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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 Apert 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 releases 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 was introduced. Given up and modifications of flaps, accepting larger skin defects and external fixateurs for soft tissue distraction were introduced. 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.

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 Bern, we identified 136 patients with proven NEC (1977–2010). Median birth weight was 1520 g, median gestational age was 32 weeks and mean age at diagnosis was 12 days ± 11.58%. 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 40% 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.

Validation of a postoperative parental pain assessment tool in a French-speaking patient 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 48 h after surgery. Children age ≥6 filled their own FPS-R. Parents and children 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 48 h 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.

Mind the gap

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Introduction: Between 2009 and 2013 seven newborns were treated for Type III esophageal atresia at the University Children’s Hospital Zürich. Here we present these cases with regard to the therapeutic algorithm.

Methods: Chart review was performed retrospectively. Postnatal diagnosis was based on babygram and tracheobronchoscopy. After confirmation of the Type III 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.
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.

Results: The cumulative follow up on biologics represented 244 years at diagnosis [range 5 months to 15.2 years]. Median duration of immunosuppression from diagnosis to start of the first BA was 18.2 months [range 0.0 to 11.5 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 Methylprednisolone alone. 14 patients were treated with valaciclovir or acyclovir, of whom 6 intravenously. 54% continued IS treatment during varicella/HZ. 14 patients (64%) were treated for juvenile idiopathic arthritis, 1 for a polyarticular 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). 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 heterogeneous, with 4 of the total 16 children showing a complicated course. Thorough assessment of VZV infection 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.

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 randomly assigned 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.

Clinical course and therapeutic 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 the clinical presentation of common pediatric diseases like infections with varicella zoster virus (VZV). There are little data concerning clinical course, potential complications and therapeutic approach in this situation.

Methods: In this retrospective multicentre study, we assessed the clinical course and therapeutic 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 polyarticular 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 Methylprednisolone 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 heterogeneous, with 4 of the total 16 children showing a complicated course. Thorough assessment of VZV infection 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 biological 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.

Methods: To assess drug survival of biological agents (BA) in SJIA patients and to describe reasons for switching or discontinuing a biologic treatment.

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
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 appropriate approach for treatment of SJIA to increase the probability of inactive disease. Inactive disease was less often observed as the only BA. This subpopulation did not significantly differ in baseline characteristics from the group that continued treatment. Asphyxia. The mean midazolam dose was 0.11 ± 0.05 mg/kg. Efficacy and safety as first-line treatment of neonatal seizures. A retrospective study is needed.

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 no intrinsic myorelaxant 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 in 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-asphyxial group, 41% (n = 18) responded to midazolam, 69% (n = 9) to the hemorrhagic group. Intrenasal route was 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, especially 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.
Feasibility of multiple-breath washout in inexperienced preschool children

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Introduction: Multiple breath washout (MBW) has gained increasing interest as a 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 (N2) MBW measurement in preschool children in a setting not experienced with MBW measurement.

Methods: N2-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 N2-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 exclusion of children (n = 12): breath 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 (5.6 ± 0.8 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.

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 together, and the SPQ to assess the level of schizotypal traits, which typically emerge during adolescence and may be altered in ADHD, were applied to a Basel-based cross-sectional study on Empathy Development group: typical adolescents and youths with conduct or autism spectrum disorder.

Results: We first found a lower level of RFQc (Z = –3.25, p = .00) and a higher of RFQc (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 (r(124) = –.216, p < .05) and negative (r(124) = –2.06, 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 into the enhancement of mentalizing skills.

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. In particular, the most current diagnostic scheme (DSM-5) explicitly states that neurophysiological or imaging based biomarkers which might 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 as 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".

References


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

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Introduction: Paediatric kidney transplantation is surgically challenging. Urological 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 transplantsations 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 (UTIs): 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 UTIs 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 urological 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 refluxing transplanted kidneys since this technique was successful in all patients.

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 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.
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: Chromometer 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.

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: 2024 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 = 0.03). 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.

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, FRCmax) at the age of 5 weeks and 6 years. Additionally, at the age of 6 years we measured FRC using plethysmography (FRCpleth). 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) FRCmax was 101 (16.4) mL at the age of 6 years, mean (SD) FRCpleth (n = 86) was 651 (139) mL, and FRCmax (n = 87) was 1094 (198) mL. Regression shows a weak tracking for absolute FRC values (FRCmax, R= 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.

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

![Graph showing prevalence of cough](image-url)
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. 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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Young Researchers’ Day / Clinical Researchers’ Day (Swiss PedNet)

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. The aim of this study is to explore brain neurotransmitters and brain function. The aim of this study is to explore brain neurotransmitters and brain function.

Methods: A multi-modal neuroimaging study was conducted combining Proton Magnetic Resonance Spectroscopy (1H 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 BSD population (GABA, r = -0.52, p < 0.05, 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 (Nacc = 16; Nrec = 18).

