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Innovations in the surgical treatment of hands in Apert syndrome

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Introduction: Apert syndrome is a congenital disorder characterized by craniosynostosis, midface hypoplasia and complex malformations of the extremities, including complex syndactylies that must be corrected to improve hand function. Lately, several conventions were given up and modifications of flaps, accepting larger skin defects and external fixateurs for soft tissue distraction were introduced.
Methods: Ten consecutive children with Aperts hands who were operated at our institution between 2004 and 2014 were reviewed retrospectively. Emphasis was given to complications, particularly after introducing new techniques.
Results: 36 syndactyly release operations were performed in ten children. Syndactyly release was performed in a two-stage operation in all cases, performing zigzag flaps in 12 and straight-line incisions in the last 24 cases. Soft tissue distraction with external fixateurs before syndactyly release of the 3rd web space was performed in 3 hands. No major complications necessitating reoperations or resulting in functional deficits occurred, irrespective of the technique. Avoiding triangular flaps in syndactyly release resulted in a natural looking distribution of pigmented and glabrous skin.
Conclusion: Straight-line incisions for syndactyly release in Apert's hands are easy to perform, give optimal cosmetic results and have minimal complication rates. The use of external fixateurs in selected complex syndactylies allows soft tissue distraction so that a five fingered hand can be reconstructed in virtually all hands. Interdisciplinary cooperation, particularly with the craniofacial surgeons, allows performing the major craniofacial and hand corrections before the age of 18 months, aiming at minimizing the negative impact on the child's development.

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Building up a National Swiss Necrotizing Enterocolitis registry: pilot projects

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Introduction: Many aspects of necrotizing enterocolitis (NEC) are barely known such as epidemiology, etiology, disease severity modifiers and outcome, making NEC an important candidate for multicenter analyses. Therefore, we would like to build up a Swiss NEC registry, including all preterm and term-born NEC patients. In a pilot study we analyzed the currently available national neonatal dataset (Swiss Neonatal Network) and a local NEC-registry.
Methods: First, we evaluated the number of all NEC patients registered by the Neonatal Network and second, we performed a pilot study in Berne. After approval from the local hospital ethics committee, we developed a database and identified retrospectively all NEC patients since 1977. Their data were extracted from medical records for a first analysis.
Results: The Neonatal Network registered 231 patients (birth weight <1500 g) with proven NEC (2000–2012). Of those 32% (N = 75) deceased. In Berne, we identified 136 patients with proven NEC (1977–2010). Median birth weight was 1520g, median gestational age was 32 weeks and mean age at diagnosis was 12 days ± 11.9. 58% of all patients were treated conservatively and 42% underwent surgery. Total mortality of the Bernese cohort was 23% (N = 31).
Conclusion: Since the Neonatal Network registers only patients <1500 g, almost 50% of all NEC patients may not be registered. According to the extrapolation of data we expect to register retrospectively ~300–500 patients (2000–2013) and prospectively ~30–40 patients per year. The establishment of this registry will provide the basis for systematic research and possible improvement of diagnostics and treatment of NEC in Switzerland.

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Validation of a postoperative parental pain assessment tool in a French-speaking population

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Aim: Evaluation at home of children's postoperative pain by their parents represents a real challenge. Recently, Parent's Postoperative Pain Measurement (PPPM) score has been validated in English and subsequently translated in several languages. The aim of this study is to validate it in French before its implementation.
Method: First, PPPM was forward and backward translated. Following Ethics approval and parental consent, we recruited between 2009–2011 bi-parental families of children age 2–12 admitted to our institution for outpatient surgery. Non French-speaking families and children with chronic diseases were excluded. Face Pain Scale-Revised(FPS-R) tool was used as the comparator. Each parent received randomly either FPS-R or PPPM to assess pain 24 and 48h after surgery. Children age ≥6 filled their own FPS-R. Parents and child were instructed not to discuss the evaluation.
Results: 279 families were recruited and complete data was obtained for 99.Mean age was 74 ± 34.4 months, 66.7% were boys. Surgical procedures included ENT (55%), uro-genital (19%), abdominal wall (11%), orthopedics (8%) and skin/subcutaneous tissues (7%). Factorial analysis confirmed a single dimension on PPPM. Spearman's rho showed good correlation (0.657; p = 0.01) between the parent's tools. Correlation was also good (0.579; p = 0.01) between parent's PPPM and child's FPS-R. Paired t-test for 24 and 48h postoperative showed significant (p <0.001) decrease in pain on PPPM and FPS-R. Internal consistency was confirmed by Cronbach's alpha (0.84). Repeated measurements ANOVA did not reveal evidence for a difference in pain assessment between child's gender/age and parents' gender.
Conclusion: Our results support the construct validity and reliability of the translated PPPM and promote its implementation in clinical practice.

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Mind the gap

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Introduction: Between 2009 and 2013 seven newborns were treated for Type I/II esophageal atresia at the University Children's Hospital Zurich. Here we present these cases with regard to the therapeutic algorithm.
Methods: Chart review was performed retrospectively. Postnatal diagnosis was based on babygramm and tracheobronchoscopy. After confirmation of the Type I/II esophageal atresia, children were treated according to the following algorithm: First a Stamm gastrostomy was performed. After consolidation of the gastrostomy, a contrast study to elucidate the pouch anatomy was done. Depending on the length of the gap several axial approximations and delayed end-to-end anastomosis of the esophageal pouches were performed.
Results: All patients underwent prenatal ultrasound. In five fetuses a polyhydramnios was documented. As of today, five children were successfully treated with primary end-to-end anastomosis of the esophagus. One child is waiting for the final operation. One child died due to congenital heart disease before esophageal continuity could be established. 10 days after surgery 4/5 patients showed an initial leakage in the contrast study. Another 10 days later, all leaks had disappeared spontaneously. Further follow-up was done at our hospital in four patients. Esophageal stenosis requiring dilatation was necessary in 4/4 and GER was diagnosed in 2/4 patients.
Conclusions: Longitudinal approximation and generous waiting time have a distinct positive effect on lengthening of the esophageal pouches and allow a successful delayed primary end-to-end anastomosis and thus salvage of the patients own esophagus.

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Retroauricular versus inguinal full-thickness skin grafts in syndactyly repair

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Objectives: Hand malformations in children require surgical correction using full-thickness skin grafts. Different donor-sites can be chosen. This study was conducted to evaluate whether changing the surgical technique from harvesting grafts from the groin to the retroauricular region results in a significant improvement regarding surgical outcome as well as patients' and parents' satisfaction.
Methods and patients: Our cross-sectional study includes 26 children (mean age 8.08, ranging 1 to 16 years), who underwent full-thickness skin transplantation on the hand with a graft from the groin (15 pts.) or retroauricular region (11 pts.) at the Children's Hospital Zurich. All patients were examined in the outpatient clinic. Pigmentation and vascularization of the skin grafts and the surrounding skin were evaluated with the Pantone Skin Tone Guide. Patient, parent and observer opinion were assessed with questionnaires based on the Patient and Observer Scar Assessment Scale (POSAS).
Results: Two sample t-tests were performed to assess the new technique. It showed a significant improvement of the pigmentation match ($p = 0.003$). Observer assessment was improved for the variables pigmentation ($p = 0.000$), general opinion ($p = 0.000$) and the total score ($p = 0.000$) on the recipient site, while parent's general opinion on the donor site improved as well ($p = 0.017$). However, no significant difference for the parent's questionnaire on recipient site was found.
Conclusion: Our results suggest that changing to retroauricular skin grafts improves the surgical outcome and parents' general opinion significantly. The better color match between skin graft and the recipient site contributed mostly to the improved parents' satisfaction.

Young Researchers' Day / Clinical Researchers' Day (Swiss PedNet)

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Randomised controlled trials in very preterm infants: does inclusion in the study result in any long-term benefit?

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Introduction: Since the introduction of randomised controlled trials (RCT) in clinical research, there has been discussion of whether enrolled patients have worse or better outcomes than comparable nonparticipants. The objective of this study was therefore to investigate whether very preterm infants randomised to a placebo group in a RCT have equivalent neurodevelopmental outcomes to infants who were eligible but not randomised (eligible NR).
Methods: In the course of an RCT investigating the neuroprotective effect of early high dose erythropoietin on the neurodevelopment of very preterm infants, the outcome data of 72 infants randomised to placebo were compared with those of 108 eligible NR infants. Our primary outcome measures were the mental (MDI) and psychomotor (PDI) developmental indices of the Bayley Scales of Infant Development II at 24 months corrected age. The outcomes of the two groups were considered equivalent if the confidence intervals (CIs) of their mean differences fitted within our ± 5 point margin of equivalence.
Results: Except for a higher socioeconomic status of the trial participants, both groups were balanced for most perinatal variables. The mean difference (90% CI) between the placebo and the eligible NR group was –2.1 (–6.1 and 1.9) points for the MDI and –0.8 (–4.2 and 2.5) points for the PDI (in favour of the placebo group). After adjusting for the socioeconomic status, maternal age and child age at follow-up, the mean difference for the MDI was –0.5 (–4.3 and 3.4) points.
Conclusions: Our results indicate that the participation of very preterm infants in an RCT is associated with equivalent long-term outcomes compared to non-participating infants.

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Clinical course and therapeutical approach to varicella zoster virus infection in children with rheumatic autoimmune diseases under immunosuppression

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Introduction: Children with autoimmune diseases are frequently treated with immunosuppressive medication to decrease disease activity. In the last decade, an growing number of children are treated with biological agents (BA). This iatrogenic immunosuppression (IS) may also modulate susceptibility and clinical presentation of common pediatric diseases like infections with varicella zoster virus (VZV). There are little data concerning clinical course, potential complications and therapeutical approach in this situation.
Methods: In this retrospective multicentre study, we assessed the clinical course and therapeutical approach to varicella and herpes zoster (HZ) in children under IS. Eligible for inclusion were children with rheumatic autoimmune diseases followed in a Swiss centre for pediatric rheumatology and treated with disease-modifying antirheumatic drugs and/or BA treatment and occurrence of varicella or HZ between 2004 to 2013.
Results: 22 patients were enrolled, presenting in 16 patients as varicella and 6 patients with HZ. 20 patients were treated for juvenile idiopathic arthritis, 1 for a polyglandular autoimmune syndrome type III, 1 for uveitis. Median age at VZV disease was 6.1 years (range 2 to 17 years). The median interval between start of IS and varicella or HZ was 11 months (range 6 to 63 months). 2 patients had been vaccinated (1 dose each) prior to IS. 10 patients were treated with Methotrexate (MTX) alone, 2 with BA monotherapy, 10 with a combination of BA and prednisone, MTX or Leflunomide. In the varicella group, clinical course was uncomplicated in 12 patients. 4 patients (25%) experienced complications: cellulitis in 1 patient treated with MTX, and cellulitis, sepsis and cerebellitis (1 each) in 3 patients treated with BA and MTX combination therapy. 5 children were hospitalized (range of duration 4 to 9 days). No complications occurred in patients showing HZ, with 1 patient being hospitalized. 14 patients (64%) were treated with valaciclovir or acyclovir, of whom 6 intravenously. 54% continued IS treatment during varicella/HZ.
Conclusion: The clinical course of varicella and HZ in children under IS is heterogenous, with 4 of the total 16 children showing a complicated course. Thorough assessment of VZV infection, vaccination history and correct VZV vaccination according to national guidelines at diagnosis of a rheumatic autoimmune disease is essential to minimise VZV complications during a later immunosuppressive treatment.

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Drug survival and switching of biological agents in systemic juvenile idiopathic arthritis

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Introduction: Several biologic agents have become available for the treatment of systemic juvenile idiopathic arthritis (SJIA) over the last decade. Prescription strategies may depend on the heterogeneity of the disease course and other factors.
Objectives: To assess drug survival of biological agents (BA) in SJIA patients and to describe reasons for switching or discontinuating a biologic treatment.
Methods: Retrospective observational study on SJIA patients treated between 2005 and 2012 in a French pediatric rheumatology reference center using the CEMARA register.
Results: 77 SJIA patients were included, with a median age of 3.8 years at diagnosis [range 5 months to 15.2 years]. Median duration from diagnosis to start of the first BA was 18.2 months [range 0.0 to 11.5 years]. The cumulative follow up on biologics represented 244

patient-years. First-line BA were anakinra (ANA) in 51, etanercept (ETA) in 12, canakinumab (CAN) in 10, tocilizumab (TCZ) in 2, abatacept in 1 and adalimumab in 1 patient. Switching of biological therapy was common, with 44.2% switching to a second, 24.7% switching to a third, and 5.2% to a fourth BA. Reasons for switching were lack of efficacy in 63%, loss of response 24%, adverse event 7%, convenience of use 4% and patient's choice in 2%. Drug survival for ANA/CAN/ETA/TCZ at 3 and 12 months of *first-line* treatment was 82/100/75/100% and 63/100/58/100%, respectively. Drug survival for ANA/CAN/ETA/TCZ at 3 and 12 months of *second- or third-line* treatment was 71/95/75/93% and 75/63/43/82%, respectively. The rate of inactive disease at last follow up was 48.1/55.8/63.6% with a first/second/third BA, and increased to 64.9% after switch to a fourth BA. ANA, CAN and TCZ did not differ in achieving inactive disease as a first, second, third or fourth biotherapy; however, ETA treatment led to a significantly lower rate of inactive disease when compared to ANA/CAN. Biologic treatment was stopped in 11 patients (14.3%) after a median of 36 months (range 8.9 to 69.3), all of them treated with ANA as the only BA. This subpopulation did not significantly differ in baseline characteristics from the group that continued treatment.
Conclusion: Switching to a second, third or fourth BA is an appropriate approach for treatment of SJIA to increase the probability for inactive disease. ANA, CAN and TCZ showed comparable overall rates of inactive disease. Inactive disease was less often observed with ETA treatment when compared to ANA or CAN.

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Pathogen identification in paediatric sepsis results in streamlining of critically important antimicrobials

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Introduction: International guidelines emphasize the need for empiric broad-spectrum antibiotic therapy in cases of sepsis. However, it is recommended that antibiotics are rapidly streamlined when microbiological results become available. We assessed the extent of streamlining in a Swiss cohort of children with sepsis.
Methods: Data on 322 episodes of pediatric (age >1 month–17 years) culture-proven sepsis included in a prospective Swiss national pediatric sepsis cohort from September 2011 to November 2013 were analysed. Exposure to three critically important antibiotic classes (CIAs – penicillins plus beta-lactamase inhibitor, higher-generation cephalosporins, and carbapenems) was assessed, as those are among the main targets of antimicrobial stewardship.
Results: Information on causative pathogens and exposure to CIAs is shown for 312/322 episodes with complete data below.

	Exposure to CIAs during definitive therapy		
Exposure to CIAs during empiric therapy	**Yes**	**No**	**Total**
Yes	161 (51.6%)	66 (21.2%)	227 (72.8%)
No	15 (4.8%)	70 (22.4%)	85 (27.2%)
Total	176 (56.4%)	136 (43.6%)	312 (100%)
Gram negative pathogens		127/312 (40.7%) episodes	
Gram positive pathogens		178/312 (57.1%) episodes	
Fungal		7/312 (2.2%) episodes	
[Exposure to CIAs and pathogen spectrum]			

Streamlining resulted in a 23% reduction of CIA regimens from empiric (72.8%) to definitive (56.4%) therapy (p <0.01). The 66 patients switched from CIAs to alternatives were most commonly definitively treated with penicillins (61%), glycopeptides (12%), and penicillins plus aminoglycosides (8%).
Only 6.9% (9/131) of Gram-negative sepsis episodes, but 32.8% of Gram-positive sepsis episodes were never exposed to CIAs (p <0.01). Definitive CIA-therapy was administered in 82.4% (108/131) of Gram-negative, but only in 39.1% (68/174) of Gram-positive sepsis episodes (p <0.01).
Conclusion: As broad-spectrum agents, CIAs are widely used for the empiric therapy of sepsis in Swiss children. Streamlining to non-CIA regimens occurred in 1/5 of children initially started on CIA regimens. Identification of Gram-negative bacteria more often results in definitive CIA regimens.

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New combined serum creatinine and cystatin C quadratic formula for glomerular filtration rate assessment in children

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Introduction: Estimating glomerular filtration rate (eGFR) is important in clinical practice. In order to find the best formula for eGFR, this study assessed the best model of correlation between sinistrin clearance (iGFR) and the solely or combined cystatin C (CysC) and serum creatinine (SCreat) derived models. It also evaluated the accuracy of the combined Schwartz formula across all GFR levels.
Method: Two hundred and thirty-eight iGFRs performed between January 2012 and April 2013 for 238 children were analyzed. Regression techniques were used to fit the different equations used for eGFR (i.e.: logarithmic, inverse, linear and quadratic). The performance of each model was evaluated using the Cohen's Kappa (κ) correlation coefficient and the 30% of accuracies were calculated.
Results: The best model of correlation between iGFRs and CysC is linear, but presents a low κ coefficient (0.24) and is far below the KDOQI targets to be validated with only 84% of eGFRs reaching the accuracy of 30%. SCreat and iGFRs showed the best correlation in a fitted quadratic model with a κ coefficient of 0.53 and 93% of accuracy. Adding CysC significantly (P <0.001) increases the κ coefficient to 0.56 and the quadratic model accuracy to 97%. Therefore, a combined SCreat and CysC Quadratic formula is derived and internally validated using the Cross-validation technique. This Quadratic formula significantly outperforms the combined Schwartz formula which was biased for iGFR ≥91 ml/mn per 1.73 m².
Conclusions: This study allowed deriving a new combined SCreat and CysC Quadratic formula that could replace the combined Schwartz formula which is only accurate for children with moderate chronic renal failure.

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Midazolam efficacy and safety as first-line treatment of neonatal seizures. A retrospective study

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Introduction: Midazolam is used as first-line treatment for neonatal seizures in the University Hospital of Lausanne (CHUV) and many other hospitals. It offers a short elimination half-life, has a rapid effect and is easy to administer (i.e. intranasal) compared to phenobarbital, widely considered the first-line antiepileptic drug (AED) for this indication. No clinical trial has investigated the efficacy and safety of midazolam as a first-line treatment of neonatal seizures.
Methods: We retrospectively studied 75 patients with neonatal seizures from the neonatology unit of the CHUV from 2007 to 2013 who received midazolam as first AED. Gestational age, diagnostic groups, timing of seizures and midazolam administration, dosage, route and need for others AEDs were studied during the 72 hours following admission. Midazolam efficacy was defined as seizure cessation without any recurrence and absence of need for another AED within 5 hours. Respiratory and hemodynamic parameters before and after midazolam administration were evaluated.
Results: The studied population consisted of mainly term infants with asphyxia. The mean midazolam dose was 0.11 ± 0.05 mg/kg. Efficacy was observed in 39 patients (52%). In the post-asphyxic group, 41% (n = 18) responded to midazolam, 69% (n = 9) in the hemorrhage group. Intranasal route was only used for term infants. There were no major side effects.
Conclusion: Midazolam appears a simple, quick and well tolerated therapy for the management of neonatal seizures before IV phenobarbital. In order to better delineate the position of midazolam compared to phenobarbital in the management of neonatal seizures, a prospective study is needed.

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Feasibility of multiple-breath washout in inexperienced preschool children

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Introduction: Multiple breath washout (MBW) has gained increasing interest as lung function test in recent years among others based on ERS/ATS guidelines. In certain lung diseases (CF and PCD), MBW is becoming established in clinical routine. However, feasibility in preschool children with and without lung disease using commercially available equipment is unknown. We aimed to assess feasibility of nitrogen (N_2) MBW measurement in preschool children in a setting not experienced with MBW measurement.
Methods: N_2-MBW measurements were performed in 67 children (22 females) aged (mean ± SD) 5.4 ± 1.0 years with various lung diseases. A preset time frame of 20 minutes was applied. All children used a mouthpiece and nose clip and watched a soundless movie to ensure relaxed tidal breathing.
Primary outcome was feasibility of N_2-MBW using standardized quality control criteria.
Results: At least 1 valid MBW trial was possible in 55 (82%) of 67 preschool children. Main reasons for failure leading to exculsion of children (n) were leaks (n = 6), irregular breathing pattern before or during washout (n = 5) and washout target not reached (n = 1). 16 children achieved 1 good trial (5.4 ± 0.9 years), 21 children achieved 2 good trials (5.5 ± 1.0 years) and 18 children achieved 3 good trials (5.8 ± 0.6 years).
Conclusions: MBW using commercially available equipment can be successfully performed in the majority of young children. This suggests that even in a MBW-inexperienced center, MBW can be performed in young MBW-naïve children on a routine basis.

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Certainty about mental states in adolescence: association with age and schizotypal symptoms

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Introduction: Mentalizing – attending to mental states in oneself and others – is the basis of healthy relationships and self-awareness as well as an essential common factor among psychotherapeutic treatments. Mentalizing is considered as a developmental achievement, easily hampered in self-disorders. However, investigations have for now mostly focused on borderline personality disorder in adulthood. With this in mind, the present study aims to assess mentalizing in oneself and others related to age and schizotypal traits, which typically emerge during adolescence and may predict severe mental disorders in adulthood.
Methods: 125 adolescents (66 female, Mage = 15.88, SDage = 1.88) and 252 adults (163 female, Mage = 23.20, SDage = 2.68) from the community participated in this study. They fulfilled the RFQ to evaluate their degree of certainty (RFQc) and uncertainty (RFQu) about mental states in oneself and others together, and the SPQ to assess the level of positive, disorganized and negative schizotypal traits.
Results: We first found a lower level of RFQc ($Z = -3.25$, $p = .00$) and a higher of RFQu ($Z = -5.73$, $p = .00$) in adolescents than in adult group. Second, in the adolescent sample, the level of RFQc was negatively associated with disorganized ($rs(124) = -.216$, $p < .05$)) and negative ($rs(124) = -.216$, $p < .05$)) dimensions of schizotypal traits.
Conclusions: Consistent with what we might expect from recent neuroimaging findings, the development of mentalizing skills is still on-going beyond childhood. Moreover, we provide original data suggesting that the degree of certainty about self and other mental states might provide resiliency against negative and disorganized schizotypal personality traits in adolescence. Adolescents might therefore benefit from a clinical practice that draws attention onto the enhancement of mentalizing skills.

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Making paediatric neuroimaging child's play – (f)MRI research studies in children and adolescents

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Introduction: The advent of magnetic resonance imaging (MRI) has led to an increase in the use of functional and structural neuroimaging techniques investigating the neural basis of perception, cognition and behaviour. Although advances in MRI methodology and techniques are apparent, research studies in younger age groups remain less frequent.
Methods: All (f)MRI data presented is analysed using customized analysis tools and standard software packages (e.g. spm5). We here review strategies, analysis and imaging protocols suitable for neuroimaging in non-sedated children/adolescents on the background of two large-scale research studies: The *Boston Longitudinal Study of Dyslexia* (BOLD: characterizes behavioural/neuronal phenotype of children with and without a familial risk for dyslexia from age 5 on) and a Basel-based cross-sectional study on *Empathy Development* (target group: typical adolescents and youths with conduct or autism spectrum disorder).
Results: Applied examples and recommendations for pediatric neuroimaging studies presented here indicate the potential of (f)MRI in research and clinic and suggests a tool to potentially identify at-risk children early on. Particularly, study findings thus far suggest that the development of characteristic networks fundamentally differs in at risk children or clinical cases when compared to typically developing peers.
Conclusion: An increased understanding of the characteristics of childhood disorders is essential for developing novel and improving existing intervention programs, and may therefore prevent negative clinical, psychological and social outcome. Overall, increased use of pediatric neuroimaging in research studies may help bridging the gap in knowledge about brain structure, function and development in younger age groups.

Ausgewählte Abstracts fPmh / Abstracts sélectionnées fPmh

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Translating neurophysiology and imaging in ADHD

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Introduction: Attention-Deficit/ Hyperactivity Disorder (ADHD) is a highly prevalent persistent disorder with childhood onset. Despite heterogeneity and developmental changes, neurophysiology and imaging reveal systematic alterations in ADHD patient's brain systems for state regulation, attention, inhibition and motivation. Translating these findings into diagnostic or subtyping aids into clinical routine has been encouraged by the reliability of brain markers for development and attention. However, the most current diagnostic scheme (DSM-5) explicitly states that neurophysiological or imaging based biomarkers which may be altered in ADHD are not diagnostic.
Methods and Results: Despite much research and commercial efforts and some opposing claims, most promising markers have not proven sufficiently diagnostic or prognostic for clinically defined ADHD in typical settings [1, 2]. This holds for simple and multimodal imaging, tests, and pattern classification approaches. Similarly, ADHD treatments based on consistent deviance such classical neurofeedback may not sufficiently tap into disorder-specific mechanisms.
Conclusions: One road to progress is to focus on homogenous neuroscience-based subtypes and clarify their predictive power for individualized treatment [3], while ensuring that the current expertise with ADHD as a heterogeneous disorder is not "lost in translation".

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Urological complications following paediatric kidney transplantation – challenges in management and outcome

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Introduction: Paediatric kidney transplantation is surgically challenging. Urologic complications do occur and can be harmful to patients and graft survival.
Methods: We performed a retrospective study evaluating urological complications, their management and long-term outcome after paediatric kidney transplantations performed at our institution between 2000 and 2013.
Results: 75 patients underwent kidney transplantation. 38 (51%) transplanted kidneys were from living donors. Mean age at the time of transplant was 11.2 years (range 2.4–20). 3 patients required a second renal transplant. 3 patients died, 2 due to multi-organ-failure, 1 due to vascular complications. None of the deceased patients had urologic complications.
Urological complications requiring surgical management occurred in 10 patients (13%). 2 ureteral obstructions: 1 managed by JJ stent and ESWL for urolithiasis, 1 by percutaneous nephrostomy and percutaneous drainage of a perirenal lymphocele followed by laparotomy and open marsupialisation. 7 patients with VUR needed surgical management for recurrent febrile urinary tract infections (UTI's): 3 were managed by open ureterocystoneostomy (UCN), 4 had subureteric Deflux-injection. Following endoscopic treatment only 1 patient had no further episodes of UTI. The remaining 3 had persisting VUR and UTI's were managed conservatively. UCN was successfully performed in 3 patients, 1 combined with nephroureterectomy of the left remaining kidney. 1 UCN was performed pre-emptively before planned implantation of an artificial sphincter for urinary incontinence. Transient postoperative obstruction occurred in one patient requiring JJ insertion. Currently 1 further patient is awaiting UCN in combination with bladder augmentation and Mitrofanoff-stoma.There was no graft loss secondary to urologic complications requiring surgery.
Conclusions: Early identification of complications and individualized treatment in the context of underlying urologic pathology is critical to preserve graft survival. Therefore experienced multidisciplinary teams are required with close coordination between the paediatric nephrologist, transplant and paediatric urologic surgeons. Subureteric injection of bulking agents for VUR into the transplanted kidney is technically feasible, but was not very successful in our hands. Primary UCN may be considered for reflluxing transplanted kidneys since this technique was successful in all patients.

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Lost in transition? What happens when childhood cancer survivors become adults

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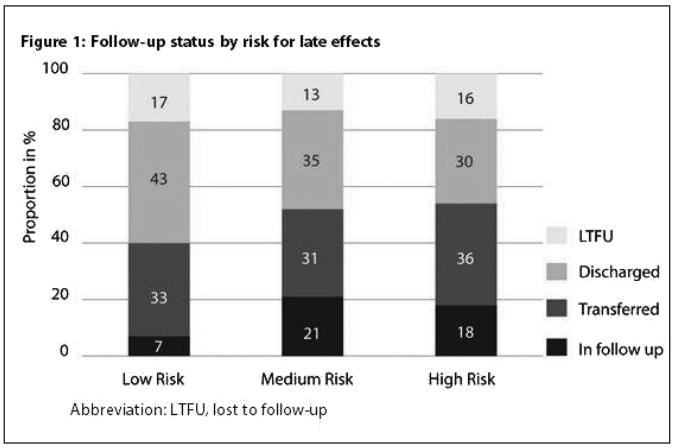
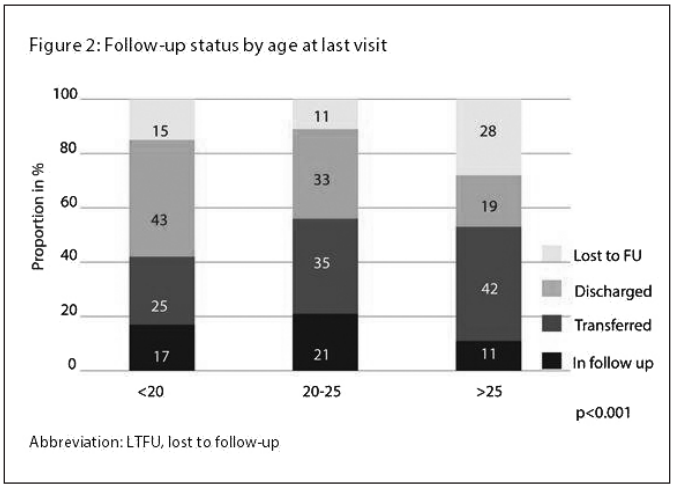
Background: Transition to adult care in pediatric oncology is important to avoid loss to follow-up. It is however particularly challenging because patients are often healthy individuals with a higher risk to develop late-effects sometime in the future. Despite its importance, it is still unclear what happens in terms of care when childhood cancer survivors (CCS) become adults in Switzerland. We aimed to describe 1) the follow-up status (FUS) of CCS (in pediatric follow-up, transferred, discharged without referral or lost to follow-up); a. by risk for late-effects, b. by age at last visit and 2) the transfer destination if transferred to adult care.





Methods: We conducted a chart review of a random sample of CCS registered in the Swiss Childhood Cancer Registry (SCCR) aged ≥16 years at study, diagnosed <16 years and who survived ≥5 years since diagnosis. For the analysis we obtained clinical variables from the SCCR. We coded risk for late-effects in low, middle and high according to diagnosis and treatment.
Results: We included 746 survivors (availability rate = 70%; mean age at last visit = 19.1 years, SD = 4.1). Overall, 134 survivors (18%) were still in pediatric follow-up, 231 (31%) had been transferred, 269 (36%) discharged, and 112 (15%) were lost to follow-up. FUS did not reflect the risk for late-effects: 46% of survivors in the high-risk category were discharged or lost to follow-up (fig. 1). FUS by age at last visit showed that 18% of survivors were still in pediatric follow-up in adult age and that the proportion of survivors lost to follow-up increased with age (28%; fig. 2). When transferred, survivors were most often sent to a general practitioner (55%), followed by adult oncologists (20%), neurologists (13%), or other specialists (12%).
Conclusions: Our findings showed that follow-up does not take into account the risk for late-effects or patient's age speaking against the systematic transition of CCS from pediatric to adult care. Earlier, risk-oriented transition could help to organize care and avoid loss to follow-up. Not to forget is the apparent important role of general practitioners.

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Pigmented dermo-epidermal skin substitutes in a long-term *in vivo* assay

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Purpose: Human pigmented dermo-epidermal skin substitutes are being constructed and tested in animal models since several years. Yet, little is known about the long-term biology of the epidermal and dermal compartment after transplantation. In this experimental study we constructed human melanocyte-containing dermo-epidermal skin substitutes and studied them in a long-term animal experiment.
Methods: Keratinocytes, melanocytes, and fibroblasts from human skin biopsies of various pigmentation types were isolated, cultured, and expanded. Melanocytes and keratinocytes were seeded in a ratio of 1:5 onto collagen gels containing fibroblasts. These skin substitutes were then transplanted onto full-thickness wounds of immuno-incompetent rats and investigated up to 15 weeks after transplantation.
Results: Chromameter evaluation showed a clear-cut color difference between light and dark pigmented skin substitutes but a consistent skin color over several weeks after transplantation for each single transplant. Histological analyses of the substitutes showed a mature epidermis in a homeostatic state, melanocytes in the epidermal basal layer in a physiological ratio, and melanin localized in keratinocytes in a normal supra-nuclear position 15 weeks after transplantation. Dermal components such as fibrillin and elastin showed a developmental stage near to that of normal skin.
Conclusion: These data suggest that pigmented dermo-epidermal skin substitutes show a promising development towards achieving near normal skin characteristics and tissue homeostasis in both epidermal and dermal compartments. In particular, data also suggest that melanocytes remain in a physiological epidermal position, function correctly over several months, and yield a skin pigmentation that resembles the original color of the donor skin.

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Reduction of drooling after crysdale procedure in children with neurological disability

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Introduction: Many children with neurological disability, also present with severe drooling. Therapeutic means used to radically suppress drooling and therefore salivation, bring along many side effects. Crysdale surgery seems to be a good therapeutic option to reduce drooling while preserving salivation.
Objective: To evaluate the benefits after Crysdale procedure (submandibular ducts relocation and sublingual glands resection) in children with neurological disability presenting severe drooling.
Methods: Retrospective cohort study involving children <18 years old, who underwent Crysdale procedure between 2002 and 2012. A questionnaire was designed, based on the drooling rating scale (Suskind, 2002) and regarding the intensity of drooling and its effects on quality of life and social interactions with the child's living environment.
Results: 20/24 questionnaires were collected (3 non-response, 1 death). Median age was 13years old, with a 1.5/1 = M:F sex ratio. 13 patients (65%) had constant drooling before surgery, vs 30% after ($P < 0.01$). 50% of parents confessed drooling before surgery was limiting the child in doing activities outside his home, whereas after surgery 70% of them were mildly or not limited ($P = 0.05$). Other items including number of bib or shirt changes per day as a result of excessive drooling, wiping frequency, drooling while eating or at night, and during activities involving attention, all showed improvement after surgery.
Conclusions: Following Crysdale procedure, there is significant decrease of drooling with subsequent improvement in certain aspects of daily functioning in children with neurological disability

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Lung growth in healthy children from infancy to early childhood – longitudinal data from a Swiss Birth Cohort (BILD study)

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Background: Knowledge regarding lung growth during early childhood is limited. This study was undertaken to describe the physiologic increase of lung volume during early childhood, and the impact of gender, growth and exposure to environmental tobacco smoke (ETS) during the first year of life.
Methods: Term-born children were recruited for a prospective birth cohort study (BILD study: Basel & Bern Infant Lung Development). We measured Functional Residual Capacity (FRC) repeatedly in 210 children using Multiple Breath Washout (MBW, FRC_{MBW}) at the age of 5 weeks and 6 years. Additionally, at the age of 6 years we measured FRC using plethysmography (FRC_{pleth}). We investigated potential influencing variables using regression.
Results: High quality measurements of n = 140 children (67 females) were analysed, mean (SD) age was 34.4 (3.9) days and 6.1 (0.2) years respectively. At neonatal age (n = 140), mean (SD) FRC_{MBW} was 101 (16.4) mL. At the age of 6 years, mean (SD) FRC_{MBW} (n = 86) was 651 (139) mL, and FRC_{pleth} (n = 87) was 1094 (198) mL.
Regression shows a weak tracking for absolute FRC values (FRC_{MBW}: $R^2 = 0.29$); higher increase of lung volume was found in boys ($p = 0.02$), children with higher weight gain ($p < 0.001$), and exposure to ETS ($p = 0.001$).
Discussion: This is the first study showing tracking of lung volume in a longitudinal data set from infancy to early childhood. The findings of higher lung volume increase in boys, children with higher weight gain and children exposed to ETS indicate an impact of genetic as well as environmental factors on lung growth.

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Prevalence of cough throughout childhood: a cohort study

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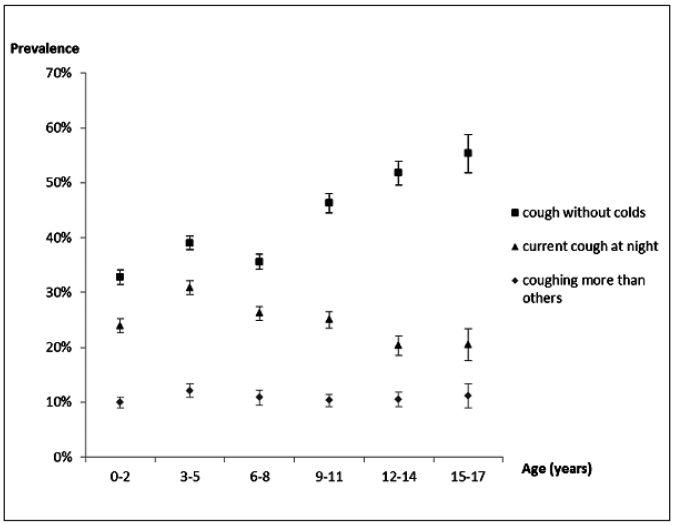
Introduction: Cough in childhood is an important cause of ill health and primary care visits. However, data on the natural history of cough in unselected children are scarce. This study describes prevalence of parent-reported cough throughout childhood, assessed by standardized questions.
Methods: In a population-based cohort study in Leicestershire, UK, we analysed respiratory outcomes from five questionnaire surveys, performed in 1998, 2001, 2003, 2006 and 2010. We investigated the following variables: night cough, cough with or without colds, cough triggered by exercise, dust or pollen, and coughing more than others. We calculated prevalence of cough at ages 0–2, 3–5, 6–8, 9–11, 12–14 and 15–17 years.



Prevalence of cough throughout childhood

Results: We analysed data from 6,808 children (response rate of 78% in 1998). Prevalence of cough without colds increased from 33% in the youngest to 55% in the oldest age group (figure). Similarly, prevalence of cough triggered by exercise, dust and pollen increased with age. Throughout childhood around 70% of children had cough with colds and around 11% were reported to cough more than peers. Prevalence of night cough was highest in 3-5 year-olds (31%), decreasing to 20% in 15–17 year-olds.

Conclusion: We found significant changes of cough prevalence from infancy to adolescence, depending strongly on the wording of the questions relating to cough. These changes might reflect developmental or environmentally mediated influences, which should be taken into account when planning studies and in the clinical approach to patients.
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A prospective study on the impact of biometric and environmental co-factors on human rhinovirus infections in healthy infants

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Introduction: There is increasing evidence that human rhinovirus (HRV) infections have detrimental effect on respiratory health, but studies on HRV-associated morbidity and the relationship with host and environmental co-factors in otherwise healthy populations, especially infants, are scarce.
Methods: Nested in a prospective birth cohort of infants of unselected mothers, respiratory health was assessed weekly in 41 subjects by telephone interviews during the first year of life. Concomitant weekly nasal swabs were obtained from the infants to determine prevalence of HRV. We used a multilevel logistic regression model to assess which biometric and environmental co-factors influence this association.
Results: We found a high prevalence of HRV in our study cohort (363/1,325 samples tested positive for HRV), associated with a seasonal effect mainly during autumn months and older siblings. Among infants with HRV detection, 51% were clinically asymptomatic. Respiratory symptoms during HRV infections were less likely during the first three months of life. A maternal history of atopy was independently associated with less symptoms during HRV infection.
Conclusions: HRV is highly prevalent in unselected term-born infants during the first year of life. While environmental factors have significant impact on HRV prevalence, individual co-factors are associated with respiratory morbidity during HRV infections. These findings shed new light on risk factors contributing to HRV-associated infant morbidity and will help to improve treatment and prevention strategies.
Authors N. Regamey and P. Latzin contributed to this abstract equally.

