing factors is mandatory especially for multiple endocrine neoplasia (MEN). Interdisciplinary tumor evaluation and therapy planning is necessary for an appropriate approach. But surgery often remains the only curative approach.

PedChir-O 4

An extremely rare congenital malignant cystic lung tumor in a newborn

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Introduction: Pleuropulmonary blastoma type I is an extremely uncommon malignant neoplasm in the newborn. We report on a newborn girl with this exclusively rare condition.
Case: A babygirl born at 41 4/7 weeks gestational age presented two days after birth with tachypnoea, fever, and rising inflammatory parameters. The chest radiography showed a hyperinflated right middle lobe of the lung. A bronchoscopy was performed in which stenosis of the middle lobar bronchus was diagnosed and a congenital emphysema was suspected. As respiratory deterioration occurred, an emergency thoracotomy was performed. Intraoperatively, a cystic tumor was enucleated and a partial resection of the upper lobe was performed. The macroscopic aspect was highly suggestive for CCAM. Histopathologic workup finally revealed the unexpected diagnosis of a type I pleuropulmonary blastoma. Complete surgical removal of the upper and middle lobe had to be carried out to achieve a R0 resection. The postoperative recovery was complicated by recurrent pneumothoraces. After leaving ICU and neonatal care a neoadjuvant chemotherapy was added.
Conclusion: Although pleuropulmonary blastomas type I are extremely rare tumors in newborns, they must be part of the differential diagnosis of congenital cystic lung lesions. Yet, as of today there is no means to reliably recognize this condition clinically or radiologically. The definitive diagnosis is made by histology.

PedChir-O 5

An extremely rare benign cystic lesion of the pancreas

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Introduction: Acinar cell cystadenoma of the pancreas is an extremely rare entity. As for today there have been 12 cases mentioned in literature. We report on a 13 year old girl with this unusual diagnosis.
Case: A 13 year old girl with severe mental and physical retardation was diagnosed with a 5cm diameter cystic abdominal lesion in 2004. Ultrasound follow-up didn't show any progression. In 2010 a CT scan showed a cystic lesion of 20x13x7 cm. A mesenteric cyst or retroperitoneal lymphangioma was postulated because of "classical" appearance and relatively common occurrence. As the child suffered of increasing abdominal discomfort and constipation, elective resection of the lesion was performed. Intraoperative findings showed a giant cyst located retroperitoneally. Astonishingly, the tumor extended cranially as far as behind the duodenum, allowing direct view on the intact common bile duct and the pancreatic head after removal of the cyst. Postoperatively, the girl presented with borderline hemodynamic parameters and a progressively distended abdomen. Re-laparotomy on the 6th postoperative day showed massive ascites and a diffuse oozing from the resection site. Secretion of chylus from small lymphatic vessels was suspected. Therefore a sclerosing agent (OK432) and fibrin-glue were applied. Post revision there was massive secretion from the easy-flow drainages, which showed highly elevated pancreatic enzymes. The final histological workup revealed the unexpected diagnosis of a benign acinar cell cystadenoma of the pancreas. Only the retrospective review of all imaging data revealed evidence that the lesion might actually originate from the pancreas.
Conclusion: This is a typical example of a missed diagnosis that was both very challenging to establish and extremely rare. A higher index of suspicion by all experts involved might have led to a correct preoperative diagnosis. The differential diagnosis of cystic lesions in direct proximity of the pancreas must include cystic pancreatic lesions irrespective of their extreme rarity.

PedChir-O 6

Open versus laparoscopic appendectomy in children. Should be considerate the laparoscopy the first choice?

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Introduction: Laparoscopic appendectomy is gaining popularity in comparison with the conventional open approach in the pediatric surgical community. In the last 5 years in our institution we have performed mostly laparoscopic appendectomies in children and rarely

an open approach. We perform the laparoscopic appendectomies in children using a technique with 3 trocars of 5-mm. The purpose of this study is to analyze the trends of laparoscopic versus open appendectomy in our institution and also the results with our laparoscopic technique of appendectomy in children.
Methods: The files of all children operated in our institution from 1999 to May 2010 for appendectomy were analyzed. Two groups of patients were created: Group 1, Open Appendectomy (OA) and Group 2, Laparoscopic Appendectomy (LA). Our pediatric surgery service is relatively young since it was created 11 years ago and the surgical procedures are performed by 2 full-trained pediatric surgeons and 1 senior resident.
Results: 230 patients were included in the study (130 boys and 100 girls). Range of age was between 2 and 16 years old. Between 1999 and 2004, 54 open appendectomies and 34 laparoscopic appendectomies were performed. Between 2005 and May 2010, 8 open appendectomies and 134 laparoscopic appendectomies were performed. Twenty-four children of the LA group had complicated appendicitis with perforation, generalized peritonitis or abscess. The rate of post-operative complications in both groups were 9,1% (13 cases in the LA and 8 cases in the OA). The rate of post-operative peritoneal abscess was 2,9% in the LA group and 3,2% in the OA group. The rate of post-operative abdominal wall infections was 1,1% in the LA and 3,2% in the OA. There were 2 patients with intestinal occlusion secondary to post-operative adhesions in the LA group, resolved by laparoscopy with an excellent out-come. Conversion rate in the LA group was 5,3% (7 patients).
Conclusion: Our technique of laparoscopic appendectomy is feasible, sure, gives great exposure of the abdominal cavity and allows to displace the 5-mm telescope and the 5-mm instruments according to the necessity of exposure and work, especially in complicated cases of appendicitis. The rate of complications is low and offers good short and long term results. The esthetics is excellent. Moreover, in institutions with pediatric surgeons with good laparoscopic skills the laparoscopic appendectomy should be considered the gold standard to treat children with acute or complicated appendicitis independently of the age.

PedChir-O 7

SILS-port Nephrectomy

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Introduction: Single port laparoscopy is an exciting technique that is gaining popularity. Several reports start to get published in the general adult laparoscopic community, however in children and adolescents little experience exist. In this abstract we present a case of a SILS-port right nephrectomy.
Methods: The patient is a 16-year-old girl with a non-functional dysplasic right kidney with episodes of lumbar right pain and hematuria. For the nephrectomy a SILS-port was placed in the umbilicus and the surgery was performed using 5-mm straight and bending instruments and a 5-mm 30° telescope.
Results: The operative time was almost 240 minutes and no complications occurred. The patient started liquid diet the night of surgery and was discharged to home in the second postoperative day. No further treatment was needed.
Conclusion: Single port laparoscopy can be used successfully for a nephrectomy in adolescents. Good skills in laparoscopy are mandatory. One of the main advantages seems to be lower post-operative pain and a great improved cosmesis

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PedChir-O 8

Fetal Surgery in Zurich: The First Successful Open In Utero Repair of Myelomeningocele

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Background: The recently published MOMS-trial suggested that in utero repair is a novel standard of care for select fetuses with MMC.
Case Report: We report on the first European fetal MMC repair performed at the Zurich Center for Fetal Diagnosis and Therapy. Evaluation, operative procedures, and postoperative care were performed according to the MOMS-trial. A 37 y-old 4G3P was diagnosed with a fetus demonstrating a lumbosacral MMC. The operation was successfully performed at GA 24+5. The postoperative course was uneventful except for leakage induced oligohydramnios and consecutive breech position. C-section was performed at GA 35. During the first 5 weeks there was no need for a VP-shunt and CIC. A bilateral hip dislocation required casting. There was a decreasing renal insufficiency possibly due to hypodysplastic kidneys.
Conclusion: Basically, we have successfully adopted the novel treatment option resulting from the MOMS-trial. However, a definitive outcome statement can not be made yet.

PedChir-O 9

Skingineering: Engineering and transplantation of large-scale skin analogues in a pig model

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Background: Extended full thickness skin defects still represent a considerable therapeutic challenge. As tissue engineering of whole skin seems to be an attractive and ambitious approach, our aim was to engineer autologous porcine grafts and to develop an appropriate large animal transplantation model for pre-clinical studies.
Materials and Methods: Porcine epidermal keratinocytes and dermal fibroblasts were isolated and expanded on culture dishes. Subsequently, hydrogels containing fibroblasts were prepared and keratinocytes seeded on top. The resulting *in vitro* generated porcine skin substitutes were analysed. For *in vivo* studies, full-thickness skin defects measuring 7.5 x 7.5 cm were created. Dermal regeneration templates and cultured autologous skin analogues were applied to the wound bed shielded by a silicone chamber. The transplants were covered by a sophisticated dressing and results documented photographically and histologically 3 weeks thereafter.
Results: As for the *in vitro* study, dermal fibroblasts grown in collagen/fibrin hydrogels showed a good proliferation and formation of a 3D-network. When these constructs were seeded with keratinocytes, a multilayered and partly stratified epidermis developed. As for the *in vivo* study, the dermal regeneration templates healed perfectly, while the cultured skin analogues showed complete take of the dermal compartment but spots of missing epidermis. The chamber proved effective in preventing spontaneous ingrowth of keratinocytes from the wound edges and the dressing efficiently kept the operation site intact and clean.
Conclusion: Our study shows that pig cell-derived, autologous skin analogues with near normal skin anatomy can be engineered *in vitro*. We also show a valid pig model permitting the transplantation of laboratory-engineered skin analogues in a clinically relevant size. Hence, this animal model will be used to establish standardized transplantation procedures to perform pre-clinical studies.

PedChir-O 10

Routine pin tract care in external fixation is unnecessary: A randomised, prospective, blinded controlled study

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Introduction: Pin site infections are seen in up to 40% of External Fixators (ExFix) and are therefore the most common complication with this device. There is no consensus in the literature as to the appropriate regimen for pin tract care and infection prevention. This study is the first intra-subject, randomised, prospective and blinded controlled trial comparing daily pin tract care to no pin tract care at all.
Method: Consecutive patients series (56 patients, 16 female, age 4–68y, mean 24y, in total 204 pins) recruited in the National Hospital in Honiara (Solomon Islands) during 2 years. Exclusion criteria were application of ExFix for less than two weeks or a non-standard ExFix. Pin treatment was allocated into groups anatomically, proximal and distal. Randomisation was intra-subject and intra-group: 101 pins had daily pin site care and 103 had no treatment at all. Endpoints: soft-tissue interface, stability of the pins, torsional stability as determined with a torque meter, osteolysis and pain. Assessment of pin sites blinded. Statistical analisys using the paired t test for parametric data and the Wilcoxon rank test for non-parametric data (Stat View).
Results: No significant difference between the two groups. Soft-tissue interface 36% vs. 35% (granulation/secretion), stability 20 vs 25 Pins with loosening. No significant osteolysis (7 vs. 6 Pins) Torque: mean 0.75Nm, max.: 3,05Nm vs. 0,60Nm, max.: 3,55Nm, no significant difference. No differences in demographics (age, localisation, sex, time of fixation).
Conclusion: This study shows that routine pin tract care is unnecessary in external fixation treatment of injuries.

PedChir-O 11

Biliary complications after pediatric liver transplantation: experience over a 20-year period

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Introduction: Analysis of biliary complications (BC) following pediatric liver transplantation (LT) over a 20 year period at the Swiss national reference center for pediatric LT in Geneva.
Methods: Retrospective study of all pediatric LT-patients of a single institution, from May 1990 to April 2011, median follow up of 104 months (range 3–240 m).
Results: One hundred and six patients received 113 LT, i.e. 11 living (LD) and 102 deceased donors (DD). BC occurred in 24 patients (21.2%): in 4 LD (36% of LD-LT) and 20 DD (19.6% of DD-LT). Amongst 11 choledocho-choledochal anastomoses (CCA) were 5 (45%) BC: 1 twist, 2 leaks, 2 primary strictures (at 6 and 11m). Amongst 102 entero-biliary anastomoses (EBA) were 19 (18.6%) BC: 2 accidental ligations of secondary bile ducts (both LD), 2 obstructions, 7 leaks (median 4d, range 1–57d), 8 primary strictures (median 5m, range 9d-46m). Accidental ligations, twist and obstructions were all treated with surgical redo of the anastomoses, 1/5 developed a secondary stricture. Of 9 leaks, 4 healed with drainage alone without sequelae, 5/9 had a primary redo (2/5 developed a secondary stricture, 1/5 a releak (death 3m after LT due to MODS). Of 13 strictures (10 primary, 3 second after redo) 1 had no treatment and cleared spontaneously, 3/13 had a primary redo without sequelae, 9/13 had 1-6 rounds (median 1, range 1–6) of percutaneous balloon dilatation (PBD) with finally 3 redos (after 1 or 2 sessions); overall 6/13 (46%) strictures had a redo. Only 1/9 sessions of PBD was performed before 2000. In total 15/24 BC-patients had 16 anastomosis redos (63% of BC-patients, 67% of all BC).
Conclusion: Compared with the literature we report a satisfying incidence of BC after pediatric LT. LD grafts and CCA are associated with more BC than DD grafts and EBA, respectively. Biliary strictures are the most frequent, bile leaks the most frequent early BC. Management for strictures has changed since 2000: PBD is now the treatment of choice. Yet, many redos are seen in this series. This high rate may be due to aggressive, early surgery rather than more expectant management preferred by other centers.

PedChir-O 12

Long-Term Cosmetic Results of Split-Thickness Skin Harvesting from the Scalp: Far from Scar?

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Introduction: The scalp is a well-established donor site for split-thickness skin grafting in pediatric burn patients. However, scientific data of long-term observations of functional and cosmetic aspects of the scalp are missing. Therefore, we studied patients up to 30 years after harvesting split-thickness skin from the scalp in childhood.
Material and Methods: Our follow-up study was performed of patients older than 30 years, which underwent split-thickness skin grafting from the scalp in childhood after thermal injury. Patients were asked to complete a standardized questionnaire and were invited to our outpatient clinic where the scalp was assessed regarding scarring, pigmentary or structural abnormality, and hair growth pattern. All results were photodocumented.
Results: Seventy-one patients met inclusion criteria for this study. Fifty-eight of these could be identified and thirty-two (eighteen men and fourteen women, between 31 and 45 years old) agreed to participate. They were between 1 and 15 years old at the time of split-thickness skin grafting and the scalp was harvested up to 5 times per patient. Clinically, none of the patients developed scars. Hypopigmented scalp areas were found in 9%, *changes* in *skin texture* in 3%, and small alopecic spots (2-20mm in diameter) in 8% of the patients. Subjectively, all patients were free of complaints and described their hair growth as normal and comparable to the hair growth of same-sex relatives.
Conclusion: There was no clinical evidence of significant functional or cosmetic alteration. Subjectively, all patients were satisfied with the result and free of complaints. Therefore, these long-term results are a further important argument for split-thickness skin grafting from the scalp in pediatric surgery.

PedChir-O 13

Appraisal of adult genitalia after hypospadias repair. Do laypersons mind the difference?

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Introduction: Men with corrected hypospadias often report a negative genital appraisal and many of them suffer from sexual inhibition and fear of being ridiculed or rejected by potential partners. Nevertheless, no investigations have been done on the appraisal of genitals after hypospadias repair by laypersons. The aim of this study is to assess the appraisal of adult genitals after hypospadias repair in comparison with circumcised genitals.
Methods: Standardized photos were taken of genitals of 20 men who were operated for hypospadias and 18 men who were circumcised. 175 non-afflicted men and women in different age-groups (age-range: 16–20; 25–30 and 40–45) were asked to rate a selection of these pictures with a structured questionnaire.
Results: Our results showed that healthy men and women rated adult genitalia after hypospadias repair significantly less positively than those of circumcised men (e.g. sum-score of overall genital perception, $p = 0.001$). However, effect size was very weak (Cohens'd of sum-score of overall genital perception = 0.15), so that the difference does not appear to be clinically relevant. Interestingly, no significant differences were found between the appraisal of hypospadias-patients' genitals and those of circumcised men regarding potential rejections as sexual partners.
Conclusion: These findings suggest that the fear of men with corrected hypospadias to be ridiculed or rejected by sexual partners is unfounded. Although the difference of genital appearance between corrected hypospadias and circumcised penis is visible for laypersons, the difference does not appear to be relevant and potential rejection as sexual partners could not be found in this study.

PedChir-O 14

Disorders of sex development: Karyotype and gender assignment

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Introduction: Children with disorders of sex development (DSD) are an enormous challenge as for their management. Multidisciplinary teams that manage these patients have to focus on the predictability of the gender identity as well as gender role from infancy through adulthood, without knowing what the individual really is. The goal of surgical genitoplasty in the management of children affected by DSD is to achieve a phenotype that can offer the patient the best possible quality of life. When choosing the type of surgical genitoplasty, the crucial question is: What should be privileged in choosing a certain gender over the other: Karyotype or sex of rearing?
Case presentation: A 2-year-old 46-XY-DSD girl and a 16-year-old 46-XX-DSD adolescent boy were addressed from abroad to our clinic for genital surgery. A multidisciplinary team analyzed the contemplable surgical options, taking into account 1) the karyotype, 2) the etiology of their DSD, 3) the development of external genitalia, 4) the potential for adult fertility and sexual function, 5) parental support, 6) cultural factors, 7) presumed gender identity and, last but not least, 8) rearing sex of these quite old children. Consequently, we choose to perform a feminizing and masculinizing genitoplasty, respectively, respecting the sex of rearing, independently of their karyotype.
Conclusion: Reinforcing the sex of rearing by the surgical achievement of a concordant phenotype is an important tool in managing DSD children and their families, in an attempt to avoid gender dysphoria when these children reach adulthood. In that way, family environment, and in particular the mother's feeling about the situation, reflected by the sex of rearing in our cases, are of outmost importance to take the ultimate decision. Only long term follow-up will confirm if we did right.

PedChir-O 15

2 years' review of pediatric emergencies admitted in the resuscitation room of a Swiss university hospital

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Introduction: Paediatric resuscitation is an intense, stressful and challenging process performed in a specific surrounding. In the

resuscitation room (RR), a dedicated pediatric team is not always available and its composition varies according to local resources. A regular review of the children admitted in the resuscitation room and the assessment of various outcome measures are the basis of quality control (QC). The epidemiology of Potentially Life Threatening Paediatric (LTP) emergencies admitted in a Swiss university hospital has never been reported. The aims of this study were to review the LTP emergency population with regards to origin, patients' demographics, reason for admission and final diagnosis, treatment modalities, critical events and outcome.
Methods: A retrospective observational cohort study of prospectively collected data was conducted, including all LTP emergencies admitted over a period of 2 years in the RR of a Swiss university hospital functioning as a tertiary level referral centre. Multiple variables including indication for transfer, mode of pre-hospital transportation, diagnosis and the time spent in RR were assessed. Data assessment took place 2 years after the implementation of a quality control (QC) team assessing the pediatric resuscitations occurring within the institution on a monthly basis.
Results: Out of 60 939 pediatric emergencies treated in Lausanne University Medical center over 2 years, a total of 277 LTP emergencies (0.46%) were admitted to the RR, including 160 boys and 117 girls, aged 6 days to 15.95 years (mean 6.69 years, median 5.06). The table below illustrates in more details the identified problems, average age, time in hospital and outcome of both surgical and medical groups of patients.

	Surgical group (n = 121)	Medical group (n = 156)
Age (years)	8.4 (0.6–15.8)	5.3 (0.02–15.9)
NACA score	4 (3–6)	4 (3–6)
GCS score	12 (3–15)	10 (3–15)
Time in RR (min)	52 (5–165)	52 (4–192)
Transfert to PICU	29	85
Transfer to OR	35	2
Death in RR	4	4
Death after 48 hours	2	7
Length of stay in hospital (days)	8 (1–194)	9 (1–145)
Transfusion	3	–

Conclusions: With the need for health care quality improvement and financial restrictions, an excellent knowledge of the characteristics of LTP emergencies is unavoidable. A thorough understanding of the resuscitation process and humans resources involved can be achieved with a systematic review of the cases. A dedicated quality control team evaluating LTP emergencies in a hospital will identify areas for improvement. A LTP registry at the national level would be of great value in Switzerland.

PedChir-O 16

Intranasal fentanyl reduces acute pain in children in the emergency department: a safety and efficacy study

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Background: Provision of rapid, painless and effective analgesia to children remains problematic in the prehospital and emergency setting. Intranasal fentanyl has the potential to eliminate many of the problems of narcotic administration in children. It is standard of care in many countries but not in Switzerland. The aim of this study was to evaluate the safety and efficacy of intranasal fentanyl in children before implementing its use in our tertiary paediatric emergency department.
Methods: Children in acute pain aged between one and 16 years were enrolled on presentation to the emergency department. Routine observations and pain scoring was undertaken prior to the child receiving 1.5 µg/kg intranasal fentanyl and after 15 min. We assessed the safety and efficacy of intranasal fentanyl and evaluated the satisfaction of the patients, their families and the providers.
Results: Thirty children were enrolled with a mean age of 6 11/12 years. 21 patients (88%) had a fracture. Early and significant reduction in pain was achieved in most cases. There was no significant alteration in pulse rate and respiratory rate or oxygen saturations. There were no negative side-effects. Satisfaction was rated highly by both patients and providers.
Conclusions: Intranasal fentanyl is safe and easy to use. Early and significant reduction in pain is achieved especially in limb injuries. It closes the time gap to intravenous administration of narcotics or makes it even superfluous. Intranasal fentanyl is now part of the standard of care at our institution.

PedChir-O 18

Forearm fractures in children: Split opinions about splitting the cast

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Background: Fractures of the forearm are the most common fractures in childhood. Various methods of cast immobilisation have been recommended. Currently there is no clear consensus on the optimal method, especially regarding the need for splitting the cast.
Methods: We designed a randomized and controlled trial from June 2008 to September 2009 at the University Children's Hospital Zurich. Children younger than 16 years presenting to the Emergency Department with a closed fracture of the forearm needing reduction were eligible for random assignement to immobilisation in a closed or split circumferential above-the-elbow cast. The primary outcome was the incidence of cast-related problems like compartment syndrome, neurovascular compromise, saw burns or skin lacerations. The secondary outcome was the stability of the fracture.
Results: One hundred patients were randomly assigned to one of the two procedures and analyzed. Follow-up was completed in 98 patients. No compartment syndrome was observed in any group. Moderate skin laceration (<2 cm^2) occurred in two patients, one in the closed cast and one in the split cast group. Secondary splitting was necessary in one patient because of a reversible lymphedema. Significant secondary displacement of the fracture was more common in the split group (6 of 48 patients [13%] versus 4 of 50 patients [8%] in the closed cast group) but did not reach statistical significance.
Conclusions: No significant difference in the incidence of cast-related problems was observed between the groups. Fracture stability was comparable in both groups. We suggest that closed circumferential semi-rigid casts are a safe and effective immobilisation technique for fractures of the forearm in children and splitting can be omitted.

PedChir-O 19

Gait analysis as an evaluation tool in paediatric orthopaedics – a case study

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Introduction: 3D gait analysis (3DGA) is used as an investigation tool to objectify gait patterns before and after treatment. Thereby it can improve monitoring of treatment in children with orthopaedic diseases. In contrast to a CT study, which does not reproduce the effective loading conditions in the hip, knee and ankle joint, 3DGA offers the opportunity to do a dynamic analysis by demonstrating joint loads and muscle activities.
Methods: Gait analysis is performed with a motion analysis system (Vicon, eight cameras), two force plates (AMTI) and a 12-channel EMG telemetry-receiver system (Noraxon). The PlugInGait marker set for the lower extremity is used. All children usually walk barefoot at a self selected speed. The standard gait analysis includes kinematics, kinetics, surface electromyography, a muscle function, and a passive motion test. The calculation of the moments which act on the joints can be made out of the means of kinematic data and the ground reaction force.
Results: A male child aged 15 years (tibial torsion right 21°, left 16°) underwent 3DGA before and one year after derotation osteotomy of both tibiae. Preoperatively, the foot progression angle was highly internally rotated, while the knee was externally rotated. After improving tibial torsion by 15° on the right and 20° on the left side, gait data shows a physiological foot progression and knee rotation angle. The postoperative kinetic data shows an external extension moment during terminal stance phase, which was absent before surgery, as well as a more typical loading in the frontal plane of the knee.
Conclusion: Gait analysis is a valuable and additional tool for quantifying pre- and postoperative results. It is also helpful to demonstrate precise pathological gait patterns and muscle activities, helping to treat the patients and to explain their pathology. Gait analysis should be used more often in clinical settings.

PedChir-O 20

Coxa retrotorta in pediatric population – Diagnostics from clinical examination to gait analysis for preoperative evaluation

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Introduction: Coxa retrotorta is well known to be a prearthrosis of the hip joint and therefore should be corrected operatively. Clinical examination and conventional X-ray are diagnostic standard tools, whereas computertomography (CT) allows quantification of femoral retroversion leading to the precise diagnosis. Nevertheless children are rarely symptomatic and decision making for children and parents is difficult. Gait laboratory analysis gains relevance in diagnosis of unphysiological movement patterns in patients with coxa retrotorta. Reproducible analytic findings causing disadvantageous pathologic impact on the musculoskeletal system of the lower extremity may offer important information for surgical indication and preoperative planning.
Methods: We studied five patients (15.5 ± 2.8) with clinical and CT-diagnosed coxa retrotorta using gait analysis (VICON, 8 MX cameras, Plug-in Gait). Focused were changes and abnormalities of the gait pattern of the patients compared to the physiological gait pattern.
Results: All patients presented with reduced or nullified internal rotation of the hip joint (<10°) and CT-diagnosed pathologic reduced anteversion (<5°) or absolute femoral retrotorsion (<0°). Kinematics in gait analysis showed in all patients an increased adduction of the hip in the coronal plane. Three of five patients had an increased external rotation of the knee joint of the affected leg in the transverse plane according to the clinical picture of kneeing out. The analysis of kinetics indicated a pathologic external valgus moment in all patients and in three of five patients a pathologic external flexion moment within the first 20 % of the stance phase.
Conclusion: We summarize the diagnostic pathway of coxa retrotorta including clinical examination, image-guided tools and gait analysis. Kinematics and kinetics of the gait analysis highlighted unphysiologic movement patterns in patients with coxa retrotorta. Bretin et al. showed in a cadaver study an increased valgus joint alignment in diminished anteversion of the hip joint. Our preliminary results support these findings in showing increased valgus moment in the knee joint in all our patients. It might therefore be discussed, if coxa retrotorta does not only affect the hip, but also the knee joint. Taking these changes in account, gait analysis can serve as an important diagnostic and preoperative planning tool in coxa retrotorta. Further studies will be necessary to analyze the reproducibility of these observations in a larger patient population.

PedChir-O 21

Treatment of congenital anomalies of the forefoot

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Congential anomalies of the toes occur with analog patterns and with a similar incidence as those on the fingers. However, malformations of the forefoot find relatively little interest in publications and meetings and treatment standards are still not well established. The aim is to present a series of children who were treated for malformations of the forefoot and to suggest treatment guidelines. Between 2000 and 2011 we have operated 85 children, among which the polydactylies were by far the largest group. Eleven had a duplication of the great toe, two a central polydactyly, 27 a postaxial polydactyly and 24 a postaxial polysyndactyly. Simple syndactylies were separated only in the 1st web space because the adjacent toes were of unequal length in 6 patients. The preferred age for surgery was around the first birthday. Predisposition to hypertrophic scar formation is an inherent problem and therefore we suggest routine postoperative silicone application. Good results with low complication rates can be achieved in most polydactylies of the foot. In postaxial polydactylies it is not always the most lateral toe that needs to be excised, because preservation of the sixth toe may sometimes better maintain the contour of the foot. Management of preaxial polydactylies can be highly demanding and a congenital hallux varus of the remaining great toe can often be observed despite attempted primary correction. We advise collaboration with a pediatric orthopedic surgeon for complex malformations of the forefoot, particularly for preaxial polydactylies.

PedChir-O 23

Familial clubfoot: three different treatment methods

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Introduction: Club foot treatment has changed from manipulation and dressing to casting, from major surgery to minor surgery and has become subject to trends and 'fashion'. We report the experience of three offspring of one family, all boys with severe congenital bilateral clubfeet, each treated by using a different method.
Patients and Methods:

Patient	Casting	Physiotherapy	Surgery	Splints
*1990	1st 7 m	5 m daily	1 y extensive release	+
*1997	1st 4 m	Weekly, before casting	4 m: TAL R, L 3 y extensive release, R only	+
*2004	taping	DFT: 1st 2 m daily, then 6 m 2× / week	–	–

y: years; m: months; R: right; L: left; TAL: Achilles Tendon Lengthening; DFT: dynamic functional treatment, based on the pathoanatomy of the clubfoot, emphasizing the reduction of the joints in the hindfoot, performed by physiotherapists.

Results:

Type of treatment	Invasive (splints, operation)	Conservative (DFT)
Patient	1 and 2	3
Dorsiflexion/ Plantarflexion	L 10-0-30 / R 20-0-30	L 25-0-50 / R 25-0-50
Eversion	L 10° / R in midfoot only	L 25° / R 25°
Pain	R knee after sport	no pain
Shoes / orthoses	normal shoes / no orthoses	normal shoes / no orthoses
Sport	intensive soccer	soccer, running
Subjective quality of life	excellent	excellent
Parents' satisfaction	more distressing	better handling of the child

Conclusion: All three boys are pleased with their cosmetic and functional results and feel no limitations in their activities. However, using objective assessments of the results, they appear to be superior following DFT. Today both parents would choose the less distressing DFT – although time-consuming –, since the baby seemed to be happier with more freedom of movement and the parents were less distressed by surgery and casting.

PedChir-O 24

Case report: angiosarcoma arising in a benign vascular malformation

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Introduction: Angiosarcoma (AS) represents less than 2% of vascular neoplasms in children, usually of visceral locations or appearing following radiotherapy on benign vascular lesions, with a 5-year survival between 20 and 50%. We report the first pediatric case of an AS arising in a vascular malformation.
Case report: A 4-year-old otherwise healthy former premature girl (29 weeks gestational age, 1140 g birth weight) without risk factors for AS presented with a three month history of a painless subcutaneous ovoid mass on the lateral side of the left thigh, proximal to the knee, measuring 4.5 × 2 × 2 cm, compressible, with cutaneous hyperthermia and redness, but no thrill or surface ulceration. No palpable nodes, distant vascular lesions or masses were noted. Radiological findings on US, MRI and angiography in this growing lesion were not completely in keeping with a vascular malformation, thus warranting surgical removal. The pathology report described in-toto excision of the lesion and identified an area of AS: solidly cellular sheet-like area with occasional spindled cells, presenting mild nuclear atypia, a mitotic rate of 5/10 high-power fields, and immunostains consistent with endothelial differentiation. Of note, the AS region was only in the

deepest area of the vascular malformation and would not have been recognized with a biopsy only; an incomplete excision (biopsy) would have upgraded the AS. A post-operative PET-scan was negative for metastases or residual local tumor activity. The patient underwent a secondary wide excision of the soft tissue and fascia underlying the lesion and of the scar from the first operation. There was no residual lesion identified at histopathology. Because of initial complete resection and reassuring radiological findings, no adjuvant therapy was considered necessary. The patient is completely asymptomatic 20 months after initial presentation, with a normal MRI at 6 postoperative months.
Conclusion: This case suggests that rapidly growing vascular lesions, whose benign nature cannot be formally confirmed by radiology, should undergo complete excision whenever possible.

fPmh-O 1

Health-related quality of life and behavior of triplets at adolescent age

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Background: Higher order multiples are known to be at risk for short and long term developmental impairments. Little is known about the long-term quality of life and behavioral characteristics of surviving triplets.
Aims: To assess the health-related quality of life (HrQoL) and behavior of a cohort of triplets in adolescent age compared to gestational age-, birth weight- and sex-matched singletons.
Methods: We examined 54 triplets of 19 sets and 51 gestational age-, birth weight- and sex-matched singleton controls at an age between 13–16 years regarding their self-rated HrQoL. Proxy reports were obtained from parents and teachers. HrQoL was measured by the Kidscreen 52 (KS 52) child and parent form, and behavior was measured by the Achenbach's Child Behavioral Check List parents (CBCL) and teachers (TRF) form.
Results: All subjects were born prematurely (gestational age range 27 0/7–35 6/7 weeks, birth weight range 750–2500 grams). Self-rated HrQoL was similar in triplets and controls. Parent-reported HrQoL was better in triplets than in controls, particularly for the areas of "Moods and emotions", "Autonomy", "Parent relations and home life" and "Peers and social support". Average CBCL composite global scales ('Internalising-, 'Externalising- and 'Total behavior problem scale') and TRF 'Internalising behavior problem scale' were significantly lower in triplets compared with controls. Compared to community norms both HrQoL and Behaviour measures in triplets were in the normal range. Multivariate analysis showed that socioeconomic status, monochorionicity and gestational age influenced outcome stronger than other perinatal factors.
Conclusions: HrQoL and behavior in triplets at adolescent age is satisfactory and in many aspects better than in matched singleton controls. Socioeconomic factors, monochorionicity and gestational age play a major role in the prediction of long term outcome of these children.

fPmh-O 2

Lessons learned after 2 full scale disaster exercises in a Swiss pediatric hospital

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Introduction: Following a disaster, up to 50% of mass casualties are children. The number of disaster increases worldwide, including in Switzerland. Following national order, the mapping of the various risks of disaster in Switzerland will be completed by the end of 2012. Pre-hospital disaster drills and plans are well established and regularly tested. In-hospital disaster plans are much less frequently tested, if only available. Pediatric in-hospital full scale disaster exercises have never been reported in Switzerland. Based on our local constraints, we set up and evaluated a disaster plan during two full scale exercises.
Methods: In a university hospital treating more than 35 000 pediatric emergencies per year, two exercises involving mock victims of a disaster aged 9 to 14 years old were successively set up over a period of 3 years. The exercises were planned during the day, without modification of the normal emergency room activities. The hospital staff was informed and trained in advance. Variables such as the alarm timing and transmission, triage set-up and function, special disaster medical records utilization, communication and victims' identification were assessed. Family members participated in the second exercise. An evaluation team observed and record exercises activities, identifying strength and weaknesses.
Results: On two separate occasions, a total of 44 mock patients participated, were triaged, admitted and treated in the hospital according to usual standards of care. Alarm transmission was not appropriate during the first exercise. Triage overload occurred on one occasion. In-hospital communication needed readjustment. Identification and in-hospital tracking of the children remained problematic. Hospital employees showed great enthusiasm and stressed the positive effect of full scale exercises on their knowledge of the hospital disaster plan.
Conclusions: Performing real life disaster exercises in a pediatric hospital was very beneficial. The disaster plan was adapted to local needs and updated accordingly. An alarm transmission protocol was elaborated and tested. Triage set-up was adapted and tested. A hospital identification plan for injured children was created and tested. Full scale hospital exercises evaluating disaster plans revealed several weaknesses in the system. Practice readjustments based on local experience were made. A tested pediatric disaster plan adapted to local constraints could minimize chaos, optimize care and support in the event of a real disaster. Children's identification and family reunification following a disaster remains a challenge.

fPmh-O 3

Are Growing Pains a Parasomnia?

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Background: The so-called growing pains (GP) are affecting 4–37% of all children with a peak incidence in preschool age. The underlying cause is still unknown. Although parasomnias (e.g. sleep terrors) share several common features with GP such as age at onset, daytime of appearance, self-limited course and complete absence of symptoms on the following day, an association has not been established between the 2 conditions.
Objective: To analyse the pain characteristics of children with GP and compare the sleep characteristics of the children with and without GP in order to investigate the possibility that GP constitute a parasomnia.
Patients and Methods: The parents of 58 children with a diagnosis of GP according to the Peterson criteria filled a questionnaire about the characteristics of the GP and the sleep characteristics of their children. The study group was then further subdivided in 2 groups according to the time of pain onset: "evening GP" occurring already in the late afternoon and/or at bedtime, and "night GP" occurring only after falling asleep during the first half of the night. 38 children from a study about children's sleep patterns served as control cohort.
Results: Children with GP had more difficulties waking up in the morning ($p < 0.0001$) and re-entering sleep after waking up ($p < 0.0001$), had a lower overall sleep quality ($p = 0.0002$), used more commonly a transitional object (cuddly toy) ($p = 0.002$) and suffered more often from sleep terrors ($p = 0.005$). In a multivariate analysis the factors wake-up difficulties, difficulties with re-entering sleep after waking up, sleep terrors and transitional object remained independently associated with GP. 14 children (24%) qualified for the definition of "night GP" and 16 (28%) had "evening GP". "Night GP" was significantly more common in boys ($p = 0.009$), had fewer pain attacks during one night ($p = 0.04$), were less likely to have their pain attacks following hectic days ($p = 0.04$), had a better overall sleep quality ($p = 0.049$) and more commonly sleep terrors ($p = 0.1$) than children with "evening GP". In the multivariate analysis the factors gender, sleep terrors and occurrence after hectic days remained independently significant.
Conclusion: Children with the so-called GP have a disturbed sleep pattern. The highly significant association of growing pains, especially of the "night GP" variant, with sleep terrors supports the hypothesis of an association between these conditions and warrants further investigations.

fPmh-O 4

Physical performance limitations in adult and adolescent survivors of childhood cancer

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Objectives: There are no studies in Europe reporting on physical performance in survivors of childhood cancer. We aimed to: 1) describe physical performance limitations in adult and adolescent survivors of childhood cancer;2) identify factors associated with performance limitations; 3) assess whether survivors reached physical activity recommendations of the Centres of Disease Control and Prevention (CDC).
Methods: As part of the Swiss Childhood Cancer Survivor Study (SCCSS) we sent a detailed questionnaire to all survivors (≥16 years) registered in the Swiss Childhood Cancer Registry (SCCR) who were diagnosed between 1976–2003 at an age <16 years. Associations between potential risk factors (cancer-related, socio-economic and demographic factors) and performance limitations were assessed with multivariable logistic regression models.
Results: The sample for analysis included 1383 survivors from the Swiss Childhood Cancer Survivor Study (response rate 75%, mean age at survey 26 years, mean time since diagnosis 18 years). Overall, 172 survivors (13%) reported a limitation or handicap affecting physical activity or sports. Most handicaps were related to orthopaedic problems (5%), followed by neurological problems (3%), cardiorespiratory problems (1%), fatigue and pain syndromes (1%), overweight and endurance (1%), visual impairments (1%), and psychological problems (1%). Performance limitations were related to type of cancer (fig. 1) whereas the proportion of survivors reaching physical activity recommendations was not. 74 (43%) of those with a handicap or limitation reached physical activity recommendations compared to 56% of those not limited. In the multivariable logistic regression, being underweight (OR = 3.0; p = 0.02), French or Italian speaking (OR = 3.7; p = 0.004), type of cancer (p <0.001), and having had a relapse (OR = 1.8; p = 0.04)were all associated with performance limitations. Most at risk were survivors of bone tumours (OR = 16.7), brain tumours (OR = 9.5), neuroblastoma (OR = 5.6), and retinoblastoma (OR = 4.4).
Conclusion: Overall only 13% of survivors reported limitations for sporting activities, mainly due to cancer treatments which had been inevitable to save the patients' lives. Despite their limitations, a large proportion of survivors managed to reach physical activity recommendations.

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fPmh-O 5

Moving between families: Behavioural and relationship problems in foster children

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Introduction: Children in foster care often unify early traumatic experiences with additional psychosocial and biological risk factors. So far, research in this field has hardly been carried out in Switzerland. This study examines behaviour and relationship problems in foster children and their association with distress in their caregivers.
Methods: 394 children in foster care and a control group of 344 children from the general population in the German speaking part of Switzerland were included in this epidemiological study. The parents completed the following research instruments: Parental Stress Scale, parents' form of the Essener Trauma Inventar für Kinder, Relationship Problems Questionnaire, Child Behavior Checklist (CBCL) and a purposely designed questionnaire that assesses educational challenges, problems with the child's biological family and other sources of parental distress.
Results: Foster parents reported significantly higher CBCL scores and relationship problems as well as more exposure to interpersonal trauma in their children than parents of the control group. Foster children who had experienced earlier interpersonal trauma showed significantly higher CBCL scores and more relationship problems than foster children without traumatic experiences. Although foster parents reported much more psychological problems in their children than parents of the control group, the difference in parental distress was only moderate between the two groups.
Conclusion: The high number of traumatic experiences in foster children and its association with behavioural and relationship problems indicate the urgent need for child psychiatric support and regular counselling for their caregivers.

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fPmh-O 6

Observation versus early CT-scan in mild closed head injury: a prospective randomized study

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Introduction: Traumatic brain injury is the leading cause of trauma-related death in children. In Geneva, head trauma is found in 16% of injured children, or about 1000 emergency department (ED) visits yearly. While there is good evidence supporting the management of moderate and severe brain trauma in children, it is more difficult to identify children with apparently benign trauma who can safely be discharged without CT scan. This prospective study had two goals: to identify the yield of routine CT scan in children with mild brain trauma (GCS 14-15) and signs of severity and to assess the long term clinical outcomes for these patients.
Methods: One-year prospective study, including all children <16 years visiting the University of Geneva Pediatric ED with a diagnosis of mild head trauma and for whom informed consent was obtained. Patients with signs of severity were randomized to have routine CT vs. optional CT, requested upon clinical course. All participating children were followed up to 90 days after discharge with phone assessments for persistent symptoms.
Results: Mild head trauma without signs of severity (MHT) was found in 511 participating children. Five of them had a head CT, normal in all cases. Signs of severity were present in another 120 patients (MHTSS), 57 of whom were randomized into the routine CT group, which was abnormal in 5, none requiring neurosurgical intervention. Of the 63 patients randomized into the optional CT group, two patients met clinical criteria for a CT, which was normal in both cases. There was no difference between groups for intracranial hemorrhage. Telephone follow-up for all patients revealed that patients with signs of severity had significantly (p <0.01) more headaches and noise intolerance. The prevalence of headache, noise intolerance, fatigue, irritability, and sleep disturbance tended to decrease with time, while vomiting tended to increase.
Conclusion: routine head CT did not modify the clinical course in our patients with MHTSS. Post-concussion symptoms can persist as long as 3 months after trauma.

Free communications INoPSU

INoPSU 1

The Swiss Paediatric Surveillance Unit *SPSU*: Highlights of 15 years of operation

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Introduction: In 1995 the Swiss Paediatric Surveillance Unit (*SPSU*) was launched jointly by the Swiss Society of Paediatrics (SSP) and the Swiss Federal office of Public Health (SFOPH) to collect nationwide data on rare diseases or rare complications of more common diseases of public health importance in hospitalized children. By joining the International Network of Paediatric Surveillance Units (INoPSU), *SPSU* is also internationally integrated.
Methods: *SPSU* is a network of all Swiss paediatric teaching clinics (currently 33), which participate in study protocols and report cases by a card mailing system. The system is well accepted by Swiss pediatricians as the 100% return rate of initial reporting cards puts into evidence.
Results: The results of 15 years of surveillance by *SPSU* and 18 studies lead more than once to changes in public health policy in Switzerland: 1) In 2003 the results of the surveillance of vitamin K deficiency bleeding in newborns demonstrated, that the guidelines needed to be adapted by implementing a third dose four weeks after birth. 2) Due to the overall low prevalence of congenital toxoplasmosis and the lack of scientific evidence regarding the effectiveness of

secondary prophylaxis treatment of pregnant women wild diagnostic testing during pregnancy was abandoned in 2009. 3) Some *SPSU* studies are targeted at the evaluation of the impact of the national vaccination programs such as the surveillance of congenital rubella and acute flaccid paralysis as a marker for poliomyelitis. 4) A recently completed study showed that 1 in 3000 children younger than 1 year is affected by the shaken baby syndrome, which increased attention to a serious public health problem. 5) The most recent study addresses the prevalence of extendend spectrum beta-lactamase (ESBL) producing gram negative bacteria in children. Preliminary results give rise to the suspicion that infections with ESBL-positive bacteria are predominantly acquired in the community rather than in the hospital setting.
Conclusion: The *SPSU* with its focus on rare and severe pediatric illnesses is an important surveillance system complementary to the mandatory notification system and the voluntary Sentinella system. *SPSU*'s role as an important source of evidence for public health policy in Switzerland has been demonstrated. The collaboration of *SPSU* with INoPSU leads to an exchange of first line information on results of ongoing studies, potentially unnoticed issues, and allows an international benchmarking in evidence-based policy making.

INoPSU 2

The Belgian Paediatric Surveillance Unit "PediSurv"

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Introduction: In 2003 the Belgian network PediSurv (**Pedi**atric diseases **Surv**eillance) was developed in order to collect information about some vaccine preventable diseases. Mumps, measles, acute flaccid paralysis (AFP) were included. Invasive pneumococcal disease (IPD) was added in 2006 following introduction of the 7-valent pneumococcal conjugate vaccine, congenital rubella syndrome (CRS) in 2007 and Hemolytic Uremic Syndrome (HUS) in 2009. During the Influenza A(H1N1)2009 pandemic, a subset of paediatricians participated influenza surveillance of children <5 years old.
Methods: All Belgian paediatricians (and GPs working in Brussels) were invited to participate to this network. Participation is voluntary and requests a monthly reporting of the diseases even in case of a zero case notification. Zero notification and reporting are possible by returning the monthly paper form or by clicking a link in a monthly email reminder.
Results: In 2010 approximately 500 clinicians participated monthly (with 35% of the Belgian paediatricians). Measles incidence remained above the elimination threshold of <1 per million and measles outbreaks are ongoing in 2011. Non-polio AFP rate ranged from 0.06 to 0.50, meaning the surveillance system is missing cases. Incidence of IPD in children <2 year decreased significantly after introduction of the PCV7 vaccine and was estimated at 104/100 000 cases in 2009. Vaccine effectiveness on vaccine-serotypes was estimated at 97%. We demonstrated serotype replacement, especially with serotypes 1, 7F, 19A and 5. HUS study highlighted the predominance of *E. Coli O157:H7* as the principal aetiology. During the influenza A(H1N1)2009 pandemic we demonstrated that hospitalization rate was highest among children <1 year.
Conclusion: Although PediSurv is not an exhaustive surveillance system, the stable participation allowed to observe trends and provided valuable information. Information is being used for decision-making in public health, such as the recommendations for the use of new PCV covering more serotypes and to monitor progresses in different elimination goals. Data is used at international level by the ECDC and WHO.

INoPSU 3

The BPSU – 25 years of informing vaccination policy?

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Introduction: The British Paediatric Surveillance Unit (BPSU) has facilitated the study of rare paediatric conditions over the last 25 years and informed public health policy in the UK and abroad. The aim of this paper is review vaccine preventable disease (VPD) studies carried out through the BPSU and their impact on vaccination policy.
Methods: A review of all VPD BPSU studies and description of their public health impact.
Results: Ten percent of all BPSU studies (n = 9) examined conditions with available vaccines at the time of study. Two studies are ongoing and one has concluded but not reported. Public health contribution of the studies broadly fall into the following categories:
1. Assessment of the pre-vaccine introduction burden/descriptive epidemiology – this included the varicella outcome study.
2. An evaluation of the impact of a current vaccination programme included studies on congenital rubella syndrome, Haemophilus influenza B (HiB) and acute flaccid paralysis.
3. An evaluation of the safety of a vaccine programme included studies on Guillain Barré Syndrome, MMR meningoencephalitis and subacute sclerosing panencephalitis.
The HiB studies documented a significant incidence reduction after introduction of the UK vaccination programme, but noted an increase of cases towards end of the study period, prompting a review of the vaccination schedule and introduction of a booster.
Conclusion: BPSU VPD studies played an important part in informing vaccination policy, have successfully monitored VPDs, and informed important schedule amendments, such as introducing a HiB booster.