Conclusion: We found a high prevalence of GABA in our study cohort (363/1,325 samples tested positive for GABA), associated with a seasonal effect mainly during autumn months and older siblings. Among infants with HSV detection, 51% were clinically asymptomatic. Respiratory symptoms during HSV infections were less likely during the first three months of life. A maternal history of atopy was independently associated with less symptoms during HSV infection.

Results: 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. Among infants with HSV detection, 51% were clinically asymptomatic. Respiratory symptoms during HSV infections were less likely during the first three months of life. A maternal history of atopy was independently associated with less symptoms during HSV infection.

Conclusions: HSV is highly prevalent in unselected term-born infants during the first year of life. While environmental factors have significant impact on HSV prevalence, individual co-factors are associated with respiratory morbidity during HSV infections. These findings shed new light on risk factors contributing to HSV-associated infant morbidity and will help to improve treatment and prevention strategies.

Authors: N. Regamey and P. Latzin contributed to this abstract equally.

Clarification and goal-attainment in child welfare and juvenile justice institutions: Results of the MAZ.-study and introduction to EQUAL S
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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

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.

Acknowledgement: National Institute of Mental Health (R01MH073998 – C.M.M.), National Institute on Drug Abuse (RC2DA029475 – D.K. and J.A.F.) and Swiss National Science Foundation (PBGE3P_139835 – A.A.L.).
Trials-R-Therapie als Thrapeimodul in der stationären Psychotherapie von Adoleszenten – Klinischer Einsatz von Rhythm-Rhyme-Recording-Therapy

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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 both by 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 group therapy 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 group therapy 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 group therapy 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.

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 children and their families.

Methods: 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.
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 prospectively 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 micturition and 15% showed a detrusor-sphincter discoordination.

An organic lesion was diagnosed in 4.2%. 3% showed a no dilatating reflux. UTI appeared in 24%. An obstruction 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 20%.

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%.

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 major 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.

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

Empiric Treatment

Empiric and definitive 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 ρ<0.01 for comparison between all groups (Fisher’s exact test).

Empiric Treatment | CA | HCA | HA
<table>
<thead>
<tr>
<th></th>
<th></th>
<th></th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td>Carbapenems</td>
<td>1.9%</td>
<td>2.7%</td>
<td>10.8%</td>
</tr>
<tr>
<td>Glycopeptides</td>
<td>0.7%</td>
<td>2.8%</td>
<td>4.1%</td>
</tr>
<tr>
<td>Both</td>
<td>0</td>
<td>7.2%</td>
<td>16.3%</td>
</tr>
</tbody>
</table>

Definitive Treatment

Empiric and definitive exposures to 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 empirical treatment. Swiss clinicians identify HCA sepsis as different from CA in terms of empiric treatment decisions.

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%). 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 10%). 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.

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.
Giant cervical lymphangioma – ENTRY by EXIT

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Introduction: Lymphangiomas are benign congenital tumors, involving the head and the neck, 90% occurring 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 may be used as an early indicator for BPD. Method: An observational study was performed at our pediatric ED to assess the safety of N2O 70% combined with 30% O2 application for short painful interventions. 341 patients were included. No fasting times were required. We recorded all administered drugs, the duration of N2O 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.

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). 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.

Conclusion: 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.
Case 2: 2 months old infant (‘2013), first pylonephritis 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 urethroscopy 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.

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 labia minora and labia majora. Pregnancy and spontaneous birth were uneventful, 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 pyrrierpigenisation, no synchia of the labia minora and meatus urthearie and anogenital distance were normal. Serum androgens on 1st and 4th day of life were in the normal range of female newborns, as were adenal steroid precursors (17-OH-progesteron and androstendion). The ultrasound examination presented a normal uterus and ovaries and normal intraabdominal state without tumor. The follow-up for 10 months showed a normalization of labia minora, 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 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 involvement of the clitoris, there were no signs of virilisation, no obstetric history of another disease.

Reference

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 detorsion of the cecum, the periphery 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 toward bowel necrosis. 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 of CV without ceceectomy is an option, but associated with the risk of recurrence.

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 anamnestic 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. Radiographic studies with and without contrast were inconclusive, as well as an esophagastroduodenoscopy. 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 duodenoejunostomy we observed an uneventful follow-up.

Conclusion: Duodenal 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.

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 is 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.

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 from a ankle sprain. On physical examination 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.
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 osteochondral fractures are rare but potentially debilitating. Intra-articular fragments in the ankle should always be looked for and fixed surgically when found.

**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 obstruction as the triggering cause, Hirschsprung’s disease was ruled out by histology. The child improved quickly and started 72h after laparotomy was performed immediately. To exclude intestinal perforation, laparotomy was performed immediately.

