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FM 1

Paediatric emergency: creation of an independent nurse practitioner consultation

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Consultations in the Paediatric Emergency Department (PED) continue to climb regularly. Emergency Nurse Practitioner consultations have long been created in the English speaking countries.
Since January 2013, an independent nurse consultation, under delegated medical responsibility, exists in the multidisciplinary PED of the Children's Hospital of Lausanne. The mean consultation time is the same as the medical consultation and the overall waiting time hasn't decreased yet. But a well definite working frame, a systematic approach, as well as the continual medical supervision possibility, make it a safe, efficient and appreciated consultation, by both patients and professionals.

FM 2

Impact of a protocol in the quality of call triage: management of children with fever without an evident cause

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Background: Call centers with nurses have been implanted in Europa, United States and Autralia to allow a diminution of consultations in the emergency wards. We created protocols for our call center to guide the nurses in asking specific questions and to indicate elements that motive differents orientations (asking for an ambulance, orientating to an emergency ward, temporising the consultation with an delayed appointment or giving advice).
Objectives: The principal objective is to evaluate the evolution of the rate of call triage found good before, during and after the introduction of a protocol (image 1) for a complain (fever without evident cause). The second objective will focus on the rate of anamnestic elements thought to be essential to make an adequate triage.
Method: we listened retrospectively all the phone calls with a complain of fever without evident other symptom concerning all children between 0 and 5 years old during the months of january 2012, 2013 and 2014 (before, at time and after the introduction of protocols in the call center).
Results: In our study 761 calls were included, 186 in 2012, 319 in 2013 and 256 in 2014. In comparing the results by year, we find that very few calls about fever occur for the population under than 3 months, and most calls concern children between 3 and 12 months. The time of the calls are not longer with the introduction of protocol. We observed more call during the night in 2014, but not more indications to consult in emergency. We noticed a significative increase of questions about the general state of the children (62% in 2012 and 82% in 2014), but specific questions stay poor in the evaluation of patient with fever. The adequacy of the orientation proposed in function of the general state of the children and the symptoms explained improved from 59% to 79%.
Conclusion: Our study shows that the introduction of a protocol improves the history taking and the call triage. However, a specific teaching is needed to enrich even more the anamnestic data, found to be necessary to make an adequate orientation.

FM 3

IntOKS – 2003–2013

Intoxications at the Children's Hospital of Eastern Switzerland, St. Gallen

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Background: The national "Tox-Zentrum" provides annual reports on registered intoxication cases in Switzerland.
Objective: We want to compare these statistical data's with our clinical cases (patients). Furthermore we aim to give a basis for further efforts in quality improvement.
Methods: Retrospective study of all intoxication cases (patients) in a cohort observed at the Children's Hospital of Eastern Switzerland, St. Gallen (OKS) between 2003 and 2013. Investigation of all patients by doctor's report on the basis of ICD-Codes.
Results: Between 2003 and 2013 we identified a total of 832 patients treatet in the Emergency Departement at the OKS because of an intoxication. Results stayed the same over the years. We had no cases of death. Two patients needed transfer to a more specialized pediatric centre (1 mushroom intoxication, 1 stack gas intoxication). The mean age at admission was 10.98 years (Newborn – 19.58 years). Gender distribution was male 53% to female 45% and 2% of unknown gender. In 80% of all inpatient treated patients intoxication was caused by medicaments, alcohol, other stimulants and drugs. Intoxications by alcohol provide a fourth of all cases seen on the emergency departement (total n = 203), therefrom 4 needed treatment in the intensive care unit. Since 2009 there seems to be a decline of cases with intoxication by alcohol. Substances used in multiple intoxications were medicaments and stimulants (incl. alcohol), The same pattern we see in intoxications with intent to suicide (total n = 54). Therefore used substances were paracetamol (n = 9), benzodiazepine (n = 5), antidepressant drugs (n = 4) and other opioids like Tramadol, CoDafalgan, Codicontin und Codein (n = 4). In only 32.5% of all cases after medicament intoxication there was an indication to charcoal medication.
Discussion: Our results were moreover comparable to the findings of the "Tox-Zentrum"-data's. But we do see a lot more Alcohol intoxications and intoxications with antiallergics and paracetamol. We only had a small amount on charcoal medications (32.5%) in intoxications with medicaments. Other intoxication groups had even less. Most because of patients late appearance in the emergency departement.
We aim to provide a better awareness of parents and health care professionals on intoxications in our hospital region (Eastern Switzerland). We want to reduce time to treatment and to improve prevention of intoxications. Likewise we design flyers for parents, which will guide them in situations of possible intoxications.

FM 4

Career choices of trainees from the pediatric residency program in Bern, Switzerland

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Introduction: There is a nationwide shortage of pediatricians in private practice. Considering that the Swiss Medical Association (FMH) has certified an ever increasing number of pediatricians during the past decade, the shortage of their workforce in the outpatient sector is puzzling. It is claimed that retention to tertiary care hospitals could be responsible. The Department of Pediatrics at the Inselspital in Bern is a 104-bed tertiary care facility (3'300 in-patients and 22'000 out-patients per year). Its residency program offers 38.5 full-time training positions (FTP). We assessed career choices of physicians who left our pediatric residency program within a seven year period to evaluate possible retention of trainees to the hospital sector.
Methods: Data from an in-house human resources database, publicly available data from the FMH (www.doctorfmh.ch), and public Internet data were used to assess career choices of physicians who had left our program between 2006 and 2012 after a minimum of 2 years of training in our pediatric residency program. Career choice was defined as the health care sector in which they worked as of January 2015. Private practice as career choice was counted also in pediatricians who kept a part-time appointment at a hospital.
Results: During the 7-year observation period, 88 physicians (mean 12.6 per year) had left the program, the majority (83%; n = 73) of them being FMH board certified in pediatrics as of January 2015. Fifty-seven (65%) were female. Eighty-three percent (n = 73) of trainees had left our institution permanently, whereas 15 (17%) pursued an in-house career. Of those who left our department, 34 (46%) chose to work in private practice in the outpatient sector and 31 (42%) selected the hospital sector. In 8 (12%) the career choice was unaccounted for at the time of the study. Among trainees who chose the private sector, a greater proportion were female (78% vs. 52% in the hospital sector; p = 0.042; Fisher's Exact two-tailed test).
Conclusion: Career choices of trainees who left the pediatric residency program between 2006 and 2012 were equally distributed between private practice and hospital services. Predominantly women chose the private sector. Only a small proportion of trainees pursued a career at our own tertiary care institution (17%). FMH board certification rate was high. Future analyses should also address the overall workforce of pediatricians by assessing employment levels in private practices (FTP) to better predict future needs of graduates from pediatric residency programs in Switzerland.

FM 5

Teaching with adolescent simulated patients, what can we learn from medical students? A mixed methods study

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Introduction: communication and interviewing skills are an essential part of adolescent healthcare and yet a lack of training in this field is often highlighted among health professionals. The introduction of programs with adolescent simulated patients (ASP) has been an opportunity to meet the substantial challenge of teaching these skills to medical students. Identifying ways to optimize student learning with ASP through a thorough understanding of how learning with ASP works is of outmost importance to improve students' skills in adolescent health. The purpose of this study is the in-depth exploration of the students' learning experience with ASP at the University of Lausanne (Switzerland).
Methods: Mixed methods study including two parts that will be conducted simultaneously in spring 2015. Part A consists of a qualitative inquiry using grounded theory approach. It includes semi-structured interviews, focus groups and in-field observation of workshops with ASP among fourth-year medical students, as well as ASP and teachers involved in these workshops. Part B consists of an online cross-sectional survey with both quantitative and qualitative data collection that will be submitted to all fourth- and sixth-year students.
Results: we will present advanced results of the qualitative part of the study that will allow us to understand experiences and perceptions of students about ASP. This understanding is essential to bring out barriers and positive factors contributing to an effective learning process and to build a theory on student learning with ASP.
Conclusion: the results of our study will help medical educators and teachers in adolescent medicine find ways to optimize this learning process and pay attention to the prerequisites for successful learning when implementing programs with ASP in the undergraduate medical curriculum. This is an essential step to improve the teaching curriculum of adolescent medicine and therefore to improve students' skills with adolescent patients.

FM 6

Fractures, pain and vitamin D deficiency in handicapped or otherwise healthy adolescent patients of an outpatient orthopedic clinic

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Introduction: In Switzerland, 20–60% of children have decreased Vitamin D (Vit.D) levels. Though the potential consequences, namely severe rickets, tetanic convulsions and increased infection rates have become rare in infants, in older children with Vit.D deficiency below 30 nmol/L, chronic fatigue, progressive muscle pain and weakness may occur. Particularly in paediatric orthopaedic patients with chronic diseases, antiepileptic drugs and severe handicap, bone fractures or rickets.may be due to Vit.D deficiency. In order to recognize Vit.D deficiency in orthopaedic patients and to avoid further complications, we present a diagnostic check list for bone health in immobile patients with cerebral palsy(CP) and in otherwise healthy children with chronic pain for unknown reason.
Methods: In all immobile patients with severe CP (GMFCS IV and V) between age 8 to 18 years of our outpatient orthopedic clinic, we monitored bone metabolic status by means of a check list on anamnesis, nutrition, pubertal staging and medication, filled out by parents, doctors and nurses, and measured serum calcium (Ca), 25-OH Vit, D, phosphate, magnesium, alk. phosphatase, PTH and Ca/Crea ratio in urine once a year. The same diagnostics were performed in otherwise healthy patients presenting symptoms and signs of Vit.D deficiency.
Results: Out of 37 CP patients (mean 15.5 years, 23 male), 36 had insufficient vitamin D levels <75 nmol/L. In one third of them, pathological bone fractures or pain or deterioration of mobility were found. Antiepileptic drugs were given in 51% and tube-feeding in 32%. Mean BMI was decreased to –2.3 SD and hypogonadism present in 11%. Vit. D was restored carefully with 600 IU/day, adequate nutritional calcium intake assured and both administered permanently.
In 2 healthy adolescents with bone pain, hypocalcaemic rickets were detected and in one accompanied by bone edema in MRI, resolving after Vit.D therapy at a higher dose (2000–4000 IU/day and 1000–2000 mg Ca for 3 months, then Vit.D prophylaxis).
Conclusion: Our serial examination confirmed that bone health often is impaired in severely handicapped patients by symptomatic Vit. D deficiency and additional risk factors as anticonvulsive drugs or intestinal, liver and kidney diseases; the fracture rate was as described previously (4–8%). To avoid further important medical problems, these patients should obtain Vit. D prophylaxis according to Swiss recommendations and be screening for bone health periodically. Normalization of Vit. D levels achieved a release of chronic pain in healthy adolescents. Our checklist, comprising specific history, status and lab control, helps to monitor metabolic bone diseases in an orthopaedic setting.

FM 7

Fundraising for a mother-infant unit in Burkina Faso

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The "Centre Médico-Chirurgical Pédiatrique Persis" (CMCPP), is situated in Ouahigouya, Burkina Faso, in a subsaharian region with 1'380'000 inhabitants. It is a private pediatric hospital, with social goals. Since 2002, a collaboration was introduced between the pediatricians of this hospital and of our hospital. The management of this hospital has, with the association "Persis Burkina", the project to develop a neonatal and maternity ward. This project is also recommended by the Ministry of health of Burkina Faso in a strategy of private public partnership. The association "Persis Valais", founded in 2008, has the purpose to initiate, share or support humanitarian actions or projects, raise funds for the CMCPP. With others association, in Switzerland and in France, it supported the development of several projects in the CMCPP and now is fully committed with this project. The aim of this presentation is to present how to develop a project description and documents for fund raising and to give some tips and addresses useful for request to social and humanitarian associations.

Reference: www.persis-valais.ch

FM 8

Childhood obesity therapy: who benefits from an information technology (IT) supported treatment?*

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Introduction: In Switzerland, multiprofessional therapy programs for obese children and adolescents and their families, including physical activity, nutritional and behavioral interventions, have been shown to be effective, in group (1, MGP) as well as in individual (2, MIT) settings. However, due to limited personal and financial resources, less than 1% of children affected participate in such programs. Health IT systems (HIS) have the potential to improve therapy assisting families in different settings. Therefore we designed a novel mobile application that accompanies obese adolescents and their parents during everyday situations.
Methods: A HIS offering physical activity, mood, photo documentation and eating speed services on a tablet PC was designed by specialists and adolescents, with a direct data transfer between home and centre (via patient privacy service). In a pilot observational study usage as well as physical and psychological outcomes were explored for 7 months in each 6 children with HIS and group therapy (HIS-MGP), individual therapy with HIS (HIS-MIT) and without HIS (C-MIT). Satisfaction of therapists with HIS was assessed by questionnaires. HIS data on usage of all services were assessed as previously described (3); examination of physical and mental health and of motivation, eating disorders, well-being and parenting were performed as in the Swiss national study (1).
Results: In all groups likewise, adolescents were extremely obese (total mean ± SD: BMI-SDS 2.9 ± 0.5, age 13.2 ± 3.4y). Both in HIS-MGP and C-MIT, BMI-SDS decreased by 0.3 and 0.2 SD, resp., but not in HIS-MIT. Before and after therapy, there were no major group differences in the main outcome parameters. The trend ($p = 0.07$) to reduce obesity under higher emotional strain was not reflected by HIS usage. Despite contracts with children and parents, HIS, mainly activity and mood services, were only used by those patients who were closely supervised by therapists or parents. Therapists found HIS to be helpful in coaching the patients.
Conclusion: MGP and MIT are effective therapies. This pilot study cannot demonstrate that mobile Apps alone improve obesity therapy.

Only under close supervision, HIS did simplify communication between therapist and patients. Further randomized controlled studies in less severely obese patients will prove whether a HIS with an automatic SMS reminder system in addition to a closely supervised therapy program can support lifestyle changes.

1 KIDSSTEP Group, Paediatrica 2013;23:27–30. 2 Maron L, et al. Swiss Med Wkly 2014;144(Suppl. 203):20. 3 Kowatsch T, et al. ecis2014.eu/E-poster/files/0438-file1.pdf; * SNF grant CR10I1_135552

FM 9

Realities und Challenges in Implementing "Shared-Decision Making" in Pediatrics

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Introduction: Shared decision-making (SDM) describes a partnership between health care providers and patients, in which each contributes equally to decisions about treatment and care. The concept has been discussed and studied extensively in adult medicine. In the the pediatic context, however, still only few research is available. The exchange of information in a parent- as well child-appropriate way is time consuming and dependent on particular skills. Neverthless, there are several reasons why SDM can and should become an integral part in pediatrics. To provide a ground for further discussions and studies we explored how health care providers, patients and parents in the specific context of pediatric oncology make sense of SDM.
Methods: In the context of pediatric oncology at the University Hospital of Zurich, Switzerland we included 16 multiprofessional health care providers in three focus groups and interviewed 5 families including their children. Exploring the sense-making, the underlying construction and the practical use of a concept (i.e. SDM), we used a modified analytical tools based on interpretative phenomenological analysis (IPA) and a hermeneutic-reconstructive analysis. In open, minimally structured talks we asked the participants about their meaning, their needs and experiences concerning SDM. All interviews and focus groups were audio-recorded, transcribed, anonymized and analyzed by three of four authors.
Results: Parents and patients showed wide satisfaction with communication and participation. Contributing factors identified by parents and patients were interpersonal relations with, and empathy and reachability of health care providers. A particular form of partnership between health care providers, patients and parents seemed to be a fundamental premise for decisions about treatment and care. However, we could not find evidence that those partnerships are bound to the idea of equality. Health care providers even showed open reservation and scepticism about SDM mostly because of its presumably time-consuming implementation in the already challenging daily practice.
Conclusion: Included parents and patients show wide satisfaction with communication in our oncolgical setting. However, satifaction in communication was not built on equality-based SDM but rather associated with an unquestioned acceptance of a hospital-related "normality". Rather than elaborating a mutual partnership, satisfying communciation seems reminiscent of paternalistic methods. In that light we also interpret our finding of refusal or skepticism of health care providers regarding SDM as a pragmatic act: They try to lead parents and patients as troublefree as possible through the complex world of care and choice concerning a serious medical problem, without making communication more difficult as "necessary". Based on these results we conclude that a sound application and promotion of SDM needs further theoretical and empirical analyses.

FM 10

Biochemical bypass steroidogenesis pathway in HSD3B2 deficiency: lesson from an experiment of human nature

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Background: Deficiency of 3β-hydroxysteroid dehydrogenase (3βHSD) is a rare disorder of sexual development (DSD) affecting both sexes and overall steroidogenesis. There are two isozymes of 3βHSD, encoded by separate genes *HSD3B1* and *HSD3B2*. Human mutations are only known for the *HSD3B2* gene which is expressed in the gonads and the adrenals.
Aim: To describe the molecular genetics, the steroid biochemistry, the (immune-) histochemistry and the clinical implications in a patient harboring a severe, homozygote HSD3B2 mutation from birth to 16 years of age.
Methods and Results: Genetic analysis of the *HSD3B2* gene revealed a known homozygote c.687del27 deletion. Further work-up by RT-PCR on RNA from testis tissue revealed a shorter cDNA product corresponding to the deletion while Western blot from testis protein extract showed no HSD3B2 protein expression. Urine steroid profiling revealed low overall steroid production for mineralocorticoids, glucocorticoids and sex steroids with typical precursor metabolites for HSD3B2 deficiency in early life. However, at pubertal age, some sex steroid metabolites appeared likely through conversion of precursors by unaffected HSD3B1 enzyme activity in the peripheral tissues. Accordingly, the 46,XY boy presented at birth with severe undervilization of the genitalia but was able to virilize further at puberty. Remarkably, however, he developed enlarged breasts (Tanner stage 4) through production of estrogens in the peripheral tissues from androgen precursors through aromatase activity. Testis histology and immunohistochemical studies at late puberty revealed arrested spermiogenesis, presence of Leydig cells in stroma, but no evidence of neoplastic changes (as seen in other types of 46,XY DSD).
Conclusion: This is the first longitudinal detail description of a patient with a severe human HSD3B2 mutation. Extensive laboratory studies on human biomaterials allowed obtaining further insight into the function of HSD3B2 and the bypass steroid pathways forced by its loss. In addition, we are able to show that there is no risk for malignancy risk for malignancy in testes in HSD3B2.

FM 11

Eliminating measles in Switzerland: preliminary data

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Introduction: Switzerland and all European WHO Member States have the goal of eliminating measles by the end of 2015. To achieve this goal, the Swiss confederation, the cantons and medical professional organizations such as SSP are coordinating their efforts specified in the National strategy for the elimination of Measles 2011–15. Measles can be eliminated if ≥95% of the population are immune against the virus, which has been achieved by many countries. In Switzerland in 2011–13, only 86% of two year old children had received 2 doses, and important gaps in immunisation coverage prevailed in the under 50 years old. Main objectives of the strategy are: 1. to raise immunisation coverage of two year olds with 2 doses of MMR/measles vaccine to ≥95%; 2. to close the gaps with catch-up vaccinations for anyone born after 1963; and 3. to bring any measles outbreak rapidly under control using uniform standards and procedures. Here, we present current data and evaluation results for measures of the national strategy.
Methods: The advancements in achieving the goals are constantly monitored by surveillance systems (such as mandatory reporting of suspected measles cases and monitoring of immunisation coverage) as well as by specific evaluative studies until the end of 2016.
Results: Immunisation coverage with 2 doses of MMR/measles vaccine is increasing in all age groups. Reported confirmed measles cases dropped significantly over the last three-year periods: from 3391 in 2006–08, to 1825 in 2009–11, to 262 in 2012–14. Median age in 2012–14 was 15 years. An enquiry among GPs and pediatricians (Sentinella preliminary data for 2014) showed an extrapolated 33'500 catch-up doses were given to 2 to 50 years old patients, in 9 of 10 catch-up vaccinations, the initiative came from the physician and quite a few parents were vaccinated in the pediatrician's offices. The so called "Baseline-study" conducted in 2012 showed that 34% of parents of unvaccinated children (age 3–16) would consider to protect their child with a catch-up vaccination, and that 43% of unprotected adults would do so. Finally, the evaluation of the "Stop Measles Campaign" showed that >50% of the interviewed adults who noticed the campaign got their own and their children's vaccination cards checked or intended to do so.
Conclusion: Recent data indicate good progress and that Switzerland is quite close to eliminating measles. However, since the goal is not reached yet, all efforts should be continued. Verification of vaccination status and completion to 2 doses is crucial for every patient.

FM 12

Influence of Structured Training in Diagnosis and Management of Patients with Sepsis

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Introduction: The clinical outcome of septic shock is strongly dependent on the swift diagnosis and beginning of treatment and stabilization of the patient. To improve our response time especially in the emergency room, we began to establish special training in recognizing the signs of septic shock for our nursing and medical staff. The purpose of this study was to evaluate the effectiveness of our training program and to reach the objectives of the surviving sepsis campaign.
Methods: We did a retrospective analysis of all children (44 weeks of gestational age to young adults) entering our emergency department with sepsis and/or septic shock and with positive blood cultures from January 2007 to December 2014. We began an informal and sporadic training for the nursing and medical staff in July 2009 (cohort 2) and expanded the training in January 2012 to a formal, regular and mandatory training (cohort 3), in combination with a pocket-sized checklist. Then we compared the patients with the cohort before the training (cohort 1) to see if our training improved our outcome. The main objectives were, our response time to start volume administration, to take blood cultures and to administer the first dose of antibiotics.
Results: In total, we included 79 patients, 22 in cohort 1, 20 in cohort 2 and 39 in cohort 3. There was no significant difference in death (cohort 1: 2, cohort 2: 1, cohort 3: 2). Intensive care unit (ICU) admission was 41% in the first, 35% in second and 51% in the third cohort. Time to fluid administration and time to antibiotics was significantly shorter in cohort 2 versus cohort 1. In cohort 3, only significant improvement in early volume therapy was observed.
Conclusion: We could improve our septic management with informed training and sensitization of our staff. With more structured training, the further improvement was less than expected. Nurses and doctors are more alert for sepsis. That's why we observed an earlier transfer to the ICU in the third cohort. There is still a relevant latency between drawing blood cultures and the administration of antibiotics.
The time objectives of the early goal-directed therapy have not been achieved in all three cohorts.

FM 13

Impact of the introduction of an algorithm for the management of pneumonia with effusion in children

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Introduction: Optimal management of pneumonia with effusion in children is still controversial and varies a lot among different centers. We developed an algorithm for the management of pleural effusion and introduced it in May 2011 in our pediatric department.
Methods: We designed a retrospective cohort study to compare management and evolution of pneumonia with effusion before and after the introduction of the algorithm. We included every child aged 2 months to 18 years and hospitalized with pleural effusion between June 2008 and May 2013.
Results: Ninety-five patients were eligible, among whom 81 are available for intermediate analysis (52 before and 29 after the introduction of the algorithm). Median age was 4.3 years (range 0.6–16.1 years) without significant difference between the groups. The introduction of the algorithm was associated with a non-significant decrease in thoracic drainage (11/52 (22%) before versus 4/29 (14%) after the introduction of the algorithm (p = 0.5). There was no difference in the frequence of radiological investigations, length of hospital stay or readmission rate. The median duration of antibiotic treatment (median of 14 days, IQR 3-13) remained unchanged. Morphine treatment (68% versus 35%, p = 0.01) and tachypnea at discharge (41% versus 15%, p = 0.02) were more prevalent after the introduction of the algorithm, while weight loss diminished (median 0% versus 3% of the admission weight, p = 0.02).
Conclusion: The introduction of an algorithm for the management of pneumonia with effusion was associated with a trend of less pleural drainage, without impact on length of hospital stay, antibiotic duration, or readmission, despite a greater proportion of tachypneic patients at discharge. The main benefice of the algorithm was the improvement of pain and nutritional control.

FM 14

Medium term follow-up of children with Kawasaki disease at the CHUV

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CHUV

Introduction: Kawasaki disease (KD) is an acute systemic vasculitis with a particular involvement of the coronary arteries. Coronary artery aneurysm develop in 20% of untreated children and can lead to stenosis and myocardial infarction. Early treatment with intravenous immunoglobulins (IG) and aspirin (ASA) can decrease this risk to 5%.
Objectives: Medium term follow-up of all patients with the diagnosis of KD at our Institution between 1981 and 2014. Determination of risk factors for unfavorable outcome and comparison with the literature.
Methods: Retrospective review of all patients <16 years of age with the diagnosis of KD after Ethics Committee approval. Creation of a database with epidemiologic, clinical data, treatment , complications and follow-up. Statistical analyses.
Results: 207 pts were diagnosed with KD during this time. Medium age at diagnosis was 32 months (minimum 1 month, maximum 15 years). 159 pts were less than 5 years old (76.8%). 122 were male (58.9). There were 61 pts with incomplete KD (29.5%). The most frequent symptom was polymorphous rash (n = 176, 85%) followed by changes in lips and oral cavity (n = 161, 77.8%), conjunctival injection (n = 159, 76.8%), changes of the extremities (n = 144, 69.9%) and cervical adenopathy (n = 136, 65.7%). Treatment with IG was done in 191 cases (92.3%), and 201 received ASA at anti-inflammatory doses (97.1%). 3 pt did not get treated (1.4%). Medium time to treatment was 7 days (minimum 1, maximum 39). 108 pt had coronary anomalies (52.2%), 67 on left coronary (32.4%), 6 on the right coronary (2.9%), 35 on both coronary arteries (16.9%). 89 pts had complete regression of coronary aneurysms (82.4% of pts with coronary anomalies) while 19 had persistent coronary anomalies (17.6% of pts with coronary anomalies) at the end of the study period. 1 pt died of KD (0.5%). 32 pts had medium term complications during the follow-up (15.4%). The most frequent complication was persistent coronary aneurysm (n = 24, 11.6%), followed by recurrence of KD (n = 5, 2.4%), and myocardial ischemia (n = 3, 1.4%). Boys had more cardiac sequelae (p = 0.01) and pts less than 1 year and more than 10 years had higher risk of medium term complications (p <0.0001.)
Conclusions: Our study confirms that medium term complications are more frequent in boys and in pts outside the normal range for KD. Medium term outcome in our pts is better than described in the literature, probably due to early recognition of signs and symptoms of KD by the pediatrician leading to early treatment in these pts.

FM 15

Efficiency of CPAP therapy in newborns with respiratory distress in a regional center

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Introduction: The cantons Hospital of Fribourg (approx. 3'000 births per year in the canton) has opened a neonatal care unit for newborns older than 32 completed weeks of gestation offering CPAP therapy. The aim of our retrospective review is to evaluate the efficacy of this non-invasive treatment modality and to correlate it with the disease and the birth weight of the patients.
Methods: We obtained anonymized data of the electronic patient chart and retrospectively analysed variables of all newborns with respiratory distress hospitalised in the neonatal care unit between May 1st, 2012 and February 28th, 2014.
Results: During the study period of 22 months, CPAP treatment was required by 46 newborns (28 m, 18 f, 2440 g median birth weight, min-max: 1200–3890 g, 35 2/7 median GAW, 30 1/7 min 41 1/7 max). 37 (78%) patients could be treated locally and 9 (22%) were transferred. 1 transfer was due to lack of space. The median duration of the CPAP treatment of the locally treated patients was 42h (2–175h), of the transferred patients median 15h (4–48h). The median duration of hospitalization was 14 days (4 to 51 days) in locally and 1day (0–2d) prior to transfer in transferred patients
No correlation between birth-weight (p 0.9), gestation week (p 0.6) or APGAR (p 0.8) and transfer likeliness could be found. A correlation

between diagnosis and necessity of transfer could be established for wet-lung and membrane hyaline disease. Of the newborns diagnosed with pneumonia (n = 5) 0% (p 0.26), the ones with wet-lung (n = 27) 7% (p 0.032), the ones with broncho-aspiration (n = 3) 33% (p 0.7), the ones with infection (n = 2) 50% (p 0.6), and the ones diagnosed with hyaline membrane disease (n = 5) 80% (p 0.023) had to be transferred.
Maximum O_2 concentrations ranged from 35–80% (median 60%) in transferred patients and 21–100% (median 30%) in locally treated patients (p = 0.007). One (2%) CPAP-related complication in the form of a bilateral pneumothorax was documented.
Discussion: Four out of five patients with respiratory distress syndrome requiring CPAP, could be treated locally. While pneumothoraces at our institution were below rates found in the literature, the availabitliy of CPAP allowed a big decrease of postnatal transports.

FM 16

Modulation of SIRT-1 expression plays a protective role against oxidative stress induced endothelial progenitor cells dysfunction in preterm infants

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Introduction: Low birth weight, notably preterm (PT) birth, is considered as a major risk factor for cardiovascular-related diseases, notably hypertension (HT) at adulthood. One of the putative mechanisms of such effect is an altered vascular tree development that may pave the way to increased arterial resistance and later HT. We recently showed that PT infants endothelial colony forming cells (ECFCs), which are now considered as biomarkers of endothelial dysfunction (ED), display an accelerated stress-induced senescence *via* a decreased expression of SIRT1. In cardiovascular diseases, SIRT1 has been demonstrated to regulate cellular protection against oxidative stress (OS) which plays a role in various vascular complications of preterm birth, such as retinopathy and bronchopulmonary dysplasia. We hypothesized that OS is involved in the ECFCs dysfunction observed in PT infants.
Methods: ECFCs isolated from venous umbilical cord blood of 25 PT and term neonates (CTRL) were pre-treated ± resveratrol (R) or ± transient SIRT1 overexpression (tSIRT1). Measures of: expression of antioxidant defenses: superoxide dismutase (SOD1), catalase (CAT) by western blot; superoxide anion (SA) production using hydroethidine ± apocynin (Apo), N-nitro-L-Arginine (L-NNA) were performed.
Results: In ECFCs from PT *vs.* CTRL infants: expressions of CAT and SOD1 were decreased (p <0.01). SA production was increased (p <0.001), and pretreatment with Apo and L-NNA decreased SA levels. Pretreatment with R or tSIRT1 restored CAT and SOD1 expression, normalized SA levels.
Conclusion: Manipulating SIRT1 expression (by R or tSIRT1) is protective against OS. It may therefore improve angiogenic capacity and open a new perspective in deprogramming the ED observed in PT infants.