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Investigation of youths with bipolar spectrum disorders: a multi-modal neuroimaging study

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Introduction: Bipolar Spectrum Disorders (BSD) have a prevalence of around 4%, and more than half of patients report an onset prior to 20 years of age. In pediatric populations the symptoms may differ, making the diagnosis challenging. Neuroimaging provides a non-invasive tool to explore brain neurotransmitters and brain function. The aim of this study is explore GABAergic neurotransmitter function as well as brain function at rest in youth with BSD.
Methods: A multi-modal neuroimaging study was conducted combining Proton Magnetic Resonance Spectroscopy (^{1}H MRS) of the Anterior Cingulate Cortex (ACC), quantifying brain metabolite levels including GABA, with resting state functional Magnetic Resonance Imaging (rs-fMRI), investigating brain connectivity patterns. Children and adolescents with BSD, who were exhibiting manic symptoms (n = 16, average age = 12.12 ± 2.47 years) and typically developing children (TDC; n = 33, average age= 12.00 ± 2.56 years), were recruited.
Results: Anterior Cingulate Cortex GABA values were lower in BSD youths compared with TDC (0.075 ± 0.012 (N = 11) vs. 0.083 ± 0.013 (N = 11)), although this difference was not statistically significant. GABA and Glutamate levels correlated negatively with the Young Mania Rating Scale (YMRS) in the BDS population (GABA, r = –0.52, p <0.15, N = 11) (Glu, r = –0.54, p <0.05, N = 16). Connectivity analysis based on rs-fMRI acquisitions revealed stronger connections between the ACC and the Left Superior Frontal Gyrus in BSD population compared to TDC (N_{BSD} = 16; N_{TDC} = 18).
Conclusion: Our findings show reduced GABA levels in the ACC as well as a relationship between manic state and GABA and Glu, signaling a possible dysfunction in the GABAergic and glutamatergic systems in this population, related to mood state. In addition stronger connections between the ACC and the LSFG (Brodmann Area 10) for the BSD population; could indicate a possible dysfunction in the executive and attention circuits of BSD patients, resulting in cognitive impairment.
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Clarification and goal-attainment in child welfare and juvenile justice institutions: Results of the MAZ.-study and introduction to EQUALS

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Background: The juvenile criminal law in our country was reformed in 2007. From then, youth welfare and the juvenile justice institutions are required to clarify the mental health status and the personal situation of the inmates at admission and perform a standardized yearly evaluation of the progress. The purpose of our study "MAZ." was the development of assessment tools to comply with these requirements and to describe the psychopathology of children, adolescents and young adults in care.
Methods: The study aggregates epidemiologic descriptions and a pre-post analysis. Several computer-based screening questionnaires and clinical interviews were administered in a sample of 592

adolescents (32% female and 68% male) aged 6 to 26 years (M = 16.1; SD = 3.07) from 64 child welfare and juvenile justice institutions in the German, French and Italian speaking part of Switzerland. Data was collected from August 2007 to February 2011.
Results: Findings revealed that the vast majority of adolescents have been exposed to multiple risk factors such as adverse family background, different forms of trauma, history of delinquency and other risk factors. 76% of the sample was rated as clinically impaired in the CBCL. 44% had comorbid diagnoses.
Conclusion: The findings on multiple risk factors as well as psychiatric and social impairment confirm the theoretical assumptions that adolescents in residential care represent a highly vulnerable population. For the intake of educational measure it is important to find diagnostic criteria to identify early indications in order to offer availability for adequate psychiatric treatment but also to prevent placement breakdowns. The challenge is to raise the standard of the cooperation between the youth welfare institutions and the Child and Adolescent Psychiatry/Psychotherapy.
Consequently, 25 Swiss and German child welfare and juvenile justice institutions entered into further cooperation with the Department of Child and Adolescent Psychiatry/Psychotherapy University Basel and will thus continue to apply the computerized assessment tools, continue data collection and further training of their key workers: www.equals.ch.

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Traumapädagogik – sichere Orte für betreute Heranwachsende und Mitarbeitende schaffen

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Vorstellung des vom Bundesamts für Justiz geförderten Projekts: Modellversuch Traumapädagogik – ein Projekt in stationären, sozialpädagogischen Institutionen der Deutschschweiz.
Mehrere Studien zeigen, dass Traumatisierungen bei fremdplatzierten Kindern eher die Regel als die Ausnahme sind. Interpersonell traumatisierte Kinder und Jugendliche konfrontieren sozialpädagogische Fachkräfte mit maladaptiven Beziehungsangeboten, weshalb diese die Kinder und Jugendhilfe vor besondere Herausforderungen stellen.
Traumapädagogische Konzepte orientieren sich an den Erkenntnissen der Psychotraumatologie und versuchen, spezifische nicht erlernte Fertigkeiten und Resilienzfaktoren gezielt zu fördern. Der zentralste Aspekt der Traumapädagogik ist, dass die Organisationsstrukturen und die Unterstützung der Mitarbeitenden ein zentraler Bestandteil des Konzeptes sind.
Im Rahmen des Modellversuchs werden im Zeitraum von 2012 bis 2015 die Mitarbeitenden und Leitungskräfte von fünf stationären, sozialpädagogischen Institutionen separat in traumapädagogischen Konzepten geschult und beim Implementierungsprozess für ihre institutionsinternen Projektziele begleitet. Der Verlauf der Massnahme wird mit qualitativen und quantitativen Methoden über vier Jahre hinweg in den fünf Modellinstitutionen und neun Kontrollinstitutionen untersucht.

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Cancer in children and adolescents: the combination of psycho-oncology and psychotraumatology in preventing trauma-related sequelae

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Childhood malignancy with frequent hospital stays, invasive treatment procedures and long-term separation from peers as well as age appropriate activities present a continuous burden for the entire family system. These are among many other high risk factors for the acute and long term development of a trauma-related sequelae in the child or the parents.
Methods: Psycho oncology according to the AWMF guideline 025/002; interventions in the field of psychotraumatology, focusing on resilience. Pediatric psycho-oncology takes place within a comprehensive medical treatment setting (diagnosis, treatment, remission, relapse, palliative care). Especially when receiving the diagnosis and in cases of relapse, the patients are highly susceptible to potential stress reactions. Well functioning interdisciplinary cooperation is mandatory for efficacious interventions. The main focus of the therapy lies on cumulative stress factors as well as interactions of potentially traumatizing factors within the family system. The life threatening disease causes profound fear and anxiety in parents, often leading to a destabilization within the family system. This in turn enhances the fears and anxiety of the sick child or adolescent. Even though the appropriate intervention is age-dependant, it always takes the entire family system into account. In certain cases, parents might require more support than their sick child. This is particularly true for younger children. Siblings are often neglected or feel like they're being neglected.
Clinical experience has shown that stabilization, the activation of resources and strengthening of resilience are focal points the interventions. Acute interventions and specific trauma-therapeutic methods are important parts of the psycho-oncologic treatment concept. Quality of life studies clearly show a positive influence on treatment outcome if quality of life is improved.

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Triple-R-Therapie als Therapiemodul in der stationären Psychotherapie von Adoleszenten – Klinischer Einsatz von «Rhythm-Rhyme-Recording»-Therapy

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Einleitung und Fragestellung: Jugendliche leben in entwicklungsgebundenen Subkulturen, die ihren Ausdruck mitunter in spezifischer Musikalität finden. Jede Subkultur hat ihren eigenen «Sound». Dieser Sound ist emotional hoch besetzt und dient der Identifikation und Abgrenzung des Jugendlichen. Er ist also im weitesten Sinne persönlichkeitsbildend. Wie lässt sich dieses für den Jugendlichen hoch besetzte Feld therapeutisch nutzbar machen? Wie lässt sich der persönliche Musikstil eines Jugendlichen als therapeutischer Zugang in einem ressourcenorientierten tiefenpsychologisch fundierten Setting verwenden?
Material und Methoden: Gearbeitet wird im Einzelsetting in einem akustisch eigens dafür optimierten Musikraum (Tonstudio) mit Digitaler Audio Workstation (Aufnahmecomputer). Der Jugendliche gestaltet in einem ersten Schritt («rhythm») seinen «Beat» bzw. sein individuelles Rhythmuspattern, allenfalls ergänzt mit Melodien oder Harmonien mittels virtueller oder nichtvirtueller Instrumente. In einem zweiten Schritt («rhyme») rappt oder singt er einen assoziativ entstehenden und iterativ überarbeiteten selbst verfassten Text dazu. Der Therapeut handelt in der Rolle eines Musikproduzenten und leitet den schöpferischen Prozess des Jugendlichen («recording»). Wo nötig ergänzt er mittels Editieren (digitale Nachbearbeitung) und erstellt zuletzt einen Mix des Musikstücks.In einem klinischen Pilotprojekt wurde mit 11 adoleszenten Jungen im Alter von 15 bis 17 Jahren gearbeitet.
Erste Ergebnisse: Bisher konnten mit einer ersten Gruppe Jugendlicher positive Erfahrungen gesammelt werden. Die Jugendlichen haben hochmotiviert ihre Songs aufgenommen und waren dabei auf implizite und später explizite therapeutische Themen nachhaltig gut ansprechbar. Die Speicherung des «Eigenen», die Modifizierung im sprachlichen wie im musikalischen Teil und die persönliche Zueignung (Schweigepflicht, kein geplanter Auftritt, keine soziale Verbreitung des Materials) waren wichtige Faktoren des Erfolges.
Diskussion: Über das Konzept einer aktiven Musiktherapie hinaus wird mit der Aufnahme der Musik («recording») des Jugendlichen gearbeitet. Das Aufnehmen und das folgende Editieren ermöglichen es dem Jugendlichen, auf technischem Wege der musikalischen Gestalt seines Werkes näher zu kommen und sich damit besser identifizieren zu können. Die resultierenden Aufnahmen werden im psychodynamischen Sinne als Produktion verstanden und für den therapeutischen Prozess nutzbar gemacht.

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The Marte Meo Method helps to improve interactions and social-/emotional development

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Does the Marte Meo Method (MMM) improve parental skills in challenging situations such as premature-, handicapped-, emotionally-stressed children? First results of an evaluation after a three-week intervention "training with MMM" at the German parent-child rehabilitation Fachklinik Bromerhof are shown. The study focus is interaction improvement parent-child with a sample of approximately 50 and a control group of about 200 families. An international working group of doctors, nurses and therapists (Austria, Croatia, Germany, Nordic countries, Switzerland) use MMM in different medical fields to improve interaction-moments between professionals, patients and relatives and thus to enable social- and emotional child development. MMM is a video supported mikro-communicational method developed in the seventies in the Netherlands by Maria Aarts. German and Swiss professionals of this international medical group present with films the method and case studies of premature born, emotionally distressed and handicapped children/juveniles.

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Mother- and father-infant-relationships in infants referred to a specialised interdisciplinary service for infants and toddlers

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Background: Infant mental health problems include difficulties to regulate emotions or attention, crying, sleeping or feeding issues. Developmental trajectories of these problems are complex, arising out of multiple interacting and co-evolving factors. One important factor may be parental mental health and parent-infant-interactions.
Method: Infants between 1 and 36 months of age (N = 280) who were referred to an specialised interdisciplinary service for infants and toddlers because of excessive crying, sleeping or feeding problems and their parents were examined and compared with an age-matched community-based control sample with no current problems. Four groups, according to DC: 0-3R, Regulation Disorders of Sensory Processing, Sleep Behaviour Disorders, Feeding Behaviour Disorders, and controls, were compared with regard to the quality of mother- and father-infant relationship (PIR-GAS), and mothers' and fathers' perception of her own psychological state (EPDS and SCL-90).
Results: In comparison with the general-community sample, mother- as well as father-infant relationships were more often distressed or disturbed among referred triads than among controls. Moreover, mothers as well as fathers in all three referred groups scored similarly high on depression, anxiety, and hostility.
Conclusion: The findings suggest that behavioural problems in infancy have a negative impact on parent-infant-relationships, which may function as a maintaining or impairing factor. Dynamic interactions between parental resources and crying, sleeping or feeding disorders compromise intuitive parenting and put families at long-term risk for a disturbed parent-child-relationship. Therefore treatment must be relationship-based, concentrating somewhere between the 'infant with a problem' and the caregivers as a main influencing factor on the behavioural problem of the infant.

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Postpartum depression: infants' irritability is associated with mothers' parenting stress and poor postpartum bonding

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Objectives: Postpartum Depression (PPD) implies an impairment of maternal adaptation. Moreover, there is increasing evidence that children of mothers with PPD are at risk for developmental abnormalities, including emotional/behavioural difficulties in later life. The aim of this study was to investigate in the association between maternal psychopathology, mother-infant-relationship and infant irritability three months postpartum. We expected that mothers with PPD would report more parenting stress and more difficulties in postpartum bonding, and that these difficulties would be associated with more infant irritability.
Methods: In a prospective study a total of 127 mothers (mean age: 33.3 years; 43% primipara) completed a series of questionnaires related to postpartum depression, sleep quality, parenting stress, maternal sleep quality, postpartum bonding and infant irritability. Of the 127 participants, 21 reported to suffer from PPD.
Results: Compared to mothers without PPD, mothers with PPD reported increased parenting stress, delayed postpartum bonding, and higher bonding anxiety. They were more likely to consider their infants as stressors, and they reported more infant irritability. Poor maternal sleep was associated with increased parenting stress and with impaired bonding.
Conclusion: The pattern of results suggests that mothers suffering from PPD are at increased risk to report poor sleep, high parenting stress, and difficulties in postpartum bonding, as compared to mothers without PPD. Mothers' behaviour and infants' irritability are associated. We claim that treatment of PPD should include support of parental competences, nocturnal sleeping behaviour and mother-infant-bonding.

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The heart and soul of change: importance of common factors in a group therapy for parents with mental disorders

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Introduction: Parent psychopathology is strongly associated with offspring psychiatric disorders, a risk transmitted through both genetic and environmental mechanisms (McLaughlin et al., 2012). Multimodal interventions attempt to minimize the developmental risk for these children and their families.
Particularly, interventions in parents with mental disorders appear to be effective in preventing mental disorders and psychological symptoms in their offspring (Siegenthaler et al., 2012).
Beside measuring effectiveness, common factors play an important role in psychotherapeutical processes (Lampropoulos, 2000).
Methods: In this contribution we present an evaluation of a manualized grouptherapy for parents with mental disorders (Kupferschmid & Koch, 2014), taking place in groups of four to five parents in the University Hospital of Child and Adolescent Psychiatry, Bern.
This grouptherapy comprises six modules concerning different topics of parenthood with a focus on communication with the child and coping with mental illness as a familial system.
Subsequent to the therapy, patients (N = 16) completed a self-provided questionnaire including a graduated Likert scale concerning variables of the intervention.
Results: The items of the questionnaire were pooled to questions concerning "psychoeducation and support" and "therapeutic relationship", known as two common factors of crucial importance for therapeutic processes. High levels of satisfaction have been ascertained for both categories.
Conclusions: Our findings suggest that in this grouptherapy different common factors are balanced well. Further research should combine outcome and procedural measures as well as transgenerational assessment in order to provide information about the ways of therapeutic change in parents with mental disorders and their children.

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Mentally ill children – mentally ill parents: an integrative treatment approach

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Background: Longitudinal studies indicate that up to 50% of all preschool children with persistently irritable or angry mood, temper tantrums and/or verbal or physical aggression which are not adequate to the situation or the child's developmental stage are at high risk for a negative life-time persistent psychopathology. In order to prevent the development of external or internal behaviour disorders (e.g. conduct disorder, depression or anxiety disorders) early intervention approaches are strongly recommended, particularly when biological as well as psychosocial risk factors are both present at the same time. The Clinic of Child and Adolescent Psychiatry of the University of Basel is one of the first units in Switzerland offering a day hospital for preschoolers with mental health problems.
Method: The diagnostic and therapeutic concept (individual child therapy, child-parent video-therapy, home-treatment) will be presented and data on the interventions' effectiveness will be discussed.
Results and conclusion: Clinical experience and first data confirm the usefulness of a day clinic approach in preschool children with mental health problems and underline the necessity to realize multimodal integrative intervention strategies between the interface of child and adult psychiatry, paediatrics and educational systems in order to identify and treat children at risk as early as possible.

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Incontinence: a prospective survey over 11 Years at the Children's Hospital of Luzern

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Aim: Evaluation of 773 children with urinary incontinence from January 2002 to December 2012 in our clinic regarding voiding symptoms, frequency, volumes and residual volume, defecation frequency, urinary tract infections, kidney malformation and neurological status.
Method: All children with urinary incontinence assigned to our clinic were registered propectively with parental consent.
A general examination, peripheral neurological status and genital inspection, as well as a ultrasound of the kidneys, uroflowmetry with residual urine were carried out in all patients. Therapy and success according to the criteria of the ICCS as well as age and sex of the patients were documented.
When required further investigations like MRI, szintigraphy, vesico-cysto-urography, cystoscopy, urodynamics were carried out. Clinical follow up examinations were done in all patients.
Results: 773 patients were seen. 52% boys, 48% girls. 57% were 6–10 years old. 36% suffered from detrusor overactivity. 16% postponed micturation and 15% showed a detrusor-sphincter discoordination.
An organic lesion was diagnosed in 4.2%. 3% showed a not dilatating reflux. UTI appeared in 24%. An obstipation had to be co-treated in 14%.
Therapy consisted in toilet training, if necessary antimuscarinic drugs, neuromodulation or bio feedback therapy. Therapy was effective in 93%.
Conclusion: Urinary incontinence in school aged children is stigmatazing and can be associated with significant morbidity. A good clinical examination without invasive investigations permits a correct diagnosis. Different therapy options adapted individually provide a success rate over 90%.

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Healthcare-associated community-presenting sepsis is different from community-acquired sepsis in pattern of treatment using carbapenems and glycopeptides

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Background: The epidemiology of healthcare-associated community-presenting (HCA) bacterial sepsis differs from community-acquired (CA), but may correspond to that of hospital-acquired (HA) sepsis. We explored whether differences in pathogen spectrum of CA, HCA and HA sepsis were anticipated by clinicians when choosing treatment with glycopeptides and carbapenems in a Swiss cohort.
Methods: 322 episodes of pediatric (age >1 month–17 years) blood culture (BC) proven sepsis included in a prospective Swiss national pediatric sepsis cohort from September 2011 to November 2013 were analysed. Episodes were classified as CA when BC was obtained ≤48 hours from hospitalisation, as HCA in children with specific risk factors (primary or secondary immunodeficiency, chronic underlying disease, previous maior surgery, central venous catheter in situ) and as HA when BC was obtained >48 hours after hospitalisation.
Results: Information on isolated pathogens and antibiotic treatment was available for 312/322 episodes. Pathogen distribution and treatment strategies are shown in table 1.

Pathogen	CA (n = 149)	HCA (n = 83)	HA (n = 80)
Gram positive	96 (64%)	43 (52%)	39 (49%)
S. aureus	27 (18%)	9 (11%)	22 (27%)
Staphylococci, coagulase neg.	0	9 (11%)	7 (9%)
Gram negative	54 (36%)	39 (47%)	34 (44%)
E. coli	27 (18%)	19 (23%)	9 (12%)
K. pneumoniae or P. aeruginosa	2 (1%)	10(12%)	16 (20%)
Fungi	0	1 (1%)	6 (7%)

Empiric and defintive exposures to carbapenems and glycopeptides are shown in table 2. The differences between CA, HCA and HA empiric and definitive treatment were statistically significant with p <0.01 for comparison between all groups (Fisher's exact test).

Empiric Treatment	CA	HCA	HA
Carbapenems	1.3%	21.7%	30%
Glycopeptides	0.7%	21.7%	41.3%
Both	0	7.2%	16.3%
Definitive Treatment			
Carbapenems	0.7%	19.3%	30.0%
Glycopeptides	0.7%	13.3%	18.8%
Both	0%	1.2%	8.8%

Conclusion: Treatment with carbapenems, glycopeptides, or both in definitive therapy was highest for HA, intermediate for HCA and lowest for CA sepsis, and is partially explained by variations in pathogen spectrum and probably additional variations in susceptibilities. This trend was also observed for empiric treatment. Swiss clinicians identify HCA sepsis as different from CA in terms of empiric treatment decisions.

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Unplanned intensive care unit admissions from the ward

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Introduction: Children with an unplanned ICU (intensive care unit) admission from the ward are known as a particular vulnerable patient group. The aim of this work is to characterise this group of children and to compare it with the rest of ICU admissions regarding demographic, anamnestic, and outcome parameters. Further we want to discuss a possible need for critical care outreach and early warning tools.
Methods: The study was set in an interdisciplinary 18 bed ICU at the University Children's Hospital Zurich. There are close to 7000 annual admissions to hospital and around 1200 admissions to the ICU. Children with an unplanned admission to our ICU from a ward within the hospital were compared with the rest of ICU admissions. Data for years 2009 and 2010 was prospectively collected and analysed using Mann Whitney U test and Chi square test.
Results: Out of 2363 admissions to the ICU 147 (6.2%) were unplanned from the ward. This group had a higher predicted (median: 1.58% versus 1.40%) and observed mortality (6.1% versus 2.3%). Most frequent reason for admission in this group was respiratory problems (27.9%). Even though mechanical ventilation in the first hour was less often necessary in the unplanned group (10.2% versus 44.9%), the median duration of ventilation was distinctly longer (2.31 days versus 0.94 days), just as the median length of ICU stay (2.38 days versus 1.71 days). Further, the unplanned admitted patients had to be quite often readmitted to the ICU after discharge (12.9% versus 1.0%). Three out of 147 unplanned admitted patients had a cardiopulmonary arrest on the ward.
Conclusions: Children who are unplanned admitted to ICU from the ward have an increased risk of dying, prolonged ICU stay and high readmission rate. Our findings should further sensitize for this high risk group, but the low rate of cardiopulmonary arrest and the low number of ventilated children in our unplanned from ward group imply that most deteriorating children were identified and discharged timely. The prolonged duration of ventilation and length of stay further indicate that these children mostly were severely ill in progress. Accordingly implementation of medical emergency or rapid response teams and paediatric early warning scores might not improve outcome in our circumstances.

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Impaired executive functions in complex tasks in children and adolescents born very preterm

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Introduction: Many studies have found impaired performance in executive functions (EF) in patients formerly born very preterm (VPT) [1]. Most evidence derives from studies in early and middle childhood. EF play a pivotal role for academic achievement and personal autonomy. Demands in both domains and, in parallel, the dependence on EF ability continuously increase in later childhood and adolescence [2]. This study therefore aims to investigate the pattern of EF deficits in older children and adolescents born very preterm and to compare them with healthy term-born peers.

Methods: Forty-one VPT children and adolescents (age: M = 12.9 [SD = 1.7, range = 10.4–16.6] years) with normal general cognitive abilities and 38 healthy term-born (TB) peers (age: M = 12.7 [SD = 1.8, range = 10.0–16.9] years) were examined. Various EF components (i.e., planning, inhibition, working memory, cognitive flexibility) were assessed with a computer-based test battery (Cambridge Neuropsychological Testing Automated Battery, CANTAB). Additionally, parents reported on their children's EF ability in the school and home environment using the Behavior Rating Inventory of Executive Functions, BRIEF (German version).
Results: Mixed-model ANOVAs with birth status (VPT vs. TB) as between-subjects factor and difficulty levels of EF tasks as within-subject factor revealed a significant interaction between birth status and difficulty level for planning ability ($F(3, 231) = 3.120$, $p = .03$), working memory, ($F(2, 154) = 2.307$, $p = .10$) and inhibition ($F(3, 216) = 3.946$, $p = .03$): Performance of VPT and TB participants was comparable in lower difficulty levels but poorer for VPT participants in higher difficulty levels. Parents of VPT participants rated EF abilities of their children poorer than parents of TB participants (e.g., global EF ability: $t(78) = 3.002$, $p = 0.004$) with more VPT participant's scores lying above the cut-off for clinically relevant EF problems ($\chi^2(1) = 7.671$, $p = .006$).
Conclusions: Executive function deficits persist into adolescence in VPT patients with normal general cognitive ability. Particularly, when tasks become more complex, VPT participants experience more EF difficulties than their peers. As EF demands become increasingly more complex in later childhood and adolescence, EF deficits may hinder optimal development in former VPT patients.

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High-concentration nitrous oxide 70% is a safe agent for procedural sedation in children

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Introduction: Nitrous oxide 70% (N20 70%) is an attractive medication for analgosedation in a pediatric emergency department (ED). It can be used for short painful procedures because of its rapid onset and offset. The purpose of this study was to provide information about the safety of N20 70% in combination with other analgetics and its adverse events.
Method: An observational study was performed at our pediatric ED to analyse the safety of N20 70% combined with 30% 02 application for short painful interventions. 341 patients were included. No fasting times were required. We recorded all administered drugs, the duration of N20 70% application, the patient's behaviour during the intervention and immediate adverse events. A few days later we called all families and asked about possible side effects.
Results: 341 patients were included, mostly for fracture reduction of the forearm or fingers. No severe adverse events were noted. The most common side effect was vomiting (7.5%). In 86% the patients had no complaints at all and the satisfaction rate was 98.5%.
Conclusion: High-concentration nitrous oxide 70% is a safe agent for procedural sedation in children. Only minor adverse events like vomiting were infrequently noticed.

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Prospective study on plasma pro-endothelin-1 (CT-proET-1) in predicting bronchopulmonary dysplasia

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Background: Very preterm infants are at high risk for developing bronchopulmonary dysplasia (BPD). Today, BPD is defined as the need for supplemental oxygen for at least 28 days of life (DOL), and its severity is graded according to the respiratory support required at 36 weeks gestational age (GA). Previously we found high plasma CT-proET-1 levels in newborn infants with respiratory distress when measured at DOL3. The objective of this study was to examine CT-proET-1 levels in the late course of BPD evolution.
Methods: Plasma levels of CT-proET-1 were prospectively measured at DOL28 and at 36 weeks GA in 110 very preterm infants born before 32 weeks GA. Non-parametric descriptive statistics were used. The study is registered at ClinicalTrials.gov: NCT01644981.
Results: CT-proET-1 levels at DOL28 were significantly higher than at 36 weeks GA (median 184 pmol/L, interquartile range (IQR) 149-233 vs. 150, 118-188, $p < 0.01$) and both values were related ($Rs = 0.274$; $p < 0.05$). Infants with BPD (all grades, $n = 51$) had elevated CT-proET-1 levels at DOL28 compared to infants without BPD (median 210 pmol/L, IQR 158-301 vs. 172, 143-259, $p < 0.01$), whereas CT-proET-1 at 36 weeks GA did not differ.
Conclusions: CT-proET-1 levels decreased during postnatal development. While CT-proET-1 measured at DOL28 indicated BPD, differences disappeared until 36 weeks GA. In summary, CT-proET-1 may be used as an early indicator for BPD.

Gong Show

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Giant cervical lymphangioma – ENTRY by EXIT

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Introduction: Lymphangiomas are benign congenital tumors, involving the head and the neck, 90% occuring in children less than 2 years of age.
EXIT procedure (ex-utero intrapartum treatment) has been developed to reverse temporary tracheal occlusion in children who had undergone fetal surgery for severe congenital diaphragmatic hernia. Its use was expanded for deliveries where difficulty in securing the airway was expected due to large neck masses, thoracic or cardiac lesions or congenital high airway obstruction.
Case report: A prenatal ultrasound at 22 weeks of gestation revealed a cystic neck mass. EXIT procedure was planned at 37 5/7 weeks in a multidisciplinary setting as this mass increased in size and the developing polyhydramnios suggested an impairment of fetal swallowing. This mass was confirmed a huge lymphangioma postnatally. Primary treatment was done with the sclerosing agent OK 432 (Picibanil®), being injected into the lymphatic cysts at the age of 8 and 23 days. Due to lack of decrease in mass size, a debulking procedure and tracheostomy was performed 4 weeks after the second OK 432-application. After further treatment with sildenafil (age 6 to 12 months) and propranolol (age 22 to 26 months) a residual macroglossia persists. A reduction of the mass of the tongue is planned.
Conclusion: The EXIT procedure is a safe method that allows time to secure the upper airways during delivery in patients with a large neck mass that could lead to life-threatening upper airway obstruction. In the multidisciplinary care of extensive cervical lymphangiomas there is a broad spectrum of therapy methods to consider.

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Cowper's syringocele: case presentations

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Introduction: Cowper' syringocele is an uncommon cystic dilatation of the paired ducts of the bulbourethral glands of Cowper. Distension of the ducts and the glands may cause urethral compression or may result in an obstructive membrane. Syringoceles may cause symptoms of infravesical obstruction similar to posterior urethral valves (PUV). Evaluation should be done with ultrasound and voiding cystourethrography (VCUG).
Patients and methods: Case 1:7 days old newborn (*2008), prenatal diagnosis of bilateral hydroureteronephrosis, VCUG and perineal ultrasound suspicious for syringocele. In addition: trabeculated bladder, urinoma left kidney, vesicoureteral reflux (VUR) grade III left, elevated serum creatinine.Transurethral endoscopic unroofing was performed.

Case 2: 2 months old infant (*2013), first pyelonephritis at age of 1 month, VCUG showed an infravesical obstruction, suspicious for posterior urethral valves and no VUR. Cystoscopy revealed a Cowper's syringocele. Unroofing was done transurethrally.
Results: Case 1: Due to persistent obstruction in the 3 month follow-up VCUG an open resection of the syringocele and urethral reconstruction was performed. Follow-up of 5 years: decreasing of bilateral hydronephrosis, bilateral kidney growth, 2 urinary tract infections (UTI) after 2 years, bladder diverticula, normal serum creatinine, urinary continence.
Case 2: Short term follow-up was uneventful, 3 months follow-up with VCUG and urethrocystoscopy are planned.
Conclusion: Although syringoceles are uncommon, they are a relevant differential diagnosis in newborns with infravesical obstruction. Paediatric urologists must be prepared to recognize them during urethroscopy for suspected posterior valves, even if preoperative VCUG may not be suggestive for syringoceles. Initial therapy is urethroscopic unroofing, followed by open resection and urethral reconstruction if needed.

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It looks like ambiguous genitalia - hypertrophy of clitorial hood as presenting sign of neurofibromatosis 1

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Introduction: Ambiguous genitalia at birth need immediate evaluation by a team of specialists. A newborn girl was presented for evaluation of an enlargement of clitoris and labia maiora. Pregnancy and spontaneous birth were unconspicuous, as was medical history of the family.
Diagnostic results: Besides a 1.5 cm swelling of clitoral hood without involvement of the clitoris, there were no signs of virilisation, no hyperpigmentation, no synechia of the labia minora and meatus urethrae and anogenital distance were normal. Serum androgens on 1st and 4th day of life were in the normal range of female newborns, as were adrenal steroid precursors (17-OH-progesteron and androstendion). The ultrasound examination presented a normal uterus and ovaries and a normal intraabdominal state without tumor. The follow-up for 10 months showed a normalization of labia maiora, but no significant reduction of the swelling of the preputial skin, in the presence of normal growth parameter and neurologic examination. Then, other case series[1] reporting similar hypertrophy of clitorial hood as a sign of neurofibromatosis (NF1) prompted us to search actively for dermatologic signs of NF1. Six café au lait spots and the beginning of an inguinal freckling were found in the patient as well as many more spots in the father. The same skin manifestation was described in his mother.
Conclusion: In cases with isolated hypertrophy of the preputial skin, not the surgical intervention, but the extensive search for the aetiology of assumed genital ambiguity was essential and led to early diagnosis of another disease.

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An abdominal twist of fate – a case of cecal volvulus in a 9-year-old boy

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Introduction: Cecal volvulus (CV) is extremely rare in children and only approximately 40 cases are reported in the English literature. Herein we present a case of CV in a 9-year-old boy and review the literature regarding clinical presentation, treatment options and outcome after CV in children.
Case description: A 9-year-old boy consulted our emergency department for acute abdominal pain for 2 hours. He showed marked abdominal distension with tenderness and a palpable mass in the right lower abdominal quadrant. Plain abdominal X-ray revealed an enormous dilatation of a colonic segment, suggesting an acute mechanical obstruction. An emergency exploratory laparotomy was performed. The intraoperative situs confirmed CV with a massively dilated, non-ischemic dolicho-cecum, without intestinal malrotation. After derotation and decompression, persistence of a huge mobile cecum justified resection of the proximal dilated colon. The postoperative course was uneventful.
Conclusion: CV can be a life-threatening condition due to progression towards colonic ischemia and perforation. Therefore early diagnosis and treatment are crucial and influence the outcome. The preferred operative treatment is primary resection and end-to-end anastomosis. Simple detorsion with or without cecopexy is an option, but associated with the risk of recurrence.

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Duodenal web and Down syndrome – a case report

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Introduction: The association between duodenal atresia and Down syndrome is well known. However, there are patients with extraordinary presentations.
Methods: We present the case of a girl with Down syndrome and a duodenal web in pars-3, that showed uncharacteristical radiographic and anatomic findings.
Case report: A newborn girl with delivery at term shows typical signs of Down syndrome, otherwise healthy, presented with repeated bilious vomiting during the first days of life. Radiogaphic studies with and without contrast were inconclusive, as well as an esophagogastrodoudenoscopy. Ultrasonographic findings were suspicious for a duodenal web in pars-3. During surgery no duodenal dilatation was apparent, just a tiny notch in pars-3 proximal to the ligament of Treitz was visible. A longitudinal incision showed a dense membrane with an almost centered "target-like" opening. After performing a partial resection of the membrane and a side-to-side duodenojejunostomy we observed an uneventful follow-up.
Conclusion: Doudenal web in pars-3 is a rare subgroup of the duodenal obstruction. Descriptions in literature are lacking. Clinical presentation resembles other intestinal obstructions, however radiographic diagnosis is difficult due to inconclusive findings. Definitive diagnosis is achieved by surgery.

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Acute pancreatitis in children: a report of three cases

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Objective: Studies suggest an increased incidence of acute pancreatitis over the last decades. There are few papers that review the incidence in pediatric patients. We would like to discuss three clinical cases from our hospital between September 2013 and January 2014.
Methods: The first patient was a 7 year old boy with an acute abdomen. An explorative laparotomy and the following MRI showed a hemorrhagic pancreatitis. The second patient was an 8 year old boy who developed acute pancreatitis after a blunt abdominal trauma. Patient number three was a 12 year old boy presenting with a new episode of biliary pancreatitis due to gallstones after cholecystectomy at the age of 6 years.
Results: Patient number one had a pancreatitis of unknown etiology until today. He is currently getting better with use of antibiotic therapy, temporary fasting and watchful waiting. The second patient had a pancreatic pseudocyst as a complication, which was decreasing size under antibiotic treatment and abstinence from food. He was discharged home after 3 weeks. Patient number three relapsed two weeks after conservative treatment and required an endoscopic retrograde cholangiopancreatography (ERCP) with papillotomy and antibiotic therapy, after which he was discharged home.
Conclusion: Clinical diagnosis of acute pancreatitis can be challenging due to the variety of symptoms. Depending on the etiology of the pancreatitis, conservative and interventional treatments are viable and effective options, surgery should be considered in selected cases. To assess whether the incidence in pediatric patients is increasing epidemiological studies would be required.

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Talar dome fracture with ankle sprain – a rare but potentially debilitating lesion not to be missed

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Introduction: Osteochondral lesions are infrequent in children, preferentially affecting the knee and elbow, and more rarely the talus. Trauma by inversion and dorsiflexion of the ankle is frequently seen in a pediatric emergency department, although very rarely associated with an osteochondral talar fracture.
Case report: We present a case of a 13 year old girl who fell while playing volleyball and suffered from a ankle sprain. On physical exam she had significant swelling of the right ankle with inability to bear weight on her right foot. AP and lateral radiographs of the ankle performed on 2 occasions were considered as normal and conservative treatment with a posterior splint was applied. Further review of the radiographs suggested a non displaced Salter I fracture of the distal fibula with loss of continuity of the talar dome and a lateral intra-articular fragment. Computed tomography (CT) confirmed the diagnosis of an inverted lateral osteochondral talar dome fracture. The

patient underwent prompt open reduction and internal fixation of the displaced talar fragment. Her postoperative course was uncomplicated and she is slowly regaining right ankle function.
Discussion: Lesions of the Talus account for 4% of all osteochondral defects. Patients often present with ankle swelling, chronic ankle pain, weakness, stiffness, and instability. When associated with ankle sprain, the diagnosis by plain radiographs can be challenging and CT is often required for better delineation of the fracture. Intra-articular osteochondral talar fractures following acute injury are most commonly treated surgically.
Conclusion: Following an ankle sprain, Talar dome osteo-chondral fractures are rare but potentially debilitating. Intra-articular fragments in the ankle should always be looked for and fixed surgically when found.

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The football and its kick-off

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Introduction: We would like to present a rare case of intestinal perforation in a neonate.
Material/Methods: A female neonate was born at 31 weeks of gestation and developed on day 3 of life an abdominal deterioration with bilious vomiting and football sign in babygramm. Due to suspected intestinal perforation, laparotomy was performed immediately.
Results: Surprinsingly, a perforated Meckel diverticulum (MD) was found, resected and histologically confirmed. To exclude intestinal obstruction as the triggering cause, Hirschsprung's disease was ruled out by histology. The child improved quickly and started 72h after surgery with oral nutrition.
Discussion: MD is a rare cause for intestinal perforation in neonates. Hirschsprung's disease is reported as predisposing factor. Because of its scarcity we would like to present this case.

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A long journey to a unique tumour

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Introduction: To define the benign or malignant character of a hepatic lesion is crucial before initiating any treatment. We herein present a case with a giant liver tumor, of particularly challenging diagnosis and management.
Clinical case: A giant intra-/retrohepatic abdominal mass was discovered incidentally in a 4 year-old boy with no past medical history. Open superficial biopsy was in keeping with mesenchymal hamartoma, although identification of adipose tissue was unusual. Since the histological and radiological aspects of the tumor (homogenous periphery, heterogenous center) were uncommon for mesenchymal hamartoma, deep needle biopsies were performed showing only mature-appearing adipose, and thin fibrovascular septae. Molecular techniques spoke against liposarcoma and angiomyolipoma; matured lipoblastoma was considered without definite diagnosis. Lipoblastoma asked for tumor enucleation, liposarcoma for absolute R0-resection and thus liver transplantation, since the tumor was intimately adherent to the portal and hepatic veins. To avoid over-treatment, a two-step approach was chosen and enucleation performed first, meant to be followed if indicated by liver transplantation. Laparotomy revealed a 1.45 kg encapsulated intra-/retrohepatic tumor of 19 x 14.5 x 7.5 cm, easily cleaved from the adjacent liver. Final histological diagnosis was lipoblastoma, supported by identification of a t(8;14) translocation involving the *PLAG1* gene. Long-term follow-up is mandatory to monitor any recurrence, as R0-resection was not feasible due to tumor's proximity to major vessels.
Conclusion: Lipoblastoma is a benign tumor of young children usually located in extremities, neck, trunk or omentum. This is the first description of a hepatic lipoblastoma. Biopsies assessing only the mature component of the tumor rendered preoperative diagnosis extremely challenging.

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Henoch-Schönlein purpura with penis involvement

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A 3-year-old male, diagnosed with Henoch-Schönlein purpura (HSP) 2 weeks earlier, presented with acute occurrence of painful swelling of the penis and urinary retention. A tender, fire-red tumefaction of the shaft, glans and prepuce of the penis was noted, associated with typical purpuric skin lesions to the lower extremities and buttocks and swelling of his ankle joints. Simple analgesia lead to spontaneous micturition. A 48-hour course of oral prednisone was initiated and the patient could be discharged home after 23 hours with a diagnosis of penile vasculitis associated with HSP. After 48 hours, the penile oedema and pain had completely resolved. Penile involvement with HSP, the commonest systemic vasculitis of childhood, is well described and must be distinguished from paraphimosis, which occurs when the foreskin is retracted behind the coronal sulcus with oedema caused by venous stasis. The use of steroids, usually reserved for abdominal involvement, remains controversial.

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The heartbreaking story of NEC and Botalli

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Introduction: Case presentation of a preterm baby born at 30 weeks of gestation, birth weight 1260 g. The newborn presented with persistent arterious duct of Botalli (PDA). Closure of the patent duct was tried with Ibuprofen unsuccesfully. Due to relevant hemodynamic problems, duct closure was achieved through open ligature on day 18. At day 17 the baby developed severe deterioration due to fulminant necrotising enterocolitis (NEC), requiering emergency laparotomy. We present the case of a preterm patient with patent duct and NEC and we discuss the current evidence in literature.
Aim is to discuss the risk of NEC in preterm patients with a patent hemodynamically relevant duct, and whether closure of the duct should be achieved surgically before the patients may develop a NEC.
Results: In the literature, the relation between NEC and patency oft he duct is questionable. The group of Wariki MN et al from Tokyo says that a patent duct is a risk factor for developing NEC. On the opposite, the groups of Yee WH et al from Scotland and Sulemanji MN et al from Boston suggest that primary closure oft he duct cannot be recommended to avoid NEC.
Conclusion: In our case there seems to be a strong correlation between patency of the duct and NEC. Therefore a critical awareness for potential NEC in patients with PDA should be anticipated.