**Results:** Surprisingly, a perforated Meckel diverticulum (MD) was found, resected and histologically confirmed. To exclude intestinal perforation, laparotomy was performed immediately. Due to suspected intestinal perforation, laparotomy was performed immediately. 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.

**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; mature 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 overtreatment, 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.
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 all patients ≥2 nights, 7 females, 1 male, 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 hypoglycaemia and therefore optimising patient instruction and treatment.

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.

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


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 (35.6%) 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.

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 Europe by 2015. Risk allows timely immunization. Switzerland will remain of central importance to eliminate measles in Europe by 2015.

Introduction: Measles encephalitis (n = 3). One child each developed acute pneumonia with oxygen requirement (n = 1). One child each developed acute pneumonia with oxygen requirement (n = 2). One child each developed acute pneumonia with oxygen requirement (n = 2). One child each developed acute pneumonia with oxygen requirement (n = 3). One child each developed acute pneumonia with oxygen requirement (n = 3).

Main complications were pneumonia with oxygen requirement (n = 19), bacterial infections of the base of the skull (n = 2) and 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, in the canton Lucerne between 2006 and 2009 were included. Course, complications, immunization rates and costs of the hospitalised 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 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.
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 ρ, −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.
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 developed 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.

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 thoracoscopic in adults.

Methods: 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 thoracoscopic 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 Claus-Debern-Horner syndrome. 28% of postoperative transient hypohidrosis. 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 exercise , 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.

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, thoracoscopic 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.
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 the Germanic (1st ZLS, 1955-77) and Romanian region (Sempé1979), and for BMI only, German references (Kromeyer 2001). The new standards for children aged 0–5 years include data of healthy breastfed 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 (<0.01 cm differences, n = 20), with the exception of the measuring rods used in infants, producing errors between +0.24 and –0.7 cm.

8007 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 = 0.95 and 0.93) 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 stunted 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 stunted 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 stunted 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.

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 78.3 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?).
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, continuing reduction of obesity in 70% of children until 1 year after the end of therapy. Since physical inactivity, 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, 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.

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) significantly differed compared to obese MIS children in MGP (11.2 ± 2.1 years; 44% girls; 62% morbidly obese with BMI >99.5th percentile), MGP and MIS therapy, obesity was significantly reduced (ΔBMI-SDS = −0.19 and −0.33, respectively, p <0.000). Morbidly obese children showed significantly higher in mental health difficulties compared to less obese ones (p = 0.018). At T2, an improvement in total SDS score was more prominent, the higher the initial score was. Diet composition was neither associated with BMI nor the children’s 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.

Anti-exocrine pancreatic and proteinase-3 antineutrophil cytoplasmatic 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 cytoplasmatic antibodies (pANCA) is commonly used in patients with inflammatory bowel disease (IBD) to help distinguish between Ulcerative Colitis (UC) and Crohn’s Disease (CD). However, there is some overlap, reducing their specificity. Recent studies postulated that exocrine pancreatic autoantibodies (PAB) and anti-proteinase-3 antineutrophil cytoplasmatic 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 10.1y, 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 15 UC-patients tested for PAB was positive. PAB was detectable in 3 CD-patients who were negative for ASCA.

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.

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 6 months. Therefore, EEN was started with good local response after 6 weeks and a weight gain of 9 kg over 3 months.

Conclusions: 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.

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%, p <0.0001).

Conclusions: In a single center prospective longitudinal cohort study, 52 children were treated...
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 < 0.00001). 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).

P14
Herlyn-Werner-Wunderlich syndrome, when embryology leads to uncommon abdominal pain
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Introduction: Obstructed hemivagina with ipsilateral renal agenesis associated to didephys 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 dysmenorrhea, 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 peribdominal and suprapubic abdominal palpation, no signs of peritonitis and no palpable mass. The external genitalia was normal.

The abdominal ultrasonound showed a pelvic mass precised by abdominal MRI which demonstrated a uterus didephys, 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 by inflammatory infection. Kept in mind, an 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.

P155
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 purpurae and an exanthema. She was pancytopenic and febrile, so she was treated for meningoencephalitis and sepsis in our ICU. With diagnosis of Mullerian duct anomalies. After menarche a hematocolpos develops with subsequent development of dysmenorrhea, abdominal pain and pelvic mass, sometimes associated with urinary retention and/or constipation.

Conclusion: 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).

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 propythiouracil. 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 purpura, dyspnea, nausea and joint-swelling. She had been treated with carbimazole (1.15 mg/kg/d) and propylthiouracil since 24 days. 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 plummeting with application of high dosed iodine before thyroidectomy is effective within few days, if required.
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 administered, 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-exposure.

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

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Introduction: Fertility counselling and preservation in post-pubertal patients remains a big challenge for the medical team. Paediatric oncologists should be aware of this topic.