FM 17

Renal tissue oxygenation as measured with BOLD-MRI in children with vesico-ureteral reflux in comparison with healthy controls

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Background and objective: Vesico-ureteral reflux (VUR) in children is a risk factor for the development of renal scarring and chronic kidney disease (CKD), yet the underlying pathophysiology is incompletely understood. Renal hypoxia might be one of the underlying mechanisms contributing to the progression of CKD in these children. We measured cortical and medullary oxygenation in children with VUR, and compared the results with those of healthy controls using blood oxygenation level dependent magnetic resonance imaging (BOLD-MRI).
Methods and design: BOLD-MRI was performed under standardized hydration conditions, before and after the administration of furosemide. Combination sequence was used to acquire T2* weighted images of four slices. The mean R2* values (= 1/T2*) were calculated for the cortex and medulla of each kidney, a low R2* indicating a high tissue oxygenation.
Results: A total of 37 children (19 patients and 18 controls) participated to the study. Cortical and medullary R2* levels were significantly higher in healthy controls than in reflux kidneys, suggesting higher renal oxygenation in kidney with VUR. Furosemide-induced changes in medullary R2* were significantly (p = 0.042) smaller in children with VUR disease. In healthy children, cortical oxygenation was higher (R2* lower) in girls than in boys (16.6 ± 0.8 vs 17.7 ± 1.8, p = 0.015).
Conclusions: These data suggest that VUR is not associated with chronic renal hypoxia in children. Oxygen balance in the diseased kidney is likely more complex. A decline of glomerular filtration rate (GFR) may decrease tubular solute delivery and solute transport which in turn would reduce oxygen utilization. The significant furosemide-induced decreases in medullary R2* levels in healthy children point towards intense renal sodium transport in this group, compared to children with VUR.

FM 18

Neonatal oxygen exposure leads to vascular, renal and metabolic diseases in adulthood

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Introduction: Preterm (PT) birth, affecting 6–15% of all birth worldwide, is a risk factor for cardiovascular, notably hypertension (HT) and metabolic diseases at adulthood. PT infants are exposed to neonatal oxidative stress (OS), notably when oxygen supplementation is necessary, due to immature and less inducible antioxydant defenses. Disorders associated with PT birth, such as retinopathy, bronchopulmonary dysplasia, periventricular leukomalacia and necrotizing enterocolitis are important complications linked to OS. However, the long-term consequences of oxidative injury induced by oxygen exposure in the neonatal period are not well established. The purpose is to unravel some mechanisms that might underlie the damage induced by neonatal oxygen exposure and the long-term risk of developing vascular, renal and metabolic diseases.
Methods: Sprague-Dawley pups were kept with their mother in 80% O_2 (O_2) or room air (RA) from day 3 to 10 of life. ***Cardiovascular parameters at 4 weeks and adulthood*:** tail blood pressure (BP), vascular reactivity (*ex vivo* carotid rings) to Angiotensine II (AngII) and acetylcholine were studied; nitric oxide (NO) production was performed; vascular OS by measuring superoxide anion production; microvascular density was assessed on tibialis anterior muscle sections; pulse wave velocity (PWV) was measured by echodoppler; nephron were counted after hydrochloric acid digestion. ***Metabolic parameters:*** body weight has been measured during the growth. In adulthood, body composition, glucose tolerance were evaluated.
Results: ***At 4 weeks of age***, any difference was observed regarding BP, vascular reactivity, and OS indices, but in rats O_2 *vs.* RA (n = 6–8 /group), microvascular rarefaction was present.
At adulthood, in rats O_2 *vs.* RA (n = 6–8/group): **i**) systolic and diastolic BP was increased; **ii**) vascular reactivity to Ang II was increased and decreased in response to acethylcholine; **iii**) NO production was decreased; **iv**) vascular superoxide anion level was increased; **v**) microvascular density was decreased; **vi**) PWV was increased; **vii**) nephron count per kidney was decreased; **viii**) body weight was less during growth, but a catch up growth was observed in adulthood, body composition was similar; i**x**) the glucose tolerance was decreased in adults.
Conclusion: These results support the hypothesis of developmental programming of vascular, renal and metabolic diseases in adulthood, after oxygen exposure during the neonatal period.

FM 19

Feasibility and usefulness of rapid 2-channel-EEG-monitoring for acute CNS-disorders in the pediatric emergency ward

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Aims: To evaluate feasibility and usefulness of rapid 2-channel-EEG-monitoring in the management of patients with acute encephalopathy admitted to a pediatric emergency ward
Methods: Standard monitoring of vital parameters in the pediatric emergency setting was supplemented by 2-channel-recording of fronto-parietal EEG-activity in all patients with manifest or suspected

acute encephalopathy. Nursing staff was trained in applying EEG-electrodes and in displaying the monitor settings of the signal for patients in all age categories; emergency physicians were trained in interpreting relevant findings (normal/abnormal background activity, ongoing epileptic discharges, relevant side difference, artifacts). Consultant neuropediatricians helped assessing the patient and the EEG either directly if in-house or indirectly via smartphone-video of patients symptoms and monitor display of EEG-signal. Impact of this information for further patient management was assessed prospectively.
Results: Within a 9 months period, emergency EEG-monitoring was applied to 16 patients with acute CNS-disorders. Nurse staff soon became familiar with the placing of electrodes. An interpretable EEG-signal could be achieved in few minutes in every patient and proved to be very helpful in cases with suspected convulsive disorders. It was especially diagnostic in infants with occipital lobe seizure symptoms, in whom either ongoing subclinical epileptic discharges could be detected or abnormal body posturing could be classified as non-epileptic and anticonvulsive over-treatment could be avoided. 2-channel-EEG gave sufficient information about cortical activity improving management of patients cases with other acute CNS-disorders.
Conclusion: Integrating 2-channel-EEG-monitoring in the pediatric emergency setting is feasible and proved to be very helpful for the management of acute encephalopathies. It turned out to be a suitable alternative to standard EEG-recording and saved time and effort.

FM 20

From 1993 to 2012: Does cerebral palsy still affect preterm children?

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Background and aims: *The incidence of cerebral palsy (CP) in preterm children has been reported to decrease. The objective of this study is to compare the epidemiology as well as the severity of CP between 2 ten- year periods in a tertiary care center.*
Methods: Retrospective analysis of a cohort of very preterm infants hospitalized in our level III NICU and born in 1993–2002 (n = 873) and 2003-2012 (n = 987). There was no change in mean birthweight in the 2 periods, but mortality decreased from 14.4% to 10.3% (p = 0.07). Follow-up was offered to the 1632 (87.7%) survivors and performed in 88% of patients. Cerebral palsy was diagnosed at the 18 months visit and was confirmed at a later appointment. In the first years children were evaluated with the Griffiths Mental Development Scales, and with the Bayley Scales, 2nd edition later on. Severity was assessed with cognitive as well as sensorial function, and with the Gross Motor Function Classification system.
Results: 1446 children were evaluated, 68 presented with cerebral palsy (4.7%), among them 32% with spastic quadriplegic, 33% spastic diplegic, 32% spastic hemiplegic and 3% ataxic subtypes. Rate of CP was 2.9% in period 1, 4.2% in period 2, with no difference in birthweight or gestational age between the 2 periods. Assessment with the GMFS showed that 66% in the first period and 80% in the second period had scores 1-2, (p = 0.158), with age at walking significantly lower in period 2 (21.5 months) versus period 1 (30.0, p = 0.03). Preliminary results show a diminishing severity of CP across the years.
Conclusions: Despite improvements in neonatal care and increased survival, CP incidence could have slightly increased, but its quality and severity have changed, and need to be discussed.

FM 21

High prevalence of pathologic copy number variants detected by chromosomal microarray in Swiss-Italian children with autism spectrum disorders

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Introduction: While the vast majority of causes of Autism Spectrum Disorders (ASDs) is still unexplained, 2–20% of ASDs patients presents pathologic Copy Number Variations (CNVs) detected by array comparative genomic hybridization (array-CGH). We collected data from 21 children presenting ASDs. Array CGH detected pathologic CNVs in 6/21 (29%). Our results strongly suggest the application of array CGH as first tier genetic investigation in patients with ASDs.
Materials and methods: Data of children referred to our institution because of ASDs were prospectively collected over 2 years. They included sex, age at first evaluation, results of Autism Diagnostic Observation Schedule and Autism Diagnostic interview (ADOS/ADI), metabolic screening, serologic screening for celiac disease and thyroid function, EEG, brain MRI and array-CGH testing.
Results: Between 1st January 2010 and 31st December 2012, 21 children with ASD were referred. None of them presented characteristic dysmorphic traits suggesting a specific genetic condition, so that all 21 patients underwent array-CGH studies. 6 out of 21 patients (29%) presented pathologic copy number variations. Thyroid function tests, celiac and metabolic screening as well as brain MRI were normal in all 6 children with pathologic array-CGH testing. EEG showed abnormal activity in 3 out of 6 patients (50%) with pathologic array-CGH results, while only 2/14 (14%) children without pathologic array-CGH presented abnormalities at EEG (3 children with normal array-CGH results did not undergo EEG-testing). While comparing patients with and without pathologic array-CGH results, no significant difference was identified in ADOS and ADI scales.
Discussion: In this study, pathologic copy number variations were detected in 29% of children with ASDs undergoing array-CGH analysis. This approach might represent a first tier investigation tool in children with autism spectrum disorders of otherwise unexplained etiology. Noteworthy, in the present case series, most of the investigations performed in children with ASDs (brain MRI, metabolic, celiac and thyroid function screening, ADOS/ADI scales) failed to predict pathologic results at array-CGH analysis.

FM 22

Cardiac follow-up of pediatric oncology patients after cardiotoxic chemotherapy

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Introduction: Anthracyclines (in particular doxorubicine) are commonly used in the treatment of childhood leukemia, lymphomas and other malignant tumors. Their use is however limited by their cardiotoxicity. It is therefore important to monitor the survivors of childhood cancer having received anthracyclines to detect signs of cardiac dysfunction and heart failure
Objectives: To evaluate long term cardiac outcome of all patients treated in the pediatric oncology unit of our institution with anthracycline for childhood cancer and followed in the pediatric cardiology unit between 1991 and 2013.
Methodology: Retrospective study after Ethics Committee approval. Creation of a database containing demographic, oncologic and cardiologic data for all patients less than 16 years of age treated with anthracyclines for childhood cancer and followed in pediatric cardiology. Anthracycline doses were standardized to doxorubicine doses. High doses were defined as doses >300 mg/m^2. Cardiac follow-up was done by echocardiography with measurement of cardiac shortening and ejection fraction.
Statistical analysis of the data.
Results: 305 pts were treated with anthracyclines during this period at the CHUV and underwent long term cardiac follow-up. 3 pts developed cardiac dysfunction. The risk of adverse cardiac evolution is significantly increased with a total high-doses doxorubicine treatment (p = 0.001), with radiation therapy (p = 0.004) and with a combination of radiation and high-dose doxorubicine (p = 0.003). This occurs most often in pts with sarcomas are these patients receive the most frequently this type of treatments. High dose doxorubicine treatment significantly increases cardiac treatment (p = 0.005).
Conclusions: Cardiac follow-up of patients having been treated with anthracyclines is mandatory. Close cardiac follow-up is of particular importance in pts receiving high-dose anthracyclines, pts undergoing radiation therapy, pts needing a combination of high-dose anthracyclines and radiation therapy, in particular pts with sarcomas. Regular follow-up as is done at our institution has permitted early detection and treatment of cardiac dysfunction.

FM 23

Clinical manifestations in Primary Ciliary Dyskinesia: a systematic review

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Introduction: Primary ciliary dyskinesia is a rare genetic disease, characterized by abnormal ciliary ultrastructure and beating pattern, which results in impaired mucociliary clearance. PCD affects primarily the upper and lower respiratory tract but it is a multi-organ disease and can lead to various clinical manifestations. There are many published studies about pathophysiology and diagnostics, but data on clinical manifestations are scarce, particularly for adults. We want to describe the prevalence and severity of various clinical manifestations stratified by age.
Methods: We performed a systematic review searching Embase, PubMed and Scopus for studies describing clinical manifestations of ≥10 patients with PCD.
Results: 48 studies fulfilled our inclusion criteria. They described a total of 1930 patients (mean 40 per study, range 10-257). 21 studies originated from pediatrics or pediatric pulmonology, 10 from ENT and 2 from adult pulmonology departments, 12 came from other clinics. 14 studies described only children, 3 only adults, and 3 did not report the age of their patients. Of the remaining 28 studies that included a larger age range, only 8 stratified their results by age. The eligible studies reported an overall mean prevalence across the studies of 53% for situs anomalies (median 53%, range 11–100%), 61% for bronchiectasis (median 56%, range 9–100%), 79% for cough (median 94%, range 14–100%) and 70% for sinusitis (median 67%, range 10–100%). 20 studies described also PCD connected manifestations affecting other than the respiratory system (e.g. infertility).
Conclusion: Our systematic review showed a widely ranging prevalence of symptoms associated with PCD. The variability of results was mainly explained by the highly selected patient population of the clinics the papers originated from. To characterize the clinical phenotype and its progression over the lifetime, we need larger studies reporting on unselected populations and a wider range of clinical manifestations.
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FM 24

Are electronic cigarettes harmful? The youths' point of view

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Introduction: Electronic cigarettes (EC) are becoming increasingly popular among adolescents, even among those who do not smoke. The objective of this study was to assess whether young people considered EC harmful.
Methods: As part of a larger qualitative study on EC use including 42 adolescents/ young adults (16–26 y.o., 19 females) interviewed in 8 focus groups (FG), participants were asked whether they thought ECs were healthy/harmful. FGs were audio-recorded and transcribed verbatim. Transcripts were analyzed according to a thematic analysis procedure.
Results: Four main results emerged from the analysis. First, overall youths considered EC as less harmful than traditional cigarettes, especially because it was considered as a way to stop smoking (*It is always better to change to electronic cigarettes than to continue to smoke, that I am convinced of* [20 year-old male]). Second, the fact that they were perceived as less harmful also rendered them more socially acceptable (*I am less ashamed,to vap than to smoke a cigarette* [18 year-old female]). Third, EC advertising was reported to very often give a message of wellness, showing people who care for their health (*There is the image of wellbeing anyway. In the sense that I see a lot of people who use electronic cigarettes and explain to smokers for 20 minutes that they take care of themselves, that they don't destroy their health* [18 year-old female]). Finally, youths also indicated that they were aware that there was not enough perspective to assess whether EC has any secondary effect on health (*So far there is no proof that it is harmful, and we don't know in the long term* [21 year-old female]).
Conclusion: Overall young people consider ECs as less harmful than traditional cigarettes even though they were aware of the lack of evidence of secondary effects. The fact that ECs are presented through publicity as a model of wellbeing and that they are socially more acceptable than cigarettes may render them especially attractive to young people. As it is the case for other substances, advertisement regulations should be put in place urgently to cease the progression of this positive image.

FM 25

Hyponatremia in bronchiolitis – a harmful complication not to be ignored

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Introduction: Bronchiolitis is frequent in infants. Against the background of self-limiting course in most cases, severe and potentially life-threatening events should be kept in mind.
Case report 1: A six-week old female infant was admitted with RSV-positive bronchiolitis, poor feeding and recurring apneas. She was term born with regular birth weight and uneventful medical history so far. Initial electrolyte measurements were normal (Na^+ 138 mmol/l). Despite oxygen supply and symptomatic therapy she suddenly deteriorated on day five. After vomiting, with consecutive apnea and bradycardia she presented with prolonged, generalized convulsions with delayed response to medical therapy during bag mask ventilation. Blood gas analysis demonstrated severe hypercapnia (pCO_2 11 kPa) and hyponatremia (Na^+ 113 mmol/l). The patient was intubated, intravenous sodium substitution was started and she was transferred to the ICU. Due to recurring bradyapneas mechanical ventilation was continued for eight days before complete recovery. Sodium substitution was required for two days. Urine sodium levels always remained normal.
Case report 2: An eight-week old female infant was admitted with RSV-positive bronchiolitis and poor feeding. She was a pre-term born twin (35 2/7 weeks) with regular birth weight and otherwise uneventful medical history. The night before admission, she presented first apneas. Initial electrolyte measurements were normal (Na^+ 137 mmol/l). Treatment was started with oxygen supply and symptomatic treatment, including chloralhydrate for sedation. Due to persisting respiratory distress, blood gas analysis was performed depicting hyponatremia (Na^+ 121 mmol/l). Despite sodium substitution hyponatremia persisted and she presented prolonged generalized convulsion with delayed response to medical therapy. She was transferred to the ICU after she additionally developed a Cheyne-Stokes respiratory pattern. Sodium substitution was raised up to 12.3 mmol/kg/24h until serum levels slowly normalized over the following three days.
Discussion: Children hospitalized with RSV bronchiolitis develop apnea in 2–4% and seizures in 1–3%. Both are at least in part attributed to SIADH. The current hypothesis is that hyperinflation of the lungs induces hypovolemia leading to increased ADH release resulting in volume overload and hyponatremia.
Conclusion: We suggest repeated monitoring of sodium levels in infants with bronchiolitis demonstrating apneas or other neurological symptoms as well as in patients with prolonged severe respiratory distress. Hyponatremia should be treated early to avoid life-threatening events.

FM 26

Different indices of inert gas washout measurements define physiological phenotypes

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Background: Gas washout measurements are used to detect ventilation inhomogeneity (VI) in patients with lung disease. Our aim was to assess whether washout tests can help to define physiological phenotypes in children with different lung diseases.
Method: 80 school age children performed three different gas washout measurements, nitrogen multiple and single breath washout (N2-MBW and N2-SBW) and double-tracer gas single breath washout (DTG-SBW). We included each 20 children with cystic fibrosis, with primary

ciliary dyskinesia, who were former preterm and healthy controls. Outcome parameters to represent global VI were lung clearance index (LCI) derived from the N2-MBW and Slope III of the N2-SBW. Specific VI were Scond (diffusion and convection dependent) and Sacin of the N2-MBW, SIII-CO2-DTG and SIII-DTG of the DTG-SBW (acinar VI). Z-scores were calculated based on healthy controls. Clustering of different gas washout indices was performed using hierarchical ward clustering.
Results: Mean values of all indices were elevated in PCD and CF, whereas in former preterm only SIII-CO2-DTG was significantly elevated on a group level (0.6 z-scores). In CF especially LCI and Scond (4.4, 3.4 z-scores) were increased. In PCD beside LCI and Scond also Sacin (1.4 z-scores) was increased. We focused on four to six clusters carrying different characteristics of the respective groups, partly independent of lung disease.
Conclusion: The combination of different indices from gas washout tests can be used to define physiological phenotypes. Longitudinal data using larger numbers will show whether the clusters are disease course-specific or can identify physiological phenotypes independent of age or disease condition.

FM 27

Do Smartphones change adolescents' sleeping habits?

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Introduction: Smartphones allow to be connected to the Internet continuously from almost anywhere. As it is a small device, youths can use it very discretely if needed, even when they are supposed to be asleep. The objective of this study was to assess whether having a Smartphone had an effect on the sleeping habits of adolescents.
Methods: A sample of 208 adolescents (100 females) who did not have a Smartphone at age 14 (T0) were divided in 2 groups depending on whether they owned a Smartphone (Owners, N = 153) or not (Non-owners; N = 55) at age 16 (T1) and compared on sleeping hours on schooldays and vacations at T0 and T1 controlling for socio-demographic and personal variables.
Results: There were no differences between groups in age, gender, family structure, nationality, residence, socioeconomic status, emotional wellbeing, academic achievement, sleeping problems or owning a personal computer or a tablet. No differences in sleeping hours between groups were noted at T0 on schooldays (Owners: 8.2 ± .20; Non-owners; 8.6 ± .12; P = .08) or on vacations (Owners: 9.8 ± .19; Non-owners; 10.2 ± .24; P = .23). However, a difference was observed at T1, with Owners sleeping fewer hours than Non-owners on schooldays (7.5 ± .09 vs. 8.0 ± .19; p <.05) but not on vacations (9.6 ± .12 vs. 9.4 ± .30; p = .55). In a logistic regression controlling for age, gender and sleeping hours on schooldays at T0, Owners were significantly less likely to sleep than Non-owners (Odds Ratio: .44; 95% Confidence Interval: .27:.71; P = .001).
Conclusion: Overall, mean sleeping time decreases as adolescence progresses and young people sleep less than recommended on schooldays. Owning a Smartphone adds to this lack of sleep on schooldays but no difference is observed on vacations. This finding implies that Smartphone owners do not recuperate their insufficient sleeping time on vacations. Health professionals should screen for Smartphone use and sleeping hours of adolescents, especially those complaining of tiredness. Parents should be advised not to allow Smartphones (and other small devices to access the web) in their adolescent's room at night.

FM 28

Co-sleeping in school-aged children with neuromotor disabilities

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Introduction: Co-sleeping is a common practice in infants and pre-school children, dependent on the familial and cultural environment. For older children, health conditions can influence its practice. The aim of this study was to compare the prevalence and determinants of co-sleeping in school-aged children with a neuromotor disability (NMD) versus an age-matched representative general population (GP) sample.
Methods: We sent a written questionnaire on sleep habits and co-sleeping, including the Sleep Disturbance Scale for Children (SDSC), to parents of children aged 4 to 18 years followed in our tertiary neurorehabilitation clinic and through public schools. All descriptive and explanatory analyses were performed with STATA® and significance values set at a p ≤0.05.
Results: We collected 275 NMD and 3083 GP responses (response rates 39% and 27% respectively). Significant demographic differences were found between both populations (NMD vs GP: male/female 1.55 vs 1.02, p-value <0.01; mean age 10.7 years vs 9.6 years, p-value <0.01). There was a trend towards increased regular co-sleeping, defined as occurring at least once weekly, in children with NMD (11% vs 8%, p-value = 0.06). Children with NMD had a significantly higher mean SDSC score, reflecting poorer sleep, further increased when co-sleeping was present.
Conclusion: In our NMD and GP samples, regular co-sleeping was not a frequent practice. It was significantly associated with poorer sleep, which may reflect the special needs of children with NMD during the night. Sleep arrangements within the family should be specifically queried in order to provide appropriate counselling.

FM 29

The maturation of sleep in the first 3 months of life in term and preterm infants

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Introduction: Sleep undergoes major developmental changes during the first years of life. Newborn infant's sleep architecture, lacking classification criteria for NREM and REM sleep, is divided into quiet and active sleep instead. Ongoing brain maturation evolves in the detection of sleep spindles after 4 weeks as well as K-complexes and slow waves by 6 months. Especially slow wave activity has been shown to be a major electrophysiological marker for brain maturation amongst others. Our aim is to map sleep maturation topographically by using EEG at birth and 3 months of life in term and preterm infants, especially finding specific cortical areas with early maturation of slow wave activity. The known delayed brain maturation in preterm infants might lead to differences in the amount or configuration of slow wave activity.
Methods: 20 preterm infants (<32 weeks of gestation) and 20 term born infants are being enrolled. EEG (18 Electrodes) are performed during a natural morning sleep period at term equivalent age (preterm infants) or at the 2nd/3rd day after birth (term born infants) and at 3 months (corrected age for preterm infants). For a preliminary analysis, 9 term (7 boys) and 8 (3 boys) preterm infants are analysed regarding sleep states.
Results: Mean gestation age was 28 5/7 (range 24 3/7 – 30 4/7) weeks for preterm infants and 40 2/7 (range 38 5/7 – 41 4/7) weeks for term infants. Mean birth weight was 1187 g and 3443 g for preterm and term infants respectively. Total sleep time was 145.2 (stderr 19.3) and 134.1 (stderr 22.9) minutes for preterm and term infants, respectively. Infants spent 58.6% and 64.4% in quiet sleep and 40.6% and. 35.6% in active sleep for preterm and term infants, respectively. There were no significant difference between the two groups.
Conclusion and outlook: In a preliminary analysis of the first 9 term and 8 preterm infants, there was no difference between the two groups regarding sleep states. As expected, infants spent about 60% of total sleep time in quiet sleep. In a further analysis the emphasis will be laid on the amount and spatial distribution of slow wave activity and sleep spindle.

FM 30

Co-sleeping in healthy school-aged children

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Introduction: Co-sleeping is a common practice in infants and pre-school children, dependent on the familial and cultural environment. Its prevalence is less known in older children, as are its effects on the sleep quality of the children and their parents. Therefore, we wanted to investigate its practice in a population of healthy school-aged children.
Methods: We sent a written questionnaire on sleep habits and co-sleeping, including the Sleep Disturbance Scale for Children (SDSC), to parents of children aged 4 to 18 years through public schools in and around Lausanne. All descriptive and explanatory analyses were performed with STATA® and significance values set at a p ≤0.05.

Results: We collected responses from 3083 children (response rates 27%, male 48.8% and mean age 9.6 years ± 3.5 years). Co-sleeping was reported for 35.16% children. Frequent co-sleeping, occurring at least once weekly, for 8.01%. Co-sleeping occurs 81.% of time in the same bed. SDSC total scores have a mean of 42.5 ± 8.5, median 41. Through regression analyses, we found that increasing age and co-sleeping practice are significantly associated with lower total SDSC scores (p <0.01 for both), sex not (p = 0.10). Parental educational level and employment are positively associated with the sleep quality (p = 0.04 and P <0.01 respectively), but there is no significant association with the co-sleeping.
Conclusion: We described in a large sample of healthy school-aged children the prevalence of co-sleeping, and its associated factors. We can thereafter compare these data with a population with a neuromotor disability.

FREE COMMUNICATIONS – SSSSC

FM 31

Sleep slow-wave activity reveals developmental changes in experience-dependent plasticity

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Experience-dependent plasticity, the ability of the brain to constantly adapt to an ever-changing environment, has been suggested to be highest during childhood and to decline thereafter. However, empirical evidence for this is rather scarce. Slow-wave activity (SWA; EEG activity of 1–4.5 Hz) during deep sleep can be used as a marker of experience-dependent plasticity. For example, performing a visuomotor adaptation task in adults increased SWA during subsequent sleep over a locally restricted region of the right parietal cortex, which is known to be involved in visuomotor adaptation. Here, we investigated whether local experience-dependent changes in SWA vary as a function of brain maturation. Three age groups (children, adolescents, and adults) participated in a high-density EEG study with two conditions (baseline and adaptation) of a visuomotor learning task. Compared with the baseline condition, sleep SWA was increased after visuomotor adaptation in a cluster of eight electrodes over the right parietal cortex. The local boost in SWA was highest in children. Baseline SWA in the parietal cluster and right parietal gray matter volume, which both indicate region-specific maturation, were significantly correlated with the local increase in SWA. Our findings indicate that processes of brain maturation favor experience-dependent plasticity and determine how sensitive a specific brain region is for learning experiences. Moreover, our data confirm that SWA is a highly sensitive tool to map maturational differences in experience-dependent plasticity.

FM 32

The local regulation of sleep is related to behaviour in children with disorders of consciousness

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Electroencephalographic slow wave activity (SWA, 1–4.5 Hz) during non-REM sleep is known to be regulated in a use dependent manner: globally after prolonged wakefulness and locally after intensive training involving specific brain areas. In the present study we investigate the regulation of SWA in children with disorders of consciousness. We propose to quantify sleep regulation using the build-up of SWA at the transition into non-REM sleep episodes across the night.
We used high-density electroencephalography (128 electrodes) to record sleep in 5 children (4–14 years old) with disorders of consciousness (sub-acute: 1–13 months after brain injury). For all transitions into non-REM sleep following more than 1 min of wake, N1 or REM sleep we calculated SWA in 1-minute intervals. The build-up of SWA from non-REM sleep onset to the subsequent SWA peak was estimated using a linear fitting approach The behaviour was assessed using the Coma Recovery Scale – Revised (CRS-R). Compared to age and gender matched healthy children, the patient's build-up of SWA after transitions into non-REM sleep was significantly decreased over parietal (62% ± 16, p = 0.005) and frontal brain areas (62% ± 14, p = 0.006). Comparing patients with the same aetiology (two patients with stroke, two patients with traumatic brain injury), a higher build-up in these areas was related to a higher behavioural score in the CRS-R.
In conclusion, the build-up of SWA after transitions into non-REM sleep might be a promising marker for residual behaviour in patients with disorders of consciousness. In the future, this marker might provide new diagnostic and prognostic information.

FM 33

Effects of *COMT* genotype and tolcapone on the waking EEG during sleep deprivation

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Background: Catechol-O-methyl-transferase (COMT) regulates dopamine (DA) neurotransmission in the prefrontal cortex and can be selectively inhibited by tolcapone. Previous research indicated that the functional Val158Met polymorphism of *COMT* (rs4680) may influence sleep-wake regulation. To examine a causal role of DA neurotransmission in sleep-wake regulation, we studied the combined effects of SD and tolcapone on EEG activity in healthy volunteers.
Methods: Thirty young men (10 Val/Val, 10 Val/Met, 10 Met/Met genotypes) completed a sleep-deprivation (SD) protocol (40 hours of wakefulness). In randomized, placebo-controlled and double-blind manner, they received 2 x 100 mg tolcapone or placebo after 11 and 23 h of prolonged waking. At 3-h intervals, Karolinska drowsiness tests were performed. Artefact-free recordings with eyes open were analyzed. Effects of *COMT* genotype and SD, and their modulation by tolcapone, on EEG power spectra and specific frequency bands (delta, theta, alpha, beta) were investigated (C3/A2 and O1/A2 derivations).
Results: Prolonged wakefulness increased EEG power in the delta, theta and beta ranges in a *COMT* genotype-dependent manner. In the 8–10 Hz band and in the 13–20 Hz range, the increase was largest in both derivations in the *Val/Met* genotype, whereas between 11–13 Hz the effect was most pronounced in *Met/Met* homozygotes. In the C3/A2 derivation, beta activity was generally higher in the *Val/Val* genotype. When compared to placebo, tolcapone did not modulate the effects of SD on EEG power spectra.
Discussion: The Val158Met polymorphism of *COMT* modulated the effects of SD on EEG theta, alpha and beta activity. This finding indicates that genetically-determined differences in DA neurotransmission interacts with sleep-wake regulation. By contrast, the COMT inhibitor, tolcapone, did not modulate the consequences of SD on the waking EEG.

FM 34

Circadian and sleep homeostatic regulation of cerebral correlates of working memory: time of day matters

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Objectives: Sleep loss-related detrimental effects on cognitive performance are dependent on time of day. Even though a steep decline can be observed in the biological night, cognitive performance has the potential to recover if wakefulness is further extended into daytime. Circadian wake-promoting mechanisms counteracting increasing sleep pressure levels putatively contribute to this recovery. However, the cerebral correlates of such mechanisms remain virtually unexplored.