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Nocturnal hypoglycaemia in diabetic children: the role of continuous glucose monitoring (cgms)

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Background: Hypoglycaemia is the most common acute complication of diabetes mellitus type 1. Nocturnal hypoglycaemia is mostly unrecognised and asymptomatic, but recurrent mild hypoglycaemia at night can lead to hypoglycaemia unawareness and reduced performance during the day.
Aim: To evaluate frequency and duration of nocturnal hypoglycaemia in type 1 diabetic children by continuous glucose monitoring (cgms) for 6 days.
Patients/methods: 59 children with type 1 diabetes for >6 months were included. The glucose record of 50 patients (29 male, 21 female, mean age 12.1 y, range 2.4–17.6 y) was complete and comprehended at least 5 nights. Patients were asked to perform 4 capillary blood glucose measurements per day and to document sleeping time, carbohydrate intake, insulin administered and symptoms of hypoglycaemia. Nocturnal hypoglycaemia was defined as any glucose level <3.7 mmol/l during nighttime (period between bed time and wake up time).
Results: 119 nocturnal hypoglycaemic events were found, only 6 of them being symptomatic. In 97 out of 292 nights hypoglycaemia occurred once or more (33% per night). No nocturnal hypoglycaemia was found in 7 patients (14%), one episode in 13 (26%), 2 episodes in 9 (18%), 3 episodes in 8 (16%) and >3 episodes in 12 (26%). Duration of a hypoglycaemic episode ranged from 5 min to 665 min, 35% of the episodes lasted <1h, 33% 1–3h, 28% 3–6h and 4% >6h.
Conclusion: Nocturnal hypoglycaemia is a relevant complication in diabetic children: it is frequent, mostly asymptomatic and often prolonged. Cgms constitutes a useful tool in detecting nocturnal hypoglycemia and therefore optimising patient instruction and treatment.

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Ambulatory arterial stiffness index in obese children

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Introduction: Altered arterial stiffness is a recognized risk factor of poor cardiovascular health. Ambulatory Arterial Stiffness Index (AASI, defined as one minus the regression slope of diastolic on systolic blood pressure values derived from a 24 h arterial blood pressure monitoring, ABPM) is an upcoming and readily available marker of arterial stiffness. We tested the hypothesis that AASI is increased in obese children compared to healthy subjects.
Methods: AASI was calculated from ABPM in 101 obese children, 45% girls (BMI-SDS median 2.9 (interquartile range (IQR) 2.4–3.8), median age 11.5 years (IQR 9.0-13.4) and compared with an age- and gender matched healthy control group of 71 subjects (49% girls) with BMI SDS median 0.0 (IQR –0.75–0.5), median age 12 years (IQR 10.0-14.0). Multivariate regression analysis was applied to identify significant independent factors explaining AASI variability in this population.
Results: AASI was significantly higher in obese children compared to the controls (0.388 (IQR 0.253–0.499) versus 0.190 (0.070–0.320), p <0.0001), whereas blood pressure values were similar (p = 0.18). In a multivariate analysis including obese children only, AASI was independently predicted by BMI and daytime systolic blood pressure (p = 0.04); and in a multivariate analysis including obese children and controls BMI and pulse pressure independently influenced AASI (p <0.001).
Conclusions: This study demonstrated increased AASI, a surrogate marker of arterial stiffness, in obese children. AASI seems to be influenced by BMI independently to blood pressure values, suggesting that other factors are involved in increased arterial stiffness in obese children.

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Paediatric rheumatology in Switzerland: data from the Swiss Pediatric Rheumatology Registry

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Background: Musculoskeletal pain is frequently reported in childhood. Among this population, the frequency of inflammatory diseases and chronic musculoskeletal pain is probably underestimated. To investigate the epidemiology of rheumatic diseases in childhood in Switzerland and evaluate the need in structures for adequate care for these patients with chronic diseases and potentially poor long-term outcome, we created the Swiss Paediatric Rheumatology Registry.
Objectives: To evaluate the prevalence and incidence of inflammatory and non-inflammatory rheumatologic conditions in the pediatric population of Switzerland, and describe their characteristics.
Methods: All children seen between 2004 and 2012 in the 10 main paediatric rheumatology clinics in Switzerland have been included. Data collected comprised diagnosis, treatment and demographic data.
Results: 4631 patients were included: mean age 7.78 years at presentation and 8.80 years at diagnosis, male/female sex ratio 1/1.4. 2972 patients (64.2%) had an inflammatory disease with an annual incidence of 23.5 for 100'000 paediatric patients; among them 1598 (53.8%) had juvenile idiopathic arthritis, 180 (6.1%) connective tissue diseases, 109 (3.7%) vasculitis, 520 (17.5%) infectious or post infectious arthritis, 263 (8.8%) periodic fever syndromes, 190 (6.4%) ocular disease, and 112 (3.8%) different other conditions. 1659 patients (35.8%) had a non-inflammatory disease; among them 753 (45.4%) had orthopaedic problems, 654 (39.4%) chronic musculoskeletal pain, 252 (15.2%) different other conditions.
Conclusions: These data show that a substantial number of children (more than 3.5 per 1000) are referred to a pediatric rheumatology clinic in Switzerland for a chronic rheumatologic condition. Among these patients, a majority (64.2%) presented an inflammatory disease with a mean incidence of 23.5 per 100'000, which represents more than 370 new patients per year. Therefore an early identification and adequate care are crucial to prevent long-term disabilities, and enough medical facilities should be provided in Switzerland to achieve this goal.

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Measles epidemic in a highly developed country: Low mortality, high morbidity and extensive costs

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Background: Vaccination with two doses of >95% of the population is necessary to eliminate measles. In Switzerland and especially in the central part, vaccine coverage is low (2006: 65%). This led 2006–2009 to a measles epidemic with thousands of cases and high costs. One death was noted in a formerly healthy 12 year old girl.
Patients and methods: All measles cases, either hospitalized or reported to the authority, in the canton Lucerne between 2006 and 2009 were included. Course, complications, immunization rates and costs of the hospitalized children were analyzed.
Results: A total of 1041 cases of measles were recorded; 758 (73%) were children <16 years of age. 56 (6%) of the patients were admitted to hospital; half of them were children (n = 26, admission rate 3.4%). Main complications were pneumonia with oxygen requirement (n = 19), bacterial infections of the base of the skull (n = 2) and acute measles encephalitis (n = 3). One child each developed acute appendicitis and diabetes mellitus type 1. No death was noted. Median hospitalisation costs were 18'780 CHF. The surveillance system was incomplete: Every third admitted child was not correctly reported to the authority.
Conclusion: Due to low vaccine coverage measles still account for epidemics with high morbidity and extensive costs. Instant reporting of all cases is crucial for disease control. Early identification of persons at risk allows timely immunization. Switzerland will remain of central importance to eliminate measles in Europe by 2015.

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Adrenal insufficiency in children with cancer treated with glucocorticoids

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Aim: To detect whether children with cancer have a sufficient adrenal function at presentation with fever in neutropenia (FN).
Patients and methods: In a prospective observational single-center study, serum was sampled at presentation with FN in pediatric patients with cancer presenting with FN, and stored at –20 °C. Cortisol concentration was measured by a commercially available ELISA. It was correlated to different clinical characteristics, including cumulative doses of past corticosteroid therapy. Cortisol concentrations <500 nMol/L were considered insufficient in the stressful FN situation.
Results: Serum samples were available in 21 (49%) of 43 FN episodes, from 14 patients aged 1.2 to 16.5 years. Patient characteristics and outcome were comparable in patients with and without serum samples. Freezing time was not significantly associated with cortisol. Median cortisol was 435 nMol/L (IQR, 262 to 653; range, <28 to 1301), with 11 concentrations <500 nMol/L (52%; exact 95% CI, 30% to 72%). There was a trend for cumulative doses of corticosteroid therapy within one month preceding FN to be associated with cortisol (Spearman's rho, –0.39; 95% CI, –0.85 to 0.07, p = 0.080), while earlier doses were not. Cortisol was not significantly associated with patient characteristics, temperature at presentation, or outcomes (adverse events, duration of hospitalization and intravenous antimicrobial therapy).
Conclusions: At presentation with FN, about one half of pediatric patients with cancer had an insufficient adrenal stress response, which was associated with past corticosteroid therapy. Larger prospective studies of adrenal response in FN are warranted.

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«Beziehungen helfen leben» – Suizidprävention für Jugendliche. Das Freiburger Modell, praktisch und konkret

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Einleitung: Rund 4% der männlichen und 8% der weiblichen Jugendlichen im Alter von 16 bis 19 Jahren geben in einem anonymen Fragebogen an, in ihrem Leben bereits mindestens einen Suizidversuch gemacht zu haben (Michaud P.-A. et al., 2002). Tatsächlich ist Suizid in der Schweiz die häufigste Todesursache der jungen Männer im Alter von 19–24 Jahren. Wir versuchen, mit unserem Freiburger Projekt zur Sensibilisierung, Information und Fortbildung – durchaus Evidence-basierten Vorgehensweisen – von primären Kontaktpersonen (Lehrerinnen) einer potentiellen Risikopopulation beizutragen.
Methoden: Die Lehrpersonen sämtlicher Oberstufen- und Berufsschulen des Kantons Freiburg werden mit einer Broschüre bedient, welche sie auf potentiell suizidgefährdete Jugendliche und junge Erwachsene aufmerksam machen soll. In Ateliers wird die Thematik vertieft. Diese Broschüre wird vorgestellt und diskutiert.
Resultat: Die Rückmeldungen von Schuldirektionen und Lehrpersonen sind positiv. Baseline und Outcome sind allerdings zu wenig präzis , als dass klare Auswirkungen, i.S. einer Verminderung von Suizidversuchen oder Suiziden in der Zielpopulation, belegt werden könnten.
Schlussfolgerung: Angesichts der gerade in der Schweiz hohen Suizidalität von jungen Männern sind konkrete und pragmatische Präventionsprojekte weiterhin wichtig und sinnvoll.

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Home support of obese children and adolescents by means of health information technology system: a pilot study for a psychosomatic therapy concept

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In Switzerland, 50% of obese children and adolescents show signs of psychological disorders. So far, existing interventions prove limited effectiveness and sustainability. IT-enhanced interventions have the potential to make the multi professional therapies more effective and less expensive. They aim not only modifying the patient`s behavior but also to positively influence their family system.
Methods: In cooperation with therapists, patients, their parents and communication scientists, a mobile health information System (HIS) was developed, consisting of a tablet pc and an activity meter with cooking and shopping support, relaxation tools, and the ability to measure emotional parameters. With the help of a goal-setting service, the HIS supports physical activity, relaxation exercises, emotional self-reflections and monitoring of eating speed in everyday situations. Excessive HIS use is prevented by measuring the screen time consumed.
The HIS was developed in several iterations and evaluated on 10 patients and their parent by the means of validated tests.Results of this pilot study show the usefulness of this HIS (Score 4 ± 0.8). It allows the patient verify and change his behavior at any time, while allowing the family system to find specific solutions and to promote cooperation within itself. The therapist is able to learn more about emotional burdens which may lead to problematical eating habits, visualize them in conversation and work them out with the patient.
Conclusion: Often impaired self perception of obese patients can be objectified with HIS, but to examine the long-term effectiveness of HIS, a randomized controlled trial has been initiated.
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Sexual behaviour problems in children and adolescents: proposal for assessment and management in the clinical practice. A help for the decision taking clinician

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Introduction: Challenging behaviour and testing out limits are part of the developmental process of a child. Therefore, presentation of sexual behaviour problems and inappropriate sexuality amongst juveniles have long been labelled as "exploratory" or even been overlooked (Becker et al., 1993). However, the last decades have focused more critically on children and juveniles whose conduct is sexually challenging. Offense statistics especially from America added their share to the discussion. Juveniles committed 22 percent of all sex crimes in the U.S. in 2007 (Christiansen and Vincent, 2013, U.S. Department of Justice, 2007). Recidivism rates in adolescent sex offenders of 5.6–10% are being reported (Caldwell, 2010). Although numbers of adjudications for adolescent sexual crimes seem to decrease (Swiss Federal Statistical Office, 2014), we are far from a comprehensive and insightful assessment and management of these children and adolescents, much less an "easy-to-use" guideline.
Methods: We reviewed the recent literature with a focus on sexual behaviour problems in children and adolescents, their assessment, management, recidivism rates, predictors and typologies (e. g., Christiansen and Vincent, 2013, Driemeyer et al., 2013, Riser et al., 2013, Chu and Thomas, 2010). We also validated (qualitatively) our clinical cases in regards to the same parameters. This we did with our forensic population and our general psychiatric population.
Results: We came up with a proposal for a pragmatic nonetheless professional decision taking path.
Conclusion: This suggestion can be used when faced with sexually challenging or violating behaviour of children or adolescents in the pediatric or psychiatric clinical practice.

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Outcome of patent processus vaginalis incidentally diagnosed by laparoscopy

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Introduction: Laparoscopy is frequently performed for children with intestinal or gynecological pathologies. Concomitantly patent processus vaginalis (PPV) might be observed in presence of normal groin and genitalia. The aims of this study were to determine the frequency and the natural history of PPV, i.e. to determine the risk of inguinal hernia in children with and without PPV.
Patients and methods: From 10/2000–10/2005 all children (<16y) undergoing a laparoscopic procedure for other reasons than PV-related pathologies were prospectively included. Exclusion criteria: previous history of inguinal/genital anomaly or inguinal surgery, abnormal physical exam of the inguinal area. The internal inguinal rings were assessed and documented during initial laparoscopy (IL). Follow-up was made by phone inquiry and medical visit if needed. Median follow-up was 10.5 years (range 1.4–12.8 years).
Results: A total of 416 patients were included in the study. IL was performed for 349 appendicitis, 26 gynecological affections, 22 other intestinal diseases, 19 other reasons. Median age at IL was 12.4 years (range 3 days–18.1 years). In total 42 PPV were found in 37 patients (9%), i.e. 32 unilateral and 5 bilateral PPV. No child with closed PV developped an inguinal hernia during follow-up; 1 child showed, at the age of 16.2 years, a hydrocele where the PV was observed to be closed at IL (3.4 years after IL); 4 children (3 boys and 1 girl) with PPV at IL (10%) presented with an inguinal hernia on the concurrent side (at a median age of 15.9 years (range 11.7–17.3 years) at a median of 22 months after IL (range 11–50 months), as compared to 1% in the whole study population.
Conclusion: The observed 10% occurrence of hernia development in children with PPV might justify its prophylactic closure during IL, if easily feasible. If left patent, parents of and children with incidentally diagnosed PPV must be carefully informed about possible inguinal hernia development within several years after IL.

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Thoracoscopic sympathectomies for primary palmar hyperhidrosis in children and adolescents

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Introduction: Primary palmar hyperhidrosis has been proven to be best treated by transaxillary bilateral thoracoscopy in adults.
Method: Review of 57 sympathectomies in 28 patients between 1998 and 2013. Mean age 14.0 years (4.4 to 21.0 years), sex ratio M / F 1:4.2.
Technique: Transaxillary bilateral thoracoscopy performed with a monopolar 3 mm hook without port and a 5 mm telescope in prone-lateral position.
Results: 28 bilateral sympathectomies T2–T3 (27 in a single step) and 1 left unilateral T1–T2 complement for an insufficient result. Length of hospital stay: 1.4 days.
No conversion, no Claude-Bernard-Horner syndrom. 28% of postoperative transient hypoesthesia. Brief unilateral thoracic drainage for 7 pneumothorax (23%). 13 minor pneumothorax not requiring any drainage.
Mean follow up of 6.4 years: both dry hands in 93% of cases. 2 asymmetrical results with one wet hand requiring successful additional sympathectomy T1–T2. 6 Compensatory sweating (21%) (especially in the back), 3.5% only during exercice , 42% absent. 64% patients complained preoperative axillary hyperhidrosis sweating as well. It disappeared or was significantly reduced in all cases.
Conclusion: Primary focal hyperhidrosis is a life-altering condition best treated by thoracoscopic sympathectomy even in children with few complications. It may be followed by a compensatory sweating, which is very well tolerated.

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Thoracoscopic approach to mediastinal masses

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Introduction: Tumors of the mediastinum reflect the nature of tissue in the particular section of the mediastinum in which they arise. Those include neurogenic tumors, esophageal duplication, bronchogenic cysts, bronchogenic cysts, lymphoma, germ cell tumors, teratoma, and others.
Thoracoscopic approach to those entities has been increasingly advocated.
We report on selected cases and discuss criteria for the indications of minimal invasive surgery.
Methods: Selected cases are presented where thoracoscopic approach to a mediastinal mass had been anticipated with short videos to illustrate surgical approach.
Evidence for minimal invasive access will be discussed based on the review of current literature.
Results: In selected cases, thoracoscopy proved to be safe and effective for the approach to mediastinal tumors. However in doubt for patient's safety and/or clear margins, conversion to thoracotomy has to be considered.
Conclusions: Thoracoscopic approach to mediastinal masses offers appropriate surgical access for a wide range of entities and provides well known advantages.
However removal of mediastinal tumors is advanced surgery. Therefore it requires critical appraisal and further long-term outcomes.

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DSD surgery in Cameroon

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Introduction: The management of children presenting with a Disorder of Sex Development (DSD) requires a multidisciplinary team, allowing for optimal management which often shows to be very difficult. This is often the case in our countries – but what about children living in Sub-Saharan Africa? Herein we describe a project of an international medico-surgical cooperation (France, Switzerland, Cameroon) managing these children.
Patients: Since 2009, 9 missions were conducted, of 10 days each, when 252 children were examined.
Results: Diagnoses: 46,XY DSD: 104 (of which 92 hypospadias). 46,XX DSD: 27 (14 Congenital Adrenal Hyperplasia (CAH) and 13 46,XX ovotesticular DSD). Sex chromosomal DSD: 2. Non-hormonal DSD: 13 (of which 12 exstrophy-epispadias complexes). Others: 106. 121 procedures were performed: 65 hypospadias, 25 cystoscopies, 9 feminizing genitoplasties, 22 others. Three children were operated in Geneva because they required complex recontructive surgery or specific equipment. Mean age at operation was 7 2/12 years. There were 5% complications: 3 boys with dehiscence and 3 fistulas after hypospadias repair. Not seen were any systemic infectious nor metabolic complications, in particular in children presenting CAH.
Conclusions: In Sub-Saharan Africa, DSD create complex management difficulties. There is a considerable need for diagnosis (genetic and endocrinological) as well as medical-surgical management. Appropriate management seems feasible as part of a well-structured North-South cooperation.

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Pin it to win it? An analysis of pediatric distal forearm fractures

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Introduction: Distal forearm fractures are frequent injuries during childhood. Due to the high potential of remodeling in pediatric fractures, they usually can be treated nonoperatively with closed reduction and casting. Secondary dislocation may occur and requires additional closed reduction, cast wedging and /or osteosynthesis with percutaneous pinning. The aim of the study was to find predictive parameters, which indicate an increased risk for secondary dislocation and therefore operative treatment.
Methods: Retrospective chart review of all patients with dislocated forearm fracture and/or distal radial fracture seen in our clinic from 1.1.2013 – 31.12.2013. Inclusion criteria were closed injury and open distal radial physis treated initially nonoperatively. Patients have been followed up according to a standard protocol at our institution, including long-arm-splint, clinical and radiological follow up. If a secondary dislocation was seen, either a secondary closed reduction and percutaneous pinning were performed or the natural course was observed.
Results: Our study presents the results of more than 100 children with distal forearm or radial fracture respectively. They were treated either with closed reduction and casting or just with casting. 22 patients out of those treated initially with closed reduction and casting, the fracture healed with angulation ≥10°. 5 patients required additional closed reduction due to secondary loss of reduction and out of them 4 patients have then been treated operatively. In 11 patients the fracture healed without any intervention with an angulation ≥10°.
Conclusion: Our results allow us to classify risks of secondary dislocation in distal forearm or radial fractures, and thereby prevent unnecessary or delayed surgical procedures for children as well as help surgeons in their decision making process.

Posters SGP A / Posters SSP A: Growth, metabolism, endocrinology, gastroenterology, adolescents

P108

New Swiss growth charts: their influence on diagnosis of over- or underweight and of growth disorders

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In 2011, nationwide growth charts were introduced in Switzerland, replacing different references in Germanic (1st.ZLS,1955-77) and Romansh region (Sempé1979), and, for BMI only, German references (Kromeyer 2001). The new standards for children aged 0–5 years include data of healthy breast fed children on five continents. The references for children aged 5–18 years are re-constructed from data of the WHO and the US-American NCHS growth curves. The aim of this study was to introduce new charts in the paediatric department, to examine their influence on the prevalence of nutrition and growth related disorders and to check the quality of auxologic measurements.
Methods: Calibration of anthropometric [1] equipment and measurement procedures were verified according to standard quality management. Accuracy and variation coefficients measuring height / length were corrected (<0.01cm differences, n = 20), with the exception of the measuring rods used in infants, producing errors between +0.24 and –0.7 cm.
6007 anonymised weight and height datasets of children measured between 2000 and 2012 were included. The degree of deviation of the "new" from the "old" percentile charts was assessed by the kappa measurement of agreement for different age groups.
Results: "New Swiss" WHO-percentiles for length/ height show a broader normal range and a good agreement with ZLS charts (K = 0.88 and 0.83) in boys and girls. Yet 2 to 9% of boys aged 2 to 15 years are now classified as normal, while they would have been defined as short statured before. Female height charts are broader now and, except for the age of puberty, classify more girls as normal, namely 4.7% of those having been short statured and 1.4% of those classified as too tall before.
WHO-weight percentiles, of boys and girls, are shifted to a higher normal range than ZLS curves.
In the first year of life, the new BMI-percentiles find 4% less obese infants, but at school age, obesity was identified by WHO-BMI-references in up to 10% more boys and 2% more girls than with references from Kromeyer.
Conclusions: Auxological measurements were of a high quality, except for the use of the measuring rod. With respect to the fact that new Swiss percentiles find less short statured and more obese school aged children, the use of ZLS curves and further markers of growth or obesity should be added, such as parental target height or waist circumference.

P109

Demonstration of the effectiveness of zinc in diarrhea of children aged 2 months to 5 years in Lausanne Childhood Hospital

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Introduction: The effectiveness of zinc in childhood diarrhea has been demonstrated in developing countries. It helps to decrease the duration and severity of diarrhea. There is currently no sufficient data to justify its use in developed countries, where there is a priori no zinc deficiency.
Methods: We designed a prospective randomized clinical trial zinc vs placebo in healthy children aged 2 months to 5 years consulting in our emergency unit with diarrhea (>3/day for less than 72 hours). 20 mg (≥6 months old) or 10 mg (<6 months old) of Zinc sulfate as a dispersible tablet was prescribed once a day for 10 days. Measured outcomes were the duration and severity of diarrhea.
Results: 87 patients (median age 14 months, range 3.1–58.3) were analyzed in an intention-to-treat approach. 42 patients took zinc and 45 placebo. There was no difference in the duration of diarrhea (median duration of 67 hours, IQR 27-94) nor in the frequency of diarrhea between day 2 and 4 of treatment (median of 7 diarrhea, IQR 3-9). However only 5% in the zinc group still had diarrhea at 120h of treatment compared to 20% in the placebo group (p = 0.05). 31 patients (13 zinc and 18 placebo) were available for perprotocol analyses. There was a statistically significant difference in the duration of diarrhea between zinc group (median of 47.5 hours, IQR 18.3–72) and placebo group (median of 76.3 hours, IQR 52.8–137) (p = 0.03). The frequency of diarrhea was also lower in the zinc group (3 diarrhea between day 2 and 4 of treatment, IQR 1-8) compared to placebo group (9 diarrhea between day 2 and 4 of treatment, IQR 7-9) (p = 0.02).
Conclusions: Zinc treatment is associated with a decrease in diarrhea frequency and severity in children aged 2 months to 5 years old in a developed country. However, poor compliance results in a questionable clinical significance (intention-to-treat analysis).
A different dosage form should be considered (oral rehydration solution?).

P110

Individual therapy equals group therapy in significantly improving mental and physical health in obese children

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Long term efficacy of family-based behavioral therapy for obese children in group programs has been demonstrated by randomized controlled studies, confirming reduction of obesity in 70% of children until 1 year after the end of therapy. Since physical incapacity, psychological co-morbidity or parental occupation hinder obese children in participating in therapy groups, the aim was to examine whether treatment in a multiprofessional individual setting (MIS) is also effective in improving the health of obese children.
Methods: In a single center prospective longitudinal cohort study, 52 children were treated within nationally certified group programs (MGP: 88 h/12 months of therapy for children and parents, each 1/3 of time by dietician, psychologist and sport's teacher) and 96 children in an individual setting (MIS: about 22 h/12 months for children and their parents on a similar multiprofessional basis). At therapy start (T0) and one year thereafter (T2), BMI and psychological and nutritional parameters were assessed by validated, standardized questionnaires. (strengths and difficulties, SDQ; adiposity-related eating cognitions and disorders, AD-EVA) and by food frequency lists (FFL).
Results: At T0, physical and psychological parameter in MIS children (11.4 ± 2.9 years; 54% girls; 66% morbidly obese with BMI >99.5th percentile of WHO references, BMI-standard deviation score (SDS) 2.8 ± 1.1) were not significantly different from MGP patients. Both after MGP and MIS therapy, obesity was significantly reduced (ΔBMI-SDS = –0.19 and –0.33, respectively, p <0.000). Morbidly obese children scored significantly higher in mental health difficulties compared to less obese ones (p = 0.018). At T2, an improvement in total SDQ score was more prominent, the higher the initial score was. Diet composition was neither associated with BMI nor with therapy outcome. While preclinical eating disorders such as vomiting were rare and even decreased until T2 (p = 0.03), scores of emotional eating, craving for food and preoccupation with body shape were more pathological than in the normative sample, but all improved during therapy, mainly craving (p = 0.048).
Conclusions: The study shows equal effects of multiprofessional group and individual settings on weight loss and improvement of eating habits in obese children, without producing side effects such as eating disorders. Mental health problems of obese children need additional attention.

P111

Anti- exocrine pancreatic and proteinase-3 antineutrophil cytoplasmic antibodies in paediatric patients with inflammatory bowel disease – a single center experience

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Introduction: Serological testing using the classical antibodies anti-Saccharomyces cerevisiae antibodies (ASCA) and perinuclear anti-neutrophil cytoplasmic antibodies (pANCA) is commonly used in patients with inflammatory bowel disease (IBD) to help to distinguish between Ulcerative Colitis (UC) and Crohn's Disease (CD). However, there is some overlap, reducing their specifity. Recent studies postulated that exocrine pancreatic autoantibodies (PAB) and anti-proteinase- 3 antineutrophil cytoplasmic antibodies (PR3-cANCA) are of additional value in indeterminate cases. Our aim was to evaluate these antibodies in a single center.
Methods: We retrospectively analyzed the antibody profiles of patients who underwent antibody testing at our pediatric IBD clinic.
Results: Antibody profiles from 56 pediatric IBD patients were included. Of those, 34 had CD (median age at diagnosis 10.9y, 17 males), and 22 suffered from UC (median age at diagnosis 10.1 y, 8 males). 21/31 (68%) CD-patients were ASCA IgA and IgG positive. On the other hand, 4/17 (24%) UC-patients tested for ASCA were either ASCA IgA or IgG positive. PAB-positivity was found in 9/17 (52%) CD-patients. In contrast, none of the 13 UC-patients tested for PAB was positive. PAB was detectable in 3 CD-patients who were negative for ASCA.
13/19 (69%) UC- and 7/31 (23%) CD-patients had pANCA by indirect immunofluorescence (IIF). All pANCA positive CD-patients had colonic disease. Four patients (10%, 3 males) were positive for PR3-cANCA determined by IIF as well as by PR3-ANCA-ELISA. None presented with any features of vasculitis. Two were diagnosed with CD and two with UC.
Conclusions: Our analysis supports the assumption that PAB-testing is of additional value for distinguishing patients with Crohn's Disease from those with Ulcerative Colitis. Regarding PR3-cANCA we cannot confirm its postulated specificity for UC. More data are needed in order to draw a clear association with UC.

P112

Exclusive enteral nutrition and its potential in perianal Crohn's disease – two cases

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Exclusive enteral nutrition (EEN) is an established treatment modality for patients with Crohn's Disease (CD) with small bowel involvement; in this group of patients remissions rates of EEN are similar compared to a course of oral steroids. The benefit of EEN in patients with fistulizing perianal disease is not well established.
Case 1: A 12 year old male patient with perianal abscesses with fistulas, requiring surgical intervention a year before, presented to our hospital for workup of CD. No complete perianal healing was achieved despite antibiotic therapy. Additionally, he had lost 3 kg over the last year. Treatment with exclusive enteral nutrition (EEN with Modulen) for 6 weeks was started via naso-gastric tube. After 2 weeks, almost complete healing of perianal disease and weight gain of 1 kg was achieved. After six months, there was no recurrence of perianal disease and he was back on his pre-disease-percentile with a weight gain of 4 kg.
Case 2: A 15 year old male patient with Crohn's disease under therapy with Infliximab developed a perianal abscess with fistula. Surgical intervention and standard drug therapy did not result in complete healing of the perianal disease during 8 months. Therefore, EEN was started with good local response after 6 weeks and a weight gain of 9 kg over 3 months.
Conclusion: EEN was effective and led to resolution of perianal disease in both patients. Steroids and anti-TNFa-therapy with all their known side effects could be avoided in one patient. In the other patient, clinical remission was induced and maintained with EEN, after having lost response to Infliximab.
Therefore, we postulated that EEN may have an important and underestimated value in fistulizing Crohn's disease. EEN should be offered to patients with perianal disease. Presumed mechanisms include alteration of microflora and immune response as well as treatment of malnutrition.

P113

Drug prescription to obese paediatric patients in ambulatory care in the United States

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Introduction: Excess bodyweight has a devastating impact on health, especially when already present in childhood. Obesity is associated with a higher prevalence of chronic diseases requiring pharmacotherapy like dyslipidemia, type 2 diabetes and arterial hypertension.
We sought to analyze the prescription patterns in obese children and young adults compared to lean pediatric patients based on the National Ambulatory Medical Care Survey (NAMCS).
Methods: Clinical data of children, adolescents and young adults aged 2 to 21 years entered into the NAMCS 2006–2010 were obtained from the Centers of Disease Control and Prevention. Data analysis included presence of chronic diseases, reason for visit, number of medications coded and drug category (vaccinations and topical agents excluded). Prescriptions were stratified by age (2–5 yr, 6–12 yr, 13–17 yr, 18–21 yr) and presence of obesity. Comparative analyses were performed between lean and obese patients.
Results: Fifty-nine percent of obese patients were prescribed medications compared to 67% of lean patients. Thirty-two percent of the obese suffered from at least one chronic condition compared to 17% of the lean (p <0.0001). Obese patients had more medical consultations for preventive care (35 %) or a chronic problem (31%) compared to lean patients (25% / 24%) whose major problems were acute (46%, in obese 31%, all p <0.0001).

The most frequently medications used in obese patients were central nervous system (CNS) agents (22%), respiratory agents (19%) and psychotherapeutic agents (13%) as compared to CNS agents (25%), anti-infective (23 %) and respiratory agents (23%) in the lean group. There was a higher use of psychotherapeutic agents (13 % vs. 7%), metabolic agents (6% vs. 1%) and cardiovascular drugs (5% vs. 2%) in obese patients (all p <0.0001), whereas anti-infective agents were more often prescribed to lean patients (23 % vs. 13 %, p <0.0001). Metabolic agents were significantly prescribed more often to obese adolescents (>12 yr) and cardiovascular agents to obese young adults (>17 yr).
Conclusions: The drug prescription pattern to obese pediatric patients is different due to the chronic comorbidities that require pharmacotherapy compared to lean children who seek medical advice more often for acute conditions that require treatment (e.g. acute infections).

P114

Herlyn-Werner-Wunderlich syndrome, when embryology leads to uncommon abdominal pain

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Introduction: Obstructed hemivagina with ipsilateral renal agenesis associated to didelphys uterus is a rare condition known as Herlyn-Werner-Wunderlich syndrome (HWWS). HWWS represents a minority of Mullerian duct anomalies. After menarche a hematocolpos develops with subsequent development of dysmenorrhoea, abdominal pain and pelvic mass, sometimes associated with urinary retention and/or constipation.
Case Summary: We describe a 12 year old female adolescent who presented with increasing abdominal pain associated with pain by passing stools. She mentioned menarche at age eleven. This febrile girl showed a tender periombilical and suprapubic abdominal palpation, no signs of peritonitis and no palpable mass. The external genitalia were normal.
The abdominal ultrasound showed a pelvic mass precised by abdominal MRI which demonstrated a uterus didelphys, duplicated vaginal cavities with a left sided obstruction associated with hematocolpos and ipsilateral renal agenesis, fulfilling the diagnostic criteria for HWWS.
The patient was treated with resection of the vaginal septum and 200 mL of a hemo-purulent discharge was drained allowing quick relief of her symptoms. Oral antibiotic treatment with amoxicillin/clavulanic acid was started. Klebsiella pneumoniae was identified.
Conclusion: Herlyn-Werner-Wunderlich is a rare cause of abdominal pain. The latter is due to blood retention (hematocolpos) possibly complicated with secondary infection. MRI seems to be the best imaging diagnostic tool. The presence of menstruations does not necessarily rule out hematocolpos.
Gynecological, obstetrical and nephrological follow-up are warranted in all patients with HWWS.

P115

When the best for the child turns to the worst: severe malnutrition in a moribund child

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Introduction: Fully breastfed infants of vegan mothers are at known risk of deficiencies of trace elements and vitamins. A delayed diagnosis of a severe vitamin-B12-deficiency can be deleterious.
Case report: The eight month old female infant was presented to our emergency department in a moribund state: The girl was in a reduced general state, altered consciousness, had multiple petechiae and purpurae and an exanthema. She was pancytopenic and febrile, so she was treated for meningoencephalitis and sepsis in our ICU. With a history of being the second child of a strictly vegan mother and still fully breastfed we simultaneously administered Vitamin-B-12 (1000 µg Vitarubin® s.c.). Subsequently the neurologic state improved rapidly: On the next day she began to note her surroundings, on the third day she began to play with things. She was fed parenteral and began on solid food. The mother continued breastfeeding her.
We subsequently proved the severe cobalamin deficiency (Hyperhomocysteinaemia and -uria, Hypermethylmalonacidaemia and -uria and non-detectable level of cobalamin). Furthermore we found a zinc deficiency and a hyperparathyroidism.
The severe malnutrition was a consequence of a whole ideology: The family fed strictly vegan and saltfree, feared electromagnetic fields and the children were totally unimmunized.
Conclusion: It's not only the medical intervention but also the psychosocial aspects of our patients that we have to consider in order to provide sustainable healthcare for the sake of the wellbeing of the child.

P116

Antithyroid arthritis syndrome associated with antithyroid therapy in a 13 year-old girl

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Introduction: Antithyroid drugs are considered first-line treatment for most children with Grave's disease. Carbimazole is the best option in children, due to the elevated risk of hepatotoxicity of propylthiouracil. Several minor and major adverse reactions can occur. Antithyroid arthritis syndrome (AAS), a major side effect, has a frequency of 1–2%, although only a few cases have been reported in children.
Case report: A 13 year-old girl was referred to our emergency department with generalized pruritus, dyspnea, nausea and joint-swelling. She had been treated with carbimazole (1.15 mg/kg/d) and propranolol since 14 days due to recently diagnosed Graves' disease. Clinical examination showed swollen and painful joints (knees, ankles, elbows, wrists and fingers). Laboratory results showed no inflammation, positivity for antinuclear antibodies (ANA) but negativity for antineutrophil cytoplasmic antibody (ANCA).
Adverse drug effect was suspected and carbimazole was withdrawn; arthritis disappeared rapidly. After 5 weeks without treatment, she was admitted to the hospital with a severe hyperthyroid state which required rapid thyroidectomy. Euthyroid status preinterventionally was achieved after 5 days of treatment with potassium iodide (Lugol solution 5%) and oral dexamethasone, which were both well tolerated. Six months later, the girl was free of complaints under treatment with levothyroxine.
Conclusion: Polyarthritis is a rare adverse effect of antithyroid therapy and can be part of AAS or associated with ANCA-vasculitis. Once recognized, the medication should be stopped immediately because of the potentially life-threatening course. A rapid plummering with application of high dosed iodine before thyroidectomy is effective within few days, if required.

P117

A longinatudinal study assessing an IT-supported neurological feedback in obestity intervention for children and adolescent emotional self control

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New approaches in the treatment of obesity in childhood and adolescent are necessary. Therefore we developed a mobile health information system (MHIS), which is able to help patients controlling there emotions in concern of their eating habits. We used neurological biofeedback to identify individual items that are able to support the patient to overcome the strong impulse of uncontrolled eating. To identify the right item we used a skin conductance approach.
IT-supported emotional interventions promise to help therapists and patients with objective real-time data in real-life situations of patients. As a result, therapists are able to guide patients more effectively without the need for several investigative hours of consultations. The usefulness of a mobile health information system in this point has already been shown in a preliminary study but results from a longitudinal investigation are still missing. A one-year longitudinal study has therefore been started in December 2013. Four objectives are addressed in this study: (1) evaluation of the technical feasibility of the MHIS in a real-life intervention setting, (2) evaluation of the practical application of the MHIS-supported intervention, (3) evaluation of the feasibility of the evaluation instruments and, (4) preliminary identification of significant predictors and medical effects of MHIS use. 13 patients in the eastern part and 6 patients in the western part of Switzerland participate in MHIS obesity intervention. In addition, seven patients have been assigned to a control group without MHIS support. It is assumed that the MHIS increases the individual and shared understanding in therapist and patient teams with regard to health knowledge, which, in turn, is expected to increase medical outcome parameters.
Funded by: SNF Grant Cr 10/1_13552
Keywords: Interdisciplinary research, obesity, therapy, prevention, children, protocol, longitudinal study, adolescents, mobile health intervention system

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Food protein-induced enterocolitis syndrome caused by cultivated mushroom: a case report

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Introduction: Food protein-induced enterocolitis syndrome (FPIES) is a potentially severe non-IgE- mediated food hypersensitivity**.** Symptoms may be chronic while the food is regularly consumed or present acutely after an occasional ingestion. The most common causative foods are cow's milk and soy, typically diagnosed before one year of age. A wide range of solid food has also been reported to cause FPIES, especially rice and oat.
Case report: We report the case of a non-atopic 9-years-old girl, who had experienced two episodes of profuse vomiting few hours after ingestion of cultivated mushroom when she was 7 years old. Since then, mushrooms have been avoided from her diet. In order to confirm our suspicion of FPIES, a diagnostic oral food challenge (OFC) was performed two years after the initial reaction. Tree hours after ingestion of 22.5 grams of cooked cultivated mushrooms, she experienced two episodes of vomiting with mild lethargy. The complete blood count showed a typical increase in neutrophils count from an initial value of 3180 to 7240 cell/mm³ at four hours. An intravenous bolus of normal saline and a dose of methylprednisolone were administrated, leading to a resolution of the symptoms within an hour. According to Powell's diagnostic criteria, we confirm a mushroom induced-FPIES and strict avoidance of this food was recommended.
Discussion: To our knowledge, this is the first reported case of FPIES to mushrooms. Diagnosis of solids FPIES can be challenging and is often delayed because of a low index of suspicion, particularly in older children. Indeed, while classic FPIES presents in infancy, there are an increasing number of reported cases in older children and even adults. Delayed diagnosis and misdiagnosis is common and can lead to incorrect treatment, invasive treatment, or both. Thus, it is important to consider FPIES in children of any age presenting characteristic digestive symptoms several hours after food ingestion. The diagnosis is usually based on a typical clinical history and confirmed by an OFC performed under medical supervision due to the risk of severe reaction upon re-exposition.

P119

Standardized program for fertility counselling in post-pubertal adolescent patients – a single centre experience

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Infertility is a severe complication of many cancer treatments and might strongly impair the quality of life of long term survivors. Advances in reproductive technology enable some post-pubertal patients to preserve fertility before the initiation of treatment for cancer Despite a number of guidelines published by leading professional organizations addressing cancer and fertility, recent research has shown that fertility preservation is not routinely discussed with paediatric patients and their parents. The lack of a local institutional algorithms approaching this issue represents one of the most relevant barriers to overcome this problem.