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 oncological 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.

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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 conservative management. In the third 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 near total obstruction of the 3rd duodenum. A nasojugal tube was placed distal to the obstruction.
Watermelon seed rectal bezoar in a 9-year-old boy

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Introduction: Watermelon seed rectal bezoar is a very rare cause of rectal obstruction and was described only in two pediatric cases up until now.

Case report: A 9-year-old boy 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 unsalted sunflower seeds. Disimpaction by digital evacuation was performed under general anaesthesia followed by saline irrigation. Anoscopy showed a circumferential ulcer of the anal canal, coated with a fibrin exudate with small areas of necrosis. The tonsus of the anal sphincter improved over the following weeks and the boy subsequently required continence.

Conclusion: Watermelon seeds are very popular but are 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 unsalted sunflower seeds. Disimpaction by digital evacuation was performed under general anaesthesia followed by saline irrigation. Anoscopy showed a circumferential ulcer of the anal canal, coated with a fibrin exudate with small areas of necrosis. The tonsus of the anal sphincter improved over the following weeks and the boy subsequently required continence.
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 (1340 (±107) kCal/d vs. 1340 (±124) kCal/d, respectively; p: NS).

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.

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 subfebrile 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, pCO2: 22.4 mm Hg, HCO3: 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 underestimated 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.

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 2003, 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 12.10 IU/L, free thyroxine 9.3 pmol/L), high levels of antibodies to thyroglobulin thyroid peroxidase and hypocholesterinaemia 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.
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?f=1) the description of testicular torsion with detailed instructions about closed detorsion manipulation,... and performed it correctly (rotating the right testicle antockwise) 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 introduce a medical website (http://www.allodocteurs.fr/actualite-sante-qu-est-ce-qu-une-torsion-de-testicule–1030.asp?f=1), can be used as diagnostic tool in the emergency department.

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 abdomen. One neonate needed brief bag/mask-ventilation with complete recovery. The other three required full resuscitation including chest-compressions. These infants were severely acidic at 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, renal diseases, 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. Possible 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.
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 bpm, swelling of lips, tongue, and periordial tissue. He was given norepinephrine (10 ug/kg i.v.), antivenin (Viperafav™ 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, periordial swelling and shock with systolic pressure of 60 mm Hg, heart rate 93 bpm, and generalised oedema. She required norepinephrine (6.4 ug/kg i.v. followed by 0.1 ug/kg/h during 16 hours). With 1 vial of antivenin (Viperafav™) 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 viper bites [1]. However, acute anaphylactic-like reactions are described in some first-bite victims [1]. The pathophysiology is unclear, but a direct autoantimicrobial 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.

References

The benefits of nebulised PTX in EP infants at risk of CLD need to be confirmed in definitive large trials.

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 the administration of nebulised pentoxifylline (PTX) in reducing the duration of oxygen supplementation in extremely preterm (<28 gestational age, GA) 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 normal saline (n = 40). 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 0.28, 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.
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 renal growth, therefore, our findings provide new molecular insight into the pathogenesis of altered renal development in chronically ventilated preterm neonates.

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**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 report on 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. They 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 88% 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.

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**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). Weaning 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 report on 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. They 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 88% 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.

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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 immediate surgery followed with the diagnosis of necrotizing fasciitis.

**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 ventilation 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 a decrease in 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 Pountales 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.

**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 postpartum, this was the case for 43% respectively 41% at 4 months. At 6 months, only 50% respectively 47% of mothers still breastfed.

**Conclusions:** The current high breastfeeding rates at discharge from maternities confirm the validity of the BFH at large. We believe new measures have to focus now on further support of breastfeeding in the first months postpartum.
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 laryngotraceoscopy 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 epiglottogeal 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.

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, hepatitisplenomegaly, 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 severe hepatosplenomegaly. 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.
Dacryocystitis following a congenital dacryocystocele

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Introduction: Acute dacryocystitis may be a complication of dacryocystocele, a rare variant of nasolacrimal 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 occurring 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 ophthalmologist with a good response.

Discussion: Dacryocystoceles are thought 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 ophthalmologists 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 referred to an ophthalmologist before occurring of complications (dacryocystitis, cellulitis, meningitis or brain abscess, systemic infection…).

A conservative approach can be initially attempted for non complicated dacryocystocele; probing can be performed if medical management fails.

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 sialorrhoea. 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 tape and flexible plastic part, when ingested, can produce severe symptoms, with possible misleading free intervals and significant complications.

This case emphasise the necessity of endoscopic investigations of persistent aerodigestive symptoms.

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-acetylmorphin (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=2-25 min), which is metabolized into morphine (T1/2=2.3h). 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.
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.

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.