Methods: Thirty-one healthy young participants underwent a 40-hours sleep deprivation (SD) and multiple nap (NP) protocol. Blood-oxygen-level-dependent (BOLD) activity was assessed during an n-back task scheduled to the end of the biological day (13h and 37h after habitual wake-up) and night (21h after habitual wake-up). Nap sleep efficiency (SE) assessed in the evening immediately after scanning under NP was considered to reflect the participants' strength of circadian wake promotion while the individual NREM delta sleep rebound after SD was assessed to estimate the individual's sleep pressure build-up.
Results: Performance was worse during SD compared to NP, particularly during the night, but recovered during the second biological day. Similarly, the underlying BOLD activity decreased under SD compared to NP in a widespread cortical network at night, whereas an additional extension of wakefulness did not result in a further activity decline. Nap SE in the biological evening negatively correlated with BOLD activity in a postero-lateral hypothalamic area (p <0.05). Higher activity in this area was furthermore associated with the ability to maintain performance at baseline levels under sleep loss during daytime (p <0.05). During the night, BOLD activity in this region covaried with the individuals' NREM delta sleep rebound in response to sleep loss: the lower the BOLD activity, the higher the rebound (p <0.05).
Conclusions: Our data indicate that the impact of sleep loss on cognitive brain correlates largely depends on circadian phase. Overall, our results indicate an important role of hypothalamic structures for the integration of circadian and sleep homeostatic mechanisms to control for human neurobehavioral functions under challenging sleep loss conditions.

FM 35

Heritability of the sleep EEG in early adolescence: Preliminary results from a twin study

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Introduction: Previous studies have shown that the sleep EEG power density spectrum in adolescence is highly variable across subjects. Furthermore, the sleep EEG spectrum undergoes significant changes during this period. In adults, twin studies have found that the variability in the sleep EEG spectrum is largely due to genes. The aim of this study was to exam the degree to which the sleep EEG spectrum during adolescence, a time of significant cortical restructuring, is heritable. To this end, we recorded sleep EEGs in monozygotic and dizygotic adolescent twins between the age of 12–14 years.
Methods: All-night high-density (64 channel) sleep EEG recordings were performed in three monozygotic (MZ; n = 6) and three dizygotic (DZ; n = 6) twin pairs. Sleep EEG spectra were calculated for derivation C3 (average reference) separately for NREM and REM sleep and divided into the following frequency bands: delta, theta, alpha and sigma. Heritability, h2, was defined the correlation between DZ pairs minus the correlation for MZ pairs times two and calculated for each frequency band during NREM and REM sleep.
Results: We found greater similarity between MZ as compared to DZ pairs in all frequency bands. This was true for both NREM and REM sleep.
Conclusions: Our preliminary findings show high heritability of the sleep EEG even during a period of significant cortical development. Many psychiatric disorders have their onset during adolescence and are accompanied by a sleep phenotype. By understanding how genes and environment contribute to the sleep EEG during this period we can open up new avenues for research.
Support: This work was supported by a grant from the Jacobs Foundation (to LT)

FM 36

Identification of subjects with high probability of sleep-disordered breathing in a non-sleepy general population: The NoSAS score

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Introduction: Sleep disordered breathing (SDB) is a highly prevalent disease associated with sleepiness and increased cardiovascular risk. Although sleepy subjects are commonly referred for respiratory polygraphy or polysomnography (PSG) to diagnose SDB, it is more challenging to identify non-sleepy subjects at risk for SDB who should undergo a nocturnal investigation. Our aim was to develop a simple score allowing physicians to identify subjects with a high probability of SDB in a non-sleepy population.
Methods: We used data from the population based HypnoLaus study, in which all subjects underwent complete PSG recordings at home. We estimated the probability of SDB among non-sleepy subjects (Epworth sleepiness score ≤10) from easily available clinical features, taking into account their specific effect size. Using ten-fold cross validation technique, the performance of the proposed score was evaluated in terms of discrimination and predictive values, and was compared to existing scores (adjusted neck circumference, STOP-BANG and Berlin questionnaire).
Results: The score was derived from a sample of 1412 non-sleepy subjects with complete data from the 2121 HypnoLaus participants. 51% were men, with a median age of 57 years [interquartile range 49–68] and a mean body-mass index (BMI) of 25.6 kg/m^2 (± 4.1). The optimal model was based on **N**eck circumference, **O**besity, **S**noring, **A**ge and **S**ex ("**NoSAS**" score). This score allocates 4 points if neck circumference >40 cm; 3 points if BMI ≥25 and <30 kg/m^2; 5 points if BMI ≥30 kg/m^2; 2 points if snoring is reported; 4 points if age >55 years and 2 points for men. Based on its ability to identify subjects with a significant apnea-hypopnea index (>20/h), the score was considered positive if ≥8 points. In terms of area under the curve (AUC), NoSAS performed significantly better than the other scores: AUC = 0.74 for NoSAS versus 0.70, 0.67, 0.63 respectively for adjusted neck circumference, STOP-BANG and Berlin score. Negative predicted values were 0.90, 0.88, 0.95 and 0.80 and positive predicted values were 0.47, 0.43, 0.35 and 0.44, respectively.
Conclusion: NoSAS score is easy to implement in general clinical practice and allows identifying individuals with a high probability of SDB among asymptomatic subjects in the general population.

FM 37

Naps not as effective as a night of sleep at dissipating sleep pressure

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Introduction: The two-process model of sleep posits that two processes interact to regulate sleep and wake: a homeostatic (Process S) and a circadian process (Process C). Process S compensates for sleep loss by increasing sleep duration and intensity. Process C gates the timing of sleep/wake favoring sleep during the circadian night. In this study we examined whether taking six naps throughout a 24-h period would result in the same amount of dissipation of homeostatic pressure at the end of the day as a night of sleep, when time in bed (TIB) is equivalent.
Methods: Data from 46 participants between ages 10 and 23 years were analyzed (mean age = 14.5 (±2.9); 25 females). Slow wave energy (SWE), normalized to account for individual differences in slow-wave activity (SWA; delta = 0.4 to 4.6 Hz), was used as a measure of sleep homeostasis. SWE was defined as accumulated SWA during NREM sleep. In the nap condition SWE of six naps each of 1.5 hours in duration distributed equally during a 24-hour period (TIB = 9 h) was calculated. In the baseline condition, SWE was measured after 9 hours TIB. A paired t-test was used to compare nap and baseline conditions. A linear regression was used to examine whether SWE in nap and baseline conditions varied as a function of age.
Results: SWE was greater during baseline than the nap condition ($t(45) = 4.28$; $p < 0.0001$). No association between age and SWE was found for either the baseline ($r = 0.05$; $p = 0.78$) or nap ($r = –0.07$; $p = 0.66$) conditions.
Conclusions: Our findings indicate that multiple naps throughout the day are not as effective at dissipating sleep pressure as a night of sleep. This is likely due to the influence of the circadian system, which staves off sleep during certain times of day.
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FM 38

Effects of sleep extension on the regulation of food intake in young obese short sleepers

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Introduction: Epidemiological and experimental data indicate that short sleep is a risk factor for obesity. Our study aims to determine whether an extension of time in bed (TIB) in obese short sleepers has beneficial effects on total sleep time (TST) and on the regulation of food intake.
Methods: Thirteen (2 women) short sleepers (<7 hours/night) aged 18 to 25, overweight or obese (28<BMI<35) were studied under two cross-over randomized experimental sessions (8 nights with habitual sleep duration versus 8 nights with TIB increased by >1 h/night). For each of these sessions, sleep was recorded every night by actigraphy and by polysomnography on days 1, 5 and 8. Validated questionnaires of hunger and appetite were administered daily on morning and evening, a food diary was completed during the last 4 days and an ad libitum buffet was served on the last day of each session.
Results: The simple extension of TIB, increased TST from ~6 hours (5:49 ± 4 min), a sleep duration associated with an increased risk of obesity in epidemiological studies, to ~8 hours (7:42 ± 6 min, p <4.10-6), a sleep duration associated with the lowest risk of obesity. This increase in TST was accompanied by a reduction in appetite for fatty and sugary energy rich food (–19%, p <0.04), a reduction in snacks (–46%, p <0.03) and in caloric intake during the buffet (–104 kcal; p <0.05). Furthermore, more the TST was increased during the TIB extension condition, more appetite for energy rich food and caloric intake during the buffet were decreased (rSp = –0.68, p <0.02; rSp = –0.75, p <0.01). Finally, increasing the TST during the TIB extension condition was strongly correlated to the increase in TST on weekend versus week day under usual conditions (rSp = 0.73, p <0.01).
Conclusion: An extension of TIB in obese short sleepers has beneficial effects on the regulation of food intake. These effects would be particularly important for individuals who increase considerably their TST on rest days.

FM 39

High prevalence of thoracic aortic aneurysm in patients with moderate to severe obstructive sleep apnoea

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Background: The incidence of thoracic aortic aneurysm (TAA) is estimated to be six to ten cases per 100,000 patient years. Sixty percent of TAA involve the ascending aorta (AA). Recent findings from observational studies suggest a possible link between obstructive sleep apnea (OSA), thoracic aortic dilatation and aortic events in patients with Marfan's syndrome. Whether there is a high prevalence of thoracic aortic aneurysm in patients with OSA is currently unknown.
Methods: Patients who underwent in-hospital respiratory polygraphy between 1996 and 2014 and had an echocardiography within the same year were included in this retrospective study. The diagnosis of TAA was defined by the following parameters: AA diameter >42 mm for females and >46 mm for males. OSA was defined as mild (apnoea-hypopnoea index (AHI) 5–15/h), moderate (AHI <15–30/h) and severe OSA (AHI >30/h).
Results: 206 patients were included in the analysis. 55 patients (27%) had mild OSA, 48 patients (23%) were diagnosed with moderate OSA and 77 patients (37%) had severe OSA. TAA was identified in 63 of the 206 patients (31%). The prevalence of TAA was significantly higher in patients with an AHI >15/h (39%) when compared to patients with no or mild OSA (21 %) (P = 0.0072).
Conclusion: This is the first study to determine a high prevalence of TAA in patients with moderate to severe OSA. Prospective controlled cohort studies are needed to investigate whether OSA is associated with faster progression of aortic dilatation and aortic events.

FM 40

P-wave duration and dispersion during intrathoracic pressure swings

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Background: A high P-wave dispersion (Pd) has been reported to be a prognostic factor for the occurence of atrial premature beats and atrial fibrillation. There is preliminary evidence that intrathoracic pressure swings which occur, i.e. during episodes of obstructive apnoeas, may promote arrhythmias. Therefore we tested the hypothesis whether an increase of Pd and p-wave duration (Pwdu) can be induced by respiratory manoeuvres in healthy subjects and patients with paroxysmal atrial fibrillation (PAF).
Methods: 12-lead-electrocardiography (ECG) was recorded continuously in 13 healthy subjects and 23 patients with PAF, while simulating obstructive apnoea (Mueller manoeuvre, MM), obstructive hypopnoea (inspiration through a threshold load, ITH) and during normal breathing (NB) in randomized order. The Pwdu was measured manually by using dedicated software for ECG-analysis. The difference between the maximum and the minimum p-wave duration is defined as Pd.
Results: Pd and Pwdu significantly increased during MM and ITH in healthy subjects (+28.6% and +11.9% during MM, +12.2% and +7.1% during ITH; P-value <0.05 for all comparisons). We observed similar findings in the 23 patients with PAF (+17.9% and +4.1% during MM, +4.8% during ITH; P-value <0.05 for all comparisons).
Conclusion: Intrathoracic pressure swings promote an increase of Pd in healthy subjects and in patients with PAF. Our findings imply that intrathoracic pressure swings prolong the intra-atrial conduction time and therefore seem to be an independent trigger factor for the development for atrial premature beats or atrial fibrillation.

FREE COMMUNICATIONS – SWISS PEDNET

SPN 1

Intensity of delivery room resuscitation in preterms does not influence neurodevelopmental outcome at 2 years corrected age

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Introduction: Neonates born below 33 weeks of usually require medical support immediately after birth, the so called "delivery room (DR) resuscitation". We have graded DR resuscitation since 2005 into three levels of intensity. Our study aimed to analyze the predictive value of the intensity of DR-resuscitation on neurodevelopmental outcome at the corrected age of 18–24 months of life.
Material and methods: Retrospective single center cohort study. Neonatal data has been collected prospectively. Included were all lifeborn neonates <33 weeks born between 2005 and 2010 without major malformations. Intensity of DR-resuscitation was classified into three levels: I-mild: stimulation, aspiration; II-moderate: respiratory support by nasal CPAP, mask ventilation; III-heavy: invasive and non-invasive mechanical ventilation (including CPAP) for more than 30 minutes, medication with vasoactive drugs, chest compression. Neurodevelopment at 2 years was assessed with the Bayley scales of infant development II (MDI and PDI scores). The main predictor was the level of DR-resuscitation. Therefore we performed a one way analysis of variance.
Results: 655 neonates met inclusion criteria, median gestational age was 29.6 (± 2.3) weeks and median birthweight was 1344 (± 461) g. All neonates required DR-resuscitation as follows: Level I 5.4%, Level II 23.7%, Level III 69.4%. 323 children had a neuro-developmental

assessment at 18–24 months of corrected age. Mean MDI and PDI scores were 90.34 and 84.70 respectively, without significant differences between intensity levels of DR-resuscitation. Subgroup analysis for neonates born below 28 weeks (n = 61) showed mean MDI and PDI scores of 90.98 and 85.92 respectively, also without difference between the only two levels of DR-resuscitation observed (II and III).
Conclusion: Intensity of DR-resuscitation in preterm neonates defined by our three levels had no independent predictive value for neurodevelopmental outcome at corrected age of 18-24 months. Heavy resuscitation at delivery was not predictive of worse outcome but a more detailed grading of level III DR-resuscitations may be necessary.

SPN 2

Transient dystonia at 6 months of corrected age of very low birth preterm children: correlation with MRI at term and follow up at 18 months

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Introduction: Approximately 7.5/100 births in Switzerland are premature with 0.4% of preterm less than 28 weeks of gestation. Prematurity itself is a risk factor for neonatal complications, such as neurological impairments, in relation with delayed and immature brain development of the premature infant's. Even without visible cerebral injuries, preterm children are at risk of later neurological troubles. One of them is a tone anomaly called transient dystonia, which is present usually at 6 months of corrected age and resolves around 12 months. This anomaly presents itself with truncal hyperextension, toe standing, hyperreflexia and saccadic motricity.
Aims: Define prevalence of transient dystonia in a group of very low birth preterm children. Identify perinatal and antenatal risk factors, correlate this tone trouble with term MRI and neurodevelopmental outcome at 6 and 18 months. Finally, compare these data between dystonic and non-dystonic children.
Methodology: Prospective study from June 2007 to December 2010, University Center Hospital of Lausanne and University Hospital of Geneva including: preterm newborns <29 GA, BW <1500 g without major brain lesions. They all had an MRI at term equivalent age. ADC (apparent diffusion coefficient) and FA (fractional anisotropy) values in different regions of interest (ROIs) of white matter and posterior limb of internal capsula (PLIC) were measured. According to the 6 months evolution, the dystonic and non dystonic preterm infants were recorded. Each baby had a neurodevelopmental evaluation at 6 and 18 months of corrected age with Bayley II. The 2 groups were compared in terms of risk factors and quantitative MRI values.
Results: 114 patients, 34 with transient dystonia (which represent 28.8% of the total groups) and 80 without. 2 were excluded because of cerebral palsy in the follow up at 18 months. The dystonic patients have significantly more perinatal complications, a more abnormal and delayed WM at term with significantly higher ADC values (p = <0.05 for all ROIs of white matter) and finally, significantly poorer mental and motor performances at 18 months. The same was seen for the FA values.
Conclusions: Transient anomalies of tone are frequent in this population and might reflect cognitive and motor disturbances in the long term in very preterm infants.

SPN 3

Very preterm infants show earlier emergence of 24-hour sleep-wake rhythms compared to term infants

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Background: Previous studies show contradictory results about the emergence of 24-h rhythms and the influence of external time cues on sleep-wake behavior in preterm infants compared to term infants.
Aims: To examine whether very preterm infants (<32 weeks of gestational age) differ in their emergence of the 24-hour sleep-wake rhythm at 5, 11 and 25 weeks corrected age compared to term infants and whether cycled light conditions during neonatal intermediate care affects postnatal 24-hour sleep-wake rhythms in preterm infants.
Study design: Prospective cohort study with nested interventional trial.
Subjects: 34 preterm and 14 control term infants were studied. During neonatal hospitalization, preterm infants were randomly assigned to cycled light [7 am–7 pm lights on, 7 pm–7 am lights off, n = 17] or dim light condition [lights off whenever the child is asleep, n = 17].
Outcome measures: Sleep and activity behavior recorded by parental diary and actigraphy at 5, 11 and 25 weeks corrected age.
Results: Sleep at nighttime and the longest consolidated sleep period within 12 pm–6 am was longer (mixed model analysis factor group: p = 0.02, resp. p = 0.01) and activity at nighttime was lower (p = 0.005) at all ages in preterm compared to term infants. Cycled light exposed preterm infants showed the longest nighttime sleep duration. Dim light exposed preterm infants were the least active.
Conclusions: Preterm infants show an earlier emergence of the 24-hour sleep-wake rhythm compared to term infants. Thus, the length of exposure to external time cues such as light seems to be important for the maturation of infant sleep-wake rhythms.

SPN 4

Chronodisruption in Obese Children

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Background: Altered circadian and ultradian blood pressure (BP) and heart rate (HR) rhythmicity has been described in many diseases with increased cardiovascular risk. We tested the hypothesis that rhythmicity in obese children is changed, compared to healthy subjects.
Method: Circadian and ultradian BP and HR rhythmicity was assessed with Fourier analysis from 24-h ambulatory BP measurement (ABPM) in 75 obese children, 45% girls, BMI-SDS median 2.79 (interquartile range (IQR) 2.54–3.41), median age 11.6 years (IQR 9.0–13.6) and compared with an age- and gender matched healthy control group of 150 subjects (45% girls) with BMI SDS median 0.32 (IQR –0.39–1.13), median age 11 years (IQR 8.0–13.0). Multivariate regression analysis was applied to identify significant independent factors explaining rhythmicity variability in this population. Subgroup analysis of non-hypertensive participants was performed.
Results: Prevalence of 24-h and 6-h BP as well as 12-h HR rhythmicity in obese group was lower (p = 0.03, p = 0.02 and p <0.0001). Prevalence of 8-h HR rhythmicity was higher in obese children (p <0.0001). Prevalence of BP rhythmicity excluding hypertensive participants showed comparable results with lower prevalence for 24-h and 6-h BP rhythmicity in obese participants (p = 0,02 and p = 0,03). 24-h BP and HR acrophase was delayed in obese children (p = 0.004, p <0.0001), 24-h BP amplitude was comparable (p = 0.07), 24-h HR amplitude was flattened (p = <0.0001). BP Mesor in obese cohort was higher (p = 0.02), HR Mesor was comparable (p = 0.1). Multivariate regression analysis failed to identify anthropometric or blood pressure parameters explaining the variability of BP and HR rhythmicity.
Conclusion: We showed altered prevalence and parameters of circadian and ultradian BP and HR rhythmicity in obese children compared to healthy controls. This was independent of anthropometric and blood pressure values, suggesting other factors being involved in altered cardiovascular rhythmicity.

SPN 5

Subclinical vitamin B12 deficiency in newborns

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Background: In the past years, we diagnosed subclinical vitamin B12 deficiency in infants with hypotonia, motor delay and/or infant malaise. These infants had elevated methylmalonic acid (MMA) in plasma and urine and plasma B12 in the low normal range for adults. They all responded very well to cobalamin supplements, with resolution of symptoms and catching up motor milestones within a few weeks.

Hypothesis: Subclinical vitamin B12 deficiency may be an unrecognized cause of delayed motor development in early infancy and may originate already in pregnancy. Aim of this pilot study: knowing the incidence of subclinical B12 deficiency in newborns.
Methods: we measured MMA in one drop of blood collected on filter paper in the 4th day of life of 100 healthy newborns at term. For 26/100 infants, we also measured MMA in urine.
Results: 7 newborns had elevated MMA in blood spot; in all of them, as well as in their mothers, vitamin B12 deficiency was confirmed by further investigation. All infants were supplemented with vitamin B12 and normalized their MMA level within 4 to 8 weeks.
Conclusions: 1) Subclinical vitamin B12 deficiency is relatively frequent (7%) in healthy newborns and starts already in pregnancy; 2) B12 deficiency should be considered in differential diagnosis of motor delay and infant malaise, even in the absence of anemia; 3) Screening for B12 deficiency is possible at the pediatrician praxis by MMA measurement in dried blood spot, which seems to be more sensitive than urine analysis.
Outlook: in order to demonstrate the impact of B12 deficiency on development of symptoms, we plan a larger prospective, randomized, placebo-controlled interventional study using dried blood spot MMA as a marker of B12 deficiency and with a clinical follow-up to 24 months of life.

SPN 6

Serum Metabolome of Children with Celiac Disease

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Introduction: Celiac disease (cd) is a complex immune-mediated disorder. Clinical features encompass chronic intestinal inflammation and metabolic alterations. We aimed to analyze the metabolic serum profile of children with cd in order to find undetected deficiency symptoms.
Methods: The serum metabolome of fasting infants with cceliac disease (5 male, 10 female, mean age 9.12 ± 0.88 years) was compared to two age-matched control groups (one fasting: 8 male, 8 female, mean age 7.63 ± 1.19 years; one non-fasting: 7 male, 10 female, mean age 7.46 ± 0.88 years) by ESI tandem mass spectrometry (ESI-MS/MS) using a targeted metabolomics assay (AbsoluteIDQ™ p180, Biocrates Life Science AG).
Results: Comparison of the fasting serum metabolom in cd with the respective control group revealed significant differences in the concentrations of acylcarnitines (e.g. decrease in C2, C16 and C18:1) and phospho-/sphingolipids (e.g. decrease in PC aa C34:1, PC aa C36:4, SM C18:0, SM C18:1). Moreover, we observed significant differences of further metabolite concentrations in these profiles based on the choice of control group (fasting, non-fasting).
Conclusion: For the most part, the cd serum metabolome of our pediatric cohort matched the profiles published for affected adults. However, we identified novel cd-specific dysregulations of serum metabolism exclusively in children, so far unknown in affected adults. These results are promising, as they might lead to future improvement of targeted treatment in pediatric patient care.

SPN 7

Longterm neurocognitive development of children with orofacial clefts

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Introduction: Few studies have focused on the consequences of clefts on cognitive development in children although oral clefts are the most frequent congenital craniofacial anomalies with a worldwide incidence estimated to 1.2–2 per 1000 birth. Anxiety and distress are often mentioned by parents, sometimes worried about the impact of this defect on their child's development.
Objective: The purpose of this research was to evaluate the mental development of five-year-old children affected by orofacial clefts. We aimed to assess whether there is a difference between subtypes of cleft.
Methods: This is a prospective descriptive study of a cohort of patients operated at the CHUV between 2002 and 2008 for clefts. At the ages of five years old, children were evaluated with a thorough history, a detailed neurological examination, and cognitive development was assessed with the Wechsler Scales (WIPPSI-III), a standardized test, which entails subscales of performance, language as well as an intellectual quotient.
Results: The mean values of the different quotients of the WIPPSI-III test for the children with cleft palate (n = 25), cleft lip and palate (n = 19), or isolated cleft lip (n = 10) were in the normal range, and without statistically significant differences between subtypes of clefts. 61% of children used speech therapy and 95% went to normal school.
Conclusions: Children with orofacial clefts have normal longterm neurodevelopment, parents can thus be reassured.

SPN 8

Epidemiology of sepsis in neonates and children in Switzerland– the Swiss Pediatric Sepsis Study

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Introduction: Sepsis remains one of the leading causes of childhood mortality worldwide, yet there is a lack of epidemiological data on culture-proven sepsis in children. The ongoing Swiss Pediatric Sepsis Study prospectively evaluates the epidemiology of blood culture proven sepsis in children in Switzerland.
Methods: Multicentre prospective observational study of newborns and children <17 years with culture-proven sepsis recruited at five paediatric university hospitals and five regional neonatal and paediatric hospitals between 01.09.2011 and 31.08.2014. Patients presenting with signs of systemic inflammatory response syndrome and culture proven bloodstream infection were included in the study.
Results: 822 episodes of culture proven sepsis were registered in the study period. *S. aureus*, coagulase-negative Staphylococci, *E. coli*, and *S. pneumoniae* were the most prevalent pathogens, detected in 479 (58%) episodes. 263 (32%) episodes occurred in patients without previously known risk factors. 324 episodes (39%) were classed as severe sepsis, and in 135 episodes (16%) patients presented in septic shock. In 166 (20%) episodes patients required admission to the intensive care unit due to sepsis and in 54 (6.6%) episodes inotropics were administered. In 55 (6.7%) episodes patients died, 5 deaths occurred in patients without previously known risk factors.
Conclusion: One third of patients with sepsis were previously healthy. Case fatality rate in this patient group was low.

SPN 9

New guidelines for the diagnosis of high-grade vesico-ureteral reflux in children under three months with first febrile urinary tract infection?

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Introduction: The Swiss Society of Pediatrics (2013) recommends a renal ultrasound (US) and voiding cystourethrography (VCUG) to identify high-grade (≥III) vesicoureteral reflux (VUR) in all children less than 3 months with a first episode of urinary tract infection (UTI). Objective: Build a probabilistic diagnostic algorithm of high-grade (≥III) vesicoureteral reflux according to the organism causing the infection and the results of the renal US
Patients and methods: We included retrospectively all the children aged less than three months who were hospitalized between January 2009 and December 2014 with a first UTI (sampling by bladder single catheterization ≥10^5 germs/ml) explored with renal US and CUM without known congenital abnormalities of kidney and/or urinary tract. The criteria for abnormal renal US included renal pelvic anteroposterior diameter >5 mm, calyceal, or ureteral dilatation.
Results: 122 children are included. The average age is 4 weeks. The sex ratio is 97/122.The prevalence of high grade VUR is 10%. For children with *Escherichia coli (E. coli)* UTI (n = 88 or 72% of urine cultures) and renal US abnormalities (n = 13), the likelihood of high-grade VUR is 15%. When renal US is normal, the likelihood of high-

grade VUR is lower than 1%. For children with *Non-E. coli* UTI (n = 34 or 28% of urine cultures) and renal US abnormalities (n = 9), the likelihood of high-grade VUR is 55%. When renal US is normal, the likelihood of VUR grade ≥ III is 15%.
Conclusion: In children <3 months with first febrile urinary tract infection when E.coli is responsible and renal ultrasound is normal, the likelihood of high-grade vesicoureteral reflux is expected to be extremely low (<1%) and CUM is not necessary.
Depending on the pathogen causing UTI and the result of the ultrasound, it is possible to reduce indication of over 60% of CUM (75/122) without underdiagnosing high-grade VUR (likelihood <1%).

SPN 10

Pain Assessment and Management Practices in a medical and surgical pediatric department

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Introduction: Despite international standards for the assessment and management of paediatric pain, pain remains poorly treated. The aim of this study was threefold: a) to establish the prevalence of pain, b) to describe pain management practices, c) to identify barriers to pain management perceived by nurses.
Methods: Following ethics approval, pain prevalence data were collected prospectively in six pediatric and one neonatal unit of a tertiary referral hospital in Western Switzerland over one day (8 am to 8 pm) J0 and retrospectively in the previous 24 hours J-1. Patients had to be hospitalized for at least 24 hours to be included. Participants' medical and nursing notes were reviewed to describe pain management practices, including pain assessment documentation, analgesics prescriptions and administration. A Pain Management Index (PMI) was calculated. The online *Nurses' Perceived Obstacles to Pain Assessment and Management Practices (NPOP)* and the *Pediatric Nurses' Knowledge and Attitudes Survey Regarding Pain (PNKAS)* questionnaires were emailed to all nurses working in the pediatric department.
Results: 83 patients with a median age of 4.1 years (IQR 11.5) were included. Pain prevalence at J0 was 21.7% and 27.4% at J-1. A median of 4 (min-max 0-12) pain assessments per patient and per day was documented. Undertreatment and inadequate prescribed analgesia indicated by a negative PMI was found in 3% and 9% of the participants, respectively. A total of 258 nurses completed the questionnaires (response rate 52.4%). 185 (71.7%) had ≥5 years of nursing experience. Out of 44 obstacles that interfere with good pain management practices, five were identified by more than 30% of nurses, including difficulties in assessing pain in cognitively-impaired children and other non-verbal patients, difficulties of older children to use self-report measures, and difficulties to obtain medical prescriptions for pain relief. Out of 36 pain knowledge items, 16 (44.4%) were under the minimum 70% good response rate and related to pharmacological knowledge of nurses
Conclusion: Pain prevalence rate reported in our study compares well with recent published reports. The discrepancy between prescribed and administered analgesics is explained by analgesia prescribed not adequately administered. The difficulties in assessing pain in non-verbal children can also contribute to inadequate treatment. The results of this study highlight areas for improvement, especially in nursing pain education.

POSTERS SGP/SSP

P 1

Acute hemorrhagic edema of young children: prospective experience with 15 cases

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Introduction: Acute hemorrhagic edema of young children, also known as Finkelstein-Seidlmayer disease or Henoch-Schönlein syndrome of early childhood, is an annular leukocytoclastic vasculitis. Distinctive features characterize this vasculitis, including non-pitting edema and targetoid lesions that develop within 24–48 hours in a well-appearing infant. The condition is considered rare because no more than 400 cases have been so far reported.
Methods: We sought to prospectively address this issue in Bellinzona and Mendrisio. For this purpose our staff members were given a talk twice a year detailing the presentation of acute hemorrhagic edema.
Results: Between September 2007 and December 2013, the suspected diagnosis of acute hemorrhagic edema was suspected in 17 cases by staff members. The diagnosis was confirmed by an experienced senior pediatrician in 15 well doing children (11 boys and 4 girls, 4–35 months of age). They presented with low grade or even absent fever, large red to purpuric targetoid lesions that do not change location within hours, non-pitting indurative swelling and absent mucous membrane involvement or scratch marks. Signs for articular, abdominal or kidney involvement were never detected. Acute hemorrhagic edema was often preceded by a febrile disease (upper respiratory tract illness; N = 5, lower respiratory tract illnes; N = 2, simple febrile illness; N = 2, diarrhea; N = 1, urinary tract infection; N = 1) or by an immunization (N = 1). The cases were managed symptomatically as outpatients and fully resolved within a few weeks. No recurrence or familiarity was noted.
Conclusions: This is the first prospective evaluation of acute hemorrhagic edema of young children. Our experience emphasize its distinctive tetrad: a well-appearing child; targetoid lesions that not change location within hours; non-pitting, sometimes tender edema; complete resolution without recurrence.