We developed in our institution a standard. It contains an algorithm which defines the patient population and the timeframe for possible interventions.
According to the "Swiss Childhood Cancer Registry", there were 190 newly cancer cases diagnosed in Switzerland in 2012; of those 33 were diagnosed at our institution. Six of thirty-three (18%) newly diagnosed patients were post-pubertal and 4/6 (66%) had fertility counselling prior to oncologic therapy.
After establishing the SOP in 2013 all newly diagnosed cases (32 patients) at our institution who were post-pubertal 9/9 (100%) benefited from fertility counselling. The patients' characteristics and details on fertility preservation are described in table 1. The introduction of a SOP for fertility counselling and preservation allowed us to offer fertility preservation counselling and preservation within two days of diagnosis to all potential candidates.
Fertility counselling and preservation in post-pubertal patients remains a big challenge for the medical team. Paediatric oncologists should be aware of this topic.

N	Age	Gender	Diagnosis	Proposed fertility preservation	accepted	refused	successful
1	13	f	Osteosarcoma	Ovarian tissue preservation	y		y
2	14	f	Ewing sarcoma	Ovarian tissue preservation		y	
3	13	f	Germ cell tumor	Ovarian tissue preservation		y	
4	22	f	ALL	Ovarian stimulation for oocyte vitrification		Lack of time	
5	21	f	AML	Ovarian stimulation for oocyte vitrification		Lack of time	
6	16	m	Germ cell tumor	Cryopreservation of sperm after TESE	y		y
7	17	m	T-lymphoblastic Lymphoma	Cryopreservation of sperms	y		Y
8	16	m	Germ cell tumor	Cryopreservation of sperm after TESE	y		Y
9	16	m	M. Hodgkin IVB	Cryopreservation of sperm after TESE	y		Azoospermia

Patients characteristics

P120

Eine verzwickte Angelegenheit – a tricky matter: recurrent superior mesenteric artery syndrome in an adolescent female

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Introduction: Superior mesenteric artery syndrome (SMAS) is a rare phenomenon resulting from compression of the 3rd portion of the duodenum between the superior mesenteric artery (SMA) and the aorta [1–3]. Incidence is estimated about 0.1% to 0.3% with a female predominance among teenagers and young adults [3]. In most cases conservative management is successful with rates ranging from 52% to 83% [1, 2].
Case presentation: A 15 year-old female high performance athlete (BMI 17.6 kg/m²) presented following bilious vomiting and was suspected to have SMAS with significant dilatation of the duodenum on abdominal ultrasound. Symptoms resolved after nasogastric tube aspiration and IV-rehydration over 48h. The patient relapsed 15 months later and responded to the same management. At the 3rd episode, midline sagittal reformatted MIP image from an abdominal CT angiographic study shows an angle of 8° (N = 28–65°) between SMA and aorta. Upper endoscopy confirmed near total obstruction of the 3rd duodenum. A nasojejunal tube was placed distal to the obstruction

delivering high caloric nutrition (completed with parenteral nutrition) for 4 weeks. Symptoms resolved after 6 weeks and a weight gain of 8 kg (BMI 19.8 kg/m^2). Follow up of 6 months shows no recurrence so far.
Conclusion: Nutritional management aimed at increasing the mesenteric fat pad and resolves SMAS in most cases. Surgery should be postponed until sufficient weight gain has been seen. Long term medical follow up is needed, since recurrence may occur.

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P121

High rate of chronic functional constipation in first-born children

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Introduction: A link between birth way, the acquisition of intestinal flora and the development of chronic constipation has repeatedly been suspected. The aim of our study was to evaluate birth circumstances in our cohort of children with chronic functional constipation.
Methods: Retrospective study on charts of patients with chronic functional constipation seen at the pediatric gastroenterology consultation of the cantons hospital of Fribourg between 1996 and 2010, assessing: gestational age, way of birth, birth weight, gender, breast feeding duration, age at first symptoms and first consultation. Of the 231 patients, 70 were excluded for incomplete data, prematurity or comorbidity.
Results: 161 term newborns, 88 male (54.7%), mean birth weight 3300g (SD 437), 40 caesarean sections (24.8%), mean duration of breast feeding 4.31 months (SD 5.6), mean age at first symptoms of constipation 29.4 months (SD 28.1), at first consultation 60.8 months (SD 41.2). Of these children 91 were first born (56.5%), 46 were 2nd born (28.6%), 24 were 3rd and more than third (14.9%). In comparison to the cantonal family structure 2nd born are significantly underrepresented in our cohort of children with chronic functional constipation.
Conclusion: 56.5% of the children with chronic functional constipation are first born. Does parental inexperience play a role?

P122

Growth hormone deficiency in an infant with Noonan syndrome: an unusual presentation

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Case: We report the case of a 14 months old boy with a Noonan Syndrome who presented with an isolated general tonico-clonic seizure and severe hypoglycaemia at 0.8 mmol/l. The diagnosis of Noonan syndrome was established during the first months of life by geneticians on phenotype. He was treated with Propranolol for a pulmonary stenosis and followed for a failure to thrive despite hypercaloric diet. Paroxystic upward deviation of the eyes, periods of poor visual contact were episodically observed and transient episode of pallor as well. Growth hormone deficiency GHD was assumed to explain height below the 3rd Percentil of Noonan Growth Curves. EEG was moderately abnormal without explaining the episodes of eyes deviation and poor visual contact. Brain MRI was considered normal. All the biological investigations, including IGF-1 were within normal limit. Measured during the episode of seizure associated with severe hypoglycemia, 0.8 mmol/l, GH was found very low and sustained the diagnosis of GHD. Conventional growth hormone treatment improved height and neurological symptoms disappeared.
Conclusion: In infant with neurological symptoms and repetitive hypoglycemia, metabolic disorder as GHD should be considered. Dosage of GH after severe hypoglycemia was as valuable as conventional GH stimulation tests.

P123

Watermelon seed rectal bezoar in a 9-year-old boy

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Bezoars are solid masses formed in the GI tract as a result of the consumption of indigestible or poorly digestible substances. They are classified according to their composition: hair (trichobezoars), vegetable matter, such as seeds, pits or fibers (phytobezoars) or medications (pharmacobezoars). Rectal seed bezoars are a common cause of fecal impaction in children in the Middle East where the custom of eating baked and salted sunflower, pumpkin and watermelon seeds is very popular, but is rare outside the Middle East region.
We describe the case of a 9-year-old boy, born in Switzerland, with Israeli origins, who was admitted to our hospital complaining of lower abdominal pain, excruciating rectal pain, constipation and encopresis. Rectal examination showed total anal sphincter atony. Five days before, he had eaten about 200 grams of salted unshelled sunflower seeds. Disimpaction by digital evacuation was performed under general anesthesia followed by saline irrigation. Anoscopy showed a circumferential ulcer of the anal canal, coated with a fibrin exudate with small areas of necrosis. The tonus of the anal sphincter improved over the following weeks and the boy subsequently reacquired continence.

P124

Cushing syndrome without origin

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Introduction: Cushing syndrome is a rare entity in children and generally it presents a female to male predominance. The most common cause in childhood is iatrogenic Cushing syndrome related to chronic administration of glucocorticoids or ACTH. Endogenous Cushing syndrome is divided into ACTH dependent (ACTH overproduction due to pituitary adenoma or ectopic production) and in ACTH independent causes (surrenal adenoma, carcinoma or bilateral hyperplasia).
Case report: We describe a case of a previously healthy 10 year old boy presented with a rapid weight gain associated with growth retardation, facial pletora and acne. He also complained of palpitations, sleep disruption and muscular weakness.
On clinical evaluation he presented central obesity, hypertension, excess sweating, acne, moon facies and hirsutism.
Biochemical investigation showed an elevation of ACTH, cortisol and other androgen hormones. A low dose dexamethasone suppression test was performed and showed a persistent high level of cortisol and ACTH and loss of cortisol circadian rhythm.
Cerebral and abdominal magnetic resonance imaging was performed and both resulted normal. Bilateral inferior petrosal sinus sampling has been done and did not demostrate the presence of an ACTH pituitary microadenoma. Total body octreotide scanning and repeated positron-emission tomography were also normal and have failed to determine the source of ACTH ipersecretion.
A therapy with ketoconazole was started with gradual increase of the dose with a good clinical and biochemical response for two years. But at the dosage of 1200 mg/die (maximal adult dose) the patient presented relevant bone retardation, hypertension and hyperinsulinism with a high possibility of multiorgan failure. Finally a bilateral total adrenalectomy was done and an adequate corticosteroid replacement therapy was started with a good outcome.
Conclusion: However the origin of ACTH ipersecretion, that is most likely cancerogenous, remain still unknown.

P125

Accuracy of Schofield's equation to predict resting energy expenditure in children with inflammatory bowel disease and in healthy controls

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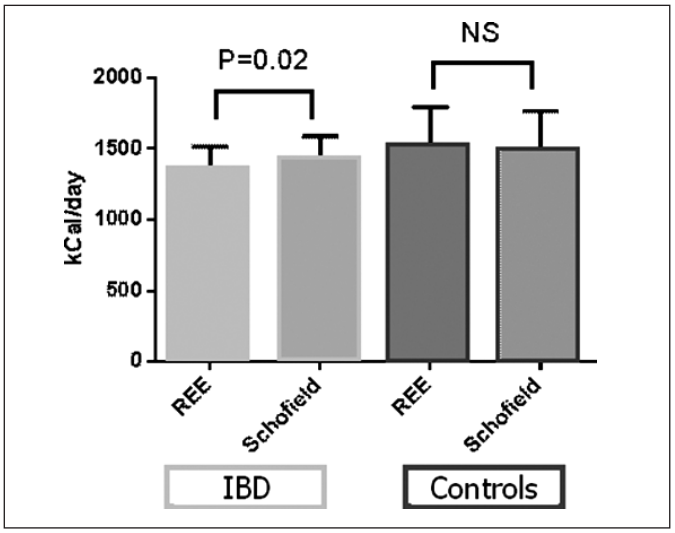
Introduction: Resting energy expenditure (REE) represents the amount of calories required for a 24-hour period by the body during a non-active period: it depends on age, body weight, and body composition (especially fat-free mass). It can be measured using indirect calorimetry or estimated by means of equations, which are not validated in children with inflammatory bowel disease (IBD). Indirect calorimetry is expensive, not easily available and often used for research purpose only: this is why equations to predict REE in sick children could be interesting.
The objective of our study was to assess the accuracy of Schofield's equation to predict resting energy expenditure (REE) in children with IBD and in healthy controls compared to REE measured by indirect calorimetry (QUARK RMR).
Methods: Twenty-one patients (11 girls; mean age: 14.8 ± 1.3 years (range 12–16)) with IBD (Crohn's disease n = 15, ulcerative colitis n = 6) and twenty-nine healthy controls (12 girls; mean age 13.1 ± 2.0 years (range 10–16.5)) were enrolled. Estimated REE was calculated using Schofield equation and compared to the value measured by indirect calorimetry. Paired t-test was performed and p-values <0.05 were considered statistically significant.
Results: Schofield's equation has a tendency to overestimate REE in children with IBD (1429 (±161) kCal/d vs. 1362 (±154) kCal/d, respectively; p <0.02), whilst it is very accurate in healthy controls (1505 (±262) kCal/d vs. 1521 (±273) kCal/d, respectively; p: NS).



Results

Conclusion: Schofield's equation can accurately predict REE in healthy children, but is not reliable in assessing REE in children with IBD. This may be explained by changes in metabolism and body composition in children with IBD.

P126

Dehydration: an underestimated causal factor of cerebral venous thrombosis?

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Introduction: Cerebral venous thrombosis is a rare disorder, which requires prompt diagnosis and treatment to prevent short and long- term complications and to reduce mortality. We report the case of a 10 year-old girl who presented dehydration, complicated by cerebral venous thrombosis.
Case description: A 10 year-old girl, known for developmental delay with autistic features, is admitted to our unit for Type C dehydration with compensated shock signs due to uncontrollable vomiting, accompanied by headache and subfebricular diathesis. Na+ value is 134 mmol/L. Hemodynamic and hydration state is rectified through intravenous rehydration. However, persistent fatigue, headache and recurrent vomiting do not allow perfusion weaning.Respiratory alkalosis (pH: 7.59, pCO_2: 22.4 mm Hg, HCO_3^-: 21.3 mmol/L), yawning and hyperpnea without tachypnea are also observed on the 4th day of hospitalisation, strongly evocating a SNC pathology. Cerebral MRI reveals transverse and left sigmoid sinus and internal jugular vein thrombosis. Family history is free, auto-immune disease and metabolic testing are normal, infection, hemoglobinopathy and renal diseases are excluded and there is no evidence of neoplasia or prothrombotic disorder. The patient receives anti-coagulation therapy for 3 months. Clinical evolution is satisfactory and cerebral MRI after 3 months of anti-coagulation is normal, permitting discontinuation. Thrombophilia test results are negative. Cerebral venous thrombosis due to severe dehydration is concluded.
Conclusion: Dehydration is an under-estimated yet preventable causal factor of cerebral thrombosis in the paediatric population and should not be neglected in the differential diagnosis. Nevertheless, other concomitant factors ought to be tested.

P127

Hashimoto's thyroiditis 6 years following Graves' disease

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Introduction: Graves' disease and Hashimoto's thyroiditis are rather uncommon in childhood. We describe the case of a girl who developed Hashimoto's thyroiditis 6 years after the diagnosis of Grave's disease.
Case report: In 2007, an 11-year-old girl presented with tremulousness of hands, palpitation, excessive sweating, goiter and exophthalmos. The laboratory work up disclosed the characteristic features of Graves' hyperthyroidism including low thyroid-stimulating hormone, high free thyroxine and high concentration of antibodies to the thyroid stimulating hormone receptor. On therapy with methimazole, euthyroidism was achieved within 4 months. Medical treatment was progressively tapered over 4.5 years. Eighteen months after discontinuation of treatment, the girl presented with asthenia. The diagnostic work up disclosed signs of latent hypothyroidism (thyroid-stimulating hormone 1210 IU/L; free thyroxine 9.3 pmol/L), high levels of antibodies to thyroglobulin thyroid peroxidase and hypoechogenicity on ultrasound. The diagnosis of Hashimoto's thyroiditis was made, and a therapy with levothyroxine was started. The girl is currently well on this therapy.
Conclusions: The history of Graves' disease results in hypothyroidism in ≈20% of patients previously treated with antithyroid drugs by different mechanisms. The present case indicates that in these patients hypothyroidism sometimes results from Hashimoto's thyroiditis.

P128

Self-performed closed testicle detorsion maneuver based on Google teaching: see it,… DO IT!

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Introduction: Internet is an important source of knowledge for teenagers, including for subjects about their own health. We present a case where Google helped a teenager to save his testicle.
Case: A 14 years old boy presented in the emergency department reporting the abrupt onset, 3–4 hours before the consultation, of pain and swelling of his right hemiscrotum. The physical examination revealed a moderately swollen, slightly painful, not ascended right testicle. The cremasteric reflex was present. There was no fever nor history of trauma, and the patient had not been sexually active. Despite these findings, the history was suggestive of a testicular torsion. Further history taking revealed that the teen, facing his own signs and symptoms, searched the web on his smartphone, found on a medical website (http://www.allodocteurs.fr/actualite-sante-qu-est-ce-qu-une-torsion-de-testicule--1030.asp?1=1) the description of testicular torsion with detailed instructions about closed detorsion manipulation,… and performed it correctly (rotating the right testicle anticlockwise) on himself in the school's lavatory.
As there was at the moment of the consultation no sign of testicle compromise, an elective surgical fixation was scheduled.
Conclusions: In this case, the informations found on the net, and especially with Google helped this patient to identify and treat his own testicular torsion, and likely save the testicle.
This highlights an important source of knowledge on their health for teenagers. We suggest that this potential, including Google, YouTube and other popular sites remains underexploited.

P129

Serum marker S100B is associated with cognitive outcome and ongoing cognitive symptoms in children after mild traumatic brain injury

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Objective: Mild traumatic brain injuries (mTBI) occur very often in childhood. Although most children recover completely within 3 months, roughly 10% suffer ongoing attention and memory problems. Due to the high incidence rate of mTBI, it is desirable to find biomarkers indicating children who are at risk for developing cognitive problems. Thus, the aim of this study was to explore whether acute serum marker S100B is related with cognitive performance as well as with post-concussive symptoms (PCS) four months after the injury in children after mTBI.
Methods: We prospectively investigated children (age 6–16 years) with mTBI (GCS 13–15, n = 38, mean age at injury: 10.9 years) and children with orthopedic injuries (OI, fractures of the adominant upper extremity and below the knee, n = 30, mean age at injury: 10.3 years) as control group. Serum S100B concentration was measured acutely. Four months after the injury, we assessed neuropsychological outcome and collected data about parent rated PCS.
Results: Preliminary results show that S100B serum concentration did not differ in children after mTBI and OI. Furthermore, four months after the injury, no group differences existed concerning the PCS rating or the neuropsychological performance. However, group specific Spearman correlations (controlled for age at injury) indicated significant relations between S100B concentration and cognitive PCS ($r = .55$, $p = .00$), verbal learning ($r = –.40$, $p = .01$) and verbal memory ($r = –.44$, $p = .01$) in children after mTBI, but not in children after OI. Furthermore, in both groups, no correlations were found between S100B serum concentration and somatic PCS.
Conclusions: Findings indicate that acutely measured S100B serum concentration was comparable between children after mTBI and OI. However, only in children after mTBI, elevated S100B was associated with more cognitive symptoms and a lower learning and memory performance, suggesting that S100B is an acute marker of degree of brain injury. Interestingly, no relations were found regarding S100B and somatic PCS, suggesting either a specific association to cognitive functions or being not sensitive enough to detect mild somatic problems after mTBI. We will further analyze the predictive ability of S100B on cognitive outcome to investigate whether S100B can be used as diagnostic tool in the emergency department.

P130

Four cases of sudden unexpected postnatal collapse in healthy newborn term Infants

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Introduction: Sudden unexpected postnatal collapse (SUPC) of apparently healthy term infants within a few hours of birth is rare, but can have serious consequences. High rates of mortality and neurological sequelae are reported in the literature. Incidence estimates vary in recent studies (2.6–5/100.000) and might be even higher than considered.
Methods: We report four cases of SUPC admitted to our intensive care unit over an 18-month period.
Results: The four term neonates (3 males, 1 female) were delivered vaginally, had excellent Apgar scores and normal umbilical artery pH. All pregnancies and births were uneventful and did not indicate the need of intensified postpartum observation. Three mothers were primiparous. The SUPCs occurred within the first 90 minutes after birth with all newborns found in a prone position on mothers breast or abdomen. One neonate needed brief bag/mask-ventilation with complete recovery. The other three required full resuscitation including chest-compressions. These infants were severely acidotic after the event and developed signs of hypoxic ischemic encephalopathy. One neonate underwent therapeutic hypothermia treatment. Further investigations were done to rule out underlying conditions, such as congenital cardiac anomalies, metabolic diseases, infection and cerebral pathology. One infant died, two survived with neurological impairments and one had no disabilities at discharge.
Conclusions: How to balance the need of postpartum observation of an apparently healthy newborn without negatively interfering with the obvious positive effects of mother-child bonding, is an important, yet, unanswered question. Prone positioning of the infant, absence of staff and primiparity have been recognized as possible risk factors for SUPC in previous studies. Although an extremely rare event, SUPC can be devastating and lead to death or severe neurological impairment. Teaching of health professionals and parents regarding SUPC seems to be important.

P131

Sexual disparity of copeptin plasma concentrations in newborn infants

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Background and aims: Arginine vasopressin (AVP), also known as anti-diuretic hormone, regulates water balance and blood pressure, and plays a role in social cognitive processes. Healthy adult men as compared to women have higher circulating level of AVP and of its surrogate marker copeptin, the C-terminal portion of the AVP precursor. We set out to investigate whether sexual disparity is already present at birth.
Methods: Plasma copeptin concentrations, determined by a CT-proAVP-luminescence-immunoassay, were measured at birth in venous umbilical cord blood of 399 infants born between 24 and 42 weeks of gestational age (GA).
Results: Multivariable linear regression analysis revealed vaginal delivery (regression coefficient β: 0.621), cord blood acidosis (β: 0.291), and male gender (β: 0.091) as significant and independent determinants of copeptin at birth (model R^2 0.624). In infants delivered vaginally copeptin was higher in boys than in girls (median 869 pmol/L (IQR 400–1428 pmol/L) vs 340 (107–975), respectively, $p < 0.05$). In infants delivered by elective C-section, that is without birth stress, copeptin was higher in boys than in girls (5.5 (4.4–10.2) vs 4.8 (3.6–5.8), respectively $p < 0.01$). When analyzing only healthy infants born by elective C-section after 34 weeks GA (n = 81), male gender was the only significant determinant of copeptin at birth (β: 0.243; R^2 0.145).
Conclusion: Sexual disparity of copeptin is already present at birth indicating increased activation of the AVP system in newborn boys as compared to girls.

P132

Severe anaphylactic-like reactions in two children following indigenous viper-bite

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Introduction: Two indigenous snake species in Switzerland are venomous, Vipera aspis and Vipera berus. Most cases of viper-bites show a mild clinical course, however, some patients may develop severe symptoms rapidly.
Cases:
Patient 1: A 15-year-old boy was bitten in the thumb. Within 15 minutes he developed shock with systolic pressure of 55 mm Hg, heart rate 112/', swelling of lips, tongue, and periorbital tissue. He was given norepinephrine (10 ug/kg i.v.), antivenin (Viperfav™, 1 vial), and steroids. Hypothermia 34.9 °C, centralisation, thrombopenia (80 G/l) and lymphangitis developed. He recovered without sequelae.
Patient 2: A 3-year-old girl was bitten in the left foot. She rapidly developed respiratory distress, periorbital swelling and shock with systolic pressure of 60 mm Hg, heart rate 93/', and generalised oedema. She required norepinephine (6,4 ug/kg i.v., followed by 0,1 ug/kg/h during 16 hours). With 1 vial of antivenin (Viperfav™) local swelling ameliorated, although a mild thrombopenia (118 G/l), leucocytosis and elevated D-dimers up to 2,36 mg/l developed. Both children had never been bitten by a viper before, there were no known allergies, but the boy had shown a slightly pronounced local reaction to hymenoptera.
Discussion: The occurrence of anaphylactic reactions is well known in patients with repeated viper bites and is IgE-mediated [1]. However, acute anaphylactic-like reactions are described in some first-bite victims [1]. The pathophysiology is unclear, but a direct autopharmacological effect of the venom with increased capillary permeability or a cross-reactivity with hymenoptera venom are discussed.
Conclusion: Vipera aspis and berus mainly cause local swelling, but occasionally an anaphylactic-like reaction can rapidly lead to life threatening systemic symptoms, also in first-bite victims.

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P133

Implication of the sympathetic nervous system in the development of hypoxia-induced pulmonary hypertension in rats

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Rational: While pulmonary hypertension (PH) is a multifactorial disease, the involvement of the autonomous nervous system is still under debate. By inhibiting the sympathetic pathway, we aim to demonstrate his potential role in the progress of hypoxia induced PH in rats.
Method: 30 males Sprague Dawley rats were assigned randomly to the following four groups. Control group received oral NaCl daily while they were exposed to air during 3 weeks. Animals in the other groups were exposed to hypoxia, (FiO_2 = 11%) during three weeks, and received orally daily either NaCl (Hypoxia Group), a sympathetic inhibitor, Carvedilol 15 mg/kg (Carvedilol Group), or Bosentan 100 mg/kg (Bosentan Group), considered as the positive control group. Echocardiography was performed at the beginning of the experiment and three weeks following exposure. The eccentricity index during diastole and systole of the left ventricle was performed. After three weeks, pulmonary pressure was measured via a pressure-volume catheter introduced in the right ventricle. The heart was removed and fixed for histological investigations. Right ventricular trophicity was estimated with thickness ratio defined as: left ventricle/ (septum + right ventricle). Data were analyzed with ANOVA and a multiple comparison test of Scheffe, or with paired t-test as appropriate. Results are expressed as mean ± SE, and significance is admitted with a p value <0.05.
Results: Both Hypoxia and Carvedilol groups exhibited PH with a ratio of pulmonary arterial pressure over systemic arterial pressure significantly higher than the control group (0.64 ± 0.07, 0.59 ± 0.5 vs 0.35 ± 0.2 respectively, p = 0.004 and p = 0.022). Bosentan, was able to blunt increase in the former ratio (0.38 ± 0.4, p = 0.007 and 0.041 vs group Hypoxia and Carvedilol respectively). Eccentricity index failed to detect difference between groups. However, Hypoxia Group increased this index during diastole between day 0 and day 21 (0.99 ± 0.1 vs 1.08 ± 0.3, p = 0.05). Finally, there was an increase in right ventricle thickness in the Hypoxia group as expressed by the decrease in ratio thickness (0.62 ± 0.01 vs 0.74 ± 0.02 in group Control, p = 0.021).
Conclusion: These preliminary results demonstrates that while Bosentan, was able to prevent the hemodynamic and histological changes due to hypoxia-induced PH, sympatholysis with Carvedilol failed to show a preventive role in pulmonary arterial pressure increase, but prevented right ventricular hypertrophia.

P134

Nebulised pentoxifylline for reducing the duration of oxygen supplementation in extremely preterm neonates – a randomised, double-blind, placebo-controlled trial

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Background: Chronic lung disease (CLD) is a significant complication in preterm neonates. Prevention of CLD with postnatal glucocorticosteroids may be associated with long-term neurodevelopmental impairment in this population. Limited poor quality evidence indicates that pentoxifylline, a non-steroidal immunomodulatory agent, may be beneficial in prevention of CLD.
Objectives: To evaluate the efficacy and safety of nebulised pentoxifylline (PTX) in reducing the duration of oxygen supplementation in extremely preterm (<28 w gestational age, EP) neonates at high risk of CLD.
Methods: 79 EP infants requiring endotracheal ventilation or ≥30% supplemental oxygen between 3 to 7 days of age were randomly allocated to nebulised PTX (10 mg/kg; n = 39) or an equal volume of identical placebo (normal saline; n = 40) for 10 days. Primary outcome was duration of oxygen supplementation at 40 weeks corrected age or discharge. Secondary outcomes included mortality, duration of ventilation, severity of CLD, need for postnatal glucocorticosteroids, and adverse effects.
Results: Median gestational age (25.0 vs. 25.0 weeks) and birth weight (728 vs. 720 g) were comparable between PTX and placebo groups. Median duration of oxygen supplementation [2160 vs. 2013 hours, adjusted hazard ratio: 0.65 (95% CI 0.45, 1.21), p = 0.173] was not significantly different between groups. Median duration of ventilation was significantly shorter [264 vs. 443 hours, adjusted hazard ratio: 0.53 (95% CI 0.26, 0.66), p = 0.017] in the PTX vs placebo group. There was no difference in other secondary outcomes. No adverse effects were noted.
Conclusions: The benefits of nebulised PTX in EP infants at risk of CLD need to be confirmed in definitive large trials.

P135

Mechanical ventilation increases expression of Insulin-Like Growth Factor Binding Protein 5 in renal cortex of preterm lambs

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Background: Preterm birth occurs at a time of ongoing nephrogenesis. Our group showed that mechanical ventilation (MV) leads to reduced surface density of glomerular capillaries compared to gestational references. Insulin-like growth factor binding protein 5 (IGF-BP5) has distinct spatial and temporal expression in glomerular mesangial cells of the developing kidney and therefore is a candidate protein for glomerular angiogenesis. We hypothesized that renal IGF-BP5 protein expression is altered by ventilation after preterm birth. Therefore, we compared the effects of MV versus non-invasive high-frequency nasal ventilation (HFNV) on IGF-BP5 protein abundance in renal cortex.
Methods: Preterm lambs were delivered at ~128d gestation (~28 wk human gestation), intubated and given surfactant and caffeine citrate. They were managed on MV or weaned to and managed on HFNV for 3d. A group of weaned preterm lambs (3d of MV, followed by 3d of HFNV) lived for 3 or 6mth (~2 and ~6yr, respectively, in humans). Unventilated fetal lambs delivered at ~128d gestation and age-matched term lambs at 3 and 6mth were references (n = 4/group). Renal cortex was analyzed by immunoblot for IGF-BP5. Statistical differences were tested by ANOVA and Fisher's PLSD.
Results: IGF-BP5 protein abundance was significantly greater in the renal cortex of ventilated preterm lambs compared to unventilated fetal reference lambs. IGF-BP5 protein abundance was significantly greater

in the MV group compared to the HFNV group. IGF-BP5 protein abundance remained higher in the renal cortex of former preterm lambs at 3 mth postnatal age compared to unventilated age references. At 6mth postnatal age, however, IGF-BP5 protein abundance was equal in former ventilated and reference lambs.
Conclusions: Our results indicate that IGF-BP5 protein expression is increased in the renal cortex of ventilated preterm lambs. The increase is greatest in preterm lambs supported by MV. Our results also show that the increase in IGF-BP5 protein abundance persists for at least 3 mth after preterm birth and MV for 3d. Upregulation of IGF-BP5 is associated with inhibited renal growth. Therefore, our findings provide new molecular insight into the pathogenesis of altered renal development in chronically ventilated preterm neonates.

P136

Look me in the eye, baby!

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Introduction: An ophthalmologic examination by the pediatrician is part of the routine examination of any newborn. Subconjunctival hemorrhages are very often found; retinoblastoma is uncommon but its early diagnosis is of utmost importance. Malformations like congenital cataract, colobomas or glaucoma should also be excluded. Seldom are iris cysts seen after birth.
Case: We describe two cases of newborn, in which routine eye examination after birth showed an irregularity of the inner border of the iris. Both patients were diagnosed with primary central pigment epithelial cysts of the iris, none had their central visual field affected, and no treatment was yet necessary.
Discussion: We discuss the different types of primary iris cysts. They can be stromal or epithelial. Cysts of the pigment epithelium represent 83% of primary iris cysts and can be classified as central (3%), midzonal (9–21%), peripheral (59–73%), or dislodged (3%). They may be bilateral and can be very small or big enough to affect central vision. Primary iris stromal cysts represent 16% of all childhood iris cysts. Most cysts have a benign clinical course and treatment is rarely necessary.
Conclusion: All pediatricians need to be comfortable performing the red reflex examination in a newborn, a powerful test to detect ophthalmologic abnormalities. Although iris cysts can easily be missed, exact identification of the type of cyst should be made to differentiate from intraocular malignancies.

P137

Ventilation decreases capillary surface area in renal outer cortex of preterm lambs beyond the neonatal age

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Background and objectives: Preterm birth happens at a time when the kidneys are developmentally immature and glomerular vascularization is ongoing. The normal development of capillary surface area in renal glomeruli and the impact of ventilation on their development are unknown. We hypothesized that ventilation after preterm birth reduces surface density of glomerular capillaries (SVgc). We compared SVgc in unventilated fetal and term reference lambs (for ontogeny), and 3 groups of preterm lambs: intubation and mechanical ventilation (MV) versus non-invasive high-frequency nasal ventilation (HFNV), and a group of weaned preterm lamb at 2mth of age after a consecutive course of MV and HFNV.
Methods: Reference fetal lambs were delivered at 128, 130, 133, 136 and 141d of gestation (term ~150 d) and immediately killed. Term reference lambs were either killed 1d after spontaneous delivery or lived for another 3 or 10 wk (n = 5/group). Preterm lambs were delivered at ~130 d gestation (~29 wk human gestation), intubated, and given surfactant and caffeine citrate. They were ventilated by MV for 3d or weaned to and managed on HFNV for 3d. Weaned lambs were ventilated by 3d of MV followed by 3d of HFNV and lived for additional 2 mth (equivalent to 2y in humans) after weaning. Kidney sections were immunostained with the endothelial surface marker CD31. Stereological techniques were used to calculate SVgc. Statistical differences were tested by ANOVA and Fisher's PLSD.
Results: SVgc was constant from 128d of gestation to term in lambs. At 3wk after birth at term, SVgc was significantly greater (ontogeny). Among preterm lambs, 3d of MV significantly reduced SVgc compared 3d of HFNV and compared to 133d fetal reference lambs. At 2 mth of age, SVgc remained significantly reduced compared to the age reference group.
Conclusions: Our results are novel, because they provide one of the first quantitative assessments of normal prenatal and postnatal development of SVgc. Our results also show that MV after preterm birth reduces SVgc, and that this negative effect persists long term after a consecutive course of MV and HFNV. Because SVgc represents filtration surface area, our findings suggest that MV of preterm neonates is associated with lower glomerular filtration capacity in the short and long term, which could lead to poor renal function later in life.

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To be or not to be a baby-friendly maternity: a prospective comparative cohort study

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Introduction: Many studies have shown the benefits of the Baby-friendly Hospital Initiative (BFHI) on duration of breastfeeding. We compared breastfeeding rates in two close and similar but distinct maternity wards, of which only one had the Baby-friendly Hospital (BFH) certification, to test the impact of the BFH guidelines on today's breastfeeding-favourable population.
Methods: A questionnaire about maternal socio-economic characteristics and breastfeeding practices during their hospital stay was proposed to all mothers delivering in the maternity wards of Pourtalès and La Chaux-de-Fonds corresponding to our inclusion criteria (term delivery, healthy mother and baby, French speaking) from 01.01.2010 to 19.03.2010. A comparison with a standardized BFH questionnaire filled by midwives for all mothers, including those non-participating in our study, allowed to detect any selection bias. Participating mothers were contacted at 2, 4 and 6 months postpartum to inquire on their breastfeeding practices and reasons for weaning.
Results: The overall participation rate was 32% (110/340 births) with 97% breastfeeding at discharge. While 68% of infants from the BFH and 56% from the non-BFH were fully breastfed at 2 months post-partum, this was the case for 43% respectively 41% at 4 months. At 6 months, only 50% respectively 47% of mothers still breastfed. Reasons given for weaning were apparently related to medical issues rather than resuming work.
Conclusions: The current high breastfeeding rates at discharge from maternities confirm the validity of the BFHI at large. We believe new measures have to focus now on further support of breastfeeding in the first months postpartum.

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Duplicity of events. And: the mother is always right!

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Introduction: Fever is a common complaint in children and the single most frequent reason for presenting to a pediatric emergency department. Viral infections are the major cause for febrile illness in young children. The challenge for emergency physicians is not to miss the serious bacterial infections, to look for the needle in the haystack.
Case report: A 7-month old boy was brought by his parents to the emergency department, the reason was high fever for 4 days and the mother's observation of favouring one arm. He appeared well in himself and the only findings were coryza, cough and a red throat. He was discharged home with the diagnosis of a viral infection. The family represented 4 hour later with concerns about the ongoing fever. The boy was kept in observation overnight by request of the worried parents. He left after an uneventful night, the fever had subsided. The following day he presented in poor general condition with a red, swollen arm and a temperature of 41 °C. Magnetic resonance showed an abscess of the extensor muscles and inmediate surgery followed with the diagnosis of necrotizing fasciitis.
Within less than a week another infant presented with fever and a painful leg. This time and with the previous case in mind investigations ensued, ultrasound showed a subcutaneous fluid collection and surgery confirmed the diagnosis of necrotizing fasciitis.
Conclusion: Duplicity of events and mothers knowing best are two well accepted concepts in medicine with absolute lack of evidence.

P140

Nicotine intoxication in an adolescent

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Introduction: Nicotine is one of the most readily available substances. Nicotine is rapidly absorbed through skin, alveoli, pharyngeal mucous membranes, and gastrointestinal mucosa.
Case report: A 15 y.o teenager came to the Emergency Department (ED) after nicotine intoxication. He first smoked a piece of cigar before drinking the remnant as a herbal infusion. Nausea, dizziness, headache, visual hallucinations and tinnitus appeared ten minutes later. He presented to the ED with dyspnea and chest pain one hour later. Physical examination was normal, except for tachycardia and high blood pressure. Due to the known risk for vasospasm, cardiac enzymes were checked and were within normal limits. He was monitored overnight and recovery was uneventful.
Discussion: Nicotine is a potentially lethal substance. As little as 10 mg of pure nicotine is a fatal dose in children, and 40–60 mg in adults. One cigarette contains 15–25 mg of nicotine but many brands of cigars contain at least 10 to 50 times higher amount. When smoked most of the nicotine is burned so the risk of acute intoxication for a smoker is unusual. Poisoning can occur through skin exposition (patches), excessive smoking (including water pipe), ingestion or a combination of them. Small doses lead to agitation and gastrointestinal hyperactivity. Large doses lead to central nervous depression, and cardiac failure. Death may occur within 5 min to 4 hours. Increased serum or urine level of nicotine or cotinine confirm nicotine overdose. Treatment modalities include activated charcoal and/or gastric lavage to reduce nicotine intestinal absorption; atropine sulfate to treat parasympathetic effect; and mecamylamine as specific antagonist.
Conclusion: Nicotine intoxication should not be underestimated. Aggressive treatment may be required to support respiratory function, protect the airway, and prevent cardiovascular collapse.

P141

Life-threatening subgaleal hematoma

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Introduction: Subgaleal hematoma (SGH) in newborns is caused by rupture of the emissary veins due to shearing forces to the scalp during delivery. The reported incidence is 4–6 cases in 1000 live births. Instrumental delivery, primiparity, prolonged second stage of labour and bleeding disorders are considered main risk factors. Ongoing, unrecognized bleeding is associated with hypovolemic shock, hence SGH can be devastating and potentially fatal. Mortality in severe SGH is high with reports up to 25% in case series.
Case: We report a term female infant with severe SGH, occipital skull fracture, cerebral contusion and intracerebellar hemorrhage due to a consumption coagulopathy.
Pregnancy was uneventful. Because of protracted delivery, vacuum extraction was deemed necessary, but proved difficult with multiple dislocations of the vacuum.
The infant required resuscitation with bag-mask ventilation after birth. In obvious hemorrhagic shock one hour after birth, the infant required emergency transfusion with 0 negative blood. Despite initial stabilisation, the condition worsened over the next few hours due to continued bleeding and consumption coagulopathy. Further packed red cells, fresh frozen plasma, platelets and clotting factors were necessary. At the age of 18 hours the infant developed generalized seizures requiring intubation and anticonvulsive therapy. A cerebral CT showed large intracerebellar and brain stem hemorrhage. The patient was extubated five days later. A hydrocephalus occlusus developed due to compression of the aqueduct, requiring insertion of a Rickham reservoir for regular cerebrospinal fluid taps. The patient was discharged at five weeks of age showing minor neurological impairment.
Conclusion: Cases of consumption coagulopathy after large SGH have been described in the literature. This might be seen as a first manifestation of inherited bleeding disorders. Close monitoring of infants after difficult instrumental births is important. Infants with large SGHs require intensive care admission with close monitoring of hematology and coagulation parameters.

P142

Vocal cord dysfunction in newborn – does it exist?

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Introduction: We present a newborn with similar clinical and endoscopic findings to classical vocal cord dysfunction (VCD).
Case report: A full-term newborn girl was investigated for repeated episodes of inspiratory stridor and marked cyanosis lasting a few minutes, beginning on the first day of life. Delivery was uneventful and clinical examination in asymptomatic periods was completely normal. Chest X ray and cranial ultrasound showed no abnormalities. Echocardiography revealed an atrial septum defect with otherwise normal cardiac anatomy and normal origin of the great vessels. On the second day of life flexible laryngotracheoscopy under local anaesthesia and spontaneous breathing showed normal bilateral vocal cord movements and no abnormality of the larynx region or the entire trachea. During visualisation of the larynx from the epipharyngeal region two episodes with acute severe inspiratory stridor with adducted vocal cords and only a small dorsal opening part were seen without evidence of increased laryngeal secretion or prior aspiration. Spontaneous resolution occurred until discharge on day 8. Clinical follow up during two months showed regular neuromuscular development without any respiratory or feeding problems.
Discussion: Paradoxical vocal cord movement (PVCM) in newborn without vocal cord palsy has been described by Omland et al 2008 in 4 cases. Our case seems to fit well into this observation. Pathophysiological explanation is difficult, laryngeal irritation or other stimuli including discomfort may be the cause.
Conclusion: Vocal cord dysfunction (or PVCM) seems to occur in neonates and is perhaps an underdiagnosed differential diagnosis of intermittent neonatal inspiratory stridor.