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 asent 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 posterolateral 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.
Unexpected diagnoses in cyanotic newborns

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-tomometry revealed decreased p50-values in both cases, compatible with a low-affinity-hemoglobinopathy.

In case 1, an abberant 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 was noted. Chest X-ray and echocardiography were normal. Given the association of marked intravuterine growth retardation and cyanosis, we propose a flow-sheet of differential diagnosis.

Conclusion: Cyanosis caused by hemoglobin variants should always included as a differential diagnosis in newborns. P50 values and oxygen saturation should be measured early in the clinical course.

Discussion: Neonatal cyanosis is a challenging diagnostic problem. A careful history and physical examination are crucial. Additional tests such as blood gas analysis, pulse oximetry, and echocardiography may be necessary. Early intervention can prevent serious complications and improve outcomes.

Cleft palate: isolated or syndromic?

Introduction: Cleft palate is a common congenital malformation. It is usually associated with various syndromes. The aim of this study was to report two cases of isolated cleft palate and to discuss the differential diagnosis.

Case report: Case 1: A 4-month-old boy was referred to our hospital due to a unilateral cleft palate. Physical examination revealed a bilateral cleft lip and palate. The patient was otherwise healthy and the family history was unremarkable. Case 2: A 6-month-old girl was referred to our hospital due to a bilateral cleft palate. Physical examination revealed a bilateral cleft lip and palate. The patient was otherwise healthy and the family history was unremarkable.

Conclusion: Isolated cleft palate can be associated with various syndromes, including chromosomal abnormalities. Early diagnosis and appropriate management are crucial for the best possible outcome.

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

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 discontinued.

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.

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 specialty (NTE).

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

Introduction: Serious sequelae after ingestion of a lithium disc battery are rarely described in the literature. We report the case of a 20 months old girl who ingested a lithium disc battery and developed serious complications.

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 oesophageal 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 6 months of conservative treatment (nasal oxygen and gastrostomy) 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 imaption in the oesophaeus 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.

Graph 1

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 analysed using multivariable linear regression. We quantified the strength of long-range correlations of body temperature in 103 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).

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.

Table 1: Demographic data of study participants

<table>
<thead>
<tr>
<th>N = 147 infants (62 female, 85 male)</th>
<th>Mean</th>
<th>Min</th>
<th>Max</th>
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<tr>
<td>Gestational age at birth (weeks)</td>
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<td>Birthweight (g)</td>
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<tr>
<td>Weight at study date (g)</td>
<td>4239</td>
<td>2640</td>
<td>6800</td>
</tr>
<tr>
<td>Z-Score of weight at study date</td>
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<td>-3.769</td>
<td>3.94</td>
</tr>
<tr>
<td>Postconceptional age (weeks)</td>
<td>44.83</td>
<td>41.86</td>
<td>51.86</td>
</tr>
</tbody>
</table>

**P155**

Early infant form of galactosialidosis – a case report
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Introduction: Galactosialidosis 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. Echocardiographic 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) and 36 preterm infants with BPD (PT+BPD) a total of 244 sighs were examined. For demographic information of study participants see Tab 1.

![Graph 1](image)

**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.
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 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 postdural 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 (SpO2) <90%, SpO2 <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 time taken for SpO2 recording in a subgroup was 5 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.

Safety of a cluster regimen for subcutaneous 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 maternal records of 21 HDM-allergic children/adolescents (age 72 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.

<table>
<thead>
<tr>
<th>Cluster day</th>
<th>IR/ml</th>
<th>Dose, ml</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>1.0</td>
<td>0.1 0.3 0.6</td>
</tr>
<tr>
<td>2</td>
<td>10.0</td>
<td>0.1 0.2</td>
</tr>
<tr>
<td>3</td>
<td>10.0</td>
<td>0.4 0.8</td>
</tr>
<tr>
<td>time between treatment days:</td>
<td>1 week</td>
<td></td>
</tr>
<tr>
<td>time between injections:</td>
<td>30–60 minutes</td>
<td></td>
</tr>
</tbody>
</table>

adapted from Pfaar O et al. Int Arch Allergy Immunol 2009
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 with history of HUS and severe central nervous system (CNS) involvement.

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

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 were 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) introducing 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 [0–75] 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.
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, or with retardation in growth and development, screening should be done earlier. The aim of screening is the early detection of coarctation of the aorta. She underwent aortoplasty with a pericardial patch. Postoperative course was uneventful. At the time of discharge, she was on aspirin medication and will be followed up in the adult cardiology clinic.

Case 1: A 9 year old girl presents because of pathological blood pressure values. 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 with 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 values. 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 with multiple well developed collaterals and the patient underwent end-to-end anastomosis. Postoperative course was uncomplicated and is currently treated with Metoprolol.