P 2

Efficacy of hydrochychloroquine in three children with interstitial lung disease

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Introduction: Hydroxychloroquine has been used as a second line anti-inflammatory agent in interstitial lung disease (ILD), when corticosteroids produce morbidity or minimal improvement. However, experience with this drug remains anecdotal.
Methods and results: We present three children with persistent respiratory distress after birth, unresponsive to conventional management, thus weaning from mechanical ventilation was not possible. ILD was confirmed by lung biopsy in two cases. Treatment with hydroxychloroquine was initiated. Both infants showed clinical improvement, extubation and discharge with oxygen supplementation was possible. In one child diagnosis of surfactant dysfunction (ABCA3 deficiency) was confirmed by testing for surfactant gene mutations. In the other child histological findings of lung tissue revealed pulmonary intestinal glycogenosis. In the third case, lung biopsy was declined by parents because of a previous fatal exacerbation of disease after lung biopsy in a sibling. In this patient, diagnosis of ILD was based on family history, clinical manifestation as well as findings in high resolution computertomography. Because of clinical deterioration we started a trial with hydroxychloroquine at the age of 8 years. This treatment was followed by an impressive clinical improvement with weaning from oxygen, increase of weight, hight and exercise tolerance. Lung function parameter of restrictive lung disease improved within few weeks.
Discussion: "chILD" represents a heterogenous group of rare lung diseases with high morbidity and mortality. Our three children share an impressive clinical benefit to treatment with hydroxychloroquine after unsuccessful therapy with systemic corticosteroids. We advocate a controlled trial with hydroxychloroquine in children with ILD.

P 3

Is it ovarian torsion? A systematic literature review and evaluation of prediction signs

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Objectives: This study aims to identify, through systematic literature review, the most reliable clinical, biological and radiological signs of ovarian torsion in the pediatric population and to compare their diagnostic value.
Methods: Systematic review of the literature, searching Medline, Embase and Cochrane Databases for articles published between January 1990 and January 2014.
Results: From the 946 references initially identified, 14 retrospective publications fulfilled the inclusion criteria, involving a total of 663 episodes of ovarian torsion. Sudden onset abdominal pain with nausea and/or vomiting is the most frequent symptom of ovarian torsion. It can occur at any age, not only in menarchal or perimenarchal patients. Abdominal tenderness is present in 88.4% of patients, whilst only 24% have a palpable mass. Blood tests are commonly requested (51.4% of cases) but are not diagnostic. Abnormalities on plain abdominal radiograph include masses, calcifications and ossified images. Ultrasound has sensitivity for ovarian torsion of 79% and CT scan of 42.2%. There is a significant diagnostic delay at 101.8 hours (median).
Conclusion: Abdominal pain in children and adolescents is difficult to evaluate and the diagnosis of ovarian torsion remains a challenge. Due to its potential complications, we need effective clinical tools. From our review of the literature, it was not possible to develop a diagnostic algorithm. Further research is needed to improve our practice and shorten the delay to diagnosis. Considering the low incidence of ovarian torsion, a multicentre prospective study would be required.
Key words: Ovarian torsion, adnexal torsion, acute pelvic pain, pediatric, children.

P 4

A multidisciplinary, general paediatrics approach to managing pervasive refusal syndrome: a case report

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Background: Pervasive refusal syndrome (PRS) describes children with dramatic social withdrawal who become unable to walk, eat or care for themselves. Literature on PRS is limited and has little focus on management. It is not known if these children can be managed on general paediatric wards and if an integrative medicine approach is useful.
Case report: A 7 years and 4 months old, sensitive girl developed progressing gait disturbances until bedridden, social withdrawal, refusal to eat and weight loss, reduced speech at barely audible voice. Comprehensive diagnostic evaluation at two neurological centres found no evidence of somatic illness. No traumatic events were elicited. She was hospitalized on a general paediatric ward that integrates conventional care with elements of Waldorf special education and Anthroposophic therapies. She was fully integrated in the structured activities of the ward even if she would only sit and watch. She received massages, movement therapy, colour light therapy and Anthroposophic remedies. All therapies were offered as for a younger child, accepting her regression. The only threat was placement of a nasogastric tube in case of further weight loss. Parents were fully integrated in the care. Over the course of four weeks the girl began to talk again, had increased appetite and gave support when moved passively. She made a full recovery within 4 weeks after hospital discharge.
Conclusion: Treatment of children with pervasive refusal syndrome seems possible in a general paediatric ward setting if a multi-disciplinary approach can be offered. Full integration of parents and an integrative medicine approach providing a variety of comforting sensorial experiences appeared helpful in this case.

P 5

Psychosocial development of maltreated children: A prospective study

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Introduction: It is generally accepted that child maltreatment has a potentially devastating impact on the well-being of children. But there are also long-term consequences, including vascular diseases or auto-immune diseases. Previous research has shown that traumatic stress associated with maltreatment can also have severe negative effects on the development of the central nervous system. Furthermore, child maltreatment is associated with an increased risk for problems such as substance abuse, depression, high-risk sexual behaviours, etc. in adulthood. A potential moderating factor is quality of life. But research on psychosocial development and quality of life of maltreated children is lacking. Past studies have been limited to retrospective designs and school-aged children. The present study addresses these limitations by providing prospective data on changes of health-related quality of life, behavior and mental health in the aftermath of maltreatment.
Method: The design is based on three data points: Baseline is collected soon after the child has been reported to a child protections team (CPT), follow-ups take place one and two years after baseline. The sample is drawn from in- and outpatients reported to the CPT at University Children's Hospital Zurich and Children's Hospital Baden, including children from birth up to the age of 15 years. Due to the intrusive topic, difficulties in recruitment and a high attrition rate are to be expected. Therefore, a two year recruitment period is considered. Each participant is matched with a control from the hospital population regarding gender, age, nationality, socioeconomic status and severity of medical treatment. Normed and established instruments will be used for psychosocial assessment of children and caregivers. Inclusion criteria are sexual abuse, physical maltreatment, psychological maltreatment and neglect. Not included are children with a Munchausen Syndrome by proxy. The maltreatment must be substantiated or indicated. The primary caregiver must be informed on the report to the CPTs and must be resident of the German-speaking part of Switzerland. So far, tree maltreated children are included. The study is approved by the ethical review boards and started in February 2014 in Zürich and in October 2014 in Baden.
Results: The study has recently started. Preliminary findings will be available after baseline data collection.
Conclusion: Child maltreatment is a serious and prevalent public health problem, responsible for substantial morbidity and mortality. Results on the long-term impact on children will support policy stakeholders nationally and internationally with arguments to provide more resources to prevention of child maltreatment. This study will be the first to provide data on changes in HRQoL in the aftermath of maltreatment.

P 6

Kawasaki disease: Unusually severe phenotypes

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Introduction: Kawasaki disease (KD) is a systemic vasculitis particularly affecting the coronary arteries whereas systemic arterial injury is rare. Systemic inflammation is also reflected by increased tumour-necrosis factor. Intravenous immunoglobulin (IVIG) reduces the frequency of coronary artery aneurysms from 25% to 5%. Up to 10–20% of KD patients show IVIG-resistance with higher risk of coronary aneurysms. The Kobayashi score was developed for Japanese patients to predict primary unresponsiveness to IVIG. Of the six children (4 males; age range 4 months – 5 years) admitted 2014 with KD, two presented with unusually severe phenotype. We wanted to test retrospectively how the score would have performed, and discuss the most severe cases.
Case reports: A four-month-old female was repatriated from abroad with fever (14 days >38.5°), unresponsive to extended broad intravenous antibiotics. The girl had shown all criteria of KD during inflammation, but the signs were not synchronous. At admission, she was afebrile with no overt clinical signs and a Kobayashi Score of 2; CRP was high (149 mg/l). Echocardiography showed bilateral aneurysms (4 mm) of the coronary arteries. Additional ultrasound revealed dilatation of the visceral and major limb arteries (up to 7 mm). IVIG, high-dose aspirin and oral steroids were administered with rapid drop of CRP within 3 days (28 mg/l). Two months later, ultrasound showed normalized abdominal vessels; steroids were stopped.

Follow-up at 4 months, on low-dose aspirin, showed a persistent left coronary aneurysm (2.7 mm). An eighteen-month-old male presented with fever (5 days), typical clinical signs of KD, high CRP (253 mg/l) and a Kobayashi Score of 1; echocardiography showed mild ectasia (2.5 mm) of the left coronary artery. After IVIG and high-dose aspirin, fever and high CRP persisted for 36 hours; thus a second IVIG dose was given and oral steroids were added. Six days later, he presented again with fever, oligoarthritis (hips, knees) and high CRP (81 mg/l). A single dose of intravenous infliximab (6 mg/kg) induced rapid regression of arthritis and drop of CRP (31 mg/l). At follow-up 2 months later, the boy was asymptomatic with normal echocardiography; thus aspirin and steroids were stopped. Detailed family history revealed Japanese origin (maternal great grandfather) as a possible reason for IVIG- and steroid-resistance.
Conclusion: Few KD patients present with either oligosymptomatic or severe phenotype. IVIG-resistance or systemic vessel involvement is rare in Swiss KD patients. Detailed family history and routine ultrasound revealed Japanese origin and systemic vessel injury in one patient each. IVIG-resistant KD requires additional treatment, e.g. steroids or infliximab, to interrupt the inflammatory process. Current guidelines define "high-risk patients" and recommend intensified first line therapy to reduce complications.

P 7

Trimethylaminuria in a patient with cystic fibrosis

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Case report: A 2 9/12-year-old boy with cystic fibrosis was referred to our emergency unit by his pediatrician due to moderate dehydration after displaying symptoms of acute gastroenteritis for four days. The most striking finding on physical examination was a strong foul and fishy odor emitted by the patient himself. Our diagnostic workup detected *adenovirus* in the stool specimen, a deep throat swab revealed a co-infection with enterobacter cloacae and *klebsiella pneumoniae* but no evidence pointing towards a recurrent colonization with pseudomonas aeruginosa. Liver failure, kidney failure and urinary tract infection were ruled out as possible causes of the odor. A thorough medical history disclosed that the striking odor had been noticeable since birth, making an underlying persistent infection unlikely. We therefore measured the urinary concentration of free trimethylamine, which was markedly increased and thus highly suggestive of the metabolic disorder trimethylaminuria. Due to the autosomal-recessive inheritance pattern of trimethylaminuria we aimed to confirm our suspected diagnosis by genetic testing, allowing genetic counseling for the family and justifying an initiation of treatment.
Discussion / Conclusion: Strong odors in patients with cystic fibrosis must primarily lead to exclusion of pulmonary bacterial colonization and secondary organ dysfunction. When infections (especially of the respiratory and urinary tract), organ dysfunction, poor hygiene and unfavorable dietary habits have been ruled out, metabolic disorders as dimethylglycinuria and trimethylaminuria must be considered, especially since symptoms of trimethylaminuria might be significantly reduced by several therapeutic measures. Our poster provides an overview of the clinical characteristics, diagnosis and management of this rare metabolic disorder.

P 8

A rare presentation of Kawasaki disease: bilateral aneurysm of the humeral arteries

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Introduction: Kawasaki disease (KD) is a vasculitis of medium-sized vessels, most commonly diagnosed in young children after a prolonged febrile course and typical clinical signs. The most feared complication of KD is the development of coronary artery aneurysms leading to stenosis and/or thrombosis, potentially to myocardial ischemia and even death. In approximately 2% of the patients, KD vasculitis also affects peripheral arteries. We report here a case of KD with an usual presentation. The boy presented with bilateral aneurysms of the humeral arteries, which retrospectively lead to the diagnosis of KD.
Case: An 18-months-old boy of Swiss and Thailandese origin was referred for bilateral pulsatile "balls" in the axillary region (fig. 1). Cardiac MRI revealed bilateral fusiform aneurysm of the humeral arteries (fig. 2). Thereafter, the boy was sent to cardiology clinic. Echocardiogram showed dilatation of the left and right coronary arteries, measuring maximum 4 mm. Cardiac function and ventricular kinetic was normal. The boy was born full term and had been treated during the first months with antibiotic prophylaxis because of pyelocalyceal dilatation. At 3,5 months of age he was admitted to hospital because of persistent fever. There was no other symptoms a part from a loss of appetite and a generalized rash, particularly on the chest, the hands and feet. Urine culture was positive for Klebsiella oxytoca and the boy received intravenous antibiotherapy with ceftriaxone. Despite the treatment for urinary infection, the fever persisted and the boy underwent echocardiography to exclude coronary aneurysm related to Kawasaki Disease. Echocardiography was normal and the fever ceased. During the following days, he developed a cough and rhinitis. Finally, a diagnosis of viral illness and urinary infection was retained.
Conclusion: Peripheral arterial aneurysms related to KD have been previously described as late complication. This is the first reported case of Kawasaki Disease diagnosed retrospectively because of symptomatic peripheral arterial aneurysm.

P 9

Zufriedenheit von akut hospitalisierten Jugendlichen im Kantonsspital Freiburg

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Introduction: In Switzerland, age-appropriate facilities for adolescents admitted to hospital are rarely provided. Usually, they are either surrounded by children or adults and their specific needs might not be met. The aim of this study was to describe the satisfaction of hospitalized adolescents (age 11–17) in acute settings of a Swiss hospital, to explore potential differences between adolescents hospitalized in pediatric or adult settings and to explore associations between their satisfaction and variables like age, education, the reason for hospitalization.
Methods: We conducted a descriptive survey. Adolescents aged between 11–17 years, being hospitalized in an acute care setting of a Swiss tertiary hospital were invited to participate. We developed an internet-based survey that was adapted from the questionnaire for children in two Hospitals in Switzerland (Chappuis et al, 2008). Our survey was available in French and German. Inclusion criteria in the study were: being aged between 11 and 17 years, understanding and speaking French or German, having spent at least 24 hours in hospital, being hospitalized for an acute reason and had signed the informed consent. Participants rated 41 items investigating 3 domains: nursing care, the emotional dimension and infrastructures on Likert like rating scales (ranging from very satisfied to not satisfied at all) and visual analog scales. Descriptive and correlational statistics were performed for the whole sample and in sub groups of age (11–14 vs. 15–17) and settings (pediatric vs. adult).
Results: The data were collected between January 2013 and December 2013. Out of 107 screened patients, 102 adolescents participated on this survey. Overall, the adolescents evaluated their hospitalization positively and 45 adolescents were satisfied and 35 very satisfied. Lower satisfaction could be observed for infrastructures, specially the decoration in the pediatric unit, the rooms without WLAN and inappropriate games. Food was another item adolescents rated as less satisfying (38). The only differences in satisfaction between the two age-groups we found was a lower satisfaction for adolescents at age 15–17 with their hospital stay (39 from 102, $p = 0.35$). No differences in satisfaction with settings could be identified. Instead 51% of the adolescents could imagine to be hospitalized in a unit particular for adolescents.
Conclusion: The results of this survey revealed that most of the adolescents are very satisfied with their hospital stay. Some minor investments in the infrastructures could improve the already high level of satisfaction.

P 10

Hematocolpos at the pediatric emergency department: a case report

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We present a case of hematocolpos diagnosed in our paediatric emergency department.
Case report: a 14 year-old girl initially presented twice to the emergency department with lower abdominal pain associated with nausea, a decreased appetit, dysuria, irregular transit and rectal bleeding. On physical examination, a palpable mass was found in the left lower and the suprapubic quadrants. Constipation was diagnosed

and treated with an enema and oral laxative. On the third consultation, pain was described as crampy, present 24 hours a day. It had begun a month earlier, with lower back pain, nausea, tenesmus, pollakiuria and it was increased after defecation. Patient was not sexually active and had not started menses yet. Mother and aunts had had a late menarche too. On physical examination, a regular well defined round and painful mass was palpable up to the umbilicus, without peritoneal signs. On external gynaecological examination, a painful protrusive hymeneal membrane with hematoma was visible at the vaginal introitus. On abdominal ultrasound, a massive hematocolpos was confirmed, with a small uterus, haematomata and dilated fallopian tubes. Under general anaesthesia, patient underwent a hymenotomy using a cruciate incision which allowed the evacuation of 700 ml of blood, and a hymeneal reconstruction.
Conclusion: Hymeneal imperforation is present in 0.1% of female patients and a hematocolpos must be included in the differential diagnosis of abdominal pain in adolescent patient consulting the emergency department.
Key words: hematocolpos, pelvic pain, paediatric.

P 11

Generalized lymphadenopathy and B-symptoms – it's not always cancer

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Background: Generalized lymphadenopathy combined with B-symptoms (fatigue, night sweats, weight loss) require the exclusion of a lymphoma. However, the discrimination between EBV associated lymphoproliferation and malignant lymphoma can be challenging.
Case presentation: A 16-year-old male was referred to the hospital after a CT scan of neck/chest/abdomen and a lymph node biopsy with primary suspicion of lymphoma. These investigations were performed because of progressive generalized lymphadenopathy, fatigue and weight loss. He had a history suspicious of an EBV-infection one year ago; fatigue and splenomegaly persisted thereafter. The lymphadenopathy had had a fluctuating character and the first manifestation coincided with the start of isotretinoin for severe acne and improved after discontinuation of therapy one month before presentation at the hospital. The patient presented in good general condition with moderate lymphadenopathy and splenomegaly. The weight loss was attributable to a voluntary diet. Laboratory investigations revealed a discrete anemia, slightly elevated LDH and bilirubin and raised ESR. Together with the positive Coombs test this was compatible with an alloimmunhemolytic process. Congruently the bone marrow aspirate showed a markedly stimulated red cell line with no signs of atypical cells. Serology showed an elapsed EBV infection, however, EBV-PCR was highly positive. A discrete lymphocytopenia was present, with slightly decreased T- and NK cells. Total IgM was markedly decreased, likely to be secondary. Extensive histopathology of the lymph node with an initial differential diagnosis of an angioimmunoblastic lymphoma was consistent with a chronic-active EBV-infection (CAEBV). EBV persistence was not colocalized to the aberrant T cell population. T cell receptor clonality was not detected.
Discussion and conclusion: As a lymph node biopsy should give a clear diagnosis, every result needs to be interpreted in the context of age, symptoms and clinical findings. Equally, if a patient presents with symptoms and signs of a lymphoma, alloimmunohemolysis can be a sign of a non-malignant process, such as infectious diseases. EBV-PCR in the blood is necessary to search for EBV persistence, even if serology shows an elapsed infection pattern and gives no hint of an active disease. However, the rare NK/T-cell lymphomas of childhood are important differential diagnoses.

P 12

Failure to thrive and signs of neglect – expression of a genodermatosis?

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We report on a 3 months old female who was referred to our institution with failure to thrive and a presumptive oral thrush. She had not been drinking well from the bottle, been increasingly cranky, and eventually stopped gaining weight. The white coverings on the tongue, the palatine, and the buccal mucous membranes had resisted antifungal treatment over three weeks. On admission, the otherwise inconspicuous baby seemed somewhat neglected in the bodily hygiene: waxy hair, scurf, retroauricular scaling, and long arched fingernails. Moreover, the mother's fingernails caught the attention with a yellowish, grotesquely thickened and brittle aspect. These had been assigned by clinical diagnosis to the dominantly inheritable condition pachyonychia congenita (PC). We suspected this disorder of keratinisation to have been transmitted to our patient. Indeed, the early appearance of oral leukokeratosis has been described as a characteristic trait of one of the subtypes of pachyonychia congenita (PC-K6a). Our patient and her mother were connected with the patient registry led by an international PC consortium, which offers genetic testing and aims at favouring studies on this rare disease. To date, mainly symptomatic treatment options are at hand to alleviate the stigmatising cosmetic effects and functional impairment caused by nail thickening, painful keratoderma or, as was the case in our patient, oral involvement. She immediately profited from a softer nipple with an enlarged opening and started gaining weight again.

P 13

Sudden headache in children: don't forget bleeding

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Case report: A previously healthy 15 y.o. girl was admitted for sudden severe frontal headache, vomiting, photophobia and nuchal pain which appeared the day before, while she was dancing. There was no trauma nor fever. She had neck stiffness without other neurological anomaly. Blood tests showed no inflammation. Lumbar puncture showed 32'000 G/L erythrocytes, 40 G/L leucocytes and negative gram coloration. She received one dose of ceftriaxone. The day after, persistence of vomiting and intense headache motivated an injected cerebral CT-scan, which showed a subarachnoid hemorrhage (SAH) coming from an aneurysm of the anterior communicating artery. She was then transferred to a neurosurgical unit where aneurysm was coiled. She recovered well.
Discussion: SAH presents with acute and severe headache, which may occur during a physical exertion, and can be associated with a brief loss of consciousness, nausea and/or vomiting, and meningismus. Most aneurysmal SAH occur between the age of 40 and 60, however young children and elderly people can be affected too. Hypertension, tobacco smoking and family history are the most witnessed risk factors. Differential diagnosis includes meningitis, migraine crisis and acute cervicalgia. Lumbar puncture reveals an elevated opening pressure (>20 cm H_2O) and an elevated red blood cell count (>10'000 G/L), with or without xantochromia. Precoce native cerebral CT-scan shows a subarachnoid hemorrhage in 95% of cases. Cerebral angiography should be made to seek anatomic cause of bleeding: ruptured aneurysm (85% of cases), arteriovenous malformation or tumor. Treatment is endovascular coiling or surgical clipping of the aneurysm. It has to be performed during the first 72 hours. SAH is associated with a consequent mortality rate (22%) but pediatric patients usually have a good recovery.
Conclusion: Sudden intense headache with meningismus should raise the suspicion of SAH. Diagnosis is made by native cerebral CT-scan. Angiography is necessary to specify the cause and guide the treatment.

P 14

Protein-Losing Enteropathy, a Severe and Challenging Complication in a Child With Fontan Physiology

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Introduction: Protein-losing enteropathy (PLE) is a rare, but severe long-term complication of Fontan procedures. The pathogenesis and pathophysiology are not yet completely understood and the treatment mainly consists of dietary restrictions. Despite different therapeutic approaches, it remains a severe condition with an overall estimated 10-year survival about 70 %. In some patients hemodynamic lesions can be identified and surgical or interventional therapy to relief venous or arterial obstruction is indicated. We will present a case, where PLE could be treated effectively, by enlarging an obstructed pulmonary vein.
Case: A 4-year-old boy with left atrial isomerism and single right ventricle presented to our emergency department. He had a complex surgical history ultimately leading to a Fontan physiology with bidirectional, bilateral cavo-pulmonary connections and connection of the liver-veins to the pulmonary artery. Since two weeks he had periorbital edema and increasing abdominal distension. He was also coughing for the last 6 days and presented with mild tachypnea and

dyspnea. A chest- X-ray showed right-sided consolidation, leading to the diagnosis of an atypical pneumonia, which was treated by antibiotics. Ascites and pleural effusion were detected by sonography. Hypoproteinemia and hypogammaglobulinemia strengthened the suspicion of a protein-losing enteropathy. Enteric protein loss was documented by abnormal stool alpha 1-antitrypsin. Under treatment with diuretics and low-fat/protein-rich diet edema, ascites and pleural effusion nearly disappeared within three days and the boy was discharged without gastrointestinal or respiratory symptoms. Echocardiography one month later showed again a slightly accelerated flow in the common right-sided pulmonary vein ostium. Despite clinical improvement we decided to perform a cardiac catheterization because of suspicion of a pulmonary vein stenosis. An obstruction of the right pulmonary veins was confirmed by catheterization and was initially treated interventionally with stent implantation in the common right pulmonary vein ostium to rapidly relieve the stenosis. Compression of the pulmonary vein by the conduit was suspected and a surgical intervention was performed few weeks later. By replacement and extension of the extracardiac cavopulmonary connection and enlargement of the pulmonary vein, relieve of venous obstruction could be achieved. At 3 months-follow up he hasn't shown recurrence of the protein-losing enteropathy.
Conclusion: Protein-losing enteropathy is a rare but severe complication of Fontan procedures with poor prognosis and limited therapeutic options. It's crucial to rule out hemodynamic causes amenable to surgical or interventional treatment.

P 15

Water intoxication in patients with anorexia nervosa – a potentially life-threatening condition not to be missed

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Introduction: Water intoxication with severe hyponatremia is perceived as a rare complication of psychogenic polydipsia in patients with anorexia nervosa, with only three published case reports. The aim of this abstract is to present potential life-threatening consequences of psychogenic polydipsia. We report on an adolescent patient suffering from anorexia nervosa, who was assigned to our clinic in a critically ill condition after a seizure of unknown etiology.
Case report: A 15 year old girl developed a generalized seizure while swimming in a public pool. At the time of hospital admission there was no additional information about the patient's past medical history. At the emergency department, the patient complained about visual disturbances while we observed an altered level of consciousness in combination with severe bradycardia. We interpreted these signs as raised intracranial pressure and decided to secure the airway via intubation. The cranial computed tomography showed no signs for a tumor, cerebral bleeding, oedema or cerebrovascular insult. The patient was transferred to the intensive care unit, where first laboratory results revealed a severe hyponatremia of 122 mmol/L of unknown origin. When her mother arrived, we were told that this girl had been suffering from anorexia nervosa with a history of excessive water intake and even possibly higher water intake before the weight control at the family doctor's practice on the day of admission. Following controlled correction of hyponatremia the clinical condition improved and remained stable under normal fluid intake.
Discussion: Both, psychogenic polydipsia and excessive water intake, before weight measurements to fake a satisfying weight is a common behaviour in patients with anorexia nervosa. As a consequence, the risk of severe hyponatremia and water intoxication might be higher than expected from the few published case reports. Measurement of specific urinary weight is not a reliable test to detect excessive water intake, as measurement during daytime reveal low values also in healthy people depending on their fluid intake.
Conclusion: This case report shows the potential life-threatening complication of excessive water intake in a girl with anorexia nervosa. Based on our case report and our experience with other patients, testing sodium is recommended even beyond the refeeding treatment period at least at larger intervals for both outpatients and inpatients.

P 16

An atypical case of myocarditis

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Indroduction: Myocarditis is a disease of adult and pediatric patients, and is defined as an inflammatory disorder of the myocardium that is typically caused by a viral infection. Symptoms and signs are variable, with a clinical presentation ranging from subclinical signs to cardiogenic shock, arrhythmias, and sudden death. We report a case with an acute, atypical presentation.
Case report: A previously healthy 15 year old boy was admitted presenting symptoms of intermittent thoracic pain over the last 2 days. Upon the day of admission the pain has changed to a persistent, constant pattern. Clinically, the patient was afebrile and presented with a stabbing precordial thoracic pain, that worsened with respiratory movements and was associated with prodrome symptoms of fatigue and myalgias 2 days prior. A pathologic ECG showed ST-segment elevations on leads V4, V5, V6. Pathological laboratory findings included an elevated CK (365 U/l), CKMB (17 µg/l) and troponine T (0.348 µg/l), which were significally elevated in the following two hours [CK (809 U/l), CKMB (48 µg/l) and troponine T (0.894 µg/l)], suggesting a heart inflammatory disease. Echocardiography demonstrated diminished ventricular function with a left ventricular ejection fraction of 38% and mild mitral insufficiency. Further work-up with an MRI scan showed a left ventricular ejection fraction of 43% with involvement of the anterior, inferior wall of the left ventricle and an associated pericardial effusion. Based on these findings treatment was initiated that consisted of monitoring, supportive measures and Milrinone in order to perevent circulatory collapse, while immonuglobulin and corticosteroids were administered. This treatment approach led to complete remission.
Discussion: The clinical presentation of myocarditis is variable and may include a variety of presenting signs and symptoms ranging from heart failure to nonspecific signs, such as fatigue or nausea. Mean age at diagnosis is 9.2 years of age. Typical ECG findings consist of sinus tachycardia, low voltage complexes and ST segment and T wave changes. Laboratory tests may reveal elevated cardiac enzymes. The reported case is atypical because of the age of presentation, the hyperacute onset, the initial absence of typical symptoms and the overall benign, presenting general condition of the patient.
Conclusion: Even though clinical myocarditis is uncommon in infants and children, it should be suspected in children with an acute onset of heart failure and no associated structural heart disease, especially after a viral prodrome.

P 17

Lipschütz ulcer: a case report

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A 14-year old girl presented with rapid onset multiple vulvar ulcerations associated with intense pain. She was febrile, and had coryza and sore throat since 3 days. She had regular menses. She denied being sexually active or sexually abused. She never had presented oral aphthous ulcers or other significant diseases. On physical examination she was in good general condition. Cardiopulmonary auscultation was normal. She had an hyperemic pharynx. She had normal female genital morphology with 3 deep kissing ulcerations on the labia minora, with the greatest one of about 1 cm diameter. Blood cell count showed normal values, C-reactive protein was 46 mg/L. Serologies for Epstein–Barr virus (EBV), citomegalovirus (CMV), herpes simplex virus (HSV) and syphilis were negative as well as the urine and the blood cultures, the culture and the polymerase chain reaction (PCR) for HSV from ulcer swabs. The PCR assay for respiratory pathogens performed on the nasopharyngeal swab was positive for parainfluenza virus 3 (PIV3) Supportive treatment included anti-inflammatory drugs and opioids to control the pain. We administered prophylactic broad-spectrum antibiotic (amoxicillin/clavulanic acid). The wound care was performed with topical anaesthetics and sulfadiazine. Given the severe dysuria, it was necessary to place a urinary catheter for six days. The patient showed a good clinical progress and was discharged in 9 days, with a total resolution and re-epithelisation of the vulvar lesions in 4 weeks. Acute genital ulceration, or Lipschütz ulcer, is a rare entity characterized by painful, necrotic ulceration of the vulva or lower vagina that usually occurs in sexually inactive adolescent girls. It's been linked to several infectious causes (e.g. EBV, CMV, influenza A virus, mumps virus, salmonella, mycoplasma and with Lyme disease)

but, in most cases, it's impossible to identify a pathological agent. The disease is characterized by the acute onset of single or multiple, large and deep ulcers with raised borders, covered by exudate or adherent eschar. Kissing lesions on opposing surfaces are characteristic. Intense pain and dysuria are almost always present. The onset of the ulcers may be preceded by influenza-like or mononucleosis-like symptoms. Diagnosis is mainly clinical. Laboratory tests are useful to exclude other causes of genital ulceration. Biopsy is not helpful and the histologic findings are nonspecific. It may be necessary in case of suspicion of specific skin disease (eg, pyoderma gangrenosum). The differential diagnosis is extensive and includes veneral and non veneral infections, noninfectious causes, traumatic lesions and malignancy. Treatment of acute genital ulceration is mainly symptomatic based on topical anaesthetics, superpotent topical corticosteroids, and oral analgesics. A short course of oral corticosteroids systemic corticoids is reserved to patients failing to respond to topical agents. A broad spectrum antibiotic should be considered to cover bacterial superinfection or vulvar cellulitis.