P143

Blueberry muffin and bronze baby syndrome – a rare presentation of congenital cytomegalovirus infection

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Introduction: Cytomegalovirus (CMV) infection is the most common congenital infection. 5–10% present clinical features at birth, such as microcephaly, affection of the central nervous system, chorioretinitis, hepatosplenomegaly, icterus and petechiae. We report a case of a neonate with congenital CMV infection presenting with many of its typical features and additionally rare blueberry muffin spots and bronze baby syndrome.
Case report: The male infant was born to a healthy 26-year-old by spontaneous vaginal delivery at 35 5/7 weeks due to premature contractions. First skin examination revealed petechiae due to severe thrombopenia and violaceous maculopapular lesions all over the body, known as blueberry muffin spots. Furthermore, microcephaly and a severe hepatosplenomegaly were observed. Due to icterus praecox intense phototherapy was initiated, during which he developed a bronze baby syndrome. This rare complication of phototherapy occurs when modified liver function is present, particularly cholestasis, which was apparent in our case and is known as one of the complications of CMV infection. Moreover he showed affection of the central nervous system such as cortical dysplasia, polymicrogyria, subependymal cysts and lesion of the white matter. An antiviral therapy with ganciclovir and valganciclovir was initiated, the severe thrombopenia required several thrombocytes-transfusions and cholestasis was treated with ursodeoxycholic acid and substitution of vitamins. He was discharged at the age of 6 weeks in a stable health condition.
Summary: This case illustrates how variable clinical manifestations of CMV infection can be. Differential diagnosis of blueberry muffin spots and bronze baby syndrome should always include infectious causes such as CMV.

P144

Dacryocystitis following a congenital dacryocystocele

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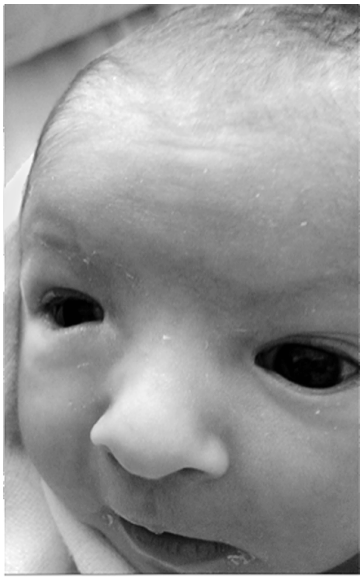
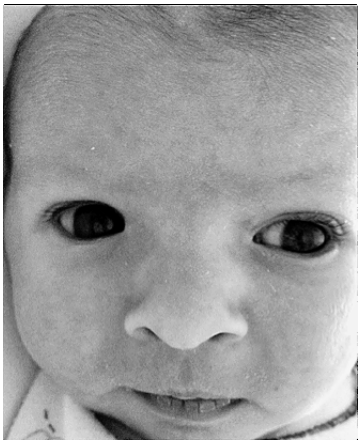
Introduction: Acute dacryocystitis may be a complication of dacryocystocele, a rare variant of nasolacrymal duct obstruction (NLDO).
Congenital dacryocystoceles may resolve with conservative measures, but when infected systemic antibiotic treatment is necessary, and most need surgical intervention. Referral in the early neonatal period may allow early intervention before complications occur.
Case report: We report a new case of congenital dacryocystocele diagnosed within the first day of birth, dacryocystitis occuring 15 days later. The infant girl was initially treated only by massage. At admission she presented erythema, swelling, warmth and tenderness of the lacrimal sac with purulent discharge. Blood CRP and CBC were normal.
The infant required intravenous antibiotic therapy, and a probing performed by an ophtalmologist with a good response.
Discussion: Dacryocystoceles are thougt to be a result of a combination of accumulation of mucus, amniotic fluid, tears, and bacterial colonization.
The appropriate timing, and management of congenital dacryocystoceles vary greatly in the ophthalmic and pediatric literature. The strategy varies from conservative treatment to early surgical management. Early surgical intervention is recommended by ophtalmologists in cases of respiratory compromise, dacryocystitis, large dacryocystoceles, recurrent dacryocystoceles and for cases in which conservative measures have failed.
Conclusion: Children with congenital dacryocystocele should be refered to an ophtalmologist before occuring of complications (dacryocystitis, cellulitis, meningitis or brain abcess, systemic infection…).
A conservative approach can be initialy attempted for non complicated dacryocystocele; probing can be performed if medical management fails.



Acute dacryocystitis in a 15-days-old infant girl. The photos are published with the informed consent of the parents.



Evolution after antibiotic therapy and probing.

P145

Playing with a tissues pack…

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Case report: 11 m.o. girl was playing with a tissues pack and ingested the little auto-adhesive plastic tape used to close the pack. She then refused to eat, had initial sialorrhea. First advice at emergency department was to observe stools and to wait, because symptoms had regressed. 5 days later, he developed fever, rough voice, rhinitis and persistant alimentary refusal. At intubation for endoscopy, the tape was found stuck in the lower hypopharynx.
Discussion: Foreign body ingestion/inhalation is frequent in childhood. Physical properties of ingested foreign bodies can present specific risks. Ingestion of more than one permanent is associated with a major risk of bowel perforation. Ingestion of button batteries can cause severe esophagal burns. Long or sharp objects can cause mechanical injuries. Vinyle gloves harden and develop sharp edges producing gastric bezoars. Some water-absorbing materials can expand and result in bowel obstruction.
The diagnosis was made after five days and four medical visits, despite the fact that the mother of the child witnessed and signaled the ingestion at the very beginning of the story. Adhesive and stiffness properties of tape and flexible plastic part, when ingested, can impact in unusual sites and produce episodic symptoms, thus delaying diagnosis.
Conclusion: Adhesive tape or flexible plastic pieces are not benign foreign bodies. They can impact in unusal sites. Even tiny pieces can produce severe symptoms, with possible misleading free intervals and significant complications.
This case emphasise the necessity of endoscopic investigations of persistent aerodigestive symptoms.

P146

Heroin intoxication of a 2-month old infant

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Case report: A 2 m.o. infant present with altered general status and feeding difficulties, no sign of infection, miosis or bradypnea. Due to parental history of drug addiction, urine toxic screening was made. Opiates were found. Specific investigations showed 6-mono-acetymorphin (6-MAM) proving heroin absorption.Investigation revealed that the infant had been entrusted by his mother to a drug addict friend who confessed having smoked heroin in presence of the baby.The poisoning of the child occurred by passive inhalation.
Discussion: Opioid intoxication in infants can occur in very different situations (erroneous administration of codeine at home, morphine overdose in hospital, accidental or intentional poisoning in drug addicts circles). History is often difficult, especially if parents give voluntarily illegal substances for sedative purposes or if they fear a justice referral. Clinical examination should be highly detailed and the classic triad: miosis, bradypnea and impaired consciousness should be researched carefully. Lack of recognition could lead to neurological sequelae or death. It is essential to carry out all the necessary tests to identify the cause of intoxication. Heroin is rapidly metabolized to 6-MAM (T1/2:6-25 min), which is metabolized into morphine (T1/2:3.2h). A positive opiate urine screening may indicate recent use (up to 12 h) of heroin, morphine or codeine. The presence of 6-MAM (chromatography coupled with mass spectrometry) confirms heroin use, its absence does not exclude it.
Conclusion: Substance abuse should be suspected in any child with parents suffering of addiction, even if specific signs may be incomplete or lacking. Specific toxicological tests must be done to identify causal substances.

P147

Evaluation of the application of the integrative management of childhood illnesses (IMCI): the fever example

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Introduction: In developed countries, the Integrated Management of Childhood Illnesses (IMCI) has reduced morbidity and mortality of common severe childhood illnesses. The objective of this study is to evaluate the application of IMCI adapted to our context for the management of fever.
Methods: We observed pediatric residents during consultation of febrile patients (2m-5yo, low severity: Australasian Triage Scale 4 to 5). We filled in a standardized evaluation chart and gave scores for every part of the consultation. Results of this first evaluation were presented to the residents and an IMCI-type formation was given. After the formation, a second evaluation was made and was finally compared to the first one.
Results: Fifty-four consultations, 26 for the first part and 28 for the second part were analyzed. The median age was 25.5 months (IQR 12–44 months) with no difference between groups. The most frequent diagnosis was upper airway viral infection (23/54 consultations). The performance before the formation wasn't optimal with a mean score of 65% (IC 95% 59–70) by consultation. After the formation, the score by consultation increases to 72% (IC 95% 65–78) (p = 0.05). We saw an improvement of almost all elements that had initially a score below 66%.
Conclusions: We developed an efficient tool for fever management evaluation adapted to our epidemiological context. IMCI application can improve the performance of young physicians. Our formation could however be enhanced. Other studies are necessary to prove the effectiveness of this measure on a longer period, in more severely ill patients and for other pediatric diseases.

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Facial trauma: don't forget nasal septal hematoma

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Case report: A previously healthy 5 y.o. boy presented, after minor facial trauma (simple fall), an epistaxis during 1–2 min. He was evaluated 3 times (12h, 2 and 5 days post trauma) because of progressive nasal obstruction, pain and febrile rhinorrhea. ENT advice 6 days post trauma confirmed the diagnosis of nasal septal hematoma (NSH) and indicated an immediate drainage. Silastic splints for 2 days were placed to prevent recurrence. Co-Amoxicillin was administrated for 10 days, followed by corticosteroid nasal spray for 1 month. Follow-up showed no deformity of the nasal pyramid, but a deviated septum associated with intermittent nasal obstruction.
Discussion: The nose is the most frequently injured facial structure in children. Rarely, NSH may occur even with minor nasal trauma. Early diagnosis and treatment are important to prevent complications. Child abuse should not be forgotten. The most common symptoms are nasal obstruction (95%), pain (50%), rhinorrhea (25%) and fever (25%) and appear within the first 24 to 72h. Inspection with nasal speculum reveals asymmetry of the septum with a bluish fluctuating mass or abnormal bulging of septal mucosa in the anterior part of the nasal cavity. Immediate hematoma drainage and antibiotics are indicated. In late diagnosed NSH, inevitable infection leads to abscess formation and necrosis of the septum. Complications may be meningitis, intracranial abscesses, orbital cellulitis and sinus thrombosis.
Conclusion: NSH must be considered in all children who have acute onset of nasal obstruction and history of nasal trauma. Clinical re-evaluation 48 to 72h after nasal trauma is mandatory to identify NSH, in order to minimize the risk of nasal deformity and to prevent septic complication.

P149

Case report: a rare cause of vomiting in a 14-day-old infant: congenital paraesophageal hernia, with gastric volvulus

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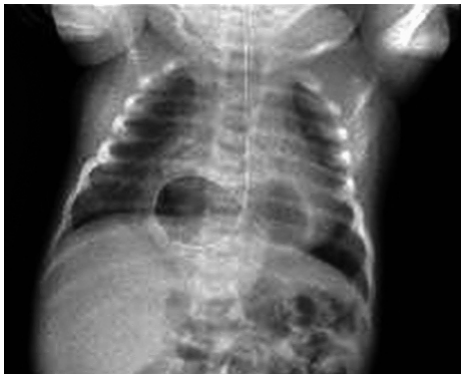
Background: Differential diagnosis for the vomiting infant is wide and include obstruction of the gastrointestinal tract due to congenital malformation. Delay in surgical management in case of volvulus may have dramatic consequence. We report a rare pathology: a congenital paraesophageal hernia.
Case report: A 14-day-old girl was admitted after 6 episodes of projectile non-bilious vomiting and weight stagnation since the 5th day of life. Her physical examination was unremarkable. She was full term born, without any complication. Owing to the persisting episodes of vomiting, the baby was fasted with naso-gastric aspiration. Two abdominal ultrasounds performed at 24h interval were described as normal. Upper gastrointestinal contrast series performed 36h after admission showed the ascent of the entire stomach into the chest through a large esophageal hiatus. An emergency surgical management was required because of gastric volvulus. No digestive necrosis was observed.
Discussion: Congenital Paraesophageal Hernia is uncommon in newborn. Anatomy is similar to hiatal hernia seen in adult with a muscular defect around the esophageal hiatus. Clinical presentation is nonspecific with pulmonary and/or gastrointestinal symptoms, but unlike congenital diaphragmatic hernia, in which there is a postero-lateral defect, there is no associated pulmonary hypoplasia described. Gastric volvulus is the most frequent complication and can be fatal.
Conclusion: Radiologic studies should be performed in newborn presenting with vomiting when there is a suspicion of mechanical occlusion. However, ultrasound is not a sensitive exam and upper gastrointestinal contrast series must be quickly performed if clinical suspicion is high.

Upper gastrointestinal series



Thorax X-ray

P150

Unexpected diagnoses in cyanotic newborns

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Introduction: Neonatal cyanosis is frequent and has a wide spectrum of differential diagnosis. We present two unusual cases of neonatal cyanosis caused by hemoglobin variants and propose a flow-sheet for the work-up of cyanosis in newborns.
Case Reports: Two term born babies were referred to our NICU in suspicion of a congenital heart defect because of a distinct cyanosis. Both children showed decreased preductal saturation values, non-responding to oxygen-support. A remarkable brown blood color was noted. Chest X-ray and echocardiography were normal. Given the physical finding of cyanosis without respiratory distress, a hematological work-up was initiated. O2-tonometry revealed decreased p50-values in both cases, compatible with a low-affinity-hemoglobinpathy.
In case 1, an aberrant band was detected by isoelectric focussing. Hemoglobin gene sequencing resulted in a heterozygous mutation of the alpha chain, known in literature as Hemoglobin type M Iwate, an extreme rare disease.
In case 2, the hemoglobin variant wasn't classifiable, but blood values normalized within the first three months of age. A HbF hemoglobinopathy is therefore presumed.
In both cases, transcutaneous and oximetric oxygen measurement were useless because of the aberrant and therefore falsely detected absorption spectra of the hemoglobin variants.
Currently both patients show healthy condition.
Conclusion: The cyanotic newborn is a challenge. Differential diagnoses range from harmless diseases to critical conditions. Cyanosis may also be caused by abnormal forms of hemoglobin. Although it is infrequent, an early detection is mandatory to prevent unnecessary investigation and delay in management.

P151

Snoring and face swelling: about the importance to look into the nose

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Introduction: We report the case of a 12 year old boy who was complaining during the last three weeks of progressive snoring and breathing difficulties at night. He never complained about pain, headache, nausea. The family noted a swelling of the right hemiface. He was treated for an allergic sinusitis the last ten days, a treatment introduced by general practitioner during holydays in France.
Case: The patient eventually visited the emergency room for impossibility to breathe through the nose. At that moment the patient presented with a striking face deformity, with an exophthalmia of the right eyes and nose deviation to the left. No nasal breathing possible. A complete systematic somatic status was done and was normal, except the nose inspection.
Nose inspection revealed a bulging blue-red mass, firm in consistency, without bleeding. An injected cerebro-facial CT scan revealed an extra-cranial facial expansive processus localized in the sinuses and choanes, sparing the skull base and its vessels. Oncological work up in the reference center identified an Ewing sarcoma of the face.
Conclusion: The signs and symptoms and the diagnosis delay participate to the striking presentation of this case. It underlines the importance of each part of the general paediatric status, and in particular to look carefully at the region of interest even if it represents a "zone of influence" of another medical speciality (NTE).

P152

Neonatal suppurative parotidis

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Introduction: Neonatal suppurative parotidis (NSP) is rare, in one report from Italy, the prevalence is 3.8/10 000 admissions.Only 32 cases of NSP have been described in the English language literature over the last 35 years, and the causative agent in most cases was Staphylococcus aureus.
Case report: We report a new case of NSP in a 7-day old infant girl. The diagnosis is made on the basis of parotid swelling, a purulent exudate from a Stenon duct, and the growth of staphlococcus aureus in pus from the orifice of Stenon duct and blood culture. The patient improved after 2 days of intravenous antibiotic therapy; the course is continued for 10 days.
Discussion: NSP affect newborns with risk factors, the healthy one can also be infected. The infection can result from hematogenous seeding or ascending infection from the oral cavity. The predominant organism is Staphylococcus aureus. The typical signs of NSP are parotid swelling, erythema, tenderness, and warmness; the presence of pus in the orifice of Stenon duct is pathognomonic of NSP. A rise of CRP and WBC with neutrophilic predominance are suggestive. The ultrasound examination show parotid swelling, cellulitis and even an intraparotid mass, including an abscess. Most patients respond to intravenous antibiotic therapy, failure to improve after 48 hours of appropriate antibiotic therapy should prompt repeat imaging with ultrasonography to identify abscess formation; incision and drainage is required infrequently and recurrence is uncommon.
Conclusion: NSP is very rare but must be managed quickly, surgery is rarely needed and the outcome is good.

P153

Cleft palate: isolated or syndromic?

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Introduction: Cleft palate can be isolated or associated with many syndromes, among which a 22q11 microdeletion. Other chromosomal anomalies can present with cleft palate.
Case report: Delivery was induced at 38 5/7 weeks because of growth retardation. Birth weight was 2030 g, length 44 cm and head circumference 31.2 cm (all parameters <P3). Cleft palate was diagnosed at day 2 and examination showed mild dysmorphic features. Cerebral and abdominal ultrasounds, as well as auditory evoked potentials were normal. Given the association of marked intrauterine growth retardation and cleft palate, a molecular karyotype (or array-Comparative Genomic Hybridization, aCGH) was performed and revealed a **de novo** 4p16.3 deletion together with an 8p23.3p23.1 duplication. At 4 months, in the context of fever, several absences with myoclonic jerks in both arms were noticed, compatible with seizures. EEG was diffusely slow and asymmetric, without an epileptic pattern. At 6 months, growth and psychomotor development were severely delayed.
Discussion: This infant has a double chromosomal anomaly associating a 4p16.3 deletion, typical of Wolf-Hirschhorn syndrome (WHS), together with a partial trisomy 8. Patients with WHS are small for gestational age, and malformations (heart, eye, brain) are very common. Evolution is marked by growth retardation, seizures, and by moderate to severe delayed cognitive development.
Conclusion: Cleft palate associated with growth retardation and/or other malformations should lead to a broader differential diagnosis, including other chromosomal anomalies than microdeletion 22q11. As the resolution of a standard karyotype is limited, it is best nowadays to perform an aCGH.

P154

Serious sequelae after ingestion of a lithium disc battery in a 20 months old girl

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Introduction: Disc battery ingestions in young children is becoming increasingly common and demand fast removal because severe tissue damage due to continuous hydrolysis can occur within few hours. Tracheo-oesophageal fistulae (TOF) are a recognized complication and can be life threatening.
Case: A 20 months old girl presented in a regional hospital with coughing and vomiting after disc battery ingestion of a large lithium cell (20 mm). The battery was located in the upper oesophageus but removal was initially hindered due to strong adherence to the mucosa already 3 hours after ingestion. After transferring the child to our hospital, the battery could be removed (delay of 7 hours after ingestion), showing massive inflammatory changes to the surrounding tissue. Oesophagoscopy on day 4 showed an oesophago-mediastinal fistula leading to a mediastinal emphysema and on day 10 a TOF as well as an increasing oedema and severe necrosis. The girl was transferred to a University children's hospital for further treatment: after 4 months of conservative management (parenteral nutrition) without effect, a surgical closure of the TOF was attempted but without success. At the time of writing this abstract, the child was still hospitalized.

Conclusion: When battery impaction in the oesophageus occurs, the time between ingestion and removal is critical: without fast removal within two hours, a high morbidity and mortality rate must be expected. Due to their high electrical current flow, outcomes are significantly worse for large-diameter lithium cells. Disc batteries should be preventively be stored out of reach from children.

P155

Early infant form of galactosialidosis – a case report

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Introduction: Galactosialdisosis is a rare lysosomal storage disease with defect of the protective protein cathepsin A, which leads to a deficiency of β-galactosidase and α-neuraminidase enzyme. Three forms of the autosomal-recessive disease are described: the early and late infantile form and the juvenile/adult form. The early infantile form is associated with foetal hydrops, ascites, oedema, hepatosplenomegaly, skeletal dysplasia, mental retardation and early death. We report a case of the early infantile form presenting all of these features.
Case description: During routine control in the 35th week of pregnancy of a 27-year-old healthy woman, hydrops fetalis of unknown origin was detected. Five days later the baby girl was delivered by emergency caesarean section. She presented with signs of generalized oedema, ascites and hepatomegaly. Echocardiografic findings showed dilated cardiomyopathy. Radiological investigations revealed dysostosis multiplex. Infectious, cardiopulmonary and haematological causes were excluded. Furthermore, diagnostic research on metabolic diseases suggested galactosialidosis, showing reduced enzyme activity of β-galactosidase and no activity of α-neuraminidase in cultured fibroblasts of skin biopsy. Symptomatic treatment was initiated including repeated abdominal punctures, substitution of albumin and diuretic therapy. At the age of five weeks the patient could be discharged in a stable condition. Regular follow-ups have taken place. Unfortunately, she died at the age of four months of unknown cause of death.
Summary: This case illustrates one of the few cases of the early infantile form of galactosialidosis that has been described worldwide. Unfortunately, prognosis is very poor, patients typically die of cardiac or renal failure. No curative therapy is known thus far. Further research on this topic is recommended.

P156

The influence of bronchopulmonary dysplasia and preterm birth on the morphology of sighs in newborn infants

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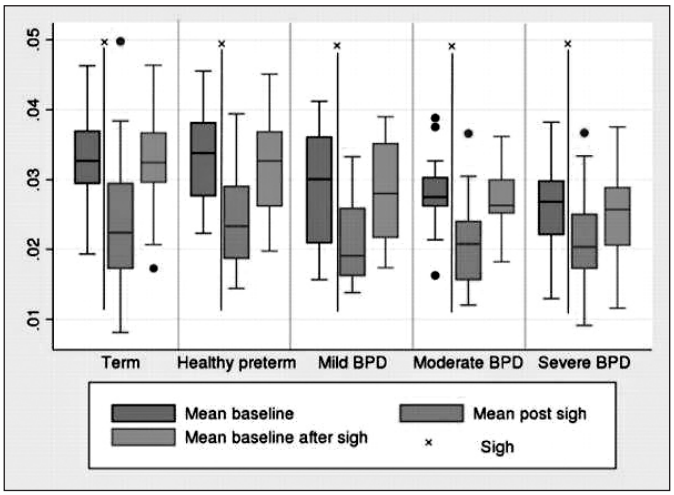
Introduction: Sighs play an important role in the plasto-elastic stretching of lung tissue and breathing muscles which results in improvement of elasticity, reduction of airway resistance and recruitment of lung capacity. To date it is not known how severe lung disease as bronchopulmonary dysplasia (BPD) is affecting the morphology of sighs.
Methods: We analysed tidal breathing measurements, performed in quiet, natural sleep at the (corrected) age of 4–6 weeks performed in the context of the BILD-cohort study.After automated sigh-detection amongst others the following parameters were analyzed: mean tidal volume at the beginning ("Mean baseline") and the end ("Mean baseline after sigh") of a measurement and just after a sigh ("Mean post sigh").
Results: In 86 term born infants (T), 23 preterm infants without BPD (PT-BPD) and 36 preterm infants with BPD (PT+BPD) a total of 244 sighs were examined. For demographic information of study participants see Tbl 1.

Table 1: Demographic data of study participants

N = 147 infants (62 female, 85 male)			
	Mean	**Min**	**Max**
Gestational age at birth (weeks)	35.39	24.14	41.86
Birthweight (g)	2505	430	4915
Weight at study date (g)	4239	2640	6800
Z-Score of weight at study date	–0.475	–3.769	3.94
Postconceptional age (weeks)	44.83	41.86	51.86

PT+BPD show a different breathing pattern in response to sighs compared to healthy infants (T, PT-BPD). Graph 1 shows the "Mean baseline", "Mean post sigh" and "Mean baseline after sigh" for the different BPD-groups.



Graph 1

Conclusion: Infants with severe BPD react significantly different to a sigh compared to healthy infants. The diminished change of the tidal volume upon a sigh might be due to their limited structural variability. As the status of BPD was more significantly associated with our findings as the gestational age at birth we assume the observed differences to be due to changes in lung development rather than to altered neuroregulation.

P157

Temperature regulation in preterm infants – a prospective observational study

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Background: Instability of body temperature is a major problem in preterm infants. Early quantification of the dynamics and complexity in body temperature may improve our understanding of autonomic temperature control in this population. We aimed to test the feasibility of characterising long-range correlations of body temperature in preterm/very low birth weight infants using the detrended fluctuation analysis (DFA).
Methods: We recorded 3h-time series of body temperature measurements in incubator-nursed preterm infants on day one of life. Body temperature was measured using a skin electrode attached to the trunk and positioned between infant and mattress. Time series of temperature data were extracted from the control unit of the incubator in 10-s intervals (Thermocare Vita, Weyer GmbH, Kürten, Germany). We quantified the strength of long-range correlations of body temperature by calculating the scaling factor alpha. Data were analysed using multivariable linear regression.
Results: We obtained valid measurements from 19/23 (83%) infants (male, n = 10; female, n = 9) with a mean (range) gestational age (GA) of 28.9 (24.0–37.4) weeks and a mean (range) birth weight of 1100 (510–1650) g. In univariable analysis, the scaling factor alpha was positively associated with GA (R^2 = 0.32, p = 0.011) and negatively associated with female sex (R^2 = 0.43, p = 0.002) and birth weight z-score (R^2 = 0.14, p = 0.122). A linear regression model including GA, sex and birth weight z-score predicted 72% of the variability in the scaling factor alpha.
Conclusions: Long-range correlations of body temperature can be quantified by DFA in incubator-nursed preterm infants on day one of life. Autonomic control of body temperature in these infants is strongly influenced by maturity, intrauterine growth and sex.

P158

Pulse oximetry screening in a paediatric emergency department to detect congenital heart disease in infants under 3 months of age

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Studies have shown that up to 24% of congenital heart disease (CHD) is detected after discharge from birth hospitalization and up to 8% of these present with cardiac decompensation. In the published literature, pulse oximetry screening for CHD is well described in in the neonatal setting but not beyond this period.
We examined the feasibility of pulse oximetry screening in a pediatric emergency department (PED).
Infants <3 months presenting to our PED for any reason between November 2012 and May 2013 were eligible for prospective pre- and postductal pulse oximetry screening. Exclusion criterion was known CHD. Primary outcome was the detection rate of CHD in the PED. A positive screen was defined as any oxygen saturation (SpO_2) <90%, SpO_2 <95% in both extremities or >3% absolute difference between the right hand and left foot on 3 occasions.
None of the 394 of 1451 eligible infants who underwent pulse oximetry screening had a positive test. 4 infants were identified as having CHD by clinical assessment (cardiac murmur and features of heart failure). Sensitivity was 0% and specificity 100%. The median total time taken for SpO_2 recording in a subgroup was 3 minutes 33 seconds (range, 50 seconds to 5 minutes).
Pulse oximetry screening for CHD in the PED is feasible, but we could not demonstrate that this is more effective than standard clinical assessment. Larger or multicentre studies are needed to examine the utility of pulse oximetry for screening for clinically undetectable CHD in the PED.

P159

Assessment of radiological investigations in hospitalized children under three months for the first urinary tract infection

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Introduction: The use of radiological studies in children with UTI is debatable. The guidelines (Recommandation SSP: 2008) used in Hospital de l'Enfance (HEL) recommend systematic renal ultrasound (RUS) and voiding cystourethrography (VCUG) for all children under 3 months old presenting with a first episode of urinary tract infection (UTI).
The objective of our study is to evaluate the performance of RUS and VCUG in detecting vesicoureteral reflux (VUR) in children under 3 months old admitted for a first episode of UTI.
Methods: We conducted a retrospective study between January 2011 and November 2013.
All children under 3 months of age who presented with a first episode of UTI admitted in our institution were included.
Results: We included 48 patients. The mean age was 52 days. Male to female ratio was 38/10. RUS was normal in 41(85%) and abnormal in 7(15%) of cases, 5(11%) with unilateral dilatation and 2(4%) with bilateral dilatation. VCUG was performed in 42/48 patients. VCUG was normal in 34 (81%) of cases. Prevalence of vesicoureteral reflux (VUR) was 1 (2.4%) for grade I, 2 (4.9%) for grade II, 2 (4.9%) for grade III, 2 (4.9%) for grade IV and 1 (2.4%) for grade V. The risk of high-grade VUR (IV-V) was increased when the RUS was abnormal (Odd Ratio = 13(1-179)).
Conclusion: The risk of RUS abnormalities and a high-grade VUR in children less than 3 months admitted for the first episode of UTI is low and similar to older Children.
Therefore systematic RUS and VCUG are also not recommended in this population.

P160

Chester step test to estimate maximal oxygen consumption in lean and obese children

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Introduction: Obese children have low cardiorespiratory fitness and exercise tolerance; therefore maximal exercise testing may be difficult to perform. The aim of this study was to elaborate a model to predict maximal oxygen consumption in lean and obese children using the sub-maximal Chester step test (ST).
Methods: We performed a maximal ST in 169 lean and obese children aged 11.5 ± 2.0 years (range: 7–16). The ST was performed as follows: 2 min stages in an increasing intensity (from 15 cycles.min^{-1}, 5 cycles. min^{-1} increments) to exhaustion. Oxygen consumption (VO_2) was measured by indirect calorimetry. Statistical Tobit model was performed in order to predict VO_2 from age, sex, body mass index (BMI) z-score and intensity levels. Estimated VO_2max was then determined from the heart rate (HR) – VO_2 relationship extrapolated to HR max (220 beats.min^{-1} minus age).
Results: VO_2 can be predicted from the following equation: VO_2 = 22.82 – 0.68*BMI z-score – 0.46*Age (years) – 0.93*sex (male = 0; female = 1) + 4.07*intensity level (stage 1, 2, 3 etc.) – 0.24*BMI z-score *intensity level – 0.34*sex*intensity level. VO_2 was lower in subjects with high BMI z-scores and in female subjects. When extrapolated to similar age and BMI, in adults, the values calculated from our equation correspond to those reported in the literature.
Conclusions: The ST can be proposed to test cardiorespiratory fitness in lean and obese children and VO_2max can be estimated from the equation developed in the present study in a maximal or sub-maximal test.

P161

Safety of a cluster regimen for subcoutaneous house dust mite immunotherapy in children

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Introduction: Allergenspecific immunotherapy (AIT) is the only therapeutic treatment for allergic respiratory diseases with well documented clinical efficacy. House dust mite (HDM) subcutaneous immunotherapy (SCIT) shows with a high evidence that asthma symptom and medication scores improve. SCIT usually requires a titration phase over several weeks to months. In cluster SCIT 2–3 injections per treatment day are applied resulting in an accelerated achievement of the maintenance dose.
The aim of this study was to investigate the safety of a cluster protocol in children.
Methods: The medical records of 21 HDM-allergic children/ adolescents (age 7,2 to 18,5 years, median 11,2 y) from 2006 to 2013 were analysed retrospectively. All patients suffered from allergic asthma, except for one (95,2%) and received a standardized depot extract adsorbed to calcium phosphate (Phostal D.pter/D.far 50/50%, Stallergènes) according to a 2-week titration cluster by the subcutaneous route (table 1).
Results: All patients completed the cluster-desensitization and reached the full cumulative dose. 2 patients experienced systemic adverse reactions, one mild (Rhinitis) and one moderate (Urticaria, mild dyspnea). Most patients had local swellings of the upper arms, but with a diameter ≤5 cm.
Conclusion: The cluster protocol for induction of a HDM-AIT is a safe and well tolerated procedure in children. In comparison with data from the literature on conventional and cluster protocols in adults, the incidence of adverse reactions is not higher, neither in asthmatic children. The short duration of the cluster protocol is more convenient for the patients and results in a better compliance and less costs.

Table 1: Administration schedule HDM-cluster.

Cluster day	IR/ml	Dose, ml
1	1,0	0,1 0,3 0,6
2	10,0	0,1 0,2
3	10,0	0,4 0,8
time between treatment days:	1 week	
time between injections:	30–60 minutes	
		adapted from Pfaar O et al. Int Arch Allergy Immunol 2009

P162

Neurodevelopmental long-term outcome in children after hemolytic uremic syndrome

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Background: Haemolytic uremic syndrome (HUS) is a multiorgan and life-threatening disease, leading to acute renal injury, and may result in long-term renal and extrarenal sequelae. Data on neuromotor outcome are scarce and limited to information on impaired fine motor skills in children with history of HUS and severe central nervous system (CNS) involvement.
Material and methods: A single-center retrospective cohort of 47 children was examined at a median age of 10.6 years (6–16.9) and a median follow-up of 7.8 years (0.4–15.3) after HUS. Intellectual and neuromotor performance were assessed with the German version of the **Wechsler Intelligence Scale 4th version** and the Zurich Neuromotor Assessment, respectively. The occurrence of neurological symptoms during the acute phase of HUS was evaluated retrospectively.
Results: Mean IQ was within the normal range (median full-scale IQ 104, 54–127). Neuromotor performance was significantly poorer in the domains "adaptive fine", "gross motor", "static balance" (all p <0.05) and "associated movements" (p <0.001). Only the "pure motor" domain was within normal range. Neurological symptoms occurred in 16/47 patients (34%) during the acute episode of HUS. Neurodevelopmental outcome was not significantly different between children with versus without CNS involvement.
Conclusion: Follow-up of children after HUS showed a favourable cognitive outcome. Neuromotor outcome, however, was impaired. Neurological impairment during the acute episode of HUS was not predictive of outcome. Long-term cognitive and neuromotor examination of children with a history of HUS might be important for early detection of motor

P163

Ignoring syncope during exercise could cost young patients their life

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Introduction: We present 3 cases of young athletes presenting with syncope during exercise and potentially life-threatening cardiac anomalies.
Case presentations: A 13-year-old competitive soccer player presented with recurrent chest pain and syncope on heavy exertion. Exercise test reproduced the symptoms and showed monomorphic ventricular tachycardia from the right ventricular outflow tract (RVOT) after 18 minutes on the treadmill. ECG, echocardiogram, Holter and cardiac MRI were normal. An electrophysiologic study confirmed a RVOT tachycardia, which was successfully ablated.
A 15-year-old competitive soccer player sustained sudden syncope while playing a match. He previously had been complaining of faintness during training. ECG, echocardiogram and exercise test were normal. Coronary CT-scan showed left coronary coming from the right coronary sinus and passing between aorta and pulmonary artery with compression between the 2 vessels during exercise. He underwent successful left coronary reimplantation.
A 12-year-old competitive basketball player complained of chest pain, pallor and sweating followed by syncope at the end of a match. He was pulseless for several seconds then bradycardic. He had previously experienced similar although less severe episodes during training. Echocardiogram, ECG and exercise test were normal. Coronary CT-Scan showed left coronary from the right sinus and intramural trajectory between the great vessels. His left coronary was successfully unroofed.
Conclusions: Young athletes with chest pain or syncope on exertion should always be referred to a paediatric cardiologist. Although rare, cardiac causes may be associated with sudden cardiac death, which makes accurate diagnosis essential for appropriate management and counselling.

P164

The "squeeze and wheeze" sign: a useful clinical sign in lung sounds analysis of children with acute cough

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Introduction: Cough is a very frequent symptom in children. Young children are often difficult to examine. They can breathe superficially and expiratory wheeze can thus easily be overlooked.
Objective: To report a new clinical respiratory sign and to measure its prevalence in young children cared for in a general paediatrics practice.
Methods: In a prospective case series analysis, all children aged 6 months to 6 years with acute cough as main symptom (patients) or without any symptom or chronic disease (controls) were studied. Lung sounds were analysed during classical chest examination and during "squeeze and wheeze" maneuvers (SWM, see video) where the patients' chest is manually compressed antero-posteriorly during expiration. The prevalence of expiratory wheeze during classical lung auscultation and the SWM was analysed.
Results: 124 children were examined, 68 healthy controls and 56 with acute cough. SWM was feasible in 108/124 (87%) children. None of the control children wheezed either during classical auscultation or during the 62/68 feasible SWM. Four of the 56 children with cough wheezed during classical auscultation, 46/ 56 SMW were feasible and 8/52 (15%) coughing children without wheeze during classical lung examination were positive during the SWM.
Conclusions: A maneuver to improve lung sounds study in young children is described. In children with acute cough, it can reveal expiratory wheeze that could be overlooked by classical chest auscultation. Inter-observer reproducibility and the clinical significance of this easily performed maneuver warrants further study.

P165

Urokinase: the magic bullet for pneumonia with pleural effusion?

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Background: Although pneumonia with pleural effusion is a well-known disease, little consensus for its specific management is found. In 2010, according to international guidelines, we introduced in our management the use of urokinase in cases drainage was needed. No other changes in treating this group of patients was made. The aim of this retrospective chart analysis was to evaluate the benefit of urokinase in this group of children with regard to morbidity.
Methods: We retrospectively analyzed data of 68 children (66% male) hospitalized with pneumonia with pleural effusion, all either receiving a pleural drainage or VATS (video assisted thoracoscopy), over a period of 11 years. We emphasized in this analysis the duration of hospitalization and duration of oxygen supply comparing the group before (n = 39) and after (n = 29) introduction of urokinase therapy.
Results: Children had a median (range) age of 4.1 (0.6–16) years. Duration of hospitalization in the urokinase group was significantly shorter compared to children treated without urokinase (12 [9–16] vs. 15 [14–19] days, p = 0.002). There was no significant difference with regard to duration of oxygen requirement (1.0 [0–7.5] vs. 0.5 [0–5.2], p = ns). We observed possibly complications due to our interventions in 3 patients (4.4%).
Conclusion: Children treated with urokinase as add-on therapy on either pleural drainage or VATS stayed on average 3 days shorter in the hospital compared to those not receiving urokinase. However, it did not affect duration of oxygen supply. Of course, further analyses are needed to exclude bias in patient selection.

P166

Two cases of late diagnosis of coarctation of the aorta

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Background: Since many years the Swiss Society of Pediatrics recommends screening of blood pressure starting at the age of 6 years. In special circumstances such as heart or renal disease, obesity, intrauterine growth retardation or prematurity and in case of symptoms, earlier and more frequent blood pressure measurements are warranted. Coarctation of the aorta is estimated with an incidence of 1 in 2500 live births. Typically it is diagnosed neonatally or during infancy but there are significant numbers of late diagnosis up into adult life. Survival is significantly affected by the age of operation and therefore early repair is advocated.
Case 1: In a 13 years old male adolescent high blood pressure values were discovered after an episode of syncope. The patient reports a good physical performance but less endurance compared to his friends. At presentation he was hypertensive with a remarkable systolic difference between the upper and lower extremities. Physical examination revealed a systolic murmur and abdominal bruit in the right upper quadrant and pulses in the dorsalis pedis artery were present. Renal ultrasound showed symmetrically *flattened* arterial pulse curve and by echocardiography the diagnosis of a severe coarctation of the aorta was made. CT angiography showed an almost interrupted aortic arch witch multiple well developed collaterals and the patient underwent end-to-end anastomosis. Postoperative course was uncomplicated and is currently treated with Metoprolol.
Case 2: A 9 year old girl presents because of pathological blood pressure screening. She did not report about decreased tolerance of physical activity. Physical examination showed absent inguinal pulses and a systolic murmur. Echocardiography confirmed the diagnosis of coarctation of the aorta. She underwent aortoplasty with a pericardial patch and 4 years later is currently on chronic antihypertensive treatment with candesartan and lecarnidipine.
Conclusion: Late detection of coarctation of the aorta leading to late surgical repair has been shown to negatively influences survival of these patients. In our opinion it remains crucial to screen children for hypertension according to the Swiss Society of Pediatrics recommendation in order to facilitate early diagnosis and therapy with the aim to avoid long term complications since hypertension in childhood may remain asymptomatic and is mostly secondary to an underlying condition

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P167

The association between the head circumference and the central venous pressure in children undergoing the Fontan procedure

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Introduction: High central venous pressure (CVP) may cause communicating hydrocephalus and macrocephaly. In children with single-ventricle physiology the brain is exposed to high venous pressure after completion of bidirectional cavopulmonary anastomosis (BDCPA) when CVP equals the pulmonary arterial pressure (PAP). We sought to asses if high PAP leads to an augmentation of head circumference (HC).
Methods: Seventy-four children with single-ventricle physiology were included in a retrospective analysis. CVP and PAP were measured during the routine catheterization before BDCPA and Fontan completion as well as the occipital-frontal circumference which was compared with longitudinal age dependent percentiles in normal children measured in our institution.
Results: Median age at BDCPA and Fontan were 4.8 (1.6–12) and 27.9 (7–40.6) months respectively. HC was more than 1 SD below 50th percentile at the time of BDCPA and showed significant growth between pre-BDCPA- and pre-Fontan catheterization (7(0–100) vs. 36 (3–56)th percentile, **p** <0.001), while PAP decreased significantly (14.8 ± 2.7 vs. 13.2 ± 2.1 mm Hg, **p** = 0.012). There was no correlation between PAP and HC pre-Fontan (R^2 = 0.001). Children with lower differences in CVP pre-BDCPA and PAP pre-Fontan showed increased growth of HC.
Conclusions: Moderately elevated CVP in children with single-ventricle physiology does not lead to macrocephaly. Between BDCPA and Fontan, HC increased significantly and achieved values close to norm for age. The lack of direct correlation between PAP before Fontan and HC may be explained by a catch-up growth of HC in patients with better pulmonary vascular bed. Further studies with focus on high PAP are needed to exclude or prove a direct correlation.