INoPSU 4

The Importance of engaging with patient support groups: education, research and influencing national policy on rare disease

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Introduction: Research into rare diseases is difficult due to lack of awareness of the importance of rare diseases, lack of research funding and infrastructure. Paediatric Surveillance Units (PSUs) produce important epidemiological and clinical information about rare diseases which is usually not available from other sources.
Methods: Australian Paediatric Surveillance Unit conducts surveillance for up to 16 different rare childhood conditions simultaneously, with monthly reporting by ~1350 paediatricians (~80% by e-mail). Over 50 conditions have been studied. APSU recognised the need to engage with and inform parents, carers, the community and policy-makers about new knowledge generated through surveillance studies.
Results: The APSU is actively engaged with a number of policy and patient support groups including the Steve Waugh Foundation, SMILE Foundation and the Association of Genetic Support Australasia. These partnerships have been very powerful in raising awareness of rare diseases in the community, because these orgnisations help disseminate results of surveillance. Our engagement with these groups has also led to development of a draft national plan for rare diseases and a funding proposal to the Australian Research Council to study the impacts of rare diseases on families, health services and paediatricians. The paediatricians' survey will determine their educational needs and preferences for mode of delivery of education and resources, and will further raise their awareness of rare diseases. APSU already provides information for parents and carers in lay language, including a summary of rare conditions, the rationale for the APSU study and links to educational resources and family support groups (www.apsu.org.au). Information sheets may be printed by paediatricians and given to patients for education. The APSU website also contains lists of publications about rare diseases we have studied for access by parents and paediatricians.
Conclusion: Paediatric surveillance units produce extremely useful information about rare diseases which is relevant to clinicians, policy makers, parents and carers. It is important to ensure that new knowledge produced by PSUs is widely and easily accessible. Parents and carers are important allies when developing new studies and when raising awareness of the results of surveillance studies.

INoPSU 5

Paediatric Emergency Department Surveillance of Injuries Associated with Baby Products, 1990–2008

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Introduction: Baby products (BPs) are commonly used items in most Canadian households with children. A wide array of products and designs are available and as such there is considerable potential for various injury scenarios. It is not uncommon to see recalls of various BPs. In the literature, there are few reports related to individual products (e.g. baby walkers, strollers) but no comprehensive studies examining the full range of BPs.
Methods: Canadian Hospitals Injury Reporting and Prevention Program (CHIRPP) include emergency departments of 11 paediatric and 4 general hospitals. The entire database was searched for cases involving injuries associated with BPs (cribs, high chairs, strollers, baby walkers, bouncy chairs, infant swings, changing tables, toilet training seats, jumpers, playpens, car seats, carriers, crib mattresses, bath seats and soothers/pacifiers).
Results: Over 25,000 cases was identified, accounting for about 4% of all incidents involving children less than 72 months; 60% of the incidents involved infants (0–11 months). Cribs and strollers were the most frequent BPs involved (31% of all cases); fall from a BP was the mechanism of injury in 60% of incidents. Overall rate of malfunctioning/breakage was 2% with significant variations between BPs and the most frequent being "Jolly Jumpers" (32.1%) and infant swings (14.1%). Overall, 5.5% required hospitalisations and 71.1% reported injuries to the head, face and neck region, significantly higher than the overall CHIRPP proportion of such injuries in the same age group (51%, p <0.001).
Conclusion: A wide range of BP-related injuries were identified in CHIRPP. The high proportion of head, face and neck injuries as well

as the potential for some BPs to malfunction raises concerns. CHIRPP continues to report injuries associated with baby walkers following a 2004 ban on the sale, advertisement and importation of these products. This reinforces the importance of ongoing surveillance, together with the ban and recall of baby products.

INoPSU 6

Alcohol Intoxication among children in The Netherlands in the period 2007–2010

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Aim: To study the number and characteristics of children who are admitted to the hospital because of alcohol intoxication and to create a prevention system in line with primary care.
Setting: All Hospitals in the Netherlands
Methods: All Paediatricians of the country report monthly by standardised interview, the parameters of the children who were hospitalised in the paediatric wards.
Results: During the years 2007–2010 an increase could be seen in the total number of admitted children (fig. 1). After a period of diminution; the mean age of this group increased in the years 2009 and 2010 (fig. 2).

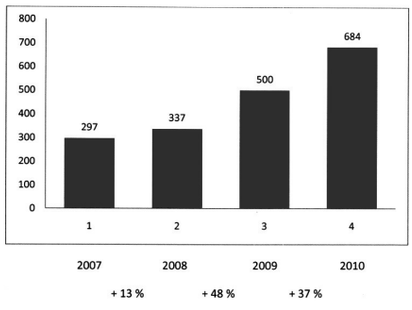

Figure 1

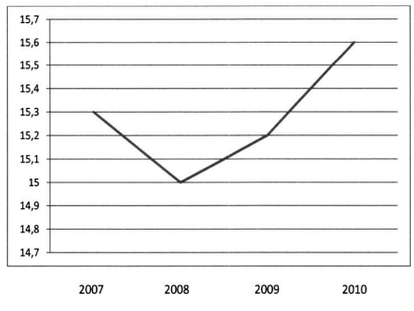

Figure 2

Conclusion: The number of children admitted with alcohol intoxication in The Netherlands is still rising significant. This study shows the seriousness of adolescent intoxication.
Key words: children, alcohol, intoxication, hospital admission

INoPSU 7

The 3rd national survey on childhood celiac disease in the Netherlands: still changing epidemiology and clinical presentation.

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Introduction: Two national surveys on childhood celiac disease (CD) in the Netherlands, performed between 1975–1990 (S1) and 1993–2000 (S2), showed a rising incidence and changing clinical presentation of CD. We investigated the epidemiology and clinical presentation of CD in the Netherlands and compared them to previous findings.
Methods: Children newly diagnosed with CD were identified through the National Pediatric Surveillance System. Data on gender, age, and CD were anonymously collected.
Results: In 2010, 301 cases of CD were reported. We collected and analyzed completed questionnaires on 230 cases (76%). There were 66% girls (S1 58%, S2 62%), with a median age of 4.4 yrs. (S1 1.5 yrs, S2 2.1 yrs). Cases were diagnosed on clinical grounds (78%), through screening in associated disorders (13%) and in family members (9%). The incidence rate was 0.91/1000 live born infants (S1: 0.22, S2: 0.81). Chronic diarrhea occurred in 22% (S1 72%, S2 41%), distended abdomen in 25% (S1 76%, S2 48%), failure to thrive in 19% (S1 63%, S2 24%), lassitude in 20% (S1 unknown, S2 12%), abdominal pain in 44% (S1 7%; S2 16%), and anorexia in 34% (S1 0%, S2 5%).
Conclusion: The epidemiology of childhood CD in the Netherlands is still changing. The percentage of females, age at diagnosis and incidence are rising, compared to both earlier surveys. Chronic diarrhea, distended abdomen and failure to thrive occur less often, and abdominal pain, lassitude and anorexia occur more often. We suggest that these findings may partly be explained by better recognition of less typical clinical presentations, in addition to active case finding in relatives and associated disorders.

INoPSU 8

Shaken baby syndrome (SBS) in Switzerland: results of a prospective follow-up Study 2002–2007

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Objective: Since the incidence of SBS in Switzerland is not known, we conducted a nationwide retrospective study covering 5 years.
Method: Data collection through SPSU system. Inclusion criteria: Presence of 1) ≥2 clinical symptoms (altered consciousness, convulsions, respiratory irregularities, bulging fontanel) or 2) 1 eye findig (retinal haemorrhages, vitreous haemorrhages) or 3) 1 MRI/CT finding (subdural haematoma, subarachnoid haematoma, parenchymatous lesions) or 4) history of shaking; all these findings in a child ≤6 years. Exclusion criteria: age >6 years or documented accident/disease explaining symptoms/findings. Outcome measurement following **K**ing's **O**utcome **S**cale for **C**hildhood **H**ead **I**njury (KOSCHI); (Crouchman M, et al. Arch Dis Child. 2001;84:120–4).
Results: Of 56 reported cases 49 met inclusion criteria. Cases reported from only 13 cantons; unexplained high incidence in 2005. High male preponderance (31 m, 18 f), mean age 4.0 months (1–58). Clinical symptoms present in 42 patients, retinal/vitreous haemorrhages documented in 39, brain imaging pathologic in 46 patients. Shaking confessed in 18 cases. Outcome (n = 47 patients, 2 patients lost for follow-up): Death (KOSCHI 1) 8 (16%), vegetative state (KOSCHI 2) 0, severe disability (KOSCHI 3) 11 (28%), moderate disability (KOSCHI 4) 14 (36%), good recovery (KOSCHI 5) 14 (36%).
Conclusions: 1) According to our data incidence of SBS in Switzerland is 14 in 100 000 live births, which corresponds to incidences in other western countries. 2) Demographic characteristics and outcome of our patients are comparable to published studies. 3) Lack of adequate reporting in half of Swiss cantons.

INoPSU 9

Hemolytic uremic syndrome in Switzerland: Twelve years nationwide surveillance

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Background: Hemolytic-uremic syndrome (HUS) is a multisystem disorder associated with significant morbidity and mortality, predominately affecting infants and young children. Beside single center studies, nationwide data on childhood HUS in Switzerland have not been available so far.
Methods: Prospective, national surveillance through the Swiss Pediatric Surveillance Unit (SPSU) covering the period April 1997 through March 2003 and April 2004 through March 2010, both including a follow-up evaluation at one year.
Results: During the first period, 114 cases (median age 21, range 0–161 months, 88% ≤5 years old; 50% boys) were reported by 38 pediatric units (annual incidence rate 1.42 per 105 children aged 16 years or less). Eighty-nine percent of cases were associated with Shiga-toxin (Stx) producing *Escherichia coli* (STEC) and half of the remaining cases were due to infection with *Streptococcus pneumoniae*. Mortality was 5.3%; severity of thrombocytopenia and central nervous involvement significantly (P <0.01) correlated with fatal outcome. Strong association (P <0.01) was found between the incidence rate of Stx-HUS and indicators of rural density (i.e. livestock breeding farms per population, number of cattle per cultivated area). Preliminary analysis of the second period reveals similar results: 135 patients (median age 22, range 0–181 months, 44% boys). Association with STEC was found in 83%, in 6% HUS was due to invasive infection with *Streptococcus pneumonia*e. Mortality was 4.4% resulting in an overall mortality of 4.8% taken both periods together.
Conclusions: By means of the SPSU, reliable clinical, epidemiological, and bacteriological data on childhood HUS in Switzerland have been established: HUS is frequent, especially in young children and is mostly associated with STEC infection. The

incidence rate of Stx-HUS significantly correlates with indicators of livestock farming intensity, supporting the impact of direct and indirect contact with animals or fecal contaminants in transmission of STEC to humans. The present association between thrombocytopenia, central nervous system involvement and worse outcome needs confirmation and if any, further evaluation.

INoPSU 10

Guillain-Barré /Fisher syndrome surveillance in the UK; is there a temporal association with influenza infection or vaccination?

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Introduction: The objective is to identify new cases of Guillain-Barré syndrome /Fisher syndrome (GBS/FS) in UK children aged 16 years or less & to detect any temporal association with influenza infection or vaccination.
Methods: Questionnaires are sent to paediatricians who report cases via the British Paediatric Surveillance Unit (BPSU) office, asking about the clinical history, relevant physical findings & results of investigations in children with GBS/FS. The HPA sends a questionnaire directly to General Practitioners asking about vaccinations.
Results: September 2009 – January 2011. Surveillance stops in September 2011. 79 children met the criteria for Guillain-Barré syndrome & 2 for Fisher syndrome. 61 of these 81 children had clinical or laboratory evidence of an infection in the 3 months before their GBS/FS symptoms as follows: upper respiratory tract infection: 24, gastro-enteritis:17, swine influenza:6, pyrexia of unknown origin:4, chicken pox:2, "flu-like symptoms":2, cytomegalovirus:2, Ebstein Barr virus:2, influenza A:1, influenza B:1. The HPA has confirmed that 3 children with GBS were given HINI vaccinations and 3 were given seasonal influenza vaccines as follows: *H1N1 vaccine;* one child was given 2 doses of Celvapan (Baxter) 4 weeks before the onset of GBS: this child was also given chicken pox vaccine (Varilix). The other 2 H1N1 vaccinations were given 6 & 7 months before the onset of GBS. *Seasonal influenza vaccine;* one child was vaccinated 3 days before the onset of GBS & the other 2 were vaccinated respectively 4 & 7 months before the onset of GBS.
Conclusion: These results are preliminary, awaiting confirmation of all 81 notified childrens' vaccination details via the HPA. The data suggest that GBS/FS cases are strongly associated with previous infections. There is little evidence of an aetiological link between H1N1 vaccinations & GBS/FS in the vast majority of children who received H1N1 or seasonal influenza vaccinations.
Acknowledgements: funded by the English Department of Health [PRP: 0190047] and carried out via the BPSU methodology.

INoPSU 11

Vitamin D deficiency rickets in Australia: need for screening and early treatment in high risk groups

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Introduction: Vitamin D deficient rickets can lead to poor growth, motor delay, bone fragility and hypocalcaemic seizures. There is also increasing evidence for non-skeletal roles of Vitamin D including autoimmune disease, cardiovascular disease and cancer. The incidence of this potentially serious and preventable condition is not known in Australia.
Methods: Prospective surveillance study using the Australian Paediatric Surveillance Unit (APSU) with monthly reporting by ~1350 paediatricians (80% by email) from January 2006 to July 2007. Paediatricians were asked to report all children ≤15 years of age with vitamin D deficiency rickets ie. 25-hydroxy vitamin D level <50 nmol/L, and elevated alkaline phosphatase levels, and/or radiological evidence of rickets.
Results: Over an 18 month period APSU was overwhelmed with 851 notifications suggesting that Vitamin D deficiency rickets is not rare in Australia as previously thought. As the APSU system is designed specifically for the study of rare conditions, it was felt that adequate data had been collected and the study ceased at 18 months into the collection period. Of the 851 notification, 156 were duplicate cases, 243 did not meet the inclusion criteria and 54 children had missing biochemical and/or radiological confirmation of simple vitamin D deficiency rickets. 398 children met case definition criteria (55% male), median age of 6.3 years, (range 0.2–15). The incidence was estimated at 6.6 per 100 000 children aged <15 years per annum. Of 95 children who had a wrist x-ray, 67(70%) had rachitic changes. 98% had dark or intermediate skin colour and with 18% of girls partially or completely veiled. 86% of the cases were of African background, while 75% were refugees. Preventative vitamin D treatment was given in 4% prior to diagnosis.
Conclusions: Vitamin D deficiency rickets is a significant problem in Australia among known high risk groups. Public health campaigns to screen for and prevent vitamin D deficiency are essential. The many reports made in error suggest that there may be many children who have low vitamin D levels without the confirmatory biochemical or radiological signs, and may be at risk of developing rickets.

INoPSU 12

Can active surveillance improve reporting of serious and life-threatening adverse drug reactions? – The Canadian Paediatric Surveillance Program Experience

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Introduction: Adverse drug reactions (ADRs) are an important cause of death and illness. Health-related accreditation bodies estimate that 95% of all ADRs are not reported.
Methods: A survey was conducted in December 2007–January 2008 to assess the value of an active surveillance methodology in overcoming documented barriers to reporting found in passive surveillance and in identifying solutions to improve ADR recognition and reporting, and to evaluate the effectiveness of the CPSP collaborative model. The survey was pilot tested via phone interviews of eight CPS Board members and information was collected via online and mailed questionnaires and 700 responses were received.
Results: The survey response rate was 28%. Almost all (90%) of respondents participate in the CPSP monthly; 68% felt that serious and life-threatening ADRs were rare in children, 24% reported such an event to the CPSP and 83% felt the study should continue. Identified barriers included heavy workload (53%), time to complete the detailed questionnaire (39%) and difficulty determining whether the problem is associated with that drug versus a disease (53%). Fear of legal liability and patient confidentiality were not seen as barriers to reporting. Solutions to support increase ADRs reporting included greater feedback (61%), Tips of the Month (60%), increased training (61%) and a simplified questionnaire (53%).
Conclusion: This evaluative survey has documented that collaboration with a national specialty active surveillance program is a very effective way to promote awareness and buy-in of participants to the ADR reporting process.

INoPSU 13

Severe Hyperbilirubinaemia of the Newborn: A Problem in Switzerland?

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Aim: To determine the incidence of severe hyperbilirubinaemia in late preterm and term infants in Switzerland.
Patients and methods: A nationwide prospective study was performed in collaboration with the Swiss Paediatric Surveillance Unit. Data of all newborn patients ≥35 0/7 weeks of gestational age (GA) with at least one value of total serum bilirubin (TSB) exceeding the upper limit of exchange transfusion (ET) were registered in a centralized database. We here present data of the study period 2007-2008. Comparative numbers about live births were supplied by the Swiss Federal Office of Statistics.
Results: We report on 60 cases exceeding the upper limit of ET, representing an incidence of 41/100 000 live births ≥35 0/7 weeks GA of which 68.3% were born at term and 66.7% were males. In only three infants TSB was >510 µmol/l (30 mg/dl). Blood group incompatibility was the most frequent aetiology. In several patients severe hyperbilirubinaemia could have been avoided if anamnestic risk factors and clinical signs were assessed and interpreted correctly. Although indication for ET was given in all patients, only in 13 cases (21.7%) ET was performed, without any correlation between performed ET and severity of hyperbilirubinaemia.
Conclusions: With a better application of the existing recommendations, some cases of severe hyperbilirubinaemia could have been avoided. Nevertheless, severe neonatal hyperbilirubinaemia has a lower incidence in Switzerland compared to other countries, probably due to a longer postnatal hospitalization duration and a well developed outpatient care system. In 2012, case payment (Swiss-DRG) will be introduced in Switzerland for all hospitalized patients. This change risks to induce a shortening of the duration of perinatal hospitalization for economic reasons and potentially a disturbance of the so far well functioning health care system for neonates. Therefore, the incidence of severe hyperbilirubinaemia in neonates needs to be kept under close surveillance over the next years.

INoPSU 14

A lasting legacy? A BPSU study case review to evaluate public health impact

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British Paediatric Surveillance Unit

Introduction: The British Paediatric Surveillance Unit (BPSU) has facilitated the study of rare non-infectious paediatric conditions over the last 25 years. We argue that many studies had measurable impact on UK and international public heath policy and this paper reviews two high impact BPSU studies to demonstrate their role in informing public health policy.
Methods: We present two examples of rare conditions, where BPSU studies have led to significant policy changes – Vitamin K deficiency and Reye's syndrome (rare non-inflammatory encephalopathy with hepatic dysfunction). We review and describe the policy changes and the role that BPSU studies had in shaping these.
Results: The study on Reye's syndrome demonstrated an association between this condition and use of Aspirin in adolescents, prompting the UK Committee on Safety of Medicines to issue warnings of Aspirin use in the under 12 year olds. Long-term BPSU surveillance showed the virtual elimination of Reye's in children in the UK. The finding of an association between Vitamin K deficiency and haemorrhagic disease of the newborn (HDN), led to the UK recommendation of Vitamin K administration in neonates, followed by a cancer controversy. Subsequent BPSU studies (ending 2009) reconfirmed that intramuscular injection is safe and effective to prevent Vitamin K associated HDN.
Conclusion: BPSU studies continue to have a large impact on public health policy. Two case studies demonstrated that surveillance of rare diseases led to significant findings, resulting in public health recommendations with measurable population impact demonstrated by monitoring through BPSU follow-up studies.

Free communications Young Researchers

YR-O 1

Antibody responses to natural influenza A/H1N1/09 disease or following immunization with adjuvanted vaccines, in immunocompetent and immunocompromised children

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[1]Centre de Vaccinologie et d'Immunologie Néonat; [2]Plateforme de Recherche Pédiatrique; [3]Centre de Recherche Clinique; [4]Laboratoire de Virol. et Centre Nat. de l'Influenza, Hôpitaux et Université de Genève

Background: Before/during the influenza A/H1N1/09 pandemic of autumn/winter 2009/2010, we compared antibody responses elicited by the infection and by immunization with adjuvanted vaccines, in immunocompetent and immunocompromised children.
Methods and Results: Prospective parallel cohort field study enrolling children with confirmed influenza A/H1N1/09 infection or immunized with 1 (immunocompetent) or 2 (immunocompromised) doses of influenza A/H1N1/09 squalene-based AS03-(Pandemrix®, GSK) or MF59-(Focetria®, Novartis) adjuvanted vaccines. Antibody titers were measured by hemagglutination-inhibition (HAI) and microneutralisation (MN) assays 4-6 weeks post-vaccination/infection. Vaccine adverse events were self-recorded during 7 days. Antibody titers in 48 immunocompetent children after a single immunization (HAI and MN seroprotection rates: 98%; HAI-geometric mean titer (GMT): 395, MN-GMT: 370) were as high as in 51 convalescent children (seroprotection rates: 98% (HAI), 92% (MN); GMT: 350 (HAI), 212 (MN). Twenty-seven immunocompromised children reached slightly lower seroprotection rates (HAI: 89%, MN: 85%) but similar antibody titers (HAI-GMT: 306, MN-GMT: 225) after 2 immunizations. Adverse events increased with age (P = 0.01) and were more frequent with Pandemrix® than Focetria® (P = 0.03).
Conclusion: In immunocompetent children, similarly high seroresponses may be expected after a single dose of adjuvanted vaccine as after infection. Two vaccine doses were sufficient for most immunocompromised children.

YR-O 2

Double tracer gas single breath washout – validation and comparison with conventional lung function tests in children with and without cystic fibrosis

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Background: Small airway disease occurs early in Cystic Fibrosis (CF), and may cause ventilation inhomogeneity (VI). Inert tracer gas washout tests over single (SBW) or multiple breaths (MBW) are sensitive for VI. Due to time and effort required for measurements these tests are currently underutilised in clinical settings. The aims of our study were (i) to develop and validate a simple technique for a new tidal SBW of double tracer gas and (ii) to assess if this SBW is feasible in children, and provides information on VI in early CF lung disease.
Methods: The double tracer gas mixture contained 5% sulfur hexafluoride (SF_6) and 26.3% helium (He), had similar total molar mass (MM) as air, and was applied for a single normal tidal breath. MM was measured using an ultrasonic flowmeter, SBW-MM curves were plotted vs. expired volume. We fitted indices describing the shape of the SBW-MM curve: MM-slopes of tidal phase II and III (MM_SII; MM_SIII), and area under the SBW-MM curve (AUC). (i) MM was compared to mass spectrometry. SBW reproducibility was assessed in 13 healthy adults. (ii) In a cross-sectional study we investigated SBW reproducibility and association with conventional lung function tests in CF and healthy children.
Results: (i) MM reflected SF_6 and He washout patterns measured by mass spectrometry. Linear regression R^2 was 0.98. Coefficient of variation (CV%) of AUC was 6.8%. (ii) Seventy CF children, mean (SD) age 11.7 (3.5) years, and 42 healthy children aged 11.5 (3.9) years performed nitrogen MBW and SF_6/He SBW. CV% for MM_SIII, MM_SII, and AUC was 15.5%, 15.3%, and 8.7%, respectively. All three CV% were similar between CF and healthy children. MM_SIII and MM_SII differed significantly between CF and healthy children. MM_SIII and MM_SII were associated with the MBW derived lung clearance index (p <0.001 for both).
Conclusion: The new tidal SBW of double tracer gas is a reliable, fast and easy to perform lung function test. We identified SBW indices characterizing VI and separating healthy from CF children. Furthermore, detailed analysis of the SBW may provide additional and yet unknown information on VI, possibly of interest in children with CF.

YR-O 3

Glucagon-like peptide 1 and peptide YY show a significant increase in obese children compared to normalweight children

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Background and Aims: Food intake is related to the release of satiety hormones of the gastroentero-endocrine cells. The physiology of these hormones during normal meal intake remains unclear. Further it is known that impaired insulin sensitivity and diabetes mellitus type 2 is a consequence of overweight. This study compares the response to meal intake of three gut hormones (glucagon-like peptide 1 (GLP-1), peptide YY (PYY) and ghrelin) between lean and obese adolescents. In parellel we investigated the insulin sensitivity of these children.
Methods: A total of 16 obese (8 males, 8 females; mean BMI 31.2 kg/m^2) and 14 lean (5 males, 9 females; mean BMI 20.7 kg/m^2) adolescents, aged 8–16 years, consumed a mixed 500 kcal meal (bread, butter and chocolate milk) during which plasma was obtained for measurement of ghrelin, GLP-1 and total PYY. Fasting glucose and insulin levels were used to calculate insulin sensitivity using the HOMA index.
Results: Fasting ghrelin tended to be lower in obese than in lean subjects (not statistically significant); after food intake ghrelin levels fell in both groups. Fasting GLP-1 and PYY levels were similar in both groups; after food intake PYY and GLP-1 levels were significantly attenuated in the obese group (p <0.05, respectively). Obese adolescents showed higher fasting and postprandial glucose and insulin levels (p <0.05, respectively). Postprandial glucose levels in the lean subjects had returned to baseline by 120 min after the start of the meal. Glucose levels at 120 min. in the obese subjects still remained

on average higher than those at baseline. As expected, the HOMA index was significantly higher in obese compared to lean subjects ($p < 0.01$) suggesting insulin resistance.
Conclusions: 1. Meal-related changes in ghrelin, GLP-1 and PYY in obese subjects suggested a significantly different hormone release pattern compared to normal weight subjects. This disturbed release could contribute to the development or worsening of obesity. 2. Obese adolescents have an increased insulin resistance.

YR-O 5

Estradiol and progesterone strongly inhibit the innate immune response of newborn mononuclear cells

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Introduction: Newborns are particularly susceptible to bacterial infections due to qualitative and quantitative deficiencies of the neonatal innate immune system. However, the mechanisms underlying these deficiencies are poorly understood. Given that fetuses are exposed to high concentrations of estradiol and progesterone during gestation and at the time of delivery, we hypothesized that these hormones impair neonatal innate immune responses.
Methods: Umbilical cord blood was collected on placentas from healthy term newborns and adult blood was taken from healthy volunteers. Cord blood mononuclear cells (CBMCs) and adult peripheral blood mononuclear cells (PBMCs) were extracted by Ficoll Hypaque gradient density centrifugation. Monocytes were isolated from mononuclear cells using magnetic microbeads coupled to an anti-CD14 antibody. Cells were incubated for 16 hours with estradiol, progesterone, hydrocortisone and steroid receptor agonists before stimulation with microbial products. Production of cytokines, activation of intracellular signaling pathways and expression of pattern recognition receptors, estrogen receptors and progesterone receptors were measured by bioassay, ELISA, RT-PCR and Western blotting.
Results: At concentrations present in umbilical cord blood, estradiol and progesterone are powerful inhibitors of NF-κB activation and cytokine production by CBMCs and newborn monocytes exposed to endotoxin (LPS), bacterial lipopeptide (Pam3CSK4) and Escherichia coli and Group B Streptococcus, the two most common causes of neonatal sepsis. Remarkably, estradiol and progesterone reduce cytokine production by CBMCs as efficiently as hydrocortisone. Moreover, CBMCs and newborn monocytes respond to lower concentrations of estradiol and progesterone than adult PBMCs and monocytes. The increased sensitivity of CBMCs to the anti-inflammatory effects of estradiol and progesterone is associated with higher expression levels of estrogen and membrane progesterone receptors but is independent of a downregulation of TLR2, TLR4 and MYD88 in newborn cells.
Conclusion: Elevated blood concentrations of estradiol and progesterone acting on mononuclear cells expressing high levels of steroid receptors contribute to impair innate immune responses in newborns. Therefore, intrauterine exposure to estradiol and progesterone may participate to increase susceptibility to infection during the neonatal period.

YR-O 6

The RNA-binding protein RBM3 is involved in hypothermia induced neuroprotection

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Introduction: Induced hypothermia is the only therapy with proven efficacy to reduce brain damage after perinatal asphyxia. While hypothermia down-regulates global protein synthesis and cell metabolism, low temperature induces a small subset of proteins that includes the RNA-binding protein RBM3 (RNA-binding motif protein 3), which has recently been implicated in cell survival.
Methods: First, RBM3 expression was determined in the developing brain of mice. Second, RBM3 induction in response to hypothermia was studied in cortical organotypic slice cultures (COSCs), primary neurons and rat pheochromocytoma cells(PC12). Third, we investigated whether in PC12 cells neuroprotection by hypothermia might involve RBM3, employing siRNA knockdown and RBM3 over-expression experiments.
Results: Immunohistochemistry of the developing postnatal murine brain revealed a spatio-temporal neuronal RBM3 expression pattern very similar to that of doublecortin, a marker of neuronal precursor cells. Mild hypothermia (32 °C) profoundly promoted RBM3 expression and rescued neuronal cells from forced apoptosis as studied in primary neurons, PC12 cells, and cortical organotypic slice cultures. Blocking RBM3 expression in neuronal cells by specific siRNAs significantly diminished the neuroprotective effect of hypothermia while vector-driven RBM3 over-expression prevented activation of caspase-3, internucleosomal DNA fragmentation, and finally LDH release also in the absence of hypothermia.
Conclusions: Together, neuronal RBM3 up-regulation in response to hypothermia apparently accounts for a substantial proportion of hypothermia-induced neuroprotection. In addition, the expression of RBM3 in neuronal progenitor cells seems promising, which might suggest a function for cell renewal and repair to the CNS.

YR-O 7

Anti-*Aspergillus* defense is NET-Calprotectin dependent and reconstituted after Gene Therapy in human Chronic Granulomatous Disease (CGD)

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Introduction: Infection by *Aspergillus spp* is a potentially lethal disease in patients with neutropenia or impaired neutrophil function. We showed previously that *Aspergillus* hyphae which are too large for neutrophil phagocytosis are growth inhibited by reactive oxygen species-dependent formation of neutrophil extracellular traps (NETs). NETs are composed of chromatin (DNA and histones) and intracellular antimicrobial substances, liberated by activated neutrophils for trapping of microbes and concentrated antimicrobial defense. This process is defective in chronic granulomatous disease (CGD) due to impaired phagocyte nicotinamide adenine dinucleotide phosphate (NADPH) oxidase function.
Methods: Antifungal activity of free and NET-released calprotectin (S100A8/A9) was assessed by incubation of *A. nidulans* with purified calprotectin, induced NETs from control and CGD neutrophils after gene therapy (GT) in presence or absence of Zn^{2+} or α-S100A9 antibody, and with induced NETs from wild type or *S100A9*$^{-/-}$ mouse neutrophils.
Results: The antifungal agent responsible for *A. nidulans* growth inhibition within NETs has not been characterized. We identified the host zinc-chelator calprotectin as neutrophil-associated antifungal agent expressed within newly formed NETs after reconstitution of NADPH oxidase function by GT for human CGD. Calprotectin prevents *A. nidulans* growth reversibly at low concentration, and leads to irreversible fungal starvation at higher concentration. Reconstituted NET-formation was associated with rapid cure of pre-existing therapy refractory invasive pulmonary aspergillosis *in vivo.*
Conclusion: These results demonstrate the critical role of NET-associated calprotectin in human innate immune defense to invasive *Aspergillus* infection.

YR-O 8

Deficient anti-viral control in cystic fibrosis airway epithelial cells upon virus infection

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Background: Viral infections play an important role in cystic fibrosis (CF) respiratory morbidity but their contribution to lung disease progression is poorly understood. We hypothesized that upon viral infection there is a deficient production of antiviral mediators [type I and III interferons (IFNs) and interferon stimulated genes (ISGs)] and an exaggerated pro-inflammatory response in CF. We therefore studied the anti-viral and inflammatory response of the CF airway epithelium upon infection with respiratory viruses.
Methods: Epithelial CF and non-CF cell lines (IB3-1/-S9, UNCCF2T/UNCN2T, 16HBE14o-/CFBE14o-) and primary airway epithelial cells (nasal, bronchial) from CF patients and healthy controls were used in *in vitro* studies. Epithelial cells were cultured and infected with respiratory viruses (RV-16, RV-1B, RSV) at a multiplicity of infection of 2. Viral replication was analyzed by real-time (RT)-PCR and HeLa titration. IFN (IFN-β and -λ) mRNA expression and production were analyzed by RT-PCR and ELISA respectively. To study downstream signalling pathways RT-PCR was carried out to assess the induction of ISGs. Exogenous IFN-β and -λ (100 pg/ml) was added before and after virus infection. Cell viability was assessed by flow cytometry and LDH assay. Levels of pro-inflammatory cytokines (Gro-α, IL-6, IL-8, IP-10, MCP-1, RANTES) were measured by Multiplex Suspension Array Technique.

Results: Viral replication was significantly increased in airway epithelial CF compared to non-CF cells after infection. Examination of innate immune responses revealed profound impairment of virus-induced IFN production and ISG expression in CF airway cells. The addition of exogenous IFNs significantly reduced virus replication in infected CF cells by up-regulating IFN production and ISG expression. All respiratory viruses induced significantly increased cytokine releases, in both CF and non-CF airway epithelial cells. However, there was a marked trend towards lower cytokine production associated with increased cell death in CF cells.
Conclusion: Cystic fibrosis airway epithelial cells have a deficient innate immune response to viral infection, characterized by an impaired IFN production and ISG expression resulting in increased viral replication. Our results regarding the inflammatory response indicate a diminished inflammatory response to viruses in the CF airway epithelium associated with increased cell death. Addition of exogenous IFNs, in particular of IFN-β, restores anti-viral pathways and virus control in CF cells. These mechanisms may contribute to the increased respiratory morbidity after virus infection in CF and suggest the potential use of IFNs in the treatment of virus-induced CF exacerbations.

YR-O 9

Ectopic lymphoid tissues develop in normal and Leishmania-infected Il7 transgenic mice

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Introduction: Antimicrobial adaptive immune responses are generated in secondary lymphoid organs such as spleen, lymph nodes (LNs) and Peyer's patches (PPs). In addition, inducible ectopic lymphoid tissues form in inflammatory lesions of chronic infections, autoimmune diseases, allergic reactions and chronic graft rejection. These so-called "tertiary lymphoid organs (TLOs)" can function as inductive sites for adaptive immune responses against self and non-self antigens.During embryogenesis, the development of secondary lymphoid tissues is regulated by the interactions of $IL7R^{+}CD4^{+}CD3^{-}$ lymphoid tissue inducer (LTi) cells with mesenchymal VCAM-1-expressing stromal cells. Interleukin 7 (Il7) is an essential survival factor for LTi cells. In this study we used Il7 transgenic (tg) mice and Leishmania major infection as a model to investigate the role of LTi cells in the formation of TLOs.
Methods: The effect of transgene expression on immune cells and lymphoid tissue formation was measured by flow cytometry and immunohistochemistry.
Results: Il7 overexpression in mice leads to the spontaneous formation of TLOs in various non-lymphoid organs such as salivary gland and lung. The presence of LTi cells, naïve follicular B, T and dendritic cells in TLOs correlates with the specific expression of chemokines. Using various knockout mice we show that the development and the organization of TLO require the presence of lymphotoxin and LTi cells. Finally, the infection with Leishmania major triggers the *de novo* formation of ectopic lymph nodes in Il7 tg but not in WT mice.
Conclusions: Altogether, we show that Il7 regulates the development of TLO via LTi cells and secondly, that Il7, LTi cells and inflammation collaborate in *de novo* formation of ectopic LNs. It will be important to investigate whether targeting of LTi cells can modify chronic inflammation and TLO developmentin humans.

YR-O 10

Individual caspase-10 isoforms play distinct and opposing roles in the initiation of death receptor-mediated tumour cell apoptosis

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Introduction: The cysteine protease caspase-8 is an essential executioner of the death receptor apoptotic pathway. The physiological function of its homologue caspase-10 remains poorly understood, and the ability of caspase-10 to substitute for caspase-8 in the death receptor apoptotic pathway is still controversial. Here we analysed the particular contribution of caspase-10 isoforms to death receptor-mediated apoptosis in neuroblastoma cells characterized by their resistance to death receptor signalling.
Results: Silencing of caspase-8 in TRAIL-sensitive neuroblastoma cells resulted in complete resistance to TRAIL, which could be reverted by overexpression of caspase-10A or caspase-10D. Overexpression experiments in various caspase-8 expressing tumour cells also demonstrated that caspase-10A and caspase-10D isoforms strongly increased TRAIL and FasL sensitivity, whereas caspase-10B or caspase-10G had no effect or were weakly anti-apoptotic. Further investigations revealed that the unique C-terminal end of caspase-10B was responsible for its degradation by the ubiquitin-proteasome pathway and for its lack of pro-apoptotic activity compared to caspase-10A and caspase-10D.
Conclusion: These data highlight in several tumour cell types, a differential pro- or anti-apoptotic role for the distinct caspase-10 isoforms in death receptor signalling, which may be relevant for fine tuning of apoptosis initiation.

Posters with short oral communication SSP/SGP

PED-P 1

Pancreatic fat fraction is increased in obese adolescents and associated with metabolic syndrome

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Aims of the study: Little is known about pancreatic fat accumulation and its possible associations with metabolic syndrome (MetS) and glucose metabolism. The aim of this study was to quantify pancreatic fat fraction (PFF) in lean and obese adolescents and explore its relationship with metabolic parameters.
Methods: This case-control study was conducted in a tertiary care center, where 25 lean and 24 obese adolescents were recruited. PFF and visceral fat (VAT) were determined using MRI. We measured blood pressure, fasting glucose, insulin, liver enzymes, leptin and lipids levels. Obese subjects underwent an oral glucose tolerance test.
Results: PFF was significantly higher in obese than lean subjects (4.8 ± 1.2 vs. 3.6 ± 0.9; $p < .001$) and was associated with VAT, gamma-GT, triglycerides, HDL-cholesterol, leptin concentrations, and MetS ($p < 0.05$ for all). None of the obese subjects had glucose intolerance, but when adjusted for VAT, the following 3 parameters correlated negatively with PFF: fasting, 30 and 120-minute insulin levels. We divided subjects into 3 groups: I) lean without MetS; II) obese without MetS and III) obese with MetS and observed that PFF increased gradually among groups (I: 3.56 ± 0.88; II: 4.70 ± 1.06; III: 5.34 ± 1.49 %; $p < .001$).
Conclusion: Obese adolescents accumulate fat in the pancreas. PFF correlates with the presence of MetS. Even in absence of glucose intolerance, pancreatic fat deposition is associated with impaired insulin response to glucose overload. This suggests that beta-cell dysfunction is already present in non-diabetic obese adolescents, mirroring what has been shown in adults, and that pancreatic fat accumulation may participate in obesity-associated pancreatic endocrine dysfunction.

PED-P 2

Analysis of a program for risk and adverse event management used in a unit of pediatric hematology-oncology

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Introduction: The Medical Surgical Department of Pediatrics at the University Hospital CHUV started in 2005 a program for management of risks and adverse events called ProGREI. It consists of a database where adverse events observed in the department are reported and registered. The purpose of the current study is to analyze after 4 years

of use the incidents that came up in the pediatric hematology-oncology unit.
Methods: Announced events recorded between November 2005 and 2009 were analyzed. The evaluation of the listed adverse events was performed by characterization of medical errors based on the taxonomy of the NCC MERP (National Coordinating Council for Medication Errors Reporting and Prevention). The most relevant data were analyzed by descriptive statistics.
Results: There were 113 adverse events registered, among which 87 declared as drug errors in an in-patient setting. Thirty-seven out of 87 (43%) arose during drug administration, 14/87 (16%) during transcription of medical orders and 10/87 (11%) during medical prescription. By order of frequency, errors of dosing were in the first position with 27/87 (31%) cases, followed by omission of drug doses in 21/87 (24%), and by either wrong infusion rate in 9/87 (10%) or wrong technique of drug administration in 7/87 (8%) cases. The clinical consequences were mild in 73/87 (84%) cases and moderate in 9/87 (10%). There were no severe consequences and for 5/87 (6%), the gravity of the error could not be determined. The declaration of errors was performed by nurses in 79/87 (91%) and by doctors in 12/87 (14%) cases. Parents were informed about the drug errors in 30%. The announcers kept their anonymity only in 40% of declarations.
Conclusion: The majority of the announced incidents are mostly minor drug errors which are by definition preventable. The majority of events concern dose errors and dose omission which occur during the administration or during drug transcription. Thus, the computerization of the drug treatments seems mandatory to avoid errors of prescription and transcription There is need for a better training of the doctors in the declaration of errors committed and the information to be given to the families.

PED-P 3

10 years follow-up in a series of children suspected or with confirmed child abuse

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Introduction: A 1999 epidemiological study of pediatric child abuse in the County of Vaud identified 17 pediatricians following 30 children with confirmed or having a significant risk of being abused. A new study was performed 10 years later to review and update the data, assess the children's outcome as well as the pediatricians experience with child abuse.
Methods: Data collection was done by a medical student for her master's degree. Review of medical records, interviews and telephone calls with involved pediatricians were performed. Initial children's demographics, risk factors and type of child abuse, socio-economic context as well as legal issues were updated. Two semi-directive discussion scaffolds were used for pediatricians' interviews.
Results: Follow-up information was obtained for 19 of the 30 children initially involved in the first study. A significant improvement of the situation was noted in 56% of the cases. Stable social and family contexts, as well as child abuse declared by the mother were associated with a favorable outcome. A familial conflict, frequent moves or social instability were aggravating factors. 13 pediatricians accepted the interviews. Once child abuse was suspected and revealed, the parents-doctor relationship was interrupted only in two cases. Following child abuse declaration, the main obstacles to adequate care by the pediatricians were a lack of feed-back information and poor collaboration with the other institutions involved with the cases.
Conclusions: The parents' doctor relationship is rarely lost following declaration of a suspected case of child abuse. Pediatricians have a pivotal position in detecting and treating children exposed to violence or abuse. Improved communication between the various institutions involved in the management of child abuse is necessary. A favorable outcome in more than half of the cases supported the time and efforts shared by the various care givers.

PED-P 4

Acceptance of two liquid vitamin D_3 formulations among Swiss mothers with newborn infants: a randomized, single-blind trial

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Introduction: Swiss infants are prescribed vitamin D_3 during the first year of life. For this purpose liquid preparations are traditionally used, which contain vitamin D_3 dissolved in alcohol. Unfortunately, many families do not adhere to the recommendation.
Subjects and methods: The acceptance of the traditional vitamin D_3 preparation (alcoholic vitamin D_3) was compared with that of a new preparation, which contains vitamin D_3 dissolved in medium-chain triglycerides (oily vitamin D_3) in 42 healthy newborn infants (20 girls and 22 boys) aged between 2 and 7 days. Their neonatal body weight ranged between 2.225 and 4.150 kg and the gestational age between $36^{1/7}$ and $41^{3/7}$ weeks. The blinded mothers rated the facial reaction of their children by pointing on a facial hedonic scale. The comparison was not commercially supported.
Results: Thirty-eight of the 41 mothers, who brought the comparison to completion, assigned a better score to the oily vitamin D_3 with no difference in the remaining three cases ($P < 0.0001$). The acceptance for the oily preparation was significantly better both among mothers whose baby was initially presented the alcoholic preparation and among mothers whose baby was initially presented the oily preparation. Furthermore, the acceptance for the oily preparation was better irrespective of gender of infant or parity of mother.
Conclusion: In children, appreciating the preference for drug preparations is crucial. From the perspective of mothers, Swiss newborns infants prefer the taste of the oily vitamin D_3 preparation over the alcoholic preparation.

PED-P 5

Infant lung function and subsequent respiratory morbidity during the first year of life in preterm infants

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Introduction: Compared to term-born infants, preterms reveal more respiratory symptoms and alterations in lung function. We aimed to investigate whether in preterms lung function parameters obtained near term correlate with respiratory morbidity during the first year of life.
Methods: Lung function (tidal breathing flow-volume loops, multiple-breath washout) was measured using an ultrasonic flowmeter in a cohort of N = 263 unsedated preterm infants at mean ± SD 44.9 ± 0.2 weeks of gestational age during natural sleep according to latest standards. The cohort included preterm infants (28% without, 22% with mild, 32% moderate, 16% severe BPD) selected randomly mainly due to logistic constraints. We retrospectively assessed respiratory morbidity in the first year of life (wheeze, rehospitalisation, inhalation and oxygen therapy) using a standardized questionnaire (recall rate 70%). We computed uni- and multivariable logistic regression adjusted for known confounders.
Results: We found sporadic associations between lung function parameters and outcomes that reached statistical significance, but no clear overall relationship. The strongest association was found for TPTEF/TE e.g. with inhalation therapy: OR 0.92 CI 0.87–0.98.
Conclusions: In this cohort of preterm infants lung function tests performed without sedation do not result in additional benefit for the prediction of respiratory morbidity during the first year of life. Reasons for that might be the pre-selected cohort, the limitations of retrospective assessment of symptoms or the large variability of the lung function measurements reflecting the clinical status.

PED-P 6

The Swiss Paediatric Renal Registry (SPRR): 1970–2010

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Purpose: End-stage renal failure in children is a rare but severe condition. In order to optimize management and surveillance of long-term follow-up, central registration of these patients is essential. We describe the Swiss Paediatric Renal Registry (SPRR).
Methods: Data collection started in 1970 at the University Children's Hospital Zurich and was extended to form the SPRR in 1980. Since 2008, the database is located at the Institute of Social and Preventive Medicine in Berne. In 2009 collaboration was established with the European registry of the European Society for Paediatric Nephrology (ESPN). Included were children and adolescents <16 years undergoing dialysis for more than 3 months and/or renal transplantation (TPL). Main data collected were primary renal disease,

concomitant diseases, demographic data, laboratory values, treatment and long-term outcome. Data were annually collected by a central data manager visiting all centres.
Results: From 1970–2008, 432 patients were registered (11.4 patients per year). Most frequent primary diseases were renal dysplasia (16%), hypoplasia (10%), focal segmental glomerulosclerosis (10%), nephronophthisis (8%), other hereditary nephropathies (8%) and hemolytic uremic syndrome (7%). Treatment consisted of (1) dialysis (564 times), (2) TPL (354 patients; living donation: n = 241 (68%), cadaveric donation: n =113 (32%); preemptive TPL: n = 60 (14% of all TPL)). Seventy-one of all registered patients (16%) died.
Conclusion: This national registry is a valuable and important resource for national surveillance of incidence, treatment and long-term outcome in children and adolescents with end stage renal failure. Furthermore provision is given to compare data with other countries and to collaborate in international research projects.