P 18

Congenital melanocytic nevus, a case report

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We report the case of a newborn with a giant congenital melanocytic nevus (CMN). No clinical problems or use of drugs during pregnancy, no family history for skin lesion. Physiological neonatal adaptation. At birth, a hyperpigmented black patch of skin was present on his back and left buttock. It was a smooth nevus except for a cerebral-like surface on the left gluteus, without hair, with irregular edges, around 20 x 20 cm in size. Physical examination also revealed several smaller disseminated ‹satellite› nevi, mostly spread on both legs. We managed the diagnose in collaboration with the Kinderspital in Zurich. The baby is going to be followed with serial neurologic examinations and developmental assessments and at the age of five months magnetic resonance imaging (MRI) will be performed. At the moment no indication for a surgical approach is given. Congenital melanocytic nevi occur in 1 to 3% of births, large (>20 cm in adulthood) or giant (>40 cm in adulthood) CMN occur in about 1 in 20,000 and are frequently accompanied by ‹satellite› nevi. The nevus could enlarge in proportion to the child's growth and could change in appearance. For this reason the differential diagnosis with melanoma is very important. The risk of a malign transformation into melanoma is correlated with the size of the giant nevus but is not critically higher than in general population. The posterior axial location and the presence of several satellite nevi increase the probability of malignant degeneration. Another complication is the proliferation of melanocytes in the central nervous system known as neurocutaneous melanosis that can be asymptomatic or account for neurologic symptoms (seizures, hydrocephalus). The risk raises with the size of the nevus and the coexistence of satellite nevi. For this reason these patients should be followed with neurologic examinations and screened with a gadolinium enhanced MRI of the CNS during the first six months of life. Surgical removal is often desired for large nevus because of their psycosocial and cosmetic outcomes. Nevertheless, recent studies show that surgical approach is not the best therapeutic option, expecially by dermoabrasion. This procedure cannot affect or modify the penetrance of the nevus that interests the deepest layers of the skin since birth. In addition it cannot prevent a possible malignant trasformation and the resultant scar might mask it. At last, repigmentation is often observed after dermoabrasion and most of giant nevus have a spontaneous depigmentation with child's growth.

P 19

Cardiovascular risk factors among men 18–20 year of age in Southern Switzerland: preliminary data

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Introduction: Atherosclerotic cardiovascular diseases start during childhood and adolescence. To further investigate this issue, we promoted the cross-sectional study "Cardiovascular health among men 18–20 year of age in Southern Switzerland".
Methods: Eligible for the study, which had been approved by the local Ethics Committee, were the Swiss male citizens living in Southern Switzerland, who obligatorily undergo a thorough physical and psychological evaluations to assess fitness for recruitment into the army (except for those affected with chronic diseases or permanent disabilities).
Results: Between April and December 2014, a first group of 258 individuals (among the 1080 who underwent the obligatory evaluation for recruitment into the army) agreed to participate the study. After signing an appropriate consent form, they were subjected to a variety of supplementary evaluations. Preliminary data-analysis indicates the following: cigarette smoking in 89 (34%); body mass index ≥25.0 kg/m^2 in 64 (25%); blood pressure ≥140/90 mm Hg in 55 (21%); pulse wave velocity ≥7.50 m/sec (95th centile) in 53 (20%); uric acid ≥450 µmol/L (N = 53, 20%); cholesterol HDL ≤1.0 mmol/L in 42 (16%), total cholesterol ≥5.2 mmol/L in 16 (6%); and 25-OH-vitamin D_3 ≤50 nmol/L in 38 (15%) individuals. Questionnaire information on lifestyle was also obtained but has not been, so far, analyzed.
Conclusion: We are expecting to investigate a total of 700–800 participants (end of the study presumably spring 2016). The preliminary data-analysis indicates the frequent existence of cardiovascular risk factors among men 18–20 year of age living in Southern Switzerland.

P 20

Lympho-monocytosis: think about Fetal Hemoglobin!

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Introduction: Juvenile Myelomonocytic Leukemia (JMML) is a rare disease (incidence 1.3 case per million). Its initial presentation with non-specific clinical (high fever, cough, rash and mild hepatomegaly) and biological (leukocytosis, monocytosis) findings can easily be mistaken, furthermore when in combination with a confirmed infection.
Case report: A 12-month-old girl was hospitalized in the intensive care unit for acute respiratory insufficiency with high fever, cough, rash and mild hepatomegaly: first diagnosis is pneumonia. Blood findings showed important leucocytosis of 55 G/l with monocytosis of 12% (11 G/l), myelocytes of 2.5% (1.4 G/l) and lymphocytosis of 24.5% (13.5 G/l). Blood smear showed significant dysplastic features in all three lines. Hemostasis was abnormal (prolonged aPTT) explained by positive Lupus anticoagulant. All these elements were correlated with the positive Mycoplasma pneumonia (MP) found in bacterial analysis. Subsequently she was treated with 10 days oral erythromycin. Due to persistent leukocytosis with monocytosis, we conducted a large differential diagnosis which led us to complete more analysis; viral PCR or serology (CMV, EBV, Parvovirus B19, HSV-6) and bacterial culture (toxoplasmosis, Cat-scratch disease) turned up to be negative. This misleading MP infectious path ended as soon as the result of the Fetal Hemoglobin (HbF) returned 22%, highly abnormal for her age, and suspicious of JMML. Indeed, 50% of JMML patients present an elevated HbF for age and 65% with monocytosis higher than 5 G/l. Findings fulfilled the JMML diagnosis criteria (no BCR/ABL fusion, no blast in the bone marrow analysis and positive heterozygote PTPN11 mutation). She received hematopoietic cord blood stem cell transplant 4 months after the initial pneumonia.
Conclusion: It appears again that JMML is a rare and difficult disease to identify. In front of poor prognosis with the only curative treatment (overall survival rate of 52% after hematopoietic stem cell transplantation), it definitely requires a rapid diagnosis. Persistent monocytosis higher than 1G/l (one of the diagnosis criterias), even with concomitant infection, should "always" lead to search for an elevated Fetal Hemoglobin.

P 21

Cushing syndrome after bilateral lensectomy

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Iatrogenic Cushing syndrome induced by oral and parenteral corticosteroid administration is a well-known complication and necessary precautions have to be taken. Cushing syndrome, however, following treatment with glucocorticoid-containing eye drops is a very rare complication. To the best of our knowledge there have been only 4 reported cases in the literature. Herein, we present an infant boy who developed Cushing syndrome due to dexamethasone-containing eye drops following bilateral cataract extraction to prevent post-operative inflammatory complications. Three months after initiation of

dexamethasone therapy, the patient presented with cushingoid facies, nephrocalcinosis, and failure to thrive. The diagnosis of iatrogenic Cushing syndrome was made and dexamethasone-containing eye drops were significantly reduced aiming for discontinuation. Follow-up examinations revealed catch-up growth and a slightly less pronounced nephrocalcinosis.
Conclusion: Ocularly administered corticosteroids may have substantial systemic side effects in infants.

P 22

Brain-lung-thyroid syndrome – update on the clinical spectrum of a heterogeneous disorder

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Background: Brain-lung-thyroid syndrome (BLTS, OMIM# 610978) is caused by mutations in the NK2 homeobox 2 (NKX2-1; TTF1) gene affecting the three NKX2-1 expressing organs brain, lung and thyroid. The hallmark of the syndrome is the triad of benign hereditary chorea (BHC), infant respiratory distress syndrome (IRDS) and congenital hypothyroidism (CH). However, the clinical spectrum and severity of symptoms vary widely, even within families. Regarding the increasing number of published mutations and heterogeneous phenotypes a clinical synopsis is needed for all specialists involved to be aware of the different symptoms and signs of BLTS.
Objectives: Summarizing all available cases of NKX2-1 related disorders to provide a detailed clinical overview of BLTS.
Methods: We performed a systematic review of literature by Medline search. All subjects with proven NKX2-1 mutations and description of at least one symptom of BLTS were included. For genotype-phenotype association studies, we compared different functional domains of the protein and type of mutation with specific phenotypes by Fishers exact test.
Results: We identified 227 subjects (balanced sex distribution) with 129 different mutations. Brain was affected in 94% of cases, lung in 56% and thyroid in 68%. In 16 patients death was reported, thereof 15 patients died of lung disease (12% mortality of 127 patients with lung disease). Patients with brain involvement suffered from choreoathetosis (87%), developmental delay (60%) and muscular hypotonia (38%). Patients with lung disease showed IRDS (42%), recurrent lower respiratory tract infection (57%) and chronic interstitial lung disease (21%). Only 50% of patients with CH were detected by neonatal screening (mean TSH 100 mU/L), while 50% were diagnosed later (mean TSH 37 mU/L). In 184 subjects, information on all 3 organs was available for genotype-phenotype analysis: Only 47% suffered from the complete triad of BLTS, brain-thyroid-phenotype occurred in 27% and 16% showed isolated brain involvement. In a detailed genotype-phenotype analysis, we found no clinically relevant correlation between type or localization of the mutation with brain, lung, or thyroid disease or the combination of the phenotypes.
Conclusions: Regarding the increasing number of published cases, a clinical update is needed for all pediatricians to be aware of the different symptoms and signs of BLT syndrome. We provide a detailed clinical overview with new insights in incidence of heterogeneous symptoms of the syndrome.

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About an adolescent boy with 45,X/46,XY karyotype

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Introduction: The 45,X/46,XY karyotype is rare with an estimated incidence rate of less than 1/15'000 live births. It represents a wide spectrum of phenotypes, from Turner females to phenotypically normal males with varying degrees of genital ambiguity. Prenatal studies showed that 95% of 45,X/46,XY male fetuses will have normal male genitalia. It can be assumed that most of them live undiagnosed as normal males.
Case report: A 14-year-old adolescent boy was referred to our outpatient clinic. Due to an increased level of alpha-fetoprotein in prenatal screening, an amniocentesis was performed, which showed a karyotype 45,X[9]/46,XY[21]. During childhood no further investigations were performed. Clinical examination showed no anomalies except a mild Madelung deformity and a spontaneous onset of puberty with a Tanner Stage 3 and testicular volumes 6 ml and 8ml respectively. The boy was at the 10th percentile for height with a height SDS corrected for parental target height around –2.0 SDS. Gonadotropins and free testosterone levels were within normal pubertal ranges. The karyotype in a blood sample was 45,X[1]/46,XY[9], FISH nuc ish (DXZ1x1,DYZ3x0) [13/200], ish(DXZ1+,DYZ3y)[5/100]. All further investigations were normal, including a screening for celiac disease and hypothyroidism, echocardiography, abdominal and testicular ultrasonography.
Discussion: Overall, clinical knowledge about patients with a karyotype 45,X/46,XY is limited, especially in male patients without any signs of genital ambiguity. It is well known, that most of the male adult patients with a karyotype 45,X/46XY have short stature. Treatment with growth hormone is controversial and likely with a poor response. Improved growth velocity was seen only when growth hormone therapy was started early in life. Most male children experience spontaneous pubertal onset and all normally virilized males have no need for testosterone treatment after spontaneous pubertal onset. Tumor risk seems to be significantly related to the gonadal differentiation pattern. In individuals with mild undervirilization and bilaterally descended testes the risk seems to be low.
Conclusion: Since the relative distribution of both cell lines in different organ-systems is impossible to know at the time of clinical evaluation, all 45,X/46,XY children should undergo evaluation similar to that performed in Turner Syndrome. The management should be individualized for each patient. A close follow-up through puberty with regard to growth and pubertal development is recommended as well as annually testicular ultrasounds. A growth hormone treatment is not indicated in our patient.

P 24

Hyperparathyroidism primarily presenting with arterial hypertension and brown tumor in an adolescent girl

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Primary hyperparathyroidism (PHT) may present with mild asymptomatic hypercalcaemia only. However, with rising serum calcium levels typical clinical signs are bone pain, nephrolithiasis, peptic ulcer disease as well as psychiatric conditions. We report an 17-year old adolescent girl who presented clinically with arterial hypertension and a brown tumour as additional signs eventually caused by PHT. She was referred to our Outpatient Clinic by an oral surgeon to exclude PHT based on the presence of a histologically confirmed brown tumour of the jaw. The patient had been complaining of unspecific bone and abdominal pain and chronic fatigue, during the preceding months. The year before referral the patient was started on candesartan because of arterial hypertension that unfortunately was not investigated further. Eventually, blood and urine chemistry as well as intact-PTH level confirmed the diagnosis of PHT.

	Units	Reference	date 04.02.15	date 28.10.14
total calcium	mmol/l	2.1–2.55	3.67	4.08
ionised calcium	mmol/l	1.13–1.3	2.01	2.01
inorganic phosphate	mmol/l	0.8–1.55	0.63	0.72
creatinine	µmol/l	45–84	59	63
alcaline phosphatase	U/l	<187		1179
iPTH	pg/ml	15–65	1116	978.9
25-OH-D3	nmol/l	49–134		30
urinary calcium	mmol/l			3.77
urinary creatinine	mmol/l			1.95
urinary calcium/creat	mmol/mmol	<0.6		1.93

Further, the radiograph of the left wrist showed osteitis fibrosa cystica. The echography of the neck showed one single hyperdense structure suggesting a parathyroid adenoma, that eventually was confirmed by parathyroid scintigraphy (99mTc-MIBI, SPECT). Genetic testing excluded MEN type 1 / 2. Surgical excision of this adenoma is planned. Secondary arterial hypertension is much more common in children and adolescents than in adults. Therefore, secondary causes of arterial hypertension, like PHT, should always be excluded before starting antihypertensive treatment.

P 25

A 19-year-old adolescent with short stature and scrotal tumour

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Introduction: Primordial short stature can have many different causes. In addition to maternal factors (e.g. placental insufficiency), genetic or skeletal disorders may be found in the child. It is important to identify the underlying aetiology in time, since not only the risk of diabetes is increased, but also the prevalence of insufficient gonadal function and malignancy, e.g. in mixed gonadal dysgenesis (MGD).
Case report: A 19 year old adolescent presented with primordial short stature (2500g, 47 cm at birth, GA 40 weeks). History revealed urinary tract infections in earlier childhood and double kidneys as well as mildly impaired intelligence. Puberty was delayed with an onset at the age of 16 years. Phenotype was male with well-proportioned short stature, except for a broad chest. He presented dysmorphic signs like low-hairline, low-set ears, multiple pigmented naevi; there were no cardiovascular or renal anomalies. The external genitals showed a normally formed penis with two intra-scrotal hypotrophic testes with a volume of 5 ml each, in the presence of nearly adult pubertal stages P5, G4-5. The palpation of the scrotum revealed an indolent scrotal tumour well separated from the testes. Endocrinological investigations identified a mixed gonadal dysgenesis with 46,XY(75%)/45,X(25%) karyotype in lymphocytes and normal testosterone production. Elevated FSH was indicative of testicular dysgenesis. Ultrasound investigation of the tumour showed a tubular structure suspected to be a hypoplastic uterus. Surgical exploration showed a circumscribed tumour histologically proved to be a rudimentary uterus without evidence of malignancy. Because of sufficient gonadal function and male phenotype the testes remained intra-scrotal and will be checked by ultrasound regularly.
Conclusions: In male and female children with primordial (or non-familial) short stature and even discreet dysmorphic signs of Ulrich-Turner's syndrome, a karyogram should be performed in time, since several types of gonadal neoplasms have been described in MGD. MGD conditions are accompanied by a wide phenotypical range and comprise the entire spectrum from normal testes or ovaries, unilateral streak gonads with contralateral testis, ovary or uterus to bilateral streak gonads. This is the first report of an intra-scrotal uterus which is well delimited from the testis in a male patient with MGD. Furthermore, there is a controversial discussion of the proceeding in male patients with MGD because of the increased risk of testicular malignancy. Though fertility may be already impaired, we decided not to perform orchiectomy in this patient since there were no indicators of malignancy, a well-accessible intra-scrotal location of the testes and an otherwise normal gonadal function. Regular clinical and ultrasound examinations should detect malignant degeneration in time.

P 26

Combinations and correlations of diabetes specific antibodies at diagnosis of IDDM

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Introduction: Insulin dependent diabetes mellitus (IDDM) is a typical diagnosis in children and ado-lescents, classically presenting with clinical signs of polydipsia, polyuria and ketoaci-dosis. To confirm the suspectetd diagnosis we measure the known autoantibodies of this autoimmune disease with destruction of beta-cells in the pancreas. In a retrospec-tive study of all new patients with suspected IDDM between 2000 and 2008 we evalu-ated the presence of diabetes specific antibodies (DSAB) at time of diagnosis and the following two years and studied the diagnostic value of the presence and different combinations of these DSAB.
Methods: We evaluated all charts of new IDDM patients diagnosed in KKJ KSA between year 2000 and 2008, concerning age (0–16.2 y), sex and DSAB (cytoplasmatic islet cell AB, **ICA**, AB against Glutamatacid decarboxylase, **GADA**, AB against Tyrosin phosphatase IA-2, **IA2A** and AB against insulin, IAA). The results and correlations of DSAB are seen in 3 different age groups for boys and girls. Analyzed datas include all parameters at start as well as one and two years after diagnosis.
Results: Out of 103 new patients in these 9 years 93 (f: 34/m: 59) were included in the study. 24% were in the infant group, 26% in scholar age and 50% in the pubertal group. In 61 patients all 4 DSAB were measured initially, beeing all positive in 5%, 3 AB pos in 31%, 2 AB pos in 34%, 1 AB pos in 18%, but in 12% no AB was pos despite typical clinics. To confirm the diagnosis of IDDM at least one of the tested AB should be positiv, which was the case in 88% if we had all 4 AB measured. If only 3 AB have been tested the combination of **GADA**, **IA2A** and **IAA** showed the highest sensitivity and at least one AB was positve in 87%. The tested combination of the 2 AB **IA2A** and **GADA** measu-red at least one of the AB positive in 84%. If only one of the AB ist tested the sensitivity of beeing positive was 66% for **IA2A**, 54% for **GADA**, 51% for **ICA** and 30% for **IAA**.
Conclusion: Because the sensitivities of the tested AB combinations vary only little, it can be deba-ted if all 4 AB should be analyzed to confirm the diagnosis of IDDM or if the testing of 3 AB **(GADA, IA2A, IAA)** or only 2 AB **(GADA, IA2A)** is sufficient in the clinical routine.

P 27

Aldosterone synthesis defect in 2 sibling infants

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Case: We describe the case of preterm monochorionic diamnioniotic twin girls with an aldosterone synthesis defect. The pregnancy was complicated by a twin-twin transfusion syndrome, which was successfully treated by laser therapy at 18 weeks gestational age. Following premature rupture of membranes at $28^{2}/_{7}$ gestational age, the twin girls were delivered by cesarean section at $30^{1}/_{7}$ weeks. Twin A (former acceptor) with a birth weight of 1490 g, was intubated for about 36 h and given surfactant for respiratory distress syndrome, while twin B (former donor) with a birth weight of 1240 g, required nasal CPAP. On the 9th day of life following an uneventful postnatal course, both twins were found to have hyperkalemia (venous) with a potassium level of 7.9 mmo/L and 7.7 mmol/L as well as hyponatremia with sodium values of 124 mmol/L and 122 mmol/L, respectively. Blood glucose values were normal and the 17-OH progesterone in the newborn screening on the 4th day of life was normal for both infants. Treatment with furosemide and sodium bicarbonate as well as sodium substitution was started and further investigations performed. Both twins had a grossly elevated renin level with a relatively low aldosterone level and normal to low ACTH level consistent with isolated mineralocorticoid deficiency. Renal sonography excluded kidney malformations and showed normal adrenal glands in both infants. Fludrocortisone therapy was started and sodium substitution at high dose was continued. Both the hyperkalemia and the hyponatremia resolved under therapy and plasma renin level normalized. The diagnosis of an aldosterone synthesis defect (likely reflecting an aldosterone synthase deficiency) was made and genetic analysis to identify the specific underlying genetic mutation is pending.
Discussion: Electrolyte disorders are quite common in VLBW infants during the first days of life. However, when hyperkalemia and hyponatremia occur simultaneously and typically in the second week of life, high suspicion is warranted for a genetic adrenal disorder such as congenital adrenal hyperplasia possibly causing life-threatening mineralocorticoid and glucocorticoid deficiency. In isolated aldosterone deficiency due to mutations in the CYP11B2 gene, the biosynthetic terminal steps of mineralocorticoid production are defective, so that aldosterone levels are low or inappropriately normal, while plasma renin activity and aldosterone precursors (18-hydroxycorticosterone) are increased. Fortunately, the therapy is quite simple, and the needs of fludrocortisone and sodium supplementation decrease with age, reflecting the physiological reduction in circulating renin and aldosterone observed in healthy infants.

P 28

Fertility preservation in paediatric and adolescent patients in Switzerland – where do we stand today?

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Infertility is a severe consequence of many cancer treatments with a major impact on quality of life of long term survivors. Despite advances in reproductive technologies and a number of guidelines fertility preservation is not routinely discussed with paediatric patients and their parents. We conducted a survey concerning fertility preservation counselling and applied fertility preservation measures (FPM) in all 9 paediatric oncological centres of Switzerland. All nine centres

answered. Four (4/9) centres had a standardized program for fertility preservation. Two of them (2/4) for both pre- and postpubertal patients. The two other institutions (2/4) had a program only for postpubertal patients. In one half of the centres, counselling was by haematologist alone. Centres with an available SOP (N = 4) offered an interdisciplinary counselling. Time point of counselling was within the first week of treatment. In the other centres no schedule was defined. The Swiss Childhood Cancer Registry reported in 2013 230 new cases of cancer in children and adolescents. In 125 (54%) of them FPM were performed. The most frequently applied technology in females was cryopreservation of ovarian tissue (19 prepubertal and 12 postpubertal girls). In boys, the most frequently applied methods, was sperm banking (62 postpubertal boys). Lack of time and insufficient know-how were the most frequently mentioned reasons for not recommending and applying FPM. In conclusion fertility preservation counselling and measures are performed roughly in one half of paediatric oncological centres in Switzerland, leaving a large number of patients not taking benefit from this opportunity. Better education of the staff, introduction of a standardized algorithm, availability of structured time slots to perform FPM and financial support are needed to improve and facilitate the fertility preservation in paediatric oncology patients.

P 29

Chest pain and fatigue – when lung and bowel meet together

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Introduction: Involvement of the respiratory tract is a rare but well known extraintestinal manifestation of inflammatory bowel disease (IBD). Pulmonary complications usually follow the onset of IBD, however, in some cases respiratory symptoms precede the gastrointestinal symptoms.
Case: A fourteen-year-old boy presented in our emergency with chest pain and fatigue for one month, but without fever or cough. Chest x-ray (CXR) showed a basal infiltrate and pleural effusion on the left side. Blood tests revealed mildly elevated inflammatory markers (CRP 24 mg/l, ESR 24 mm/h) and hypochromic microcytic anaemia (HB 124 g/l). Suspecting pulmonary tuberculosis (TB), sputum was induced, but neither acid fast bacilli nor other pathogens were detected; a tuberculin skin test and an interferon gamma release assay were normal. The child was treated with amoxicillin/clavulanate but without any improvement: inflammatory markers and pulmonary infiltrates on CXR remained unchanged. CT-scan only revealed bilateral pleural effusion and infiltrates. Bronchoscopy demonstrated discrete mucosa inflammation, and BAL did not reveal any pathogens. Biopsy of the lung showed mildly elevated eosinophils in the alveolar septum, but no signs of TB; blood tests revealed persistent eosinophils (16%, 2400 (ul). After ruling out other conditions associated with eosinophilia (ABPA, Churg Strauss, SLE, etc.) an eosinophilic pneumonia was suspected and oral steroids (prednisolone, initial 1 mg/kg) were started. Within a short time, the boy was asymptomatic, but lung function as well as CXR improved slowly. Steroids were tapered after two weeks, but continued for one year as inflammation markers persisted on a low level. During treatment a weight loss of 15 kg was observed and chronic diarrhoea developed. Consequently, an upper and lower gastrointestinal endoscopy was performed with the diagnosis of ileo-colic Crohn`s disease. As a consequence, a treatment with Infliximab and Azathioprin was started. At the same time, bronchoscopy was repeated and demonstrated multiple aphthous mucosa lesions. BAL only revealed elevated neutrophile granulocytes but no eosinophils.
Conclusion: Among the extra-intestinal manifestations of Crohn's disease pulmonary complications are relatively rare but can precede the onset of IBD. In patients with otherwise not explained chronic pulmonary disease investigations towards IBD should be evaluated.

P 30

Establishment of a patient oriented bowel preparation protocol before colonoscopy

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Background: Protocols for bowel preparation before colonoscopy in children vary between centers. Many use polyethylene glycol (PEG) solution because of its good cleaning results. However, due to its salty taste and high volume requirement, administration via nasogastric tube is often necessary. In 2012, we introduced a new protocol using sodium picosulphate (Picoprep®) in two doses a day before colonoscopy, with additional intake of clear fluids of patients' own choice. The aim of this study was to assess the acceptance of the new protocol by the patients.
Methods: In 2014, consecutive patients admitted to our day care unit for colonoscopy were asked to answer an anonymous questionnaire.
Results: Twenty-two patients (median age 13 y [range 5–17]; 15 (68.2%) male) returned the questionnaire. 21 found the protocol comprehensible, and all patients were satisfied with the information provided concerning the bowel preparation. The taste of Picoprep® was acceptable for fourteen. The mean oral fluid intake (mostly water, followed by broth and syrup) was 3.7 l [range 1–7.5], 13 (72%) children >9 y drank ≥3 l and 2 (66.7%) children <9 y drank ≥2 l. No patient needed nasogastric tube administration of the laxative or the required fluid volume. 21 (94.5%) patients complied with dietary recommendations three days before colonoscopy and 12 (54.5%) patients took Movicol® three days before colonoscopy. The average time between Picoprep® intake and the first bowel movement was 2.3 hours [range 0.3–6.6]. 12.8 hours (range 4–21.9) were between the first and the last bowel movement after Picoprep® intake. No colonoscopy had to be cancelled due to inappropriate bowel cleaning. In all patients the terminal ileum could be intubated.
Discussion/Conclusion: Our new bowel cleaning protocol was well-received by the patients. The laxative Picoprep® was well-tolerated and allowed bowel preparation at home. All colonoscopies were successfully completed. With the overall good acceptance and sufficient bowel cleaning, we will continue to use the new protocol.

P 31

When a grunting hides a battery

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Introduction: Button battery ingestion's incidence has increased in the last years with expanded use of electronic devices requiring such batteries. When lodged in the esophagus, they are a cause of serious injuries via three different mechanisms. The first one being electrical discharge, the most clinically significant with mucosal injury appearing in less than 2 hours. The second mechanism, as for every foreign body, is pressure necrosis. The third mechanism is leakage of alkaline electrolyte (happens mostly in an acidic environment such as in the stomach). For larger batteries, the upper esophagus is the typical site of impaction with an often atypical clinical presentation leading to a delay in diagnosis.
Case presentation/summary: A 13 months old, healthy and fully vaccinated patient presents to our ER with a history of 5 days of continuous fever. Other than a refusal to swallow solids (liquids are tolerated), an occasional grunting in his sleep, and an impression of discomfort according to the parents, the history was inconclusive. We also had a completely normal physical examination, with an afebrile child who was playing in the examination room. The intermittent grunting and fever prompted a plain chest X-ray which revealed a 2 cm diameter metallic object lodged in the upper third of the esophagus. We transferred the patient to a university hospital for endoscopy. Upon extraction of the button battery, a posterior erosion of 4 cm of the proximal esophagus and transmural necrosis was found. A CT-scan showed air in the parapharyngeal space but no signs of mediastinis. The patient received IV amoxycillin and clavulanic acid and a total parenteral nutrition with relay by nasogastric tube. TOGD follow-ups showed a good evolution with complete resolution of the esophageal tear 17 days after the incident, allowing reintroduction of non solid food. Reintroduction of soft solids was difficult due to repetitive episodes of blockage and occasional choking on liquids. This is why endoscopy was performed 8 weeks after the incident and showed persistence of the posterior tear with cicatricial stenosis requiring esophageal dilation. Long term follow-up will tell how often such dilatation will need to be repeated.
Conclusion: Ingestion of batteries is a medico-surgical emergency and they should therefore be removed as quickly as possible. Known complications to esophageal lesions upon battery ingestion include local perforation, tracheoesophageal fistulas, esophageal strictures, ex-sanguinations after fistulization into large blood vessels and death. In this case, the clinical presentation was extremely poor and aspecific, despite extensive esophageal lesions caused most likely by the electrical discharge. The duration of the presence of the battery is unknown, as the ingestion went unwitnessed but more than 5 days ago. Luckily for our patient, there were no signs of fistulizations. Most

batteries <12–15 mm pass into and through the stomach and therefore run a spontaneously benign course through the gut. Batteries >12–15 mm in diameter are more likely to become lodged in the upper esophagus and therefore have a great and rapid potential of serious injury. Most of such ingestions lead to rapid onset of dysphagia, hypersalivation, thoracic pain, cough, fever and parents are usually able to date the time of ingestion. However, absent to poor clinical presentation can render the diagnosis difficult, especially in young nonverbal children with unwitnessed ingestion. **Clinician's vigilance must remain high and x-ray should rule out any suspicion of battery impaction. General prevention of these accidents is still largely insufficient, as button batteries are found in most household utensils.** Two batteries with a diameter <12 mm could replace the large batteries, and be safer for children.

P 32

Abdominal pain and constipation in children as initial presentation of lymphoma

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Case report: A 12 y.o boy known for chronic constipation was seen by his pediatrician with a history of 6 months of abdominal pain getting worse since 1.5 month. A laxative treatment was tested for some days but was unsuccessfull. One week later, detailed history showed that during the same period he presented few episodes of vomiting, progressive asthenia, pallor, and weight loss. A new physical examination was relevant for abdominal mass without any palpated adenopathy. Echographic workup revealed a 10x8 cm pseudo-cystic heterogeneous subumbilical mass. An abdominal MRI showed an important thickening of distal ileum associated with retroperitoneal adenopathies. A mass biopsy confirmed abdominal diffuse large B-cell lymphoma (DLBCL).
Discussion: Abdominal pain in children is a frequent consultation motive. Constipation is a common etiology. In the presence of persistant abdominal pain, it is recommended to review the presence of any red flags (ie abdominal mass, deceleration of linear growth,…). Persisting abdominal pain in itself, even though not a recognized alarm sign, could indicate further diagnosis workup including echography. DLBCL are classified among non-hodgkin lymphoma and account for approximately 10% of them. Mean age at first presentation of DLBCL is 11.4 y.o. Symptoms are non-specific and related to primary localization. Of note, only 14% of children with this disease will present B-symptoms (fever, weight lost, night sweats). A fast workup is necessary in order to stop progression and potential complications (ileus, digestive perforation).
Conclusion: Common symptoms such as abdominal pain and constipation can also hide uncommon diagnosis. Persisting abdominal pain can be initialy the only presentation for abdominal lymphoma. It requires active look up for red flags and should be investigated.