Late detection of coarctation of the aorta leading to late growth retardation and intellectual disability. In children with coarctation of the aorta, the aortic arch is narrowed, leading to high blood pressure in the upper body and low blood pressure in the lower body. If left untreated, this can lead to heart failure and strokes.

Conclusions: Late detection of coarctation of the aorta leading to late growth retardation and intellectual disability. In children with coarctation of the aorta, the aortic arch is narrowed, leading to high blood pressure in the upper body and low blood pressure in the lower body. If left untreated, this can lead to heart failure and strokes.
Electrolyte abnormalities in cystic fibrosis: systematic review of the literature

S.A.G. Lava, O.D. Simonetti, S.A.G. Lava, P.B. Fara, M.G. Bianchetti

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 and Meta-Analyses statement.

Results: We found 54 cases (ranging in age from 2 to 37, median 9.0 years; male to female ratio: 1:1.7) 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 14 days or less after starting the medication, following complication mostly develops in subjects managed with the intranasal formulation. 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.

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

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Methods: We recently made the diagnosis of cystic fibrosis in an infant with normal neonatal screening, cough, hypnatremia, hypokalemia and hyperbicarbonatemia. 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 and Meta-Analyses statement.

Results: The reports included 172 subacute and 90 chronic cases with a 1:1.57 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 hyponatremia, 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.

Skeeter syndrome, a frightening but benign mosquito bite reaction

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Methods: We report the case of an healthy 8 years old child, who developed renal tubular dysfunction on treatment with deferasirox and review the corresponding literature

Case summary: We report the case of a 8 years old child, who developed renal tubular dysfunction on treatment with deferasirox and reviewed the literature. 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.

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 IgE/IgG antibodies. 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 IgG antibodies were negative.

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.
Primary hyperparathyroidism due to parathyroid carcinoma

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Background: Primary hyperparathyroidism in childhood is a rare disease with severe hypercalcemia. A parathyroid adenoma is the most often underlying cause.

Case report: A 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 lumbar spine. Family history revealed primary hyperparathyroidism with a documented genetic mutation (HP-JTHHRPT-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×P 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 96 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), Hypercalcruia (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 hypochogenic kienyges and renal displasibility. Electromyography were normal. MIBI-Single photon emission computed tomography (SPECT) and ultrasound revealed one hypochogenic, 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 subhumanization 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.

Discussion and conclusion: Severe congenital hyperbilirubinemia may be observed in SpHUS with a variable disease prognosis. Mechanism and pathophysiology of cholestasis seem 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.

Dilated cardiomyopathy in a patient with propionic academia: 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 academia, 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 propionylcarcinoline in a dried blood spot and secretion of 3-OH-propionate and methylcitrate in urine, typical findings in propionic academia. Enzymatic tests in leukocytes 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 un specific. 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 academia in previously healthy adolescents.
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 improved 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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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 (PM10) 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 PM10 exposure were obtained from a background monitoring station. The mean of PM10 was then calculated for the last trimester of pregnancy and for the last 30, 14 and 7 days before delivery. The association between PM10 and umbilical cord blood cells was assessed using a multivariable linear regression analysis, adjusted for possible confounders, such as gestational age, mode of delivery, stress factors during delivery, prenatal smoking exposure, season of birth, maternal atopy.

Results: PM10 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 PM10 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 PM10 may influence fetal immune development leading to an increase of cord blood dendritic cells, particularly mDCs, 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 (1.3±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 7.3±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 fulfill 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 TRECks (T-Cell receptor excision circles) from dried blood spots (DBS) is a sensitive and specific screening test for SCID and severe T cell deficiency. TRECks are a reliable marker of the number of circulating naive 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 TRECks in the original DBS of babies with confirmed SCID.

Results: TREC copy number were measured from an 1.5 mm DBS of 8 patients with confirmed SCID; 0.8, 0, 0–9; and 47 controls; 136, 118, 27–329; mean, median, range, respectively.

Conclusion: The TREC assay we tested is a reliable assay, easy 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 care 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 helminth 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.
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 cephalhaematoma. A systematic literature review is added correlating clinical presentation, laboratory results, therapies and outcomes of previously reported cases of infected cephalhaematoma 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 markers, 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 38%, fluctuance in 34% and skin erosion in 21%. Sixty-two percent of the infants presented with fever. E.coli was isolated from 66% of the haematoma. The mean time of antibiotic treatment was 27 days (range 7–67). Only three infants healed without surgical intervention.

Conclusions: Diagnosis of infected cephalhaematoma remains challenging. Neither clinical nor inflammatory markers are definitive. Imaging has limited power in differentiating liquefaction versus abscess formation. Infection should be suspected if there is secondary enlargement of the haematoma, erythema, fluctuance, 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.

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 ankle 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.