PED-P 7

Adrenal cortex insufficiency in a child due to inhaled corticosteroids

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We present the case of a 6-year-old girl with partial adrenal insufficiency due to long-term asthma treatment with inhaled corticosteroids (ICS). Because of recurrent wheezing episodes and increased specific airway resistance by interruptor technique (RINT), long-term preventive therapy with Montelucast and inhalation of Salmeterol and Fluticason was initiated elsewhere below the age of one year. Recurrent wheezing episodes made a stepwise increase of ICS up to 500 µg Fluticasone daily (= 45 µg/kg/d, 943 µg/m²) necessary over a period of 2 years. The first suspicious clinical sign occurred months earlier with hypoglycaemic seizures. After several episodes of weakness of unclear origin, the girl was seen in our endocrinologic clinic for further investigation. The 6 year old girl (weight 12.3 kg, P3, length 101 cm, <P3) was in good general health status. Routine blood work up was normal. Only serum-cortisol was very low taken at noon with also normal ACTH. Cortisol in 24 h urine was <60 nmol/24 h. During ongoing investigation, the girl complained of abdominal pain, vomiting and fatigue, suggestive signs of an Addison crisis. She was admitted to the hospital. After substitution therapy with corticosteroids the patient's adynamia improved and blood glucose was within normal limits. MRI of the head ruled out pathologies of the pituitary gland. Six days later the girl was discharged with hydrocortisone 13.5 mg/m²/day. Short acting inhaled beta-mimetics on demand was initiated as asthma treatment. The oral substitution with hydrocortisone will be reduced carefully to allow restitution of the adrenal function. In summary, insufficiency of the adrenal gland due to inhaled corticosteroids is a rare but severe complication in asthma treatment. Our patient received Fluticasone at a dose of at least 500 µg/day for 2 years. As weight and length were ≤P3, relatively high doses of ICS (943 µg/m²) were uncritically administered. The use of cortisol measurement in routine follow-up in children with long-term and high-dose ICS should be discussed. If preventive treatment with ICS is needed, this should be prescribed for a limited time of 6–12 weeks only. In case of longer-term treatment, dose of ICS has to be decreased stepwise at regular control visits to the lowest level needed for adequate asthma control.

PED-P 8

Optimization of diagnosis of urinary tract infection in children

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Introduction: Urinary tract infections (UTI) are among the most frequent bacterial infections and a frequent cause of fever in children. Urine sampling technique and, most importantly, interpretation of immediately available urine leukocyte numbers are critical for diagnosis and initiation of empiric antibiotic treatment, while awaiting confirmatory urine culture results.
Methods: Review of bacterial culture and leukocytes count results of consecutive urine samples from children evaluated for UTI in our Emergency Department over a 5-months period. UTI was defined as bacterial growth of ≥10000 cfu/ml in urine culture plus decision by the caring physician for and completion of antibiotic treatment for UTI. Results plus sampling technique were compared in children with and without UTI.
Results: Among 275 children (166 girls) with a median age of 10 months (range, 7 days – 17 years) a UTI was present in 65 children (24%). Overall, urine was collected by catheterization in 169 (58%) and by clean-catch in 106 children. Urine cultures yielded growth of ≥10000 cfu/ml in 29%, growth of >2 species or <10000 cfu/ml, thus suggesting contamination in 34%, and no growth in 36%. Sterile cultures were found in 48% of catheter urines and 17% of clean-catch urines (p <.001), growth compatible with contamination in 18% and 58% (p <.001), and growth of ≥10000 cfu/ml in 33% and 25% (p = .18). The specificity of leukocyturia (defined as >10 cells/ml) in children >4 weeks of age was 64% (catheter 73%, clean-catch 53%). Increasing the threshold for leukocytes to 50 cells/ml resulted in a specificity of 88% (catheter 94%, clean-catch 81%). Conversely, the sensitivity was >95% independently of a leukocyte threshold value of 10 or 50 cells/ml and the urine collection method.
Conclusion: The reliability of microbiological UTI diagnosis was significantly higher investigating urine collected by catheterization than by clean-catch, and setting the threshold for leukocyturia at >50 cells/µl increased UTI diagnosis specificity without loosing sensitivity. Thus, urine collection by catheterization and increasing the threshold value for leukocyturia optimizes diagnosis of UTI in children and has the potential to reduce unnecessary antimicrobial treatment and follow-up investigations.

PED-P 9

Growth response and insulin sensitivity in children born SGA under 4 years of GH treatment

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Background: GH treatment is well accepted for children born small for gestational age who fail catch-up growth. As GH reduces insulin sensitivity, and low birth weight is associated with increased risk of diabetes type 2, the effect of GH on glucose metabolism is of concern in these patients.
Objective: We evaluated glucose tolerance and insulin sensitivity during 4 years of GH treatment in SGA children.
Patients/ methods: In 20 SGA children treated with GH (33 µg/kg/d) an oral glucose tolerance test was performed at baseline and yearly for 4 years. Insulin sensitivity was calculated using the homeostasis model assessment (HOMA) and the insulin sensitivity index (ISI).

	Pre-GH	1 year	2 years	3 years	4 years
Age	7.9 (4.5–14.3)	9 (5.6–15.3)	10.1 (6.6–16.3)	11 (7.6–17.3)	12.1 (8.9–18.4)
Height SDS	–3.24 (-4.2 to -2.31)	–2.44 *** (-3.13 to – 1.46)	–2.02 *** (-3.11 to – 0.8)	–1.61 *** (-2.59 to -0.4)	–1.24 *** (-2.38 – 1.12)
BMI SDS	–1.67 (–4.9 – 0.49)	–1.68 (–4.6 – 1.06)	–1.5 (–4.47 – 1.74)	–1.24 * (-3.67 – 1.97)	–1.2 ** (-3.9 – 1.09)
Fasting glucose (mg/dl)	78 (66–91)	83.5 * (72–99)	83.9 * (69–104)	87.3 *** (73–101)	86.1 *** (76–94)
Glucose 120min (mg/dl)	95.5 (50–140)	103.2 (67–141)	105.4 (75–141)	102.8 (55–167)	105.4 (59–140)
HbA1c (%)	5.4 (5–5.9)	5.47 (4.9–5.9)	5.44 (5.1–6.1)	5.51 * (5.1–5.9)	5.54 * (5.1–5.9)
HOMA	0.92 (0.26–3.16)	1.46 (0.38–4.24)	2.07 (0.54–7.1)	2.01 ** (0.58–6.46)	2.66 *** (1.13–9.82)
ISI	13.5 (4.5–43.5)	7.01 *** (1.61–14.6)	6.9 * (1.11–20.7)	6.14 ** (0.97–24.3)	4.53 *** (1.38–8.62)

Results: (Significance vs baseline: * p <0.05, ** p <0.01, *** p <0.005) A significant decrease of insulin sensitivity was measured after one year of GH treatment and persisted thereafter. Glucose tolerance became borderline at most in only few cases.
Conclusion: In SGA children treated with GH, insulin sensitivity decreases rapidly but fortunately it does not worsen with increasing treatment duration.

PED-P 10

Skin disorders encountered in a Swiss pediatric emergency department

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Background: Skin disorders are commonly seen in pediatric emergency departments (PED) and may cause diagnostic difficulties. We aimed to evaluate the frequency and spectrum of skin disorders in an urban PED at a Swiss University Hospital and the need for dermatological advice.
Methods: The medical records of all patients attending the PED

during the first 10 days of each month over a 1-year period (total 120 days) were retrospectively analyzed. The dermatological findings were evaluated regarding their incidence, patient age and gender, way of referral, diagnosis, frequency of dermatology review, treatment and hospitalization rate. A questionnaire completed by the PED doctors additionally assessed the need for dermatology support.
Results: In 1572 (17.4%) of the 9041 attendances skin findings were recorded. The skin problem was the primary cause for the PED visit or related to the primary complaint in 853 (54.3%) and 335 (21.3%) cases, respectively. In 373 (23.8%) patients a diagnosis was only suspected or remained unknown. Inflammatory and allergic disorders were most commonly encountered (42.9%), followed by skin infections (31.8%), physically induced skin lesions (11.9%) and congenital disorders (2.3%). Viral exanthema was the most common diagnosis (13.1%), followed by anogenital dermatitis (7.7%), gingivostomatitis (7.1%), petechiae (6.4%), burns (6.0%), urticaria (5.0%) and insect bite reactions (5.0%). Eighty-one (5.2%) of the patients with skin conditions were hospitalized. The PED doctors estimated that they wished for dermatological advice in 25% of the patients with skin findings.
Conclusions: This study shows a high frequency, broad spectrum and diagnostic difficulties of pediatric emergency skin conditions. We highlight the need for improved educational measures and a closer collaboration between the two specialities for the benefit of these children.

PED-P 11

Haemoglobinopathies in Cambodia

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Background: Thalassemia and other hemoglobinopathies are a common cause for anaemia in Southeast Asia. As in many developing countries insufficient data exist about the prevalence and morbidity caused by haemoglobinopathies in Cambodia. Since 1991, with the help of donations from Switzerland and other countries along with the support of Cambodian authorities Kantha Bopha hospitals provide specialised paediatric care to a majority of Cambodian children. In order to validate the laboratory diagnosis and to generate data about the presentation of haemoglobinopathies, we analyzed blood samples from patients admitted to the Kantha Bopha hospitals.
Methods: Initial screening for anaemia and haemoglobiopathies was performed locally (Kantha Bopha and Pasteur Institute, Phnom Penh). Blood samples on filterpaper were shipped to our laboratory.
We screened for the three most common Southeast Asian α-thalassaemias and, for the presence of β-thalassemia we sequenced the β-globin-gene.
Results: Between March and July 2010 we analyzed samples from 53 children. While in three of 15 cases compound Haemoglobin E-Thalassaemia was misdiagnosed for heterozygote Hb E, the majority of beta-globin haemoglobinopathies was diagnosed accurately. α globin mutations were found in 10 children (5 children with HbH disease and 5 samples with a single heterozygeous α globin mutation) In 12 children (and in nearly 45% of compound HbE-Thalassaemias) a Hb-level below 5 g/dl was found.
Discussion: Severe anaemia was found in a signifcant proportion of children. Although an accurate diagnostic work up was performed compound thalassaemia-HbE were missed in a small proportion. No β-thalassemia maior was found and it is unclear if these children do not survive. In addition a relatively low incidence of α-thalassaemia was found with a few children with HbH disease presenting with a rather mild anaemia. Further studies are required to define the prevalence of haemoglobinopathies in Cambodia, to find treatments for severely anaemic children with compound HbE-thalassaemia and to better characterize the aethiology of anaemia in children without haemoglobinopathies.

PED-P 12

Therapeutic hypothermia in term infants with perinatal encephalopathy: the last 5 years in Switzerland

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Background: Therapeutic hypothermia (TH) following perinatal hypoxic-ischemic encephalopathy in term or near-term infants improves mortality and neurodevelopmental outcome (1). In Europe most neonatal units perform active cooling using cooling devices such as water mattresses, in Switzerland, however, passive cooling is predominantly used.
Aims: To evaluate the temperature variation between different cooling methods used in Switzerland and to assess how neuromonitoring is provided compared to other European neonatal centres.
Methods: All notes of infants with perinatal encephalopathy who underwent TH between March 2005 and August 2010 were retrospectively reviewed. Active cooling techniques included cooling devices such as cooling matresses or cooling wraps. Passive cooling technique was defined if natural cooling was performed by turning off all heating systems. Passive cooling technique in combination with gel packs or ice was defined if in addition to natural cooling gel or ice packs were added.
Results: A total of 150 infants with median (range) gestational age of 39 (35–42) and median birth weight of 3269 (1990–4920 g) were cooled. 109 (73.6%) infants were outborn and 28 (18.9%) infants died due to perinatal asphyxia. Active cooling was performed in 27 (18.2%) infants, 87 (58.8%) infants were cooled passively in combination with gel packs or ice and 34 (23%) infants were cooled passively. There was a significant difference between the three cooling methods in terms of temperature variation over time. Passive cooling had a significant higher variation of temperature (SD of 0.88) than both passive and cold packs (SD of 0.78) and active cooling (SD of 0.73). aEEG before TH was obtained in 35.8% and 86.5% had a full EEG. At least one cranial ultrasound was performed in 95.3% and MRI was performed in 62.2% of the infants.
Conclusion: Target temperature can be achieved with all three cooling methods, however passive cooling seems to have the highest variation of temperature. Continuous neuromonitoring should be improved.

1 Edwards AD, BMJ 2010; 340:c363

PED-P 13

Tetanus vaccination status in refugee children in Geneva, Switzerland

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Background: data on tetanus vaccination coverage in recently arrived refugee children in Switzerland are missing, and are essential to formulate vaccination strategies.
Materials and methods: we retrospectively collected data from refugee children followed in our hospital for general care. Tetanus antibodies were measured by ELISA either at arrival or one month after booster vaccination, according to local recommendations.

Table 1
Number of patients according to tetanus antibody titters after booster vaccination and length of immunological protection.

Age (years)	Number of patients	<500 U/l	500–1000 U/l	1000–10 000 U/l	>10 000 U/l	Immediate vaccination needed	Protection <5 years	Protection 5–10 years	Protection >10 years
<2	2	0	0	1	1	0	1	1	0
2–5	12	0	0	3	9	0	2	8	2
5–10	19	1	1	6	11	2	1	14	2
10–16	22	0	0	8	14	0	0	12	10

Results: 92 children were evaluated between January 2009 and May 2010. Most were from Eastern Europe and Africa. According to reported history, only 30% were up-to-date with their vaccination at arrival, and only 4% had available vaccination records. Antibody levels against tetanus were measured at baseline in 13 patients, and after booster vaccination in 55 children. Few patients (6% of the total children, 15% at baseline, and 3.6% post-booster) were insufficiently immunized despite prior reported tetanus vaccination in their own country. Measuring antibody levels after booster vaccination saved children, in average, from two unnecessary additional vaccines.
Conclusion: Immunologically proven protection against tetanus in refugee children is heterogeneous, but most children in our study are sufficiently immunized. Vaccine-related antibody measurement allows individual tailoring of vaccine schedules and avoids risk of hyperimmunization.

PED-P 14

Medical conditions in immigrant children in Geneva, Switzerland

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Background: Because of poor medical follow-up in their own country, high exposure to infectious diseases, confrontation with war, violence, and exile, refugee children are at higher risk of physical illness and psychological difficulties. We analyzed the medical conditions of immigrant children in Geneva.
Materials and methods:
We retrospectively collected data from recently immigrated children who were followed in our hospital for general pediatric care. Past medical history, physical examination, tuberculin test (Mantoux test, which was followed by chest X-ray and interferon gamma essay if positive), and parasites screening in stool samples were reviewed.
Results: 92 paediatric patients aged 6 months to 16 years old (median 4.7 years) were evaluated between January 2009 and May 2010. Most were from Eastern Europe and Africa. 13 patients (14%) had a positive Mantoux test. Amongst those, 3 were diagnosed with latent tuberculosis and 2 with active tuberculosis. The most frequent other infectious diseases were cutaneous infections (tinea, pediculosis, impetigo, and scabies) in 8.7% of patients and digestive parasitosis in 7.6% of patients (mainly Giardia Lamblia). The main non infectious medical conditions were dental cavities, and psychological difficulties (eating, sleeping and anxious disorders) in respectively 42% and 32% of the patients.
Conclusion: The general physical health condition of refugee children in Switzerland seems acceptable. However, tuberculosis remains highly prevalent in this population. Detection of intestinal parasitosis, dental cavities, and psychological difficulties should be improved.

PED-P 15

Guillain-Barré syndrome and adjuvanted pandemic influenza A (H1N1) 2009 vaccines – a multinational case control study in Europe

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Aim: To investigate a potential association of the 2009 pandemic influenza A (H1N1) with Guillain-Barré syndrome (GBS).
Methods: Five European countries participated in a prospective case-control study between November 2009 and March 2010 using a common protocol and a distributed data model. GBS case status was classified according to the Brighton Collaboration definition. Controls were matched to cases on age, sex, index date and country. Those who received pandemic influenza vaccines in the 42 days before the index date were considered exposed Conditional logistic regression analysis was adjusted for influenza like illness, upper respiratory tract infections (ILI/URTI) and seasonal influenza vaccination. Country-specific data were pooled using a fixed-effects model.
Results: 104 cases were matched to one or more controls. Case recruitment and vaccine coverage varied considerably between countries; the most common vaccines used were adjuvanted (Pandemrix® and Focetria®). The unadjusted pooled risk estimate for all countries was 2.8 (95% CI: 1.3–6.0). After adjustment for ILI/URTI and seasonal influenza vaccination pandemic influenza vaccines were not associated with an increased risk of GBS (OR_{adj}: 1.0, 95% CI: 0.3–2.7).
Conclusion: This study shows that a potential increase in relative risk of GBS after pandemic influenza vaccine is unlikely to exceed 2.7. The unadjusted increase in GBS risk following pandemic influenza vaccines could be fully explained by confounding by ILI/URTI and seasonal influenza vaccination. When assessing the association between pandemic influenza vaccines and GBS, it is therefore important to make statistical adjustments for exposure to ILI/URTI and seasonal influenza vaccination.

PED-P 16

Procalcitonin Guidance To Reduce Antibiotic Treatment Of Children With Lower Respiratory Tract Infection – The Propaed Study

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Abstract: Background: Clinical trials in adults have shown that procalcitonin (PCT) guided antibiotic (AB) treatment of lower respiratory tract infection (LRTI) markedly reduces AB use. We investigated whether this applies to children as well.
Methods: Previously healthy children, 1 mo to 18 yrs, presenting with LRTI to emergency units of two pediatric tertiary care hospitals in Switzerland between 01/2009 and 02/2010 were randomized to AB treatment according to internationally recognized guidelines or PCT guided treatment. In an intention-to-treat complete case analysis, logistic regression was used for AB exposure within 14 days of randomization (primary endpoint) and the combined safety endpoint of complications or disease specific failure, and linear regression for the cumulative AB treatment duration (secondary endpoint). We investigated effect modification according to the underlying case definition.
Results: Of 337 children with LRTI with mean age 3.8 yrs (range 0.1–17.5), 60.2% were hospitalized. The odds ratio (OR) for AB use in the PCT compared to the guideline group was 1.26 (95% CI 0.81–1.95) and for the combined safety endpoint 0.81 (95% CI 0.45–1.46). In patients without community acquired pneumonia (CAP) the OR for AB use under PCT guidance was 2.34 (95% CI 1.24–4.49). There was evidence that PCT guidance had a different effect in CAP patients (OR 0.21; 95% CI 0.07–0.56). The OR in CAP patients was 0.48 (95% CI 0.22–1.03). PCT guidance reduced the mean cumulative duration of AB treatment by 4.11 days (95% CI 3.18–5.03).
Conclusions: PCT guided treatment of LRTI overall was not shown to reduce AB prescription rates but in children with CAP PCT guidance may reduce AB prescriptions. If antibiotics are prescribed, PCT guidance reduces the duration of AB exposure.

PED-P 17

Effects of multiprofessional group therapy on health-related quality of life, mental health status, and eating behaviour in overweight children*

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Background: The high prevalence of obesity in children warrants continuing public health attention. The purpose of this analysis was to show the effects of standardized multiprofessional group therapy on health-related quality of life, mental health status and eating habits in overweight children and adolescents.
Methods and Results: We present here data of 342 children (12.2 ± 2.2 years) of the ongoing national multi-centre cohort study, collected between 03/2009 and 12/2010 in 22 certified programs in Switzerland. The primary outcome was change in BMI-SDS; secondary outcomes were improvements of health related quality of life (HRQOL measured by Kidscreen 52), mental health problems (SDQ) and eating behaviour (AD-EVA) after one year (T2). At T0, HRQOL of overweight children was significantly worse than in healthy Swiss peers except for school environment; notably physical well-being, self-awareness and social acceptance by peers ($p < 0.001$) were impaired. Furthermore, obese children had much higher rates of emotional and behavioral problems at T0 (38.2%) than normative references (10%). Mean change in BMI-SDS from baseline was -0.19 ± 0.36 ($p < 0.001$). Until T2, significant improvements of HRQL were found in physical and mental well-being, moods, positive self-awareness, autonomy and social acceptance by peers ($p < 0.01$). Emotional and behavioural difficulties decreased by half (17.6%). In comparison to the references, study participants showed higher means ($p < 0.01$) for craving and compulsive overeating, emotional eating, and preoccupation with weight and shape which improved significantly during therapy ($p < 0.01$), with the exception of emotional eating. Preclinical eating disorders, e.g. purging to control weight were rare at T0 and T2 and not different from healthy peers.
Conclusion: Mental health and quality of life are often impaired in overweight children and clearly improve during family-based group therapy, as does the degree of adiposity. Improvements in psycho-social functioning are thought to be pre-requisites to sustain a healthy lifestyle and a beneficial weight in the long turn. Health professionals should also take into account mental health care in overweight children and adolescents.

PED-P 18

Epidemiology of Uveitis in Children over a 10 years period

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Introduction: Uveitis is an inflammation of the vascular tunic of the eye (uvea), usually idiopathic, infectious or related to a rheumatologic disease. In children it is a responsible for more than 17% of unilateral legal blindness.
Purpose: To investigate the distribution, clinical features, complications and loss of visual acuity in paediatric uveitis in the French speaking part of Switzerland treated according to a multidisciplinary approach.
Methods: Retrospective cohort study of all patients diagnosed with uveitis under the age of 16 years presenting to two tertiary referral centres in Lausanne (uveitis and paediatric rheumatology clinics), between 2000 and 2009.
Results: 79 children (37 girls) were identified, 62 lived in Switzerland, 11 in Italy. Mean age at first symptoms was 9.0 years (1.5–15.8 years) with a mean follow-up time of 1.9 years (0–8 years). 51 had involvement of both eyes (64.6%). The course was acute in 26.5%, chronic in 51.9%, recurrent in 8.9% and not specified in 12.7%. Anterior uveitis occurred in 40.5%, intermediate in 32.9%, posterior in 25.3% and panuveitis in 6.3%. The three main diagnoses were idiopathic uveitis (34.2%), juvenile idiopathic arthritis-related (22.8%) and toxoplasmosis (15.2%). More than the half of the patients was treated with systemic therapies: corticosteroids 39, methotrexate 17, azathioprine 11, cyclosporine 4 and biotherapies 13. The visual acuity of most patients remained stable (37.5%), or improved (36.3%) during the follow-up. Only 7.5% eyes had a loss of visual acuity (one or more lines) during the follow-up. 26.3% patients presented one or more ocular complications. The two commonest were cataract (12.5%) and ocular hypertension (10%).
Conclusion: Our patients show a better visual outcome than previously published cohorts. Thus, early recognition and prompt and aggressive treatment are essential to reduce the rate of complications and improve the visual outcome. Collaboration between ophthalmologists and rheumatologists enables a better management of systemic treatments and prompt access to immunosuppressive agents and biotherapies.

PED-P 19

Is an early onset of anorexia a factor of poor prognosis?

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Introduction: Young adolescents suffering from anorexia nervosa remain a big challenge for child psychiatrists and pediatricians. An early onset of the disease is sometimes associated with poor prognosis.
Methods: The aim of this study was to evaluate the outcome of female adolescents affected by anorexia nervosa before the age of 16 and treated at the Children's Hospital of Geneva between 1997 and 2006. Standardized interviews based on the Morgan Russell scale were used to evaluate somatic, social and psychological outcome.
Results: Among 63 patients initially enrolled in this study, 33 young women were finally interviewed. No differences regarding the age of onset, the initial body mass index (BMI), the medical complications and the duration of follow-up were found between the patients who were interviewed and those who weren't. The outcome among the 33 patients (average age of 14.4 at diagnosis and at 66 months follow-up) was globally good. No cases of death were registered. However, 21.2% had a poor outcome and nearly 50% of the cohort still experienced psychological problems. Those who had an early onset of anorexia, i.e. before the age of 14, had a significantly better outcome, as well as those who had a higher initial BMI.
Conclusions: Our results emphasize the need for detecting and treating young adolescents with anorexia as fast as possible, before they lose too much weight.

PED-P 20

Air pollution is associated with longer duration of lower respiratory tract infections in healthy infants

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Rationale: There is increasing evidence that short-term exposure to air pollution has a detrimental effect on respiratory health, but data in unselected populations, particularly infants, are scarce.
Objective: To assess the association of air pollution with respiratory symptoms and infections in healthy infants and relate this impact to other known risk factors.
Methods: In a prospective birth cohort of unselected infants, respiratory health status was assessed weekly by telephone interviews during the first year of life. Daily mean levels of PM_{10}, NO_2 and O_3 were obtained from local monitoring stations. The association of the preceding week's pollutant levels with symptom scores was assessed using generalized additive mixed model with an autoregressive component. Timing effects were estimated by stratifying the analysis by trimesters. The analysis was stratified by presence of lower respiratory tract infections to assess the impact of air pollution on prevalence and duration of infectious episodes.
Measurements and Main Results: In 366 infants (19,106 observations), we show that air pollution has a significant association with overall respiratory symptoms, particularly in the first 8 months of life. This association is small compared to the impact of other risk factors and strongest in the week after lower respiratory tract infections. During times of elevated air pollution, there was no higher prevalence of lower respiratory tract infections, but the duration increased by 20%.
Conclusions: There is a significant impact of air pollution on respiratory symptoms, which is relatively small compared to that of other risk factors. Air pollution did especially lead to longer duration of infectious episodes.

Posters

P 1

Characteristics epidemiologic, clinical, paraclinic and etiologic of the nonparalytic frontal axial deviations of the knee in children

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Introduction: The study aimed to analyze the characteristics epidemiologic, clinical, paraclinic and etiologic of the frontal axial deviations of the knee in children.
Material and method: It is about an exploratory study, led January 2008 to December 2008 and concerned the townsmen children from 0 to 15 years.
Results: On 65 children retained for this study, 39 were *infants*, 22 were old 2 years and half at 5 years and 4 5-years-old children to 15 years, with an average age of 2 years 8 months. 43 patients were male (66.2%) and 22 were female (33.8%). The sex-ratio was 1.95. The socio-economic level of the parents was high in 23%, means in 70.8% and low in 6.2%. 43.07% of the children were exposed little to the solar rays. Gen varum was the head with 63.08%, follow-ups by genu valgum, 26.15% and windswept deformity, 10.77%. 34.48% of the children were classified with the 4th degree according to LADITAN and 85.71% at the stage II of BATESON. The deviations were idiopathic in 43.08% of the cases and rachitics in 40% of the cases. The phosphocalcic assessment was frequently abnormal and consisted of a hypocalcemy, a hypophosphoremy, a hypoprotidemy and an alkaline hyperphosphatasemy.
Conclusion: Deviations of the knee, as a sequel to rickets; they are associated with lack of sunlight and, and significant relationship between those deformity and phosphocalcic perturbation.
Key words: Genu varum, genu valgum, etiology.

P 2

Rapidly progressive scoliosis in children: Don't forget to check the eye movements!

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Horizontal gaze palsy with progressive scoliosis (HGPPS) is a rare autosomal recessive disorder characterized by absence of conjugate horizontal eye movements, preservation of vertical gaze and convergence and progressive scoliosis developing in childhood and adolescence. It is caused by mutations in the ROBO3 gene which plays an important role in axon guidance and neuronal migration.
We present a 12-year-old female child who was admitted for corrective surgery for scoliosis. Her neurological examination showed absence of conjugate horizontal eye movements, preserved vertical gaze and convergence. Scoliosis was first noted at the age of 7 years with a progressive course resulting in severe thoraco-lumbar scoliosis. Neuro-ophthalmologic examination revealed small angle esotropia, synergistic convergence, crossed fixation and a low amplitude high frequency pendular nystagmus. T2-W sagittal and axial images (MRI) of the brainstem showed depressed floor of the fourth ventricle, hypoplastic pons, absence of the facial colliculi, anterior and posterior clefts of the pons and medulla oblongata. Somatosensory and motor evoked potential studies demonstrated uncrossed sensory and motor pathways. The patient is homozygous for p.Arg704Pro mutation in the ROBO3 protein. This case highlights the importance of examination of oculomotricity in children with progressive scoliosis. Further on, gene analysis for *ROBO3* mutations is recommended for genetic counseling.

Abstract presented to the EPNS congress.

P 3

Vitamin D is related to pancreatic fat fraction and insulin concentration in obese adolescents

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Aim of the study: The role of vitamin D in the development of type 2 diabetes (T2DM) has been described in adults. Pancreatic fat fraction (PFF) appears to be related to the development of impaired insulin secretion in obese adolescents. Vitamin D is known to be decreased in obese subjects. The aim of our study was to explore the relationship between serum vitamin D levels, PFF, and metabolic parameters in obese and lean adolescents.
Methods: We recruited 25 lean and 24 obese adolescents (mean age 13.6 ± 1.5 yrs). Pancreatic fat fraction (PFF) and visceral adipose tissue (VAT) were determined using MRI. We measured 25-OH vitamin D, fasting glucose, insulin, leptin and lipids levels. Obese subjects underwent an oral glucose tolerance test.
Results: Vitamin D was significantly different between lean and obese subjects (18.5 ± 7.4 vs. 9.9 ± 6.4, $p < 0.001$). As vitamin D was strongly related to VAT, we performed linear and multiple regressions adjusted for this variable. Results showed that vitamin D was only associated with PFF (R^2-change: 0.061, $p = 0.039$). In obese subjects vitamin D was related to fasting insulin (R^2-change: 0.261, $p = 0.007$), HOMA-IR (R^2-change: 0.215, $p = 0.0017$) and inversely to PFF (R^2-change: 0.358, $p = 0.001$). In lean controls Vitamin D was only related to serum ALT (R^2-change: 0.241, $p = 0.011$) but not to PFF or insulin level.
Conclusion: Vitamin D concentration is very low in obese adolescents probably due to its accumulation in visceral adipose tissue. Obese adolescents with the lowest vitamin D concentration show the highest pancreatic fat deposition and the lowest fasting insulin level. These findings may suggest a novel mechanism for the development of obesity-related glucose intolerance.

P 4

The Swiss Paediatric Surveillance Unit *SPSU*: Highlights of 15 years of operation

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Introduction: In 1995 the Swiss Paediatric Surveillance Unit (*SPSU*) was launched jointly by the Swiss Society of Paediatrics (SSP) and the Swiss Federal office of Public Health (SFOPH) to collect nationwide data on rare diseases or rare complications of more common diseases of public health importance in hospitalized children. By joining the International Network of Paediatric Surveillance Units (INoPSU), *SPSU* is also internationally integrated.
Methods: *SPSU* is a network of all Swiss paediatric teaching clinics (currently 33), which participate in study protocols and report cases by a card mailing system. The system is well accepted by Swiss pediatricians as the 100% return rate of initial reporting cards puts into evidence.
Results: The results of 15 years of surveillance by *SPSU* and 18 studies lead more than once to changes in public health policy in Switzerland: 1) In 2003 the results of the surveillance of vitamin K deficiency bleeding in newborns, demonstrated that the guidelines needed to be adapted by implementing a third dose four weeks after birth. 2) Due to the overall low prevalence of congenital toxoplasmosis and the lack of scientific evidence regarding the effectiveness of secondary prophylaxis treatment of pregnant women diagnostic testing during pregnancy was abandoned in 2009. 3) Some *SPSU* studies are targeted at the evaluation of the impact of the national vaccination programs such as the surveillance of congenital rubella and acute flaccid paralysis as a marker for poliomyelitis. 4) A recently completed study showed that 1 in 3000 children younger than 1 year are affected by the shaken baby syndrome, which increased attention to a serious public health problem. 5) The most recent study addresses the prevalence of extendend spectrum beta-lactamase (ESBL) producing gram negative bacteria in children. Preliminary results give rise to the suspicion that infections with ESBL-positive bacteria are predominantly acquired in the community rather than in the hospital setting.
Conclusion: The *SPSU* with its focus on rare and severe pediatric illnesses is an important surveillance system complementary to the mandatory notification system and the voluntary Sentinella system. *SPSU*'s role as an important source of evidence for public health policy in Switzerland has been demonstrated. The collaboration of *SPSU* with INoPSU leads to an exchange of first line information on results of ongoing studies, potentially unnoticed issues, and allows an international benchmarking in evidence based policy making.

P 5

Human umbilical vessels relaxation is impaired in newborns with intrauterine growth restriction

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Introduction: Numerous epidemiological studies have shown that limitation of nutrients or oxygen supply to the fetus, resulting in a small body weight for gestational age, was linked to an increased risk to develop cardiovascular diseases in adulthood. Intrauterine growth restriction (IUGR) affects approximately 8% of all pregnancies and is associated with considerable perinatal mortality and morbidity. The mechanisms implicated in the development of IUGR are not yet elucidated. We previously demonstrated, in a murine model, that a transient exposure to hypoxia during the perinatal period resulted in altered regulation of the pulmonary circulation in adulthood. We postulate that a reduced oxygen supply *in utero* could influence regulatory mechanisms in other vascular beds, like in the umbilical circulation, which present some similarities with the pulmonary circulation.
Hypothesis: Reduced delivery of oxygen and/or nutrients to the fetus in case of placental insufficiency could be associated with functional and/or structural alterations in the umbilical circulation, contributing further to the impairment of the maternal-fetal exchanges and therefore to the development of IUGR.
Objective: We will characterize the modifications occurring in umbilical cord of growth-restricted newborns compared to controls, in order to devise potential novel therapeutic strategies to prevent or limit the development of IUGR.
Methods: Umbilical cords were harvested just after birth. The vasoreactivity of isolated umbilical vessels was tested in organ chambers. Relaxant response to the nitric oxide (NO) donor DEA/NO was tested in preconstricted vessel rings. Expression of protein kinase G (PKG), a mediator of NO-induced relaxation, was evaluated by Western blot.
Results: Growth parameters, placental weight and umbilical cord diameter are significantly decreased in newborns with IUGR compared to controls. Umbilical vessels of growth-restricted infants display a

significantly reduced NO-induced relaxation, as well as a lower PKG protein relative content compared to controls.
Conclusion: Umbilical vessels relaxation is impaired in growth-restricted newborns. The observed defect in PKG protein expression probably contributes to the altered relaxation, and could serve as potential target for further therapeutic interventions.

P 6

Post-streptococcal reactive polymyalgia in a 13-year-old girl: a case report

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Introduction: Polymyalgia due to acute poststreptococcal infection has been rarely described. Tenderness of skeletal muscles without any signs of arthritis can last for several weeks up to months. The prognosis is good and selflimitating without any residuum. Treatment is symptomatic with NSAR, rarely Glucocorticoids.
Clinical Presentation: A-13-year old girl was admitted to our hospital suffering from severe symmetrical myalgia of thighs, shoulder girdle muscles and backpain. She was completely immobilized or walked inclinated. Two weeks earlier she was treated with oral penicillin for 10 days for acute streptococcal tonsillitis. Physical examination showed mild rhinitis and cough without fever. There were no signs of arthritis but marked tenderness of muscles. CRP (28 mg/l) and ESR (46 mm/1h) were slightly elevated, leucocyte count and creatine kinase were normal. To exclude an osteomyelitis, bone fracture or lumbar spondylodiscitis conventional radiologic examinations and MRI were performed which turned out normal. *Borrelia burgdorferi* antibodies were positive for IgM and negative for IgG, therefore doxycycline was given for 10 days. After 2 weeks of therapy our patient showed no significant improvement, the ESR was still elevated (40 mm/1h) and there was no seroconversion (Borrelia-IgM and -IgG negative). Therefore we suspected a poststreptococcal reactive polymyalgia which was confirmed by elevated antistreptolysin O (ASO) titre 326 IU/ml (normal <200 IU/ml). Paracetamol and diclofenac were prescribed. One month later her thighs were less painfull, she could walk again without any help and restarted normal activity. Five months after hospitalization the patient had no muscle pain and showed normal life-activity including sports without any further drug treatment. ESR decreased slightly (39mm/1h) but was still elevated.
Conclusion: Polymyalgia resulting from acute streptococcal infection should be considered as a possible diagnosis in patients with severe chronic muscle pain, persistent elevated ESR and normal values of creatine kinase.

P 7

Acute interstitial nephritis (AIN) due to Diclofenac in an adolescent with acute rheumatic fever

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Introduction: Drug induced AIN is characterized by an idiosyncratic acute renal failure after exposure to certain drugs like NSAID, antibiotics, antiepileptic drugs, diuretics or others. Approximately 85% of AIN are drug induced, whereas the rest is either infectious, idiopathic or due to systemic diseases or malignant infiltration of the kidney. AIN results in an oliguric or non-oliguric renal failure within days to weeks after exposure. The role of steroid treatment in AIN is still controversial.
Case presentation: 13-year-old male adolescent who presented in the emergency room with signs and symptoms of acute rheumatic fever. Fullfilling the Jones criteria the diagnosis was confirmed: two major criteria (endocarditis, polyarthritis) and three minor criteria (fever, increased ESR and CRP, long PR interval). The antistreptolysin O titer was 2160 U/ml and the antistrepto-DNase B titer was 447 U/ml. He subsequently was treated with diclofenac, a cyclo-oxygenase inhibitor, and penicillin. From day 3 of treatment, we observed increasing microscopic hematuria (max. 322 Ec/ul), glucosuria (while normal blood glucose levels), proteinuria and cell casts (white cell and red cell) as well as a sterile leucocyturia (max. 322/ul). Urinary protein/creatinine ratio was increased to 218 mg/mmol (normal <25 mg/mmol), serum creatinine remained normal as arterial blood pressure. Diclofenac was stopped and treatment was switched to prednisolone, which resulted in rapid normalization of the urinary findings within one week.
Conclusion: AIN can be diagnosed by monitoring urine parameters closely. After cessation of the probably causing agent (diclofenac) and steroid treatment the patient showed rapid and complete remission. Alternatively penicillin could provoke AIN but obviously not in our patient. There was no need for biopsy because our patient never showed signs of renal failure.

P 8

Biocard® Celiac-test: a simple and reliable screening test for celiac disease

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Background: Celiac disease (CD) is a genetically determined autoimmune enteropathy characterized by lifelong gluten intolerance. Including silent forms, its prevalence reaches 1/131 in Switzerland but still remains undiagnosed in a large number of patients. The gold standard for diagnosis remains small intestinal biopsies, showing villous atrophy, crypt hyperplasia and intraepithelial lymphocytosis. However, standard serological testing including total IgA and anti-transglutaminase IgA (anti-TTG IgA) is usually performed as first step screening in children with typical symptoms of CD or failure to thrive. The aim of our study was to perform standard serological test and a newly available capillary rapid test (Biocard® Celiac-test) and to compare with small intestinal biopsies (gold standard).
Patients and methods: Fifty-nine patients (median age 6.6y, range 10m – 16.4y; 19 boys, 40 girls) with suspicion of CD (typical symptoms, failure to thrive, positive standard serological test, or decreased total IgA) underwent duodenal biopsies and Biocard® Celiac-test from April 2009 to April 2011.
Results: Small intestinal biopsies from 33 patients (55.9%) showed villous atrophy, confirming CD. All of them had increased standard serological test and 28 (84.8%) had positive Biocard® Celiac-test. Biopsies were normal in 26 patients (44.1%). Eleven of them (42.3%) had increased standard serological test and one of them had a positive Biocard® Celiac-test (3.7%). The Biocard® Celiac-test and standard serological test showed respectively a sensitivity of 84.8% and 100%, specificity of 96.2% and 57.7%, a positive predictive value of 96.6% and 75.0%, and a negative predictive value of 83.3% and 100%, compared to small intestinal biopsies.
Conclusion: Biocard® Celiac-test is more reliable than standard serological test to diagnose CD, with better specificity and positive predictive value. Both tests have similar sensitivity and negative predictive value. Biocard® Celiac-test appears to be a simple, cheap and reliable screening test for CD, in order to determine if small intestinal biopsies are required. It can easily be performed by pediatricians and generalists in private practice, with a very small amount of capillary blood.

P 9

Methotrexate neurotoxicity with stroke-like syndrome – a case report

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Introduction: Methotrexate (MTX) is a folic acid antagonist and as an antimetabolite it is used to treat patients with a variety of autoimmune and neoplastic disorders. It is an important agent used in acute leukaemia to treat CNS disease and prevent CNS relapse. MTX toxicity, especially acute toxicity, is a common side effect and in high-dose MTX i.v. treatments, the antidote folic acid needs to be used to prevent life threatening complications. Recently, triple intrathecal chemotherapy with cytarabine, MTX and prednisolone has been introduced into the German AML-BFM 2004 protocol to prevent CNS relapse. We present the case of a 14 year old girl with high risk acute myeloid leukaemia (AML), FAB M1, who suffered from subacute MTX neurotoxicity 10 days after receiving her fourth dose of intrathecal MTX.
Case report: On the tenth day after receiving her last intrathecal chemotherapy of the AI/2-CDA protocol (MTX, cytarabine and prednisolone), she suddenly developed left-sided motor hemiparesis with brachiofacial emphasis and anarthria. We performed a cranial MRI within two hours after onset of the symptoms. There were no signs of cerebral ischemia, haemorrhage or infectious encephalitis. Doppler sonography of the cranial vessels was normal and no thrombi were detected in echocardiography. An EEG showed focal slowing over the right fronto-parietal lobe. Since subacute MTX neurotoxicity can present as a stroke-like syndrome, we subsequently treated the girl with oral dextromethorphan, a NMDA-receptor antagonist, for a total of 14 days. A second cranial MRI, 29 hours after the first one, showed bihemispheric changes in the diffusion-weighted images typical for MTX neurotoxicity. There was an impressive amelioration of the neurological symptoms within the first 72 hours after the onset and complete resolution within three weeks.
Discussion: Methotrexate neurotoxicity is not an uncommon adverse effect in patients under intensified chemotherapy with high-dose i.v. and/or intrathecal MTX, which can present as stroke-like leukencephalopathy. Dextromethorphan can be an effective treatment of subacute MTX neurotoxicity. This confirms the role of excessive NMDA receptor stimulation in the pathogenesis.

P 10

Lipschutz genital ulceration

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Introduction: The Lipschutz genital ulceration is a rare and little-known disease for the pediatrician and also for the gynecologist.
Clinical presentation: We present the clinical history of a young girl with genital ulcer, without found STDs (Sexually Transmitted Diseases). This patient presents a good evolution with a symptomatic treatment.
Lipschutz disease: This pathology is rare, affecting mainly virgin girls. It can be associated with EBV infection and may be one of the acute symptoms of Salmonella infection. The differential diagnoses include major STDs and aphtous diseases (for example Behçet's disease). The onset is abrupt with pain and fever. The ulcer is a punched-out lesion, with a red border and a fibrino-purulent membrane, more often localized on the internal face of the labium minus. General signs can be present or not.
The evolution is spontaneously favorable and non-recurring. Local disinfection and analgesics are the only treatment necessary, with sometimes antibiotics for the possible secondary infection.
Conclusion: The diagnostic of Lipschutz's disease can be suggested for every genital ulcer, once aphtous diseases and STDs have been eliminated. It is a diagnosis of exclusion, which can be certainly only made a posteriori, when no recurrence can be seen. It's important to keep in mind that the recovery is spontaneous and the treatment only local and symptomatic.

P 11

Choriocarcinoma and foetomaternal hemorrhage: a new case with new questions

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Introduction: Intraplacental choriocarcinoma (IC) is rare, but the most aggressive form of gestational trophoblastic disease (GTD), seldom diagnosed at the time of delivery. We describe a case of neonatal asphyxia associated with a massive feoto-maternal hemorrhage (FMH). Placental examination led to the fortuitous diagnosis of IC.
Case report: A 3450 g full-term boy was born to a 34 years old Greek G2P1 O+ mother known for hypothyroidism and a previous miscarriage. Increased nuchal translucency (NT) in the first trimester motivated chorionic villous sampling (CVS) with normal karyotype. Gravidic cholestasis over the last month required treatment with ursodeoxycholic acid and the induction of labor at 38 6/7 weeks gestation. APGAR score was 2, 2, and 5 at 1, 5 and 10 minutes respectively. Umbilical cord blood gas showed arterial pH 6.89, venous pH 6.93, lactates 14.3 mmol/l, blood analysis revealed Hb 94 g/l, erythroblastosis 4%, Kleinhauer-Betke test 45/1000, no hemolysis. Both infant and mother's metastasis work-up was negative.
Investigations: A 3.2 cm central mass noticed on the sectioned placenta. Immunohistochemistery confirmed IC.
Conclusion: FMH occurred in 37.5% of 40 cases of IC, and is reported to be an early sign of IC, emphasizing the need of a systematic anatomopathological placental examination in case of FMH. Most likely IC incidence is underestimated. Ethiophysiopathology remains unexplained. Could increased NT be a warning sign of IC or GTD? Does CVS play a role for the development of IC? Is gravidic cholestasis associated or fortuitous?

P 12

Breath-holding spells caused by an alcoholic vitamin D_3 preparation in infancy

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A frightening or emotionally upsetting event or a painful experience may induce a blue breath-holding spell. Typically, the child cries out, breathes out, and then stops breathing. Shortly afterward, the skin begins to turn blue, and the child becomes unconscious. After a few seconds, breathing resumes and normal skin color and consciousness return. Breath-holding spells are quite rare before 6 months of age, peak as children enter the twos, and usually disappear by about age five. Swiss infants are prescribed vitamin D_3 during the first 12 months of life. For this purpose liquid preparations are used, which contain vitamin D_3 dissolved either in ethanol 65% or in middle-chain triglycerides. Currently the oily vitamin D_3 preparation is unavailable in Switzerland owing to preparation difficulties: hence, this widely prescribed preparation has been replaced by the alcoholic preparation. Three families with a negative history of breath-holding spells recently noted that their infants (two girls and one boy aged 5, 8 and 10 weeks), who had been initially given the oily vitamin D_3 preparation, stopped breathing, lost consciousness for a short period and the skin began to turn blue immediately after administering the alcoholic vitamin D_3 preparation. We suspected the diagnosis of breath-holding spells caused by burning sensation in the throat induced by ethanol 65% and confirmed the hypothesis by observing characteristic blue breath-holding spells after the administration of the medicine in the office. No breath-holding spells were noted when the medicine was administered diluted with little milk to render it palatable. In children, appreciating the preference for drug preparations is crucial. The present experience with breath-holding spells precipitated by unpleasant gustatory sensations confirms that, from the perspective of infants, the taste of oily vitamin D_3 preparations is considerably better than that of alcoholic vitamin D_3 preparations.