P 33

Exclusive breastfeeding of a vegetarian mother leads to symptomatic cobalamin and other vitamin deficiencies in a 9 month-old child

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Introduction: The most common cause of vitamin B12 deficiency in infants is insufficient dietary intake. Humans do not synthesize cobalamin, which is mostly found in food of animal origin. Vitamin B12 participates together with folic acid in DNA synthesis and is essential for hematopoiesis and for the development of the nervous system. A strictly vegetarian diet of a breastfeeding mother can cause pernicious anemia and serious neurological abnormalities in exclusively breastfed infants. These manifestations are only reversible in case of early therapeutical intervention.
Case report: Here we present an exclusively breastfed 9 month-old girl, who exhibited fatigue and irritability since 1 month, episodic vomiting during the last 3 weeks, and presented to hospital because of the appearance of edemas on feet, hands and face. Food diversification did not succeed because the girl refused any food other than breast milk. She did not receive any vitamin D prophylaxis. The patient has two healthy older brothers. The mother is vegetarian since 20 years and was supplemented with iron during the third trimester of her third pregnancy. During clinical examination the girl appeared slightly hypotonic, tired although alert, and pale. Weight and height were within normal percentiles. Neurological examination showed moderately delayed locomotion, decreased movements of lower limbs and lack of the right Achill tendon and both patellar reflexes. Clinical and radiological signs of mild rickets were also observed. Laboratory tests revealed hypoalbuminemia, macrocytic hyperchromic anemia and vitamin B12, vitamin D and iron deficiency. Urinary organic acid analysis showed a slightly increased methymalonate, an indirect sign of vitamin B12 deficiency. Other metabolic problems causing vitamin B12 deficiency could be excluded. Intramuscular vitamin B12 injections were used for 3 consecutive days, then weekly.
Discussion/Conclusion: Vitamin B12 deficiency is well known in vegetarian adults, but can also occur in infants, especially if exclusively breastfed. A vegetarian diet of the mother can lead to developmental delay already in utero, as well as to protein, calcium, and vitamin and iron deficiency. Infants have less storage capacity than adults and clinical signs of deficiency become apparent between 4 and 12 months of age. Theoretically, vitamin B12 deficiency could be detected by blood tests at birth. Therefore, obstetricians and midwifes should be aware of the importance of monitoring and prevention of vitamin B12 deficiency, particularly in vegetarian or vegan women during pregnancy and while breastfeeding. Secondly, pediatricians should recognize signs of vitamin B12 deficiency such as weakness, fatigue, irritability, pallor, vomiting, diarrhea, icterus, apathy, hypotonia, failure to thrive and developmental delay. In infants with B12 deficiency, other conditions such as a congenital deficiency of intrinsic factor, gastrointestinal diseases or intestinal malabsorption should be excluded.

P 34

Reptiles in households with young children – is the potential risk known?

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Introduction: Keeping exotic pets might be a potential risk for human health – especially if young children are living in the same household. "Reptile-Associated-Salmonellosis" (RAS) has been reported as gastroenteritis, or invasive disease especially in children younger than 6 months of age. We report of a 3 months old boy with reptile-associated non –typhi salmonella gastroenteritis due to exposure to a gecko kept indoors by his mother.
Case: A 3 months old breastfed male infant was referred to our emergency ward in late summer with fever (up to 39. 1°C) and mucous diarrhoea since 2 days. The past history was uneventful. There were no contacts to persons with gastrointestinal symptoms. On examination, he was in a good general condition; however a poor hygiene status of the child as well as mother was apparent and bloody stools were noted. Furthermore, anogenital thrush as well as eczema of the groin, neck and perioral area was noted. Inflammatory markers (leukocytes, neutrophils and c reactive protein) were not elevated. Blood culture was drawn and the child was admitted to the pediatric ward for further observation. Stool culture revealed *Salmonella enteritidis,* serovar offa. Antibiotic treatment with ceftriaxon for 4 days was started, followed by amoxicilin for 4 days according bacterial resistance. Under antibiotic treatment fever disappeared and diarrhoea as well as hematochezia stopped after 3 days. On detailed request, the mother reported of a gecko and, until recently, a tortoise living in the family's' household. The cage of the gecko was usually cleaned in the kitchen. Also, the gecko was frequently petted by the family members.
Conclusion: In the presented case, RAS related gastroenteritis is very likely. As often, owners are unaware of the potential risk of Salmonella transmission from reptiles, especially when they are kept in close contact with humans. Education of family members keeping reptiles in the household is mandatory to reduce this risk. Furthermore, if Salmonella gastroenteritis or invasive disease is detected, a detailed request to reveal "pet" reptiles as possible reservoir is essential.

P 35

Community-acquired enterococcal urinary tract infection

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Introduction: Community-acquired febrile urinary tract infection (UTI) is common in children, causing substantial morbidity and accounting for many hospitalizations particularly in infants and younger children. Most causal pathogens are gram-negative enterobacteria: The predominant pathogen is *E. coli,* whereas only ≤5% of UTI are caused

by Enterococci, mainly *E. faecalis.* All enterococci are resistant to cephalosporins. We report a case series of children with febrile UTI caused by *E. faecalis.*
Patients and methods: We retrospectively collected data from patients presenting with febrile UTI (temp >38.0°) between January 2013 and December 2014 caused by isolated growth («monoculture») of Enterococci. Urine was obtained either by transurethral bladder catheterization or midstream urine in continent children. The patients› demographic features, laboratory parameters (CRP, plasma creatinine), ultrasound of the kidneys and the urinary tract, and the clinical course was analysed.
Results: A total of 8 episodes of febrile UTI were observed in 7 boys (no girl). Median age was 0.75 years (range 0.1–5.1). Median fever duration at diagnosis was 2 days (0.5–4) and median CRP 118 (5–372 mg/l. Urinalysis (dipstick and sediment) showed mild leucocyturia (6–10 / high power field) in only one case. Initial empiric antibiotic therapy consisted of cephalosporin in 5 (switched to amoxicillin after urine grew *E. faecalis*), and amoxicillin in one; in 2 episodes, amoxicillin was started only when urine culture became positive.
5 patients had an underlying malformation: Ectopic kidney, duplex kidney, solitary kidney with megaureter, hydronephrosis and complex bilateral renal and ureteral malformation. The clinical course was uneventful in 6 patients. The boy with complex renal malformation sustained two episodes despite antimicrobial prophylaxis, the 2nd episode leading to transient acute renal failure.
Conclusion: Febrile UTI with enterococcal monoculture is rare in children. In addition, the often inconspicuous urinalysis, i.e. absent leucocyturia, is a diagnostic challenge leading to underdiagnosis. It is suggested that boys, in particular with underlying malformation of the kidneys and the urinary tract, are more prone to enterococcal UTI. Standard empiric antibiotic therapy with cephalosporins often fails to resolve symptoms in enterococcal UTI, and switch to amoxicillin is required. This study is also meant to raise the awareness of enterococcal UTI, in particular in infants and young children with persistent fever without other symptoms and normal urinalysis.

P 36

Swiss pediatricians' approach and management of streptococcal pharyngitis: a Swiss National Survey

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Introduction: Group A Streptococcus (GAS) pharyngitis is frequently diagnosed in children. Swiss guidelines recommend a microbiological test for GAS detection in the pharynx by a rapid antigen detection test (RADT), or a throat culture. If present, antibiotics are usually recommended. Different dosages, duration of treatment, frequency of administration and type of antibiotics have been extensively studied, with various conclusions regarding the efficacy of treatment. For this reason, management and treatment may differ between pediatricians within the country.
Methods: A three language online survey was mailed to all pediatricians registered in the SSP list (Swiss Society of Pediatrics), as well as to those working in University Hospitals in Geneva. It was previously tested by a group of 15 pediatricians twice, two weeks apart. The questionnaire includes six hypothetical cases on approach, management and treatment of GAS pharyngitis, as well as six general questions about GAS epidemiology, GAS clinical presentation and reason for antibiotic treatment.
Results: Of the 1977 eligible pediatricians, 459 (23%) responded to the survey and 202 (44%) had more than 10 years experience as a pediatrician. 61% followed the Swiss recommendations while 17% based the treatment on their experience and practice. The main reason to prescribe antibiotics was to prevent non-suppurative complications (64%), because of the presence of GAS in the pharynx (54%) and prevention of suppurative complications (52%). The clinical sign leading to the diagnosis of GAS pharyngitis were various; 110 (24%) used the Mc Isaac score and 235 (55%) based their decision on the result of RADT. In a 6 years old child with a sore throat for 24 hours and with a Mc Isaac score equal to five, 353 (77%) did a throat culture for GAS detection if the RADT was not interpretable, and 298 (65%) waited for the result to prescribe antibiotics. When the RADT was negative, the throat culture was performed by 38%, and 63% do not give antibiotics. In contrast, when RADT was positive, 2% performed a throat culture and 97% gave antibiotics immediately. Even if the dosage, duration and frequency of administration is heterogeneous in pediatricians, amoxicillin 50 mg/kg twice daily for 7–10 days and penicillin 1 Mio or 50000UI twice daily for 10 days were still the most commonly prescribed treatments. Treatment was not modified if the symptoms reappeared two months later or if parents did not strictly follow the treatment. In case of a cutaneous rash, macrolides were chosen in 45% of cases (once daily for 3 days for azithromycin and twice daily for 10 days for clarithromycin and erythromycin).
Conclusion: Treatment options of streptococcal pharyngitis are mostly based on the result of RADT. The antibiotic dosage and the frequency of administration for penicillin in particular as well as the length of treatment for amoxicillin are heterogeneous. We believe that the evidence-based Swiss recommendations should be promoted to enhance the treatment and management of GAS pharyngitis.

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Blind passengers and Immigration

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Introduction: The introduction of the Haemophilus influenza type b (Hib) and pneumococcal conjugate vaccine led to a decline of the incidence of bacterial meningitis in Children over the age of two months. The incidence of meningococcal disease is highest in the "meningitis belt" of sub-Saharan Africa. There are periodic epidemics during the dry season (December–June) there. Clustering people like on the hajj are known risk factors for invasive meningococcal disease, especially with serotype W135. Meningococcal invasive disease in Europe is caused mainly by serotype B, where a vaccine is under way and serotype C, which is covered by the standard vaccines. A combined vaccine covering serotypes A, C, W135 and Y is also readily available. Our patient entered Switzerland a month before symptoms after immigrating from Eritrea through Libya to Europe.
Case report: After a history of fever for four days and progressive swelling and redness of his right eye this 8 month old male infant was referred by his paediatrician in a decreased general state being irritable and sensitive to touch. A bacterial meningitis was suspected. He showed signs of meningitis such as a bulging fontanel, lethargy and touch-tenderness. He was febrile and in a very reduced general state. Kernig and Brudzinski were positive. He was treated with ceftriaxone and vancomycin empirically (DD resistant pneumococcus). Dexamethasone was added for two days (DD Hib-Meningitis). Lumbar punction confirmed the diagnosis. The blood culture showed N. meningitidis, later identified as serotype W 135. The CSF-culture was sterile due to prioritized empiric antibiotic treatment. The total course of parenteral antibiotics was 9 days, we streamlined to ceftriaxone after blood-culture positivity. The redness of the eye (diagnosed as accompanying uveitis: Uveo-Meningeal Syndrome) resolved after the first days. A hearing test was normal. The infant was discharged in a very good general condition without neurologic sequelae.
Conclusion: Neisseria meningitidis has become the leading cause of bacterial meningitis in children since routine vaccination of infants against Pneumococci and Hib was introduced. Serogroup W135 plays a role in endemic infection: In 2000/2001 the largest outbreak due to this strain occurred among more than 400 pilgrims returning from the Hajj in Mecca, Saudi Arabia. The mortality rate of untreated bacterial meningitis approaches 100 percent and even with optimal therapy, morbidity and mortality are alarming. Our case illustrates how not only people but also disease is migrating.

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The Slippery Slope of Staphylococcal Scalded Skin Syndrome

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Introduction: Staphylococcal Scalded Skin Syndrome (SSSS) is a dermatologic infection characterised by flaccid bullae formation and rupture. SSSS can rapidly involve an extensive skin surface leading to complications such as sepsis or electrolyte imbalance and cause up to 4% mortality. No clear guidelines are available for management of these patients. We present four cases of SSSS:
Discussion: In these cases, prodromal symptoms are unspecific and can lead to a delay in hospital referral. Only one lesion swab came back positive for staphylococcus and none of the blood samples showed the exfoliative toxin responsible for the bullae formation. Patient 1 was treated as an outpatient due to a good general appearance and little skin surface involvement but others showed rapid evolution and needed hospitalisation for IV antibiotics, analgesia, and rehydration.
Conclusion: Recognising SSSS at its early stage is a challenge for paediatricians. Diagnosis is clinical but can be confirmed, if specified correctly to the laboratory, either by isolation of exfoliative toxin, or by skin biopsy that shows midepidermal separation at the zona granulosa

	Patient 1, 2-month-old male	Patient 2, 19-month-old male	Patient 3, 13-month-old male	Patient 4, 19-month-old female
Before hospitalisation	Constipation since two weeks treated by microlax. Typical SSSS bullae on thorax.	Upper respiratory tract infection. Anal fissure since one month. Suspicion of impetigo treated by antibiotics.	Upper respiratory tract infection since three weeks. Suspicion of peri-infectious urticarial treated my feniallerg.	Conjunctivitis and cheilitis for two days. Suspicion of allergy treated by feniallerg.
Evolution in emergency room	Good appearance, no pain. New lesions on abdomen, axillar, popliteal and inguinal folds.	Very rapid evolution in first hours of care: oedema and positive Nicolsky sign. More flaccid bullae formation 24 hours post start of antibiotics.	Dehydrated child in severe pain. Only cheilitis and conjunctivitis at first then diffuse desquamation of body until 48 hours post start of antibiotics.	Poor general appearance and anorexia since two days. Bullae formation and rupture 12 h after later.
Laboratory work up	Superficial lesion swab: positive for MSSA resistant to penicillin G.	– Nasal and cutaneous swab: no MRSA – Blood culture: negative	– Blood culture negative – Stool culture negative – Nasal swab: positive for MSSA – Exfoliative toxin: negative	Conjunctival and skin swab: no MSSA Blood culture: negative ExfoliativeToxin: negative
Management	Outpatient treated by oral antibiotics and control after 36 hours	Hospitalisation for IV antibiotics and Morphine	Hospitalisation for IV antibiotics and rehydration	Hospitalisation for IV antibiotics, morphine and rehydration

(rarely done in the emergency room). Most cases present with little skin involvement and won't need hospitalisation, but close follow-up should be the rule as rapid deterioration can occur and hospitalisation and intensive treatment may be needed.

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Mycoplasma Pneumoniae Associated Mucositis a challenging case

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Introduction: Mycoplasma Pneumoniae Associated Mucositis or otherwise called 'atypical' or 'incomplete' or 'without skin lesions' Stevens Johnson Syndrome is a rare but increasingly recognized entity. We report a successfully diagnosed and treated case.
Case report: A 13-year-old, previously healthy, adolescent boy is admitted to the emergency department with severe odynophagia secondary to lip and oral cavity ulcerations. He also presented bilateral conjunctivitis, fever and cough. The clinical picture together with an associated M. pneumoniae lung infection suggested a MPAM. Antibiotics were started together with supportive therapy. Patient was discharged after 14 days, more than a month later, he completely recovered without any sequelae. Positive serology for M. Pneumoniae in our case points toward a probable significant role of the bacteria in this affection.
Discussion: An extensive literature search was performed on PubMed by using the keywords mentioned in the introduction. Few case reports of isolated mucous involvement triggered by M. pneumonia are published. It is believed that empiric antibiotic treatment could limit the disease duration and severity. Supportive care is crucial including systemic anti-inflammatory agents and parenteral nutrition. Though, the use of corticosteroids and IVIG are not yet indicated. No lethal cases have been reported until now.
Conclusion: MPAM, a condition not to be missed, could be a challenging diagnosis in the pediatric setting necessitating appropriate supportive care.

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Pneumococcal meningitis despite immunization with PCV23

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Introduction: Streptococcus pneumoniae is the most common cause of bacterial meningitis in infants and children older than one month of age. Despite prompt and appropriate care, it remains an important cause of childhood morbidity and mortality. The incidence of pneumococcal meningitis declined significantly since the pneumococcal conjugate vaccine (PCV) was added to the universal infant immunization schedule. However, some serotypes are not covered by the vaccine and prompt recognition and treatment of pneumococcal meningitis remains mandatory.

Clinical cases: We present two cases of pneumococcal meningitis in patients in our hospital who had been immunized with PCV13 in 2014.

Description	Vaccination	Pneumococcal serotype	Neurological Complications	Outcome
19 months old girl with history of 7 days of fever, then somnolence and neck pain	3 doses of PCV13: 2, 4 and 12 months	*S. pneumoniae* serotype 24F	Sagittal sinus thrombosis, Seizures, initial right hemiparesis	1 year later: favorable outcome
4 months old girl with 3 days of fever, then fatigue and irritability	2 doses of PCV13: 2 and 3 months	*S. pneumoniae* serotype 12F	Seizures, multifocal vasculitis Brain abscess, Hygroma pericerebralis	3 months later: Persistent axial hypotonia

Discussion: *S. pneumoniae* not only causes meningitis, but also acute otitis media, sinusitis, mastoiditis and other invasive pneumococcal diseases (IPD). PCV 7 was introduce in 2005 in the Swiss routine childhood immunization schedule, and was replaced by PCV13 in 2010. This vaccine contains thirteen pneumococcal serotypes (1, 3, 4, 5, 6A, 6B, 7F, 9V, 14, 18C, 19A, 19F, and 23F) and not those which caused the disease in our patients. Both serotypes 12F & 24F are rare serotypes in Switzerland. They are not particularly known for their invasiveness. Our cases illustrate the fact that despite pneumococcal vaccination, children with suspected bacterial meningitis can have IPD, and that follow-up is necessary even after the acute disease to better care for these children.
Conclusion: The impact of PCV in IPD is high but pneumococcal meningitis may occur despite immunization with PCV, due to non-vaccine serotypes. In the future, the development of a universal pneumococcal vaccine (serotype non-specific) would be useful.

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Management of an infant with haemophilia B, requiring major cardiac surgery

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Introduction: HEMOPHILIA B is an inherited X-linked disorder, characterized by a deficiency of blood coagulation factor IX. Neonates and infants present few symptoms and may not have bleeding until later in life when they begin walking or running. Many patients are

asymptomatic until the hemostatic system is stressed, for instance by surgery. We here report the case of a 1-month-old infant, who was incidentally diagnosed with moderate hemophilia B following the hemostatic screening prior to heart surgery.
Case report: A 1-month-old infant was admitted for elective heart surgery to correct aorta coarctation. He has already received vitamine K, according to the local recommendations. The pre-surgery hemostatic screening revealed an abnormally prolonged partial thromboplastin time (PTT) of 77.5 seconds (N: 26–37 seconds) with normal prothrombin time and normal platelet count. The PTT was corrected after mixing with pooled normal plasma, indicating a factor deficiency. Coagulation factors VIII and IX were then measured, and a factor IX activity at 1.3% was demonstrated, thus giving a diagnosis of moderate haemophilia B (normal factor IX activity at 1 month of life: 30–70%). In retrospect, the family history was notable for a maternal uncle and a maternal male cousin with mild bleeding, diagnosed with hemophilia B (with factor IX levels about 40% in adulthood). The surgery was postponed and the pharmacy's hospital was alerted to stock extra vials of recombinant Factor IX (Benefix®) for the necessary replacement therapy and in case of emergencies. We studied the kinetic response of the patient to a dose of 500 IU (100 IU/kg) of Factor IX, showing a half-life of about 4 hours (FIX activity: t 30 minutes 72%, t 2 hours 55%, t 24 hours 13%, t ½ 4h15min). This value is significantly lower than what described in the literature, but no studies have been done in children. The patient received 500 IU (100 IU/kg) of Factor IX 12 hours and 30 minutes before the surgery, in order to target an activity of factor IX of 60–80%. The intervention lasted one hour without recourse to extracorporeal circulation. No transfusion was needed. Factor IX was then administered according to the World Federation of Hemophilia (WFH) guidelines and FIX activity was checked regularly to insure the adequacy of factor replacement. The target was an activity of 40–60% from D1 to D3 post-operatively, of 30-50% from D4 to D6 and of 20–40% from D7 to D14 post-operatively. The patient didn't experience any bleeding complications and could be discharged home 14 days after surgery.
Conclusion: Our 1-month-old hemophilic patient underwent a major cardiac surgery with no bleeding complications. We strongly recommend doctors to plan elective surgeries from all viewpoints (including medication availability), evaluate the half–life of the factor in vivo in order to allow personalized replacement schedule, and follow the WFH guidelines for the management of all hemophilic patients.

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An unpleasant souvenir from Ecuador

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Introduction: In children, infections with *Dermatobia hominis,* also known as the botfly, are rarely seen in Europe. In returning travelers from endemic areas, the differential diagnosis of myiasis has to be considered.
Case report: A 5 8/12 year old boy presented after a family vacation in Ecuador with a painful swelling located on the scalp with concomitant lymphadenopathy. Because of suspected bacterial furunculosis, oral antibiotic treatment with amoxicillin/clavulanate was initiated. After two days of treatment, the swelling was progressive and Staphylococcus aureus was isolated from a culture of a swab specimen taken from purulent exsudate draining from a pore. The dose of amoxicillin/clavulanate was increased and eleven days later, four days after completion of the antibiotic regimen, the boy returned to the emergency room because of persistent swelling. The diagnosis of subcutaneous myiasis was made by ultrasonographic examination. After incision the larva of the botfly *Dermatobia hominis* was extracted. A follow up two weeks after the surgical intervention revealed an unobtrusive local wound healing.
Conclusion: In Switzerland, myiasis in children is rarely seen, and often misdiagnosed. Symptoms are often misleading to the diagnosis of furunculosis. With a history of travelling to endemic areas, myiasis should always be considered as differential diagnosis of purulent skin lesions. A careful clinical examination and an ultrasound scan are fast, easy and noninvasive diagnostic instruments. The treatment of choice is to eliminate the whole larva. To minimize the risk of damaging the larva surgical removal should be considered. Other methods, such as occlusive oinment therapy in combination with forceps extraction bare a certain risk of incomplete removal of the larva and consecutive inflammation. In case of bacterial superinfection, surgical removal is the therapy of choice to assure appropriate cleaning of the wound cavity. Although the larva produces bacteriostatic substances in its gut, bacterial superinfection is the most important complication. Other rare complications include cerebral ocular and oral myiasis.

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Early and complicated relapse of varicella-zoster virus infection in immunocompetent child: a case report of Ramsay-Hunt syndrome

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Background: Ramsay Hunt syndrome (RHS) is a rare complication of varicella zoster virus infection (VZV) in which reactivation of latent varicella zoster virus infection occurs in the geniculate ganglion, causing otalgia, auricular vesicles and peripheral facial paralysis. This syndrome usually affects adults but rarely children. However, RHS should be considered in the differential diagnosis of peripheral facial paralysis in children.
Case report: we report a child of a 10-year-old boy affected by Klinefelter syndrome admitted for a VZV infection. The patient presented with fever and an extensive painful vesicular rash with skin bacterial superinfection (S. aureus) in the left C2 and C3 dermatomes. Treatment with Co-Amoxicillin and Valacyclovir during ten and seven days respectively allowed a complete recovery and it has been stopped at this time after detection of abdominal pain and elevated transaminases. Two weeks later he was readmitted severe with a 4-day history of increasing left-sided headache, otalgia, auricular vesicles, left sided hemifacial paralysis, mandibular nerve hypoesthesia and nocturnal visual hallucinations. On examination temperature was 37.1 °C, vital parameters were within normal range. Cranial nerves were intact except the V (mandibular branch) and the VII (peripheral facial paralysis scored grade III on House Brackmann scale), pupils were equal and reactive to light. Strength, sensation, cerebellar testing and gait were normal. Ear, nose and throat examination revealed a reddened, swollen left external auditory canal with a normal tympanic membrane and vesicular lesions of the concha, pinna and mandibular area. The rest of physical examination was unremarkable. Complete blood count was normal, a positive IgG and IgM serology for VZV suggested a recent infection. Instrumental acumetry showed a partial unilateral perception hearing loss, with normal auditory evoked potentials. A brain MRI and a lumbar puncture ruled out a possible central nervous system involvement. Based on the above clinical and laboratory findings, the diagnosis of RHS was established. A treatment with high-dose corticosteroid (Prednisone) and antiviral therapy (Acyclovir) was administered with a completely recovery within two weeks.
Conclusions: RHS is one of the most frequent causes of facial of peripheral facial paralysis in children and should be considered early in case of VZV reactivation. We report here an atypical case of RHS showing a complicated relapse despite a previous anti-viral treatment. Association of acyclovir and high-dose corticosteroids improved symptoms in our patient even though treatment remains controversial. Large cohort studies are required to assess the best RHS treatment in a pediatric population.

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Pott's Puffy Tumor as a Complication of Frontal Sinusitis

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Background: The rate for complications in hospitalized patients with a bacterial sinusitis is about 5%. Among these are preseptal cellulitis, orbital phlegmonia, septical venous sinus thrombosis, meningitis, osteomyelitis, epidural empyema and intracerebral abscess. We present the case of a patient with Pott's Puffy Tumor, a characteristic swelling of the forehead.
Case: A girl of 13 years presented with nocturnal headache for several days and sensitivity to noise and light. She was in a good general condition and had a normal body temperature. The only clinical finding was a moderately painful swelling of the forehead. An X-ray of the skull showed a fluid level in the frontal sinus. This prompted us to perform CT- and MRI-scans which showed an epidural empyema, partial thrombosis of the sagittal sinus, and osteomyelitis of the frontal bone. The lumbar puncture showed pleocytosis of the CSF. The patient received treatment with i.v. antibiotics for two weeks and s.c. low molecular weight heparin. The patient underwent bifrontal craniotomy and drainage of the epidural empyema and obliteration of the fistula between the frontal sinus and the epidural space. The girl left the hospital after 2 weeks in a good condition and has stayed free of sequalae.
Conclusion: Rarely, a bacterial sinusitis can lead to serious complications. Pott's Puffy Tumor is a frontal phlegmonia associated

with frontal sinusitis. In most cases it is associated with intracranial complications which need to be diagnosed quickly and treated thoroughly. Thus, Pott's Puffy Tumor must prompt the clinician to perform urgent imaging by CT- and MRI-scans and consider a lumbar puncture.

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Thrombocytopenia as a clue to congenital CMV infection

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Case report: A premature 35 W. RCIU for height and weight, born in the context of maternal preeclampsia, presented with a global moderate hypotonia, persistent feeding difficulties, and at day, with thrombocytopenia, between 41 and 87 G/l. Spontaneous correction occurred at day 11. Urine PCR was positive for CMV and serologies were positive (IgG 105 kUI/l (N <14), IgM borderline). GGT was at 195 U/l (N <60). CMV PCR was negative in liquor. Maternal platelets were normal and the mother was immune for CMV (IgG positive, no IgM). Because thrombocytopenia remained stable, allo-immunisation was not specifically investigated, nor was there evidence for other congenital infections. A cerebral ultrasound at day 3 showed a ventricular asymmetry and no calcifications. CT Cerebral scan at day 11 was normal. Further investigations showed no chorioretinitis and normal auditory brainstem responses (ABR). Brain MRI at 1½ months however was compatible with a postinfectious course (calcifications not confirmed), so that antiviral treatment with valganciclovir (16 mg/kg/jour) was initiated at 6 weeks of age for 6 months. Psychomotor development was thereafter normal, as well as sequential ophthalmologic examination and auditory evoked potential.
Discussion: Congenital Cytomegalovirus (CMV) infection has an incidence of 1%. 10% are symptomatic at birth and can affect multiple organs. It is the main cause of sensorineural hearing loss. Symptomatic newborns can have an insidious initial clinical course. More severe infection can cause thrombocytopenia and a sepsis-like syndrome. Isolated thrombocytopenia should raise suspicion of congenital infections or materno-foetal immunization. In absence of immune thrombocytopenia a CMV infection should be researched, independently of maternal CMV immune status. Reactivation of maternal CMV infection is far less frequent than primoinfection, and sequelae in the offspring are generally less severe. Treatment is controversial. It was initiated in this case because of signs of fetal infection with hypotonia, in order to avoid later hearing and psychomotor development impairments.
Conclusion: Congenital CMV infection should be considered in the case of an unexplained persistent thrombocytopenia at birth.

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Leg ulcers in adolescence: infectious, vascular or inflammatory disease?

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A 14 y.o. boy presented with painful extensive, haemorrhagic, necrotic and deep ulcers on both legs, with pink borders. It appeared after prurigo-like lesions provoked by insect bites during sea side vacations. There was no fever and moderate elevation of acute phase reactants. He first received co-amoxicillin, without success. No micro-organism was found, including fungi, mycobacteria and Leischmania. Pathergy was present. No other organ was concerned: bowel disease, lymphoma and blood malignancy were ruled out. Biopsy showed neutrophilic necrosis, with few non caseous, eosinophilic granuloma and vasculitis, without argument for bullous disease. Diagnostic of Pyoderma gangrenosum (PG) was retained. Oral prednisone was begun, followed by slow healing during the six following months with tapering doses. No other side effect than cushingoid face and no adrenal insufficiency were observed. Follow-up was maintained.
Discussion: PG is an auto inflammatory disease of skin, rare in children and adolescents. The elementary lesions begin with nodules or pustules, secondary eroding in painful necrotic ulcer, with haemorrhagic and/or purulent exudates, surrounded by violaceous borders and erythema. Differential diagnoses are numerous: vascular, infectious, granulomatous or vasculitis-related diseases and recurrent familial arthritis (PAPA: Pyogenic sterile Arthritis, PG, and Acne). Factitial dermatitis should be kept in mind. Lacking pathognomonic marker, PG is a diagnostic of exclusion, after work-up of systemic underlying disease such as inflammatory bowel disease and malignancies. Rapid progression of lesion, pathergy (including post-traumatic exacerbation), pathologic findings, and insensitivity to antibiotic versus responsiveness to immunomodulator therapy are his hallmarks. Debridement should be avoided. Local care must be associated with systemic treatment. Healing takes usually several months, with steroid as first line treatment. Second step therapy with immunomodulator is often necessary: colchicine, dapsone, cyclosporine or methotrexate. Anti-Tumour Necrosis Factor can be considered. Side effects of therapies imply caution.
Conclusion: When deep cutaneous ulcers persist, without infectious, inflammatory, tumoral or paraneoplastic explanation, think about primitive Pyoderma gangrenosum.