P168

Arterial stiffness in children with inflammatory bowel disease

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Background: Altered arterial stiffness is a recognized risk factor of poor cardiovascular health. Chronic inflammation may increase arterial stiffness and patients with inflammatory bowel disease (IBD) are at increased risk for cardiovascular events.
We tested the hypothesis that arterial stiffness is increased in children with IBD, a chronic disease characterized by intestinal and systemic inflammation.
Methods: Arterial stiffness, expressed as carotid-femoral pulse wave velocity (PWV_{cf}), was measured in 25 children with IBD: 11 girls, 15 with ulcerative colitis (UC) and 10 with Crohn's disease (CD); median (range) age 14.1 (6–18) years and median duration of disease 2.8 years. PWV_{cf} in children with IBD was compared to 50 healthy controls matched for age and gender, and was correlated with clinical and laboratory markers of inflammation.
Results: PWV_{cf} was similar in children with IBD compared to controls: PWV_{cf} median (interquartile range) was 4.3 (4.0–5.2) vs. 4.6 (4.1–5.2) m/s, p not significant. None of the children with IBD showed pathologically increased PWV_{cf} (>95. centile). In univariate or multivariate correlation analysis, PWV_{cf} was not associated with markers of inflammation (Calprotectin, BSR, disease activity using the PUCAI Score for UC and PCDAI Score for CD), cardiovascular parameters (blood pressure), disease duration and drug therapy in children with IBD.
Conclusions: Arterial stiffness, measured as PWV_{cf}, is not increased in children with IBD.
These results do not confirm the data of a similar study performed in adults showing increased arterial stiffness in patients with IBD independently of other cardiovascular risk factors (J Hypertens 2012;30:1775–1781). A possible explanation for this discrepancy may be the longer disease duration in adults. Further studies are needed to elucidate the effect of chronic intestinal inflammation on the cardiovascular system in children.

P169

Fatal cardiac ischemia in hemolytic uremic syndrome

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Background: The hemolytic uremic syndrome (HUS) is clinically characterized by the triad of hemolytic anemia, thrombocytopenia and renal failure. In >90% of cases, the disease is triggered by an infection with Shiga toxin-producing Escherichia coli (STEC) harboring specific Shiga toxin and often presents with prodromal bloody diarrhea (STEC-HUS). Cardiovascular dysfunction in patients with HUS may be related to secondary issues such as volume overload, hypertension or electrolyte disturbances including hyperkalemia. Furthermore, primary myocardial involvement has been increasingly recognized as a potential comorbid feature of HUS.
Methods: Description of a case with a fatal cardiac complication during the early phase of HUS.
Case presentation: In a 4.5 years old boy the diagnosis of HUS was made based on bloody diarrhea and the classical HUS triad (hemoglobin 48 g/l, platelets 41 G/l, creatinine 494 µmol/l). The child remained anuric despite controlled fluid resuscitation, blood pressure and plasma electrolytes were within the normal range. Troponin T was considerably increased (0.462, norm <0.014). Suddenly he developed clinical signs of poor myocardial function (congested jugular veins, arterial hypotonia). Despite inotropic agents and application of resuscitation algorithms, he developed electro-mechanical dissociation and ventricular tachycardia/fibrillation, which did not respond to defibrillation. Death occurred after 90 minutes of cardiopulmonary resuscitation. Necropsy findings showed in addition to the typical signs of thrombotic microangiopathy in the kidneys, also myocardial and pulmonary involvement with diffuse thrombotic microangiopathy and related ischemia.
Conclusions: Previous reports of primary cardiac involvement with HUS have included thrombotic microangiopathy of the coronary vasculature resulting in myocardial ischemia, myocardial infarction or depressed myocardial function, myocarditis, congestive heart failure with dilated cardiomyopathy and pericardial effusion with tamponade. Awareness of cardiac involvement in HUS is warranted and the routine initial evaluation of myocardial function and ischemia is indicated in all cases of HUS.

P170

Electrolyte abnormalities in cystic fibrosis: systematic review of the literature

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Introduction: We recently made the diagnosis of cystic fibrosis in an infant with normal neonatal screening, cough, hyponatremia, hypokalemia and hyperebicarbonatemia. Cystic fibrosis per se can sometimes tend to hyponatremia (≤134 mmol/L), hypokalemia (≤3.4 mmol/L), hyperbicarbonatemia (≥27 mmol/L) or hypochloremia (≤100 mmol/L). This tendency was first documented 60 years ago and subsequently confirmed in case reports and small case series that were mostly retrospective.
Methods: We reviewed the literature using the principles established by the UK Economic and Social Research Council guidance on the conduct of narrative synthesis and on the Preferred Reporting Items for Systematic Reviews statement.
Results: The reports included 172 subacute and 90 chronic cases with a ♂:♀ -ratio of 1.57. Electrolyte abnormalities were associated with clinically inapparent fluid volume depletion, mainly affected patients ≤2.5 years of age, tended to recur and often were found before the diagnosis of cystic fibrosis was established. Subacute presentation included history of heat exposure, vomiting, excessive sweating and pulmonary infection. History of chronic presentation, in contrast, was often inconspicuous. The tendency to hypokalemia, hyperbicarbonatemia and hypochloremia was similar between subacute and chronic cases, with hyponatremia being more pronounced (P <0.02) in subacute rather than in chronic presentation. Subacute cases were repaired parenterally, chronic ones instead were usually managed with oral supplementation. Retention of urea and creatinine was documented in 38% of the subacute cases.
Conclusions: We wish to warn physicians to be aware of the fact that these electrolyte abnormalities occur both as a presenting and as a recurring feature of cystic fibrosis.

P171

Severe hyponatremic encephalopathy secondary to desmopressin treatment for enuresis: a systematic review

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Background: Dilutional hyponatremia is a serious adverse effect of desmopressin, a vasopressin analog that is widely prescribed to manage monosymptomatic enuresis. The presentation of hyponatremia, largely related to cerebral dysfunction, can include severe signs like altered mental status and seizures.
Methods: We reviewed the literature dealing with altered mental status or seizures in enuretic subjects on desmopressin. For this purpose, we used the principles underlying the UK Economic and Social Research Council guidance on the conduct of narrative synthesis and the Preferred Reporting Items for Systematic Reviews and Meta-Analyses statement.
Results: We found 54 cases (ranging in age from 2.0 to 37, median 9.0 years; male to female ratio: 1.71) of severe hyponatremia (<135 mmol/L) secondary to desmopressin treatment presenting with altered mental status or seizures. In most cases the complication developed 14 days or less after starting desmopressin. An intranasal formulation had been used in 47 patients. Excess fluid intake was documented as a contributing factor in at least 22 cases. In 6 cases severe signs of hyponatremia developed in the context of intercurrent illnesses.
Conclusion: Altered mental status or seizures are very rare but recognized complications of desmopressin in enuresis. This complication mostly develops in subjects managed with the intranasal formulation 14 days or less after starting the medication, following excess fluid intake and during intercurrent illnesses.

P172

Hyperchloremic metabolic acidosis induced by the iron chelator deferasirox (Exjade®): a case report and review of the literature

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Background: Deferasirox (Exjade®) is a new treatment of iron overload that is administered orally once-a-day, resulting in better acceptance among patients. We describe the case of a girl who developed renal tubular dysfunction on treatment with deferasirox and review the corresponding literature
Case summary: A 17-year-old female adolescent with b-thalassaemia on deferasirox 30 mg/kg daily since 6 months presented with isolated hyperchloremic metabolic acidosis (bicarbonate 12.9 mmol/L, sodium 137 mmol/L, chloride 111 mmol/L, potassium 3.6 mmol). Creatinine, urea, uric acid, total calcium, inorganic phosphorous and urinalysis were normal. Acidosis resolved after withdrawing deferasirox. The Naranjo adverse drug reactions probability scale indicated that a relationship between deferasirox and hyperchloremic metabolic acidosis is likely.
Review of the literature: Eight cases of metabolic acidosis have been reported in patients treated with deferasirox. In most cases, acidosis was associated with further features of proximal renal tubular dysfunction (such as hypophosphatemia, hypokalemia, hypouricemia, glucosuria in the face of a normal blood glucose level, generalized hyperaminoaciduria and mild proteinuria). In 3 further cases signs of renal tubular dysfunction were noted that were associated with a normal acid-base balance.
Conclusion: We describe herein a case of metabolic acidosis in the setting of treatment with the deferasirox. Our case and the literature indicate a potential risk of kidney toxicity on this agent.

P173

Skeeter syndrome, a frightening but benign mosquito bite reaction

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Introduction: Skeeter syndrome is a uncommon reaction due to an often not identified mosquito bite. Symptoms can be disturbing for parents as for the medical staff, and often lead to unnecessary additional investigations or treatment.
Case report: We report the case of an healthy 8 years old child, who consulted because of a skin lesion following a suspected insect bite on the forearm. He described a localized redness within 24 hours after the bite, with subsequent increase of the redness with induration and blisters. Because cellulitis was suspected, a treatment with Coamoxicilline was started. One day later, blood tests including CRP, procalcitonin, and CBC were normal.
The child repeatedly visited outpatient emergency due to clinical worsening characterized by progression of lesions and erythema of the forearm, with pain during mobilization. He remained afebrile. In presence of repeated negative blood tests, we were able to exclude inflammation or infection, and antibiotics were finally stopped.
Evolution was slowly favorable. Two weeks later, IgE and IgE i71 (mosquitos) were negativ.
Discussion: The clinical history and the follow-up suggested the diagnosis of Skeeter Syndrome, a late-onset reaction which might be associated with IgE and IgG, usually appearing within hours following a mosquito bite. Lesions are itchy, sometimes with blisters, bleeding or central necrosis. An erythema and induration around the lesion might be present.
The clinical course is generally spontaneously favorable in about 3 to 10 days.
Conclusion: Skeeter Syndrome may be difficult to differentiate from a bacterial infection. Diagnosis is based on a precise history (time to onset of lesions). Costly and unnecessary investigations might result from ignoring existence of this condition.

P174

Primary hyperparathyroidism due to parathyroid carcinoma

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Background: Primary hyperparathyroidism in childhood is a rare disease with severe hypercalcaemia. A parathyroid adenoma is the most often underlying cause.
Casereport: 14 years old boy with pain in knees, heels and back, loss of weight, fatigue, polyuria and polydipsia 6 months prior first examination. He presented with reduced general condition, hypertension (140/70 mm Hg), hyperreflexia, pain of both tuberositas tibiae, fingers and the upper lumbal spine. Family history revealed primary hyperparathyroidism with a documented genetic mutation (**HP-JT/HRPT-2** mutation) on mother's pedigree (mother, uncle and grandaunt, grandaunt's daughter and grandson).
Laboratory investigations: Total calcium 3.7 mmol/l (normal 2.1–2.7), Ca^{++} 2.1 mmol/l (normal 1.1–1.3), phosphate 0.8 mmol/l (normal 1.1–1.7) and magnesium 0.6 mmol/l (normal 0.7–1.0). Parathormone (PTH) 845 pg/ml (normal 10–55), creatinine 98 µmol/l (normal 54-121), estimated GFR (calculated by Schwartz formula) 70 ml/min/1.73 m², uric acid 417 µmol/l (normal 11-353). Hypercalciuria (calcium/creatinine ratio of 0.9 mol/mol, normal <0.7). Bone lesions in the distal radius and ulna, and in the middle phalanx of several fingers were shown on x-ray. Ultrasound demonstrated hyperechogenic kidneys without nephrocalcinosis. Electro- and echocardiography were normal. MIBI-Single photon emission computed tomography (SPECT) and ultrasound revealed one hyperechogenic, highly vascularised lesion (3x2x2 cm) in the left lower pole of the thyreoidea suspecting a parathyroid adenoma.
Treatment consisted surgical removal of the suspected parathyroid tumor. As there was no significant decrease of PTH, further exploration was leading to a second, larger, ectopically jugular located tumor, histologically identified as a parathyroid carcinoma. Thirty minutes after removal of the second tumor, PTH decreased from 1000 to 150 pg/ml. High substitution of calcium, magnesium, phosphate and vitamin D was applied postoperatively and tapered, but not stopped thereafter. First examination after the patient's discharge demonstrated rapid improvement of general condition, normal GFR, normal blood pressure and normal values of calcium, phosphate and PTH. Re-evaluation will be due if PTH and/or calcium will rise again.
Conclusion: Primary hyperparathyroidism is very rare in childhood. Appropriate treatment consists of removing suspected adenomas including a histological examination of the tissue in order to identify a possible carcinoma.

P175

Streptococcus pneumonia associated haemolytic and uremic syndrome with cholestasis

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Introduction: Streptococcus pneumonia associated haemolytic uremic syndrome (SpHUS) is uncommonly associated with conjugated hyperbilirubinemia. Only four cases of conjugated hyperbilirubinemia in SpHUS are reported in literature: two of them showed good evolution. We report the case of a 2½ years-old girl diagnosed with SpHUS associated with severe jaundice.
Case report: A 2 ½ years-old girl, vaccinated with Prevenar13®, hospitalized for bilateral pneumonia, presented anuria and jaundice one day after. An extensive workup revealed anemia (84 g/l) with schistocytes, thrombopenia (35 G/l) and renal insufficiency (creatinine value: 182 µmol/l) associated with increased liver enzymes and severe conjugated hyperbilirubinemia (368 µmol/l). Thoracic and abdominal computed tomography showed bilateral pneumonia with necrotizing pneumonia in left lobes, normal liver appearance with peri-portal oedema without any sign of cholestasis but with stoneless increased size gallbladder. Blood culture exposed serotype 3-*Streptococcus Pneumonia*. Peritoneal dialysis was started; however two days later, she developed peritonitis associated with abdominal cellulitis due to *Pseudomonas Aeruginosa* and *Enterococcus Faecium* requiring antibiotics therapy and catheter removal after 6 days. Cholestasis was treated by ursodeoxycholic acid. Patient evolution was rapidly favourable with normalization of creatinine value after 10 days, resolution of thrombocytopenia and anaemia after 11 and 12 days respectively, and normalisation of liver function tests after 13 days. She was discharged from hospital after 3½ weeks with a normal clinical exam.
Discussion and conclusion: Severe conjugated hyperbilirubinemia may be observed in SpHUS with a variable disease prognosis. Mechanism and physiopathology of cholestasis seemed to be due to a diminished bile flow secondary to an increased vascular permeability and a plasma extravasation induced by the inflammatory syndrome. The inefficient excretion of bilirubin associated with the important hemolysis led to hepatocellular injury. We hereby added another critical case with a very good outcome.

P176

Dilated cardiomyopathy in a patient with propionic acidemia: Why we should think of and look for the rare

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Introduction: Although rare, dilated cardiomyopathy (DCM) is the most common cause for cardiac transplantation in children. Possible causes include myocarditis, neuromuscular, metabolic and other diseases. Yet, in 66% of patients with DCM aetiology remains unexplained. We report on a patient presenting with DCM who was diagnosed with propionic acidemia, a potentially reversible cause of DCM.
Case report: A 16 years old boy with Hispanic background presented with a one week history of fatigue and breathlessness. His parents are consanguineous; further family history is unavailable. The medical history included alpha-thalassemia minor and ADHD treated with methylphenidate. Somatic growth was restricted below the third percentile. On physical examination the patient displayed tachycardia, a precordial heave and a soft systolic murmur. The liver was enlarged. Echocardiography showed normal anatomy but a markedly dilated left ventricle with severely impaired function (ejection fraction 17%) and significant mitral regurgitation. Metabolic screening revealed elevated propionylcarnitine in a dried blood spot and secretion of 3-OH-propionate and methylcitrate in urine, typical findings in propionic acidemia. Enzymatic tests in leucocytes confirmed the diagnosis. He was started on carnitine supplementation therapy and protein reduced diet.
Conclusions: Patients with DCM need to undergo a broad diagnostic evaluation including a metabolic screening – even beyond the early childhood period. The clinical picture and the echocardiography findings in DCM are unspecific. To clarify aetiology, a thorough history including family history and a specific laboratory testing algorithm are required. Although unusual, cardiomyopathy can be the first sign of propionic acidemia in previously healthy adolescents.

P177

Renal tubulopathies: rare patients, typical patterns

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Introduction: Renal tubulopathies are rare genetic diseases. Clinical presentation is highly variable whereas blood and urine tests often show specific patterns. Genetic testing allows final diagnosis. We present 4 cases with inborn dysfunction along the renal tubule.
Case 1: 3-year old boy. Initial findings: Pneumonia with glycosuria and proteinuria. Uneventful family history. Further tests: Normal plasma creatinine, tubular proteinuria, intermittent hypercalciuria and nephrocalcinosis. Genetic analysis revealed a novel nonsense mutation in the X-linked *CLCN5*-gene (c.1885C>T;p.Q629*;p.Gln629Ter) in the proximal tubule, confirming Dent's disease.
Case 2: Preterm female with growth retardation and severe polyuria. Consanguineous parents. Further tests: Hypokalaemic, hypochloraemic metabolic alkalosis with transient prerenal failure, hypercalciuria and nephrocalcinosis. Genetic analysis showed a homozygous mutation in the *SLC12A1*-gene (c.1685C< T;Ala562Val) of the loop of Henle, consistent with Bartter's syndrome type 1.
Case 3: 2-year old boy with 2 episodes of urolithiasis (100% calciumoxalate-dihydrate). Parents were first cousins. Further tests: Normal plasma creatinine, hypomagnesaemia, mildly elevated uric acid and parathyroid hormone, hypercalciuria and normal ultrasound. Genetic analysis revealed a novel homozygous mutation in the *CLDN16*-gene (c.316T;p.S106P) confirming a tight-junction dysfunction in the loop of Henle.
Case 4: 7-month old boy: Incidental finding of repeated hypokalaemia. Uneventful family history. Further evaluation: Metabolic alkalosis, hypomagnesaemia, hypocalciuria and normal ultrasound, all findings consistent with Gitelman's syndrome in the distal tubule. Genetic analysis is pending.
Conclusion: Diagnostic algorithm in renal tubulopathies includes precise history, clinical examination, renal ultrasound and targeted analysis of blood/urine metabolites. Specific patterns lead to a clinical hypothesis which can be confirmed by genetic analysis.

P178

Epstein Barr virus – associated smooth muscle tumor after pediatric heart transplantation – a complication of immunosuppressive therapy

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Introduction: Epstein Barr Virus-associated smooth muscle tumors (EBV SMT) in immunocompromised patients were first reported in the early 1990s. In children with AIDS, EBV SMT is at present the second most common type of neoplasm. We report the case of an epipharyngeal EBV SMT in a patient with immunosuppressive therapy after heart transplantation. The patient also suffered from 2 episodes of post transplantation lymphoproliferative disease (PTLD).
Methods: Case report
Results: This female patient underwent heart transplantation for dilatative cardiomyopathy (donor EBV +, recipient EBV –) at the age of 4 years. She manifested an EBV-associated PTLD of small intestine, retropharynx and glottis 7 months postoperatively which was successfully treated with rituximab. Follow up MRI at the age of 10 years revealed an epipharyngeal mass while the girl was asymptomatic under immunosuppressive treatment with mycophenolate mofetil, everolimus, cyclosporine and prednisolone. Repeated MRI showed progression of epipharyngeal mass with infiltration of circumferential musculature. Transnasal biopsy revealed EBV SMT. Subsequently the tumor was completely resected. The postoperative clinical course was uneventful. During follow up after one year PTLD recurred in the small intestine and parapharyngeal. Following repeated treatment with rituximab the patient recovered completely and MRI shows complete regression of PTLD 10 months after the last dose of rituximab. Significant reduction of immunosuppressive therapy from first to second manifestation of PTLD was not possible because of repeated subclinical rejections on endomyocardial biopsy.
Conclusions: The pathogenesis of EBV-SMT is related to the infection and neoplastic transformation of smooth muscle cells by EBV with clonal expansion occurring only in the immunocompromised host. Solid organ transplant recipients and those with EBV donor+ / recipient- constellation in particular may develop EBV-associated neoplasm under immunosuppressive treatment. The head and neck region is a preferential site of EBV SMT. Chemotherapy may slow disease progression, surgical resection and improving the host immune status are considered as first therapeutic options. It is important to differentiate EBV SMT, requiring surgery, from PTLD, requiring treatment with B-cell antibodies.

P179

2d-strain echocardiography: a sensitive marker for graft rejection in pediatric heart transplantation?

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Introduction: Heart transplantation is an established procedure in pediatric patients with terminal heart failure. Graft rejection is the major cause of morbidity and mortality. Strain echocardiography (myocardial deformation imaging) has been reported to be superior to conventional echocardiography for detection of early myocardial dysfunction and could be a promising technique in graft rejection.

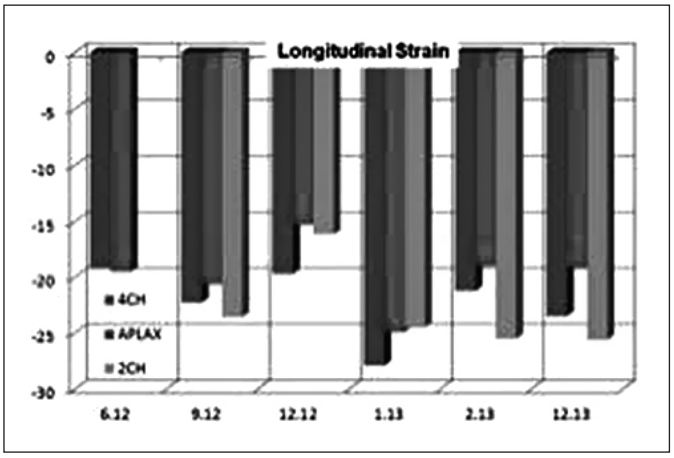

Figure 1
Longitudinal Strain.

Patient and methods: A 13 year old heart transplanted female was seen for a routine control two years after transplantation. Under immunosuppression (Tacrolimus, Everolimus, Prednisone) the course was uneventful, without signs of graft rejection. Echocardiography including ejection fraction and myocardial thickness and 2d-strain was performed, followed by endomyocardial biopsy.
Results: Conventional echocardiography was normal. 2d-strain parameters were typical for impaired left ventricular function: longitudinal 2d-strain values were decreased and radial strain showed increased thickening (fig. 1 and 2, 12.12). This finding was in line with the graft rejection proven in the biopsy (1R in ISHLT 2004). After adaptation of immunosuppressive medication, the degree of graft rejection in the control biopsy was reduced and the 2d-strain was normalized (fig. 1 and 2, 1.13).
Conclusion: This case shows the potential of strain echocardiography in early detection of left ventricular dysfunction in acute graft rejection. Hence, it might help to reduce the numbers of endomyocardial biopsies. Further studies are needed to establish this promising technique in transplanted patients.

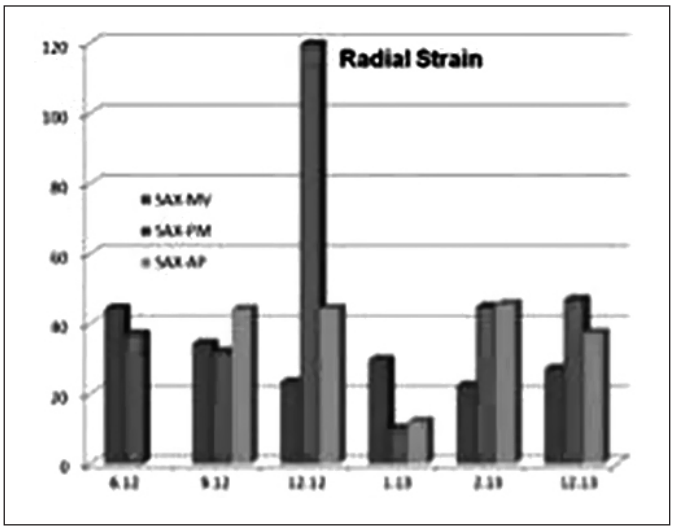

Figure 2
Radial Strain.

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Strenuous exercise and elevation of cardiac biomarkers

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Background: Current guidelines for the identification of an acute myocardial distress depend on the presence of cardiac troponin as a highly sensitive marker of cardiac damage. However, a number of studies have reported elevated cardiac troponin levels in healthy subjects after endurance exercise.
Methods: Description of a case of thoracic pain and dyspnea associated with an increase of troponin levels after a strenuous exercise in a adolescent.
Case presentation: We introduce a case report of a 16 years old male subject, with chest pain and dyspnea after a soccer game. Suspecting an ischemic event, we dosed the cardiac enzymes that were slightly elevated (Troponin I 0,41 µg/L). We made an ECG, an echocardiography and blood tests that were normal. In the course the subject didn't show any symptom and the cardiac enzymes gradually decreased. Therefore we suspect a likely functional symptomatology with a troponin increase due to the great effort.
Conclusions: A strenuous exercises can cause an elevation of cardiac biomarkers. The mechanism and significance of this observation however has not been fully elucidated. This increase could be partially attributed to cardiac stress: intensive exercise can cause an acute volume overload, with transient reductions of ventricular ejection fraction and elevations of cardiac biomarkers, all of which generally return to normal quickly.

P181

Stimulation of umbilical cord blood dendritic cells by prenatal exposure to particulate matter

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Introduction: It is known that the late prenatal period as well as the early infancy represents a critical window of immune vulnerability. It has been shown that prenatal exposure to air pollution affects lymphocyte subpopulations in umbilical cord blood.
However the effects of prenatal exposure to particulate matter <10 µm (PM_{10}) on cord blood dendritic cells of healthy neonates are unknown.
Methods: In a subset of a birth cohort of unselected healthy neonates (BILD), we performed complete blood counts (n = 284) and determined myeloid dendritic cells (mDC) and plasmacytoid dendritic cells (pDC) using a FACScan (n = 246) of umbilical cord blood. The daily values of PM_{10} exposure were obtained from a background monitoring station. The mean of PM_{10} was then calculated for the last trimester of pregnancy and for the last 30, 14 and 7 days before delivery. The association between PM_{10} and umbilical cord blood cells was assessed using a multivariable linear regression analysis, adjusted for possible confounders (sex, gestational weight, gestational age, mode of delivery, stress factors during delivery, prenatal smoking exposure, season of birth, maternal atopy).
Results: PM_{10} exposure during all considered time intervals was significantly associated with increased levels of mDC and pDC after adjustment for confounders. The mDC/pDC ratio was significantly higher with increasing PM_{10} exposure during the last trimester of pregnancy and the last 30 days before delivery. No significant association was found between absolute leukocyte counts and all considered exposure time periods.
Conclusions: The present results suggest that prenatal exposure to moderate levels of PM_{10} may influence fetal immune development leading to an increase of cord blood dendritic cells, particularly mDC's, which are known to be involved in the development of allergic disease and asthma.

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Influenzavirus B-associated acute benign myalgia cruris: an outbreak report and systematic review of the literature

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Introduction: Acute benign myalgia cruris is characterized by transient bilateral calf pain that leads to difficulty walking. An outbreak of influenzavirus B-associated myalgia cruris was observed during the seasonal influenza outbreak observed in Switzerland from week 1 to 13 of 2013.
Methods: We performed a prospective case finding among the Swiss-Italian pediatric emergency units and pediatricians. A systematic review of the literature was also performed.
Results: The diagnosis of myalgia cruris was made in 49 Swiss-Italian children aged 3.0–14 years (♂ : ♀ =1.7) and in 2 of their parents. Flu-like symptoms were resolving when bilateral calf pain began, which remitted over ≤7 days. The creatine kinase-level, assessed in 28 patients, was elevated in 25. Nose swabs were positive for influenzavirus B in 13 out of 14 cases. The blood cell count, measured in 41 cases, disclosed leucopenia in 12 and thrombocytopenia in 3. The review of the literature found 10 outbreaks of ≥10 cases of influenzavirus B-associated myalgia cruris, which included a total of 203 patients with a mean age of 7.3 years (and a ♂ : ♀ = 2.0).
Conclusions: Influenzavirus B caused a large Swiss-Italian outbreak of myalgia cruris. Epidemic influenza virus B-associated myalgia cruris affects preschool- and school-aged children, primarily boys. In characteristic cases with bilateral calf pain, preserved reflexes and sensory function, a detailed medical history, a careful physical examination and blood tests including cell count, C-reactive protein (or sedimentation rate) and creatine kinase are all that is needed for diagnosis.

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Newborn screening for severe combined immunodeficiency (SCID) – retrospective analysis of positive cases and proposed pilot screening project

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Introduction: Severe combined immunodeficiency (SCID) and severe T cell deficiency fulfil criteria for newborn screening (NBS) since these diseases are asymptomatic at birth and might be fatal within the first year of life, the confirmation of the disease is easy (numeration of lymphocyte subsets), and early hematopoietic stem cell transplantation (HSCT) is a curative treatment. Quantification of TRECs (T-cell receptor excision circles) from dried blood spots (DBS) is a sensitive and specific screening test for SCID and severe T cell deficiency. TRECs are a reliable marker of the number of circulating naïve T cells recently emigrated from the thymus and are undetectable or very low in infants with SCID or severe T cell deficiency.
Methods: In a retrospective study we have tested the EnLite™ Neonatal TREC kit from Perkin Elmer to determine the TRECs in the original DBS of babies with confirmed SCID.
Results: TREC copy numbers were measured from a 1.5 mm DBS of 6 patients with confirmed SCID; 0.8, 0.0, 0–9; and 47 controls; 136, 118, 27–329; mean, median, range, respectively.
Conclusion: The TREC assay we tested is a reliable assay, easily to be implemented into NBS programs. NBS for SCID and severe T cell deficiency is already recommended in the US and a few other countries. Since early HSCT before the occurrence of irreversible organ damage can provide cure for these patients, a proposal to the Swiss Health Ministry (BAG) regarding inclusion in the routine NBS screening program in Switzerland is underway.

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Intestinal parasitic infections among refugee children in Geneva

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Background: Intestinal parasites infections are endemic in developing countries and cause significant morbidity. Epidemiologic data are necessary to improve the screening strategy for refugee children in Switzerland.
Materials and methods: We retrospectively analyzed the results of a single microscopic stool examinations (formol-ether concentration technique) in recently immigrated children followed at the Children's Hospital of Geneva from January 2002 to December 2011.
Results: Stool exams of 615 pediatric refugees aged between 6 months to 16 years were evaluated. 161/615 (26.2%) children had ≥1 positive intestinal parasite (including *Blastocystis hominis*). The prevalence was higher in children between 3 and 5 years of age (49/138; 35.5%). African children were the most affected with a prevalence of 39.7% (77/194).
The most common parasite was *Blastocystis hominis* (76/161; 47.2%), followed by *Giardia lamblia* (61/161; 37.9%) and *Trichuris trichiura* (24/161; 14.9%). African children had a higher prevalence of both protozoa and helminths than children from other regions ($p < 0.05$). The prevalence of multiple pathogens in children was 20.5% (33/161).
Conclusion: We found a high prevalence of intestinal parasites among refugee children in Geneva. Considering the lack of sensitivity of one single stool exam for parasites, the prevalence is likely to be underestimated. A screening strategy based on demographic characteristics and origin could be developed.

P185

Cephalhaematoma and elevated inflammatory markers – is it a sign of infection? Case report and review of the literature

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Introduction: Cephalhaematoma is usually a benign condition which resolves spontaneously within weeks. Nevertheless there is a risk of primary or secondary infection and diagnosis is not easy. We tried to identify risk factors and clinical criteria and to outline appropriate methods for investigating cephalhaematomas for possible infection.
Methods: We present two illustrative cases of suspected infected cephalhaematomas. A systematic literature review is added correlating clinical presentation, laboratory results, therapies and outcomes of previously reported cases of infected cephalhaematomas to our two cases.
Results: An infant with a large cephalhaematoma after vacuum-extraction presented at the age of five weeks with fever and elevated inflammatory makers, which persisted under antibiotic therapy. Only on the second presentation E.coli was isolated from the haematoma and evacuation was performed. The second infant presented with secondary enlargement of cephalhaematoma and markedly elevated inflammatory markers at the age of seven weeks. There was a spontaneous resolution of the haematoma.
In the 47 cases of infected cephalhaematomas reviewed secondary enlargement was present in 60%, erythema in 53%, tenderness 36%, fluctulance in 34% and skin erosion in 21%. Sixty-two percent of the infants presented with fever. E.coli was isolated from 66% of the haematomas. The mean time of antibiotic treatment was 27 days (range 7–67). Only three infants healed without surgical intervention.
Conclusions: Diagnosis of infected cephalhematoma remains challenging. Neither clinical nor inflammatory markers are definitive. Imaging has limited power in differencing liquefaction versus abscess formation. Infection should be suspected if there is secondary enlargement of the haematoma, erythema, fluctulance, skin erosion or signs of systemic infection. Although aspiration is not recommended because of the risk of complications, a diagnostic tap needs to be performed in such situations.

P186

A boy with a swollen foot, large lymphnodes and little pain

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Introduction: We report on an eight-year-old boy from Eritrea who presented with a large swelling of the right hindfoot, that had started four months ago. He was limping, but despite the large swelling he had only little pain while walking. The swelling was slightly tender, only faintly red and warm to touch. Functional testing of the ancle joints did not show significant deficits. However, a remarkable lymphadenopathy was found on clinical examination, with large, indolent lymphnodes in the right inguinal and supraclavicular region. In contrast to these impressive local findings, the patient appeared generally in good health. There was no history of weight loss or night sweats. The boy was afebrile and had been in school every day. Two years earlier, upon arrival in Switzerland, an abdominal Schistosomiasis had been diagnosed and treated with Praziquantel. Other routine health check-ups upon arrival in Switzerland had been normal.

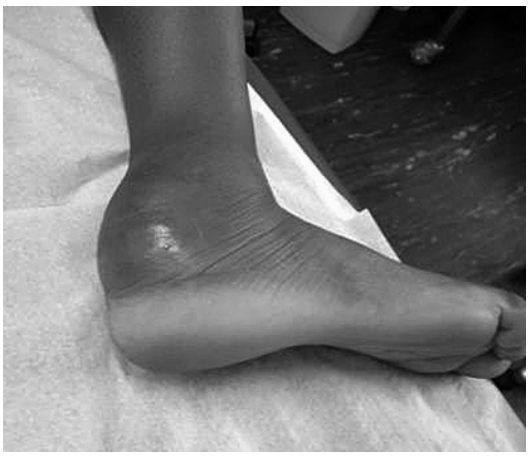

Figure 1
Photograph of the right foot upon presen.

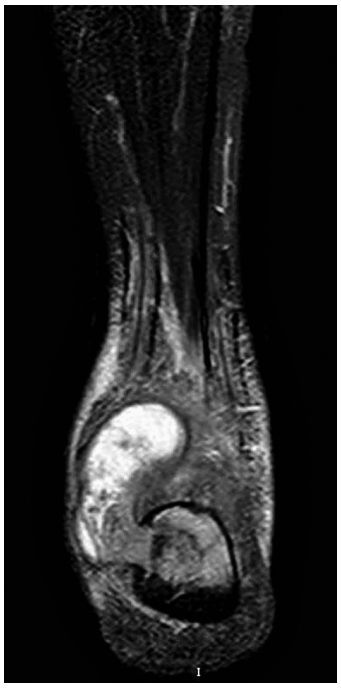

Figure 2
Magnetic resonance imaging with bone des.

Methods: X-ray- and MRI-scans of the right hindfoot revealed a large abscess of the calcaneus penetrating into the surrounding tissue. A bone biopsy was taken from the pathologic process in the calcaneus and investigations including histology, microbiology and molecular biology were performed. Additional investigations included chest-x-ray, routine laboratory testing and TB-spot.
Results: Radiologic evaluations revealed a pathologic leason in the calcaneus with cortical destruction and an abscess in the interspace between the Achilles- and Peroneus tendon.
Histological analysis of the biopsy exhibited ephiteloid-giant-cell-granuloma; microscopically, acid-fast bacili were found. Molecularbiologically, Mycobacterium-tuberculosis-complex-DNA with full sensibility to common tuberculostatica was detected. On chest X-ray mediastinal lymph node enlargement was visible, but we were not able to demonstrate a distinct primary tuberculosis lesion. Therapy included local surgical treatment and a tuberculostatic regimen was initially started with Isoniazid, Rifampicin, Pyrazinamid and Ethambutol and modified later. Eight months later the boy is doing well without significant limitation on walking.
Conclusion: This 8-year old patient from Eritrea had tuberculous osteomyelitis of the right calcaneus with large bony destruction and abscess formation, impressive lymphadenopathy and remarkably little symptoms.

P187

Chickenpox: sometimes not harmless

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Introduction: Arterial ischemic stroke (AIS) in childhood is rare and correlates with high morbidity and mortality. The causes of stroke in children are different than in adults, consisting in multiple risk factors (infections, trauma, arteriopathies, coagulopathies, metabolic diseases) for vascular pathology. Varicella zoster virus (VZV) may cause cerebral vasculitis with stroke after several months or even years after acute infection.
Case report: A former healthy seven year old boy woke up one morning with an unusual headache. After napping the mother noticed slurred speech and weakness of his right arm. Therefore, 4 hours later, they consulted our emergency department. No previous history of trauma or important illnesses was noted. PedNiHSScore was 7 points (dysarthria, right-sided facial nerve paralysis and right-sided sensomotoric hemiparesis). An acute stroke was suspected. Emergency Computed tomography scan revealed signs for an ischemic stroke in the left striatum. MR-Angiography confirmed the diagnosis and showed a vasculitis of the left middle cerebral artery. VZV Polymerase Chain Reaction (PCR) in cerebrospinal fluid (CSF) was positive. Therefore, we started treatment with acyclovir as well as high dose steroid therapy according to the vasculitis protocol (BrainWorks, sickkids.ca) as well as aspirin. Six months later he shows mild weakness of his right arm and problems with attention and concentration.

Discussion: Here, we present a case of varicella vasculitis leading to stroke 1.5 years after acute chickenpox with a positive PCR in CSF. Therefore, causative treatment with acyclovir and steroids were performed with a positive outcome. In children with acute focal neurological deficit AIS must be considered and diagnostic tests performed immediately. Thrombolysis has to be considered even though controlled studies in children are still missing and evidence is based on case reports. There is agreement between professionals in Switzerland on antithrombotic treatment with aspirin except in extracranial dissection or cardiac embolism where heparin is the treatment of choice.
Conclusion: VZV is known to be prone to affect cerebral vessels via direct affection even months or years after acute infection as shown by our case. Vaccination may help to prevent this possible devastating complication.

P188

Fever in children unvaccined, don't forget epiglottitis

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Introduction: Incidence of epiglottitis has decreased with introduction of routine *Haemophilus influenzae B (HiB*) vaccination. We report a case with an atypical presentation, the child presenting with vomiting.
Case report: A 4.5 m.o. unvaccinated boy was admitted with repeated vomiting, crying and high fever for a few hours. At arrival, he was tachycardic (220/min), tachypneic (52/min), with episodic respiratory pauses of few seconds without stridor, suggesting sepsis. Non segmented neutrophiles (15%), segmented neutrophiles (38.5%), and procalcitonin (1.35 mcg/l) were observed on biological exploration. Lumbar puncture, chest radiography and urinanalysis were normal. Fluid resuscitation and i.v. ceftriaxon were given. While performing abdominal ultrasound for persisting vomiting, he suddenly worsened with respiratory distress including chest indrawing, desaturation, and hemoptysis. Suspecting epiglottitis, oxygen, adrenalin aerosols and corticosteroids were given prior to intubation, which confirmed the diagnosis 4 hours after admission. Blood culture was positive for *HiB*.
Discussion: Despite dramatical decrease of epiglottitis incidence with vaccination, cases are still encountered. Usual symptoms consist of respiratory distress with soft respiratory stridor, minimal cough, drooling and fever. Management includes antibiotics, corticosteroids, hemodynamic support, minimal handling and rapid intubation by anaesthesiologist and otolaryngologist.
Because of epiglottitis decreased incidence, this diagnosis is less frequently evoked. In this case, lack of usual signs at admission contributed to diagnosis challenge, waste of time, and unnecessary or dangerous additional diagnostic tests.
Conclusion: Diagnosis of epiglottitis should not be forgotten in children with fever and respiratory distress, especially in unvaccinated infant. Routine vaccination remains the best protection.