P 13

The lost peanut

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A 3-year-old girl was presented at the Emergency room with fever, cough and loss of weight since 2 months. On auscultation left-sided breath sounds were diminished. Chest X-ray (CXR) revealed a pneumothorax, shift of the mediastinum to the right as well as an infiltrate in the lingula. Pneumonia was diagnosed and considered to be cause of pneumothorax. Thoracotomy tube was inserted and antibiotics given due to hemophilus influenzae cultured in tracheal secretion specimen. Mantoux as well as sweat test results were negative. The girl improved quickly and was discharged after 6 days. Three weeks later, she was readmitted due to dry cough and wheezing unresponsive to inhaled beta-mimetics. On chest auscultation there were again reduced left-sided breath sounds. CXR showed hyperinflation of the left lung with mild shift of heart and mediastinum to the right. On fluoroscopy mediastinal shift during inspiration to the left and on expiration to the right was demonstrated. This is highly suggestive of a foreign body in the left-sided airways. However in a subsequent rigid bronchoscopy no foreign body was detected. As a next diagnostic step chest computer tomography confirmed an intraluminal mass in the distal left main bronchus. Bronchoscopy was repeated, this time by the use of a flexible instrument. A foreign body with a granulomatous surface was seen and removed thereafter by use of the rigid bronchoscope. Examination confirmed a piece of a peanut partially digested and covered with granulation tissue. Children younger than 3–4 years of age are at risk for foreign body aspiration. One third of the aspirated objects are nuts, primarily peanuts. Early diagnosis may be hampered by lack of a clear history of a foreign body aspiration with sudden onset of cough and airway obstruction. Thereafter an asymptomatic interval of several weeks may be followed by complications related to the presence of a foreign body like e.g. fever, cough, hemoptysis, pneumonia and atelectasis. Pneumothorax however is a rare complication of foreign body aspiration.
Conclusion: in young children presenting with respiratory distress never forget the possibility of a foreign body even if history of aspiration is missing. Late phase complications occur due to inflammation and infection related to the presence of a foreign body. Pneumothorax is a rare complication thereof. In difficult cases a bronchoscopy using a flexible instrument in a first step may be helpful for better visualisation followed by removal of the localized object with the help of a rigid bronchoscope.

P 15

Non-traumatic spinal cord ischemia in childhood – clinical manifestation, neuroimaging and outcome

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Introduction: Spinal cord ischemia (SCI) is rare in childhood. There is very few information on clinical presentation and outcome in these children.
Methods: Retrospective analysis of patients from the last 10 years with SCI. Relocation by contact to Swiss centres of neuropediatrics. Data search: age, primary manifestation, risk factors, neuroimaging and outcome. Additional literature search of cases reports.
Results: 14 patients relocated, 8 (5 girls) gave consent to participate. Mean age was 12.5 years (10 to 15 years). 6 presented with paraplegia

preceded by pain in 4, one each with Brown Sequard syndrome and quadriparesis. Sensation levels were thoracolumbar in 7. All reported bladder, 5 bowel dysfunction. Time to maximal symptom was <12 hours in 7/8. Risk factors were surgery, minor trauma, recent infection and thrombophilia. Mean Follow up was 3.3 years. 3 patients each remained wheelchair-dependent and were ambulant without aid. Bladder function recovered fully in 5. Most affected aspects of quality of life were physical and psychical well-being and selfperception.
In all patients MR images showed pencil-like T2-hyperintensity in the sagittal and H-shaped or snake-bite-like lesion (6/8) in the axial views.
Conclusion: SCI in childhood, presenting with pain, paraplegia and bladder dysfunction has high morbidity concerning motor problems and quality of life. A H-shaped or snake-bite-like T2-hyperintensity with the clinical suspicion is highly suggestive for SCI. The Outcome is not related to either age at manifestation nor level of functional lesion. Acute arterial ischemic event in children does not seem to be different in adults, neither have the children less severe clinical pictures, nor recover they better.

P 16

Effects of farming environments on childhood atopy, wheeze, lung function, and exhaled nitric oxide

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Introduction: Whereas studies have consistently shown a protective effect of farming on atopy, results for asthma were heterogeneous so far. Thus, we studied the farming effect on wheeze phenotypes and objective markers like lung function and exhaled nitric oxide and their interrelation with atopy in children.
Methods: The GABRIEL Advanced Studies are cross-sectional multi-phase population-based surveys on the farm effect on asthma and allergic disease in children aged 6 to 12. Detailed data on wheeze, farming exposure, and immunoglobulin E were collected from a random sample of 8,023 children stratified for farming exposure. In a nested case-control design, another random subsample of 895 children stratified for asthma and atopy was drawn for spirometry including bronchodilation and exhaled nitric oxide measurements.
Results: We found evidence for several farm effects: (i) on the prevalence and degree of atopy, (ii) on the prevalence of transient wheeze and (iii) on the prevalence of current wheeze among non-atopics. Among atopics we detected (iv) less elevated exhaled nitric oxide levels and (v) less impaired lung function.
Conclusions: Farming exposure may affect distinct mechanisms during the development of atopic sensitisation and wheeze. The lower prevalence of current wheeze among non-atopic farmers suggests a genuine farming effect independent of atopic sensitisation. Furthermore, farming appears to reverse the effects of eosinophilic inflammation and to normalize airway function among atopics. However, this is functionally too small to result in a clinically relevant reduction of wheeze prevalence in atopics.

P 17

Comparison of online single-breath versus multiple-breath exhaled nitric oxide in children at school entry

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Introduction: Despite requiring less cooperation, there are no standards for online multiple-breath (mb) measurements of exhaled nitric oxide (eNO) with uncontrolled flow rate and no studies comparing them to more difficult online single-breath (sb) eNO measurements in young children.
Methods: Online eNOmb and eNOsb were measured by a chemoluminescence analyzer connected to an ultrasonic flowmeter in n = 75 children of a birth cohort of unselected children (mean ± SD age of 6.1 ± 0.2 years, 43.5% males) and in n = 7 asthmatic children during regular follow-up (mean ± SD age of 12.0 ± 4.0 years, 83.3% males). During measurements, we aimed for 20 tidal breathing manoeuvres for eNOmb and for 3 eNOsb manoeuvres according to current standards. We compared both techniques by standard comparison methods including regression analysis and Bland-Altman plots.
Results: After strict quality control, eNOmb and eNOsb measurements were acceptable in n = 63 and n = 59 children, respectively. Paired data were available for n = 52 children (48.1% males). With $r^2 = 0.94$, slope 1.0, $p < 0.0001$, eNOmb was significantly correlated with eNOsb (mean ± SD 10.3 ± 9.6 ppb) after computing NO output or after extrapolation to an expiratory flow rate of 50 mL/s ($eNOmb_{50}$, mean ± SD 10.9 ± 9.3 ppb), also on a log-log scale. The mean difference between $eNOmb_{50}$ and eNOsb according to standards was –0.5 ppb with upper and lower limits of agreement of 4.1 and –5.2 ppb, respectively.
Conclusions: At school entry, online eNOmb with uncontrolled flow rate is highly correlated with the gold standard of eNOsb measurements controlling for expiratory flow. The wide range of limits of agreement hampers eNOmb use in population-based research. Being less dependent on cooperation at this age, it might be a promising additional tool for the clinical setting to discriminate between disease groups.

P 18

A 11 year old girl with Kawasaki disease and hemolytic anemia after intravenous immune globulin therapy: A case report

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Introduction: Intravenous immunoglobulins (IVIG) is a well-established treatment of Kawasaki disease. The most common adverse events of IVIG therapy are headache, nausea and vomiting, back pain, fever and fatigue. Less frequently (2010: 5 cases in Switzerland) hemolytic anemia is reported after IVIG treatment.
Clinical Presentation: A 11 year old girl was admitted to our emergency unit with a history of fever for 7 days, enanthema, bilateral cervical lymphadenopathy (<1.5 cm), bilateral conjunctivitis and edema of hands and feet. In suspicion of a streptococcal tonsillitis penicillin therapy was initiated (positive antigen test). A normal hemoglobin concentration (126 g/l, normal erythrocyte indices) and elevated inflammatory indexes (leucocyte count 15 G/l, CRP 68 mg/l, ESR 65 mm/1h) were observed. The diagnosis of incomplete Kawasaki disease was established and treated state-of-the-art with aspirin (60 mg/kg/day) and IVIG (single dose of 1.8 g/kg over 12 hours). The penicillin treatment was continued for a total of 10 days. Initially the overall condition rapidly improved and the girl was afebril under treatment. Four days later she presented with recurring fever and three episodes of presyncope. Laboratory evaluation showed signs of hemolytic anemia with a hemoglobin concentration of 76 g/l (normal erythrocyte indices), elevated reticulocyte count (11.4%), spherocytes in the blood smear, anisocytosis (RDW-CV 20.4%), an elevated lactate dehydrogenase concentration and a positive Coombs-test. A spontaneous increase of the hemoglobin level was observed during the following four days. We dismissed the girl in good overall condition after ten days of hospitalisation into outpatient care without need of red cell transfusion.
Conclusion: Normochrome, normocytic anemia might be caused by Kawasaki disease. Hemolytic anemia is also an adverse event of penicillin therapy (most commonly after more than 14 days of treatment). However the temporal correlation between IVIG therapy and occurrence of the anemia is suspicious of an IVIG induced hemolytic anemia, which is a known, but rare adverse effect of this treatment. In our patient Kawasaki disease and penicillin treatment are considered as potential aggravating factors of the IVIG induced hemolytic anemia.

P 19

Paediatric posterior circulation arterial ischemic stroke: manifestation, risk factors, neuroimaging and outcome

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Introduction: Posterior circulation arterial ischemic stroke (PCAIS) accounts for 5–15% of childhood arterial ischemic strokes. Data about pediatric PCAIS is limited.
Methods: Analysis of clinical presentation, risk factors, radiological features and outcome in a prospective series of patients with PCAIS registered in the SNPSR (Swiss Neuropaediatric Stroke Registry) from 2000 to 2010.
Results: 40 children (25 boys) could be included. Mean age at manifestation of stroke was 8.45 years (range 1–16). Most patients presented with ataxia (23), hemiplegia (18), cranial nerve dysfunction (20), nausea and vomiting (16), impaired consciousness (16) and/or headache (15). Mean pediatric NIHSS at presentation was 6.32 (range 1–25). Risk factors, mainly coagulopathy (9), vasculopathy (8) and dissection (7), were identified in majority of patients, multiple risk factors in more than half of them. Cerebellar (18) and thalamic (16) lesions were predominant. Lesions were multiple in one third of patients, four patients had additional anterior circulation lesions.

Posterior cerebral artery was the most frequently affected vessel (21). After a follow-up period of 6 months, outcome was favorable, with 17 of 35 patients having no residual deficits and 19 patients demonstrating deficits such as ataxia (10), hemiplegia (10) and cranial nerve dysfunction (10). After a follow-up period of 2 years, 13 of 20 patients had residual deficits, mean modified Rankin score was 1.3 and mean IQ was 92.
Conclusions: Most pediatric patients with PCAIS have underlying risk factors such as coagulopathy and vasculopathy. Lesions are mainly cerebellar, nevertheless pediatric PCAIS are rarely isolated cerebellar strokes, but rather consist in multiple lesions. Despite severe symptoms at presentation recovery is favorable. Neurological outcome as measured by a modified Rankin score is slightly better than in supratentorial stroke (1.3 versus 2.0).

P 20

Vertical transmission of Hepatitis B despite adequate immunization

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Introduction: Failure of active and passive hepatitis B immunization in a newborn born to a mother with hepatitis B is uncommon (10%) and strongly related to the presence of HB_eAg and circulating HBV DNA in the mother. We here describe a patient with vertically acquired acute hepatitis, despite immunization, in the context of a Langerhans Cell Histiocytosis (LCH), further complicating the initial diagnosis.
Case report: The newborn was born at term to a 28-year-old 1G 0P mother with known hepatitis B (normal liver function tests, HB_sAb neg, HB_eAg neg, HB_eAb pos, HB_cAb pos, HCV and HIV neg), via normal vaginal delivery. The infant received within 12 hours of birth both active and passive immunization, and was discharged home without any problems. She received a second HBV vaccine at one month of age. Soon after discharge, she was also diagnosed with LCH (multiple and characteristic bone involvment). At six months of age, she was admitted with acute jaundice with no other complaints. Physical exam was normal except for jaundice, and a palpable liver (2 cm from the lower rib) and spleen (tip of the spleen). Laboratory analysis showed the following abnormal values: ALT 3339 U/l, ferritin 2156 μg/l, triglycerides 4.39 mmol/l. Because of the very high ferritin level, a diagnosis of hemophagocytic lymphohistiocysis (HLH) was considered but soon ruled out when we received the serological tests showing an acute HBV infection.
Discussion: Despite adequate immunization of infants, about 5–10% of perinatal transmission of HBV can still not be completely eliminated. Several risk factors have been identified, the most important ones being the presence of detectable HBV DNA and HB_e antigenemia in the pregnant mother. Of note, the mother of our patient had undetectable HB_eAg, and the infant received adequate immunization, which contrasts sharply with what has been reported in cases of failed immunization. This case report stresses out the importance of timely active and passive immunization in such cases, as it has been shown that recommendation are not always correctly followed and implemented. If the presence of HB_eAg and/or circulating HBV DNA in the mother requires different immunization recommendation for the child remains to be defined.

P 21

Cyclic vomiting Syndrome – are we really aware of it?

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Background: Cyclic vomiting Syndrome (CVS) is a chronic functional disorder characterized by episodes of recurrent vomiting with symptom-free intervals and typically a stereotypic pattern of episodes within individuals. Since the differential diagnosis of episodic vomiting is large and the entity of CVS is not very well known, time to diagnosis is usually long and effective treatment is delayed.
Objective: To describe the patient population, clinical course, treatment, family history and additional diagnoses in a cohort observed at the Children's Hospital St. Gallen.
Methods: We studied children aged less than 16 years who were diagnosed with CVS after presentation to our gastroenterology or neuropediatric service, emergency or inpatients department from 2004 to 2010. The clinical and treatment patterns were obtained by chart reviews.
Results: Fifteen children were identified and analyzed for this study. The mean age at onset of first episode was 6.7 years (1.5–14 years) and mean age at diagnosis was 9.7 years (2.5–14.5 years). The interval from onset to proper diagnosis was 2.7 years (mean; 0.5–12.5 years). We found a male to female ratio of 1.1 : 1. Frequency of episodes ranges from twice a month to once a year, mostly 3 cycles per year. Mean duration until recovery to oral feeds was 2.4 days (hours–7 days). Precipitating factors were infections (3/15 patients) and psychological/emotional stress (5/15 patients). Besides vomiting, abdominal pain was coexistent in 9 of 15 patients and nausea in 2 of 15 patients. Concomitant neurological symptoms were found in 7 of 16 patients (photo/phonophobia 2/15, headache 5/15). Five of 15 patients had a known family history for migraine or cyclic vomiting. Ten of 15 patients received episodic treatments such as prokinetics, antiemetics, benzodiazepines, ergot alkaloids or antacids, while 6 of 15 patients needed additional interval treatment with propranolol or amitriptyline.
Discussion: The results of this CVS cohort study in Eastern Switzerland were comparable to the findings of international studies. Patient characteristics, delay until diagnosis, precipitating factors and clinical course were similar to previous publications. Proper diagnosis and patient education may reassure families and therefore reduce unnecessary investigations and treatments and improve quality of life. We emphasize that clinicians in Switzerland should be aware of CVS and consider it in the differential diagnosis of recurrent vomiting in children.

P 23

Case report: UBO in the brain and spinal cord MRI in a 5-year-old male patient with NF 1

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Introduction: "Unidentified bright objects" (UBO) on T2-weighted sequences of brain MRI are a frequent radiological sign of NF 1 without related clinical symptoms. UBO in the spinal cord however have not been described in the literature. The clinical and MRI results of a 5 year old boy with NF1 are presented. The main question of this case report is the radiological and clinical significance of UBO in the spinal cord.
Case presentation: At the age of 7 months the patient showed 15 café-au-lait spots. At this time no UBO were detectable in the MRI of the brain. At the age of 2 3/12 years freckling in the axilla, inguina and neck was detectable. At the age of 4 5/12 years a thoracolumbar scoliosis of 20° was noticed. A progression of the thoracic and lumbar spinal scoliosis to 33° was seen in only 5 months duration.
Results: At brain MRI UBO were detectable in several regions of the brain, brainstem, cerebellum and along both optic tracts. A thickening of the pons, the medulla oblongata and the cerebellar peduncle on the right was noted. At spinal cord MRI there was an intramedullary T2w hyperintense lesion exceeding from T6/7 to T10 and a skip lesion was seen at the level of the conus.with a thickening of the spinal cord. Diffuse focal intramedullary T2w hyperintensities were also present in the cervical spinal cord especially at the level of C 6/7. There was no plexiform neurofibroma found. Follow up MRI 6 months later showed regressive cervical spinal cord lesions; the thoracic intramedullary lesions were less hyperintense, but the thickening of the spinal cord was still present. The intracerebral signal alterations were slightly regressive as well. There was no contast enhancement on both brain and spinal cord MRI's. Clinically the boy never showed any signs of cerebral or spinal cord dysfunction.
Conclusion: Focal intracerebral hyperintensities are well known in NF1 and interpreted as UBO. But expanded hyperintense areas in the spinal cord as in this patient are not described in the literature. The clinical significance was unclear, since the patient showed a progressive scoliosis without other neurological signs. The hyperintensities of the spinal cord were regressive after 6 months and no contrast enhancement was seen; the boy still didn't suffer from neurological symptoms therefore we discussed the diagnosis of UBO of the spinal cord; a low grade glioma might be under the differential diagnosis for the thoracic spinal cord lesion.

P 24

Influenza A/H1N1 Encephalopathy with Lesions of the Corpus callosum – A case report

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Introduction: Influenza A/H1N1 was the first pandemic influenza in the 21th century. In most subjects Influenza A/H1N1 presents with illnesses of the respiratory tract, but additional systemic manifestations are also reported. Neurological symptomatology ranges from headache to altered mental status and seizures. Neuroimaging of encephalitic Influenza A/H1N1 subjects reveals cerebral cytotoxic edema, lesions of the thalami and cortical regions or ADEM-like lesions. Encephalitic/encephalopathic manifestations of Influenza A/H1N1 fortunately are a rare complication.
Case: We report on a $5\ ^{4}/_{12}$ year old boy who presented with two episodes of generalized tonic seizures, with 3 days of gait instability

and 24 hours of altered consciousness before hospitalisation. A few hours hours before hospitalisation the child developed mutism. There were no fever and no symptoms or signs of repiratory tract illness. The initial neuropediatric examination revealed a impaired vigilance with a GCS of 11 recovering spontaneously. There were no clinical signs of focal neurological deficits and the boy completely recovered from his neurological symptoms.
Diagnostic findings: Nasopharyngeal sectretions were PCR positive for Influenza A/H1N1. Further analysis revealed an increasing Influenza A antibody titer (1:15 to 1:1280). Cerebrospinal fluid (CSF) showed an elevated IgG/Albumin-Quotient but neither an elevated leukocyte count nor a positive PCR or a positive serology for any virus or bacteria. EEG detected a generalised severe slowing of background activity with full recovery 14 days later. The MRI exhibited two lesions of the corpus callosum (1 in splenium and 1 in genu corporis callosi).
Discussion: Our patient suffered of an Influenza A/H1N1 infection with rapid development of neurological symptoms. Neurological recovery was complete. Encephalopathy without respiratory symptoms with Influenza A/H1N1 infection is rare. Additional unusual findings in our case are the lesions of the corpus callosum. We conclude that Influenza A/H1N1 encephalopathy is an acute but rare disease and may occur without typical influenza symptoms.

P 25

Person-to-person transmission of multidrug-resistant (MDR) *Mycobacterium tuberculosis* is reality in Switzerland

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Background: Worldwide, an increasing number of multidrug-resistant tuberculosis (MDR-TB) diseases have been reported in the last years. In Switzerland, 1.4% of all TB cases are caused by MDR strains. With previous TB-treatment, the proportion of MDR strains is even higher (4%). Either selection of drug resistant strains by inadequate treatment or, in a lesser extent, direct transmission of MDR-TB strains may contribute to this increase. Yet, no MDR-TB person-to-person transmission has been documented in Switzerland.
Case: We report a case of a healthy young female from Tibet, living in Switzerland since 2007. She never had a TB-diagnosis or TB-treatment before. She was in close contact over a period of two weeks with a Tibetan friend with acute respiratory illness. He was hospitalized and diagnosed with smear positive, pulmonary tuberculosis, which turned out to be due to a MDR-TB strain. Contacts of the index case were investigated by TST and IGRA. As the young woman had positive TST and IGRA and minor symptoms with fatigue and cough, she also had a chest X-ray and sputum examination. Chest X-ray and clinical examination were normal and initial sputum microscopy was negative. Three weeks later, sputum culture turned out positive for *M. tuberculosis*. Genotyping revealed resistance to Rifampicin (R) and Isoniazid (H). Antituberculous treatment with Ethambutol (E), Pyrazinamid (Z), Amikacin (Am), Moxifloxacin (Mfx) and Ethionamid (Eto) was started immediately.
Conclusion: The index case and the above patient do not originate from the same community in Tibet. They had close contact in Switzerland while the index patient was symptomatic with open lung tuberculosis. The infected young woman subsequently developed respiratory symptoms. Resistance patterns of both *M. tuberculosis* strains that have been identified were the same and both strains were proven to be identical by RFLP-analysis. Thus, transmission most likely occurred by direct person-to-person transmission. During the investigation among contacts of a case of suspected or proven MDR-TB, the possibility of transmission should be kept in mind. In case of secondary TB, the treatment has to be adapted to the sensitivity of the strain of the source case.

P 26

Neonatal infection or withdrawal syndrome? A case of tetrahydrocannabinol (THC) withdrawal syndrome after concealed maternal cannabis smoking

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Neonatal infection and withdrawal syndrome share common symptoms. In neonates presenting with unspecific symptoms like fever, tachypnea, altered consciousness and irritability, pediatricians should consider the possibility of undeclared maternal abuse and neonatal withdrawal syndrome.
Case report: A 36-hours-old full term male newborn was admitted for fever, tachypnea and irritability. The pregnancy and delivery has been uneventful. A presumptive diagnosis of infection was made, for which he received empiric intravenous antibiotics. However, sepsis screen and blood culture remained negative. In spite of a negative history of drug abuse, except tobacco smoking, a withdrawal syndrome was rapidly suspected based on observations of tremor, exaggerate suction and sudation. Finnigan score at admission in neonatal unit was as high as 16. On further questioning, the mother finally admitted daily smoking of THC during the last 3 months of pregnancy. Drug screening in newborn urine was positive for THC, negative for other drugs. The patient was successfully treated with oral morphine for 4 days. Breast-feeding was continued, but the mother was advised to cease cannabis use. Pedopsychiatric support was offered. Both parents showed very surprised that cannabis could be harmful for their infant.
Discussion: Little is known about short and long-term effect of prenatal exposure to THC, despite the frequency of the problem. It may be linked with long term behavioural troubles and cognitive disorders. Neurobehavioral dysfunction after birth is described, but neonatal withdrawal syndrome needing treatment seems to be uncommon.
Conclusions: 1. Neonatal withdrawal syndrome may occur after chronic use of THC during pregnancy. 2. Effort should be made to identify use and advise regarding the dangers of THC consumption during pregnancy. 3. Pediatricians should be alert to non specific withdrawal signs, even in case of a negative maternal history.

P 27

Hypercortisolemia without hypercortisolism: A classical pitfall in adolescence

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Most of the hormones in the circulating blood are bound to specific binding proteins. Only the free fraction of specific hormones exert endocrine activity. Routine determination of total hormone levels representing the sum of the free and bound fraction may be misleading in clinical situations, when the concentration of binding proteins is altered. A 16-year-old girl was investigated for possible anorexia nervosa by the pediatrician. A blood test at 08.00 a.m. revealed an excessively high cortisol value (1455 nmol/l; N <600). At referral, the patient's history was uneventful. She was performing intensive daily exercise, showed no weakness or documented weight increase in the last months. Menses were regular under oral contraception (OC). Clinical examination did not reveal signs of Cushing's syndrome nor hypertension. At 04.00 p.m. an elevated total cortisol value was confirmed (785 nmol/l, N <400) indicating a residual circadian rhythm but on an elevated level. ACTH was not suppressed and androgenes (androstenedione, dehydroepiandrosterone) were normal. Low-dose dexamethasone test revealed an incomplete inhibition of the hypothalamo-pituitary-adrenal axis, while 24-hour urinary free cortisol excretion was completely normal. OC intake is well known to cause elevated corticosteroid-binding globulin (CBG) leading to isolated increase of total cortisol between 600–1000 nmol/l in healthy women. Nevertheless, total cortisol levels as high as in our patient are rare. Drug-induced hypercortisolemia in patients without hypercortisolism can be diagnosed by measurement of normal levels of free cortisol in blood or urine and increased CBG if discontinuation of OC is not desired.

P 28

Robust prediction of later asthma in symptomatic toddlers: a novel approach

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Aim: Many children have asthma-like symptoms in early life, but few develop asthma. Several models for predicting later asthma in symptomatic toddlers have been built, but some included factors that are difficult to assess, and methods used were prone to overfitting, leading to selection or exaggeration of irrelevant factors. We aimed to identify predictors for later asthma avoiding previous limitations.
Methods: In a population-based cohort, we selected 1–3 year-olds with respiratory symptoms (current wheeze or recurrent cough) and related healthcare visits. Asthma (current wheeze and treatment) was assessed 5 years (N = 1226) and 8 years (N = 866) later. The included factors are easy to assess in clinical practice: family history, symptoms at baseline, demographic and perinatal data. We used lasso penalized logistic regression to select predictors. This minimizes the number of included predictors while maximizing area under ROC curve (AUC).
Results: Main predictors selected in the model for asthma 5 yrs later (AUC = 0.76) were ≥4 wheezing attacks in the past 12 mo (OR = 1.65), wheeze causing breathlessness (3.1) and activity disturbance (2.4), eczema (1.5) and male sex (1.5). Other predictors (OR <1.5) were: non-viral triggers for wheeze or cough, parental history of asthma,

older age at baseline and low birth weight. The results for asthma 8 yrs later (AUC = 0.72) were similar.
Conclusion: Among factors easy to assess in symptomatic toddlers, wheeze severity, eczema and male sex are main predictors of asthma in mid-childhood. Because our approach for variable selection avoids overfitting, the resulting prediction models should perform well with new data. However, external validation is needed.

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P 29

Sexual precocity in our modern society: case report of an eleven month old boy presenting with testosterone intoxication

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We present a situation of an 11 months old boy, born at term, with normal neonatal screening who consulted for signs of male sexual precocity: increased penis size, pubic hair, hyperpigmentation of the scrotum, frequent erections, growth acceleration from 7 month onwards and advanced bone age. Small testicular size pointed to peripheral and not central precocious puberty. Blood test showed very elevated testosterone (16,1 nmol/l, N <0.5 nmol/l) level corresponding to adult values. The parents were little concerned about the precocious puberty of their son. No personal testosterone use was reported. Hormonal and radiologic work up permitted to exclude congenital adrenal hyperplasia of late onset, adrenal and testicular tumor and rarer causes as LH receptor mutation (testotoxicose), Mc Cune Albright syndrome, bHCG producing tumors. In this situation an external source of testosterone was suspected. Finally, a close friend of the family was identified to use an anti aging cream (Receptra Hormone balance®), containing testosterone. The interdiction of close contact with this family friend showed rapid decrease of testosterone level and stabilisation of virilisation signs and growth velocity. The clinical and biological follow up confirmed the testosterone intoxication in this case. Sexual precocity in a boy needs always endocrinological evaluation and tumor exclusion. In a society where sport performance, eternal youth and beauty are promoted testosterone intoxication has to be considered. As testosterone use, doping and antiaging treatment remain a taboo subject with interdicted annotation, patient history can first be negative and erroneous! Paediatricians and adult physicians should be aware of this increasing problem.

P 30

Urogenital schistosomiasis: An unusual cause of macroscopic hematuria in Switzerland

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Background: Hematuria in Swiss children is most frequently caused by urolithiasis, bacterial or viral infections, neoplastic diseases or glomerulonephritis. Worldwide however schistosomiasis is one of the most common reasons of hematuria particularly for individuals from Africa.
Case: An 11-year-old boy from Togo who had immigrated to Switzerland 18 months earlier presented with a history of macroscopic hematuria for the last two years. His hematuria was at the end of micturation and there was no dysuria or fever. Urinalysis confirmed the hematuria with the presence of numerous red blood cells and no evidence of bacterial infection or glomerulonephritis. An ultrasound of the urinary tract detected a thickening of the upper bladder wall, both kidneys were normal. Complete blood analysis showed an eosinophilia of 1.7×10^9/l with normal white blood cell and platelet counts and a normal hemoglobin concentration. Liver and renal parameters were within normal range. Serology was positive for Schistosoma mansoni. A morning spot urine analysis did not reveal the presence of parasites. However a subsequent early-afternoon urine collection over two hours showed eggs of schistosoma haematobium, mansoni and intercalatum. Stool analysis was negative for parasites. Treatment with two doses of Praziquantel (2 × 20 mg/kg) was given. On follow up, one months later, hematuria and esosinophilia had resolved.
Conclusion: Schistosomiasis (or bilharziosis) is an unusual cause of hematuria in Switzerland, but should particularly be considered in African immigrants. Detection of schistosoma eggs is enhanced when urine is collected in the early afternoon. Treatment with Praziquantel, which is currently not licensed in Switzerland, is simple, well tolerated and effective in preventing long-term complications such as bladder cancer.

P 31

Cranial polyneuritis as a rare manifestation of neuroborreliosis in children

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Introduction: Borrelia infection, also known as Lyme disease is a spirochetosis transmitted by deer ticks. In about 15% of patients it involves the central nervous system and is then called neuroborreliosis. Neurologic complications occur more often in Europe than in the US suggesting a greater neurotropism of Borrelia garinii a subspecies prevalent in Europe. Usual manifestations of neuroborreliosis in children include headache, facial nerve palsy and serous meningitis. We present a case with abducens nerve palsy a rare manifestation of neuroborreliosis.
Case presentation: We report the case of a 10 years old girl who was referred to our emergency department with suspicion of neuroborreliosis. She complained about double vision and reported several tick bites that occured about 5 weeks before first presentation of symptoms. The neurological examination showed a palsy of the left abducens nerve. All other cranial nerves were clinically not affected. Blood serum was tested positive for Borrelia IgG and IgM (ELISA and Western blot) and negative for tick-borne encephalitis. A lumbar puncture revealed a pleocytosis (19 mononuclear cells/µl), cerebrospinal fluid with normal protein as well as an intrathecal production of Borrelia antibodies. A cerebral MRI showed signs of a neuritis of the left oculomotor nerve, the left trigeminal nerve as well as both abducens nerves. After treatment with intravenous ceftriaxone for 21days the patient recovered completely within two months.
Conclusion: Neuroborreliosis has to be considered in the differential diagnosis in children at risk for tick bites presenting with cranial nerve palsy. The facial nerve is most frequently affected but also other cranial nerves can be involved and have to be carefully examined. In cerebral MRI multiple enhancing cranial nerves can be seen not always correlating with neurologic symptoms. Despite the higher incidence of neuroborreliosis in children the clinical course is generally milder an shorter than reported for adults and has a good prognosis. It remains unclear whether children older than 8 years with isolated cranial nerve palsy, other than the facial nerve, could also be treated with oral doxycycline.

P 32

Familial cerebral cavernomatosis fortuitously disclosed during parainfluenza virus infection in a child

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Introduction: Cerebral cavernous malformations (CCM) represent 5% to 13% of all vascular malformations. A familial (genetic) form of cerebral CCM is known to affect 10–40% of patients with CCMs. Asymptomatic patients with known familial CCM are commonly encountered. However, fortuitous discovery of familial CCM in an asymptomatic patient is rare. We report the case of an infant in which lesions consistent with multiple CCM were incidentally disclosed, leading to the diagnosis of familial CCM.
Case report: A 8 months-old boy with normal development, presented with fever and bulging fontanel. CSF analysis ruled-out meningo-encephalitis. Parainfluenza virus was detected. The child rapidly improved. Cerebral sonography revealed enlarged ventricles. Fundoscopy demonstrated left retinal hemorrhagic focus without papillary edema. Brain MRI disclosed multiple cortical and subcortical lesions. Signal characteristics, in conjunction with CT findings, were consistent with small hemorrhagic foci. Findings were, therefore, consistent with multiple CCM and retinal cavernomatous malformations (CM). Family history was unremarkable except for cerebral vascular event of unknown etiology in the paternal grandmother. Father's brain MRI and fundoscopy revealed multiple asymptomatic CCM and retinal CM.
Discussion: In this case, both baby and father had multiple CCM which was sufficient to diagnose familial CCM. Demonstrating CCM in the father was important because it offers additional arguments that the lesions in the baby were multiple CCM and for genetic counseling. There is currently no clear consensus concerning follow-up strategies. Some authors recommend regular MRI and others suggest it only in symptomatic cases. This case shows a rare association between CCM and communicating hydrocephalus, suggested by the finding of a subependymal CM responsible for ventricular hemorrhage.
Conclusion: Familial MRI screening should be performed to confirm the diagnosis of familial CCM when lesions consistent with multiple CCM are detected in a child. As demonstrated by the present case, familial CCM can be asymptomatic both in children and in adults.

P 33

Cholelithiasis: obesity complication or orlistat adverse reaction?

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Introduction: Obesity is increasingly prevalent in the pediatric population. Its short and long-term health consequences are significant. Diet combined with physical activity is not always successful and orlistat, a synthetic lipase inhibitor which decreases absorption of dietery fats, is sometimes prescribed in children and adolescents, despite limited safety data in this population.
Case description: A 15 old, obese teenager (105 kg – BMI: 37kg/m^2) was admitted for abdominal pain. She had been treated by orlistat for 3 months with a weight loss of 6 kg. The patient was soon diagnosed with acute cholecystitis with cholelithiasis and operated 48 hours later.
Discussion: Obesity is a known risk factor for lithogenesis. Cholelithiasis prevalence in obese children is 2% compared to 0.1–0.6% in the non-obese. Other known risk factors are rapid weight loss and female sex. Efficacy of orlistat on weight loss is rather limited, but some studies demonstrate that it can decrease gallbladder emptying, which could theoretically promote lithogenesis. Other adult and pediatric cases of cholelithiasis/acute cholecystitis have been reported in patients treated by orlistat during clinical trials and post-marketing surveillance, but causality link could not be confirmed due to confounding factors and lack of pharmaco-epidemiological studies.
Conclusion: Orlistat could increase the risk of cholelithiasis and patients treated with this lipase inhibitor, either prescribed or over the counter, should be carefully monitored. Further data from prospective or case-control studies including pediatric patients are needed to assess this potential risk.

P 34

Chronic recurrent osteoarticular pain

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Introduction: We observed 3 cases of recurrent osteoarticular pain, in a primary care hospital, all diagnosed as Chronic Recurrent Multifocal Osteomyelitis (CRMO).
Case report: Two girls (8 and 9 years old), and one boy aged 9, were admitted to the hospital after several months of osteoarticular recurrent pain with variable symptomatology: local swelling, fever, fatigue, hepatomegaly, splenomegaly and with mild to severe biological inflammation. The boy had palmar and sole aseptic pustulosis. The girls displayed clavicular involvement. One had foot immobilized for suspected stress fracture. Leukemia was excluded by bone marrow analysis in the boy. Blood cultures were sterile. Bone scan and/or total body MRI showed anomalies in metaphyses of tubular bones, clavicles, ribs, patella and pelvis. Antibiotic were ineffective in one child but NSAID dramatically improve all 3. Biphosphonates were administrated later on successfully in one patient. During one year follow-up and despite continuous NSAID, all patients had intermittent periods of improvement and exacerbation of the disease.
Discussion: CRMO has been described in 1978 and is now classified as an auto-inflammatory disease involving multifocal osteoarticular sites as metaphyses of tubular bones, ribs, vertebrae, mandible, pelvis – and also soft tissues. Children over 5 years, adolescents and young adults may be affected. Bone scan or total body MRI are often diagnostic. Biopsies must be reserved in unilocular presentation and bone marrow aspiration when hemopathy is suspected. Histology shows lytic and/or sclerous osteous lesions, sometimes with hyperostosis. NSAID are first line treatment and biphosphonates, if necessary, may be administrated. Some patients have been treated with biotherapies. The disease has usually a several months duration but may sometimes become chronic. Adults suffering from this disease have usually skin involvement corresponding to SAPHO syndrome (Synovite Acne Pustulosis Hyperostosis Osteomyelitis).
Conclusion: CRMO must be evoked in patients with recurrent multiple osteoarticular pain. Pediatrician must be aware of this entity to perform rapidly adequate investigations and begin treatment with NSAID that could save time and invasive investigations.

P 35

Sudden death at 5 years

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Case report: A 5 years old African French girl from non related parents presented a syncope in her classroom after being reprimanded by her teacher. The delay of intervention of the Emergency Medical Service (EMS) was about 10 minutes, without Cardio-Pulmonary Resuscitation (CPR) in the meantime. The resuscitation by EMS was stopped after 90 minutes without recovery of any cardiac activity. Two months before, she had a benign cranial traumatism after a sudden fall on her back and spent a few hours in hospital for a check-up. There were no other personal or familial past event. The biologic controls and anatomical autopsy were normal but histologic study revealed arrhythmogenic right ventricular cardiomyopathy (ARVC). A heterozygous mutation was found in the gene of DSP (desmoplakine, a desmosome protein), as in her father and 2 years old sister, both presenting normal clinical, echo- and electrocardiographic examinations. Gene SCN5A and KCNQ1, containing mutations in long QT and Brugada syndromes, were normal in our patient.
Discussion: ARVC manifestations are exceptional at this age. There are often triggered by catecholaminergic stimulation (effort, emotion). ARVC is due to fibroadipous cardiomyopathy begining in the right ventricle. The molecular anomaly may possibly concerns myocyte desmosomes which are important in the intercellular signalisation, and then be characterised by autosomic dominant transmission. The risk of sudden death by ventricular arrhythmia is a reality in ARVC as in catecholaminergic polymorphic ventricular tachycardia, Brugada and long or short QT syndromes.
Conclusions: 1. Teaching widely Basic Life Support techniques may improve prognosis of life threatening events out of hospital. 2. Autopsy remains the gold standard for post-mortem diagnosis in sudden deaths. 3. Syncopes of unclear origin should be investigated by a cardiologist, particularly in a context of effort or stress. 4. Familial investigations must be performed when a genetic pathology is diagnosed to detect asymptomatic patients and discuss a preventive treatment.

P 36

Irregulary tachycardia: think about adult disease

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Introduction: In presence of tachycardia, paediatricians think about sinus tachycardia or supraventricular tachycardia (SVT), if the QRS is narrow and about ventricular tachycardia if it is wide. Nevertheless, some others diagnosis as atrial fibrillation and atrial flutter should be kept in mind.
Case report: A 15 years old boy, without any medical history except of obesity, came to the emergency room because of heart palpitations with dyspnea and dizziness. It began while having sexual intercourse. He admitted repeated alcohol consumption the last days and smoking regularly. He felt palpitations for the first time the night before, which solved spontaneously. Physical examination showed obesity (91 kg >P97), arterial hypertension (130/90 mm Hg), irregular central tachycardia (170–220/min, peripheral 60/min) and tachypnea (20/min). ECG showed irregular, tachycardia with absent P waves and narrow QRS. SVT was the first diagnosis suspected. Therefore, 6mg of adenosine, then 12 mg were given. Thereafter, ECG showed a typical atrial fibrillation. 15 mg diltiazem was then given, without any effect. Laboratory (blood count, clotting test, electrolytes, troponin T, CRP, thyroid tests) and echocardiography were normal. He received flecainide 150 mg for cardioversion. Six hours later, sinusal rhythm was restored.
Discussion: Atrial fibrillation is very rare in children and mostly due to structural heart disease (rheumatic mitral valve disease, congenital heart disease with dilated atria) or sometimes as a complication of intra-atrial surgery. Without cardiac history, thyrotoxicosis, infectious pericarditis and pulmonary emboli are to be excluded. Recently, it was also postulated that exogenous causes as ecstasy, caffeine, alcohol could also be responsible of this heart disorder.

P 37

How can otitis media still complicate our (and our patients') lives?

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Introduction: Lateral Sinus Thrombosis (LST) is a rare complication of otitis media and associated mastoiditis that still occurs in the antibiotic era. We report a case of a LST as a result of an otogenic infection, complicated with nerve VI impairment, intracranial hypertension and papilledema.
Case report: A 5-year-old boy was admitted for otalgia with repetitive vomiting, headache, photophobia and persistent fever without meningism, despite adequate antibiotic therapy for otitis media. Intravenous ceftriaxone treatment was initiated. As ocular symptoms with pain and photophobia persisted, he underwent a cerebral CT scan, which showed right sigmoid, transverse and inferior sagittal sinus thrombosis with extension to the upper right internal jugular vein, in association with a right mastoiditis. Emergency myringotomy tube placement and mastoidectomy was performed. Anticoagulation was immediately initiated. Despite this, symptoms progressed with onset of

left VIth nerve impairment and, 15 days later, with left papillary edema. MRI confirmed intracranial hypertension with bilateral papilledema. The child underwent an external ventricular drainage and was anticoagulated by acenocoumarol for a total of 6 months. He fully recovered.
Discussion: LST is an uncommon intracranial complication of otitis media and associated mastoiditis. Typically originating in the sigmoid sinus, thrombosis may propagate to other sinuses and to the internal jugular vein. Vessel occlusion ultimately obstructs cerebrospinal fluid drainage, leading increased intracranial pressure and hydrocephalus. Classical symptoms include prominent headache with otalgia, otorrhea, postauricular tenderness and fever. Treatment strategies include myringotomy tube placement, mastoidectomy, anticoagulation and intravenous antibiotics. Mortality rates range from 5 to 10%.
Conclusion: Despite appropriate antibiotic therapy, LST is still a threat to children with otitis media. Paediatricians should suspect a LST in patients with otitis media who report headache, vomiting or other neurological symptoms.

P 38

Monoxide carbon intoxication – retrospective case review

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Introduction: Inodorant, insipidant, and invisible, carbon monoxide is one of the most frequently cause of intoxication. If neurological symptoms are neglected, it can result in permanent neurological injury. The manifestations of delayed neuropsychiatric sequelae are variable and non specifics.
Case review: A retrospective study (2004–2011) identified 13 children hospitalized in CHCVs with CO intoxication. Epidemiologic and clinical data were collected and analyzed. The patient ages ranged from 7 to 14 years, with a mean of 11 ± 4.5 years. The chimney smoke inhalation was the most common cause of intoxication (54%), followed by the improperly vented exhaust produced by gas water heaters (31%) and the house fires (15%). 100% of events occurred during cold season. The most common presenting symptom was consciousness disturbance (72%). Four patients (31%) received hyperbaric oxygen therapy. One (8%) needed intubation for laryngeal edema. HbCO levels ranged from 1.5% to 30.3%.
Discussion: The presence of the non specific neurological symptoms in children during the hibernal season must lead to an exhaustive anamnesis, in order to discover a potential exposition to carbon monoxide. Hyperbaric treatment is well codified for high HbCO level. Acute neurological symptoms should be detected in order to initiate hyperbaric therapy, regardless HbCO level. Delayed therapy may compromise neurologic recovery. More information is needed regarding the long-term effects of carbon monoxide on the pediatric population related to the acute neurological symptoms and to HbCO levels at the admission moment.
Conclusion: Physicians should be aware to the non-specific symptoms in children with positive anamnesis of CO intoxication. Early identification of neurological sequelae risk factors should help to provide better care.

P 39

Successful management of developmental hip dysplasia with Tübingen harness

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Introduction: Developmental dysplasia of the hip (DDH) is a frequent entity in early childhood. The incidence of dysplasia is 2–5% in general and 1–2‰ with dislocation. Obvious risk factors are: family history, breech position, multiple gestation, orthopedic malformations and neuromuscular disease. Without treatment, DDH may lead to early coxarthrosis. Maintenance in hyperflexion and abduction is needed for a correct hip joints development. Treatment of hip dysplasia by Tübingen harness has been applied since 1980's, mostly in German-speaking areas and is easier to apply than Pavlik harness.
Methods: We performed a retrospective study of early hip echography and follow-up of patients with risk factors born in Sion, from January 2009 to April 2011. The first echography was performed within 4-6 weeks of age and, if abnormal, was repeated 4 to 6 weeks after. Treatment with orthopedic harness was started immediately in case of severe hip dysplasia or dislocation (type D or worse by Graf) or with 3 months in case persisting mild dysplasia (≥IIb by Graf). All the treated patients were referred to an orthopedist, and a control echography was made 6 weeks later and a pelvis X-ray was performed at 6 months of age and by the acquisition of walking.
Results: 699 patients were screened with1674 hip ultrasounds.124 cases with hip immaturity/mild dysplasia normalized by 3 months of age. Twelve infants were lost to the follow-up. 27 infants with DDH needed treatment (38 hips). Twenty-one were classified as Graf type II, 9 as type D and 8 as type 3, 0 as type 4. 24 children were treated with Tubingen harness, one with Pavlik harness and two with another type of harness. In all cases, except 4 still under treatment, a total correction was obtained. The average time of treatment was 11 weeks. No patient needed surgical treatment. Neither femoral head necrosis nor other complication was observed during treatment.
Conclusions: Non invasive treatment of neonatal hip dysplasia by Tübingen harness in our population was successful, simple and well accepted by parents. No complications or failure were observed. The Tübingen harness may be used in all cases of mild to moderate dysplasia in newborn and infants, and even in children with unstable hips under orthopedic control. The follow-up should be continued even after 1 year of age because of risk of the residual dysplasia.

P 40

Pneumothorax, pneumomediastinum and epidural emphysema: rare complications of bronchiolitis

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Introduction: Spontaneous pneumomediastinum is uncommon in children. It is often associated with predisposing conditions, most frequently asthma, respiratory infections, vomiting and coughing. The epidural emphysema is a benign and exceptional complication of pneumomediastinum.
Case report: A 2 years old infant hospitalised for a mild RSV bronchiolitis, has rapidly deteriorated with severe RDS and fever. A moderate inflammatory syndrome was observed (CRP 65 mg/l – PCT 0.97 microg/l). Chest x-ray followed by CT scan showed basal infiltrate, pneumothorax, pneumomediastinum, epidural and subcutaneous thoracic emphysema. Oxygenotherapy (100%), antibiotic and antitussive therapy was initiated. The patient's condition progressively improved under symptomatic treatment.
Discussion: Pneumothorax and pneumomediastinum are rare but well-known complications of bronchiolitis. The paediatric literature identifies an initial incidence peak between 6 months and 3 years, probably due to the high prevalence of respiratory infections in this group. Both conditions are usually mild and self-limited with conservative treatment. They occasionally need chest tube drainage. Epidural emphysema associated with spontaneous pneumomediastinum is also a benign, self-limited condition and no neurological complications are reported so far with this entity. Because of lack of neurological symptoms, the diagnosis is often delayed or missed until a CT scan is performed.
Conclusion: Viral bronchiolitis is one of the most common paediatric respiratory diseases and the association with pneumomediastinum could be mediated by increased pressure within obstructed airways or perhaps by tissue necrosis from parenchymal infection. It could be rarely associated with air within the spinal canal. If this last condition appear to be benign and self-limited as in our case, a follow-up of this rare entity remains necessary because of lack of experience about possible neurological complications. In the literature, this is the first case reported in child of epidural emphysema following bronchiolitis.