Reference: Ahronowitz I. Etiology and Management of Pyoderma Gangrenosum. Am J Clin Dermatol. 2012;13(3):191–211.

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Cervical lymphadenitis – always a trivial matter?

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Introduction: Cervical lymphadenitis is a very common pediatric disease that often leads to medical consultation. A thorough medical history and physical examination is important to evaluate its etiology, which is mostly associated with infectious agents (viral, *streptococcus pyogenes, staphylococcus aureus,* anaerobic bacteria, atypical mycobacteria, *tularemia, bartonella henselae*). If required, an empiric antibiotic treatment can be initiated and is mostly successful. If suppuration occurs, a surgical intervention may be needed. Further investigations are reserved for some rare cases.
Case: A 16-month-old girl, born in Switzerland, was admitted in a regional hospital with a history of persistent bilateral suppurative cervical and facial lymphadenitis. No systemic symptoms were present. A parenteral antibiotic treatment with amoxicillin/clavulanate was started but did not show any clinical improvement within the first 72 hours. A surgical incision and drainage was performed with detection of *Burkholderia cepacia complex* in microbiological cultures. According to the bacterial resistance pattern, the treatment was switched to trimethoprim/sulfamethoxazole. But lymph node enlargement and suppuration progressed, leading to a continuous fistulation. She was then referred to our pediatric infectious disease department for further evaluation. There was no known preexisting condition nor any familial inherited immunodeficiencies. Due to the unusual clinical presentation and bacterial findings, we decided to perform an immunological exam. The only pathological finding was a nitrobluetetrazolium (NBT) test showing no functional granulocytes. After repeating the NBT and conducting an additional dihydrorhodamin (DHR) test, the diagnosis of a chronic granulomatous disease (CGD) was confirmed. We started a bactericidal therapy with meropenem as well as an antifungal prophylaxis with remarkable improvement, unfortunately the largest lymph node had to be removed surgically. Histologic findings were suitable for a CGD without any signs of malignancy. Other organ involvement was excluded. After 14 days of intravenous antibiotic treatment, we were able to discharge the patient, following an oral antibiotic prophylaxis.
Background: In patients with CGD granulocytes are not able to form superoxide radicals, so phagocytosis is ineffective and can lead to granulomas in any organ system. This makes the patients susceptible for bacterial and fungal infections. The prevalence is about 1/250'000, most often there is an x-linked inheritance, so girls are rearly affected, but other genetic pathways (i.e. autosomal recessive) are known.
Conclusion: Even though cervical lymphadenitis is common and can usually be treated by the GP, differential diagnosis must be considered if the history, clinical presentation or microbiological/histologic findings are not suitable for common causes of lymphadenitis. In general the prognosis of CGD is good if optimal management with close follow-up and prophylactic treatment is achieved, leading up to the only curative treatment, namely a hematopoietic stem cell transplantation from a matched donor.

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Refusal to walk is not to banalize

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Introduction: Acute non traumatic limp or refusal to walk is a common cause for emergency department visits and can be challenging. The differential diagnosis is broad, going from traumatic injuries to infectious, inflammatory or neoplastic causes. Its localization also can be hard to define, especially in young children. We describe here the case of a spondylodiscitis in a young child with initial unclear

presentation and whose diagnosis was made after repeated examinations.
Case report: A 2-year-old girl with acute, intermittent refusal to walk. Previous trauma is unclear. Frequent night awakenings. She is initially subfebrile (disease day 2/3). Usual painkillers allow resolution of symptoms and normal gait with no limp. Repeated physical examinations are entirely normal until the apparition of sacrum tenderness on day 7. Blood tests show unspecific inflammatory findings with elevated CRP, leucocytosis and elevated ESR. Diagnosis of L4-L5 spondylodisicits/osteomyelitis with protrusion in the spinal canal at L4/L5 and L5/S1 and suspicion of an epidural abscess is made on day 8 with nuclear magnetic resonance imaging. Routine bacterial blood cultures remain negative. *Kingella kingae* is identified by PCR on a nasopharyngeal swab. No biopsy is performed. 2 weeks i.v. cefuroxime followed by 4 weeks oral co-amoxicilline proved successful.
Conclusion: A child refusing to walk or with a limp is never to banalize and needs repeated evaluations until clear clinical signs appear or complete resolution of symptoms. Spondylodiscitis (i.e. osteomyelitis, discitis and spondylitis) in young children is a challenging diagnosis. Its exact incidence is not known but is thought to represent 2–4% of pediatric bone infections. Recent studies suggest an increasing role of organisms carried asymptomatically in the upper respiratory tract, such as *K. kingae.* Common symptoms include refusal to walk or sit, diffuse abdominal pain or frequent night-awakening. However, clinical presentation is often nonspecific and can lead to diagnosis delay of several weeks. Therefore, a high index of suspicion is needed to allow early diagnosis and timely treatment. In our case, close follow-up and repeated clinical evaluation allowed reasonable time-to-diagnosis and treatment when referring to the litterature.

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Parvovirus B19: unusual presentation in a previously healthy 15-year old female

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Background: Parvovirus B19 (= PVB19) can result in a wide range of clinical manifestations, normally influenced by patient's immunological and hematological status. The major clinical manifestations that can occur with B19 infection include: erythema infectiosum, arthritis or arthralgia, transient aplastic crisis, fetal hydrous and other rare manifestations. PVB19 has a particular tropism for erytroid progenitor cells: the only known host cell of PVB19 is the human erythroid progenitor. In patients with increases destruction of red cells and acativated erytropoiesis (e.g.: sickle cell disease, hereditary spherocytosis), PVB19 acute infection can cause aplastic crisis. Neutropenia, lymphopenia ot thrombocytopenia have also been reported in acute parvovirus B19 infection; pathogenetic mechanism of these rare form of cytopenia PVB19-related are not yet known.
Case report: A15-year-old female patient presented with a prolonged and intermittent febrile illness (5-days history of high fever), general malaise, sore throat, mild gastrointestinal disturbance with a single case of vomit, general malaise and two episodes of fainting. Her past medical history was unremarkable and she denied drug consumption. Phisical examination was almost normal: we observed at least a modest cervical lymphoadenopaty and splenomegaly (confirmed subsequently by ultrasound). Nevertheless she was found to have leucopenia, lymphopenia and trombocytopenia, but a normal red cell and hemoglobin count. A full blood count showed a hemoglobin of 135 g/L, white cell and platelet counts were of 3.4 x 10^9/L and 86 x 10^9/L respectively; lymphocytes count was of 1.31 x 10^2/L with a 9.3% of large unstained cells. The morphological test, performed to exclude leukemia, revealed the only presence of reactive lymphocytes and the negativity for blasts. The C-reactive protein was 9 mg/L. Liver function tests were normal, as chest radiography and urinalysis. Antibody tests for Epstein-Barr virus and cytomegalovirus were negative. IgM and IgG for Parvovirus B19 by immunofluorescence (IF) were weakly positive. Parvovirus B19 positivity was also confirmed by PCR in a blood specimen.
Discussion and conclusions: Parvovirus B19 infection can occur with a very wide spectrum of symptoms and should be considered in the differential diagnosis of leucopenia, lymphopenia and trombocytopenia in children with aspecific symptoms (like fever or general malaise), further in patients without anemia or erythema infectiosum.

P 50

Viral meningitis after chickenpox in a 10 year old girl

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Case report: Sophia, a 10 years old girl, presented in our emergency Unit with one week history of chicken pox, as well as her sister (aged 11). Two days prior to admission she developed fever, irritability, headache, stiff neck, photophobia, nausea, lack of appetite and asthenia. On physical examination she had temperature of 37.8 °C, heart rate120 per minute, oxygen saturation 100%, blood pressure 110/64 mm Hg. She had small crust lesions all over her skin without vesicular eruption, no conjunctival hemorrhage; GCS 15/15. Cardiovascular, pulmonary and abdominal examinations were unremarkable, but we observed nuchal rigidity, walking difficulties, positive Kernig's sign and Brudzinski's sign. Considering the physical examination and the clinical history evocative for meningeal irritation, we hospedalized Sophia in order to treat her symptoms. The blood count showed hemoglobin of 128 g/L, white blood cells of 6.6 x 10^9/L, neutrophils 5.7 x 10^9/L, lymphocytes of 0.86 x 10^9/L, normal platelets and PCR of 10 mg/L. Three days after the admission, the concurrence of worsening nuchal rigidity, photophobia and fever (until 39°C) convinced us to do more exams to exclude septic complications. Lumbar puncture showed clear and colorless CSF, leukocytes 99 cell/microL, neutrophils 5%, reactive lymphocytes 9%, and CSF culture was sterile. The neuroimaging (CT scan of the head) was negative too. In the light of clinical exams negatives for a bacterial superinfection, we continued a symptomatic therapy. Only one week after the admission, we observed a real good improvement of Sophia's symptoms, so that we could discharged our patient in good general conditions.
Discussion: chickenpox is one of the most common infectious diseases of childhood, caused by the varicella zoster virus (VZV). It is usually a self-limited disease that lasts 4–5 days. However there is a risk of serious –rare- complications, not only in adults but also in children: skin bacterial superinfections, purpura fulminans, neurological presentations and pneumonias. In our case, the patient developed a meningitis following chicken pox infection. The cerebrospinal fluid (CSF) profile in VZV viral (aseptic) meningitis is indistinguishable from other causes of viral meningitis: clear and colorless, lymphocytic pleocytosis with normal protein, glucose, and lactic acid levels. The clue to most of the diagnoses of VZV aseptic meningitis, indeed, is based on the temporal relationship between antecedent or concomitant chickenpox, as happened in our patient. In most cases, there is no specific treatment for viral meningitis but some patients (in particular people with weakened immune systems caused by diseases, medications and recent organ or bone marrow transplantations) may benefit from treatment with an antiviral medication.

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Ulceroglandular tularaemia following contact with a boar: a case report

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Introduction: Tularaemia is a highly infectious zoonosis caused by the bacterium *Francisella tularensis,* mostly transmitted by small mammals like hare and rodents. An increasing number of tularaemia has been reported in Switzerland since 2007. We report on a 13-year-old boy presenting with a skin ulcer and enlarged axillar lymph nodes 6 days after contact with a boar and an ulcero-glandular tularaemia was diagnosed.
Case report: A 13-year-old Swiss boy was admitted to the paediatric ward with a digital skin ulcer and enlarged and painful satellite axillary lymph nodes. History revealed hunting and butchering a boar, followed by the occurrence of a papular skin lesion and tender axillary lymph nodes accompanied by night shivers 6 days later. He denied fever and was in good overall health. No other animal contacts were reported. He was treated by his general practitioner with oral co-amoxicillin (50 mg/kg/day) for 7 days, followed by another 2 days with increased dosage (80 mg/kg/day) as well as a topical antibiotic without any clinical improvement. C-reactive protein was 73 mg/dl on day 3 and 9 mg/dl on day 9 after onset of disease. The digital skin lesion turned to a pustule, then excreted a purulent or caseous discharge on day 9, and finally turned into an ulcer. On day 11, a standard culture of the ulcer didn't show any bacterial growth. At admission on day 13, the ultrasonography highlighted multiple deep and superficial enlarged lymph nodes with beginning abscess. A treatment of oral ciprofloxacin 1g/d was initiated and the suspected diagnosis of tularaemia was

proven by real-time PCR performed on a skin ulcer biopsy and on a swab (50'000 copies/ml Francisella tularensis). Francisella tularensis IgM and IgG serum antibodies were positive whereas those against Bartonella henselae, EBV, CMV, Toxoplasmosis and Mycobacteria, as well as blood cultures were negative. Treatment with Ciprofloxacin was continued for 14 days with a good clinical response.
Discussion: Francisella tularensis is a facultative intracellular gram-negative bacterium, it is considered to be a highly virulent organism. The infection is acquired by contact of healthy skin with infected animals, usually small rodents. It can also be transmitted by insect bites, but human-to-human transmission is rare. Symptoms include fever with chills, headache and malaise. The ulceroglandular form is the most common and bacteremia is a rare but potentially fatal condition. The incubation period vary from 1 to 14 days and symptoms usually develop within 3–5 days of infection; the disease can be deadly without treatment. The diagnosis is confirmed by microbiology, by PCR or increase in specific antibody titers. First line treatment options include gentamicin and quinolones.
Conclusion: To our knowledge, this is the first case of tularaemia after contact with a boar. Tularaemia is a rare condition that should be considered in any child presenting with skin ulcer and enlarged lymph nodes after contact with wild animals or rodents, or after contact with their dejection and faeces. In ulcero-glandular tularaemia a treatment of ciprofloxacin should be initiated as quickly as possible.

P 52

Not MOTT (mycobacteria other than tuberculosis) in the neck as expected, but active tuberculosis

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Introduction: Infections with atypical mycobacteria in pediatric patients generally present in children <5 years with cervicofacial lymphadenitis. Lymphadenopathy, however, can also be a manifestation of systemic tuberculosis. Isolated cervicofacial tuberculous lymphadenopathy is rare in children.
Case report: A two year old previously healthy boy had suffered from unilateral left cervical lymphadenopathy since two weeks. Clinical examination revealed a firm, painless mass with a purple color which grew quickly and became fluctuant over the next 10 days. Sonography of the lesion was indicative of an inflammatory process. Mantoux screening test was positive (induration: 15 m). All findings were suggestive of MOTT (mycobacteria other than tuberculosis). Because of the progressive enlargement and mainly for aesthetic reasons, the lymph node was excised. Surprisingly, tissue analysis with STA (strand displacement amplification) revealed mycobacterium tuberculosis instead of MOTT, consistent with isolated cervicofacial tuberculosis. Further examinations showed normal chest X-ray and negative culturing of gastric fluid. The boy did not suffer from cough, fever, weight loss or reduced appetite. Inflammatory markers (full blood count, CRP) were normal. As isolated lymph node tuberculosis is rare, the otorhinolaryngologists actively ruled out tuberculosis of the larynx or the tonsils. Triple therapy with isoniazid (3 mg/kg daily), rifampicin (8 mg/kg daily) and pyrazinamide (20 mg/kg daily) was administered for two months, followed by dual therapy with isoniazid (13 mg/kg daily) and rifampicin (10 mg/kg daily) for another four months. The cervicofacial lesion healed without any complications, and the boy was always in a good general condition. The source of infection, i.e. a contagious index patient could not be identified.
Conclusion: MOTT is a well-known cause of unilateral cervical lymphadenitis in younger children. Lymph node tuberculosis can be both a manifestation of a systemic disease and less common of an isolated lesion in the neck. Isolated lymph node tuberculosis can mimic MOTT, especially if the lymph node lesion is not improving on conservative or surgical therapy.

P 53

Decision-making in extremely preterm infants: a survey about the attitudes and values within a multi-cultural society (SNF project NFP67)

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Introduction: The continuous and rapid progresses in neonatal medicine over the last decades have led to an increase in survival chances for extremely low gestational age infants (ELGA). However, long-term morbidity has not decreased accordingly. Furthermore, the ambiguity of prognostication can confuse parents' perception of their child's quality of life. Several studies have suggested that physicians› perceptions of quality of life seem to be at odds with those held by society, and thus possibly parents. The goal of this representative survey was to explore the attitudes and values of the Swiss population and to offer important insights on assessments of medical decision-making and quality of life for ELGA infants.
Methods: An anonymous nationwide telephone survey was conducted to explore the attitudes and values towards extreme prematurity within the Swiss population. A random sample was drawn from the official Swiss telephone registry. Quotas were allocated for regions, age, and gender. Computer-assisted telephone interviews (CATI) were conducted with 1210 Swiss residents aged 18 to 84 years (response rate 23.7%). Statistical analysis was performed using IBM SPSS Statistics.
Results: Two main results emerged from the questionnaire. First, respondents stated that an acceptable quality of life of ELGA infants should imply: the conduct of an independent lifestyle (32%), being normal/like others (18%), a life without medical assistance (10%), or a life without a handicap (8%). Second, in terms of intensive care treatment a majority (78%) of the population preferred shared decision-making between parents and professionals. In case of dissent, 65% felt that parents should have the final word in decision-making.
Conclusion: Important insights of this study were that a majority of the population conceptualized (1) quality of life in terms of independence or normalcy and (2) that medical decision-making for ELGA infants must be based on a shared decision-making model. The shared decision making process should not be exclusively based on what could be done medically but also on a profound deliberation of the values, wishes and preferences of parents, and on the child's best interest. This study was the first step toward a better understanding how the different criteria's and values, such as quality of life (e.g. independence, normalcy) influence specific shared decision-making processes in daily practice.

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«Santé des nourrissons – feuillets d'accompagnement»: an evidence-based interdisciplinary tool positively evaluated by parents and professionals in a cross-sectional study

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Introduction: Studies show that parents of newborns need simple, reliable and accessible information, and that coordination and partnership between professionals is essential to empower them and to reduce their anxiety. Professionals of canton de Vaud (early childhood nurses, midwifes, pediatricians) developed together ten evidence-based leaflets on topics often discussed with parents: «Santé des nourrissons – feuillets d'accompagnement» (www.avasad.ch/santedesnourrissons).
Methods: After 6 months of distribution of 70'000 leaflets, we conducted a cross-sectional survey with 789 questionnaires to parents and 335 to professionals (early childhood nurses, midwifes, pediatricians) in public and in private settings. Group interviews with 41 mothers completed the information. Likert scale from 1 – strongly disagree to 5 – strongly agree was used to assess usefulness of messages, clarity, sense of knowledge improvement, and a self-efficacy indicator was created based on «Guide for constructing self-efficacy scales» (Bandura 2006).
Results: Participation rate was 26% for both parents and professionals. Only 43% of responding parents knew the leaflets, but they found them useful (median: 4.72) with clear messages (4.64) that improve their knowledge (4.35) and they felt confident to do simple care to their child (self-efficacy: 4.75). 89% of responding professionals knew the leaflets, they found them useful (median: 4.62) with clear messages (4.73) that improve parents knowledge (4.66), and that they were useful to harmonise messages amongst professionals (4.65). Several improvements were also suggested.
Conclusion: Developing in an interdisciplinary way a common evidence-based tool for healthcare professionals working with parents of newborns proved successul.

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Swiss-DRG provokes Insomnia to the Heads of Neonatal Departments

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Introduction: Swiss-DRG is a so called "self-learning system" and has been developping continuously since 2012. However, for sick newborn infants the system did not learn to finance their medical needs, instead, it did worse. Did the neonatologists miss the start of Swiss-DRG whilst snoozing or were they too lazy to teach the system?
Method: (1) The course of MDC 15 from catalogue version (V) 1.0 to 4.0 (2) Calculation of the mean cost recovery weighted on patient numbers from 13 hospitals, which sent data of all MDC15-patients of 2013 in V2.0 using the hospitals own cost recovery (own baserate). (3) Simulation with Swiss-DRG Catalogue 4.0: Four hospitals (CHUV, Kinderspital Zürich, OKS, UKBB) sent simulated data for 2015 for evaluation. (4) Counting of the applications to Swiss-DRG AG and BfS 2013 and 2014.
Results: Ad (1) The cost weight in MDC 15 decreases from 2012 to 2015. Ad (2) The Swiss-DRG Version 2.0 already showed massive deficits in cost compensation for neonatology units. 10/13 Hospitals were level III centers. 7/13 hospitals provided neonatal data without the maternity, all of them except one showed a negative cost coverage (deficit) for 2013. 6/13 hospitals provided data including their maternity units: 4/6 showed positive results, only 2/6 showed a deficit. 13/13 hospitals showed the largest deficit in ill newborns >1999 g and <1000 g birth weight. Ad (3) The simulation with catalogue 4.0/2015 using data of the four level III centers show an increasing deficit for the MDC 15 of 11.8 Mio CHF. Ad (4) In 2013 and 2014 thirty applications concerning neonatology were handed in to the institutions Swiss-DRG AG and BfS in order to improve the situation. But the results are sobering.
Conclusion: The Swiss DRG System and its mechanisms again show to be appropriate for acute adult medicine, but not for neonates. Sick newborns are an extreme particular group even in pediatrics. Our data show that DRG mechanisms applied in Switzerland do not work for this group of patients. This is not surprising since other countries, like California with larger numbers of newborns have not solved the problem over the past 20 years using similar compensation mechanisms. We urgently need adaptations and appropriate calculation mechanisms for children – in particular neonates, since negative revenues will harm the patients' treatment and safety on the long run. The actual unfit and unfair system is a nightmare for the heads of neonatal departments. Despite 30 applications there has been not enough response from the official institutions to date. Hopefully, the heads will find a deep releasing sleep with a long REM-sleep-dream where the epiphany of DRG-solution appears. But unfortunately it is probable that they won't be able to solve the problems without an intense political will and the aid of the official organs.

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Automated identification of activity state in very preterm infants – a way of cleaning diaphragmatic EMG data?

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Background: Very preterm/ very low birth weight infants are known to be a high risk population, especially during transition after birth. Diaphragmatic EMG measurements in those infants contribute to a better understanding of immature regulation processes such as control of breathing and heart rate. Those measurements are technically very demanding in preterm infants given their small body surface which leads to close neighborhood of organs, occurrence of movement artefacts, and the fragility of this population *per se*. Determining acceptable EMG data therefore is essential but demanding and time consuming.
Aims: We aimed to test the feasibility of surface diaphragmatic EMG measurement in very preterm/ very low birth weight infants under clinically relevant conditions during the first week of life. We intended to apply a video-based algorithm to separate active from non-active periods to avoid misleading interpretation of the EMG signal due to noise.
Methods: This is a prospective, single center, observational study in the neonatal intensive care unit at UKBB. We performed surface diaphragmatic EMG measurements (Polybench; Inbiolab, NL) and synchronized video recordings using Microsoft webcam (1080 HD Sensor; 15fps, 424x240 pixel). Daily measurements lasted for three hours each during the first five days of life. Video recordings were used to detect motion artefacts and disturbing influences due to parental visit or medical/nursing interventions. In a subset of 20 videos, three trained members of the research team visually analyzed the whole 3-hour video recordings. They scored active and non-active states of the infants as well as interventions in blocks of 10 seconds.
Results: Final comparison of inter-rater reliability and agreement between observers and the software is still pending but the preliminary results are promising. Conclusive results will be presented at the congress.
Conclusion: Elimination of noisy parts in EMG measurements is an important step in quality control. This is especially important in preterm infants as measurements have to be conducted in the clinical setting of an intensive care unit where motion and handling can't be avoided. Using automated analysis of synchronized video data that enables differentiation of noisy from clean data would be very time-saving. The motion–detection software will be verified with visually obtained information on activity states in 20 measurements.

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Acute femoral artery thrombosis in a very low birth weight infant

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Introduction: Sick neonates are at high risk for thromboembolic complications (TC). The most important risk factors are indwelling intravascular catheters. Incidence of TC in children is increasing but evidence supporting treatment recommendations remains weak.
Case Presentation: This male infant (twin A, spontaneous, monocygotic) was born by emergency caesarean section at $26^{3}/_{7}$ weeks of gestation, birth weight was 950 g (P50–75). The baby's postpartum adaption was delayed (Apgar 1,3 and 4 at 1, 5 and 10 minutes respectively, umbilical arterial cord pH could not be evaluated). He was on mechanical ventilation until the 4th day of life and needed circulatory support with Dopamine on his first day of life. Umbilical catheters (UVC and UAV) were installed. The second day of life, cerebral ultrasound revealed bilateral intraventricular haemorrhage (IVH) Stage III(*Papile*) without ventricular dilatation. On the fifth day of life the UAC was removed. A few minutes later, he showed clinical signs of acute arterial occlusion in the left leg (not palpable pulses, livid, mottled and cool skin, and necrotic tip of the 4th left toe). Doppler sonography confirmed occlusion of the left proximal A. femoralis communis. An intravenous treatment with unfractionated heparin (UFH) was started and antithrombin administered several times to achieve a target anti-factor Xa activity of 0.35–0.7 IU/ml, leading to immediate clinical improvement. Doppler sonography after 7 days showed well developed collateral perfusion and blood flow in the left A.femoralis communis and anticoagulation was discontinued. The tip of the 4th left toe remained necrotic and fell off after a few days without further complications.
Conclusion: Preterm babies and neonates are at increased risk of TC, associated with short- and long-term morbidities. Due to the unique haemostatic properties, the increased bleeding risk on anticoagulation therapy and the lack of evidenced based treatment guidelines in this very young population an accurate, interdisciplinary evaluation of treatment modalities is recommended. In particular, a paediatric haematologist should be involved.

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A hungry Newborn: A Case of Neonatal Hypoglycaemic Encephalopathy

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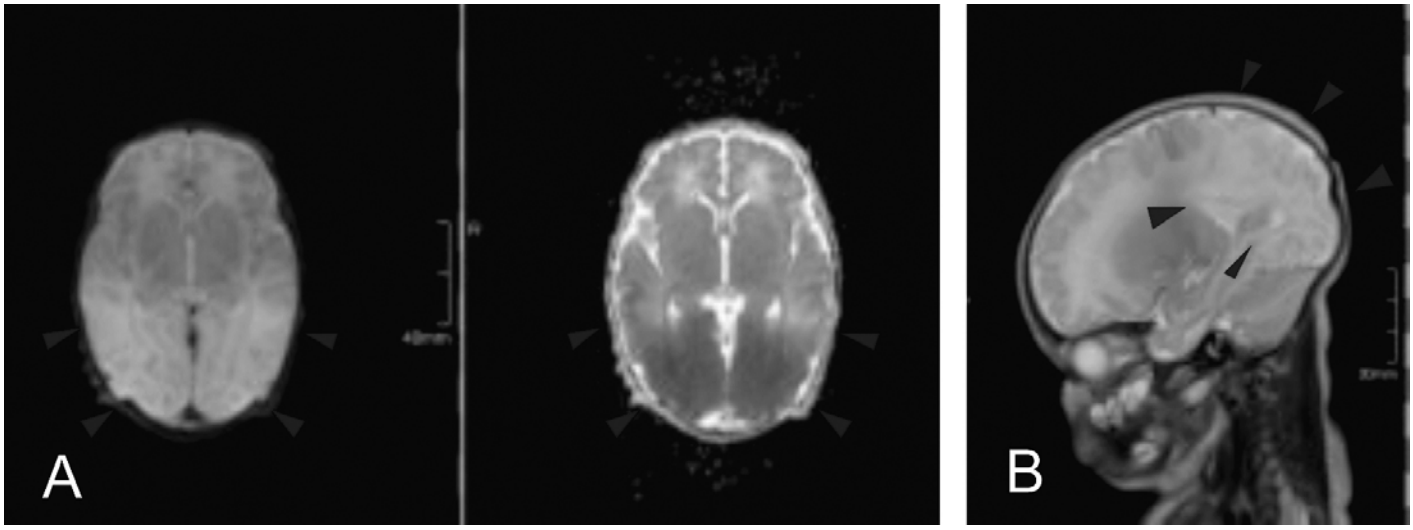
Introduction: Hypoglycaemia is common during metabolic transition to extra-uterine life. Nevertheless in a minority of cases, neonatal hypoglyceamia can lead to severe encephalopathy. According to the guidelines of Swiss Society of Neonatology (SSN), newborns with a birth weight <2500 g or <3. Percentile are at high risk for hypoglycaemia. In our case, the newborn had a birthweight <3.P but because he weighted >2500 gr (2620 gr) he was erroneously defined as "low risk" patient.
Case report: The male patient was born at term to a nondiabetic secundigravida. The mother noticed her child was feeding poor from the first day of life despite extreme hunger. Although small for gestational age (SGA) the infant did not receive complementary

feeding nor was the blood glucose monitored. At three days of age he started having seizures with minimum blood glucose of 1 mmol/L, not responding to enteral feeding. After four hours of persistent hypoglycaemia, our PICU was involved and following immediate intravenous glucose infusion, blood glucose normalized quickly. The seizures continued for eight hours upon admission despite antiepileptic medication. A brain-MRI at four days of age disclosed a cytotoxic edema (image A) and parenchymal loss (image B) in the parietal and occipital lobes, consistent with neonatal hypoglycemic encephalopathy with cortical necrosis.
Results: Extended investigations revealed no evidence of endocrinopathy or inborn error of metabolism. The 3-month neurologic follow up revealed a developmental coordination disorder and a beginning microcephaly.
Conclusions: The 2007 guidelines of SSN[1] should be revised, underlining the importance of identifying SGA (birth weight <3.P or weight-for-length <3.P) in order to ensure appropriate management of high risk patients and prevention of neurodevelopmental impairment due to hypoglycaemic encephalopathy.

[1] Betreuung von Neugeborenen ≥34 0/7 mit Hypoglykämie, SSN, TM Berger et al., Paediatrica 15, Vol. 18, No. 5, 2007.



P 59

A rare case of a ductus arteriosus and left pulmonary artery thrombus in the neonate

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Intro: Spontaneous thrombus formation in the ductus arteriosus (DA) represents a rare condition that can cause potentially fatal thromboembolism. Its diagnosis can be achieved using pre- and postnatal echocardiography. Due to the small number of reported cases there is no consensus regarding management and therapy. In particular, DA thrombus formation has been reported to be a major complication of spontaneous DA aneurysms in the neonate.
Methods: The case of a female newborn with a thrombus in the DA and left pulmonary artery (LPA) is described.
Results: Following delivery the patient presented with irregular transcutaneous oxygen saturation with SpO_2 values between 88–91%. A first transthoracic echocardiography (TTE) performed on postnatal day 5 revealed right ventricular hypertrophy with reduced contractility, a closed DA without any sign for thrombus formation. A follow-up TTE three weeks postnatally depicted a large thrombus in the DA protruding into the LPA. After initial i.v. anticoagulation with heparin, a lysis therapy with alteplase was run during 24h. These therapeutic interventions did not result in a resolution of the thrombus. Thus surgical removal was performed and consisted of a thrombectomy, surgical closure of the DA and LPA patch plastic. Postoperative TTE demonstrated no residual thrombus nor residual vascular shunt after DA ligation. The patient was discharged 5 weeks postoperatively in good clinical conditions. A thrombophilia screening is planned at the age of 3–6 months.
Discussion: This case illustrates a rare case of a DA thrombus in a newborn. We suppose that the thrombus may have been caused by an aneurysm of the DA. Given the right heart hypertrophy observed in TTE a premature closure of DA before birth by the thrombus formation may have occurred. We speculate that in the first postnatal TTE, the thrombus was not recognized as it was all imbedded in the DA; the protruding part into LPA was possibly the result of a thrombotic apposition. This case exemplifies the importance of repeated TTE assessments in patients with premature DA closure as delayed thrombus visibility might occur. Potential complications of DA thrombus formation include pulmonary as well as arterial embolism or stenosis of the pulmonary trunk. In the follow-up of cases with DA thrombus formation also a status of hypercoagulability needs to be ruled out by laboratory screenings.