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 lesion in the calcaneus with cortical destruction and an abscess in the interspace between the Achilles- and Peroneus tendon. Histological analysis of the biopsy exhibited epitheloid-giant-celgranuloma; microscopically, acid-fast bacilli were found. Molecularbiologically, Mycobacterium-tuberculosis-complex-DNA with full sensibility to common tuberculostatics 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 tuberculosis 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.

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. PedNHIScScore 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.
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 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 afebrile 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.

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 24 hours. Aside an isolated typical peripheral facial paralysis, neurological examination was normal. Because Lyme disease is frequently encountered in our region, the child was treated with prednison 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 aphous ulcers was positive, Behcet disease was evoked, but HLA BS1 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 disease1. 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

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 reported in adults may be missing.

Case: An 8 year old boy presented to our emergency department with a 12-day history of severe undifferentiated headache and 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 empirical 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 neurology 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 unpecific. Sinogenic intracranial empyema should be considered in patients with persistent headache of biphasic evolution, focal neurological signs and altered mental state. Early detection with contrast-enhanced CT or MRI scan is recommended, followed by antibiotic treatment and surgical drainage of subdural empyema and affected sinuses.

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 160 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 1986 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 different clusters 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.

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 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 occurred full years after implementation.

Results: We report two cases of IPD, which occurred in children optimally vaccinated with PCV13:
Patient 1: A 2 year-old previously healthy 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 pneumoniae was only detected by molecular work from the pleural tap with ongoing genotyping of the strain. Adequate drainage and i.v. ceftriaxone allowed for a slow recovery.

Discussion and conclusion: Serious musculoskeletal complications including septic arthritis, osteomyelitis and abscesses following varicella infection are uncommon (1/100 000 cases). Sacroilitis accounting for 1–2% of osteo-articular infections in children is therefore an extremely rare finding as a complication of varicella. The diagnosis of sacroilitis in children is therefore difficult and often delayed due to the lack of specific signs and symptoms. Optimally, diagnostic tests and management of children with septic sacroilitis remain yet to be defined.
Varicella's reinfection in immunocompetent patients

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 3-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 ecephalitis 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 family predisposition could be an argument to vaccine the other insufficient making recommendations difficult to edit. Nevertheless the Conclusion:

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 visit for some of the lesions and ulcers to heal with epithelialization. The outcome will probably leave some unesthetic scars. In conclusion severe invasive bacterial superinfection with GAS, even if reported as a rare event, must be considered in any case of varicella.

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 practitioners of this presentation, and we review the literature.
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 organisational 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.

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.
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.

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 0.68 IU/ml to 0.80 IU/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 0.45 IU/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.
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 retrobullar 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 spherocytic 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 I. We opted not to biopsy the mass and observed radiological evolution. Nevertheless, due to the spinal involvement, triple intraethalcs 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.

A rare combination of severe ADAMTS13 deficiency with acute liver failure

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Introduction: ADAMTS13 cleaves von Willebrand Factor regulating microangiopaties usually not associated with severe 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.

Transplantation

Chloromas in ALL are rare tumours, especially intraspinal ones. ALL should be included in the differential diagnosis of intramedullary masses.

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

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Introduction: Spleen 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 spherical blast. Cerebrospinal MRI showed a splenomegaly and observed radiological evolution. Nevertheless, due to the spinal involvement, triple intraethalcs 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.

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%. Here we report a relapse of disseminated Mucormycosis in a patient with relapsed AML and double cord HSCT.

Case: We report a 20 year old female patient with relapsed AML who underwent allogeneic HSCT with a HLA-matched (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 was initiated. On day +25, 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. B. cinerea was identified and 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 impairment after repeated cycles of high dose steroids. Imaging studies showed severe alveolar consolidation of both lungs with multiple pulmonary cates. 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.

Discusison: 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.
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 concentration was measured 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 ≥25th 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.

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 Preferred Reporting Items for Systematic Reviews and Meta-Analyses statement.

Results: Forty-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 metabolism and potassium balance. A significant tendency towards mild to moderate hyperchremic 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.
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 calcium channel blockers 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? S. Joyeux1, J. Llo1, J.-J. Cheseaux1, M. Russo1, R. Tabin1, C. Poloni2, J.-P. Marcoz2
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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, hyposthenia, and fecal retention, occurring 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 initiated 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) with this treatment the boy is symptom free.

Discussion: Initially 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 CIDP 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? M. Beri1, A. Donas1
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Introduction: 77'000 children annually with acute varicella infection lead to approximately 140 hospitalizations in Switzerland. In 2013 we treated five patients with 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 ecmhaya on her trunk: She had had acute varicella infection two weeks earlier on a summer holiday 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.

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Clinical and toxicological features of attempted suicides by deliberate self-poisoning in adolescents in Switzerland C. Degrandi1, K.E. Hofer1, C. Rauber-Lüthy1, H. Kupferschmidt2, A. Ceschi2
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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 (9 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. However, the remarkable frequency of antidepressants and ANS compounds, mainly with psycholeptics.