P 41

Kwashiorkor: Protein malnutrition in Switzerland

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Introduction: Kwashiorkor is a disease caused by severe protein malnutrition. It is a frequent problem in low income countries. Only a few cases have been reported from the US, Canada and Australia. To our knowledge there is no previous report about protein malnutrition in Switzerland.
Case presentation: An 11 month old boy was admitted to our emergency department with progressive feeding refusal, diarrhea, rhinitis and fever. In the newborn period he had suffered from gastro-esophageal reflux and had developed generalized eczema under formula milk. The mother had tried several different formulas and ended up with rice milk as the supposedly most suitable alternative. According to the mother the milk's sweet taste led to an initial improvement of the feeding problems. In time the boy got irritable and developed a skin rash and marked discoloration of the hair. His growth and motor development were stunted. At 11 months the boy was unable to sit or turn without support. The clinical examination showed striking symmetric edema and a cushingoid facies. Laboratory testing revealed low serum protein levels which could not be explained by renal or fecal protein loss. Serum levels of phosphate, zinc and vitamin A were below the normal range. We calculated the average percentage of the recommended daily allowance for major nutrients the child had received during the previous 7 months: 1,2 g/kgKG/d for proteins; 1,2 g/kgKG/d for fat; 21,0 g/kgKG/d for carbohydrates. Because of the fact, that the biological values of protein in ricemilk is less than for casein based formula the boy had recived insufficient nutrient levels. All symptoms were compatible with severe protein

deficiency. We initiated a slow realimentation. Electrolyte disturbances were corrected. The therapy aimed at establishing a well-balanced diet and to instruct the parents in the importance of adequate nutrition for the development of an infant.
Conclusion: We suggest that despite, or possibly because of the abundance of various foodstuffs available, cases of severe malnutrition may be not only a problem of poverty, but can also be seen in the richest countries. In a time when a lot of investments and efforts are made in preventing allergies and to impose so called healthy diets it is the responsibility of every pediatrician and medical practitioner to have an open eye for conditions caused by malnutrition from inadequate diets given for health reasons.

P 42

From the eye to the kidney – ocular abnormalities leading to the diagnosis of Alport syndrome

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Background: Alport syndrome is a hereditary disease of type IV collagen characterized by the association of renal symptoms (hematuria and possible progressive kidney failure), sensoneurinal hearing loss and ocular changes. The inheritance is predominantly X- linked and mutations affect the gene encoding the α5 chain of type IV collagen (COL4A5). Homozygote men are usually severely affected with progressive kidney failure, leading to end stage renal disease. In contrast women tend to have a mild form, often with only microscopic hematuria and normal kidney function. The distinctive clinical features occur because the α5 chain of type IV collagen is normally expressed in the basement membranes of the kidney, ear, lens capsule and retina. Particularly in children, the definitive diagnosis of Alport syndrome may be difficult and many cases remain undiagnosed.
Case: A 12-year-old boy was diagnosed to have a severe sensoneurinal hearing loss, which required hearing devices since the age of 6 years. At the age of 12 years he additionally complained of visual impairment; an ophthalmological examination revealed the presence of a lenticonus, the retina did not show any pathological abnormalities. Because the combination of hearing loss and lenticonus was suspicious for Alport syndrome, urinalysis was performed, which revealed microscopic hematuria and gross proteinuria with normal renal function and blood pressure. The mother also has a known microscopic hematuria. The kidney biopsy confirmed the diagnosis of Alport syndrome with lack of staining of the glomerular basement membrane for α5 chain of type IV collagen on immunofluorescence microscopy and irregular thinned and thickened areas of the basement membrane on electron microscopy. An antiproteinuric treatment with the angiotensin converting enzyme inhibitor ramipril was started.
Conclusion: Lenticonus, retinal abnormalities and sensoneurinal hearing loss may be the first symptomatic manifestation of Alport syndrome in childhood. In case of typical eye abnormalities comprehensive and careful diagnostic exams are needed in order to make an early definitive diagnose of Alport syndrome, which allows a prompt initiation of antiproteinuric treatment to slow renal failure progression.

P 43

Perforated Meckel's diverticulum in a female neonate

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Introduction: The incidence of Meckel's Diverticulum (MD) is about 2% in the western population with a total life time risk of complications of 4%. Most symptomatic paediatric cases occur within the first two years of life. However, perforated MD in neonates has been reported in very few cases in the English literature.
Case: A term girl (birth weight 4000 g) presented in the emergency department at the age of 18 days due to mucous, blood-stained stools and abdominal distension without vomiting. Clinical examination showed a distended abdomen. Laboratory work-up revealed normal values for blood picture, coagulation profile and C-reactive protein. Initial abdominal ultrasound showed fluid-filled small bowel and some free abdominal fluid but no signs of intussuception or malalignment of superior mesenteric vessels. Over the next few hours, non-bilious vomiting occurred. Repeat abdominal ultrasound showed non-propulsive peristalsis suggesting intestinal obstruction. Abdominal x-ray indicated *small intra-intestinal air fluid levels,* but no pneumoperitoneum. Explorative laparoscopy revealed a perforated MD with macroscopic signs of diverticulitis. Two centimeters of small bowel were resected via lower transverse laparotomy. Further clinical course and feeding were uncomplicated and the girl was discharged home on full breast feeds six days post surgery.
Conclusion: Intestinal obstruction in neonates may be due to perforated MD. Laboratory signs of inflammation may be absent in neonatal perforated MD despite presence of diverticulitis. Laparoscopic evaluation is helpful in establishing the diagnosis.

P 44

Peripheral facial nerve palsy in severe systemic hypertension: report of two cases and review of the literature

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Objective: Signs of nervous system dysfunction such as headache, vomiting or convulsions often occur in severe systemic hypertension. Less recognized is the association between severe hypertension and peripheral facial nerve palsy. The goal of this study is to present 2 cases of severe hypertension associated with peripheral facial nerve palsy and to synthesize the published literature.
Patients and Methods: The U.S. National Library of Medicine database and the web-based search engine Google, through November 2010, were used as data sources. All articles published after 1960 as full-length articles or letters were collected.
Results: We included our 2 cases and 23 patients reported in the literature with severe hypertension (= blood pressure exceeding the 99th percentile by >5 mm Hg in children or 180/110 mm Hg in adults) and peripheral facial nerve palsy. Twenty-three of the 25 patients were ≤15 years of age. Facial palsy was unilateral in 24 cases and bilateral in one case. A second attack occurred in 8 patients. The time elapsed between the first facial symptoms and the diagnosis of hypertension ranged between 2 days and 2 years. In three case series specifically addressing the complications of severe hypertension in paediatric patients, 32 further cases of peripheral facial nerve palsy out of 755 patients (4.2%) were listed but not further described in detail.
Conclusions: The association between severe hypertension and peripheral facial nerve palsy merits wider recognition and assessment of blood pressure is recommended in every case of peripheral facial nerve palsy.

P 45

Polyhydramnios and high urea levels as leading signs of neonatal Bartter syndrome: a case report

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Introduction: Neonatal Bartter syndrome is a rare autosomal recessive disorder of renal salt reabsorption. Early recognition of typical symptoms and adequate fluid replacement can be life-saving. Most patients present with polyhydramnios during pregnancy and prematurity. After birth polyuria and various electrolyte imbalances prevail.
Case presentation: We present a boy who was born at 34 2/7 weeks of gestation with a birth weight of 1995g (P50). During pregnancy polyhydramnios was present and required repeated (15!) tapping. The boy's initial symptoms were floppiness, weak suck, polyuria and failure to thrive. A Lab work-up revealed high serum urea level with normal creatinine, hyponatraemia, hypochloraemia and metabolic alkalosis. We suspected the diagnosis of neonatal Bartter syndrome. This diagnosis was supported by the findings of elevated serum-osmolarity, aldosterone and renin and hypercalciuria with nephrocalcinosis in renal ultrasound. Detection of a mutation in the SLC12A1 gene confirmed the diagnosis of neonatal Bartter syndrome Type I. Massive substitution of up to 500 ml/kg/d of enteral and parenteral fluid was needed to improve the general condition and allow weight gain. Sodium, chloride and potassium were temporarily replaced. The administration of indometacin in increasing dosage up to 3.5 mg/kg/d finally permitted a gradual reduction of fluid administration. At present our patient is 11 months old, thriving well and the indometacin dosage could be reduced to 0.5 mg/kg/d. His fluid demand decreased to 130 ml/kg/d.
Conclusion: Though a rare disease, neonatal Bartter syndrome should always be considered in severe polyhydramnios. The testing of chloride in the amniotic fluid can easily give a hint towards the diagnosis (chloride >107–109 mmol/l). This simple lab test is helpful to suspect neonatal Bartter syndrome and to start early fluid replacement. With early treatment the prognosis of the patients is improved.

P 46

A study on the acceptability of the influenza vaccine during pregnancy

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Introduction: Pregnant women and infants are at increased risk of complications due to influenza infection, which are vaccine-preventable. In Switzerland seasonal influenza vaccine is recommended for pregnant women since 2009. We conducted a study to assess the acceptability of influenza vaccine during pregnancy.
Methods: All women hospitalised in March 2011 in the post-partum unit of the Maternity of the University Hospitals of Geneva were offered a questionnaire to assess their knowledge, beliefs and acceptance of influenza vaccine during pregnancy.
Results: The questionnaire was offered to 320 women of which 255 (80%) agreed to complete the questionnaire. 77% of women were aware that influenza presents an increased risk for pregnant women but only 46% were aware that flu vaccine was recommended during pregnancy. Only 19% of women reported having been immunized during their pregnancy and only 43% recalled that the vaccine had been offered. Principal barriers to vaccination were lack of information from healthcare workers (47%), and the beliefs that the vaccine was unsafe during pregnancy (37%) and not effective (19%).
Conclusion: Identifying the barriers to influenza vaccination during pregnancy will help in designing interventions aimed at improving the knowledge about and acceptance of influenza vaccination during pregnancy.

P 47

Often harmless – but sometimes with severe complications: Three cases of intracranial complications in trivial ORL-Infections

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Introduction: Upper airway infections lead to 70% of pediatric consultations. Most of them heal spontaneously or under antibiotic treatment without difficulties. However intracranial infections can rarely occur as a severe complication.
Case Reports: We report three cases of severe intracranial complications of trivial ORL-infections:
1. A 5-year-old girl in unimpaired general condition admitted with periorbital cellulitis after antibiotic treatment of otitis media. Cranial MRI revealed sinusitis and transosseous intracranial extension with meningeal enhancement.
2. A 14-year-old boy who presented with headache, a complex focal seizure and Todd's paralysis. The CT-scan confirmed sinusitis and antibiotic treatment was started. During this treatment he developed severe cerebritis with reduced vigilance, hemiplegia, and epidural/subdural empyema, necessitating subsequent neurosurgical intervention.
3. A 18-month-old girl presenting with ataxia, nystagmus and impaired vigilance following otitis media and mastoiditis. The cranial MRI showed an epidural abscess , sigmoid sinus thrombosis and neurosurgical intervention was necessary.
Conclusion: Common ORL-infections can be complicated by severe intracranial complications, sometimes initially associated with just mild symptoms. Minor or major focal neurological signs must trigger immediate neuroradiological imaging to detect or exclude intracranial pathology and enforce proper treatment.

P 48

Ethical decision making in severe malformation syndrome

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Introduction: As physicians we are often faced with ethical questions such as whether to continue life-supporting measures or not. We report the case of a newborn presenting with a severe malformation syndrome, which led us to judge intensive care as not beneficial for the child even before knowing the genetic diagnosis.
Case report: A full-term male was born by primary caesarean because of vaginal bleedings and breech presentation in the 37 ⅐ gestational week. The ultrasounds during pregnancy and the pregnancy itself were without pathological findings. There are no genetic disorders in family history. There was an intrauterine growth retardation with birth weight of 2050 g (350 g <P3). Postpartum several malformations stood out: Trigonocephaly, club-feet, contractions and abnormal position of the upper extremities, cavernous eye sockets with near impossibility to open the lids, preauricular tags, retrognathia and a high arched palate. Furthermore there was a muscular hypotonia, a missing sucking reflex and in the abdominal ultrasound we saw two large polycystic kidneys. Findings were indicative of severe CNS-dysfunction of prenatal, most probably genetic origin. Before we got the results of the chromosome analysis there was a progressive deterioration of the boy's condition with frequent episodes of central bradyapneas requiring noninvasive ventilatory support. The situation became critical, so that we had to decide about life sustaining therapy although we did not yet have a definite diagnosis. Because prognosis was estimated to be very poor regarding survival as well as future quality of life, we and the parents came to the decision not to intubate and later on to stop ventilatory support as well. The boy died at the age of one week. The final diagnosis was an unbalanced chromosomal translocation 13;22 with partial trisomy 13 and monosomy 22q.
Conclusion: This case illustrates the importance of an individual, case-related approach to ethical decisions in our daily work. According to the "7-step-dialog" of *Dialog Ethik* our team unanimously came to the conclusion to stop life supporting measures before knowing the diagnosis. Post mortem we did a MRI for further diagnosis and from the genetic laboratory we got the final diagnosis of translocation trisomy 13, which has little chance of survival.

P 49

Primary ciliary dyskinesia: Usefulness of nasal nitric oxide measurments

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Background: Primary ciliary dyskinesia (PCD) has a prevalence of 1:15000. The gold standard of the diagnosis is an electron microscopy (EM) of nasal or bronchial cilia. Nasal nitric oxide (nNO) levels are usually very low in this disease and the nNO measurements are used as screening tool for PCD. In our clinic a cut-off level of 100 ppb was used. In the recent literature much higher nNO levels are reported in confirmed PCD patients.
Objectives: To evaluate the diagnostic pathway for PCD based on nNO.
Methods: all the children with nNO <180 ppb in the last 3 years (n = 30) were interviewed in a systematic way and retested in our outpatient clinic. The measurement were done only when the children hat no signs of upper respiratory infection. If the second measurement confirmed the low nNO level or if the clinical history remained suggestive for PCD, nasal brushing for light and/or EM were performed.
Results: A second nNO measurement was performed in 30 children. Median (range) nNO of the first measurement was 83 (6.4–180) ppb. Median retest nNO was 280 (1.4–600) ppb. 23 Patients showed values higher than 180 ppb. 7 patients had a persistently low nNO or suggestive anamnesis and underwent further investigations with light and/or EM. Ciliary beating frequency was normal in all 7 patients, the EM results are still lacking (end results are expected in June/July 2011).
Discussion: Diagnosis and/or exclusion of PCD remains a diagnostic challenge. nNO measurements can be useful as a first screening. Measurements during infection of upper airway might show false low results and should be repeated after treatment of the nose with saline and in symptom-free interval.

P 50

Familial cases of neonatal lactic acidosis, Fanconi tubulopathy and sensorineural deafness in complex III deficiency

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Background: Neonatal lactic acidosis represents the onset of different inborn errors of intermediate metabolism. If a specific organic aciduria or gluconeogenetic deficiency is early recognized, dietary treatment is usually life-saving. When neonatal lactic acidosis is due to primary mitochondrial dysfunction, for which no effective treatment exists, neurological outcome and vital prognosis are mostly poor. Molecular confirmation of mitochondrial diagnosis is often burdensome but important for genetic counseling.
Clinical report: We report a 17 year-old boy born to consanguineous Turkish parents. In the neonatal period he presented hypoglycemia, glycosuria and life-threatening lactic acidosis which completely

normalized after infancy. Hearing loss was recognized and treated from age 3 years. At 14 years, he presented with short stature, deafness, genua valga, hypophosphatemic rickets due to renal Fanconi syndrome, and cognitive function in the low normal range. A mitochondrial disease was suspected based on the clinical history, although lactate was not increased in plasma and urine. He developed mild glomerular renal insufficiency. Family history revealed that his first cousin, a 14 year-old girl, had growth retardation, renal Fanconi syndrome, genua valga, severe hypophosphatemic rickets, deafness (diagnosed at age 13 years) and mental retardation.
Results: Complex III deficiency of the mitochondrial respiratory chain was detected in muscle biopsy. Sequence analysis of BCS1L revealed the novel homozygous point mutation p.M48V in both our patient and his cousin.
Discussion: Complex III deficiency is caused most often by mutations in the nuclear gene BCS1L, coding for a CIII assembly factor. The phenotypic features presented here are in between the Bjornstad syndrome (sensorineural deafness and pili torti) and the lethal GRACILE syndrome, both caused by BCS1L mutations. Cognitive outcome may be variable and possibly linked to the timing of deafness correction. This report also highlights that lactic acidosis may be transient in this disorder with relatively good neurological outcome, unusual for a respiratory chain disorder with severe neonatal onset. Molecular diagnosis allowed appropriate genetic counseling and possibility for prenatal testing in multiple siblings, in a family with high rate of consanguinity. The precise diagnosis improved the management and follow-up of the 2 patients with multi-organ survey and supportive treatment. The genetic diagnosis brought an important psychological benefit to the family, after 16 years of strong guilt and uncertainty feelings.

P 51

Development and validation of a didactic game for food allergic children

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Background: Food allergy in children, significantly affects the quality of life. As there is no available cure for the disease, management is restricted to avoidance and emergency treatment of reactions after accidental ingestions. Education is therefore key in order to improve their quality of life and to reduce morbidity and mortality.
Objectives: The aim of our study was to develop and validate – by the validated French versions of the Food Allergy Quality of Life Questionnaires, parent's form and children's form (FAQLQ-PF and FAQLQ-CF) and the food allergy independent measure (FAIM) – a food allergy specific, didactic and interactive electronic game.
Methods: The game was based on common situations encountered by food allergic children. Food eviction, accidental ingestions, anaphylaxis management and general knowledges about food allergies were pictured in the game by 12 scenarios and 9 multiple-choice questions. The questionnaires were submitted to 8–12 years old children and their parents during an allergy visit, and analysed in term of emotional impact, food anxiety and social and food limitations *before* the children played the game, *and 2 months later.*
Results: 32 nuts, 8 to 12 years old, allergic children were included, with a girl/boy ratio of 1:1.66. Median age was 9.4 years. Most of them were allergic to only one food (62.5%). 66% suffered from mild systemic reactions and 34% from severe systemic reactions. Quality of life was improved by the use of the didactic game among children with severe reactions (Global score: 2.45 versus 1.97, p = 0.03; emotional impact score: 2.15 versus 1.51, p = 0.04; social limitations: 2.43 versus 1.87, p = 0.03) and among all children in term of their perception of their ability to manage an allergic reaction (FAIM: 2.27 versus 1.58, p = 0.02).
Conclusion: We could demonstrate that our didactic interactive game was a useful tool to improve their quality of life, especially among children with severe food allergies. Education of food allergic children is an important part of the management of this condition.

P 52

Children with food allergy: Validation of the French version of the Food Allergy Quality of Life Questionnaire-Children Form

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Background: It has been previously shown with English speaking children that food allergy clearly affects their quality of life. A health-related quality of life questionnaire for food allergic children has been validated in Dutch in 2009, however to date no such questionnaire is available in French.
Objectives: To validate the French version of the Food Allergy Quality of Life Questionnaire- Children Form (FAQLQ-CF) developed and validated in Dutch and available in English.
Methods: The English version of the FAQLQ-CF was translated to French by two different French-speaking translators and retranslated by a English-speaking translator.
We then recruited 32 patients between 8 and 12 years with an IgE mediated food allergy. Children answered the questionnaire during a regular clinic visit. The results obtained were then analysed and compared with the results of the original FAQLQ-CF and the Food Allergy independent Measure (FAIM).
Results: 32 questionnaires were fully completed and available for analysis. A Cronbach's α correlation index of 0.67 was found for the total FAQLQ-CF. Validity was demonstrated by significant correlations between FAQLQ-CF and the FAIM. (0.67; $p < 0.05$).
Conclusion: The French version of the FAQLQ-CF was validated and will provide a tool for assessment of Quality of Life in French-speaking children with food allergy. It will be an important tool for clinical research and will allow research collaboration between French and English speaking research teams.

P 53

Prevalence of streptococcal pharyngitis in a paediatric emergency department

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Introduction: Determining the prevalence or the pre-test likelihood probability of disease is the first step in evidence-based practice. It is important for clinicians to know the prevalence of disease to evaluate if a diagnostic test is necessary. We conducted a study to determine the prevalence of group A beta-haemolytic streptococci (GAHS) infection in child who presented sore throat in a paediatric emergency department.
Methods: We included in a prospective study 1500 patients aged 3–18 years with sore throat between January 2010 and March 2011. All children benefited rapid antigen detection test (RADT) and throat culture (gold standard test). We divided children into 3 subgroups according to age for more accuracy.
Results: Prevalence of GAHS is 45.3% with % IC95 (43–48) with throat culture and RADT. Prevalence is variable with age. Group of age of 5–10 years have high prevalence (55%) versus age 3–5 years (<40%) or age 11–18 years (<40%) where prevalence is low. Prevalence is also variable with season. High prevalence occurs in autumn (53%) and low prevalence in others seasons (43%).
Conclusion: This study demonstrates that children who present sore throat in a paediatric emergency department have a high probability of having GAHS pharyngitis and must be tested for GAHS infection. This is valid for all subgroups of children and for all seasons.

P 54

Megalencephalic leukoencephalopathy with subcortical cysts – a rare cause of macrocephaly and mild gross motor developmental delay

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Introduction: Megalencephalic leukoencephalopathy with subcortical cysts (MLC) is a rare autosomal recessive disease first described by Van der Knaap 1995. It is characte-rized by infantile macrocephaly, delayed motor development and early onset of seizures. Mild mental deterioration appears at a later age. Cerebral MRI shows diffusely abnormal and swollen cerebral white matter and subcortical cysts, cerebral atrophy occurs over time. In approximatly 70% of MLC patients mutations in the MLC1-gene is found.
Case report: We report a now 3-year-old boy, first child of non-consanguineous and healthy parents, presenting first in our

neuropediatric clinic at the age of 1 year, with progressive macrocephaly, with a head circumference of 35 cm (P. 50) at birth and 56 cm (6 cm >P. 97) at the age of 1 year. History of pregnancy and birth as well as motor and cognitive development within the first year were normal. Family history showed no abnormalities. Head circumferences of both parents were above P. 97. To determine if there is a genetic contribution to macrocephaly we used the "Weaver curve" with calcula-ted standard deviation scores for child and parents, which showed a head circumference 4 standard deviations above the parents' average score. Glutaraciduria type I could be ex-cluded and there were no clinical signs of elevated intracranial pressure. Because parents were reserved towards further investigations we proposed follow up every 6 months. Over the course the boy showed a normal cognitive development, but slight difficulties in balance. Head circumference rose slowly. At the age of 2½ years we remarked mild problems in motor development with minor overall hypotonia and problems in balance, so that we performed a cerebral MRI to explore the etiology. This showed an abnormal, swollen white matter and bitemporal subcortical cysts typical for MLC. A routine EEG showed unspecific abnormalities, but no epileptic discharges. Because of the desire of further children we recommended a genetic confirmation, which we are waiting for.
Conclusions: Our case demonstrates a rare cause of severe macrocephaly with only slight disturbances of motor development. Confirmation of the etiology is important for suppor-ting the child but also for family planning in consideration of the autosomal recessive inheritance. In children with macrocephaly without syndromal features but signs of developmental delay neuroimaging is warranted because there is a broad spectrum of differential diagnosis. In this context, standard scores and associated curves can be helpful to determine whether a constitutional origin for macrocephaly is likely or not.

P 55

Rapid transformation of Stevens Johnson syndrome into toxic epidermal necrolysis: early recognition of cutaneous warning signs

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Introduction: Bullous skin eruptions may be an important early marker for potentially severe drug reactions such as Stevens Johnson syndrome (SJS) and toxic epidermal necrolysis (TEN).
Results: We report the case of a 14-year-old boy who initially presented with localized grouped blisters on his neck, conjunctivitis and stomatitis after receiving sulfadiazin and pyrimethamin for 4 weeks due to a re-activation of congenital ocular toxoplasmosis. His bullous skin lesions rapidly spread to result in TEN with epidermal detachment of 60% his total body surface area (BSA), including severe mucous membrane and ocular involvement. He was successfully treated with intravenous immunoglobulins, intravenous methylprednisolone and appropriate intensive care and wound management. The lymphocyte transformation test identified pyrimethamin as the most likely causative agent.
Conclusion: Although rare in childhood, early recognition of characteristic skin findings and clues for the development of a severe drug reaction is crucial for a favourable outcome. These skin signs include acute onset of painful erythema, mucous membrane lesions, onset of blisters on the face and neck with centrifugal spreading and atypical target lesions. SJS (epidermal detachment <10% BSA) can rapidly progress to SJS-TEN overlap (epidermal detachment 10–30% BSA) and TEN (epidermal detachment >30% BSA) with high morbidity and mortality. The key to successful outcome includes early recognition, prompt withdrawal of the causative agent, treatment with immunoglobulins and corticosteroids as appropriate, transfer to an intensive care unit, fluid resuscitation and infection monitoring.

P 56

Fever in a returning traveller: a journey through differential diagnosis

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Case Report: A 7 years old girl in good condition presented with fever ten days after her return from a trip in Martinique (area that is not endemic for Malaria). It was accompanied by a non-petechial rash. A tourniquet test was positive (presence of 10 or more petechiae per square inch after inflation to a point between the systolic and diastolic blood pressures for five minutes). The other clinical features were non contributory. Nonspecific laboratory abnormalities included leucocytosis. Platelet count was normal. A serologic test confirmed the clinical diagnosis.
Discussion: Fever in travellers returning from tropical areas often presents a diagnostic challenge for clinicians. Most tropical infections have a nonspecific presentation. However, there are clinical and laboratory features that could help in diagnosing the most frequent tropical conditions including the travel destination. First of all one must rule out Malaria which is the most frequent imported tropical disease. Signs highly suggestive for Malaria are trombocytopenia, hyperbilirubinemia, enlarged spleen. When Malaria is ruled out main predictors for rickettsial infection are skin rash, and skin ulcer; for dengue: skin rash, thrombocytopenia, leucopenia and a stay in Latin America or southern Asia/Pacific; for acute schistosomiasis: eosinophilia; for enteric fever: enlarged spleen and elevated alanine amino-transferase level. Going through the fevertravel.com guidelines helps make the diagnosis. In the case of our patient, the presence of fever after a stay in Central America accompanied by a rash, was highly suggestive of Dengue Fever. Furthermore the presence of a positive tourniquet test, which determines capillary fragility, helped guide the diagnostic. The dengue NS1 Antigen was negative due to the duration of the symptoms, however the dengue serologic test was positive. Dengue is the most prevalent mosquito-borne viral disease. It's incidence has grown dramatically around the world in recent decades. It is now endemic in more than 100 countries in the tropics. WHO currently estimates there may be 50 million dengue infections worldwide every year including 24 000 deaths due to hemorrhagic complications. In Switzerland there is an increase of the recorded cases, 86 cases in 2010 versus 2 cases in 1988. This is in part due to the constant increase in international journeys. It is therefore important to recall dengue fever when faced with fever in a returning traveller.

P 57

"The girl who had the limbs on fire": erythromelalgia due to small fiber neuropathy

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Introduction: Erythromelalgia (ETM) is characterized by severe, usually paroxysmal burning pain in hands and feet with warmth and redness, relieved by cold water immersion. Secondary ETM can occur in adults with diabetes mellitus, collagen or myeloproliferative diseases, and primary ETM in children and adults harboring a mutation in a sodium channel subunit expressed in sensory neurons (familial or sporadic).
Case report: This previously healthy 6-year-old Caucasian girl presented with sudden onset of attacks of burning pain in both hands and feet after skiing. Progressive increase in severity and frequency of the episodes was noted during the following days, preventing sleep. Osteo-articular and neurological examination including sensory modalities was normal. No swelling, redness or temperature change was observed, except cold skin and maceration due to almost permanent cold water immersion which was her only relief. Analgesic drugs including morphine were inefficient. Fortunately, the child improved within 2 months on a combination of gabapentin, mexiletine and chlorpromazine. Electroneurography showed no signs of neuropathy. CSF (day 8) was normal. A large screening for autoimmune, paraneoplastic and metabolic disorders (including Fabry) was negative; search for mutation of Na 1.7 sodium channel negative. Punch skin biopsy (day 74: PGP9.5 immunolabeling with morphometric quantification) showed severe small fiber axonopathy (SFA).
Conclusion: The pathophysiology of ETM is not yet fully understood. SFA may be an important but under-recognized cause. In this case, SFA was found only thanks to a special skin biopsy interpreted by an expert. An autoimmune mechanism as in Guillain-Barré syndrome, which can also affect axons and not only myelinated fibers and more specifically sensory and autonomic nerve-endings, is postulated here. Since remission was obtained, no immunotherapy was given, but this can still be offered in case of relapse. Paroxysmal severe pain attacks are very distressing not only for the patient and family, but also for the caregivers. A psychosomatic origin may be suspected and if not thought of, the diagnosis of ETM can be very difficult.

P 58

Bright kidneys on fetal ultrasonography – a rare differential diagnosis

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Introduction: Some diseases can prenatally present with enlarged hyperechogenic kidneys, autosomal recessive polycystic kidney disease being the most frequent. We describe the case of a newborn girl with a rare disease, which infrequently may prenatally present with hyperechogenic and enlarged kidneys.
Case: At 22 weeks of gestation, fetal ultrasound of a healthy secondipara with unremarkable personal and familial history revealed enlarged hyperechogenic kidneys without any other abnormalities. At that time a tentative diagnosis of autosomal recessive polycystic

kidney disease was made. At 29 weeks of gestation, oligohydramnion, mild cardiomegaly and cystic enlargement of the septum pellucidum were additionally found. Amniocentesis disclosed a normal caryotyp e (46 XX) and fetal MRI confirmed the findings on ultrasound. Because of increasing oligohydramnion and asymmetric growth retardation, elective caesarean section was performed at 36 1/7 weeks. The hypotrophic girl had a good initial adaptation, diuresis and creatinine were within normal range and abdominal ultrasonography showed enlarged, polycystic kidneys. The girl presented a relative macrocephaly, a big and prolapsing fontanelle, flat ears and a generalized muscular hypotonia. Cerebral ultrasound demonstrated bilateral calcification of basal ganglia, an echocardiography revealed ventricular hypertrophy. On the second day of life the girl developed tachypnea, lactic acidosis with hyperammonemia and an odour of sweaty feet. Diagnosis of glutaric aciduria type II was confirmed by organic acids and acylcarnitine profile. Therapy with benzoate, carnitine and riboflavin did not improve the condition and in view of the infaust prognosis of the neonatal form of the disease, active treatment was terminated. The girl died on third day of life.
Conclusion: Glutaric aciduria type II should always be considered when enlarged kidneys are observed on fetal ultrasonography, especially if there are other organ abnormalities, such as cardial or cerebral malformations. Prenatal diagnostic possibilities to confirm the diagnosis are available.

P 59

Mimicking perinatal infection: Aicardi-Goutières Syndrome

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Introduction: Small-for-gestational age, thrombocytopenia, mild ventriculomegaly, echogenic lenticulostriate vessels and intracranial calcifications are nonspecific findings in neonatology with a broad differential diagnosis covering chromosomal anomalies, infectious diseases or they may present the first clue to rare genetic disorders. Here, we present the case of a neonate with Aicardi-Goutières syndrome.
Case report: A female infant was delivered by elective caesarean section at 35 weeks to a 29-year-old primigravida. The parents were consanguineous. The pregnancy was unremarkable until the 33rd week when mild ventriculomegaly and growth retardation were diagnosed. A fetal MRI confirmed the ventriculomegaly and showed loss of white matter. Due to a pathological fetal Doppler exam, a caesarean section was performed. The initial transition was normal and birth weight at 1835 g was on the 5th percentile for age and head circumference on the 15th percentile. Physical examination was unremarkable except for moderate generalized hypotonia and mild hepatomegaly. Laboratory tests showed a thrombocytopenia and mild neutropenia. Cranial ultrasound demonstrated small bilateral subependymal hemorrhages and slightly dilated lateral ventricles. In addition, there were echodense calcified parenchymal and echogenic lenticulostriate vessels. An MRI confirmed the ultrasound findings. Aside from small retinal hemorrhages, the ophthalmological examination was normal as was the otoacoustic screening. Congenital infections (HIV, CMV, rubella and toxoplasmosis) were excluded. Cerebrospinal fluid showed a lymphocytic pleocytosis, CSF protein concentration was 1.13 g/l. Aicardi-Goutières syndrome (AGS) was also considered in the differential diagnosis. CSF interferon-α was measured and found to be markedly elevated. The clinical and radiological findings were compatible with AGS. Genetic testing was done but to date none of the known mutations for AGS have been identified.
Conclusions: Aicardi-Goutières syndrome should always be considered in neonates with symptoms suggestive of congenital infection but negative screening for infections. Genetic counselling is essential for the families concerned.

P 60

Symptomatic congenital CMV infection after recurrent maternal infection

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Introduction: Cytomegalovirus (CMV) is probably the most common congenital viral infection. About 10 percent of congenitally infected fetuses display symptoms at birth. While the transmission rate is high in primary maternal CMV infection it is low in re-activated infections. Treatment of symptomatic infants with intravenous (i.v.) ganciclovir may decrease the risk of hearing loss, but strategies regarding route and duration of treatment differ widely due to a lack of randomized controlled trials.
Case report: We present the case of a term baby-girl with extensive petechiae, hepatosplenomegaly and jaundice at birth. Prenatal MRI demonstrated intraventricular occipital adhesions. Given that the mother had documented CMV-IgG antibodies prior to pregnancy and CMV-PCR from amniotic fluid was positive, we suspected maternal recurrent CMV infection during pregnancy. After birth, congenital CMV infection was confirmed by PCR in blood and urine samples of the infant and i.v. ganciclovir therapy via central line was started. Postnatal MRI showed distinct cerebral abnormalities (intraventricular frontal and occipital adhesions, bilateral subependymal cyst at the caudothalamic groove, and ventriculomegaly) but neither calcifications nor polymicrogyria were seen. Severe thrombocytopenia required repeated platelet transfusions. Profound hepatopathia led to conjugated hyperbilirubinaemia, coagulopathy, and progressive cholestasis within the first two weeks of life. The latter was explained by functional bile duct obstruction due to CMV rather than an adverse effect of ganciclovir. Otoacoustic emissions and acoustic evoked potentials at newborn age were abnormal. Eye examination was unremarkable except for initial preretinal bleeding spots. Liver dysfunction improved and CMV viral load in plasma decreased significantly during 3 weeks of i.v. ganciclovir therapy. Thus, therapy was changed to oral valganciclovir (for further 3 weeks) and she was discharged home with a close interdisciplinary follow-up in the outpatient clinics.
Conclusion: Symptomatic congenital CMV-infection due to recurrent maternal CMV-infection in pregnancy is rare but can be associated with severe hepatic and cerebral manifestations. Current strategies for treatment of congenital CMV infection will be discussed including route and duration of ganciclovir administration.

P 61

The effect of restricted knee extension on muscle activity during gait in healthy subjects

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Introduction: In patients with cerebral palsy (CP), spasticity and muscle weakness lead to gait deviations. If both legs are affected, crouch gait with excessive knee and hip flexion is a frequent problem endangering gait function. The purpose of this study was to determine the effect of restricted knee extension during gait in normal individuals and its influence on muscle activity.
Methods: Nine healthy adult female subjects underwent a 3D gait analysis to measure whole body kinematics and lower body kinetics. First the subjects walked with restricted knee extension of 30° bilaterally accomplished by applying a taping technique to the upper and lower leg. Then the tape was cut and after at least 10 minutes of rest, recordings continued for normal gait. To retrieve the time-frequency and intensity content of the sEMG, the wavelet analysis technique as proposed by Von Tscharner [4] was used. The duration of muscle activity (on-off phases) was calculated and expressed in percentage of gait cycle.
Results: The gait pattern with restricted knee extension of 30° resulted in increased anterior thorax and pelvic tilt, hip and knee flexion, and ankle dorsiflexion compared to normal. The duration of muscle activity over the gait cycle, and the normalized amplitude of the sEMG intensity patterns, both increased under restricted knee extension gait pattern for the majority of muscles investigated.
Conclusion: Restricting normal subjects during gait at one level only, their knee extension, resulted in substantial deviations from normal gait with compensatory adjustments observed at all levels. In contrast to the experiment, Patients with cerebral palsy walking with crouch gait usually have pathologies at more than one anatomical level as well as spasticity and muscle weakness. Although comparing our results with patient data will be difficult, the data give us insight into the changes occurring when imposing a single factor known to influence crouch gait [5]. This shows the complexity and challenge of treating patients with crouch gait.

P 62

Involvement of autophagy in severe hypoxic-ischemic encephalopathy of newborn infant: an autoptic study

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Introduction: Despite advances in neonatology, perinatal hypoxic-ischemic encephalopathy (HIE) remains of major concern for its acute and long-term neurological consequences. Developing more effective neuroprotective strategies requires a better understanding of the

neuronal cell death pathogenesis. Studies derived from neonatal animals models of hypoxia-ischemia (HI) which target both necrosis and apoptosis have demonstrated only limited neuroprotection. More recently, our and other groups have shown that autophagy, a physiological process of cellular components degradation linked with lysosomes, is enhanced and that its inhibition exhibits neuroprotection in rodent neonatal models of cerebral HI.
Aims: The objectives of this study were to determine whether changes in autophagy can be observed following a perinatal asphyxia in autopsied brains of human newborns.
Material and methods: Newborns (>36 weeks) who died between 2004–2009 after a perinatal asphyxia were selected retrospectively based on the following criteria: Apgar score ≤5 minutes, metabolic acidosis with cord arterial pH <7.00 at 1 hour of life or a base deficit >12 mmol/l, early onset of clinical encephalopathy Sarnat III and multi-organ failure. Neonates matched for gestational age and died from other conditions were selected as controls. The presence of excessive autophagy was investigated in brain sections (thalamus and basal ganglia) preserved in paraffin blocks by using immunohistochemistry and confocal microscopy. LC3, marker of autophagosomes, was used along with LAMP1 and cathepsin D, two markers of lysosomes.
Results: The brains of 7 HIE cases (4 males, 3 females) and 6 control cases (3 males, 3 females) were analyzed. Preliminary results showed more than a two-fold increase in the total number of LC3-positive dots per neuron in the thalamus of HIE cases compared to control cases. Total numbers of cathepsin D- and LAMP1-positive dots per neuron were significantly higher in HIE cases than in control. The size of cathepsin D- and LAMP1-positive dots was also significantly larger than in control suggesting an increased neuronal formation of autolysosomes.
Conclusion: These results suggest an upregulated autophagic flux in the brain of newborns with HIE, as observed in animal models. Neuroprotective strategies targeting autophagy may be considered for future therapeutic approaches in combination with hypothermia to prevent the pathological consequences of HIE.

P 63

Infantile exanthematous pustular psoriasis evolving into plaque psoriasis and successful treatment with topical tacrolimus

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Introduction: Psoriasis in infants below the age of 12 months is very rare and pustular variants particularly uncommon in this age group.
Methods: We describe the clinical course and treatment response of a female infant presenting with acute onset of an exanthematous pustular rash.
Results: A 7-month-old girl showed a rapidly spreading generalized pustular eruption predominantly affecting her palms and soles. A diaper rash had appeared 2 weeks earlier. The child was otherwise well, there were no signs of systemic infection or fever and the pustules were sterile. The differential diagnosis included pustular psoriasis, acute generalized exanthematous pustulosis or post-streptococcal pustulosis acuta generalisata. The child was on no medication, investigations were negative for enterovirus, parvovirus or streptococcus infection and the family history was negative for psoriasis. The histology of a skin biopsy was consistent with pustular psoriasis. Initial treatment with oral amoxicillin-clavulanic acid and topical mometasone resulted in improvement of the lesions. However after stopping the anti-inflammatory treatment, widespread plaque- and guttate-type psoriasis with a PASI of 11 and nail pitting developed over 8 weeks. Treatment with topical tacrolimus 0.03% ointment once daily and emollients was initiated, which rapidly lead to complete resolution of all lesions over the next 2 weeks with residual leucoderma. At the last follow-up 4 months later the patient remained symptom-free.
Conclusion: This is remarkable course of acute onset pustular psoriasis evolving into widespread plaque-type psoriasis in a young infant. Mono-therapy with topical tacrolimus was highly effective and lead to sustained clearance. Topical calcineurin inhibitors represent a useful alternative to corticosteroids in the treatment of childhood psoriasis, in particular in young children and for flexural and facial involvement.

P 64

Chronic relapsing urticaria confined to injection sites of upper arms

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Background: Identification of the course of localized recurrent urticaria in childhood may be challenging.
Case report: 8 yrs old boy with asthma, allergic rhinoconjunctivitis and atopic dermatitis for a period of 12 months repeatedly presented with urticae strictly localized to the upper arms. Beforehand, the same areas had been subject to multiple intramuscular and subcutaneous injections for immunotherapies and vaccinations. No other injection sites were reported by the parents.
Discussion: The observed localized reaction can be best explained by the pathogenetic concept of an isotopic response phenomenon, which is characterized by the occurrence of a new skin disorder exactly at the site of another unrelated and already healed skin lesion or trauma. This is in contrast to an isomorphic response (Köbner phenomenon), which refers to the appearance of new lesions of a pre-existing disorder along the site of injury. The main known events that may damage and immunologically mark a skin region, which later on shows an isotopic response include chronic lymphoedema, atypical and exaggerated immune reaction to viral antigens, to immune complex deposits or altered tissue antigens. Furthermore, a bidirectional interaction between the nervous system and the immune system may affect the secretion of neuropeptides from cutaneous nerve fibers. This may influence the number and function of T lymphocytes, monocytes, mast cells, and endothelial cells in the skin.
Conclusion: This is the first reported case of an isotopic response presenting with urticaria following multiple injections. Skin lesions due to of subcutaneous and intramuscular injections may affect the immunological, neural as well as vascular system, hereby making this area prone to the development of new cutaneous symptoms.

P 65

Neonatal Bartter syndrome-spontaneous resolution of all biochemical abnormalities

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Background: Bartter's syndrome is a group of renal tubular disorders charaterized by hypochloremic metabolic alkalosis, hypokalemia, hyperreninemia with normal blood pressure, and eventually hypercalciuria. The neonatal variant has the following additional features: prenatal onset with polyhydramnios, premature delivery, postnatal salt wasting with severe dehydration, constipation and hyperprostaglandinism. Most children with Bartter's syndrome have an inherited disorder and the causes are mutations of genes encoding proteins that transport ions across renal cells in the thick ascending limb of Henle's loop.
Method: Case report.
Results: Our female patient was born in 2006, at 34 wks of gestation by caesarean section, BW 1690g, APGAR 5/7/9. Antenatally the mother had a severe unexeplained polyhydramnios since the 31st week of gestation. At birth there were no craniofacial dysmorphisms, but the girl presented with marked polyuria and polydipsia. Biochemical investigation showed a hypochloremic metabolic alkalosis with hypokalemia, and hypercalciuria. This in combination with high plasma renin and aldosterone levels and high prostaglandin levels in blood and urine was suggestive of neonatal Bartter's syndrome. Sequencing of major Bartter genes did not reveal any mutation. The parents were not consanguineous. Renal sonography was normal. With sufficient fluid intake, sodium and potassium supplementations and indomethacin therapy, serum electrolytes normalized. The baby was discharged 4 weeks after birth. Psychomotor development and growth have been normal since. Hyperreninemia and electrolyte imbalance in urine persisted until the age of 3.5 years but then surprisingly fully normalized. That's why all medication was stopped at the age of 4.5 years. With the exception of a recurrence of mild plasma renin activity increase electrolyte and acid-base balance have remained normal.
Conclusion: This girl's history and laboratory findings were pathogonomic for the neonatal Bartter's syndrome. The evolution is quite unexpected and surprising but known from other rare reports in literature. One hypothesis might be, that the kidneys of such patients find a way to compensate for the underlying defect.

P 66

Craniofrontonasal syndrome and cor triatriatum – one pathogenesis of two rare disorders?

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Background: Craniofrontonasal syndrome is an X-linked development malformation syndrome with variable phenotypic expression. It affects females more severely than males which is quite unusual for x-linked genetic disease. The underlying cause is a mutation in the EFNB 1 gen encoding for the transmembran protein ephrin –B1. This gen regulates axon guidance, cell migration and adhesion in the developing brain. Moreover the Ephrin B family proteins plays also an important role in the formation of blood vessels.
Method: Case report.
Results: Our female patient was born at 40 5/7 wks of gestation after a normal pregnancy, BW 3150 g, APGAR 9/10/10. Dysmorphic features were brachycephaly and coronal craniosynostosis with frontal bossing, microcephaly, hypertelorism, flat and broad nose with a vertical groove on the top of the nose, highly arched palate, low posterior hairline, broad first toes with a gap between the first and second toes and longitudinal splitting of the nails. Cerebral ultrasound and MRI demonstrated an agenesis of the corpus callosum. Echocardiography showed a classic cor triatriatum with a diaphragma which subdividing the left atrium in a proximal and a true distal chamber and an atrial septal defect between the right atrium and the distal chamber. No signs of pulmonary veinous obstruction nor pulmonary hypertension were present. The detection of the heterocygote mutation in EFNB 1 gen c.196C>T/p.Arg66X confirmed the diagnosis of a craniofrontonasal syndrome. The parents are not affected. Currently the girl is developing well but multistage surgery for the heart and the craniosynostosis will be planned.
Conclusion: To the best of our knowledge it's the first description of a craniofrontonasal syndrome with a cor triatriatum. A potential link between these two conditions is the role of the function of the mutated Ephrin B.

P 67

Follow-up of a newborn with coronary artery fistula (CAF)

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Introduction: Although coronary artery fistula (CAF) is a rare anomaly, present in 0.2–0.4% of patients undergoing catheterization, it is the most common significant coronary anomaly. In 50–60% they origin from the right coronary artery, usually drainage is to the right side of the heart (up to 90%). In about 35% they are associated with other cardiac anomalies. Usually, patients remain asymptomatic until the second or third decade. The recommendations regarding timing and kind of intervention for CAF remain controversial.
Case: In a 3-week-old newborn with a continuous heart murmur a patent arterial duct was suspected. On the initial echocardiography a CAF was found, showing a dilated left circumflex coronary draining into the right ventricle. On CW-Doppler there was a LR-shunt with a maximal gradient of 45 mmHg. Initially and on follow-up; the ventricles showed a normal function (EF 70%) and no dilatation. Moreover, no signs of ischemia were seen on the ECG and values of troponin T (<0.010, normal range <0.010 µg/l), BNP (<5, normal range <80 pg/ml) remained normal. Therefore, we decided not to intervene but to follow the baby with ECG and echo. The patient had no signs of heart failure and showed a normal weight gain in 11 month of follow-up.
Conclusion: From the first surgical correction of this malformation, performed in 1947 surgical techniques have evolved. However, in the pediatric population the decision when to intervene remains difficult. Our patient was evaluated for signs of myocardial ischemia and volume overload, as these are the most frequent complications in the long term. Weighing the risk of an intervention and the spontaneous course we decided to follow our young patient and to intervene in the case of first signs of ischemia or dysfunction.