P189

Antibiotic treatment of pertussis – is a 7 day period really sufficient?

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International guidelines recommend 7 days of antibiotic treatment for pertussis. This is based on a few studies which implicated that 5 and 7 days of treatment with azithromycin or clarithromycin are sufficient to eradicate *Bordetella pertussis* (Bp) from the nasopharynx when compared to 14 days of erythromycin. However, eradication was tested only by bacterial culture on the day of last treatment. At such an early time after ceasing treatment, remaining antibiotic levels in the host may inhibit growth of bacteria which may still be viable and transmittable despite negative culture findings. We report a young infant with pertussis who remained PCR-positive for a prolonged period of time despite appropriate clarithromycin treatment.
Case: A 4-week-old newborn acquired *Bp* infection, proven by PCR, after exposure to several household members with unrecognized pertussis. Hospitalization was required due to apneic spells. Antibiotic treatment (15 mg/kg/d oral clarithromycin) was started immediately and tolerated well. The result of quantitative PCR was 7.02 log (GEq/ml) at onset of treatment (day 1) and remained at 6.26 log at the end of treatment (d7). The test was repeated on day 11 remaining positive at 7.17 log. The child progressed well and was discharged from hospital, but remained in isolation with antibiotic treatment prolonged for another week. Further PCR tests on d14 and d19 (end of 2nd antibiotic course) remained positive with 2.66 and 2.67 log.
Conclusion: 7 days of clarithromycin may be insufficient to terminate contagiousness of *Bp* infection. Prospective treatment studies using both PCR and culture are warranted.

P190

Osteomyelitis of the dens axis – a rare cause of acute torticollis

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Introduction: Acute Torticollis is a rather common diagnosis in children with a broad differential diagnosis. Osteomyelitis of the dens axis is a rare cause of torticollis. In literature only few cases are described and thereof the smallest part in children.
Case report: We report on two boys (2 8/12 and 1 2/12) who presented at our hospital with an atraumatic torticollis and progressive rest pain for two, respectively four weeks. Antiphlogistic therapy was inefficient in both so far. The boys presented both with a distinct torticollis, in a non-toxic state, one afebril the other subfebril, both with discrete cervical lymphadenopathy and without neurological deficits. CRP was slightly elevated (<20 mg/l), leucocyte count was normal, but the erythrocyte sedimentation rate was in both cases importantly elevated (70 > mm/h). Diagnosis was made by MRI. Since the localization was delicate and perioperative risk was high, an interdisciplinary panel decided in both cases against biopsy. Blood cultures in both and a throat culture in one were negative. Therefore the offending pathogens remained unknown in both cases. With appropriate antibiotic treatment the clinical course was uneventful. Both boys recovered fully and show no sign of sequelae.
Conclusion: Osteomyelitis of dens axis is a very rare cause of acute torticollis in infancy. It should however be considered when symptoms are prolonged and antiphlogistic therapy is not effective. Inflammatory markers might be only slightly elevated and children may present in good general condition. MRI is the examination in first place.

P191

Peripheral facial paralysis in herpes infection

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Introduction: Incidence of acquired peripheral facial paralysis is 8.6/100,000 children/year. Etiology includes infections, tumor, trauma and may be inflammatory or idiopathic. We present a girl whose etiological condition and treatment were not easy to determine.
Case report: A 7 y.o. girl presented with asymmetrical smile for 24hours. Aside an isolated typical peripherical facial paralysis, neurological examination was normal.
Because Lyme disease is frequently encountered in our region, the child was treated with prednisone and amoxicillin. The child did not have any pre-existing illnesses nor was bitten by a tick. Lyme serologies returned negative and amoxicillin was stopped.
Two months earlier, she had painful white spots in her mouth lasting for some weeks.
Because family history (mother and sister) for aphtous ulcers was positive, Behcet disease was evoked, but HLA B51 was negative. Swab of the white spots tested by PCR was positive for HSV1. No antiviral therapy was started. The girl had a full recovery in a couple of weeks.
Discussion: Etiology of facial paralysis is not the same all over the world. In Valais, Lyme disease is frequently found. When Lyme disease was excluded, HSV1 was suspected to be the etiological agent. Antiviral therapy has no proven effect on the course of disease[1].
Conclusion: Etiology of facial paralysis is not easy to confirm. Laboratory tests (LP or not LP) and therapeutic strategies are controversial even in Switzerland. When Lyme disease is excluded, other etiologies should be evoked.

Reference
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P192

A severe complication of sinusitis

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Introduction: Sinogenic intracranial empyema in children is a rare, but severe complication of sinusitis. It may be life threatening or lead to devastating neurologic sequelae.
The clinical triad of fever, headache and altered mental status often reported in adults may be missing.
Case: An 8 year old boy presented to our emergency department with a 12-day history of severe undulating headache and persistent fever. There was a marked worsening in his general condition and increasing frontal headache prior to admission. With marked leukocytosis in the cerebrospinal fluid and elevated CRP, meningitis was suspected and empiric antibiotic and antiviral treatment was started. A contrast-

enhanced cerebral CT scan showed pansinusitis and a small amount of subdural fluid located frontal on the right side. On admission neurological examination was unremarkable. A few hours later the clinical situation deteriorated with therapy refractory status epilepticus, requiring multiple anticonvulsiva and admission to intensive care unit. Despite improving general condition, left-sided hemiparesis and occasional seizures persisted. Due to increasing amount of subdural fluid on MRI scan 3 days later neurosurgical drainage, drainage of sinuses and partial ethmoidectomy was done, confirming the diagnosis of subdural empyema. There was no growth in the initial blood and CSF cultures, but Streptococcus pyogenes was identified as causative agent in PCR amplification of drained material. Intravenous antibiotic treatment was continued for one month. Discrete paresis of the left arm persisted throughout the hospitalization, no further seizures were observed on levetiracetam-monotherapy.
Conclusion: In the pediatric population signs of sinusitis may be unspecific. Sinogenic intracranial empyema should be considered in patients with persistent headache of biphasic evolution, focal neurological signs and altered mental status. Early detection with contrast-enhanced CT or MRI scan is recommended, followed by antibiotic treatment and surgical drainage of subdural empyema and affected sinuses.

P193

Painful neuropathy in pediatric Lyme disease – causality or coincidence?

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Introduction: Neuropathic manifestations of Lyme disease are very rare in childhood. Establishing causality is difficult, as is effective treatment.
Case report: A 13 y/o boy presented with disabling right-sided plantar foot pain after a 4-week history of flu-like symptoms. There was a history of travel in Lyme endemic regions with rash 1 month prior to onset of symptoms, but no evidence of tick bite or of trauma. Initial neurologic and orthopedic exam was within normal limits, without motor or sensory deficit. Since the father was treated for Lyme disease, it was suspected also in the boy and then confirmed by serology. Oral amoxicillin provided no symptom relief, motivating further investigations. CSF analysis revealed pleiocytosis and intrathecal IgM synthesis, confirming neuroborreliosis. Brain and spine MRI showed slight hyperintensity of spinal nerve roots at cervical and cauda equina level. Nerve conduction studies, X-ray and MRI of right foot were normal.
IV-ceftriaxone was given for 2 weeks. Oral gabapentin produced little pain reduction, but carbamazepin and amitriptylin were effective within few days. Three months later the patient was asymptomatic.
Discussion: Neurologic manifestations of Lyme disease such as cranial nerve palsies are common in children, but neuritis is very rare. Clinical diagnosis should be confirmed by CSF analysis, serology, neurophysiologic studies and neuroradiology.
Conclusion: Neuropathic pain is a rare symptom of neuroborreliosis in children. It confronts the clinician with three major difficulties: corroboration of a causal nexus between infection and pain, exclusion of other etiologies and adequate pharmacotherapy.

P194

Septic sacroiliitis: a musculoskeletal complication of varicella in childhood

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Introduction: Varicella is a common viral infection with an annual estimate of 20 000 paediatric cases in Switzerland. Complications following varicella infection are rare among immuno-competent children. Septic arthritis as a complication of acute varicella mostly involves the knee and is mainly caused by *Streptococcus pyogenes* or *Staphylococcus aureus.*
Case report: A 2.5 year-old girl developed pain in the right hip 5 days after the onset of typical varicella lesions. A reactive arthritis was initially suspected. After a 6-day history of pain new onset of fever and raised inflammatory markers the girl was referred to our hospital. On examination the child was febrile, had a diminished range of motion and limpness of the hip/lower back and palpation was painful. Magnetic resonance imaging of the pelvic area showed inflammation of the right sacroiliac joint. Intravenous antibiotic treatment with amoxicillin/clavulanic acid led to improvement within a few days. After one week treatment was changed to oral and continued for another 3 weeks with complete resolution of symptoms.
Discussion and conclusion: Serious musculoskeletal complications including septic arthritis, osteomyelitis and abscesses following varicella infection are uncommon (1/10 000 cases). Sacroiliitis accounting for 1–2% of osteo-articular infections in children is therefore an extremely rare finding as a complication of varicella. The diagnosis of sacroiliitis in children is therefore difficult and often delayed due to the lack of specific signs and symptoms. Optimal diagnostic tests and management of children with septic sacroiliitis remain yet to be defined.

P195

The Swiss National Registry for Primary Immunodeficiencies (PID): report of the first 5 years' activity 2008–2013

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Introduction: To date >250 primary immunodeficiencies (PID) with more than 180 genetic causes are known. Most are rare diseases that are often diagnosed late or not at all with ensuing organ damage or death. The overall prevalence for PIDs is estimated to be 1:1000, and 1: 10.000 for severe PIDs. In Switzerland we therefore expect 7000 patients with PID. In a first publication in 1988 only 313 patients were documented. This was the incentive to start a Swiss National Registry for PID. The aims of this registry are to enrol as many Swiss PID patients as possible, to determine the prevalence of different PIDs and to search for geographic differences or family clustering. To build up a Swiss National Registry for PID, a nationwide network would be helpful.
Methods: In 2008 we started to register Swiss patients in the online registry of the European Society of Immunodeficiency (ESID). Today there are 89 documenting centres in Europe that have registered over 18.000 subjects since 2004. The registry has been used as a platform for many translational/basic research studies because it offers a wide range of well-defined patient collectives and it has turned out to be a useful tool to connect different centres.
Results: Today all 5 university centres, 3 level A Hospitals (Aarau, Lucerne and Sankt Gallen) and 1 centre in Bellinzona participate. Most of these started to register, and 337 patients with PID are already registered. Distribution of different PIDs, age distribution and the diagnostic delay for the different diseases are similar to the statistical data of the European cohort.
Conclusions: When all centres have registered their patients by the end of 2014, the first nationwide statistical analysis will be possible. As other national PID registries the Swiss National Registry can provide a basis for both national and international investigations and activities that aim to raise physicians' awareness of PID, allow better knowledge on clinical evolution or complications, and may have an impact on therapy costs.

P196

Invasive pneumococcal disease after implementation of 13-valent pneumococcal conjugate vaccine

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Background: Invasive infections caused by Streptococcus pneumoniae continue to be a major cause of morbidity and mortality worldwide. The widespread use of a heptavalent pneumococcal conjugate vaccine (PCV7) resulted in a significant decrease of serious invasive pneumococcal disease (IPD). However, the emergence of replacement non-PCV7 serotypes, especially serotypes 1,3 and 19A, has resulted in an increase in the incidence of serious and invasive infections caused by replacement serotypes. In 2011, a 13-valent PCV was licensed in Switzerland. Yet, the impact that this vaccine will have on IPD remains uncertain.
Objective: To discuss the epidemiology of invasive pneumococcal infections in Switzerland in the PCV13 era through the presentation of two clinical cases of IPD, which occured in fully vaccinated children.
Results: We report two cases of IPD, which occurred in children optimally vaccinated with PCV13:
Patient 1: A 2 year-old previously health boy was admitted with bilateral pneumonia complicated with left-side empyema, which progressed on i.v amoxicillin treatment. Pneumococcal antigen was detected positive from a pleural tap and urine and Streptococcus pneumonia was only detected by molecular assays from the pleural tap with ongoing genotyping of the strain. Adequate drainage and i.v. ceftriaxone allowed for a slow recovery.

Patient 2: A 2.5 year-old previously healthy girl was admitted with bilateral Streptococcus pneumoniae pneumonia complicated with right-side parapneumonic effusion and bacteremia. Subsequently she developed atypical hemolytic uremic syndrome necessitating peritoneal dialysis. It is of note that associated liver dysfunction presented as both transaminitis and cholestasis. Streptococcus pneumoniae was identified as penicillin sensitive (MIC 0.06) with ongoing serotyping of the strain. Peritoneal dialysis and i.v amoxicillin allowed for slow recovery.
Conclusions: Invasive pneumococcal disease is also observed among optimally vaccinated children with PCV13. The establishment of ongoing surveillance of the serotype distribution in IPD and complicated pneumonia is crucial to document serotype replacement.

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Varicella's reinfection is not as uncommon as thought…

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Introduction: Varicella's reinfection in immunocompetent patients remains a frequent and underestimated entity. We present the case of a boy with a complicated varicella reinfection.
Case report: A three-year-old boy presented multiple itching vesicles of different age. A year before, his paediatrician diagnosed varicella rash. His young sister presented a first varicella infection at 13 months old and a concomitant reinfection. At the third day after the beginning of the second episode of varicella, he was hospitalized for **Staphylococcus aureus** cellulitis. A day after a varicella's encephalitis was suspected in front of abnormal tonus and behaviour, tremor and headache. Varicella Zoster Virus was detected by polymerase chain reaction in cerebrospinal fluid. He was treated with amoxicillin/clavulanic acid, clindamycine and acyclovir. Cutaneous and neurological status quickly improved.
Discussion: S.Hall and al report 5 to 13% of immunocompetent children present varicella reinfection. The first episode is described at median age of 3 years old in varicella reinfection compared to 6 years old in a unique episode of varicella. Risk factors for a varicella reinfection are a first episode before one year old, a mild initial infection. Hereditary predisposition is suspected as 45% of concerned patients have a family member who presented varicella reinfection. Prevalence of complications in varicella's reinfections is not increased.
Conclusion: Epidemiological data on varicella reinfection are insufficient making recommendations difficult to edit. Nevertheless the family predisposition could be an argument to vaccine the other children at 11 years old as suggested in actual recommendations for children without varicella history.

Reference

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P198

Bone abscess presenting as tendinitis with minimally elevated inflammatory markers

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Early recognition of acute bone and joint infections is difficult because no laboratory marker that is both sensitive and specific is currently available. Pus culture is often considered the gold standard even if it shows low positivity rates. A 10-year-old female presented with a sore throat and fever diagnosed a viral pharyngotonsillitis; after two days she developed pain in the right wrist of sudden onset. She had no previous medical problems, and did not complain of any other symptom. Examination of the extremities revealed the presence of swelling, redness and local heat of the right wrist with no deformity or restriction of movement. Physical examination was otherwise normal; plain radiographs were normal and ecography showed inflammation of the tendons of the flexor muscles. Inflammatory parameters were normal so suspicion was initially of tendinitis. The day after, because of worsening of the pain, she underwent MRI scan, showing a huge abscess (maximum diameters approximately 65 x 18 x 9 mm) raising the periosteum, of the right distal radius, suggestive of osteomyelitis. Laboratory tests were still mildly elevated with WBC 5.5 G/L ESR: 16, CRP: 13 mg/L, procalcitonin: 0.12 µg/L. Intravenous antibiotics were administered and surgical debridement performed; blood and wound cultures elicited growth of MSSA. Clinical response was obtained after 10 days of parenteral treatment and she was discharged on oral antibiotics to complete treatment. We describe a case of bone abscess with only mildly elevated laboratory markers, to raise awareness among practioners of this presentation, and we review the literature.

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Varicella-associated invasive Group A streptococcal disease: a case report

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Chickenpox is most often a benign childhood illness, the most common complications are secondary to bacterial infection, especially by Group A beta-Hemolytic Streptococcus (GAS), that can sometimes be life-threatening. The association of these two infections is reported more and more frequently in the literature; we review this clinical presentation. We report the case of a previously healthy 5 -year-old girl affected by varicella that was admitted because of: decline of condition, development of vesicles over the entire body with superimposed infection, painful ulcers in the mouth and throat, severe dehydration and high fever. Her 3-year-old sister presented mild varicella and a scarlet exanthema. The diagnosis of streptococcal infection was confirmed by positive strept A test, GAS grew in blood cultures and was isolated from exudate of the blisters. The clinical symptoms improved with administration of antibiotics and intravenous rehydration , but it took approximately 3 weeks from the first visit for some of the lesions and ulcers to heal with epithelialization. The outcome will probably leave some unaesthetic scars. In conclusion severe invasive bacterial superinfection with GAS , even if reported as a rare event, must be considered in any case of varicella.

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Une infection virale à ne pas ignorer

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Les pétéchies en association avec un état fébrile provoquent toujours une certain inquiétude chez les pédiatres à cause de la possibilité d'une grave maladie infectieuse ou d'une pathologie tumorale.
Notre patiente agée de 10 ans s'est présentée à notre Centre des Urgences avec un état fébrile depuis 2 jours et des pétéchies au tronc ainsi qu' aux bras d'apparition soudaine; l'état général était conservé et l'examen neurologique dans la norme. Les investigations paracliniques montraient une dépression médullaire (une leucopénie à 2.1 G/l (ANC 1575) et une thrombocytopénie à 148 G/l) sans syndrome inflammatoire (CRP 3.2 mg/l), avec cependant un compte érythrocytaire dans la norme. En raison de la clinique (pétéchies et état fébrile sans foyer) et du résultat du laboratoire (dépression médullaire débutante) la patiente fut hospitalisée. Un diagnostic d'infection virale put être rapidement confirmé par la présence d'anticorps anit-IgM spécifiques anti-Parvovirus B19. Les contrôles suivants demontraient une reprise de l'activité médullaire ainsi qu'une résolution de l'exanthème paraviral.
Parvovirus B19, un virus de petite taille découvert en 1975, est connu comme étiologie de la 5ème maladie de la petite enfance. En 1990 le papular-purpuric "gloves and socks" syndrome fut décrit par Harms et al., un syndrome confirmé le plus souvent chez des patients d'âge adulte.
Nous rapportons ce cas de présentation atypique au vu de l'âge et de la clinique.
Ainsi en présence de fièvre et d'une éruption pétéchiale sans altération de l'état général chez l'enfant d'âge scolaire et au-dela, il nous semble utile d'inclure dans le bilan initiale les investigations pour la recherche du Parvovirus B19.

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Preferences for the organization of long-term follow-up in childhood cancer survivors

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Purpose: Follow-up care is important for many childhood cancer survivors to detect and treat late effects early. We aimed to describe follow-up attendance, reasons for follow-up, and preferences for organisation of follow-up in Swiss childhood cancer survivors.
Methods: We contacted 720 survivors who previously participated in the Swiss Childhood Cancer Survivor Study. They had been diagnosed with cancer aged ≤16 years in 1990 or later, were ≥5 years after diagnosis and ≥18 years of age when they received the questionnaire. We asked about reasons for attending follow-up, the importance of medical and general aspects and the preferred organisational model using 0-3 point scales (higher score = more agreement). We used descriptive statistics to analyse data.
Results: Of the 314 (44%) responders, 150 (47.8%) reported they still attended follow-up. Clinical reasons (mean = 2.33 SD = 0.58) were more important than supportive reasons (mean = 1.61; SD = 0.71; p <0.001). Among medical aspects, survivors reported *checking for cancer recurrence* was most important (mean = 2.78, SD = 0.53), before *screen for late effects* (mean = 2.67, SD = 0.53; p = 0.001) and *information on potential late effects* (mean = 2.63, SD = 0.55; p <0.001). Regarding general aspects knowing about *risks for my children* was rated most important (mean = 2.22, SD = 0.83). Regarding the organizational model, survivors rated paediatric oncologist follow-up (mean = 2.24, SD = 0.72) and medical oncologist follow-up highest (mean = 2.17, SD = 0.69; p = 0.087). Hospital-based follow-up by multidisciplinary team (mean = 2.07, SD = 0.73), GP follow-up (mean = 1.90, SD = 0.84), or follow-up by telephone or questionnaire (mean = 1.06, SD = 0.83) were less favoured.
Conclusion: Clinic based follow-up by specialists is highly favoured by childhood cancer survivors and could provide an optimal model for organizing follow-up in future.

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Involvement of Swiss physicians in follow-up care of childhood and adolescent cancer survivors

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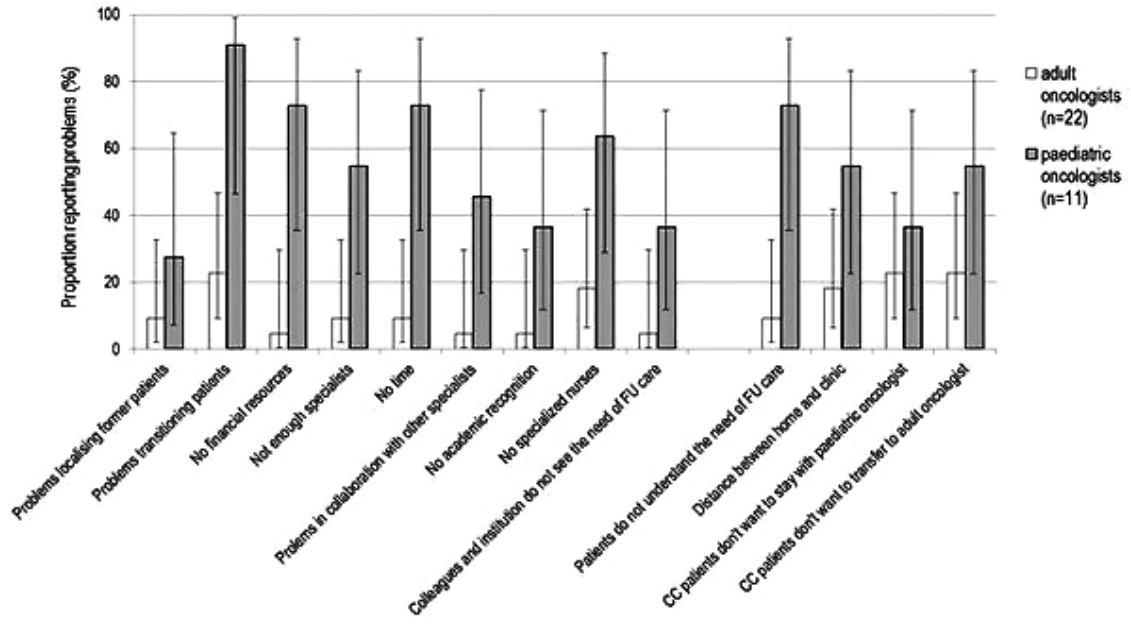
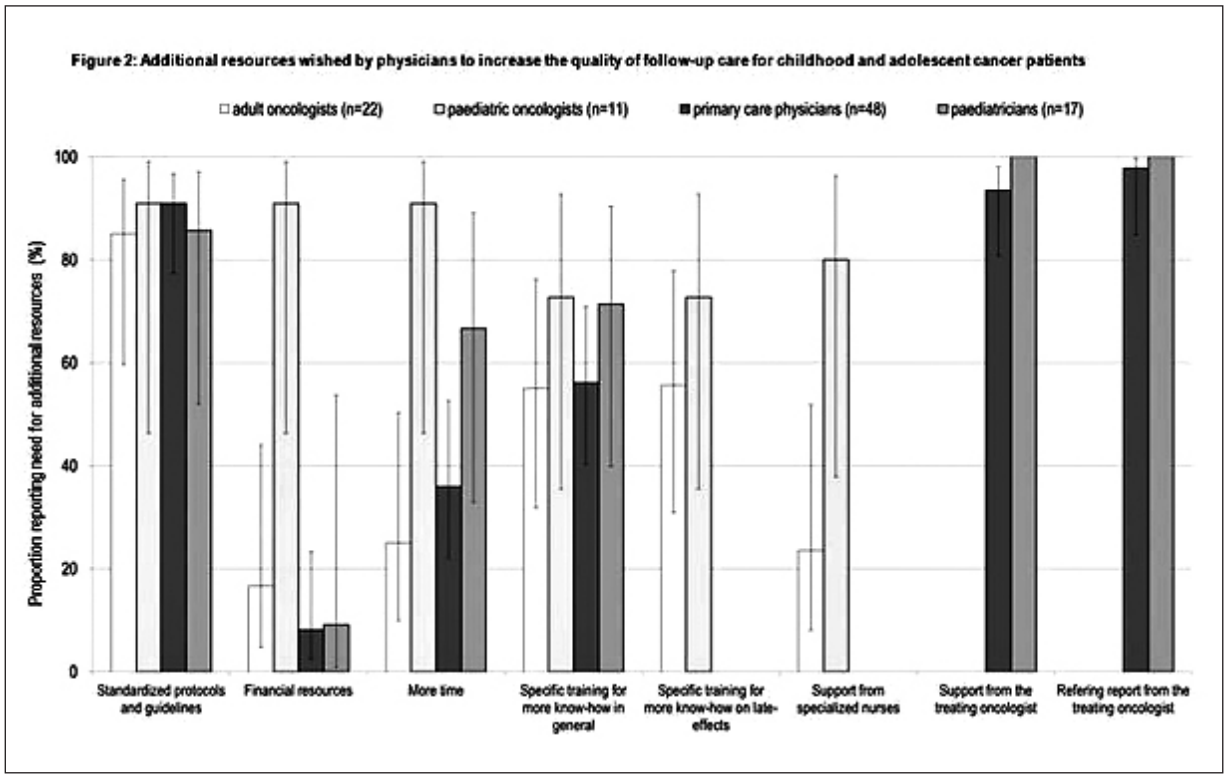
Introduction: Regular follow-up is essential for childhood and adolescent cancer survivors. We wanted to describe 1) involvement of Swiss physicians in follow-up, 2) their problems encountered, and 3) additional resources needed for optimal follow-up.
Methods: We sent adapted questionnaires to a sample of Swiss physicians including adult oncologists (AO), paediatric oncologists (PO), primary care physicians (PCP) and paediatricians (P). Only oncologists participating in follow-up were asked to report problems encountered. PCPs and Ps not involved in follow-up could indicate reasons why.
Results: 183 physicians returned the questionnaire (27 AO, 13 PO, 122 PCPs, 21 P). Involved in follow-up were 81% AO, 85% PO, 39% PCPs and 81% P. POs encountered more problems than AOs in follow-up, mostly during transition of patients (91%), because of financial resources (73%), time restraints (73%) and patients neglect of the need of follow-up (72%; figure 1). AOs reported most problems during transition (23%). Reasons of PCPs and Ps for not participating in follow-up were: not knowing patients needing follow-up (74%), oncologists taking care of (51%), and lack of experience (28%). All physicians stated the need for standardized protocols (85%–91%) and specialized training (55%–73%, fig. 2). POs additionally listed needing more financial resources and time (both 91%). PCPs (94%) and Ps (100%) wished more support from the oncologists.
Conclusions: Many physicians in Switzerland reported to be involved in follow-up of childhood and adolescent cancer survivors. A national follow-up model to increase efficiency and know-how of involved physicians as well as collaboration between specialists needs to be developed.

Figure 1: Problems with follow-up care on institution and patient level reported by adult and paediatric oncologists

Abbreviations: FU, Follow-Up; CC, Childhood Cancer





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Information provision and information needs in parents of long-term childhood cancer survivors

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Introduction: Studies on information provision and information needs for parents of long-term childhood cancer survivors are scarce. We aimed to describe 1) the information parents received during and after treatment, 2) parents' needs for information today, and to investigate 3) associations of information provision with clinical characteristics of the child's disease and late outcomes.
Methods: As part of the Swiss Childhood Cancer Survivor Follow-up study a questionnaire was sent to parents of childhood cancer survivors, diagnosed <16 years, aged 11–17 years at study who had survived ≥5 years after diagnosis. The questionnaire included questions on information provision and information needs, current follow-up, and socio-demographic information. Information on clinical characteristic of the child's disease was available from the Swiss Childhood Cancer Registry.
Results: Most parents reported to have received oral information (information on illness: oral 91%, written: 40%; treatment: oral: 88%, written: 46%; follow-up: oral: 74% written: 27%; late effects: oral: 68%, written: 19%). Many parents reported current information needs, especially on late effects (62%). The preferred source was written general (42%) or oral information (38%). In univariable regression models, we found that male parents, those higher educated and parents of children who received radiotherapy reported more often to have received information. There was a trend for less information in migrated parents, parents of children with CNS tumour, or with late effects.
Conclusion: Parents need more information especially on possible late effects. Appropriate information provision is important to allow parents take informed decisions on follow-up care of their children.

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Perioperative management in a paediatric patient with protein S deficiency

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Background: Protein S (PS) is a vitamin K-dependent natural anticoagulant acting as a cofactor for activated protein C and inhibiting activated FV and FVIII to down regulate thrombin formation. There are two forms of PS: a free active form (40%) and an inactive form bound to C4b binding protein (60%). Hereditary PS deficiency follows an autosomal dominant inheritance while acquired PS deficiency is associated with autoimmune disease. PS deficiency is a risk factor for venous thromboembolism.
Case: An eight year old girl with Moebius syndrome, a congenital facial paralysis and muscle aplasia, was scheduled for complex plastic facial microsurgery. Because of a positive personal (juvenile rheumatoid arthritis, ANA positive) and family history for autoimmune disease, thrombophilia work-up was initiated. PS deficiency with a free PS of 0.42 IU/ml (0.67–1.97) and a total PS of 0.51 IU/ml was detected with otherwise normal thrombophilia work-up.
The possibility of thromboprophylaxis in the setting of PS deficiency was considered because of the risk of both bleeding and thrombosis of vascular microanastomoses. Three perioperative FFP infusions were administered. Resulting free PS levels ranged from 0.68IU/ml to 0.80 UI/ml. There were no haemostatic complications either peri- or post-operatively.
Because of ANA positive autoimmune disease and PS deficiency in both the mother (free and total PS 0.45IU/ml and 0.52 IU/ml, respectively) and the patient, the differentiation between inherited and acquired PS deficiency remained unclear (father with normal PS levels).
Conclusion: Thromboembolic risk of PS deficiency was effectively controlled by perioperative application of FFP in an eight year old girl with complex facial microsurgery.

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Unusual presentation of an intraspinal chloroma in a child with acute lymphoblastic leukaemia (ALL)

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Introduction: Chloromas are solid collections of extramedullary leukemic cells usually described in myeloid but occasionally in ALL. Their observation might precede the development of the leukemia. Although reported at various sites such as retrobulbar areas, skin, bones and lymph nodes, spinal involvement is unusual.
Case report: A 7 y.o boy presented with back pain, urinary incontinence, confusion, fatigue and fever. Physical examination revealed painful percussion and mobilisation of the vertebral column. Neurological examination was normal. Moderate pancytopenia was observed with no peripheral blasts. Cerebrospinal MRI showed an intramedullary mass extending from T3 to T9.
A day after admission, a second blood smear revealed blasts and ALL was confirmed. CSF was negative. Standard chemotherapy was initiated and pain rapidly disappeared. Ten days later, a follow up MRI showed regression of the mass that disappeared at the end of induction.
Discussion: Chloromas have been reported as the initial presentation of leukemia. Spinal involvement is rare. In our patient, MRI revealed an intramedullary mass mimicking tumours such as meningioma or ependymoma. CSF was negative and the patient classified as CNS 1. We opted not to biopsy the mass and observed radiological evolution. Nevertheless, due to the spinal involvement, triple intrathecals were added to the standard treatment protocol. A rapid and complete regression of the solid mass was observed alongside disappearance of clinical symptoms. Treatment is ongoing but radiotherapy will not be included upfront.
Conclusion: Chloromas in ALL are rare tumours, especially intraspinal ones. ALL should be included in the differential diagnosis of intramedullary masses.

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Spontaneous splenic rupture in hereditary spherocytosis after Parvovirus B19 infection with aplastic crisis

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Introduction: Splenic rupture in hereditary spherocytosis (HS) in children is very rare. Our case bled spontaneously into the spleen after aplastic crisis in HS and parvovirus B19 infection. There is no recent literature covering this topic.
Case presentation: We present the case of a 10 year old girl with moderate HS who presented with convulsive syncope after an episode of fever. Due to an acute Parvovirus B19 infection she showed an aplastic crisis and additional haemolysis. She needed repeated red blood cell (RBC) transfusions. Compared to the sonography performed a year before, the spleen was slightly larger, but showed no other abnormality. Four days after discharge she presented with severe abdominal pain. The sonography then showed splenomegaly with a new triangular hypoechogenity subcapsular, an MRI confirmed the diagnosis of a spontaneous splenic rupture. She was treated conservatively.
Discussion: Splenic rupture is much more common in patients with viral infections, especially EBV, than in HS patients, even though they have massive splenomegaly. In our case the parvovirus B19 infection led to an aplastic crisis, combined with haemolysis, so the pre-existing splenomegaly progressed slightly. Probably due to the viral infection, the acute haemolysis and the repeated RBC transfusions the spleen became particularly vulnerable.
Conclusion: Even if spontaneous splenic rupture is rare in children with HS, there may be additional circumstances which lead to a special vulnerability of the spleen. Therefore splenic rupture is to consider in children with HS presenting with severe abdominal pain.

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A rare combination of severe ADAMTS13 deficiency with acute liver failure

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Introduction: ADAMTS13 cleaves von Willebrand Factor regulating platelet aggregation. A severe ADAMTS13 deficiency (<5%) is pathognomonic for Thrombotic Thombocytopenic Purpura (TTP), a rare disease with a high mortality rate when untreated.
Clinical case: We report the case of a 15 year-old girl with biliary atresia and acute-on-chronic liver failure. She presented with acute renal failure, petechiaes, anemia and thrombocytopenia first ascribed to presumed bacteremia. Faced with persistent anemia and thrombocytopenia, we suspected TTP and identified a severe ADAMTS13 deficiency (<5%) due to a strong ADAMTS13 inhibitor (2BU/ml) establishing a diagnosis of acquired TTP (aTTP). Management included plasma exchange, steroids and rituximab. After one week, the ADAMTS13 inhibitor had disappeared and ADAMTS13 activity increased. Eight weeks later, the patient underwent liver transplantation (LT) and the next day ADAMTS13 activity was normal. She received daily fresh frozen plasma infusion during 10 days. Post transplant immunosuppression included basiliximab, prednisone and tacrolimus. Since then, she has remained in remission with normal ADAMTS13 activity and no inhibitors.
Conclusion: In children and in solid organ transplantation, thrombotic microangiopathies are usually not associated with severe acquired ADAMTS13 deficiency, and the few cases reported to date were diagnosed after LT. To the best of our knowledge, this is the first report of aTTP occurring in the setting of acute-on-chronic liver failure prior to LT. Aggressive treatment resulted in clinical remission in due time, which allowed for successful LT without any thrombotic complications. Close follow-up is warranted as relapse of aTTP is common.

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Fatal outcome of disseminated mucormycosis in a patient with relapsed AML and double cord HSCT

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Introduction: Mucormycosis is an increasingly recognized invasive fungal infection in immunocompromised patients associated with high mortality rates ranging from 50–100%.
Case: We report a 20 year old female patient with relapsed AML who underwent allogeneic HSCT with an HLA mismatched (8/10) double cord transplant after conditioning with cyclophosphamide and total body irradiation. Engraftment was on day +30. Bone marrow on day +30 and +100 showed complete remission and full donor chimerism. On day +4, febrile neutropenia developed and empiric antibiotic therapy with meropenem was initiated. On day + 14, progressive right oculo-orbitar swelling with exophthalmia and visual impairment, right paranasal sinusitis developed. The CT scan showed pulmonary infiltrates. Antifungal therapy with liposomal amphotericin B and posaconazole was added on days +9 and +15, respectively. BAL identified *Lichtheimia corymbifera* (genera causing mucormycosis) on day +25. Surgical debulking of the right orbit and paranasal sinuses was conducted on day +36 with good clinical response. Pulmonary mucormycosis showed further progression on imaging studies therefore right middle lobectomy was performed on day +61. Postoperatively, the patient developed ARDS that required long-term ventilation. Differential diagnoses included ARDS, tension pneumothorax, viral or bacterial superinfections, progressive mucormycosis, GvHD of the lungs, and idiopathic pneumonia syndrome because of transient respiratory improvement after repeated cycles of high dose steroids. Imaging studies showed severe alveolar consolidation of both lungs with multiple pneumatoceles. On day +140 post HSCT, the patient passed away due to progressive respiratory failure. Autopsy of the lungs showed predominance of pulmonary angio-invasion by mucormycosis with obstruction of pulmonary vessels, persistent fungal mycelia, extensive destruction of alveoli, and necrosis. There were no apparent signs of GvHD or leukemic infiltration.
Discussion: Early diagnosis, rapid administration of antifungal agents, early and aggressive surgery, and reversal of underlying predisposing risk factors are crucial to control mucormycosis. However, patients post HSCT might be asymptomatic in the early course of infection especially when neutropenic, but critically ill later on making surgical intervention and definitive cure extremely difficult.

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Vitamin D status among children and adolescents on anticonvulsivant drugs in Southern Switzerland

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Introduction: Vitamin D status is often inadequate (<50 nmol/L) in epileptic children, mainly because some anticonvulsivant drugs induce the enzymes responsible for its inactivation. The purpose of the present study was to address vitamin D status among children and adolescents treated with anticonvulsivant drugs and control subjects who reside in Southern Switzerland, a high solar radiation region.
Methods: Between January and May 2013, total serum 25-hydroxyvitamin D was assessed by liquid chromatography-tandem mass spectrometry in 58 children and adolescents with epilepsy and 29 controls residing in Southern Switzerland. Dark-skinned individuals, females wearing dress styles covering practically the whole body and subjects with body mass index ≥85th percentile for age and sex were excluded.
Results: Concentration of serum 25-hydroxyvitamin D was similar in epilepsy patients (48 [37–62] nmol/L; median and interquartile range) and control subjects (53 [47–64] nmol/L). An inadequate serum 25-hydroxyvitamin D concentration was common both among patients (55%) and control subjects (34%). Serum 25-hydroxyvitamin D was significantly lower among patients treated with anticonvulsivant drugs that induce the metabolism of vitamin D (30 [21–51] nmol/L) than among the remaining patients (51 [40–65] nmol/L) and controls.
Discussion: The present study indicates a relevant tendency towards inadequate vitamin D status among children with and without anticonvulsivant drug management who reside in Southern Switzerland. This tendency is more prominent in patients treated with anticonvulsivant drugs that induce the metabolism of 25-hydroxyvitamin D. Based on these data, supplementation with vitamin D is advised both for children with and without epilepsy.

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Metabolic disturbances and renal stone promotion on treatment with topiramate: a systematic review

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Introduction: The use of topiramate, which is prescribed for the management of epilepsy, for migraine headache prophylaxis and as a weight-loss agent, has been associated with the development of metabolic acidosis, hypokalaemia and renal stone disease. We systematically reviewed all the literature.
Methods: The systematic review of the literature was realized using the principles underlying the UK Economic and Social Research Council guidance on the conduct of narrative synthesis and the **P**referred **R**eporting **I**tems for **S**ystematic Reviews and **M**eta-**A**nalyses statement.
Results: Fourty-seven reports published between 1996 and 2013 were retained for the final analysis. Five case-control studies and 6 longitudinal studies addressed the effect of topiramate on acid-base and potassium balance. A significant tendency towards mild to moderate hyperchloremic metabolic acidosis (with bicarbonate ≤21.0 mmol/L in approximately every third case) and mild hypokalaemia (with potassium ≤3.5 mmol/L in 10% of the cases) was noted on treatment with topiramate. This tendency was similar in children and adults. A single study observed that topiramate causes mild hyperuricaemia in male adults. A tendency towards hypocitraturia, a recognized promoter of renal stone formation, was noted in all patients on topiramate.
Conclusions: Increasing evidence supports the use of topiramate. Topiramate is generally well tolerated and serious adverse events are rare. Nonetheless, the current systematic review of the literature indicates that its use is linked with the development of acidosis, hypokalaemia, hyperuricaemia and hypocitraturia.