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.

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A case of familial primary congenital glaucoma S. Joyeux1, R. Boldea2, A. Botton2, J. Llo1, J.-J. Cheseaux1, R. Tabin1
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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 clinical 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 as Sturge-Weber syndrome or certain metabolic diseases. Early goniopuncture 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.

Vomiting, diarrhea, dehydration, acidosis: basal 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 academia. 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 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 ketoadidosis 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 methylmalonic acid in urine is still elevated (650 mmol/L creatinine) indicating a cobalamin-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 derailing can cause an ischemic stroke with severe neurological sequelae. Early diagnosis and prompt administration of hydroxycobalamin is related to a better prognosis.

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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1 AllkidS Allianz Kinderspitäler der Schweiz, 2 Universitäts-Kinderspital beider Basel (UKBB), Basel, 3 AllkidS Allianz Kinderspitäler der Schweiz, 4 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 Groupier 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 development of tarifal structure for hospital treatment in children's hospitals.
Discussion: These cases stress the value of genotype, in case of suboptimal serum concentrations despite good treatment adherence and without any drug interactions. The identification 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.

Conclusions: Genetic testing is a first step towards an individualized prescription, which are of particular value in situations where subtherapeutic plasma concentrations are linked to treatment failure.

Discussion and Conclusion: Genital hypoplasia is a frequent feature in patients with CHD7 mutation and, if associated with other anomalies as dysmorphic ears, CHARGD syndrome should be considered. Primary hypoparathyroidism and thyamus dysfunctions are increasingly recognised in CHARGD syndrome and mark the overlap with DiGeorge-syndrome. As CHD 7 is involved in the organization of chromatin and contributing 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.

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.

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

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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 development, anomalies of genitourinary 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 expression of several genes during embryonic development, there is a wide spectrum of clinical manifestations with variable phenotype.

Table 1: Plasma levels of clozapine and norclozapine

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<tr>
<td>norclozapine</td>
<td>109 ng/ml</td>
<td>38 ng/ml</td>
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Discussion: BrainStim is an intervention tool on working memory in patients with AN-R. AN-R patients considerably improved during the training procedure will be further analyzed.

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

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Introduction: CYP1A2, 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 where subtherapeutic plasma concentrations are linked to treatment failure. The patient was given permission to go home for three days a week. The clinical response decreased where subtherapeutic plasma concentrations are linked to treatment failure. The patient was given permission to go home for three days a week. The clinical response decreased where subtherapeutic plasma concentrations are linked to treatment failure. The patient was given permission to go home for three days a week. The clinical response decreased where subtherapeutic plasma concentrations are linked to treatment failure.
Multidisciplinary management of anorexia nervosa in a regional hospital setting

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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 used to produce an inventory (Docket and colleagues 2003) to produce important propositions about children’s experiences of hospitalization.

Results: The main quantitative result was that the mouse was the most popular chosen right, next was the right to be not hungry, and as co-therapist were designed the child adolescent senior psychiatric service.

Conclusion: 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.

The case of a dangerous fish

M. Papathanasiou Terzi1, A. Tschemia1

Introduction: 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 histamine fish poisoning was made 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”.

Discussion: Our multidisciplinary case management is effective in a regional hospital for moderate cases of anorexia nervosa.
We believe that awareness among pediatrics regarding food borne poisoning is important to differentiate these cases from allergic conditions. Furthermore case reporting remains essential to ensure optimal public health intervention.

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.

Technologiefolgenkritik und Cyberphilosophie – mediensozioologische Aspekte des pathologischen Internetgebrauchs
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Introduction: In der publizistischen und mediensozialwissenschaftlichen 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 wie weit diese offentliche Diskussion für die Konzeptualisierung von Diagnose- und Therapiestrategien bei pathologischem Internet- und Medieneinsatz nützlich gemacht werden kann, ist zu untersuchen.


Ergebnisse: Es zeigen sich drei große Linien in der aktuellen mediensozialwissenschaftlichen Literatur: 1. die technikfreundlichen Publikationen, 2. die technikkritischen Publikationen und 3. Publikationen, die versuchten einen Ausgleich herzustellen. Während die erste Kategorie primär die Nutzung sozialer Medien und der neuen Kommunikations- und interaktionellen Chancen in den Vordergrund stellt, finden sich bei der zweiten Kategorie verschiedene Autorinnen, 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 die 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 entsprechenden Publikationen kommen fast ausnahmslos aus dem amerikanischen Raum und bilden die dortige gesellschaftliche Realität ab.

Diskussion: Die weltweit geführte mediensoziale 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.

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.

References
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 separates 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 personality 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.

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.
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