P 68

Bronze baby syndrome in severe hemolytic disease of the newborn

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Introduction: The bronze baby syndrome is an uncommon side effect of phototherapy mainly described in neonates with an elevated conjugated bilirubin, possibly by transformation of Cu(II)-protoporphyrin. A similar photosensitivity has been reported in severe haemolytic disease of the newborn (HDN) because of transient neonatal porphyrinemia.
Case report: We report the case of a baby born from a first pregnancy in a Rh- mother known for RhD immunization. Because of signs of severe fetal anemia, an intra-uterine transfusion was performed at 33 weeks of gestation and the baby was delivered a week later. He was icteric at birth with a highly positive Coombs test and a total bilirubin of 215 umol/l. Phototherapy was started but the bilirubin decreased only transiently and the baby showed signs of severe hemolysis with clinical instability, low platelets, elevated carboxyhemoglobin and marked extramedullary hematopoiesis. An exchange transfusion was performed, but the bilirubin continued to increase, up to a maximum of 425 umol/l on the 3rd day of life. The procedure needed to be repeated for a total of 6 times (exchanging 15 times the circulating blood volume) until the bilirubin finally stabilized on the 7th day of life. Moreover, the phototherapy had to be stopped after 24 hours because of a purple coloration of the exposed skin, evolving later to a second degree burn. We attributed the need for repeated exchange transfusions to the fact that a major part of the bilirubin, antibodies and red cell precursors were out of reach in the extravascular compartment. As for the bronze baby reaction, the conjugated bilirubin was within normal limits, but this phenomenon was described in case reports of severe HDN, the massive extramedullary hematopoiesis leading to a transient neonatal porphyrinemia. Porphyrins assays confirmed this diagnosis. The skin lesions were treated as burns with a favourable evolution and the porphyrins decreased over the three following months.
Conclusion: Severe HDN can lead to a bronze baby syndrome when exposed to phototherapy.

P 69

Patient-derived leukemic blasts retain their leukemogenecity and induce potent donor-specific NK cell immunity in a "humanized" NOD/SCID/IL2Rγ^{null} xenotransplantation model

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Xenotransplantation models using the engraftment of human leukemic blasts or hematopoietic stem cells in the highly immuno-permissive NOD/SCID/IL2Rγ^{null} (NSG) mouse strain have been instrumental to establish new models in experimental allogeneic hematopoietic stem cell transplantation (HSCT). However, preclinical models that might allow the exploration of graft-versus-leukemia (GvL) effects in a donor-patient-specific context are missing. To establish and validate a patient-specific xenotransplantation model of human leukemia, we first engrafted 18 different bone-marrow derived B-ALL, T-ALL and AML samples from pediatric patients into NSG mice and compared their leukemogenic profile with the original leukemia. Following engraftment, NSG-derived blasts resembled leukemic cells of the patient with respect to phenotype, chromosomal aberrations, transcriptome and minimal residual disease (MRD) marker expression. More importantly, the extent of leukemic engraftment at 10-20 weeks post transplantation highly correlated with the prognosis of overall survival of pediatric B-ALL patients. To enable studies on natural killer (NK) cell-mediated, donor-patient-specific GvL effects, we subsequently transplanted NSG-derived leukemic blasts into NSG mice previously engrafted with stem cells from either killer immunoglobulin-like receptor ligand (KIRL)-mismatched or matched donors and could demonstrate a donor-patient-specific GvL-immunity to KIRL-disparate leukaemias. Thus, we here depict a complex *in vivo* model that will substantially improve the future development of highly individualized, patient-tailored treatment strategies in allogeneic HSCT.

P 70

The skin, the visible omen

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Introduction: Cutis marmorata et telangiectatica congenita (CMTC) is a rare skin disorder which commonly presents directly after birth [1] with reticulate erythema, telangiectasia, skin atrophy and/or ulceration. It is often accompanied by further anomalies. The etiology is currently not known, but body asymmetries, orthopedic disorders, glaucoma and in particular venous malformations need always to be excluded.
Case report: We present a male newborn infant, born by cesarean section for maternal placenta increta, at 36 1/7 weeks of gestation, who presented with respiratory distress syndrome within the first hour of life. First adaptation was good, with APGAR scores of 7/9/9 at 1, 5 and 10 minutes respectively. His skin was eye catching mottled, but was not interpreted as pathologic. After the first hour of life, however,

skin lesions, clearly consistent with CMTC, presented mainly localized on the left lower limb up to the left hemi-thorax with slight skin alterations on the right forearm and hand. Because of worsening respiratory distress syndrome and increasing needs in oxygen CPAP support was initiated at 5 hours of age. The chest radiograph was consistent with mild acute respiratory distress syndrome of the newborn. Echocardiography revealed an irrelevant persistent ductus arteriosus and a slightly widened v. cava superior. Blood pressure was adequate for gestational age. Laboratory results showed neither signs of inflammation nor other disorders. Sonography of the brain and abdomen were normal. The infant was extubated within 24h of life and the need of supplemental oxygen persisted until the third day of life. Because he was found to be sweating and his oxygen requirements increasing, we repeated the echocardiography. Total anomalous pulmonary venous return of the non obstructive type was visualized and confirmed by heart catheter investigation.
Discussion: Whether CMTC is an isolated dermatological condition or rather a syndrome with associated anomalies is still under debate [2, 3]. Currently the etiology is not known for this disease but a failure of the development of mesodermic vessels during the early embryonic stage [2] is posited in the literature in conjunction with a genetic predisposition. In our case of CMTC accompanied with the total anomalous pulmonary venous return and the "vessel misplacement" in the placenta increta might corroborate these two theories.
Conclusion: Our case illustrates the need for careful investigation of newborns with CMTC. The question, whether there is a common pathophysiological origin of the different diagnosis so far communicated with CMTC, needs to be researched further.

1 Way, et al. Cutis marmorata teleangiectatica. J Cutan Pathol. 1974.
2 Amitai, et al. Cutis marmorata teleangiectatica congenital: Clinical findings in 85 patients. Ped Dermatol 2000.
3 Kienast, et al. Cutis marmorat teleangiectatica congenital: a prospective study of 27 cases and review of the literature with proposal of diagnostic criteria. Clin and Experiment Derm 2009.

P 71

Delayed gene expression modifications in a newborn rat ventilation model of brochopulmonary dysplasia

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Introduction: Bronchopulmonary dysplasia (BPD) is a chronic lung disease affecting preterm infants with potential long term respiratory sequelae. Among its risk factors are premature birth, mechanical ventilation, hyperoxia and inflammation. To better understand the different molecular pathways involved in this disease, we used a newborn rat ventilation model with orotracheal intubation to study delayed gene expression modifications after extubation and a ventilation-free interval of 48h.
Methods: Systemic inflammation, often associated with premature birth, was induced by LPS injection intraperitoneally (2 mg/kg) to male rat pups on postnatal day 4 or 5. Twenty-four hours later they were anesthetized, intubated and ventilated with tidal volume of 15 ml/kg and 21% or 60% O_2. After 6h of ventilation, isoflurane-anesthesia was weaned, and when showing an efficient spontaneous respiration, they were extubated and replaced with their mothers. After a 48h ventilation-free period, gene expression was measured in their lung tissue separately in 4 groups by Affymetrix® Gene-Arrays and verified by qPCR.
Results: Expression changes were mainly found in genes involved in inflammation and extracellular matrix remodeling. Among them MMP-9 gene and several of its main regulator genes were significantly modified (table).

qPCR fold changes				
	Group I Ctrl	**Group II LPS**	**Group III LPS+MV+ 21%O_2**	**Group IV LPS+MV+ 60%O_2**
MMP-9	1	1.39 ± 0.22*	1.39 ± 0.22*	1.64 ± 0.36*
CAMP	1	2.22 ± 0.50*	2.04 ± 0.46*	2.78 ± 0.68*
NGAL	1	1.84 ± 0.08*	2.14 ± 0.21*	2.14 ± 0.20*
NP4	1	4.12 ± 0.19*	3.28 ± 0.17*	3.29 ± 0.23*
Values are expressed as mean ± SD from 9 animals/group. *: p <0.05				

Conclusion: MMP-9 gene so far known as an important factor involved in lung development, angiogenesis and tissue repairing showed in this model an enhanced expression in lung tissue of rat pups exposed to an accumulation of risk factors for BPD, even 48h after the end of mechanical. Additionally, the expression of other genes enhancing MMP-9 efficiency was also upregulated. Therefore, the role of this molecule and its regulators in the pathophysiology of BPD deserves further investigation.

P 72

Regional Perinatal Network of Lausanne: Epidemiology and Characteristics of Neonatal Transport

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Introduction: The perinatal network of Lausanne, created in the 1960s, can be seen as an example of regionalised perinatal care. It consists of a single tertiary referral centre at the CHUV, englobing both obstetrics and neonatology with its neonatal intensive care unit, ten level II units: Aigle, Morges, Nyon, Payerne, Vevey, Yverdon (Canton Vaud), La Chaux-de-Fond, Fribourg, Neuchâtel, and Sion in neighbouring Cantons and three level I units: two in Lausanne (private clinics) and one in Canton Fribourg (Riaz). A population of about 1.2 million is covered, with 12 000–14 000 births/year. This study investigated the transport activity and the profile of transported neonates over one year (2008).
Methods: This is a retrospective analysis of prospectively collected data on transported neonates regarding administrative and medical information, transport, and timing.
Results: Of the 565 patients admitted to the NICU, 31% (n = 175) were outborn and transported postnatally as emergencies to the CHUV, and 43.4% (n = 245) were retransfered to a peripheral hospital close to the parents' home after initial stabilisation at the NICU. Reaction time (from initial call to arrival at bedside) shows a median of 55 min (range 18min–4h15). 45% of the transported patients weighed ≥3000 g. Of all emergency transports 60.0% concerned term neonates, and 10.9% preterm <32 weeks. Among pathologies inducing transport, respiratory problems arose in 66.9%, complicated prematurity (29.1%), cardiovascular instability (19.4%) and infections (16.6%). Most frequent respiratory diagnoses included wet lung (28.2%), aspiration syndrome (15.4%), hyaline membrane disease (13.7%), pulmonary infections (12.0%) and asphyxia (9.4%).
Conclusion: The high number of retransfers out of the CHUV which outweighs the number of transfers to it reflects that the perinatal network ensures a good turnover as required in a regionalised system. The leading cause for emergency neonatal transfer to a higher level of care are respiratory issues. Further attention should be given to early recognition of risk factors for respiratory distress, in order to anticipate transport, ideally before birth.

P 73

Accessory mamilla and areola in childhood – diagnostic challenge and urologic marker lesion: case report from the Hochgebirgsklinik Davos

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Background: Different cutaneous lesions may mimick accessory mamilla and areola, hereby causing problems in establishing the proper diagnosis. Reports mention an association of accessory mamilla with urological disorders.
Case report: A 14-year-old boy showed on his right ventral trunk, 5 cm below of his right mamilla, a well-defined brown macule (diameter 5 mm) with a central papule. The lesion was first noticed after birth and increased slowly according to general growth. Medical history, clinical examination, laboratory analyses as well as ultrasound examination were uneventful in regard to disorders of kidney and urinary tract.
Discussion: Girls as well as boys may show mamilla, areola, and/or breast tissue on the milk line, which extends from the axillar region to the groin region. Rarely, other areas like neck, back and extremities can be affected. Accessory mamillas can be found in 0.2 to 6.0% of the general population, affecting more males and more often the left side of the body (Schmidt H. Eur J Pediatr 1998;157:821–3). Ferrara et al. (Scand J Urol Nephrol 2009;43 47–50) reported recently an association with renal malformation (>18%), pyelectasis (>16%), nephrovascular hypertension (>16%), proteinuria (10%) and other disorders. Differential diagnosis of accessory mamilla and areola includes nevus, histiocytoma, verruca vulgaris, and mastocytoma.
Conclusion: If a small-sized brown lesion is detected along the milk line, accessory mamilla and areola should be considered among other cutaneous disorders. In any case of accessory mamilla and areola, an urological screening for disorders of kidney and urinary tract seems appropriate.

P 74

Parental involvement in weight management programs for obese teenagers: Results of a qualitative study

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Introduction: Family dynamics are strongly associated with teenagers' obesity. Our aim was to assess parents' perceptions before and at the end of a six month Family-Based Behavioral Treatment (FBBT) program for obese teenagers.
Methods: We conducted a qualitative research including 13 parents in two focus groups, one at the beginning and one at the end of a FBBT program for obese teenagers (12–15 years). Four open ended questions were used to explore parents' perceptions: 1) What do they think about their parents 'participation in the program? 2) How do they live their child's obesity? 3) How are family interactions? 4) What can they say about their child's well-being? Sessions were recorded, transcribed and content analysis was performed.
Results: Preliminary findings indicated that parents felt isolated in regards to their teenager's obesity. They were scared about psychological and somatic consequences: "it is a lot of suffering," "it is very serious, and we pass near death, even if we do not think of that." Meeting other parents and sharing their difficulties and tiredness related to this chronic condition helped them to decrease guilt: "it's tiring, we tried all techniques: hiding, being nice, being mean, and blackmail," "it's been six months since I lock all the food." Parents' interaction raised questions on family functioning, bringing less attention to the food or diet of their child. Group discussions made them aware of the importance of family involvement in the weight management process. Final results will be presented.
Conclusions: Weight management programs for obese teenagers should include parents to relieve their distress of having an obese child, modify family's interactions and enhance teenager's autonomy and self-efficacy.

P 75

Fertility Preservation in Female Paediatric Cancer Patients, A Multidisciplinary Approach

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Introduction: Major advances in the treatment of childhood cancer have resulted in a 5-years survival rate of about 78%. Unfortunately, childhood cancer treatments often include chemotherapy (alkylating agents), surgery and radiotherapy which may adversely affect fertility. Of all cancer survivors 15% will have compromised reproductive function. Different fertility preservation options (FPO) are available for pubertal patients (sperm banking for males and cryopreservation of embryo/oocytes for females). Recently, ovarian tissue cryopreservation (OTC) has emerged as a new experimental option. This technique, available at CHUV and HUG, is proposed to pubertal and prepubertal females. Theoretically, two different options are available once the patient is ready to conceive: ovarian auto-transplantation (experimental method, feasible today) or in vitro maturation of primordial follicles followed by in vitro fertilization (still in research, not feasible nowadays).
Methods: In 2010, two multidisciplinary pediatric teams in CHUV and HUG were formed under the auspices of the "Reseau Romand de Cancer et Fertilité". Each team consists of paediatricians specialized in oncology, gynaecology, endocrinology, surgery, psychiatry, reproductive medicine, radiation oncology, adolescent medicine and genetics. Monthly video-conference meetings are organized during which new oncological cases are presented, with the objectives of determining 1) indications for fertility preservation and 2) the best FPO for the patient. Meetings also provide an opportunity to discuss guidelines, literature, difficult cases and to set up a patient registry to follow up the outcome on the long term.
Results: To date, 9 patients were presented and 3 OTC were realized.
Conclusion: Fertility preservation techniques are now available also for pubertal and prepubertal females and continue to develop. The opportunity to provide accurate information and hope for future fertility in young cancer patients is invaluable. In view of the paucity of cases, only unification of medical forces will enable us to offer for each of our patients the best possible care.

P 76

Determinants of Hepatitis A Vaccine Immunity in the Cohort of Human Immunodeficiency Virus-Infected Children Living in Switzerland

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Introduction: Vaccination in HIV-positive children is often less effective. The goal of this study was to assess the efficacy of hepatitis A virus (HAV) vaccine in children infected with HIV.
Methods: Children, part of the Swiss Mother and Child Cohort Study, were enrolled prospectively. Recommendations for initial / catch-up / additional HAV immunization were based upon baseline antibody titers and vaccine history. HAV IgG antibodies were assessed by ELISA and a protective cut-off value was ≥10 mIU/ml.
Results: Eighty-seven patients were included in the study (median age 11 years, range 3.4 – 21.2) between June 2006 and August 2007. Forty-five (51.7%) patients were seronegative for HAV. Vaccine responses were assessed after the priming dose in 29/35 naïve patients, and after the booster dose in 33 children. Anti-HAV geometric mean titers were 962 mIU/ml after boosting dose. Seroconversion was 97% after 2 doses of vaccine. No factor influenced significantly HAV vaccine priming response. However, baseline CD4 T cells count <750 cells/ml decreased significantly booster response (P = 0.005).
Conclusion: Despite a high rate of seroconversion, patients with low CD4 counts had low anti-HAV antibody titers, possibly predicting a shorter protection time. Thus, antibody titers monitoring and additional vaccine doses may be necessary in HIV-positive children.

P 77

Critical incidents management: when adminstration's greed adds holes in the swiss cheese

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Introduction: Avoiding medical errors is one of the greatest and most difficult challenge facing medical doctors. We here describe a patient that failed adequate medical supervision because of unusual administrative procedure, leading to a delay in a proper diagnosis and treatment.
Case report: A 2-year-old Pakistanese child, traveling with his family in Switzerland, presented in our emergency department with three days of fever (39.7 C), loose stools and one episode of vomiting. Physical exam was normal. Complete blood count (CBC) showed no leucocytosis and no left shift. CRP was 59.3 mg/l. Other tests, including malaria test, were non contributive. He was sent home with a diagnosis of probable viral gastro-enteritis, with a follow up appointment the next morning. On that day, the status was unchanged, and CBC was drawn again. There was a striking 35% left shift on CBC that went unoticed, and the child was sent home again. Because of continuing fever, the child was finally admitted to another hospital where correct diagnosis and treatment were given. With antibiotics, he made a full recovery.
Discussion and conclusion: Evaluation of medical emergencies in children is notoriously difficult, especially for junior residents. To avoid such errors, all pediatric patients encounters are reviewed by a senior physician in our department. But in our case, we discovered that the medical charts of foreign patients, for some obscure administrative reasons, were immediately filed after cash paiement, therefore escaping the review of the senior doctor in charge. This led to the abnormal values not being noticed by the resident in charge, therefore delaying the appropriate treatment. Internal procedures were changed immediately, to avoid such occurrence in the future: immediate alert of abnormal laboratory results, and review of *all* charts, regardless of the cumbersome administrative procedures.
It is extremely worrysome and ethically concerning that the administrative point of view could have taken priority on the medical point of view. Money is necessary for hospitals, but patients' life and security should stay the absolute priority!

P 78

Critical incidents management: how to improve the Guthrie test's fishing net?

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Introduction: In 24 months we identified in our regional hospital (900 live births annualy) 4 occurrences of failed universal neonatal screening (Guthrie test (GT)), involving 6 newborns. Given the potential dramatic consequences, we tried to identify the factors that

led to these critical incidents and establish a new internal procedure to avoid such errors.
Description: Failed neonatal GT were defined as follow: 1) complete absence of the GT, 2) GT performed outside the normal time frame, or 3) GT performed improperly. We encountered one occurence where 3 GT cards were mistakenly sent 2 weeks later, therefore delaying any benefits of a potential treatment. In 2 other cases, the resident assumed that the GT had been previously performed in the transferring hospital, as stated in the transfert medical letter (inter-hospital transfert). In the last case, the resident wrongly assumed that a patient transferred from nursery to the pediatric department had his GT done in the nursery (intra-hospital transfert).
Discussion and conclusion: Assuming that GT has already been performed, during inter- or intra-hospital transferts represents the most common cause of failed neonatal GT in our experience. In order to tighten the procedure safety, we have created a new algorithm. It is now mandatory for the resident to obtain the GT results for any newborn who did not have his GT performed on the pediatric ward. Moreover, a control has been introduced to ensure that the GT cards are sent the same day they are performed. We suggest that every unit dealing with neonates should review their procedures to improve their safety net procedures and avoid such pitfalls, especially during transferts.

P 79

ABCB1 gene polymorphisms and neuropsychiatric adverse events in oseltamivir-treated children during influenza H1N1/09 pandemia: a cohort study

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Introduction: Oseltamivir, a substrate of P-glycoprotein (P-gp), has been associated with neuropsychiatric adverse events (NPAE) in children. We examined the safety profile of oseltamivir and evaluated the impact of P-gp *(ABCB1)* polymorphism on the incidence of NPAE in oseltamivir-treated children during the H1N1/09 pandemia.
Methods: This prospective cohort study was conducted between October 1st, 2009 and January 31st, 2010. All children <18 y.o. presenting at the emergency department with a flu-like illness were eligible for inclusion. Adverse events were systematically recorded using a diary card, with a 30 days follow-up. Causality of oseltamivir in NPAE was performed by pharmacologists. Patients were genotyped for *ABCB1* C3435T and G2677T/A polymorphisms.
Results: Among the 42 H1N1-infected, oseltamivir-treated children who were genotyped for *ABCB1* 3435C>T and 2677G>T/A variants, 36% presented NPAE possibly related to oseltamivir. Frequency of NPAE displayed a "genotype-trend effect" with the mutant and the wild type subgroups at the two fare ends. Eleven percent of the 2677GG-3435CC individuals (wild-type homozygotes) presented NPAE, compared to 39% of the individuals being heterozygote for at least one variant allele and 67% of the 2677TT-3435TT individuals (mutant homozygotes).
Conclusion: A higher incidence of NPAE in oseltamivir-treated patients than previously reported. This could be due to the fact that we actively sought NPAE. Diplotype 2677TT-3435TT might increase the patient's vulnerability to NPAE, maybe as a result of an enhanced permeability of the blood-brain-barrier to oseltamivir.

P 80

Case report: Lethal outcome of persistent pulmonary hypertension in a newborn

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Introduction: Persistent pulmonary hypertension of the newborn (PPHN) occurs in 1–2 newborn per 1000 live birth. The majority suffers from secondary pulmonary hypertension caused by congenital heart disease or lung disease. Rarely PPHN origins from a primary cause such as lymphangioleiomyomatosis or pulmonary alveolar proteinosis (PAP). We present a case of a primary pulmonary hypertension caused by PAP.
Case report: Our patient was born at term and adapted well after uncomplicated pregnancy. After 4 hours he progressively developed respiratory distress, and was intubated. Chest radiography revealed bilateral pneumothoraces and chest drains were inserted. White cell count with differentiation and CRP were normal. Echocardiography showed a severe pulmonary hypertension with structurally and functionally normal heart. Because of persistent oxygenation problems inhaled nitric oxide (NO) was started. In the further course therapy with sildenafil, prostacyclin inhalation and different ventilation mechanisms were started without success. In CT-scan diffuse ground-glass opacity was seen corresponding to hyaline membrane disease, consistent with PAP. Because of rapid worsening of the respiratory situation and various unsuccessful treatments, therapy was withdrawn after 3 weeks and the patient was extubated. He died 2 hours later. Histopathological examination confirmed pulmonary abnormalities corresponding to pulmonary hypertension such as thickening of the small arterial vessels by smooth muscle hyperplasia. Electron microscopy findings showed evidence for alveolary proteinosis. Different mutations of the surfactant metabolism have been ruled out.
Conclusion: Pulmonary alveolar proteinosis is a rare disease with excessive accumulation of lipoproteinaceous material within alveoli. Clinical features are highly variable. Four different types are described. Beside secondary forms (infection, tumor), there are acquired forms in adults, idiopathic and congenital forms. In the latter different mutations of the surfactant metabolism are known. Two of the congenital forms present an early onset with respiratory failure in neonates refractory to any treatment. In our patient clinical presentation, histological and radiological findings are consistent with the diagnosis of congenital or idiopathic pulmonary alveolar proteinosis.

P 81

Infants with just another viral infection? Don't forget SCID!

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Introduction: Despite common occurrence in children, viral infections can sometimes be the presenting feature of immunodeficiency disorders. We report a patient with recurrent and persistent viral infections caused by a Severe Combined Immunodeficiency (SCID) syndrome and revealing the skeletal dysplasia Cartilage-Hair Hypoplasia (CHH)
Case Report: A four-month-old infant was admitted with hypoxic superinfected bronchiolitis and failure to thrive. The evolution was atypical with 25 days of oxygen supplementation before discharge. He was readmitted 4 days later with a sepsis-like clinical picture associated with *Rotavirus* gastroenteritis. He then developed respiratory symptoms again with need of CPAP and oxygen therapy. The nasopharygeal secretions showed persistence of *RSV*. The bronchiolitis was complicated with a bacterial superinfection. One week later a *Parainfluenza 3* virus was isolated in the nasopharygeal secretions, coinciding with a new respiratory deterioration. An immunodeficiency disease was suspected on the base of the recurrent viral infections with atypical evolution and failure to thrive. Laboratory investigations showed lymphopenia (below 3.154 G/L) and hypogammaglobulinemia. Further immunologic investigations revealed a T-B+NK+ SCID syndrome and bone Xrays were suggestive of CHH which was molecularly proven by RMRP gene analysis.
Discussion: Infants affected by SCID commonly present within the first six months of life with recurrent or persistent infections, e.g. gastroenteritis (*Rotavirus, Adenovirus, Enterovirus*. etc), pneumonia (*Pneumocystis Jivrovecii, RSV, Parainfluenzavirus*, etc), bacterial sepsis and failure to thrive. SCID patients are usually lymphopenic (<2 G/l). The different SCID syndromes are treated effectively by hematopoietic stem cell transplantation which is in preparation for this patient.
Conclusion: Don't lose time and think SCID when facing an infant with persistent or recurrent episodes of viral infections with atypical evolution! Lymphopenia is a consistent feature. CHH is one classical condition associated with SCID.

P 82

Campylobacter jejuni infection with two types of Guillain Barre Syndrome

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Introduction: Guillain Barre Syndrome (GBS) is an immune mediated polyneuropathy with acute onset which often follows an infection. The diagnosis relies on clinical presentation, cerebrospinal fluid (CSF) analysis and electromyography. Miller Fisher syndrome (MFS), a rare variant of GBS, is a descending paralysis that proceeds in the reverse order of GBS. It presents with the triad of ophtalmoplegia, ataxia and areflexia.
Case report: Case 1, a 15 ys old presented with progressive flaccid paralysis of upper and lower limbs and **case 2**, another 15 yrs old boy with unsteady gait, dysarthria and bilateral palpebral ptosis. In both patients, a complete workup including cranial CT yielded negative findings. CSF analysis revealed in the first case normal protein level and cell count and in the second case a cyto-albuminological dissociation. Both had positive stool culture for Campylobacter jejuni.

Both were treated with IVIG for 5 days followed several days later by gradual improvement. Despite normal CSF, case 1 was diagnosed as GBS because of the pertinent clinical history and the electromyography result. Case 2 was diagnosed as MFS because of the clinical history and the positive immunological test for ganglioside's antibodies (GQ1b and GM1).
Discussion: GBS should be suspected by clinical presentation. CSF analysis is not always relevant, it may be normal in the early phase and shows elevated protein level in 90% of patients at clinical nadir and in 50% of MFS cases. Most cases occur weeks after an infection. Campylobacter jejuni, frequently associated with these diseases, is known to have similar epitopes as the gangliosides (GM1, GD1, and GQ1b). Anti-GQ1b antibodies are present in 95% of acute MFS. Although MFS is generally a benign, self-limiting condition with almost all patients return to normal activities within 6 months, GBS has a serious long-term impact and recovery is slow and may take years.

P 83

Mitochondrial disease due to TMEM70 mutation in a neonate with early onset hypotonia

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Introduction: Mitochondrial diseases constitute a heterogenous group of disorders. mtDNA and nDNA mutations, may affect the energy producing capabilities of the mitochondrion. A relatively recent discovered mutation at the TMEM70 gene of nDNA is responsible for the diminished production of mitochondrial ATP synthase (complex V) [1]. The phenotype exhibits common traits like hypotonia, hypertrophic cardiomyopathy (HCMP) and hypospadias, with variable phenotypic severity in the newborn period.
Case report: We present a male neonate with IUGR, hypotonia, hypospadias,systolic heart murmur, right inguinal hernia and cryptorchidism. Laboratory findings included, 3-MGC-uria elevated γGT and. metabolic lactic acidosis at the age of 5 months. Skin biopsy demonstrated diminished activity (<30%) of complexV (ATP synthase). Molecular analysis showed homozygous TMEM 70 mutation in the patient, while the parents were heterozygous for the same mutation. A significant lag in weight development (P <3) was observed, despite continuous feeding by nasogastric tube and high caloric intake. Neurological evaluations showed tonus disturbances with irritability, difficulties with visual fixation and diverging strabism. Hypertrophic cardiomyopathy was diagnosed at the age of 5 months and treated with propranolol.
Discussion: TMEM70 mutation [1] should be considered in the diagnosis of neonates with IUGR, early onset muscular hypotonia, facial dysmorphy, hypospadias and lactic acidosis. Cardiological follow-up is necessary, as HCMP is a major limiting factor to the life expectancy of these patients. Blood gas, lactate, ammonia in blood and organic acids in urine, represent simple biochemical screening tests for metabolic disorders and are indicated in all newborns with unexplained illness, even in presence of minor malformations.

[1] Honzik, et al. Arch Dis Child. 2010;95:296–301.

P 84

Relapsing post-streptococcal reactive arthritis and/or vasculitis: report of two cases

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Introduction: Acute Rheumatic Fever (ARF) has dramatically declined in North America and Western Europe since the end of World War II. Post-streptococcal reactive arthritis (PSRA) and post-streptococcal vasculitis (PSV) are more frequent complications of streptococcal throat infections but their relapsing pattern is not always recognized by pediatricians.
Case report: 1. A 6 years old boy presented with 2 episodes of right foot arthritis and one episode of finger inflammation within one month. He complains also of hip pain. He had fever, elevated ESR and positive streptococcal throat swab with elevated ASLO titers (first episode: 2460 UI/ml; second episode: 3520 UI/ml). Cardiac investigations were normal. He was treated by antibiotics and ibuprofen with resolution of the symptoms. 2. A 9 years old boy presented with dysphagia and, after a few days course, with fever and diffuse arthralgias and myalgias. He had also a vasculitis of the right hand. Laboratory showed an elevated ESR and very high ASLO titers (5.634 UI/ml). Despite successful treatment with penicillin and steroids, he relapsed 15 months later, as steroids were stopped, with myalgias and a very painful vasculitis of the malar area. ASLO titers were still elevated (693 UI/ml).
Discussion: Both patients presented after a documented streptococcal infection with symptoms mimicking Jones' criteria for ARF but with atypical joints involvement and without cardiac lesions. Nevertheless, the medical literature reports a few patients developing cardiac lesions, raising the question of an antistreptococcal prevention by administration of a long term penicillin prophylaxis. Those with relapsing disease may also benefit of such a treatment as it was done in our second patient.
Conclusion: Post-streptococcal reactive arthritis, as vasculitis, may show relapsing patterns, even years after the first episode. This entity must be known by pediatricians as well as the fact that severe forms may improve rapidly following corticotherapy.

P 85

Acrodynia during hyperthermia

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Introduction: Acrodynia is mostly known for mercury intoxication. Nevertheless, even if the etiology is not well understood, viral infections and inflammatory diseases are thought to play also a role in the pathogenesis. The association with hyperthermia raises the probability of an inflammatory cause such as vasculitis.
Case report: A 13 years old boy came to the emergency room because of intense pain in the fingers and toe's extremities with fever since a week. The pain appeared and worsened with increasing of the temperature. During these crises, he was lethargic, complained about headache, weakness in the extremities and was unable to walk. On clinical examination, the patient presented swelling of the face and purpuric lesions on the extremities, which were warm but without redness. The pain was not relieved with paracetamol, NSAID nor morphine treatment. On the other hand, the patient recovered within several hours under corticosteroid. The laboratory exams showed ESR 64 mm/h and CRP 45 mg/l. Search for bacterial, viral infections and auto-immunity disease were negative. Capillaroscopy, doppler ultrasound and Allen test were also normal.
Discussion: Such clinical case with hyperthermia and pain in the extremities is a challenge for the paediatrician. Sometimes, the exact etiology remains unknown, as for our case and the evolution may help to precise the diagnosis. Nevertheless, inflammatory diseases as CAPS (Cryopyrin associated periodic syndrome) and metabolic diseases as Fabry's disease must be excluded. In inflammatory diseases, the second step of the therapy (after the NSAID), is the corticotherapy. When this doesn't work, immunosuppressive drugs such as azathioprine and cytostatic drugs such as methotrexate must be used.

P 86

Congenital dacryocystocele treated by fiberoptic endonasal dacryocystotomy

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Background: Dacryocystocele is an uncommon consequence of congenital nasolacrimal duct obstruction. The classical clinical presentation is a bluish, cystic, firm swelling below the median canthus. Two main complications can occur in neonates with dacryocystocele – severe infection and respiratory compromise due to nasal obstruction. Uncomplicated dacryocystocele is primarily treated conservatively with gentle massage on the lacrimal sac to promote spontaneous rupture. Irrigation of the nasolacrimal system and probing or silicon intubation of the nasolacrimal duct are recommended ophthalmologic interventions, if conservative treatment fails. Endoscopic endonasal probing together with surgical marsupialization of the cyst has also been described in the literature. We describe simple dacrycystotomy by nasal fiberoptic endoscopy for successful treatment in 3 newborn infants.
Case Report: We present a case of a healthy full-term born boy with a bluish swelling at the medial corner of the right eye at third day of life. Ultrasound examination confirmed a cystic lesion compatible with a dacryocystocele. After ophthalmological consultation conservative treatment consisting in local massage was started. At the 7th day of life an increase in swelling and local inflammation was observed. Probing of the nasolacrimal duct by an ophthalmologist was unsuccessful and systemic and local antibiotic therapy was started. Since spontaneous rupture did not occur and local inflammation progressed, fiberoptic endonasal dacryocystotomy was performed on the 9th day of life. Antibiotic treatment was continued for one week despite complete disappearance of all symptoms after the intervention. The subsequent clinical course was favourable. The same procedure was previously performed with the same result in two identical cases.
Conclusion: Management of congenital dacryocystocele differs greatly in literature. We report our experience with simple endoscopic fiberoptic endonasal dacryocystotomy as a successful treatment option in congenital dacryocystoceles.

P 87

The Australian Triage Score (ATS) – Impact on waiting time in our emergency department

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Introduction: The number of consultations in the emergency departments (ED) has increased worldwide over the past years. In response triage score systems were introduced in most pediatric emergency departments. Swiss pediatric emergency physicians opted for the Australian Triage Score (ATS), which was developed in EDs with up to 40'000 consultations/year. In contrast Swiss EDs are smaller and distances are shorter. The aim of this study was to examine the impact of the ATS on the waiting time.
Methods: During May 2009 data was prospectively collected through a parents based anonymous questionnaire at the ED of the Children's Hospital St. Gallen. From 600 questionnaires 320 were returned (response 53%). Aditionally, the nursing documentations from the same period were evaluated retrospectively (n = 1028).
Results: 320 questionnaires were included. A waiting time less than 30 min until the first physician contact was shown in 72% (ATS 2), 76% (ATS 3), 73% (ATS 4) and 61% (ATS 5). More than 60min waiting time for the first contact had 11% (ATS 2), 5% (ATS 3), 9% (ATS 4) and 9% (ATS 5). The time of day had no influence on waiting time. Waiting time had a strong influence on the satisfaction of the parents. 1028 nursing documentations were analyzed. Waiting time [mean (median)] to first physician contact or first drug administration was 15 (10) min (ATS 2), 21 (15) min (ATS 3), 27 (25) min (ATS 4), 34 (30) min (ATS 5). The waiting time was independent of time of day, surgical vs medical treatment or whether the patient was discharged home vs kept in hospital.
Conclusion: In our study, triage score had only a small influence on the waiting time. Overall, the waiting time was quite short - including patients with a triage score 4 or 5. Performing ATS properly is time-consuming for the nurse. Triage also leads to a delayed first physician contact. A simpler triage system would relieve the nurse and shorten waiting time. In Switzerland we have smaller EDs, shorter distances and more human resources compared with Australia. This should be considered for selecting an adequate triage system.

P 88

Parents' relationship with their child's provider: more than just the transition from pediatric to adult health care

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Introduction: Transition from pediatric to adult health care is a determinant period for adolescents with chronic diseases. Parents also have to go through multiple adjustments during this period. Knowing that they can influence transition success, parents are privileged partners for the health professionals. In this context, the aim of this study is to assess, in pre- and post-transition parents, the differences in their ease with their child's specialist provider, as a possible marker of transition success.
Methods: We divided parents in pre- (N = 311) and post-transition (N = 85) groups to compare their feeling at ease with their child's specialist provider controlling for adolescent's age, gender, recruiting hospital, specialty, illness status, disease severity, and the adolescents' ease with their specialist and readiness for transition. All statistically significant variables at the bivariate level were then included in a logistic regression.
Results: 87% of parents felt at ease with their child's specialist provider in the pre-transition group, whereas only 39% in post-transition (P <0.001). From the parents' point of view 86% of adolescents were at ease with their specialist in pre-transition, compared to 70% in post-transition (P = 0.001). The difference in parents' ease remained significant when controlling for potential confounders (Adjusted odds ratio: 0.206 [0.076–0.559]).
Conclusions: Parents in the post-transition group are significantly less comfortable with their child's specialist provider than those in pre-transition. Parents of chronically ill adolescents have to go through a double transition: health care transition occurring at the same time than their child's emancipation. This can be a source of anxiety for parents who may feel excluded from the care process. Confused feelings regarding the adult specialist and reactive overprotection of their child can preclude the transition process. Support and preparation of the parents, as well as an increased provider's awareness concerning this double transition could enhance transition success.

P 89

Cullen Sign in a 3-year-old child with acute severe pancreatitis

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Acute pancreatitis is known to be a possible complication of chemotherapy in children treated for acute lymphoblastic leukaemia. PEG L-Asparaginase, widely used today in induction and intensification of anti-leukemic therapy, is especially implicated to cause acute pancreatitis. We report the case of a 3-year-old patient with acute lymphoblastic leukaemia, who presented in severe neutropenia with fatigue and vomiting and subsequent circulatory and respiratory instability during his induction protocol on day 43 (according to ALL-BFM 2000 protocol IA) six days after stopping the prednisone and 17 days after receiving his second dose of PEG L-Asparaginase. On the second day of symptoms the boy developed periumbilical ecchymosis, the Cullen sign, which is not specific but commonly described in association with acute hemorrhagic pancreatitis in adults. There it is a sign for the expansion of the retroperitoneal inflammatory process and hemorrhage, occurs in about 3% of adult acute pancreatitis and is associated with a increased mortality. In this case the Cullen sign led to the diagnosis of acute severe exsudative pancreatitis and cardiopulmonary instability. To our knowledge this is the first report to describe the Cullen sign in a child. Clinical "textbook" signs, if known, can be very useful in guiding the diagnosis.

P 90

Interactive education for asthmatic children: Which effect on emergency visits and quality of life?

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Introduction: Many approaches of education about asthma exists but there is no evidence of their effectiveness because of the heterogeneity of practices. The aim of the study was to evaluate the impact of "Asthma School" (therapeutic education) on quality of life. We also assessed the number of medical visits in asthmatic children.
Methods: We included in a prospective longitudinal study every child aged 4 to 12 years old and their parents attending asthma school during one year and followed them at 6 months. Quality of life was evaluated through focus groups and the Pediatric Asthma Quality of Life Questionnaire (PAQLQ) at inclusion and 6 months after. Number of emergency visits, medical appointments and number of hospitalizations during the year before and after Asthma School were assessed with a questionnaire to the paediatricians.
Results: 27 children and their parents were included. Mean age was 7.02 years. Quality of life improved in all domains evaluated by the PAQLQ (score of 6.15 vs 6.78). Paediatric visits, emergency visits and hospitalizations decreased significantly between the year before and the year after Asthma School.
Conclusion: Interactive education about asthma improves clinically important outcomes and quality of life of children and their families.

P 91

MEN 2B – early recognition of this syndrome allows to prevent medullary thyroid cancer

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Background: Multiple endocrine neoplasias (MEN) include several distinct syndromes predisposing to tumors of endocrine glands, each with its own characteristic pattern. MEN syndromes are inherited in an autosomal dominant manner. MEN type 2 (MEN 2; incidence 1:35'000) is characterized by phaeochromocytoma, medullary thyroid cancer (MTC) and other manifestations. MEN 2B accounts for 20 % of all MEN 2 diagnoses. In MEN 2B diagnosis at-a-glance is possible because of its typical clinical phenotype and recognition pattern.
Case report: An 8-year-old girl presented with an enlarged thyroid gland without further clinical symptoms. In her personal history there were orthopaedic problems with a hypoplasia of the left lower leg. Additionally, the girl had a short stature and bowel complaints. Her phaenotype included a marfanoid habitus, prominent lips, small papules along the eyelids and nodules on the tip of the tongue. Blood tests showed a normal thyroid function and an elevated calcitonin, indicating C-cell hyperplasia. Ultrasound and MRI of the thyroid gland revealed bilateral tumors, the largest with 25 mm in diameter. Several cervical lymph nodes were enlarged. Since the clinical phenotype had been suggestive of MEN 2B, genetic testing for MEN 2B was performed. A mutation in the RET-protooncogene on chromosome 10q11.2 confirmed the diagnosis. This made MTC in association with MEN 2B most likely. Therefore total thyroidectomy and radical bilateral

neck dissection was performed. Histology revealed multifocal bilateral MTC and perifocal C-cell hyperplasia. Eleven out of 38 lymph-nodes were involved. In the post-operative controls calcitonin-levels rose slightly over the last 6 months. Follow-up MRIs have shown no local or systemic relapse so far.
Conclusion: We present a girl with marfanoid habitus, mucosal neurinomas of the tongue and eyelids, bone abnormalities and suspicion of intestinal ganglioneuromatosis. This typical phenotype leads to the diagnosis of MEN 2B syndrome. Early diagnosis is crucial, as all patients with MEN 2 develop MTC and preventive total thyroidectomy is indicated. In familial MEN 2B genetic testing is performed after birth and thyroidectomy is done by the age of six months. Further periodic controls are recommended for detection of phaeochromocytoma and relapse of MTC, respectively.

P 92

Screening for primary ciliary dyskinesia by nasal NO measurment

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Background: The prevalence of primary ciliary dyskinesia (PCD) is currently estimated at 1:20.000 to 1:60.000. However, recent data suggest that these figures are largely underestimated (Kuehni CE et al. 2010). Determination of nasal NO (nNO) has meanwhile been established as a non-invasive screening tool.
Methods: Between September 2010 and Mai 2011, 150 patients with chronic respiratory disorders were screened for PCD by nNO determination at the Hochgebirgsklinik Davos (HGK). In 5 individuals aging 2 to 15 yrs further diagnostic procedures were initiated as a result of repeated pathological nNO values <100 ppb. All subjects were referred to the HGK because of asthma or chronic bronchitis. PCD was confirmed in 3 cases and dismissed in one case; in one case results are pending.
Case report: 15-year-old boy with complete situs inversus, who was repeatedly treated for chronic sinusitis and otitis media since infancy. Childhood marked by complications such as hearing loss, retarded speech development and chronic tympanic colonization with P. aeruginosa, leading to mastoid surgery and bilateral tympanoplasty. In early adolescence diagnosis of asthma in the context of recurrent respiratory infections with no stabilization under repeated antibiotics and ICS/LABA. Referral to the HGK for treatment optimization of an uncontrolled asthma. Repeatedly nNO values <50 ppb with FeNO of 5 ppb, normal methacholin provocation and inconspicuous lung function. Referral to the Gemeinschaftsklinikum Koblenz for nasal brush biopsy and confirmation of suspected Kartagener syndrome by video microscopy.
Conclusion: Screening for PCD by nNO can result in optimized and earlier diagnosis and consequently to a correction of PCD prevalence. The method should be established in every paediatric respiratory center. For preschool children, measurement of nNO via a tidal breathing technique using a straw has been validated (Jung A et al. 2011). Early PCD diagnosis results in lower frequency of respiratory complications including surgical procedures, prevents unnecessary therapies and permits focused therapeutic interventions.

P 93

Pleural tap-guided antimicrobial treatment of pleuropneumonia can be limited to 14 days

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Background: Management and antimicrobial treatment of childhood pleuropneumonia are not standardised. Antibiotic treatment for up to 3 months has been advocated.
Objective: To determine the outcome of pleuropneumonia treated with amoxicillin/ clavulanic acid (amoxi-clav, 80 (po) –100 (iv) mg/kg/d) for 14 days from diagnostic pleural tap yielding bacteria susceptible to amoxi-clav or no bacterial growth.
Methods: In patients with pleuropneumonia blood cultures were prospectively collected, and pleural tap performed after chest X-ray or computed tomography (CT), and amoxi-clav was started for 14 days as per protocol. Follow-up included chest X-ray or CT after ≥3 months.
Results: 34 of 63 children first presented with pleuropneumonia (group A: median age 5 years) and 29 with pneumonia and subsequent pleural effusion (group B: after 1-11, mean 4.5 days; median age 5.7 years). 9 children (14%), all from group A, were not pre-treated with antibiotics when tapped. Pleural fluid and/or blood cultures were positive in 14 children (13 group A). Pleural fluid cultures from 4 children grew *Streptococcus pneumoniae*, from 4 *Streptococcus pyogenes*, and from 2 *Staphylococcus aureus*. Blood cultures from 4 additional children grew *S. pneumoniae*. All isolates were susceptible to amoxi-clav. 13 children from group A and 7 from group B received a chest drain (p = 0.28). Median hospitalisation in group A was 14.5 days and in group B 9 days (p <.0001). Follow-up 3–5 months later showed clinical recovery in all children, residual pleural scars in 6 patients from group A and 6 from group B, two children with pneumatocele (group B; radiologically resolved 5 months later) and one (group A) with a bulla after intubation and bronchopulmonary fistula.
Conclusion: Pleuropneumonia treated with amoxi-clav 14 days from tap clinically resolved in all but one child. Limiting antibiotic treatment to 14 days following microbiologic diagnostic pleural tap is safe.

P 94

Potential pitfalls in neonatal DRG

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Introduction: In 2012, diagnosis related financing (diagnosis related group, DRG) will be introduced in Switzerland. In newborns, DRG coding serving as a basis for reimbursement by health insurance programs is highly related to admission weight as well as diagnostic categories. We aim to assess a) potential pitfalls of the APDRG system related to coding as well as b) expenses due to insufficient cover ratio of frequent neonatal ICD codes in low DRG weights.
Methods: DRG coding was performed for neonates (age less than 28 days) admitted to our secondary neonatal care centre born between 2008–2009. Actual costs were compared to future reimbursement under DRG. Moreover, medical coding was compared with diagnoses identified in comprehensive medical charts.
Results: During the study period, a total of 733 neonates were enrolled. In one fourth substantial coding related mistakes were observed: DRG classification did not match the actual admission weight of <2500 g in 69 children (9%) due to a software problem. A total of 105 (14%) sick neonates discharged to the obstetric ward after 1.9 ± 1.2 days for better bonding with their mothers were wrongly coded as healthy children and allocated to the midwifery. Moreover, in 12 children (2%) no DRG coding was performed. With regard to expenses, insufficient cover ratio was observed in 2 common diagnoses for newborn: "Rule out sepsis, ICD P39.9" (n = 36, cover ratio: 64%) with empiric antibiotic treatment, especially due to high outliers (n = 19; 53%); and "Icterus neonatorum, ICD P59.0" (n = 68, cover ratio: 87%). Interestingly, in neonates with opiate withdrawal cover ratio was favourable (n = 13, cover ratio: 108%) even though all were high outliers (days on neonatal ward: 50.5 ± 18.2).
Conclusion: DRG system in neonates requires increased attention on correctness in admission weight registration. Moreover, strategies for early discharge to the obstetric ward for improved bonding with their mothers seem to be incompatible with DRG. Cost-effective strategies especially for neonates in low DRG weights with common diagnoses, including a shortened stay, seem to be mandatory.