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Effects of working memory training on functional network connectivity in patients with juvenile multiple sclerosis

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Background: Cognitive deficits, especially in the domains of working memory and attention, are reported in one third of patients with juvenile multiple sclerosis (JMS). So far, there is little knowledge of efficient treatment approaches in children. In adult MS, cognitive treatment effects on activation patterns in fMRI but also altered functional connectivity have been reported.
Aim: To evaluate the effects of a computerized working memory training in patients with JMS.
Methods: Five patients with clinically definite MS, aged between 12 and 18 under all medications were included so far. All patients underwent a comprehensive neuropsychological testing at baseline combined with resting-state fMRI at baseline and after a computerized working memory training that lasted 4 weeks (BrainStim). Resting-state fMRI data were preprocessed using SPM8. To analyze these data we applied an independent component analysis (ICA) and extracted resting state networks by means of the Gift toolbox. Further, we focused on components related to the default mode network (DMN), which represents an important regulatory system for cognitive functioning, and studied the executive control network (right and left) as well as the anterior salience network. Finally, connectivity was determined by applying the FNC toolbox. Due to the small number of participants at this point we decided to run a first analysis in a case-based approach.
Results: An interim analysis of the first 5 patients (m = 2, f = 3; disease duration 1 to 4 years) who completed the training so far is presented. After cognitive intervention, two patients showed increased performance in visual and verbal working memory as well as in alertness, one patient improved solely in working memory but not in other cognitive domains and two patients showed even a small decrease in performance. Patients had significantly higher connectivity between the right and the left executive control network after the training. Additionally, in patients with increased cognitive performance, higher connectivity within the DMN after the training compared to baseline was detectable.
Conclusions: First results of our interim analysis indicate that working memory training may be effective in some JMS patients. In the so-called "responders", positive training effects in several behavioral as well as MRI measures were detectable.

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Posterior reversible encephalopathy syndrome in childhood: a systematic review of the literature

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Background: Posterior reversible encephalopathy syndrome (PRES) is a generally reversible neurologic condition that is diagnosed based on distinctive clinical and radiologic findings. Imaging discloses a predominantly posterior leukoencephalopathy.
The objective was to analyze predisposing factors, clinical symptoms and outcome of PRES in childhood.
Methods: A systematic review of published case reports and case series dealing with PRES was performed using the "Medline" database. The publications retained for this analysis included patients younger than 18 years of age who were described individually, revealing as a minimum age, gender, cerebral imaging findings and outcome.
Results: In total, 196 reports were included describing 476 patients with a mean age of 10.2 years (range 0.1–18 years), 49% were female. The most frequent associated condition was arterial hypertension (45.8% of the patients), followed by medication without concomitant arterial hypertension (27.9%), and medication combined with arterial hypertension (20.8%). Two thirds of the cases due to medication were caused by calcineurin inhibitors (cyclosporine A or tacrolimus), and about 1/3 by cancer chemotherapy. Frequent symptoms were seizures, headache, blurred vision or altered mental status. In about 50% of the cases the localization of the lesions on cerebral imaging was in the posterior part of the brain (more frequently in the occipital region), but in some cases purely in the frontal lobe. In almost all cases, with an

immediate antihypertensive therapy or discontinuation of the causative medication, the outcome was described as good with complete normalization of the cerebral imaging findings and/or disappearance of the initial symptoms.
Conclusions: This comprehensive analysis shows that PRES in childhood is mainly due to a pathological (acute) increase of blood pressure or some medication like calcineurin inhibitors or cancer chemotherapy. The cerebral localization of the lesions is mainly described in the posterior region of the brain. With prompt adequate therapy and discontinuation of the causative medication, the outcome is mainly described as good.

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Chronic inflammatory demyelinating polyradiculoneuropathy or Guillain Barré syndrome?

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Introduction: We report a case of chronic inflammatory demyelinating polyradiculoneuropathy (CIDP), a rare autoimmune disease of the peripheral nervous system, with similar clinical presentation to Guillain-Barré syndrome (GBS).
Case report: A 21-month old boy presented with loss of walking, hypostenia, and fecal retention, occuring a few days after a subfebrile state with light diarrhea (C jejuni+). Areflexia and weakness in the four limbs were found. IgM were positive for Parvovirus B19. CSF showed absence of albumino-cytological dissociation. Brain and spinal cord MRI showed slightly enhanced contrast in the *cauda equina*. EMG was compatible with polyradiculoneuritis.
Treatment was initated with IgG and corticosteroids. Diagnosis of a GBS was made. Relapses of similar symptoms occurred at the ages of 2 and 3 years, prompted by febrile episodes, which led to diagnosis of CIDP. Control MRI was normal. Repeated IgG infusions (0.4 g/kg/jour) at intervals of 8 week. With this treatment the boy is symptom free.
Discussion: Initialy CIDP presents like GBS. Presence of dysautonomia at onset (moderate bowel dysfunction in this case) is suggestive of CIDP and a second episode is characteristic of this syndrome. Swift recognition of CIPD is important in order to initiate repeated IgG infusions to avoid relapses and irreversible damage. The treatment modalities are not clearly defined.
Conclusion: In front of a first episode of GBS with autonomous symptoms diagnosis of CIDP should be kept in mind. A second episode confirms the diagnosis and should introduce long term treatment.

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Varicella: time for Immunization?

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Introduction: 77'000 children anually with acute varicella infection lead to approximately 140 hospitalizations in Switzerland. In 2013 we treated five patients with serious varicella-associated complications; one formerly healthy schoolboy died. As this highly contagious disease is mostly harmless it is not yet part of the routine vaccination schedule in Switzerland.
Case reports:
– A 7 year old boy with florid varicella infection was admitted showing signs of meningoencephalitis and subsequently sepsis. Despite maximal efforts his general state deteriorated over the next hours. Diagnosis of systemic infection with listeria monocytogenes was established. Due to progressive parenchymal swelling of the brain a decompressive craniotomy was performed but the patient died.
– A 6 year old boy showed altered consciousness and aphasia during acute varicella infection. Liquor showed pleocytosis and PCR-positivity for varicella. He was treated with i.v. acyclovir and showed slow improvement.
– A 4 year old girl showed deep ecthymata on her trunk: She had had acute varicella infection two weeks earlier on a summer holida abroad. She was treated with Amoxicillin/Clavulanate and showed excessive scarring.
– A 5 year old boy was admitted with staphylococcal toxic shock syndrome with acute varicella infection. He also showed pulmonary infiltrates, was treated with Amoxicillin/Clavulanate and Clindamycin and recovered completely.
– A 3 year old girl showed a facial phlegmone with florid varicella. The swelling rapidly progressed so she couldn't see for three days. She was treated with Amoxicillin/Clavulanate.
Conclusion: After 2013 we feel that childhood immunization against varicella is worth considering and feasible.

Posters SGP G / Posters SSP G: Pharmacology, psychology, mixed topics

P215

Clinical and toxicological features of attempted suicides by deliberate self-poisoning in adolescents in Switzerland

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Objectives: Although deliberate self-poisoning (DSP) in young people is common, information on clinical and toxicological characteristics of suicidal poisonings in this age group in Switzerland is limited.
Methods: Retrospective analysis of cases of attempted suicide by DSP in adolescents (10–19y), reported by physicians to the STIC between 1997 and 2012. The severity of symptoms was graded according to the Poisoning Severity Score.
Results: 5650 cases were included: 4712 (83%) females and 898 (16%) males (40 unknown), resulting in a female/male ratio of 5.2:1. The mean age was 16.4y, with a peak at 15y in females and 18y in males. 908 (16%) patients remained asymptomatic, 3606 (63%) showed mild, 837 (15%) moderate, and 299 (5%) severe symptoms. We found a significant association between development of severe symptoms and male gender (OR 1.76; 95% CI 1.34–2.31; $p < 0.0001$). Pharmaceuticals were involved in 92%, household products in 3%, and drugs of abuse in 2% of the cases. There were 3001 multiple-agent and 2649 single-agent poisonings, mainly with psycholeptics (655), analgesics (427), psychoanaleptics (413), NSAIDs (287), and cold preparations (112). The drugs most commonly involved were paracetamol (299), mefenamic acid (154), and dextromethorphan (103). The highest rate of severe courses was observed with amitriptyline, carbamazepine, and gasoline.
Conclusions: In our country suicidal self-poisoning is more frequent in females with a peak at the age of fifteen, and mostly associated with mild to moderate symptoms. However, the remarkable frequency of paracetamol poisoning in adolescents deserves particular attention because of the need for specific treatment to prevent fatal poisoning.

P216

A case of familial primary congenital glaucoma

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Introduction: Primary congenital glaucoma (PCG), the most common type of childhood glaucoma, is usually sporadic and is due to abnormal development of the anterior eye chamber. We report a familial case.
Case report: A term newborn presented with buphthalmos and corneal clouding of the left eye. Her Indian mother had been affected by bilateral glaucoma. Ocular pressure of left eye was high (32 mm Hg), compared with right (18 mm Hg). Treatment was initiated with brinzolamide and timolol drops, with favorable outcome by day 6. Diagnostic work-up (brain ultrasound, auditory evoked potentials, electrocardiogram) was normal. In the absence of other clinicial or etiologic clues, familial PCG was diagnosed. Left eye goniotomy was performed at one month, with partial healing. A second intervention is considered, with topical treatment in the meantime.

Discussion: PCG, usually diagnosed between birth and 3 years of age, is causally heterogeneous and can be difficult to detect in newborns, especially in those with dark-colored eyes. It is often bilateral and sporadic. Presentation at birth (and/or positive family history) should raise the possibility of a genetic form. **CYP1B1** is the gene most frequently involved.
Prognosis changes if it is associated with other ocular or systemic defects, such in Sturge-Weber syndrome or certain metabolic diseases. Early goniosurgery is the preferred treatment.
Conclusion: Neonatal PCG is rare and should be actively looked for, especially if family history is positive. Syndromic conditions, in particular Sturge-Weber syndrome, are crucial to exclude. The sooner the surgery is performed in PCG, the better is the prognosis.

P217

Vomiting, diarrhea, dehydration, acidosis: banal gastroenteritis or first presentation of methylmalonyl-CoA mutase deficiency?

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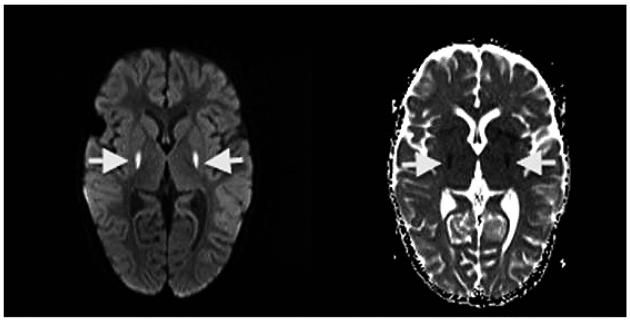
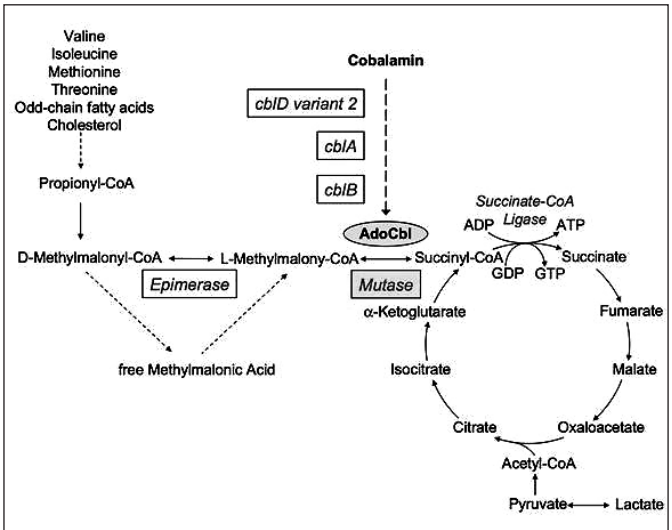
Introduction: Methylmalonyl-CoA mutase (MUT) is a mitochondrial enzyme, which in collaboration with cobalamin (vitamin B12)-cofactor, metabolises specific amino acids and fatty acids. Genetically caused dysfunction of methylmalonyl-CoA mutase (MUT) or lack of the essential cofactor cobalamin (due to inadequate uptake, transport or intracellular metabolism) leads to the clinical picture of methylmalonic acidemia. This inborn error of metabolism usually presents acutely in infancy with acidosis, vomiting, lethargy, failure to thrive and development delay, often leading to renal failure in due course.
Methods: Our female patient was born to healthy parents. She showed normal newborn screening, normal developmental milestones and followed a balanced diet. At the age of three years she was admitted in our pediatric intensive care unit in a lethargic and dehydrated state after recurrent vomiting and diarrhea. Laboratory investigations revealed hypoglycaemia, severe ketoacidosis with an increased anion gap, hyperammonemia (146 µmol/L) and increased excretion of methylmalonic acid in urine (8100 mmol/L creatinine, normal <20 mmol/L creatinine) suggestive of a cobalamin defect. We treated repeatedly with high dosis of hydroxycobalamin, low-protein diet and carnitine. Anamnestic investigation and normal cobalamin levels in blood excluded a dietary cobalamin deficiency. Fibroblast cultures showed a heterozygosity for two different mutations in the MUT-Gene. After resolving the catabolic state the girl still showed muscular hypotonia, dystonia and dysarthria. Brain MRI disclosed an *ischemic stroke* in basal ganglia, usually associated with a mitochondrial dysfunction.
Results: Following treatment the girl improved considerably. After two months of therapy methymalonic acid in urine is still elevated (550 mmol/L creatinine) indicating a cobalmin-nonresponder with a poorer prognosis.
Conclusions: The first presentation of methylmalonyl-CoA mutase deficiency in previously apparently healthy children could lead to the wrong diagnosis of a gastroenteritis. Severe metabolic derailment can cause an ischemic stroke with severe neurological sequelae. Early diagnosis and prompt administration of hydroxycobalamin is related to a better prognosis.



Diffusion/perfusion-weighted MRI Brain: Bilateral symmetric diffusion restriction in the globus pallidus consistent with ischemia.

Figure 1



Metabolic interrelationships of methylmalonic acid. mutase, methylmalonyl-CoA mutase; AdoCbl, adenosylcobalamin (cobalamin-cofactor); Ligase, succinate-CoA ligase; ADP, adenosine diphosphate; ATP, adenosine triphosphate; GDP, guanosine diphosphate; GTP, guanosine triphosphate. J Inherit Metab Dis (2008) 31:350–360, B. Fowler & J. V. Leonard & M. R. Baumgartner.

Figure 2

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Development of the Swiss-DRG Version 1.0 to 3.0 in three independent children's hospitals of Switzerland since introduction of the case-based compensation – an AllKidS study

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AllKidS (Allianz Kinderspitäler der Schweiz) founded in 2009 is an umbrella organisation of three independent children's hospitals of Switzerland (Ostschweizerisches Kinderspital St.Gallen, Kinderspital Zürich, Universitäts-Kinderspital beider Basel UKBB) in order to maintain and develop the health care system for children facing the case-based compensation of Swiss-DRG introduced in 2012.
AllKidS collected and analyzed the case data of 2012 and 2013 of all three independent children's hospitals and grouped them with the Swiss-DRG Grouper Version 1.0, 2.0, 3.0. We compared the MDC Groups over time (V 1.0-V3.0) and among institutions.
We interpreted the results and identified topics in need of further developement of tarifal structure for hospital treatment in children's hospitals.

P219

Cytochrome P450 2C19 genotyping could predict the efficacy of voriconazole treatment in children: two case reports

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Introduction: Voriconazole is used for prophylaxis and treatment of invasive fungal infections in immunocompromised individuals and has a narrow therapeutic margin. There is a clear relation between higher voriconazole plasmatic concentrations and rate of efficacy but variability in pharmacokinetics of voriconazole is large (60–70%). Voriconazole is metabolized by the cytochrome P450 (CYP) system, including CYP2C19. CYP2C19 is subject to genetic polymorphism, with the existence of "extensive", "poor" and "ultrarapid metabolizers", which impact on the pharmacokinetics of voriconazole. Subjects carrying CYP2C19*17 allele, i.e. "ultrarapid metabolizers", are frequent (observed frequency is about 20% in caucasian populations) and may be at higher risk of subtherapeutic voriconazole concentration and treatment failure.
Case report: 1) A 4 year old boy with a juvenile myelomonocytic leukemia and bone marrow transplant was treated with voriconazole 340 mg/d, but presented persistant subtherapeutic voriconazole concentrations (0.2 mg/l, 0.25 mg/l).
2) A 4 year old girl treated for pre-B LLA presented a pulmonary aspergilloma. Oral voriconazole was started at 7 mg/kg and therapeutic drug monitoring showed suboptimal concentration (0.4mg/l).
In these two cases, there was no drug interaction. Genotyping showed a CYP2C19*1/*17 genotype.

Discussion: These cases stress the value of genotype, in case of suboptimal serum concentrations despite good treatment adherence and without drug-drug interactions. The demonstration of a CYP2C19*1/*17 genotype allowed dose adjustment to achieve concentrations within the therapeutic range and favor the predictive value of genotyping in these clinical settings.
Conclusion: Genotyping tests are a first step towards an individualized prescription, which are of particular value in situations where subtherapeutic plasma concentrations are linked to treatment failure.

P220

BrainStim – efficacy of a computerized working memory training in patients with anorexia nervosa restrictive subtype

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Introduction: Patients with anorexia nervosa restrictive subtype (AN-R) often present with rigid and detail focused thinking, key features, which seem to be associated with malfunctioning in the domain of working memory. It is hypothesized that these factors contribute to the maintenance of the illness. Thus, general working memory improvement may have favorable effects on overall clinical outcome. This study will investigate the efficacy of a computerized intervention tool (*BrainStim*) targeted on working memory in patients with AN-R.
Methods: Cognitive performance will be assessed using the *RavelloProfile (RP)* and additional tests to investigate working memory functioning (Digit span and Corsi blocks, N-back (2-, 3-back) and VLMT), SDMT and PASAT before and after the cognitive intervention. Stability of effects on working memory functioning over time will be checked in a 6 months follow-up. The *BrainStim* training procedure will last 4 weeks with 16 sessions of 45 minutes duration.
Results: Data collection is still ongoing. So far we tested 18 women with AN-R, 9 in the training group and 9 without training. Almost all subjects improved significantly over time during training indicating a strong learning effect. This effect was transferred into neuropsychological outcomes after training including improvement on the Y-BOCS obsession subscale, EDE-Q shape concern, PASAT, VLMT 1-5, Corsi blocks backward, 2-back reaction time and accuracy, Rey Delayed Recall, and Tower of London test. Five out of these 9 AN-R were examined 6 months later and showed improved outcomes on the EDE-Q restraint subscale, Y-BOCS obsession and compulsive subscale, and the Corsi Blocks backward.
Conclusions: Since all participants considerably improved during training and in several neuropsychological outcome measures immediately after the training the efficacy of our intervention could be proven. Whether these effects will last over a period of 6 months and whether training effects will positively influence the general therapeutic setting will be further analyzed.

P221

Micropenis and hypoparathyreoidism in CHARGE syndrome: a prismatic case of an overlapping syndrome

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Introduction: CHARGE Syndrome is a congenital anomaly syndrome consisting of the dominant features coloboma, heart defects, choanal atresia, retarded growth and developement, anomalies of genito-urinary tract and ears and/or deafness. Since the identification of the causative gene CHD7 on chromosome 8, which encodes a chromodomain helicase DNA binding protein 7, in 2004, a wide spectrum of associated features has emerged and clinical characteristics overlap with other syndromes such as DiGeorge, Kallmann and Barakat (HDR) syndromes.
Case: We report a case of a term newborn who presented with penis hypoplasia and hypocalcemia as signs of hypogonadotropic hypogonadism and hypoparathyroidism. Despite this early manifestations, a CHARGE syndrome was only diagnosed after identification of further characteristics, namely Fallot-type heart defect, retinal coloboma, facial dysmorphic features with ear anomaly, dysphagia as manifestation of cranial nerve dysfunction and vesico-urethral reflux complicated by urinary tract infections. Additional endocrine findings raised suspicion for hypothyroidism and growth hormone deficiency. Diagnosis was confirmed by a heterozygous CHD 7 mutation. The course was complicated by an immunodeficiency syndrome.
Discussion and Conclusion: Genital hypoplasia is a frequent feature in patients with CHD7 mutation and, if associated with other anomalies as dysmorphic ears, CHARGE syndrome should be considered. Primary hypoparathyreoidism and thymus dysfunction are increasingly recognised in CHARGE syndrome and mark the overlap with DiGeorge-syndrome. As CHD 7 is involved in the organization of chromatin and contributes to time- and tissue-specific regulation of the expression of several genes during embryonic development, there is a wide spectrum of clinical manifestations with variable phenotype.

P222

Kinderpsychologie anstatt Kinderanästhesie

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Einleitung: Krebskranke Kinder erhalten im Rahmen ihrer Therapie und Nachsorge häufig eine CT-, MRT oder PET-Untersuchung. Bei jüngeren Kindern werden diese Untersuchungen in der Regel in Kurznarkose durchgeführt. Aufgrund positiven Erfahrungen in der Radiotherapie (vergl. Radiation Onkology, 2008, 4,3:17) wurden onkologisch erkrankte Kinder mit psychologischen Massnahmen dazu befähigt, diese Untersuchungen ohne Kurznarkose zu bewältigen.
Methode: Eltern onkologisch erkrankter Kinder, bei welchen eine CT-, MRI- oder PET Untersuchung durchgeführt werden musste, wurde während eines Jahres das Angebot gemacht, diese Untersuchung mit psychologischer Unterstützung (Psychoedukation, Veranschaulichung des Vorgehens mit Filmen und Bildern, Training der dazu notwendigen Fertigkeiten) und Begleitung ohne Narkose durchzuführen.
Resultate: 13 Kinder zwischen 4–10 Jahren wurden für eine CT-, MRI- oder PET-Untersuchung entsprechend angeleitet. 12 von 13 Kinder konnten ohne Kurznarkose untersucht werden. Die Kinder erleben sich als selbstwirksam, erleben weniger Angst und zeigten ein besseres Krankheits- und Therapieverständnis. Für die Familien resultierte eine Zeitersparnis und weniger Angst vor Nebenwirkungen und Komplikationen durch die Narkose. Für die Klinik bedeutete es weniger organisatorischer Aufwand und weniger Kosten.
Schlussfolgerung: Mittels Psychoedukation und Begleitung der Kinder und Eltern können bei Kindern ab 4 CT-, MRT- und PET-Untersuchungen erfolgreich ohne Narkose durchgeführt werden. Der Aufwand ist klein (zwischen 1 bis 4 Konsultationen bei einer Psychologin). Dieses Vorgehen macht überall da Sinn, wo regelmässig PET, MRI oder CT durchgeführt werden müssen. Die Methode kann auch durch eine entsprechend geschulte Pflegefachfrau erlernt und durchgeführt werden.

P223

Added value of CYP1A2 phenotyping in teenager patients on clozapine: a case report

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Introduction: Clozapine, used in the treatment of refractory schizophrenia, is metabolized into norclozapine by the Cytochrome P450 enzymes (CYP), mainly by the CYP1A2 isoform. Plasma concentration of norclozapine represents on average 50%–70% of the parent drug plasma concentration. There is a 45 times interindividual variability in plasma concentrations for a given dose influenced by age, sex, bodyweight, adherence to treatment as well as CYP1A2 activity. This activity is highly influenced by smoking, due to induction of CYP1A2 by polycyclic hydrocarbons. In case of poor treatment response, measurements of plasma concentrations are recommended and in the presence of a low level distinction between poor adherence and metabolism induction must be done.
Case report: A 15 years old smoker boy was hospitalized because of psychotic disorder resistant to amisulpride and risperidone. Clozapine was introduced, with an initial good clinical response despite concentrations below the reference intervals, but still compatible with the known interindividual variability. The patient was given permission to go home for three days a week. The clinical response decreased together with variable plasma levels and metabolic ratio (MR). The patient pretended to be compliant, but admitted to have a variable tobacco consumption. Plasma levels of clozapine and norclozapine are given in table 1. Measures were done at steady state.

Table 1: Plasma levels of clozapine and norclozapine

			11/06/2013	27/06/2013	04/07/2013
clozapine	ng/ml	250–500	131	72	101
norclozapine	ng/ml		109	38	50
norclozapine/ clozapine (MR)	%		83	53	50

Discussion: There were no concomitant interacting drugs. An increase in tobacco smoking would not explain the decrease in parent compound of the two last measures since the MR did not change accordingly. These lower levels and the poor clinical response would rather to be linked with poor adherence. The hypothesis would be further assessed by phenotyping CYP1A2 activity.
Conclusion: Because of high interindividual metabolic variability, high prevalence of low adherence to treatment and smoking in teenager population, therapeutic drug monitoring is advocated to guide dose adjustment of clozapine. In the presence of low levels, phenotyping of CYP1A2 activity is the next step to rule out poor adherence.

P224

How do children experience a stay in hospital? Translation of a Peruvian book on a study focused on the resilience and patient rights of children in hospital

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Introduction: Children in various different hospitals in Peru were explained their patient rights, and were invited to chose one in particular to illustrate in a picture.
Methods: The study analyzed 330 of these drawings and paintings using different criteria to generate eleven categories, flanked by interviews and observation of the children's playing. The various results were drawn together (Dockett and Perry, 2005) to produce important propositions about children's experiences of hospitalization.
Results: The main quantitative result was that the most popularly chosen right was right no. 1, the right to tender and loving care.
Qualitative results: What became most evident from what the children and youths expressed was how they themselves wanted to be ideally seen as individuals:
– as communicative (the children articulated their personal circumstances in a clear and intelligible manner),
– as active (in comparison to grown-ups, children have a pronounced need to move and for opportunities to play),
– and as participants (the illustrations do not show the children themselves as passive and helpless patients, but as persons actively participating in the events concerning them).
Conclusions: The children articulate their claims to love, to movement and to play as essential needs, whereby the relationship between the child and the hospital staff takes on a crucial role. The children and youths bring with them an innate resilience from their daily worlds that can be reinforced by play and amusement to form a vital part of their treatment and convalescence.

P225

Multidisciplinary management of anorexia nervosa in a regional hospital setting

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Introduction: Anorexia nervosa is associated with numerous general medical complications that are directly attributable to caloric restriction and weight loss. The complications affect most major organ systems and often include physiologic disturbances such as hypotension, bradycardia, hypothermia, and amenorrhea. The typical psychiatric and somatic signs of anorexia nervosa are presented.
For moderate cases of anorexia nervosa we saw the possibility to follow inpatients in a regional hospital setting, even by possibility we can have a conservative
Method: We report our positive experience in the management of some cases of adolescent female with anorexia nervosa in a hospital setting starting from the strictly collaboration between the paedopsychiatric service of the region and the somatic paediatric service.
In our clinical management we promoted the multidimensional approach of anorexia nervosa, the theoretical frame of the psychopathology was based on a psychodynamic reference.
During the meeting the team shared the information about the different psychoeducative activities, school activities, somatic medical and nurse clinical observations.
The adolescent was initially treated in an one per day in-patient psychotherapy setting by a child adolescent junior psychiatrist, himself supervised one per week by a senior colleague.
The parents were invited in one per week multidisciplinary meetings and informed concerning the somatic and psychological evolution.
During the in-patient period school courses were introduced after the critical asthenic period, held in the in-patient unit in the first step by a teacher.
After team meeting with regular school teachers, the adolescent began going at her normal school, step by step to higher school timetable levels according to her psychological and somatic wellbeing.
Every change step by step was discussed and searched by a shared agreement with the adolescent, teachers, parents ,senior paediatrician and individual and senior family therapist.
Family treatment with the adolescent, the parents, and when possible with siblings were also organised in the in-patient setting one per week , as co-therapist were designed the child adolescent senior psychiatric consultant and the senior paediatrician.
Discussion: Our multidisciplinary case management is effective in a regional hospital for moderate cases of anorexia nervosa.

P226

Le diagnostic communautaire participatif: une étape nécessaire pour un appui extérieur visant à une amélioration durable de la santé maternelle, néonatale et infantile dans un district rural de Haïti

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Introduction: La mortalité maternelle et infantile en Haïti est l'une des plus élevées au niveau international, hors Afrique sub-saharienne. Malgré la présence de nombreuses ONG, suite au séisme de 2010 en particulier dans les zones rurales, cette situation ne s'est guère améliorée. Pour mieux répondre à cette problématique, dans une perspective de pérennité et pour redéfinir leur stratégie dans la zone goâvienne où elle travaille depuis plus de quinze ans, MdM-CH s'est alliée à EDM pour organiser un diagnostic communautaire participatif dans le cadre d'une approche Individus Familles et Communautés (OMS).
Méthode: Entre avril et juin 2013, des tables rondes ont été organisées dans les différentes communautés. Elles étaient dirigées par des leaders locaux, formés spécialement pour l'occasion. Elles ont réuni des femmes enceintes ou mères d'enfants en bas âges et leur mari, des grand-mères, des prestataires de soins et des accoucheuses traditionnelles (AT). Quatre champs d'activité ont été analysés : 1) Comment développer les aptitudes ? 2) Comment développer les connaissances ? 3) Comment améliorer les liens entre les acteurs de santé ? 4) Comment améliorer la qualité des soins au bénéfice de la santé maternelle, néonatale et infantile.
Résultats: Il en est ressorti une méconnaissance de l'importance de consultations prénatales et des signes de complications courants des grossesses, un manque de considération par les autres acteurs de santé des AT, qui pourtant accompagnent l'essentiel des naissances ainsi que d'importants problèmes d'accessibilité aux centres de santé et de qualité des soins qui y sont offerts. Les structures de santé sont ainsi devenues le lieu de «dernier recours» avec un taux de naissances institutionnelles avoisinant les 20% au mieux.
Conclusion: Cette approche a permis de redéfinir une partie des actions de MDM-CH en se focalisant davantage sur les besoins des communautés et en renforçant le système référentiel des patients. En redynamisant le mouvement social MdM-CH et EdM espèrent placer la population au cœur de la redéfinition d'un système de santé toujours en faillite.

P227

The case of a dangerous fish

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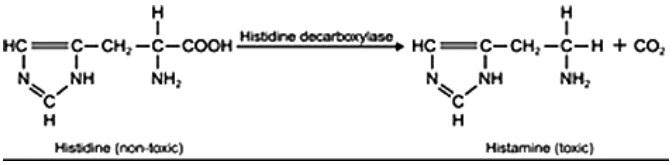
Histamine fish poisoning is an acute histamine intoxication caused by consumption of decomposed histidine-rich foods such as certain fish. We report the case of a family presenting to our emergency department after ingestion of spoiled tuna; two children presented to our emergency department with facial flushing, cutaneous rash, burning sensation of the tongue and sore throat. Moreover, one child presented with headache and abdominal pain. A diagnosis of scombroid fish poisoning was established based on the history and clinical presentation. An extensive literature search was performed on Pubmed by using keywords/MeSH terms such as "histamine fish poisoning", "fish poisoning" and "scombroidosis".
The underlying mechanism is the decarboxylation of histidine by the enzyme histidine decarboxylase occurring mostly in the Scombridae fish family when inappropriately stored. The diagnosis of intoxication is history and clinically based. In the vast majority of cases it is a mostly benign, self-limited condition requiring supportive care and observation. In Switzerland, histamine fish poisoning appears to be a rare event. From 1966 to 1991, The Swiss Toxicology Center recorded 27 physician reported cases, 18 of which were consistent with scombroid fish poisoning. Since 1997, 34 cases including 2 children were reported to the Center (age range; 12–65, mean age: 33). The Center acknowledges a possible underreporting of cases.



We believe that awareness among pediatricians regarding food borne poisoning is important to differentiate these cases from allergic conditions. Furthermore case reporting remains essential to ensure optimal public health intervention.

P228

Acute hemorrhagic edema of childhood: a vasculitis more impressive than serious

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Introduction: Acute hemorrhagic edema of childhood (AHEC) is a fairly rare clinical entity first described by Snow in 1913. Together with Henoch-Schoenlein purpura (HSP), they both fall within the scope leucoclastic vasculitis, although they differ by many points. It impressive clinical presentation, mimicking HSP or meningococcal purpura, calls for a proper and quick diagnosis, in order to avoid complications or unnecessary therapeutic.
Methods and results: We report the case of a previously healthy 3-year-old boy, who presents with high fever, in no distress. At the time of admission, he had generalized hives, which increased in the next 48 hours, together with bilateral swelling of the eyelids. Soon after admission, the skin lesions clearly became purpuric, predominantly in the lower limbs, associated with edema. Clinical follow-up showed a spontaneous improvement of the skin lesion within days. Blood cultures remained sterile, and the complete blood count was consistent with a viral infection, with normal leucocytes and transiently low platelets.
Conclusion: AHEC affects infants aged between 4 months and 2 years. Recent infection or immunization is found in ¾ of the cases. There is a striking difference between the well-preserved condition of the child, and the impressive skin lesions, characterized by inflammatory edema and ecchymotic purpura (often roundel aspect) predominantly on the upper and lower limbs and face. Renal involvement in AHEC (microscopic hematuria, proteinuria or a discrete elevation of blood urea nitrogen) is always transient and visceral involvement is rare, which distinguish it from HSP. Spontaneous and complete remission occurred in 1–3 weeks. The lack of reported complications suggests that the AHEC harbours a benign course.

Posters SGKJPP / Posters SSPPEA

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Technologiefolgenkritik und Cyberphilosophie – mediensoziologische Aspekte des pathologischen Internetgebrauchs

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Fragestellung: In der publizistischen und medienwissenschaftlichen Fachöffentlichkeit werden die Auswirkungen der modernen Medien und der neuen Internetapplikationen intensiv und kontrovers diskutiert. Multiple Publikationen, insbesondere von ehemaligen oder aktuellen Entwicklern von Technologie, erscheinen vor allem im angloamerikanischen Raum. In wieweit diese öffentliche Diskussion für die Konzeptualisierung von Diagnose- und Therapiestrategien bei pathologischem Internet- und Mediengebrauch nützlich gemacht werden kann, ist zu untersuchen.
Methodik: Die nach Verkaufszahlen wichtigsten 20 Publikationen zum Thema moderne Medien- und Internetentwicklung der letzten 10 Jahre wurden darauf hin untersucht, in wieweit Trends, Bedrohungsszenarien aber auch Lösungsansätze für die sog. digitale Revolution gefunden werden können. Hierzu erfolgte eine qualitative Inhaltsanalyse mit entsprechender Kategorienbildung.
Ergebnisse: Es zeigen sich drei grosse Linien in der aktuellen medienwissenschaftlichen Literatur:
1. die technikfreundlichen Publikationen
2. die technikkritischen Publikationen
3. Publikationen, die versuchen einen Ausgleich herzustellen.
Während die erste Kategorie primär die Nutzung sozialer Medien und der neuen kommunikativen und interaktionellen Chancen in den Vordergrund stellt, finden sich bei der zweiten Kategorie verschiedene Autoren, die insbesondere die soziale Isolation, die mangelnde Informationshoheit und den Verlust von Individualrechten in den Vordergrund stellen. Die dritte Kategorie, die eindeutig unterrepräsentiert ist, versucht für den interessierten Laien die Vor- und Nachteile intensiver Internet- und Mediennutzung abzuwiegen, ohne zu einem abschliessenden Urteil zu kommen oder Risikogruppen zu benennen.Insgesamt ist der Anteil an europäischer Literatur zu diesem Thema ausgesprochen gering, die entscheidenden Publikationen kommen fast ausnahmslos aus dem amerikanischen Raum und bilden die dortige gesellschaftliche Realität ab.
Diskussion: Die weltweit geführte medienkritische bzw. positive Diskussion bildet sich in umfangreicher und teilweise intensiv recherchierter und aufwendiger publizistischer Fachliteratur ab. Insbesondere die technikkritischen Autoren vertreten einzelne interessante Konzepte, die das Verständnis vom Prozess der Medienabhängigkeit bei Kindern und Jugendlichen erleichtern und auch für die Therapieplanung sinnvoll genutzt werden könnten.

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Personality specific description of juvenile sex offenders: standardisation of the adolescent sex offender assessment pack (ASAP) using a school sample

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Introduction: An accurate diagnostic of juvenile sex offenders has to take both personality traits (i.e. self-esteem, impulsivity or empathy) and the act (i.e. factual circumstances, attitude towards the act or approval of violence) into account. The ASAP assessed personality traits have been deemed relevant for the development and maintenance of sexually abusive behaviour based on the current state of research (Van Outsem, Beckett, Bullens, Vermeiren, Van Horn, & Doreleijers, 2006). However, to be able to adequately interpret the ASAP results of sex offenders, it is necessary to know which scores are deviant from the norm.
Method: A representative sample (**N** = 464, **M** = 15.21, **SD** = 1, **Range** = 13–18 years) of pupils in urban and rural schools was questioned.
Result: The aim of our study was to develop norm tables of German speaking pupils in Switzerland. Since sex was found to be a significant factor (**Pillai-Spur** = .08, **F** = 2.85 (10), **p** < .01), one norm table for each sex was necessary.
Conclusions: This is the first time that the trait-specific part of the ASAP was standardized using a Swiss sample. The results and the tables deviate in almost all scales from the former available standardized data from England. In addition, our study was the first to assess female subjects. Although the impact of sex difference was assumed to have a significant impact on the results of the ASAP, it could not be proved until now.

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Phenotyping healthy and impaired personality development in adolescence

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In the DSM-5 Section III the "Alternative DSM-5 Model for Personality Disorders" was proposed. This hybrid model combines the categorical and the dimensional approach to understand and describe personality disorders. The dimensional approach includes the assessment of a "Level of Personality Functioning-Scale" (LoPF-Scale, rated by the therapist) as an overall measure of the severity of personality dysfunction. The LoPF model seperates two areas: "self-related" and "social-related", both subdivided in two further dimensions of personality functioning. These four personality features were designed to summarize the core impairments by which personality disorders (PD) can be characterized fundamentally. The "self-related" domain contains "identity" and "self-direction". The "social-related" domain contains "intimacy (behavior in close relationship, attachment)" and "empathy (behavior in general social relationships, cooperativity)". As identity development is regarded as the key feature in emerging personality disorders in adolescence, we developed the self-report questionnaire AIDA (Assessment of Identity Development in Adolescence; Goth et al., 2012) as a first step to establish a reliable, valid, and time-effective inventory which represents a new dimensional concept of healthy and impaired personalilty development. In a next step we enlarged the AIDA to a complete assessment tool that covers all four areas of personality functioning (LoPF-QA). It was assessed in school and clinical samples. AIDA showed good scale reliabilities and a remarkable diagnostic validity with a differentiation between PD-patients and students ~2 standard deviations, proving adequacy of the model and the technique of self-report. Results concerning the psychometric property of the LoPF-QA will be presented.

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Can eye-tracking represent a reliable tool to aid the diagnosis of autism spectrum disorders in toddlers and children?

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Background: Autism spectrum disorders (ASD) affect 1 on 88 children. As early intensive interventions dramatically improve the cognitive and social outcome of affected people, diagnosis should be made as early as possible. Abnormal patterns of visual explorations in children with autism represent a promising tool to help identifying children at risk for autism.
Methods: Twenty-seven children aged 3.5 ± 1.8 y.o. (range: 1.0–8.1) were enrolled in the current eye-tracking study. Seventeen children had a diagnostic of ASD confirmed with the ADOS and ADI scales. Ten typically developing children had no autistic symptoms. We designed a one-minute video (inspired from Pierce et al., Arch Gen Psychiatry 2011), where biological motion and non-biological motion are simultaneously presented on both sides of the a Tobii T60XL eye-tracking screen, and measured visual preference.
Results: The children with ASD spent significantly less time looking at biological motion (40.1 ± 4.1%, mean ± SEM) than typically developing children (69.0 ± 5,2%, $t = 4.29$, $p < 0.0002$). The optimal number of false negative (1) and false positive (3) was observed when setting the threshold at 66% of the time spent looking at biological motion, yielding a sensitivity of 84.2% and specificity of 87.5% in autism screening.
Conclusions: This simple, one-minute, eye-tracking task is able to provide high accuracy in identifying ASD children. These preliminary results strengthen the hypothesis that individuals with autism lack interest and orientation to social stimuli (Chevallier et al., Trends Cogn Sci 2012). In total, our results provide strong support for the use of eye-tracking technology to identify children at risk with autism.