P 95

Familial precariousness in a pediatric emergency department: a inventory in the Children Hospital of Lausanne (CHL)

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Introduction: Inventory of the major phenomena of precariousness concerning families consulting at the Emergency department of the Children Hospital of Lausanne.
Methods: Exploratory study including 60 families. A interview focusing on medical, social, psychological, economic and cultural datas has been developed by a multidisciplinary team using a tool called «Rapid Assessment Process». Vulnerabilities have been identified and quantified in 5 key-dimensions and the overall level of familial precariousness was defined by combining the results of the 5 dimensions and classified in 4 subgroups: absent, recognized, unrecognized and absolute.
Results: Prevalence of familial precariousness is 82%. Precarious families often consult spontaneously for general pediatrics issues. Children are mostly healthy and have adequate medical supervision, but a high prevalence of overweight and obesity, increasing in proportion to the family precariousness was found. Migrant families are strongly represented in emergency department (81%) and are mostly in a precarious situation. The status of asylum seeker, sending money home, a high prevalence of precarious job and communication difficulties are the main reasons highlighted to explain the high precarious level among migrant families.
Conclusion: This study demonstrates a high prevalence of precariousness in families consulting at the emergency department of the CHL. It also provides valuable information on the socio-cultural pattern and on specific health consumption mode by precarious

families. Finally, this study opens the perspective of a familial screening tool for precariousness that could be used by health care teams after a further validation.

P 96

Implementation and evaluation of a therapeutic educational multidisciplinary program within the paediatric clinic for cystic fibrosis

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Purpose of therapeutic education (TE) is to improve the understanding of the disease, its complications and needs for treatment.
Objective: implementation and evaluation of TE in the CF pediatric department in Lausanne.
Method: Patients were followed on their regular 3-month basis over 2 years. Interventional education took place at each visit and assessment every year. At visit 1, patient and CF team agree on an educational diagnosis leading to a contract. This allows defining the content of educational program, according to patient, family and CF team expectations. We used conductors created and validated by the GETHEM, in France.
Primary endpoint: Slowing of FEV1 and FEF25–75% slope decrease. Secondary: Benefit on quality of life (CFQ®) and evaluation of disease knowledge. We compared study to non-study subjects. This was an open interventional study. All patients ≥6 years of age followed at the pediatric CF department were included.
Results: Out of 32 patients from 6 to 16 years, 17 accepted the study, 15 denied and were used as control. For FEV_1 evolution was positive over time of study with a gain of 5.3 % in the study group versus a fall of 3.6% in the control group ($p < 0.02$). For FEF 25–75%, evolution was even better with a gain of 8.3 versus a fall of 0.5% in the control group ($p < 0.5$). In the study group, scores of knowledge obtained were between 22 and 28 (maximal score), instead of 15 and 27 in the control group. For CFQ® data were not conclusive.
Conclusion: This program was very well accepted by patients and their family. Despite very small number of patients, there is a clear trend to improvement of lung functions as well as knowledge of the disease over the study period.

P 97

Possible role of Delta-like 1 homolog (drosophila) in the development of chemoresistance in Neuroblastoma

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Introduction: As in other cancers, the development of chemoresistance represents a major obstacle in the successful treatment of high grade neuroblastoma (NB). NB is the most frequent extracranial paediatric solid tumour and a particularly heterogeneous and devastating disease.
Methods: To address the mechanisms underlying the chemoresistant phenotype in NB, we analysed the gene expression profile of doxorubicin-resistant cells (LAN-1-R) compared to the sensitive parental LAN-1 cell line.
Results: This analysis allowed us to identify DLK1 as a moderately, but significantly, overexpressed gene in the resistant variants. DLK1, a member of the Notch/delta/serrate family of proteins, is expressed in several embryonic tissues and in adult adrenal glands. DLK1 is also highly expressed in neuroendocrine tumours such as NB, suggesting a possible involvement in the development of the disease. We confirmed the increase in DLK1 expression by real-time quantitative PCR in LAN-1-R vs the non resistant LAN-1 cells with a 5.2-fold stimulation. Higher amounts of DLK1 protein were detected on total LAN-1-R cell extracts as measured by Western blot, as well as released in resistant LAN-1-R cells culture fluid as compared to non resistant LAN-1 cells. Overexpression of DLK1 in the LAN-1 sensitive cell line highly influenced the proliferative behaviour of these cells without modifying their drug sensitivity. In contrast, silencing of DLK1 in the LAN-1-R cell line by lentiviral-mediated micro-adapted shRNA restored caspase-3 activation upon different drug treatments, suggesting a role of DLK1 in the drug resistant phenotype of the LAN-1-R cell line.
Conclusion: Our data which fully support a recent report, implicating DLK1 in enhanced tumorigenic and undifferentiated characteristics of NB cells, further propose a role for DLK1 in their multi-drug resistant phenotype. These observations which associate DLK1 to multiple mechanisms leading to the particularly malignant behaviour of NB deserve further investigation.

P 98

Infant colitis – keep in mind defects of innate immunity!

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Introduction: Inflammatory bowel disease (IBD) is a rare event in infancy usually called Crohn's disease, ulcerative colitis or indeterminate colitis without precise knowledge of the underlying etiology. The differential diagnoses of primary immunodeficiencies (PID) (e.g. chronic granulomatous disease, IL-10 receptor defect, IPEX syndrome) are important since patients with PID may remain therapy-refractory or steroid-dependent after conventional treatment and only allogeneic hematopoietic stem cell transplantation (HSCT) is curative.
Case Report: We present the case of a child of consanguineous origin who showed clinical signs of IBD from early infancy with steroid-dependent chronic colitis, perianal fistulae leading to multiple abdominal surgical interventions and unusual complications (e.g. intestinal perforation). Under immunosuppression, the patient developed CMV and EBV primary infections below the age of two years leading to partial lung resection. Different treatment modalities (anti-TNF-alpha antibodies, azathioprine, tacrolimus) were administered to cure IBD but steroid dependency and failure to thrive remained a major problem leading to osteoporosis and consecutive vertebral fractures. Finally, at the age of 5 years, the underlying immunodeficiency was disclosed by molecular detection of an IL-10 receptor mutation leading to insufficient IL-10 signalling. We performed allogeneic HSCT from a mismatched healthy volunteer donor with improvement of colitis despite complete tapering of steroids. After HSCT, serious complications occurred, e.g. viral infections/ reactivations (HHV6, ADV, BK, CMV).
Conclusion: Our case illustrates that defects of the innate immunity should be kept in mind as important causes of IBD in infancy. Early allogeneic HSCT may be beneficial to prevent morbidity and mortality and is often curative.

P 99

Involvement of the CXCL12/CXCR4/CXCR7 axis in the malignant progression of human neuroblastoma

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Neuroblastoma (NB) is a devastating childhood neoplasm for which no efficient treatment is available for high stage tumours. Chemokines and their receptors, in particular the CXCR4/CXCL12 axis, have been involved in tumour progression. We previously reported a tumour type-specific and microenvironment-related growth-promoting role for the CXCR4 receptor. Such growth-promoting effects were highly significant only when NB cells were orthotopically injected in the adrenal gland of nude mice. This finding highly suggested a pivotal cross-talk between the CXCR4-expressing aggressive tumour cells and the associated microenvironment. The recent description of CXCR7 as a second CXCL12 receptor, add to the CXCL12/CXCR4 chemokine/receptor axis a new player, which function remains to be determined. In search for specific microenvironment-related effects, which might cooperate with CXCR4-mediated NB tumour growth, we addressed the role and participation of CXCR7. Although reported to confer atypical properties to cancer cells, the role of CXCR7 in NB development and the cross-talk with the microenvironment is still unknown. A preliminary screening of a small panel of NB tissues of different stages and histology types for CXCR7 expression revealed a selective CXCR7 staining on the more differentiated cells of the tumour. In contrast, CXCR7 was only moderately expressed on NB cell lines, but was found to increase upon exposition of cells to differentiation agents. From these preliminary observations, we propose that CXCR4 and CXCR7 may display two distinct and atypical roles in NB. Rather than a metastatic-promoting role identified in several other tumour systems, our data favour a tumour type-specific and growth-promoting influence for CXCR4, while CXCR7 may be implicated in NB maturation. These preliminary findings open new research perspectives for the role of the CXCL12/CXCR4/CXCR7 axis in the behaviour of NB, that will be further explored *in vitro* and *in vivo.*

P 100

Reptile-associated salmonellosis in children

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Introduction: A 7-week-old girl was hospitalised for septicaemia because of reptile-associated salmonellosis (RAS). RAS is being recognised as an emerging zoonosis. *Salmonella enterica* is a natural commensal organism of reptiles and different serotypes can be isolated from a single animal. Young children seem most vulnerable to serious infection by *Salmonella*. Therefore, we reviewed RAS and potential risk factors in children.
Methods: RAS in childhood is reviewed and discussed: 167 human *Salmonella* infections with a proven link by culture of *Salmonella* species between the affected child and the exposing reptiles are published since 1965.
Results: 162 children reported with RAS are otherwise healthy, while 5 suffer from immunocompromising co-morbidity. Focused on healthy children, RAS presents in the majority with gastrointestinal disease (74%), but 25 children (16%) have invasive salmonellosis, i.e. septicaemia (13), meningitis (9), and bone and joint infection (3). The median age is 1.7 years (mean 3.1 years): 46% are <1 year and 77% <5 years, respectively. Children with invasive disease are younger (median 0.3 years) than children with non-invasive RAS (median 2.0 years, p = <0.0001). The predominant reptile exposure is to turtles (52%); interestingly, children with septicaemia or meningitis more frequently were exposed to iguanas (54% and 67%, respectively; p = <0.001). Children with exposure to pet reptiles kept indoors (e.g. iguana, bearded dragon, snake, chameleon, gecko) are younger (median 1.3 years) than those exposed to turtles which commonly are kept outdoors (median 4.2 years).
Conclusion: Pet reptiles are significant epidemiological risk factors for salmonellosis. RAS most often causes self-limiting gastroenteritis, but is associated more likely with invasive disease such as septicaemia and meningitis, especially in infants (particularly <3 months), young children and immunocompromised children. Reptiles other than turtles may be associated with RAS particularly in young children because these reptiles are kept indoors and provide close (in-)direct contact to the young child.

P 101

Hashimoto Thyreoiditis related cerebrovascular ischemic insult in children

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Background: Hashimoto encephalopathy is a rare clinical condition of unknown etiology, closely associated with Hashimotos's thyroiditis and elevated titers of antithyroid antibodies. The disease presents with a wide spectrum of neuropsychological symptoms, also stroke like episodes caused by cerebroarterial vasculitis. The disease usually shows a very good response to corticosteroids.
Case report: We report a case of a 12-year-old girl with sudden onset of headache and left sided sensomotoric hemiparesis. The cranial MRI showed a right sided cerebral infarction of the Thalamus and Capsula interna. The MR-angiography and the neurovascular ultrasound demonstrated an occlusion of left A. cerebri posterior and a stenosis of A.cerebri media and A. carotis interna. Thyroid function tests showed elevated TSH with normal FT4, FT3 and elevated thyroid antibodies (Anti TPO-AK 2313 IU/ml, Thyreoglobulin AK 46 IU/ml). After exclusion of other cause of stroke, the diagnosis of Hashimoto Encephalitis with cerebral vasculitis was made and a treatment with high dose corticosteroids was started in addition to aspirin. This led to almost complete recovery of the patient.
Conclusion: Hashimoto thyreoditis can lead to cerebral vasculitis resulting in cerebral infarction. Because there exists an highly effective treatment, thyroid function testing including anti-thyreoid antibodies should always be considered in young patients presenting with cerebrovascular insult.

P 102

Evaluation of anatomic plates for medical education in adolescents

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Introduction: Patient education is an integral part of medical care. It is also crucial for the adherence to treatment, especially in chronically ill adolescent patients. Improving patients' knowledge regarding their condition has been demonstrated in several studies to contribute to their adherence to care. We therefore need to adapt our explanations to this particular age group by developing didactic tools.
Discussion: We developed fifteen specific anatomic plates simplifying the human body and its organs. Each plate is designated to a specific disease (e.g. diabetes, cystic fibrosis, kidney disease, asthma, obesity). Doctors can directly draw or write their explanations on these plates so that the patient (or his parent) can take this document home, in order to, revise and share the information given to them. Otherwise, the document could be added to the patient›s file. The figures could also be used to explain certain more invasive procedures (e.g.endoscopies, lumbar puncture, gynecologic examinations).We would like to present this didactic tool to pediatricians and get their opinion about its use.
Conclusion: Using anatomic plates, as didactic tool, could be useful to all pediatricians. The purpose of this presentation is to allow an access to these anatomic plates to pediatricians.

P 103

Respiratory distress and brachial plexus palsy: think about the phrenic nerve!

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A one-month-old girl presented with a history of breathing difficulties during the last two weeks and poor feeding since the day before. Her chest was asymmetric with prominent left lower ribs. We noticed a tachypnea, intermittent desaturations, sub-costal depression and left basal pulmonary hypoventilation. The chest X-ray showed an elevation of the left hemi-diaphragm and the diagnosis of diaphragmatic paralysis was confirmed through a diaphragmatic fluoroscopy. Despite continuous positive pressure ventilation and nasogastric feeding, the situation did not improve and a laparoscopic diaphragmatic plication had to be performed. This patient was born at term delivered by forceps and a left brachial plexus palsy was diagnosed on the first day of life. She manifested no respiratory distress and was discharged home, her left arm improving with physiotherapy. Obstetrical brachial plexus palsy may be complicated by lesion of phrenic nerve as it emerges the brachial plexus posteriorly from C3-C5, with a subsequent diaphragmatic paralysis. Symptoms may not be present at birth and develop when the activity, strength and respiratory flow of the child increase. Depending of the respiratory status, affected newborns should be hospitalized, the aim of the management being to avoid complications like respiratory infections, atelectasis, feeding difficulties and failure to thrive. Supportive treatment consists of oxygen administration, nasogastric feeding and mechanical ventilation and a surgical intervention must be discussed depending of the lack of improvement.

P 104

Pott's Puffy Tumor – Three Cases of an Intracranial Complication of Rhinosinusitis

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Introduction: Pott's Puffy Tumor is a rare complication of sinusitis originally described after head trauma and characterized by a subperiostaeal abscess associated with osteomyelitis of the frontal bone. Clinically, it presents with fever, headache, forehead tenderness and swelling, vomiting and malaise. Causative organisms include strepto- and staphylococci, Haemophilus influenzae, and anaerobic bacteria. Timely diagnosis, antibiotic treatment and surgical drainage are crucial, in order to avoid intracranial expansion. We present 3 pediatric cases of Pott's Puffy Tumor with intracranial involvement seen at the Kinderklinken Inselspital within 1 year.
Method: Case series from a single institution.
Cases: Three children aged 11 to 15 years (1 f, 2 m) with Pott's Puffy Tumor have been seen within 1 year. All of them presented with recurring headache, fever and vomiting. Focal neurological signs were present in one, impaired consciousness in another patient. Imaging showed extensive sinusitis with multifocal extra- and intracranial abscesses. Therapeutic regimen was set by pediatric infectiologists and neurologists, neuro-, pediatric and ENT-surgeons. All 3 patients underwent surgical abscess evacuation. Antimicrobial treatment was begun right after patient admission (1 patient had been on amoxicillin/clavulanic acid before admission). Cultures of the intraoperative specimens showed growth of streptococci of the milleri group in all 3 patients. Two patients were discharged without neurological sequelae. One adolescent is still in an inpatient rehabilitation program after having suffered from recurrent abscesses necessitating multiple neurosurgical interventions. He shows signs of frontal lobe dysfunction.

Conclusion: We present 3 cases of complicated sinusitis due to infection with streptococci of the milleri group (including S. anginosus, S. constellatus und S. intermedius). All of these bacteria are known to cause bone-penetrating, purulent infections, as well as endocarditis and bacteremia. One should think of intracranial complications in a case of clinical sinusitis and concomitant neurological signs. Early operative intervention in combination with intravenous antibiotics is the key stone to successful treatment.

P 105

Not your usual newborn rash – perinatal parainfluenza virus infection in a term neonate

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Introduction: Generalized exanthema in the newborn period is a common phenomenon. Most of the cases of maculopapular rash are due to exanthema neonatorum, a benign rash of the newborn. Differential diagnosis, often with specific additional clinical symptoms, includes varicella, rubella, HIV, syphilis, and a variety of other, often postnatally acquired, viral infections. Here, we present a case of perinatal parainfluenza virus infection in a term neonate.
Case Report: A boy was born at 39 4/7 weeks gestational age with a birth weight of 3585g. He was born vaginally to a healthy G3P3 mother of 36 years. The pregnancy was uneventful, except for symptoms of a common cold 2 days prior to delivery. The Apgar values were 8/9/9 at 1, 5 and 10 minutes respectively. 2 hours after birth the baby developed respiratory distress, paleness and prolonged capillary refill time, suggestive of neonatal infection. He was admitted to the neonatal ward for CPAP therapy and sepsis work up. Empiric antibiotic treatment with amikacine and amoxicilline was started. The respiratory distress increased leading to mechanical ventilation. The boy developed hemorrhagic pneumonia, with bilateral diffuse pulmonary infiltrates, mild pleural effusions and tracheal aspirates containing blood. Coagulation tests were normal. Further laboratory tests showed a leucocytosis of 29 G/l without left shift, a CRP <3 mg/l, a normal IL-6 and mild transient thrombopenia (minimum 128 G/l). Upon admission he was noted to have a diffuse and changing disseminated maculopapular rash. This rash disappeared within hours after delivery. The tracheobronchial aspirate was positive for parainfluenza virus (without further typing). Blood cultures remained negative. The boy was extubated to room air on day 3 and antibiotic therapy ceased after 5 days. The boy was discharged home on day 7 having completely recovered.
Conclusion: This is the first described case of a perinatal parainfluenza virus infection in a term neonate, highlighting the importance of considering the recent family history of the days before delivery.

P 106

Safety of adjuvanted pandemic influenza vaccines: background rate of narcolepsy in Europe

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Aim: To calculate background rates of narcolepsy in Europe and assess rates before (<April 2009) and during (May–September) the pandemic and after the beginning of the vaccination campaign (October 2009).
Methods: Seven European countries united in the VAESCO consortium used population and health care databases to calculate background incidence rates (IRs). Countries used standardized Jerboa® software locally on common input data to produce uniform aggregated data, which could be transferred centrally for calculation of incidence rates (IR) and pooling.
Results: 193 million person years (PY) including 18 million PY in 2009 and 2010 from Finland, IPCI (NL) and GPRD (UK) were captured. Overall crude and non-validated narcolepsy rates varied between 1.00 and 2.04 per 100,000 PY per country. Age-specific incidence rates differed between countries. The pooled age standardized rate was 1.30 (95%CI: 1.10–1.52) per 100,000 PY. Overall rates remained within confidence limits of a 10-year secular trend after start of the vaccination campaign. In 2009 narcolepsy rates increased in 5–19 year olds in Finland and 20–59 year olds in IPCI while rates decreased in GPRD. Rates started increasing before the vaccination campaign.
Conclusion: Background rates show different age distributions between countries. The observed increase of narcolepsy rates in Finland and the Netherlands was significant but started prior to the immunization campaign and involved different age groups. In the Netherlands the affected age group did not correspond with the group targeted for vaccination, in Finland it did. A European case control study is underway.

P 107

Are pathogen inactivated platelet concentrates less effective than standard platelets in children receiving chemotherapy?

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Introduction: Children treated with chemotherapy because of malignancies regularly require platelet (PLT) transfusions. The Swiss blood donation centers started to produce pathogen inactivated (PI) PLT concentrates in January 2011 in order to further reduce the risk of transfusion transmitted infections. This procedure (Intercept, Baxter) causes crosslinking of DNA and RNA molecules and by this inhibits replication of infectious pathogens. As there are recent data suggesting a worse effectiveness of PI PLT transfusions in adults (Kerkhoff, BJH 2010), we compared in vitro and in vivo efficacy of standard apheresis collected to PI PLT concentrates in children.
Method: We retrospectively analyzed data of platelet transfusions in children receiving chemotherapy at the University Children's Hospital Zürich at two time periods: July to December 2010, before PI (group A) and after introduction of PI, starting in January 2011 (group B). Transfusion efficacy was estimated using the corrected count increment (CCI) at 1 hr and 2 to 24 hrs after transfusion (Charles, JCO 2001). In vitro platelet function tests (aggregometry and flow-cytometry of platelet markers in stimulated and unstimulated platelets) were performed in both types of PLT concentrates.
Results: In group A 15 patients received 57 transfusions, whereas in group B 13 patients received 42 transfusions. Patients of both groups had similar clinical characteristics. At 1 hr after transfusion CCI was higher in group A (median 14726; range 3313 to 34830; n = 10) than in group B (4939; 0 to 16430; n = 26). In group B no satisfactory response, defined as CCI ≥5000, could be found for 14 transfusions. Equally at 2 to 24 hrs CCI was higher in group A (11360; 4200 to 23540; n = 19) compared to group B (5870; 0 to 20323; n = 34). Platelet aggregometry demonstrated absent response to collagen and ADP in PI PLT concentrates (data will be updated).
Conclusion: In this group of children receiving chemotherapy, transfusion of pathogen inactivated platelet concentrates led to an inferior rise in platelet count at 1 hr and up to 24 hrs after transfusion. Next to an unsatisfactory transfusion response, our in vitro data indicates inferior platelet function of pathogen inactivated platelets.

P 108

Cardiogenic shock secondary to coarctation of the aorta responding to prostaglandin infusion in a one month-old infant

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Introduction: Prenatal diagnosis of coarctation of the aorta is a challenge especially when not associated with hypoplastic aortic arch. Histological exam shows extension of ductal tissue with contractile properties in the aortic wall, responsible for aortic constriction at the time of ductal closure and for the variable clinical manifestation among patients from the neonatal period to late childhood.
Case Report: We report the case of a one month-old infant born at term with good neonatal adaptation. The emergency team was called at the parents' home for severe respiratory distress syndrome (RDS) and hypotonia. Upon arrival, the child was very cachectic, severly cyanotic with bradypnea, bradycardia and no palpable pulses necessitating intubation and volume infusion though a intraosseous line. Upon arrival in the emergency department, the infant deteriorated with bradycardia evolving into cardiac arrest and received multiple adrenalin boluses cardio-pulmonary resuscitation with cardiac massage during 20 minutes. Two episodes of seizures related to severe hypoglycemia were treated with phenobarbital. Echocardiography performed on a non beating heart revealed normal 4 chambers anatomy with a dilated left ventricle and severly depressed biventricular function. The aortic arch was S-shaped rendering it suspicious for coarctation and prostaglandin (PGE) infusion was initiated with a large bolus. Blood gas showed severe acidosis with pH 7.13, BE –9.8, HCO3 17.8 mmol/l and lactate 8.3 mmol/l. Continuous

bedside echocardiography showed progressive improvement of ventricular contraction and widening of the coarctation site together with reopening of the ductus arteriosus and was concordant with clinical improvement. The PGE infusion was continued and the infant underwent end-to-end anastomosis repair of the coarctation 8 hours later. The brain MRI performed 48 hours after the episode was normal. Her post-operative course has been uneventful with slow recovery and improvement of the nutritional status.
Conclusion: We repot a case of severe coarctation of the aorta presenting at one month of age and responding to prostaglandin infusion which allowed for widening of the coarctation site and reopening of the ductus arteriosus. Even though it is sometimes not be possible to achieve ductal reopening, PGE may act on the aortic posterior wall, allow for partial relief of the left ventricular obstruction and some clinical improvement.

P 109

Successful lung transplantation after cardiopulmonary resuscitation with extracorporeal membrane oxygenation in a child with idiopathic severe pulmonary arterial hypertension

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Introduction: Cardiopulmonary resuscitation (CPR) using extracorporeal membrane oxygenation (ECMO) is widely used following pediatric cardiac surgery, but is seldom reported in patient with severe pulmonary arterial hypertension (PAH) suffering cardiac arrest.
Case Report: A 9 years old boy was diagnosed with idiopathic PAH who failed to respond to sildenafil and bosentan therapy. Cardiac catheterization revealed suprasystemic non-reactive PAH (PAP: systolic 170, diastolic 126 and mean 140 mm Hg) with increased right (RAP 25 mm Hg) and left (PCWP 20 mm Hg) heart filling pressures. A central venous catheter was inserted under general anesthesia for intravenous prostacyclin therapy. Following extubation, the patient presented a laryngospasm and a pulmonary hypertensive crisis, followed by a cardiac arrest unresponsive to conventional cardiopulmonary resuscitation (CPR).The arterial blood gas (ABG) showed severe acidosis with pH 6.9, BE –24.6 and lactate 15 mmol/l. After femoral veno-arterial cannulation, ECMO was started 60 minutes after CPR was initiated. An atrioseptostomy was performed to unload the RV and allow for recovery. The child presented several complications which included transient acute renal failure, left anterior leg compartment syndrome needing emergent fasciotomy and an unresolving anterior medullary syndrome at the level of L4-L5, without brain damage. After two weeks of support, the child was transferred on ECMO by air ambulance to our transplant center. There, ECMO was weaned after 5 weeks and the child underwent lung transplantation 12 weeks after his cardiac arrest. The child is now 7 months post-transplantation, on tacrolimus and low-dose methylprednisolone immunosuppressive therapy and shows constant regression of his medullary syndrome.
Conclusion: ECMO-CPR can be life saving in children with severe PAH who suffer cardiac arrest and can be used as a bridge to recovery or lung transplantation. In such a situation, a multidisciplinary approach is essential.

P 110

Critical aortic stenosis progressing to hypoplastic left heart syndrome despite technically successful fetal aortic valvuloplasty

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Introduction: Fetal aortic stenosis (AS) with a normal sized left ventricle (LV) diagnosed at mid-gestation may progress to hypoplastic left heart syndrome (HLHS). Fetal aortic valvuloplasty can improve LV size and function and may allow for biventricular physiology.
Case Report: A 22-weeks gestation male fetus was diagnosed with critical AS. The LV was of normal size and apex-forming with endocardial fibroelastosis (EFE) and severly depressed systolic function. There was decreased forward flow through the mitral valve and the shunt across the foramen ovale (FO) was bidirectional. The fetus was referred at 23 ⅗ weeks to London and underwent successful fetal balloon aortic valvuloplasty. The fetal echocardiography showed an initial favorable appearance of R-L shunt across the FO, forward flow through the mitral valve and increased flow across the aortic valve with a peak velocity of 1.4 m/s. Further echocardiograms revealed evolution towards HLHS with almost exclusive L-R shunt across the FO, a very small jet of forward flow across the aortic valve (Z-score –2.4) and a hypoplastic mitral annulus (Z-score –6.3) with very limited forward flow. The LV was becoming small (Z-score –2.4) with little LV shortening or relaxation, in part due to EFE. This combination of findings made it very likely that a univentricular circulation would be the surgical option at birth and fetal counseling at 28 weeks was made in that direction. Nevertheless, the parents decided to carry on the pregnancy until birth and opted for comfort care of the newborn. Post-natal echocardiography confirmed the diagnosis of HLHS and the baby deceased 48 hours after birth from cyanosis and cardiogenic shock.
Conclusion: Fetal AS leads to increased LV pressure and EFE responsible for diastolic dysfunction and further compromise of LV growth. Proper selection of patients suitable for fetal aortic valvuloplasty is essential in order to prevent progression to HLHS.

P 111

Cardiac Troponin I Mutation responsible for Idiopathic Restrictive Cardiomyopathy in a 4-year-old Child

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Introduction: Restrictive cardiomyopathy (RCM) is a heart-muscle disease resulting in diastolic heart failure. In the past decade, genetic studies have demonstrated that RCM is part of the spectrum of sarcomeric disease and frequently coexists with hypertrophic cardiomyopathy (HCM) in affected families.
Case Report: We report the case of a 4 year-old boy originating from Congo, known for multiple episodes of thyphoid fever and malaria, who was referred by his pediatrician for hepatomegaly, dyspnea and sudations. The chest X-Ray was remarkable for cardiomegaly. The echocardiography showed enlarged both atria, normal size of the ventricles with normal biventricular systolic function but signs of abnormal diastolic function with moderate tricuspid regurgitation and signs of increased right heart filling pressure compatible with RCM. The cardiac catheterization confirmed the elevation of the filling pressure (right atrial pressure 12 mm Hg, pulmonary capillary wedge pressure 21 mm Hg, left ventricular end-diastolic pressure 20 mm Hg) and mild pulmonary hypertension (pulmonary artery pressure 35/15/23 mm Hg) secondary to left ventricular diastolic dysfunction. The cardiac MRI showed no pericardial thickening, no myocardial fibrosis with late enhancement and abnormal diastolic function, compatible with RCM. An extensive work-up showed increased Pro-BNP and ASAT, and polycythemia (Htc 58%). A bone marrow aspiration was normal. A renal biopsy revealed mesangioproliferative glomerulonephritis with neither immunoglobulin nor amyloidosis deposits. Mutation analysis by a resequencing array harbouring the complete coding sequence of 12 HCM genes revealed a novel missense mutation (p.Arg192Cys) in the cardiac troponin I gene (*TNNI3*) very likely responsible for the cardiac disease in our patient. After multiple episodes of heart failure, the child was listed for cardiac transplantation but unfortunately died from cardiac arrest following anesthesia for a permanent central line placement after one year on the waiting list.
Conclusion: We present the first pediatric case of RCM secondary to a mutation in the troponin I (TNNI3) gene. Large scale genetic sequencing platforms represent a rapid and efficient method for molecular analysis of heterogeneous cardiac disorders in pediatric clinical practice.

P 112

Tachycardia in a newborn – cardiac failure as a consequence of an arteriovenous malformation

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Introduction: Tachycardia in a newborn can have many causes, some common ones like sepsis or supraventricular tachycardia, but also some rare ones like in our case.
Case Report: A ten days old male newborn with two right sided segmental facial hemangiomas presen-ted with tachypnea and tachycardia. Echocardiography showed dilated ventricles, but no structural malformations. Supraventricular tachycardia was suspected. Adenosine was applied for this narrow complex tachycardia and revealed sinus rhythm. Cranial ultra-sound indicated arteriovenous malformation. Angio-MRI of the head revealed multiple malformations of large and medium sized vessels. The main findings were a right sided epidural arteriovenous malformation perfused from a giant meningeal artery, aplasia of the right internal carotid artery and a widely dilated right internal jugular vein. Angio-graphy visualized a pronounced arteriovenous shunt. Additionally, abnormalities of the brachiocephalic vessels were found (truncus bicaroticus).

Arteriovenous shunting was the cause for cardiac volume overload. Cardiac insufficiency now is being treated with diuretics. At present a surgical intervention or a catheter embolisation cannot be expected to be successful due to the complexity of the malformation. Further develop-ment of the child and progression of the disease have to be expected.
Discussion: Diagnostic criteria for PHACE-Syndrome were fulfilled as the newborn presented with segmental facial hemangiomas and had intracranial vessel malformations (mainly involving the arterial system) combined with an aortic arch anomaly. The differential diagnosis of Sturge-Weber-Syndrome could be excluded as this would affect capillary size blood vessels and typically is associated with leptomenigeal angiomas which were not detected in our case. Amongst all intracranial vessel malformations the Vena-Galeni-malformation most often manifests as cardiac failure.
Conclusion: In the case of tachycardia in the first weeks of life facial hemangiomas may provide the clue to the diagnosis of a major intracranial malformation with significant arteriovenous shunting. Although typical first symptoms of PHACE-Syndrome are neurological, a clinical presentation with the symptoms of cardiac failure is possible.

P 113

Effect of respiratory dead space on lung clearance index in healthy term neonates

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Background: Lung clearance index (LCI), a measure of ventilation heterogeneity calculated from multiple breath washout (MBW) test, is being used as a marker of disease severity and disease progression in infants and children with cystic fibrosis and chronic lung disease. LCI is defined as cumulative expired volume (CEV) when the end-tidal tracer gas concentration has fallen to 1/40th of the starting concentration, divided by functional residual capacity (FRC). Respiratory dead space (V_d) may influence LCI given that V_d contributes to CEV.
Aims: To assess whether there is an association between V_d and LCI in healthy term neonates.
Patients and methods: Thirty-five healthy-term infants (gestational age 39.7 w, SD ±1.2 w, 17 males) were studied in quiet unsedated sleep using a mainstream ultrasonic flow meter (Spiroson Exhalyzer D, Ecomedics AG, Duernten, Switzerland). LCI and FRC were calculated by MBW with 5% sulphur hexafluoride. V_d was calculated from expired molar mass during tidal breathing immediately prior to MBW. Data were analysed by multiple linear regression.
Results: Linear regression revealed a positive association between dead space to tidal volume ratio (V_d/V_T) and LCI ($R^2 = 0.18$, $p = 0.011$). No other factors including birth weight, body length, gender, age at test, and FRC were related to LCI.
Conclusion: V_d/V_T is a significant predictor of LCI in healthy term neonates. Further studies are required to assess the impact of V_d/V_T on LCI in infants with lung disease.

P 114

Exanthem subitum may not always be benign

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Introduction: A previously healthy 9-year-old boy presented to the emergency room with a two day history of ataxia and lethargy. On physical examination, the patient was afebrile and lethargic. His neurological exam showed discrete nuchal rigidity, positive Romberg sign, ataxia and asymmetric osteotendinous reflexes. A lumbar puncture showed a clear cerebrospinal fluid with 45M/l leucocytes with a 98% predominance of lymphocytes, slightly elevated proteins (0.53 g/l) and normal glucose concentration (3.4 mmol/l). The gram stain was negative. PCRs for HSV-1 and HSV-2, varicella, enterovirus, parechovirus and borrelia were negative. An electroencephalogram showed diffuse slowing with spikes and spike-waves in the temporal region. Cerebral MRI was normal. Because of these findings and in the presence of a persistent alteration of consciousness consistent with herpes encephalitis, the patient received intravenous Acyclovir. A second cerebrospinal fluid analysis showed a positive PCR result for HHV6. Serologies showed a reactivation of a past infection and we concluded to a meningoencephalitis due to HHV6 (unknown variant). The boy was discharged on day 10 with a normal neurological status.
Discussion: HHV6 infection generally presents as a benign condition. However, due to its tropism for glial cells HHV6 may provoke febrile convulsions, chronic epilepsy, multiple sclerosis and encephalitis, even in the immunocompetent host. Associated neuroradiological findings include necrotizing encephalopathy, striatal necrosis, and white matter abnormalities. These potentially severe neurological complications suggest either direct invasion of the virus into the CNS or immune-mediated damage. HHV6 typical treatment is Ganciclovir.
Conclusion: HHV6 is associated with meningoencephalitis and should be screened for in a suggestive context.

P 115

Mild encephalopathy with splenial lesion of the corpus callosum associated with Parainfluenza: A case report

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Background: Mild encephalitis/encephalopathy with reversible splenial lesions (MERS) has been associated with influenza A and B virus infection, measles, mumps-, adeno-, and rotavirus. Patients typically present with neurological symptoms 1 to 3 days after a prodromal illness (fever, cough and rhinorrhea). They recover completely within a few days. Characteristically, cytotoxic edema, predominantly in the splenium of the corpus callosum is demonstrated by MRI.
Case report: A 5-year-old boy was referred to our institution with tonic-clonic convulsions. He showed a decreased level of consciousness (GCS 9) and slight nuchal rigidity. There were no focal neurological signs. Laboratory investigations revealed an elevated CRP (110 mg/l) with normal WBC, and a moderate pleocytosis (83 × 10E6) in the cerebrospinal fluid (CSF). The EEG showed slowing of background activity, but no epileptic activity. Brain MRI revealed cytotoxic edema of the corpus callosum with decreased apparent diffusion coefficient values and high signal intensitiy in diffusion weighted imaging. At admission treatment with ceftriaxon and acyclovir was initiated, but bacteriological cultures and repeated PCR for herpes simplex virus in the CSF remained negative. Other infectious etiologies for encephalitis were excluded by PCR in the CSF and blood serology. Ultimately, parainfluenza virus type 1–3 infection alone was documented by direct immunofluorescence in the initial nasopharyngeal swab. PCR for parainfluenza virus type 1–4 in the CSF remained negative. After 4 days the patient completely recovered.
Conclusion: This is – to our knowledge – the first description of MERS in association with parainfluenza virus infection. Although we could not detect the parainfluenza virus in a PCR of the CSF, we assume parainfluenza to be the aetiology of this cytotoxic edema of the corpus callosum.

P 116

A surprisingly dramatic manifestation of a common chronic disease

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Background: Cystic fibrosis (CF) is an autosomal recessive multisystem disorder caused by mutations in the CFTR-gene. There is considerable variability in both presentation and clinical course of the disease. Classical presenting symptoms include failure to thrive and recurrent respiratory symptoms.
Case report: An 11-year-old girl was admitted to a regional hospital for an acute respiratory infection with severe bronchial obstruction, hypoxemia and reduced general condition. Atypical pneumonia was suspected and treatment with oxygen, clarithromycin, salbutamol and ipratropium bromide was initiated. On day four, the child developed acute respiratory failure needing endotracheal intubation and was referred to our hospital. On arrival, the girl presented in catecholamine-requiring shock with severe hypercarbia (pCO_2 of 143 mm Hg), respiratory and metabolic acidosis, fine bilateral crackles over all lung fields and hepatomegaly. Because of unsuccessful conventional ventilation, high frequency oscillatory (HFO) ventilation was initiated. Ancillary studies revealed moderate inflammation, five-fold elevated liver enzymes and bilateral reticulonodular opacities. A putride and bloody trachea-bronchial aspirate revealed moderate growth of *Staphylococcus aureus* and profuse growth of two strains of *Pseudomonas aeruginosa* (one with mucoid colony morphology). Further laboratory investigations including antibodies for vasculitis and goodpasture syndrome remained negative. After four days of HFO and four days of conventional ventilation, the patient was successfully extubated. Low weight and clubbing were noted. Progressive hyperglycemia developed after extubation and was treated with insulin. Repeated history taking revealed one single episode of obstructive bronchitis at six months of age and loose, large volume stools, the family history was unremarkable. An abnormal sweat test confirmed the clinical suspicion of cystic fibrosis.
Conclusion: This child presented in a very unusual way with manifestation of cystic fibrosis in an advanced stage with acute near fatal respiratory failure, colonization with *Staphylococcus aureus* and *Pseudomonas aeruginosa* mucoid strains and manifest exocrine and endocrine pancreas insufficiency. CF newborn screening program started in January 2011 may prevent delayed diagnosis of CF in future.

P 117

Lactic Acidosis in Children with Severe Acute Asthma Exacerbation

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Introduction: Lactic acidosis can occur during acute asthma attacks primarily related to excessive respiratory muscle work. It has been rarely described in children. We report 2 cases in children, with acute asthma exacerbation and hyperlactatemia.
Case Report: Two children, aged 15 and 5 years, were admitted to the Emergency Department of the Children's Hospital, with acute asthma exacerbation. Both children had a past history of asthma. Initial treatment included inhaled βeta-2 agonist therapy and oral corticosteroids. Partial clinical improvement was noted with a reduction of wheezing. Both children remained markedly dyspnoeic and tachypneic, leading to an intensification of βeta-2 agonist treatment. Venous blood gas analysis showed increased lactates (10.7 and 7.3 mmol/l respectively) with normal pH, pCO_2 and O_2 saturation. During the course of treatment, 1 child developed low blood pressure (60/30 mm Hg), malaise and general discomfort. He improved after receiving intravenous volume replacement. Both children showed significant clinical improvement with normalisation of lactate levels in parallel to withdrawal of βeta-2 agonist therapy after several hours of monitoring.
Conclusion: These cases illustrate the development of hyperlactatemia during βeta-2 agonist therapy in acute exacerbation of asthma. The elevated lactate values could not be solely attributed to tissue hypoxia as observed by vigorous muscle contraction of the respiratory muscles. Several explanations have been proposed and the cause is likely multifactorial. Lactate production due to βeta-2 agonist therapy is a side effect rarely described and can play an important role in patient management when aggressive treatment is initiated. This side effect is often unrecognised and therefore under-estimated.

P 118

Pelvic Osteomyelitis Mimicking Acute Abdomen: a case report

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Introduction: Pelvic osteomyelitis is a rare but serious disease that can lead to systemic and local complications if left untreated. The diagnosis and management are often delayed because of the nonspecific clinical presentation.
Case: We present the case of a fourteen-year-old boy who presented to our Emergency Department with a 5 days history of high fever and continuous left abdominal pain exacerbated by walking and weight bearing. At arrival, physical examination reveals an elective pain in the left iliac quadrant without signs of peritonism, associated with an elevated C-Reactive protein. The initial radiological screening (abdominal, hip, testicular ultrasound and CT-scan) is normal except for the presence of intra-peritoneal fluid in small quantity. After 4 days of hospitalization without antimicrobial therapy, the patient remains highly febrile and complains of worsening abdominal pain; the inflammatory parameters increase and physical examination reveals an elective pain of the pelvic bone. An MRI is performed, and confirms signs of osteomyelitis of the ilio-pubic branch with surrounding abscess. After an unsuccessful trial of conservative treatment, a surgical drainage is necessary. The clinical and biological course is then favourable with a 6-weeks antibiotic treatment. After 2 months of follow-up, the evolution is actually uncomplicated.
Discussion: This case emphasizes the difficulty of making early diagnosis of pelvic osteomyelitis. In fact, peripelvic infections are rare but serious affections, which are often not considered in differential diagnoses for patients presenting with fever, abdominal pain, inability to bear weight and elevated inflammatory parameters. One must remember to consider it in front of the clinical presentation of our patient, and to complete the screening with an MRI.
Conclusion: In case of atypical febrile abdominal illness, bone infection should always be considered. If the clinical presentation is inconsistent with laboratory results and first-line radiological techniques, further studies are necessary. As ultrasound and CT-scan are known to be limited techniques for early diagnosis of bone disease, MRI should be considered early in the course of the disease, because of its great accuracy in evaluating bone infection and deep tissue involvement.

P 119

Outbreak investigation because of neonatal listeriosis cases in a Swiss maternity

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Objectives: Since 2008, the Federal Office of Public Health in Switzerland has reported no single case of neonatal listeriosis. For this reason, outbreak investigation was immediately started after occurrence of severe invasive infection with detection of *L. monocytogenes* in two term borne neonates within one week in a Swiss maternity. The aim was to identify potential sources of this foodborne pathogen and consecutively being able to prevent further illness.
Methods: Confronted by a foodborne disease outbreak, guidelines for investigation and control as recommended were applied.
Results: *L. monocytogenes* serotype 4 was cultured in the cerebrospinal fluid of case 1 as well as 4 days later in the blood of case 2. Both neonates had pleocytosis and responded to antibiotic treatment with high dose amoxicillin. Further investigation of listeria isolates by pulsed field gel electrophoresis typing in the national listeria reference centre identified both as type 7, a type not detected in the 20 other human isolates that has been tested before. The mother of case1showed symptoms of a viral illness 2 weeks before giving birth. Listeria infection was suspected because of a 4-fold rise in specific antibody titers. The mother of case 2 stayed seronegative even 4 weeks post partum. Neither direct or indirect contacts, nor common used subjects or food exposure could be identified except breast milk of both mothers that has been stored during one night together in one fridge. After implementation of clearly spatial separation in clean-contaminated and mother-nurses areas within the milk kitchen and individually packaged bottles and related accessoirs no further cases of listeriosis infections occured.
Conclusion: Although transmission by contaminated hands of health care workers is considered the most frequent reason for nosocomial infection, this route appears unplausible in this outbreak. In fact, breast milk was identified as the most likely source of nosocomial infection. As a consequence, system changes and structural changes concerning breast milk administration were implemented. As breast milk administration errors are not infrequent events, breast milk should be considered in foodborne outbreak investigations and strict prevention guidelines should be implemented and controlled in all maternities.

P 120

Invasive *Fusobacterium* infection after mononucleosis

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A 16 years old boy with initially uncomplicated monunucleosis developed after 10 days a peritonsillar and a large confluent abscess of the cervical lymph nodes, clearly delineated in the CT of the neck. We performed a drainage of the abscess and started an antibiotic therapy with ceftriaxone i.v. with initially fairly poor response. In the culture of the drainage fluid we could ascertain the presence of Fusobacterium necrophorum by a 16S-rRNA PCR. After adding Metronidazol i.v. the course was gradually improving. A MRI of neck and skull shows a total thrombosis of the whole internal jugular vein. We interprete this situation as a partial Lemierre syndrome. In the literature about 10% of published cases of invasive Fusobacterium necroforum infections are associated with EBV infections.

P 121

Endobronchial inflammatory myofibroblastic tumor as a rare cause of left lung atelectasis

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Introduction: Atelectasis is a key component of many acute lung diseases. In pediatric clinical settings, it is most often related to lung infection and mucus plugging. We report the case of a child presenting with chronic onset of dyspnea, wheezing and complete left lung atelectasis resulting from left mainstem bronchial tumoral obstruction.
Case Report: An 11½ years old healthy boy presenting with gradual onset of dyspnea and cough was treated for asthma without clinical improvement. One month after the onset of respiratory symptoms, he developed fever and the cough became persistent. Chest-XRay showed left lower lobe consolidation. The diagnostic of Pneumonia was established and antibiotherapy prescribed. Two months later, he was referred to our Pulmonary Unit because of unsatisfying clinical

course with complete left lung radiological consolidation. White blood cell count and blood chemistry were unremarkable, CRP was <10 mg/dl and tuberculin skin test was negative. Chest ultrasound showed pulmonary consolidation without significant pleural effusion. Thoracic CT confirmed total left lung atelectasis and raised a high suspicion of left mainstem endobronchial mass, which was later confirmed by chest MRI (size: 2.1 × 1.8 × 3.4 cm). On PET CT this mass showed increased metabolism (SUV max 21.8). No other hypermetabolic lesions were found on PET-CT and on whole body MRI. Lung volumes were markedly decreased with a restrictive syndrome (total lung capacity 55% of predicted). Bronchoscopy visualised the endobronchial tumor which was subtotally obstructing the distal end of the left mainstem bronchus. The polypoid shape of the tumor allowed total endoscopic resection with a diathermic snare combined to argon plasma coagulation, without complication. Histological analysis revealed a mesenchymal tumor more commonly called IMT (Inflammatory Myofibroblastic Tumor) which has good prognosis if totally removed. Cytogenetic analysis confirmed this diagnosis with positive characteristic rearrangement at 2p23.9 of the gene coding for the anaplastic lymphoma kinase (ALK)-receptor tyrosine kinase (found in 50% of IMT cases). Radiological studies after resection showed complete resolution of the atelectasis. Repeated bronchoscopies at 1 and 3 months were normal. Gradual recuperation with TLC at 75% of predicted was obtained at 3 months.
Conclusion: When confronted with persistent atelectasis with or without infection signs, anatomical obstruction must be considered. This case illustrates the importance of radiological studies, bronchoscopy and lung function testing in the evaluation and treatment of the endobronchial masses which remain a rare diagnosis in children. Successful endoscopic removal by interventional bronchoscopy could be performed in this child.

P 122

Pulmonary intersitial glycogenosis in an infant presenting with hypotonia, heart failure, pulmonary hypertension and interstitial lung disease

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Introduction: Interstitial lung disease of infancy is a rare entity usually presenting in the neonatal period with tachypnea, hypoxemia and respiratory insufficiency. We report the unusual case of an infant born without significant initial pulmonary compromise, but who showed in the first months of life a persisting unexplained hypotonia.
Case Report: The patient is a caucasian male born at term by C-section because of fetal bradycadia, with Apgar 3/8/9. Given a respiratory distress syndrome, he necessitated 10' bag mask ventilation followed by CPAP. Birth weight was 3400 g (P25-50), length 51 cm (P50), OFC 36.5 cm (P75-90). Discharge from hospital was on day 5. At the 1 and 3 months visits, generalized global hypotonia was noted in an otherwise alert baby. Work-up (ENMG, brain MRI, genetic testing for Prader-Willi syndrome and spinal muscular atrophy) was non contributive. Further investigations at the age of 6 months revealed significant cardiomegaly and increased vascular markings on chest X-ray. Echocardiography showed a large secundum atrial septal defect (ASD), severe dilation of the right heart chambers, biventricular systolic dysfunction and severe infrasystemic pulmonary hypertension (SPAP estimated at 70 mm Hg). Cardiac catheterization confirmed a left to right shunt across the ASD. The pulmonary to systemic output (Qp/Qs) ratio was 2.1, while the pulmonary to systemic resistance (Rp/Rs) ratio was 0.24. PAP was 53/23/37 mm Hg, decreasing to 43/19/31 mm Hg after pulmonary vasodilatation. Chest CT showed bilateral pulmonary infiltrates and ground glass opacities. Bronchoscopy revealed a pulsatile posterior extrinsic left mainstem bronchus compression. Bronchoalveolar lavage was normal and red oil stain for detecting silent aspiration was negative. Nocturnal oxymetry was abnormal with numerous and significant episodes of desaturations. Pulmonary biopsy showed increased thickness of alveolar septa with an increased number of cells containing glycogen in the interstitium, compatible with pulmonary interstitial glycogenosis (PIG). Patient underwent surgical closure of ASD with a fenestrated patch, to allow for post-operative right heart decompression. One month later, echocardiography showed regression of right heart chambers dilatation and quasi normalization of the PAP. Nocturnal oxymetry was also normal.
Conclusion: PIG was first reported by Schroeder et al. The clinical presentation is usually with neonatal tachypnea, diffuse pulmonary infiltrates on chest X-Ray and interstitial proliferation of histiocytic-type cells with minimal inflammation on lung biopsy, evolving usually for most patients towards overall general clinical improvement over time. Our case illustrates an unusual presentation of PIG in early infancy, with hypotonia, ASD, pulmonary hypertension and heart failure. In this case, ASD closure allowed for both cardiac and pulmonary improvement.

P 123

Intrapleural vincristine infusion in a 7 year-old girl with an acute lymphoblastic leukemia: a case-report and review of the literature

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Background: Despite the well-known side effects of subcutaneous diffusion of chemotherapeutic agents, there is limited data in literature concerning the management of accidental intrapleural infusion of antineoplastic drugs. While vincristine was reported as an effective sclerosing agent for malignant pleural effusion in adults, we report the case of an intrapleural infusion of vincristine from a misplaced central venous catheter (CVC) and its management.
Case-Report: A 7-year-old girl was diagnosed with a standard risk B precursor lymphoblastic leukemia and treated according to the Children's Oncology Group Protocol AALL0331. At D1 of the Induction Phase and after implantation of a CVC in the right internal jugular vein, the fluoroscopic control showed correct positioning of the CVC, but there was no blood reflux. Therefore a fibrinolytic treatment (urokinase 10000 UI) was performed, blood reflux was obtained and she received intravenous treatments (vincristine 1,5 mg/m^2, ceftazidim 2600 mg, esomeprazole 10 mg, ondansetron 8 mg, konakion 5 mg, rasburicase 4.5 mg, dexamethasone 6 mg, and D5-NaCl 0.45% with 20 mEq/l NaCO3 around 1L). Four hours after injection, the patient presented basithoracic pain with acute respiratory distress (tachypnea, desaturation) and right lung hypoventilation. Chest X-ray and US showed a right pleural effusion, and CVC angiography showed diffusion of contrast liquid directly in the pleural space, confirming our suspicion of CVC malposition in the pleural cavity. Treatment included pleural drainage with aspiration of 1.5L of liquid and washing with 0.9% saline solution. The CVC was removed. The analysis of pleural liquid showed small concentrations of ondansetron and vincristine. We didn't perform any further surgical treatment neither local application of hyaluronic acid, as proposed in literature for subcutaneous vincristine extravasations. Clinical evolution was excellent, the thoracic drain was removed after 3 days and a new CVC was inserted in the left internal jugular vein. Except a severe constipation during the week following this event there were no sequelae of this accidental intrathoracic vincristine infusion after one year of follow-up.
Conclusion: We report the rare occurrence of accidental intrapleural vincristine direct infusion. Treatment with pleural drainage and washing was effective in this case, with an excellent outcome. We hope that our experience will be helpful for potential future accidents.

P 124

Fibrous Tumors in Infancy and Childhood: from Spontaneous Cure to Mutilating Disease

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Introduction: Fibrous tumors (FT) is a rare and heterogeneous entity in pediatric oncology. Clinical course ranges from spontaneous cure to aggressive and mutilating disease. Surgery is generally considered as the primary treatment modality, however is often not feasible. Second-line treatment is therefore chemotherapy (CT) and radiotherapy (RT).
Results: We report on a case series of 13 pts. with FT, including 7 aggressive fibromatosis (AF), 2 infantile myofibromatosis (IM), 2 inflammatory myofibroblastic tumors, 1 inflammatory fibrosarcoma and 1 low-grad fibroblastic sarcoma.
For AF (n = 7) medium age at diagnosis was 3yrs. (range 0.7 to 12 yrs.). The most common location was face/neck (5 pts.), followed by spine/trunk (2 pts.). Six out of 7 pts. received CT (mean duration 15 mths.). Three pts. had surgery, 2 of them at diagnosis and 1 after 11 mths. of CT. One pt. had RT after initial surgery. All pts. are in complete remission (n = 5) or have stable disease (n = 2) with a median follow-up of 35 mths. The two girls (5 and 6 mths.) with IM presented with multiple lesions in subcutaneous tissue, muscles and bone. No visceral organs were involved. Both achieved complete or near complete spontaneous remission. The 2 pts. with an inflammatory myofibroblastic tumor the tumor was located in the bladder and in the trachea, respectively. Treatment with steroids and antibiotics in the former, and repeated endoscopic tumor resections in the latter, led to stable residual disease in both cases. One patient with inflammatory fibrosarcoma of the bladder received CT for 2 mths. followed by partial bladder resection. He is in complete remission. The last patient is a neonate who presented with a painless swelling of the right cheek at 1 mth. of age. Biopsy revealed an atypical myofibroblastic proliferation and the patient was followed without treatment. Because of tumor progression biopsy was repeated and a low-grade sarcoma was diagnosed. After 2 mths. of CT there is a slight tumor reduction.

Conclusion: There is a wide range of fibrous tumors in children and histological sub-classification might be difficult. The clinical course is often unpredictable and challenging for therapeutic decision making. Surgery plays an important role but might be often limited due to risk of mutilation. Chemotherapy, often long lasting, can be beneficial in many cases and can even lead to complete remission. Complete remission or stable disease in fibrous tumors can be achieved in the majority of patients.

P 125

Atypical presentation of Purpura Henloch Schönlein with severe intestinal involvement

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A 6 years old boy presented with worsening colicky abdominal pain and vomiting since 12 hours. No diarrhea, no blood in the stool, no fever. On admission the physical examination revealed a diffuse tenderness of the abdomen with an ill-defined mass on the left side and increased bowel sounds. WBC and CRP not alterated. Sonographically little amount of ascites, no signs of intussusception. In the next days gradual deterioration of the clinical condition with worsening of the abdominal pain; at serial sonographic examinations massive increase of ascites and thickening of the jejunal intestinal wall. Five days after the onset appearance of melena. A laparotomy showed a massive inflammatory swelling of a jejunal segment of about 40 cm without other pathologies. An empirical antibiotic therapy was started (ceftazidim and metronidazolum) waiting at the bacteriological results of blood, ascites and stool (afterwards with negative results). Postoperativelly conservative therapy particularly correction of a severe hypoproteinemia caused by a proteinloosing enteropathy. Twelve days after onset appearance of a purpuric rash who allowed to make the diagnosis: atypical presentation of Purpura Henloch Schönlein with severe intestinal involvement. A steroid therapy was started with rapid improvement of the clinical conditions. In the following days appearance of a scrotal swelling and transitory arthralgia of the left ellbow, typical signs of the illness as well. A renal involvement was not present.

P 126

Dobody mass index and physical activity levels affect spirometry results in children?

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Objectives: The literature on the influence of physical activity and Body Mass Index (BMI) on pulmonary function is inconsistent. We aimed: 1) to describe physical activity levels and BMI in a cohort of children with and without asthma; and 2) to test the independent effect of physical activity and BMI on spirometry results.
Methods: We performed spirometry (post-salbutamol FEV_1, FVC, FEF50) in a nested sample of 1141 children aged 9–13 years from a population-based cohort study in Leicestershire, UK. BMI (standardized for age),reported physical activity and asthma related factors were assessed by questionnaire at the same age. Independent associationsof BMI and physical activity with spirometry results were tested using multivariable linear regression adjusting for sex, age, height, ethnicity, socio-economic factors, current wheeze (last 12 months) as well as wheeze and physical activity at age 1–6 years.
Results: Among all children, 374 (33%) reported little physical activity (0–1 hour outdoorplay per day), 447 (40%) played 1–2 hours and 296 (26%) more than 2 hoursoutdoors. 644 (56%) of children had normal weight, 57 (5%)were underweight, 316 (28%)overweight and 121 (11%) obese. Prevalence of wheeze (226 children, 20%) and severity of wheeze defined by treatment stepwere not associated with physical activity level ($p = 0.24$) or BMI ($p = 0.84$). In the multivariable model, physical activity was not associated with FEV_1 FVC or FEF50. Increasing BMI was associated with increased FEV_1 and FVC, but not with FEF50.
Conclusion: In this population,physical activity and BMI were not associated with current wheeze. Physical activity level did not affect spirometry results but BMI was associated. Our results suggestto consider weight for standardization and interpretation of spirometry data in children.

P 127

PFAPA syndrome is linked to dysregulated IL-1β production

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Background: PFAPA is associated with aphtosis pharyngitis, and cervical adenitis found in children with a spontaneous resolution in most cases until adolescence. There is no clear aetiology found up to now and recently a familial predominance has been shown in PFAPA suggesting a genetic cause. In other autoinflammatory diseases elevated levels of IL-1β and the efficacy of the treatment with Anakinra, a recombinant human IL-1 receptor antagonist, suggests a role for IL-1β in its pathogenesis.
Aim: Evaluate the role of dysregulation of IL-1β secretion in the pathogenesis of PFAPA syndrome.
Methods: In 12 patients with confirmed PFAPA blood was drawn during and outside a febrile episode. Inflammatory parameters were measured. PBMCs were stimulated with LPS and cytokine levels were dosed. Genomic DNA was screened by PCR and sequencing for genetic variants of MEFV, TNFRSF1A, MVK and NLRP3 genes.
Results: Monocyte and neutrophil counts, ESR, CRP and SAA levels were significantly elevated during febrile episodes. PBMC's secreted more IL-1β upon LPS stimulation during fever flares ($p < 0.001$: OUT 235 ± 56 pg/ml; IN 575 ± 88 pg/ml). Serum levels of IL-6, IP-10 and caspase -1 were also increased significantly during febrile episode whereas TNFβ and MCP-1 did not show significant changes. 4 of 12 patients were found to have a heterogeneous variant in the NLRP3 gene (associated to CAPS) but no variant in the MEFV, TNFRSF1A and MVK genes. Patients with NLRP3 variants did not have a different clinical presentation in comparison to the other PFAPA patients.
Conclusion: In PFAPA patients, stimulated mononuclear cells show increased IL-1β secretion during fever episodes suggesting a role for IL-1β in the pathogenesis of PFAPA. Interestingly, we found in a subset of patients variants for the NLRP3 gene, which is implicated in the regulation of IL-1β production. These findings may open new treatment options in PFAPA.

P 128

aEEG developmental changes during the first three days after birth in very preterm infants

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Background: Maturation of amplitude-integrated electroencephalogram (aEEG) activity in the preterm infant has been shown to be influenced by both gestational and postmenstrual age. Objective: To describe the development of aEEG activity in preterm infants <32 gestational weeks over the first three days of life in relation to the presence of cerebral injuries.
Methods: 104 infants born at 25–32 gestational weeks without evidence of congenital or metabolic abnormalities had continuous two-channel aEEG during the first 72–84 hours of life. aEEG characteristics were evaluated visually applying pre-established criteria ("background pattern" after Hellström-Westas et al. 2006, "maturity score" after Burdjalov et al., 2003) and quantitatively (maximum and minimum aEEG amplitude). Cranial ultrasound was obtained at the first, third and seventh day of life and repeated on a weekly basis.
Results: A significant increase in the cortical activity was observed between day 1 and day 3 after birth with all assessment methods: a) aEEG pattern significantly changed from an immature to a more mature pattern. This trend occurred more rapidly in newborns without any brain (78) or minor lesions (20) on cranial ultrasound compared with newborns with major brain lesions (6), however, this was not statistically significant. The sleep-wake cycling activity tended to develop particularly slower in newborns with major brain lesions; b) the maximum and minimum aEEG amplitude increased significantly over time in all newborns; c) over time, the background score was well correlated with the maturity score (r^2 range = .16 – .68, $p < .001$) as well as the maturity score with the quantitative measurements (r^2 range = .05, $p < .05$ to .15, $p < .001$, and r^2 range = .25 to .43, $p < .001$, respectively).
Conclusions: aEEG activity matures significantly already during the first days of life. In the presence of overt cerebral lesions this process is delayed. The good correlation between the different aEEG assessment tools are of utility in the clinical practice.

P 129

Morning of informations and speaking with the families of children with sicklecelldisease (SCD): preparation, unfolding, evaluation. Lausanne's experiment

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Introduction: Migration and adoption confront the medical teams with the sicklecelldisease. There are different aspects in this disease: multiorganic, preventive, therapeutic, educational, school, professional, psychological and cultural. In 2010 in Lausanne 50 children are followed in the consultation.
Objectif: To propose a meeting for a better accompagnement
Method: *Preparation.* We send a letter to the parents (interests, waitings, availabilities). Topics chosen starting from the answers and difficulties perceived by the team. Next we send the program for invitation. *Unfolding.* Saturday, August 21, 2010. Three presentations: what is the sicklecelldisease and why all these follow-up examinations at the hospital?, school and neuropsychological assessment, role of the social worker. Workshops: pain, infection, games for children.
Evaluation: 46 families invited, 20 interested, 10 went (37 persons). After the meeting, we have send 10 paper-interviews, 7 came back. All wish a new meeting and propose different subjects.
Conclusion: One 2nd morning will be organized with the adult consultation and we hope to develop this type of meeting with other Hospitals in Switzerland. We would that the parents participe to an ethical Conference to speak about antenatal diagnosis of SCD in November 201. Lastly, the French's experiment (ROFSED), London's (St Thomas Hospital), Quebec's and Cameroon's (Fundation Mère Enfant C.Biya) in this field will be a good resources to continue this support.

P 130

The predictive value of preoperative B-type natriuretic peptide in children undergoing heart surgery

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Introduction: Low cardiac output syndrome is frequent in children after heart surgery for congenital heart disease and may result in poor outcome and increased morbidity. In the adult population, preoperative brain natriuretic peptide (BNP) was shown to be predictive of post operative complications. In children, the value of preoperative BNP on postoperative outcome is not so clear. The aim of this study was to determine the predictive value of preoperative BNP on postoperative outcome and low cardiac output syndrome in children after heart surgery for congenital heart disease.
Methods: We examined, retrospectively, the postoperative course of 97 pediatric patients (mean age 3.7 years, range 0-14 years old) who underwent heart surgery in a tertiary care pediatric intensive care setting. NTproBNP was measured preoperatively in all patients (median 412 pg/ml, range 12–35'000 pg/ml). Patients were divided into three groups according to their NTproBNP levels (group 1: 0–300 pg/ml, group 2: 300-600 pg/ml, group 3: >600 pg/ml) and then, correlations with postoperative outcomes were examined.
Results: We found that patients with a high preoperative BNP required more frequently prolonged (>2 days) mechanical ventilation (33% vs 40% vs 61%, p = 0.045) and stayed more frequently longer than 6 days in the intensive care unit (42% vs 50% vs 71%, p = 0.03). However, high preoperative BNP was not correlated with occurrence of low cardiac output syndrome.
Conclusion: Preoperative BNP cannot be used, in children, as a reliable and sole predictor of postoperative low cardiac output syndrome. However it may help identify, before surgery, those patients at risk of having a difficult postoperative course.

P 131

Possible H1N1 associated encephalopathy of a 12 year old male with neuropsychiatric symptoms

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Introduction: Neurological complications such as seizures, encephalopathy, encephalitis, or transverse myelitis associated with Influenza A H1N1 are well known and have all been previously reported. In contrast, there are only a few reports addressing cases with predomiantly neuropsychiatric manifestations. We describe a patient with such a presentation.
Case Report: A 12-year-old male presented with a sudden onset of disorientation, amnesia, dysnomia, and regressive behavior after a two days course of a common cold. No other neurologic findings were present. The diagnosis of a H1N1 infection was made on detection of the virus in the nasopharyngeal swab specimen. Cerebrospinal fluid (CSF) and serologic testing were negative for other infectious agents (borrelia, flavivirus, HSV). Blood tests for metabolic and autoimmune disorders were normal; A urine toxicology screen was negative. Neuroimaging (CT, repeated MRI) showed no pathological features. An unspecific diffuse bifrontal slowing was detected in an otherwise normal EEG. Apart from oligoclonal bands, the CSF examination was within normal ranges. The patient showed prolonged behavioural disturbance, motor stereotypies, and intermittent disorientation. He suffered from optic and acoustic hallucinations and had an excessive need for sleep. Symptoms occured in several waves, lasting for days. Neuropsychologic testing in a seemingly asymptomatic phase showed impaired executive function and a normal IQ. The patient required a course of neuropsychological rehabilitation.
Conclusion: H1N1-associated parainfectious encephalopathy should be considered as a differential diagnosis in children presenting with acute onset of neuropsychiatric symptoms during the influenza season. The presence of oligoclonal bands in the CFS might correspond with a parainfectious immunologic reaction also in context with H1N1 infection.

P 132

Interim results of the multidisciplinary weight-loss intervention study "Keep on moving"

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Introduction: Excess body fat is associated with long term detrimental cardiovascular and metabolic effects even in youth. "Keep On Moving" is a multidisciplinary weight-loss intervention study for obese (Body mass index (BMI) >P 97) children and adolescents from 11 to 16 years and their parents. The intervention is certified by the Swiss Society of Pediatrics, and is part of the nationwide evaluation project "kidsstep obesity".
Methods: The intervention consists of medical, psychological, nutritional, and sportive mentoring, including peer coaching. The groups of 6–8 participants meet once to twice weekly over a period of 9 months, followed by less frequent meetings for another 15 months. In 2009 and 2010, a total of 32 children were enrolled. There were 3 drop outs. Before and one year after commencing the intervention the following data were obtained: physical exam, height, weight, waist and hip circumference, tanner stage, blood pressure, fasting blood glucose, insulin and lipid profiles, ASAT/ALAT, and dual energy X-ray absorptiometry (DXA) for determining whole body fat, bone mineral density (BMD) and bone mineral content (BMC). BMI was expressed as standard deviation score (SDS_{LMS}) from the median.
Main Results: At the beginning (t0) of the intervention, the mean BMI SDS was 2.6, DXA body fat percentage was 41.0%. After one year (t1) total body fat was reduced to 37.5% corresponding to a reduction of (mean ± SD) 9.0 ± 13% of the initial body fat percentage. The mean BMI SDS was reduced to 2.5.
Conclusion: The intervention study "Keep on moving" has been able to help significantly reduce the body fat mass by an average of 9% for the first two groups in the trial. The achievements of individual participants differ widely. The reduction in %body fat is not accompanied by a reduction of BMI SDS, possibly due to an increase in lean body mass induced by more physical activity.

P 133

A child in decay: Severe malnutrition due to vegan diet

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Introduction: A strict vegan diet is known to cause severe neurological damage due to vitamin b12 deficiency. To demonstrate the harmful consequences, we present the case of boy with severe impairment because of severe malnutrition.
Case report: An 8 yrs old boy was found by the paramedic team unconscious and in respiratory distress at home. His parents called emergency services due to a continuous worsening of his general condition with difficulty breathing. The boy was exclusively breastfed for the first 21 months by a vegan mother and thereafter, he was nourished on a strict vegan diet. Until the incident, he had never been seen by a doctor nor received any vaccines. The family had no permanent residence and has been on a journey around the world living in a seclusive system. He was transferred intensive care unit. Diagnostics revealed hypovolemic shock due to severe megaloblastic anemia (Hb 28 g/l, Hkt 8%, MCV 120 fl), prerenal failure, hepatic failure, and a severe coagulation disorder. This was complicated by decompensated bilateral pneumonia with evidence of corynebacterium pseudodiphtericum and RSV. Consequence of severe b12-hypovitaminosis (40 pmol/l, 150-520), he had a pancytopenia, cerebral atrophy, and a paraplegia (above L3/4) with a sacral decubitus. Severe osteoporosis resulted in recurrent pathological fractures. After initial treatment including IV vitamin b12, he was transferred to rehabilitation services continuing the peroral supplementation of nutrients. The osteodensitometry after 11 month showed improved osteodensity, but still within the range of osteoporosis. 13 months after beginning of medical treatment, improvements of motor function have been observed. Time and further investigation will reveal the exact scale of irreversible damage.
Conclusion: Children with strict vegan diets are at great risk of severe neurological damage and need specific supplementation and substitutions. It is difficult to detect the harmful effects in the early stage; for this reason, medical personnel should pay particular attention.

P 134

Subglottic stenosis after bacterial laryngotracheitis

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Background: Bacterial laryngotracheitis is a feared infection in children. In severe cases invasive ventilation is necessary. Subglottic stenosis after intubation is a known complication.
Case Report: A four-year-old boy was admitted to the emergency service with an asthma attack and silent chest. He was admitted to ICU for intensive bronchodilatatory treatment and respiratory support. The patient showed severe wheezing and inspiratory stridor but always breathed spontaneously; intubation was not necessary. Based on the clinical picture and on blood culture results, the diagnoses of invasive streptococcus group A infection with bacteremia, pneumonia and laryngotracheitis were made. The child was treated with cefuroxim 50 mg/kg tid. Because of persistent fever and enlarged mediastinum, treatment was changed to meropenem 40 mg/kg tid. Because of persistent inspiratory stridor and hoarseness, bronchoscopy was performed sixteen days after admission and revealed a pseudomembranous alteration of the glottis and subglottic area with poor vocal cord movement. Diphtheria was ruled out. Twelve days later bronchoscopy showed a slight improvement of the vocal cord mobility and trachea opening, and because the child had otherwise fully recovered he was discharged home. Two weeks later the child presented again to the emergency room with respiratory infection, severe inspiratory stridor and obstructive breathing. Bronchoscopy showed scarred tissue in the glottis and subclottic area with a glottic stenosis and a synechia of the anterior glottis leaving only a 3mm slot for air-entry. The child was referred to the ENT-Department of Lausanne where he was treated by CO_2 laser. A glottical implant was inserted after securement of the airways by a tracheostoma. Back to our unit the child and his parents are getting trained to manage the tracheostomy. The plan is to remove tracheostoma and prothesis three month after surgery.
Conclusion: Bacterial laryngotracheitis may lead to severe subglottic stenosis in the absence of preceding intubation. Surgical procedure with CO_2 laser and maintenance of airway patency should lie in the hands of well trained surgeons.

P 135

The Collodion Baby – don't give any prognosis at birth!

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Introduction: Ichthyosis is a heterogenous group of keratinizing disorders affecting the entire skin. It is characterized by hyperkeratosis, scaling and erythema. The prognosis of the different ichthyosis forms is highly heterogenous.
Methods: We give a review of literature including the recently published Ichthyosis Consensus Classification and illustrate the topic with the case reports of two newborn collodion babies diagnosed and treated in our neonatal intermediate care unit (NICU).
Results: At birth, a possible neonatal phenotype of ichthyosis is the "collodion baby (CB)," which is characterized by a hyperkeratotic membrane covering the entire body surface. The membrane usually sheds within the first week of life. The most severe but rare phenotype with the worst prognosis is Harlequin ichthyosis (HI), characterised by an armour of hyperkeratosis covering the whole body. The clinical course of the first days is compromised in both CB and HI by high transdermal fluid losses and infections. The mortality is described to be high. HI changes in severe lamellar ichthyosis (LI) or erythrodermic ichthyosis (EI) and is usually associated with growth retardation. CB transit to milder forms of LI or EI but in up to 25% changes to the "self-healing collodion baby" with very good prognosis. Another favorable form is ichthyosis prematurity syndrome, in which babies are usually prematurely born with asphyxia; this form later develops into a mild follicular ichthyosis. A conclusive diagnosis predicting the clinical course of CB at birth is not possible. Evolution of the skin manifestation as well a genetic testing based on key clinical features are the diagnostic cornerstones.
Discussion & Conclusion: The phenotypic heterogeneity and wide overlapping between different forms of ichthyosis represent a diagnostic challenge. In a first step the Harlequin and Collodion Baby has to be stabilised in a NICU. The wide range of prognosis should be communicated to the parents. In a second phase, precise diagnosis should be established in collaboration with a pediatric dermatologist. In our experience it is of utmost importance not to alienate the parents and compromise the vulnerable phase of bonding with the communication of premature and therefore wrong diagnoses.

P 136

Food preferences and taste perception in anorexia nervosa

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Introduction: Restrictive and selective eating behavior with food preferences and aversions is a characteristic symptom in anorexia nervosa (AN). Typically fat-containing food is avoided. Furthermore, very often patients with AN replace food which is rich in calories by fruits and vegetable. The aim of the present study was to evaluate if there is a relationship between food preferences and taste perception in AN.
Methods: 15 female patients with acute AN (ANacute) (BMI 16.5 ± 1.3 kg/m², aged 20.4 ± 4.7 yrs), 21 remitted female patients with AN (ANrem) (BMI 21.4 ± 2.9 kg/m², 24.2 ± 3.2 yrs., remission period 6 ± 2.6 yrs.) and 15 female healthy controls (HC) (BMI 21.6 ± 3.1 kg/m², 24.9 ± 2.7 yrs.) were included. Taste perception was evaluated using taste strips (sweet, sour, salty, bitter). Food practice and choice were assessed by self-report. Several blood parameters including hormones and leptin were analyzed. The number of fungiform papillae was quantified using digital photography and image processing. For psychological evaluation SIAB-S, EDI-2 and ASR were used.
Results: In ANacute bitter tasting foods were significantly consumed more often when compared with ANrem and HC. However, red meat, sausages, ice cream and potato chips were rarely consumed in both ANacute and ANrem. The number of fungiform papillae was significantly decreased in ANacute. Taste perception for bitter tastes within a range of concentrations correlated with serum estrogen and leptin.
Conclusions: Our results indicate a relationship between food selection and biological factors which can influence taste perception during the underweight stage of ANacute.
This study was funded by Institut Danone Ernährung für Gesundheit e.V., Germany.

P 137

Prevalence of at-risk criteria of psychosis and help-seeking behaviour – a population survey pilot

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Introduction: In early detection of psychosis, two complementary approaches are mainly followed: (i) the 'ultra high risk' (UHR) criteria of an imminent risk including attenuated psychotic symptoms (APS), brief limited intermittent psychotic symptoms (BLIPS) and a combination of a genetic risk factor and a recent persistent significant decline in functioning and (ii) the basic symptom criteria 'cognitive-perceptive basic symptoms' (COPER) and 'cognitive disturbances' (COGDIS) that partially overlap but delineate risk of different imminence. However, based on epidemiological studies that have reported much higher prevalence and annual incidence rates of psychotic-like symptoms (PLEs) in the general population than the clinical phenotype of psychotic disorders, the clinical validity of at-risk criteria had been questioned. Yet, PLEs do not equal at-risk criteria and seem to be more common. The aim of this pilot study was to assess the prevalence of at-risk criteria in the general population in clinical interviews conducted by mental health professionals.
Methods: The sample consisted of randomly selected residents of the Canton Bern (age 16–35) years. Exclusion criteria were (i) life-time diagnosis of psychosis and (ii) insufficient language skills. 60 persons (70.5%) participated in the telephone interview, 2 met exclusion criteria. At-risk symptoms were assessed using the (i) Schizophrenia Prediction Instrument, Adult version and (ii) the Structured Interview for Prodromal Syndromes.
Results: Only one person (1.2%) fulfilled the attenuated psychotic symptoms (APS) criterion according to SIPS, none the transient psychotic symptom criterion. Furthermore, 8 persons reported APS relevant symptoms but did not meet the time, frequency and severity criteria for APS. Nobody fulfilled at-risk criteria according to the basic symptom concept, although 8 persons reported relevant basic symptoms but at an insufficient frequency or as lacking change. Thus, altogether 12 persons (14.1%) had sub-threshold at-risk criteria for psychoses.
Conclusion: At-risk criteria are not as common as PLEs reported in epidemiological studies, and thus might be able to delineate a clinically relevant psychopathological state. These results, however, have to be confirmed in a larger sample.

P 138

Prevalence and Impact of Cannabis Use Disorders in Adolescents with Early Onset First Episode Psychosis

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Introduction: Previous studies on the impact of cannabis use disorders (CU) on outcome in psychosis were predominantly based on non-representative samples, often have not controlled for confounders and rarely focused on adolescent patients. Thus the aims of the present study were to assess (i) prevalence of CU, (ii) baseline and pre-treatment differences between CU and those without CU (NCU) and (iii) the impact of baseline and course of CU on 18-month outcomes in a representative cohort of adolescents with early onset first episode psychosis (EOP).
Methods: The sample comprised 99 adolescents (age 14 to 18) with EOP (onset age 14 to 17), admitted to the Early Psychosis Prevention and Intervention Centre in Australia. Data were collected from medical files using a standardized questionnaire.
Results: Prevalence of lifetime CU was 65.7%, of current CU at baseline 53.5%, and of persistent CU throughout treatment 26.3%. Baseline CU compared to NCU had significantly higher illness-severity, lower psychosocial functioning, less insight, lower premorbid functioning and longer duration of untreated psychosis. Compared to all other groups, only persistent CU was linked to worse outcomes and more service disengagement. Effect sizes were medium controlling for relevant confounders. Medication non-adherence did not explain the association between persistent CU and worse outcome.
Conclusions: Baseline CU was associated with worse baseline characteristics, but only persistent CU was linked with worse outcome. About half of those with baseline CU reduced cannabis during treatment. For these, effectively treating the psychotic disorder may already be beneficial. However, future research is necessary on the reasons for persistent CU in EOP and its treatment.

P 139

At-Risk Criteria of Psychosis: Reliability between Interviewers and Interview Modes

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Introduction: In the early detection of psychosis, two at-risk approaches are currently mainly followed: (i) the 'ultra high risk' (UHR) criteria of an imminent risk including attenuated psychotic symptoms (APS), brief limited intermittent psychotic symptoms (BLIPS) and a combination of a genetic risk factor and a recent persistent significant decline in functioning and (ii) the basic symptom criteria 'cognitive-perceptive basic symptoms' (COPER) and 'cognitive disturbances' (COGDIS). In the prospective evaluation of at-risk criteria and early intervention strategies, particularly when spanning a larger time period, however, a re-evaluation in a face-to-face interview is not always possible; thus, frequently, both face-to-face and telephone interviews are carried out. While the validity of telephone-assessed data in comparison to a face-to-face interview has already been shown for a variety of mental disorders and problems, the reliability of telephone assessments of symptomatic at-risk criteria as well as exclusion and transition criteria (past or current psychosis) has not yet been shown. We examined the interrater reliability and the reliability of telephone interviews in comparison with face-to-face interviews.
Methods: The study was conducted on 31 psychiatric in- and outpatients as well as 16 non-clinical subjects. The two interviewers (clinical psychologists) were blind to each other's results and subjects' clinical status. To account for the order of interview modes and interviewer effects, both sequence of interviewer and interview mode were varied in a counterbalanced, 2×2 cross-over design. One week was chosen as the lag time between interviews to reduce memory effects while avoiding effects due to significant changes in psychopathology.
Results: The interrater reliability for the four symptomatic at-risk criteria (COPER, COGDIS, APS and BLIPS) showed good to excellent interrater reliability (κ = .632-1.0). Further, both interviewers led to an agreement for the presence of the 22 at-risk symptoms between 60% and 100%. With regard to the presence of the four symptomatic at-risk, κ-values also showed good to excellent reliability of telephone assessments. Further, both interview modes led to comparable results with an agreement for the presence of the 22 at-risk symptoms between 80% and 100%.
Conclusion: The interrater reliability is sufficient following training of the instruments. Further, the results indicate that the use of telephone interviews to collect data on at-risk criteria is justified.

P 140

The early detection of psychosis from the perspective of child and adolescent psychiatry

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To fight the devastating consequences of psychosis, early detection and treatment of persons at-risk of developing psychosis is currently regarded a promising strategy. Two at-risk approaches have been developed on mainly adult samples: (1) the 'ultra high risk' (UHR) and (2) the 'basic symptom' criteria. Although psychoses frequently start in adolescence, it has rarely been studied they can be transferred to children and adolescents. From the few studies on pure child and adolescent samples regard to UHR-criteria, there is indication of some attenuated psychotic symptoms being potentially non-specific in adolescents and brief limited intermittent symptoms being difficult to clinically classify in children when observable behavioural correlates are missing. For basic symptoms, preliminary results in adolescent samples indicate that, similar to results in adult populations, cognitive basic symptoms may be promising candidates for at-risk criteria. Yet, as some developmental peculiarities in children have to be considered in the assessment of basic symptoms, a child and youth version of the 'Schizophrenia Proneness Instrument' (SPI-CY) has been developed. However, only a small pilot study has hitherto systematically examined the clinical validity and predictive value of at-risk its use in children and adolescents. Thus, research is needed to examine if current at-risk criteria have to be tailored to the special needs of children and adolescents. If a 'Prodromal Risk Syndrome for Psychosis' is included in DSM-V, it will be indispensable to highlight that its suitability for children and adolescents is not yet known.

P 141

BESTEKKE – Bernese study on the influence of parental mental disorder on the development of their children

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Introduction: Psychopathology and psychiatric disorders in parents present a developmental risk for their offspring. A large body of literature of the last two decades shows that the risk of these children to develop a psychiatric disorder is increased by factor 3 compared to normal population. Authors like Cummings, Davies and Mattejat propose in frame models the influence of quality of interpersonal relationship, coping strategies and environmental factors.
Methods: In a first accounter the psychopathology of the parents was described and their health status was screened by a modified Sheehan Disability Scale (SDS) and BDI. The Strength and Difficulties Questionnaire (SDQ) was administered for the children and the psychosocial risk factors in the family were measured by the Family Adversity Index. In a second step parent and their offspring are examined together, by using the K-SADs, the dyadic coping inventory (DCI), parenting style inventory (ESI) and the inventory of life quality (ILK).
Results: Since October 2008 a full population survey has been accomplished. More than 1600 consecutively administered patients were included. 299 (18%) of them with children under the age of eighteen. In the final study group (N = 70) a high correlation was found between both negative and positive parenting style and the psychopathological burden of the child.
Conclusion: Parenting style is a mechanism of transmission on children's mental health and quality of life. In our view parenting style could be a target for prevention as well as for family-based intervention.

P 142

When acute becomes chronic: Increased blood flow in frontal and cingulate brain regions in a 17-year old boy with a chronic derealisation phenomenon after having used cannabis twice

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Background: Depersonalisation/derealisation (DP/DR) phenomena are characterized by the subjective experiences of feeling unreal and detached from oneself as well as from the external world. These symptoms are relatively common in normal (1–2%) and psychiatric populations (up to 42–91% dependent on the underlying disorder). Here we investigate the cerebral blood flow (CBF) of a 17-year-old boy with a chronic derealisation phenomenon after having used cannabis twice 2 years ago. His distressing symptomatology was characterized by feeling detached, like being in a dream, with 1–2 incidents per week of being uncertain whether or not he has actually experienced or dreamt a situation.
Method: CBF was assessed with MR-arterial spin labeling (ASL) in a 3 T MR scanner (Siemens Magnetom TRIO), once in a resting state, a second measurement under provocation of the DR phenomenon.
Results: Using ASL, we found increased regional CBF in the anterior cingulate gyrus as well as in the right inferior frontal gyrus relative to other brain regions in a resting condition. Subjectively, the patient reported 4-5/10 on a visual analogue scale for experience of DR. Under provocation of DR, 7-8/10 on the visual analogue scale, these increases were even more pronounced. The very same regions showed higher activations during the fMRI measurements under provocation (level 9/10) compared with a resting state (4/10). Here, we found activation in the very same regions found in the CBF measurement: Right Broca aequivalent, dorsolateral prefrontal cortex and anterior cingulate gyrus.

P 143

Tradition in abroad? Characteristics of suicidal behaviour in adolescent migrants in Switzerland

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Background: Suicidal behaviour is a complex event. It includes an interaction of predisposing factors (e.g. psychiatric disorders), external triggers (e.g. critical life events) and promoting environmental factors (e.g. availability of weapons). Although psychiatric disorders as the strongest predictor for suicidal behaviour there are intercultural differences pertaining power of prediction for suicidal behaviour. In this regard, Nock et al. (2008) could show in migrants of high income countries that mood disorders are the strongest predictor for suicidal behaviour, whereas in migrants from low and mid income countries higher prediction power of impulse control problems for suicidal behaviour was displayed. Recent lack of research investigating the impact of migration on suicidal behaviour in adolescents leads to inconsistent results; considering migration both as a protectoral factor and a risk factor for the incidence of suicidal behaviour.
Method: Between 2004 and 2008 182 patients aged 16 to 21 years old committing a suicide attempt (defined by WHO criteria) were included and seen or treated in the emergency department of the University Hospital of Berne. In retrospective medical chart analyses ICD-10 diagnoses and sociodemographic data have been comprised.
Results: Adolescents and young adults without migration background showed significant lower indications for impulsivity (56%) compared to adolescents and young adults from a mid or low income country (75%). Borderline Personality Disorders (39% vs. 13%) and prior mental (psychiatric and/or psychological) treatment (69% vs. 39%) are more common in adolescents and young adults without any migration background. Our results are in the line with recent results showing a distinction of suicide attempted patients with and without migrant background. Downsizing of cultural and linguistic barriers seems to be necessary to provide an equivalent health care access also for migrants.

P 144

Cannabis use disorder and age at onset of psychosis – A study in first-episode patients

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Introduction: Age at onset of psychosis (AAO) may be younger in patients with cannabis use disorders (CUD) compared to those without CUD (NCUD). Previous studies included CUD co-morbid with other substance use disorders (SUD), and many did not control for confounders.
Methods: Controlling for relevant confounders, differences in AAO between patients with and without CUD excluding those with any other SUD were analyzed in a large representative file audit of 625 first-episode psychosis (FEP) patients (age 14 to 29 years) admitted to the Early Psychosis Prevention and Intervention Centre in Melbourne, Australia.
Results: Three quarters of the 625 FEP patients had a CUD. Cannabis use started before psychosis onset in 87.6% of patients. AAO was not significantly different between CUD (without other SUD, n = 201) and NCUD (n = 157). However, AAO was younger in those with early CUD (starting age 14 or younger) compared to NCUD ($F(1) = 5.2$; $p = 0.024$; partial $\eta 2 = 0.026$). Earlier age at onset of cannabis use predicted earlier age at onset of psychosis ($\beta = -0.49$, R2-change = 0.25, pb0.001).
Conclusion: Only CUD starting age 14 or younger was associated with an earlier AAO at a small effect size. These findings suggest that CUD may exert an indirect effect on brain maturation resulting in earlier AAO potentially only in cannabis sensitive subjects.

P 145

Surveillance of early onset eating disorders – a multi-national comparison

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Introduction: The ambiguous position of early-onset eating disorders (EOED: onset <age 13) between paediatric and mental health services, together with difficulties of recognition in this age group, has led to gaps in knowledge about prevalence, presentation and management. This paper describes and compares the range of clinical features and co-morbidities at presentation collected through the national paediatric surveillance units.
Methods: Prospective active surveillance was conducted in Australia, Canada and the United Kingdom between July 2002 and May. A report card/email was sent monthly to consultant paediatricians and

psychiatrists in order to ascertain cases. Clinicians reporting a case were sent a brief questionnaire requesting de-identified information.
Results: The BPSU study gave an estimated incidence of 3.03/100,000 children, comparable to Canadian (2.06/100,000) but higher than Australian (1.38/100,000) figures. Two groups of children were identified in all three countries; 64–70% of the sample exhibited symptoms consistent with anorexia nervosa (AN), while a second group (30–36% of the sample) exhibited symptoms of weight loss, active food avoidance, and somatic symptoms but would not have been classified with an eating disorder using current epidemiological screening tools and diagnostic criteria.
Conclusion: These are the first prospective incidence studies undertaken to look at EOED. Protocol sharing has allowed us to compare and contrast data. Case definition is crucial in determining incidence and may account for differences. Anorexia Nervosa may be stable, but the incidence among young females appears to have increased, and numbers of cases presenting clinically were higher than predicted. Reasons may include a lack of developmental sensitivity in current classification systems for eating disorders. Services for children with eating disorders should reflect the clinical profile and developmental needs of this patient group distinct from their older adolescent counterparts.

P 146

Interdisciplinary systemic team education in inpatient adolescent psychiatry – effects on cooperation and distress

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Introduction: To work in an acute adolescent psychiatry unit means being confronted with very challenging situations. Internal problems as staff shortage or dysfunctional communication and external conditions like conflicting requests, e.g. help versus control, lead to dismissals, be away sick, no learning from mistakes, focusing on problems and deficits, violence and terminations of treatment. The aim of the study is to investigate whether a multi-professional team education with systemic topics, like techniques in conversation and negotiation, understanding interaction processes, recursiveness and much more, helps to reduce the mentioned problems and to raise satisfaction of everyone involved.
Methods: The trial was done in a pre-post design. Standardized questionnaires of job satisfaction, just before and half a year after team education (6 days within 6 months) and also 5 years later, were given to the team. Guided interviews (what's useful in clinical practice?) completed the survey.
Results: The multiprofessional training intensifies interdisciplinary cooperation. It results in an increased appreciation of the nurses and physicians involved. Staff burnout decreased during the research period and also 5 years later, task orientation and security within teams increased. New modes of formal meetings, e.g. systemic handing over, are helpfully. Especially the adolescents embrace in every time being member of their own case conferences.
Conclusion: Interdisciplinary team education with systemic topics seems to be suitable to improve quality of patient care and interdisciplinary cooperation and to reduce staff distress.

P 147

Treatment of a female adolescent with type 1 diabetes mellitus and comorbid anorexia nervosa

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Objective: Type 1 diabetes mellitus (DMT 1) is associated with a higher prevalence of bulimia nervosa (BN) in females. Furthermore, the development of DMT 1 in preadolescence or adolescence seems to be a risk factor in girls for subsequent development of anorexia nervosa (AN), as well. The aim of this case report is the presentation of insulin misuse in a patient with AN and the therapeutical implications. We will give a review of comorbid occurrence of DMT 1 and AN.
Methods: A 17-year-old female adolescent was admitted with a BMI of 14.3 kg/m^2 (<P3) and a HbA1c of 16.2. She fulfilled all criteria of AN (ICD-10) including bingeing and had a history of 11 years of DMT 1 with distinct malcompliance. Onset of AN was with the age of 14. Since then, inpatient and outpatient episodes in child and adolescent psychiatric services were ongoing up to admittance in Littenheid. Systemical and behavioral therapy were combined with nutritional and medical treatment. For diabetes management an endocrinologist was consulted.
Results: An increase of BMI during inpatient treatment was associated with higher levels of concern about shape and weight and consequently had implications for diabetes management. Despite malcompliance during the first weeks of treatment, the adolescent achieved a continual weight gain (BMI at discharge = 18.7 kg/m^2, <P25), decrease in bingeing and an autonomous management of insulin (HbA1c at discharge = 14.0).
Conclusions: We describe the successful inpatient treatment of a female adolescent with DMT 1 and comorbid AN and subsequently surveyed catamnestic data. Insulin-purging and features associated with DMT 1 like weight gain, dietary restraint and food preoccupation complicate the treatment of AN and will be discussed.

P 148

Neonatal Morbidity in Singleton Late Preterm Infants Compared With Full-Term Infants

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Aim: To test the hypothesis that singleton late preterm infants (34 ⁰⁄₇ to 36 ⁶⁄₇ weeks of gestation) compared with full-term infants have a higher incidence of short-term morbidity and stay longer in hospital. Methods: In this retrospective, multi-centre study electronic data of children born at 5 hospitals in Switzerland were recorded. Short-term outcome of late preterm infants were compared with a control group of full-term infants (39 ⁰⁄₇ to 40 ⁶⁄₇ weeks of gestation). Multiple gestations, pregnancies complicated by fetal malformations, maternal consumption of illicit drugs as well as infants with incomplete documentation were excluded. The results were corrected for gender imbalance.
Results: Data from 530 late preterm and 1686 full-term infants were analyzed. Compared with full-term infants, late preterm infants had a much higher morbidity: respiratory distress (34.7% versus 4.6%, p <0.001), need for oxygen administration (16.6% versus 2.3%, p <0.001), hyperbilirubinaemia (47.7% versus 3.4%, p <0.001), hypoglycaemia (14.3% versus 0.6%, p <0.001), hypothermia (2.5% versus 0.6%, p <0.001), duration of hospitalization (mean 9.9 days versus 5.2 days, p <0.001). The risk to develop at least one complication was 7.6 times higher (95% CI 6.2–9.6) among late preterm infants (70.8%) than full-term infants (9.3%) and correlated negatively with gestational age.
Conclusion: Singleton late preterm infants (34 ⁰⁄₇ to 36 ⁶⁄₇ gestational weeks) show considerably higher rate of medical complications and prolonged hospital stay than matched full-term infants and therefore need much more medical and financial resources.

The numbers refer to the pages of this supplement.