P 60

An atypical cause of neonatal hyperammoniemia

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Case report: A term newborn presented with pallor on day 1, regurgitations and refused to feed. Glycemia was elevated, 8.7 mmol/L. There was no infectious risk. CBC and CRP were normal. On Day 2, she was slightly hypotonic. Blood gas showed normal pH 7.38, BE –1.6, elevated lactate (3.7 mmol/L) and hyperglycemia (8 to 10 mmol/L). Urine control showed no glucose, no ketosis and transient proteinuria. She had no fever whereas she had once hematemesis. On day 3, she had abdominal distention, pallor and had no meconium emission since birth. Blood gas was normal, with glucose persistently elevated. Because of persistent hyperglycemia, transient hyperlactatemia and hypotonia, ammonium was controlled and was high: 347 µmol/L (N <150 mmol/L). She developed respiratory failure and abnormal movements with opisthotonos and was intubated. E. coli was found in hemocultures without inflammatory syndrome and ceftazidime was given. In 2 hours ammonium increased up to 430 and hemofiltration was discussed. Hyperammonemia was treated with a loading dose and maintenance of sodium benzoate and L-arginine, carnitine, hydroxycobalamine and 12 mg/kg/min of glucose. Normalization level of ammonium occurred in 24 hours (ammonium decreased 200 points in the first 2 hours). The treatment leaded to a prompt recovery of the encephalopathy (hypotonia, abnormal movements), abdominal distension and hyperglycemia. A metabolic disease was excluded. The newborn was released with a normal physical exam and blood tests at 3 weeks of age.
Discussion: Usually inborn errors of urea synthesis presents as encephalopathy with an important elevation of ammonium and metabolic acidosis. Prompt treatment with sodium benzoate,

L-arginine, carnitine, hydroxycobalamine and high glucose intake is crucial. Neonatal hemofiltration is difficult to place because of catheter sizes. Hyperammonemia is a neurological emergency that could be lethall. This case is atypical because hyperammonemia was caused by sepsis with an E. coli that wasn't urease negative. No metabolic disease was diagnosed.
Conclusions: Hyperammonemia is an emergency. In front of a newborn with feeding disorder, neurological abnormalities (even slight) and atypical laboratory findings like hyperglycemia and hyperlactatemia without acidosis, ammonium must be controlled. Other causes than metabolic disease, like sepsis can cause severe hyperammonemia.

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Respiratory distress and perinatal stroke

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Case report: A term infant had an excellent adaptation after C-section for intrauterine growth restriction with gestational hypertension. Mother received neomercazole for Grave's disease. At 24h respiratory distress with peribuccal cyanosis, tachycardia and multiples regurgitations was noted. Chest radiography, CBC, CRP and blood gas were normal. Antibiotic and CPAP were started. At 36h right seizures developped with chewing, eye rolling, left lateral nystagmus and loss of contact. Right side tonus was augmented with asymmetric reflexes. Brain scan showed an extended parieto-occipital stroke. At 72h EEG showed abnormal electric activity in the sagittal zone. Brain echography was normal. At 6 d, MRI showed an ischemic lesion in the left middle cerebral artery (MCA) territory, without hemorrhage or signs of vascular occlusion. Echocardiography showed patent foramen ovale with right-to-left predominant bidirectional shunt without intra cavitary thrombus. Coagulation tests were normal without thrombophilia in the family. No cause was found to explain the cerebral stroke.
Discussion: Neonatal arterial ischemic stroke (AIS) occurs in 1:2300 to 1:5000 term births. Maternal (thrombophilia, gestational hypertension, smoking, intrauterine growth retardation, infection, cocaine), fetal (thrombophilia, congenital heart disease (CHD), hypoglycemia, perinatal asphyxia, need of resuscitation) and placental (chorioamionitis, infarcts, underweight) risk factors are described. Most common presenting feature is focal seizure (69–90% of cases) in the first 3 days of life, sometimes preceded by isolated respiratory distress in 24% of cases. Other symptoms are encephalopathy, apnea, chewing, tone abnormalities and feeding difficulties. Most strokes are unilateral, in the left hemisphere and in the MCA territory.
Conclusion: Respiratory distress without pulmonary cause could evoke a neonatal stroke as in this case. On the other hand, because AIS is of high risk for persistant sequelae, it's important to identify the concomitant risk factors as prothrombotic disorders, CHD and vascular malformation in order to decrease the risk of recurrence (2%).

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Neonatal mastitis due to methicillin resistant Staphylococcus aureus (MRSA)

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Introduction: Neonatal mastitis is an uncommon infection. No guidelines exist for optimal management. We present a case with a complicated course and review the literature.
Case report: A 15 day old male neonate, term born, to a 22 year old Erythrean MRSA carrier, presented with erythema and painful swelling of the right breast. Clinical examination revealed a well-looking, febrile (38.5 °C) neonate, with a swollen, tender 7.5 x 6.0 cm right breast without discharge. Leukocyte count was 21.8 G/L with 1635/uL band neutrophils, C-reactive protein was 52.9 mg/L, procalcitonin was 8.58 microg/L. Ultrasound showed no abscess. Intravenous ceftriaxone and vancomycine were started after microbiology testing. Blood cultures were negative, breast swab returned positive for MRSA. On day 4 purulent discharge returned positive for MRSA. Ultrasound showed partial abscessification. No drainage was performed. On day 5 intravenous therapy was switched to oral clindamycine and patient dismissed. On day 9 the infant had a recurrent increase in breast inflammation with a purulent discharge, no fever or blood test anomalies. Clindamycine was changed to the intravenous route. Incision and drainage were performed on day 20. After additional 7 days of oral clindamycine the patient was cured.
Discussion: *Staphylococcus aureus* is the usual cause of infant mastitis during the first 3 months of life, with a peak incidence at 3 weeks. MRSA has been reported. Literature is controversial as to optimal management. Most often S.aureus covering intravenous antibiotics are started. Some authors advocate initial oral treatment. Also some authors advocate more conservative approach as to surgical treatment. In our case, the absence of fever and good clinical response from day 3 onwards allowed the switch to oral therapy on day 5. Surgical drainage was not performed immediately: initially because of the partial and small size abscess; after the recurrent inflammation with a slow and favorable response because the abscess entering the process of spontaneous cutaneous fistula formation.
Conclusion: Management of neonatal mastitis is empirical. This case illustrates that intravenous therapy should be considered and oral treatment can be an option in the absence of local severity or resistant germs. Surgical treatment should be considered when abscess forms. Controlled trials are needed to establish optimal management.

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Spontaneous Pneumothorax in Neonates: A Single Center Experience

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Introduction: Pneumothorax (PTX) in a neonate may turn into a life-threatening situation. Its association with respiratory distress and ventilatory support is well known. Drainage with tube thoracostomy (TT) is the most frequently used treatment modality; however single puncture aspiration (SPA) and expectant management are well established alternatives. The purpose of this study was to evaluate our patients with spontaneous PTX, to determine risk factors, and to describe clinical presentation, treatment, complications and outcome.
Methods: We retrospectively analyzed data of all neonates with diagnosis of PTX hospitalized in our perinatal centre from 01/2009–12/2014. Among the 2280 admissions throughout this time period, we identified 30 infants. Owing to congenital malformations/severe underlying pulmonary pathology eight infants were omitted from the analysis.
Results: 22 infants (5 female, 17 male) were diagnosed with spontaneous PTX in this time period, with a median gestational age (GA) of 33^{+4} weeks (mean: 34^{+0}) and median birth weight of 2150g (mean: 2304 g). The incidences (in NICU population) related to GA were 1.6% (<32^{+0} weeks), 0.7% (32^{+0} – 36^{+6} weeks) and 0.9% (≥37^{+0} weeks). 19 infants (86%) received respiratory support (CPAP, bag/mask and/or intubation) at birth, and all but one infant developed respiratory distress prior to diagnosis of PTX. The median age at diagnosis was 14 hours (range: 2 – 39.5 hours) and 80% were receiving some form of respiratory support when diagnosed. In 4 infants (18%) PTX resolved with expectant management, and in 3 patients (14%) PTX resolved with high frequency oscillation ventilation only. 4 infants were initially treated with SPA, however 3 of these patients subsequently required drainage with TT. 5 infants (23%) presented with a tension PTX (mediastinal shift) and 3 developed pulmonary/mediastinal emphysema. One patient needed re-TT owing to recurrent PTX. All patients survived.
Conclusion: Although the incidence of PTX is highest in the very low birth weight population, the condition is encountered more frequently in the group of late preterm and term infants. Hence, the pediatrician caring for this patient population should have a high level of suspicion in view of worsening respiratory distress. In a NICU caring for preterm/term infants with respiratory distress, staff trained to drain a PTX should be readily available.

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Apparent life threatening event (ALTE) in an healthy newborn in the first hour of life

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Introduction: An apparent life threatening event (ALTE) or sudden death in a presumably healthy newborn in the first hours after birth is an exceptional event. We report a case of a healthy newborn who presents a cardiorespiratory arrest in his first hour of life.
Case presentation: Our patient was delivered at 39 weeks of gestation to a 35 year-old primiparous mother known for gestational diabetes. It was a vaginal, forceps-assisted delivery because of dystocia in a context of meconium stained amniotic fluid. Neonatal adaptation was good (Apgar score 8/9/10), umbilical cord pH 7.23/7.38, suction was unnecessary. After usual primary care, he was allowed to

stay on his mother for skin-to-skin contact and was left with both of his parents in the delivery room. Fifty minutes after birth, the father noted the newborn to be unresponsiveness. The midwife confirmed cardiorespiratory arrest and started CPR. Intubation, 10 min cardiac massage and one dose of intratracheal adrenaline were needed. The first blood gas sample obtained 15 minutes after initiation of CPR showed a severe metabolic acidosis, pH 6.77, pCO_2 68.9 mm Hg, bicarbonate 10 mmol/l, lactate 16 mmol/l. Heart rate was restored after 10 minutes, and after stabilisation, he was transferred to a level III NICU. He needed few days of mechanical ventilation and presented a pulmonary hypertension, but he improved rapidly. A hypothermia protocol was not applied and he was discharged from the NICU after 9 days, back to our pediatric ward. Neurological examination was unremarkable, the electroencephalogram, brain MRI at the age of 8 days and the cardiac ultrasonography were normal. The infectious and metabolic investigations were negative. No new event occurred during the following 5 days of observation, that were necessary for breastfeeding introduction and to ensure that parents have adequate time to trust their baby before going home.
Discussion: Even if such a critical event is very rare, it remains of concern due to a mortality of >75% and frequent severe neurologic deficit in survivors. Mechanisms leading to such events remain unclear, but according to the literature, the risk is increased in newborn of primiparous mothers. Most described patients were in prone position on the mother, as they were both left alone. To our opinion, this must not lead to reconsider the early skin-to-skin contact, the benefits outweighing the risks. However particular considerations should be given to the positioning of the newborn, especially in primiparous mothers. It should be avoided in tired mothers or in those under sedation. Monitoring of the newborns in the delivery room should be considered in "at risk" situations. As most of the cases of ALTE reported in the literature presented in the first hours of life and considering the extremely poor outcome, a greater attention should be given to this problematic and nursing staff instructed.

P 65

Breastfeeding-associated hypernatremia: review of the literature

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Introduction: There are increasingly appearing reports on hypernatremia, a potentially devastating condition, in exclusively breastfed newborn infants.
Methods: We performed a review of the existing literature in the National Library of Medicine database and in the search engine Google Scholar. A total of 115 reports were included in the final analysis.
Results: Breastfeeding-associated hypernatremia was recognized in infants ≤21 days of age and ≥10% weight loss of birth weight. Cesarean delivery, primiparity, breast anomalies or breastfeeding problems, excessive pre-pregnancy maternal weight, delayed first breastfeeding, absent previous breastfeeding experience and low maternal educational level were significantly linked to breastfeeding-associated hypernatremia. In addition to excessive weight loss, following clinical findings were observed: poor feeding, poor hydration state, jaundice, excessive body temperature, irritability or lethargy, decreased urine output and epileptic seizures. Midwives' rule of thumb that newborns may lose up to 10% of birth weight is supported by the literature: daily weighing during the first 4–5 days of life is a cost-effective method for detecting dehydration.
Conclusion: Everything should be done to promote breastfeeding because its advantages are unquestionable. Therefore, it is crucial to prevent neonatal hypernatremic dehydration secondary to undetected lactation failure. When factors that predispose to breastfeeding-associated hypernatremia are observed, intervention to maximize maternal milk supply and improve infant milk intake may avert this problem. Weight monitoring and supplemental fluids in the presence of weight loss ≥10% of birth weight allow early detection and intervention.

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Levamisol induced vasculitis in a 6 year old girl with steroid-sensible idiopathic nephrotic syndrome

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Introduction: Levamisole is frequently used as adjuvant therapy in children with relapsing or steroid dependent idiopathic nephrotic syndrome.
Case report: A 6 year old girl with chronic steroid-sensible nephrotic syndrome in remission, on an oral levamisole therapy (8 mg/kg/week) for three years with an additional therapy of prednisone (1.25 mg every second day), was admitted to our emergency department. She presented with painfull skin lesions on both ears and cheeks, showing rapidly enlarging purpuric and erythematous macules, with small necrotic areas. Furthermore, she had fever and cold-like symptoms since 3 days without arthralgias. The last days she had been exposed to frosty weather outside. Laboratory tests showed leucopenia 3.22 G/l, a reduced neutrophil count 1.07 G/l and CPR of 13.2 mg/l. Antibodies to mycoplasma pneumonia and cryoglobulins were negative. p-ANCA (>5120) and Anti-PR3 (183 U/l) were strongly positive, Anti-MPO, Anti-nuclear Antibodies, Antiphospholipid Antibodies and Anti-DNA Antibodies were negative. Levamisole therapy was stopped. The skin lesions completely disappeared, but reoccurred twice in the following 12 weeks with small purpuric lesions on the ears. High p-ANCA levels persisted after 3 months.
Conclusion: We report a Levamisol induced vasculitis with persisting high titer p-ANCA. If p-ANCA are induced or constitutionally present in our patient is not known. Histologic examination was not performed because the immunoassay showed a p-ANCA pattern which is typically described in literature in patients with levamisole induced vasculitis. Normally skin lesions disappear in these patients although severe skin reactions with scarring are described in adult patients. Levamisole is an anthelminthic drug with immunomodulating effect, restoring depressed immune function, stimulating formation of antigens and T-cell activation as well as increasing neutrophil mobility and chemotaxis. In patients with nephrotic syndrome it is used because of the steroid sparing effect. Cutaneous side-effects have been infrequently discussed in paediatric patients, but more often in adults consuming Levamisole-adulterated cocaine.

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Goodpasture's disease in childhood: the results of a Web-based survey

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Background: Antiglomerular basement membrane antibody disease (Goodpasture's disease (GD)) is extremely rare in children, information about manifestation, diagnosis and especially therapy of GD in children is lacking. The purpose of this study is to gain information about management of GD in children.
Methods: We performed an international survey on GD in childhood among pediatric nephrologists. An invitation to participate in this survey was sent by electronic mail. In an effort to get as much information as possible about management of GD in childhood we analyzed the responses to 28 questions that elicited information about the following: incidence, demographics, patient's history and clinical presentation, diagnostics performed, acute and chronic therapy, course of the disease and outcome.
Results: 66 hospitals from 4 continents answered the questionnaire. Forty patients with GD have been followed during the last ten years. The hospitals following patients with GD cover more than 113 Mio (1–25 Mio/hospital) inhabitants; the calculated incidence is about 0.025 childhood cases per million inhabitants per year. For 24 patients we received more detailed information. The patient's age by the time of diagnosis varies between 2–19 years, the median age is 14 years. All patients are Caucasian and 56% are male. 75% of the patients have kidney and lung manifestations, 25% only have renal manifestation. The median age of patients with kidney and lung manifestations is 15 years, whereas the median age of patients with only kidney manifestation is 6 years ($p = 0.09$); no patients under the age of 11 years have been described with lung manifestation. At first presentation 91% have renal and 63% have pulmonary symptoms. The acute therapy included steroids, cyclophosphamide and/or plasmapheresis. No difference in the therapy strategy was noted between patients with or without lung manifestation. Moreover, the therapy was not adapted to renal biopsy results. Renal biopsy showed

in 77% of the cases >50% crescents, 36% had even more than 90% crescents. All patients with >90% crescents needed renal replacement therapy. During the acute period 79% of all the patients needed renal replacement therapy. Irreversible end stage renal disease was described in 56% of the patients. Nevertheless complete recovery of lung disease was documented in 82%. No case of death among the pediatric patients was reported.
Conclusion: This survey confirms that GD is an extremely rare disease in childhood, yet toddlers may also be affected. Lung manifestation seems to be present only from adolescence. Outcome is better than in adults with a survival rate of 100% and renal improvement also in patients with severe biopsy findings. Guidelines for treatment of GD in childhood are warranted.

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Painful menarch: are both kidneys here?

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Introduction: In girls, error's of embryological development of the (para)mesonephrotic ducts can result in renal agenesis and homolateral mullerian duct abnormalities, most often being uterus didelphis and blind hemivagina (also known as Herlyn-Werner-Wunderlich syndrome, part of the entity called "obstructed hemivagina and ispilateral renal anomaly"). If not discovered at prenatal ultrasound, this malformation can remain latent during childhood and will disclose at puberty with severe pain at menarche and presence of an increasing abdominal mass due to hematocolpos.
Case description: A 14 year-old girl without relevant medical history presented with increasing abdominal pain. She had her menarche three months before with normal discharge. Examination of the external genitalia showed a normally perforated hymen and no vulvar bulging. Because of severe pain during her menarche, an abdominal ultrasound had already been performed, which revealed agenesis of left kidney without other abnormality noticed. As pain increased with time, a new abdominal ultrasound was done, which further showed a pelvic mass. A Herlyn-Werner-Wunderlich syndrome was suspected and a pelvic MRI finally disclosed a uterus didelphis with mild distension of the left hemi-uterus, but large dilatation of the left hemi-vagina filled with a semi-liquid content, the right hemi-uterus and -vagina were of normal aspect. Transvaginal surgery was performed to resect the obstructing vaginal septum and drain the hematocolpos. Resolution of abdominal pain was immediate and the patient was discharged 2 days after surgery. Six months after surgery the girl has uneventful menstruations and, on control ultrasound, complete disappearance of the pelvic collection. Future pregnancies can be expected; women with uterus didelphis do not present difficulties to conceive and even multiple pregnancies have been reported, with one fetus in each uterus.
Conclusion: In cases of a solitary kidney in a female fetus or child, beware of hematocolpos at puberty. Severe menstrual pain should yet be investigated, especially if pain is not relieved at the end of each period.

P 69

C3-glomerulopathy caused by a dysregulation of the alternative pathway of the complement system: improvement with the anti-C5 monoclonal antibody eculizumab

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Background: C3 glomerulopathy (C3G) is a recently defined clinical entity comprising glomerular disorders characterized by C3 predominant immunofluorescence on the renal biopsy. Light microscopy shows a variable picture, ranging from mesangial proliferation to a membranoproliferative pattern and diffuse endocapillary proliferation. Recent advances have expanded the understanding of the underlying mechanisms, leading to the hypothesis that blocking the alternative complement pathway may be an effective treatment for C3Gs, as has been shown in other renal diseases driven by alternative pathway dysregulation, such as atypical hemolytic uremic syndrome.
Case report: A 7 year old girl presented with a wheezing episode, macrohematuria and generalized edema. Nephrotic-nephritic syndrome was diagnosed (hematuria, gross proteinuria (500 mg proteine/mmol creatinine), edema, arterial hypertension) and the renal biopsy showed a membranoproliferative type I pattern with C3 deposits on immunofluorescence, without deposits of immunoglobulins. Antiproteinuric therapy with the angiotensin receptor blocker candesartan, corticosteroids and mycophenolate failed to improve the nephrotic-range proteinuria. Further investigations of the complement system showed an activation of the alternative pathway with factor H deficiency; therefore therapy with the anti-C5 monoclonal antibody eculizumab was started (600 mg every week for three weeks followed by a maintenance therapy of 600 mg every 2 weeks). Corticosteroids and mycophenolate were tapered off. After 8 weeks of therapy, proteinuria improved considerably (80 mg proteine/mmol creatinine), renal function remained normal and hematuria persisted.
Conclusion: This case highlights the importance of the complement system in the pathogenesis of C3G. Treatment with the anti-C5 monoclonal antibody eculizumab seems to be beneficial in this young patient. Probably, treatment with eculizumab for C3G should be initiated early, before major sclerotic modifications have occurred in the renal glomeruli.

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Old drug, new mutation in neonatal epileptic encephalopathies

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Introduction: KCNQ2 mutations have been detected as a cause of different "idiopathic" neonatal epileptic encephalopathies and the spectrum comprises variable phenotypes. The succesful treatment of the seizures may be challenging: a variety of antiepileptic drugs is often needed.
Cases and results: 1st case: After uneventful familiy and pregnancy history, the newborn presented with repetetive partial seizures beginning at the 2nd day after birth. Synchronous to clinical overt seizures with alternating head- and eye-deviation, bedside EEG showed alternating left/right-hemispheric seizure onset and pathological background consistent with the diagnosis of severe migrating partial epilepsy. Treatment with vitamin B6, phenobarbitone, levetiracetam, clobazam, topiramate showed no effect. Introducing phenytoin led to cessation of clinical overt seizures and marked reduction of epileptic activity in the EEG without any other side-effect. Genetic analysis by NGS panel revealed a pathogenic mutation c.578C>T/p.A193V in the KCNQ2 gene. 2nd case: The girl presented at the age of 2 hours with frequent partial seizures. Her family history was positive for benign familial neonatal seizures. Interictal EEG did not show specific epileptic discharges. Therapy with carbamazepin, topiramate, phenobarbital showed no positive effect. At the age of 3 month adding phenytoin to PHB led to cessation of seizures. Genetic analysis by NGS panel showed also a mutation c.1657C>T/p.R553W in the KCNQ2 gene.
Discussion: Phenytoin was first introduced in the 1930`s as an anticonvulsant acting as a blocker of the voltage-dependent neuronal sodium channel. Its use as a second line drug for neonatal seizures has been limited due to toxicity and pharmocodynamic concerns and newer AED have emerged. Our case observations suggest that it may have a specific therapeutic effect in difficult-to-treat neonatal epileptic encephalopathies associated with KCNQ2 mutations and worth a try in comparable cases.

P 71

Relapse of herpes simplex encephalitis after surgical treatment for refractorv epilepsy

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Background: Herpes simplex virus (HSV) encephalitis reactivation after neurosurgery is a very rare condition. Early recognition and treatment is mandatory to prevent significant morbidity. Clinicians have to keep HSV reactivation in their differential diagnosis in a febrile patient after surgical procedures.
Case-Report: A 12 y.o. girl with a past medical history of HSV encephalitis at 11 months of age, complicated by mild left hemiparesy and refractory focal epilepsy underwent a selective right temporal lobectomy and amygdalohippocampectomy. The child was sent home at day 7 without seizure recurrence. On day 11 after surgery, she presented with new onset fever, headache, and scar pain. Neurological exam was stable and laboratory results unremarkable. After three days of fever and persistent headache, level of consciousness worsened. Magnetic resonance imaging (MRI) demonstrated abnormal signal

intensity with vasogenic edema distant from the resected area and compatible with an inflammatory process. Cerebrospinal fluid (CSF) testing showed 91 M/l leucocytes (85% lymphocytes), hyperproteinorachia (2.05 g/l), hypoglycorachia (1.9 mmol/l. IV acyclovir was immediately started and continued for 21 days after evidence of positive for HSV-1 by Polymerase Chain Reaction (PCR) in the CSF. Electroencephalography at day 1 of treatment revealed a diffuse slowing. At day 4 of treatment, a brain MRI revealed a malignant edema with mass effect correlated to a worsening of the level of consciousness and headache, prompting a rapid decompressive right (fronto-parieto-temporal) craniectomy. Thereafter, evolution was finally favorable with apyrexia 4 days after onset of antiviral treatment. The patient's neurological status returned to baseline before HSV reactivation except for a mild worsening of her preexisting left hemiplegia.
Discussion: HSV-1 is the most devastating viral encephalitis, most often at the time of the initial infection. Reactivation of the latent virus with relapsing encephalitis after a prolonged silent period has been described, and brain surgery identified as a potential trigger, including in children. It is not clear whether the neurosurgery itself or simply the stress of the surgical procedure causes HSV reactivation. Early suspicion and prompt initiation of acyclovir in patients presenting with symptoms of encephalitis can prevent further mortality and significant morbidity from HSV encephalitis. Prophylactic acyclovir treatment should be considered in patients with prior history of HSV encephalitis who undergo intracranial procedures.

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Robot-assisted gait training might be beneficial for more severely affected children with cerebral palsy: Brief Report

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Purpose: Robot-assisted gait training (RAGT) can complement conventional therapies in children with cerebral palsy. We investigated changes in walking-related outcomes between children with different Gross Motor Function Classification System (GMFCS) levels and the dose-response relationship.
Methods: Data from 67 children (3.9–19.9 years) with GMFCS levels II-IV were evaluated retrospectively. Every child received RAGT with the Lokomat complementing a multidisciplinary rehabilitation program. Changes in various walking-related outcomes were assessed.
Results: Walking-related outcomes did not improve differently between GMFCS level groups. Significant within-group improvements were mainly observed in children with GMFCS level IV. A dose-response relationship was present for children with GMFCS levels III and IV.
Conclusions: Our results indicated that, although children with a GMFCS level IV walked less during an average Lokomat session, they experienced significant improvements in walking-related outcomes. Further, training dose correlated with changes in walking-related outcomes. However, between-group differences in changes in walking-related outcomes were not significant.

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Guillain Barré Syndrome: Diagnosis in spite of varying presenting symptoms!

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Aim: To drive awareness on the different ways Guillain Barré Syndrome (GBS) can present, resulting in varying outcomes, from total recovery to wheelchair-bound.
Methods: Retrospective analysis of 5 patients admitted from 2011–2014 with the diagnosis of GBS.
Results:

Table 1

Patient	Age	Presenting symptoms	Areflexia	Cytoalbuminary diss.
1	18 mo	Refusal to walk, pain in legs	–	+
2	19 mo	Severe respiratory distress, acute paraplegia	+	+
3	3y3 mo	Leg weakness	+	+
4	3y4 mo	Unsteady gait	+	+
5	15 y	Central facial palsy and acute ataxia	+	+

Patients were 18 months to 15 years old at the time of diagnosis, all with a history of prodromal illness one to four weeks prior to the largely varying neurological symptoms. See table 1. All five patients received intravenous immunoglobulins. Two patients had to be transferred to an intensive care unit. All required physiotherapie at discharge from the hospital. Three patients recovered completely. Patient 5 who suffered from the Miller Fischer variant had slight motor deficits at discharge. Patient 2 is wheelchair-bound four months after the diagnosis.
Conclusion: Presenting symptoms leading to GBS were very variable. They began after a mild respiratory tract infection or gastroenteritis. Paraparesis was consistently present: In most cases an areflexia was present at some point during illness. It is highly important to consider this diagnosis in suspicious patients in order to start the required treatment as soon as possible.

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One seizure, two seizures: what a headache!

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Introduction: Cerebral Cavernous Malformation (CCM) are Central Nervous System (CNS) vascular anomalies, which are found in 0.1 to 0.5% of total population. One fourth of these lesions occur in the pediatric age group. Two peaks of incidence of CCM are reported in the pediatric population below 3 years of age and between 13–16 years old. These malformations could be responsible for seizures.
Case report: We report a case of CCM in a 15 year old female who presented with focal then focal secondarily generalized seizures, occurring during the same day. Seizures were associated to severe headache, but no fever nor other signs of infection. She had already manifested with two similar focal seizures in the past, never investigated nor treated. Family history was negative and no abuse of drugs was suspected. Physical examination, and in particular neurological examination, was normal. CT scan showed a right frontal cortical acute bleeding. MRI with angiographic sequences showed a precentral multilocular lesion (6.5x4.5x6.25 mm) surrounded by hemosiderin deposits confirming cavernoma; no arteriovenous malformation (AVM) was found. EEG was consistent and showed an irritative slowlying on the right frontotemporal area. We began an anti-epileptic drug (AED) (levetiracetam) and her evolution over six months was favorable, without any relapsing of seizures nor other acute episode.
Discussion: CCM consists of closely clustered, enlarged capillary channels with a single layer of endothelium without mature vessel wall elements or normal intervening brain parenchyma. Symptoms, which are secondary to bleeding, may include seizures, focal neurological deficit and headaches. Cavernoma itself is not thought to be responsible for these deficits. Factors that have been implicated as likely sources of neurologic changes are hemosiderosis, which is due to bleeding from the cavernoma into the surrounding brain tissue, and reactive gliosis, which is caused by recurrent bleeding. Diagnosis is established by characteristic imaging finding (especially MRI) and treatment is basically based on AED; neurosurgery should be discussed case-by-case, depending on CCM location and symptoms.
Conclusion: CCM are not uncommon and are one of the two main causes of brain hemorrhage in children with ruptured AVM. CCM can occur sporadically or as familial autosomal dominant condition with incomplete penetrance and variable clinical expression attributable to mutations in three different genes (CCM1, CCM2, CCM3). CCM may occur also in the oncology population following cerebral radiotherapy, especially in children under ten years old. It should be envisaged in front of focal seizures associated with headache. Facing focal seizures with severe headache, it is important to rule out encephalitis, but clinician must think about a brain hemorrhage even in pediatric population.

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Status epilepticus in an adolescent treated for lymphoma

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Case report: A 14 y.o. boy known for Hodgkin Lymphoma, treated with prednisone, vincristine, doxorubicine and etoposide, presented refractory status epilepticus preceded by 12 h confusion. He had mild hypertension (140/90 mm Hg). Brain CT scan performed after intubation revealed hypodensities of sub-cortical temporo-parieto-occipital areas. MRI demonstrated symmetrical bilateral multiple T2 hyper intense anomalies of the white matter (occipital and frontal), appearing as vasogenic edema. Blood exam revealed agranulocytosis (WBC 0.1G/l) and mild inflammatory signs (CRP 73 mg/l). CSF analyse was normal. P.R.E.S. (Posterior Reversible Encephalopathy Syndrome) was diagnosed. Complete recovery occurred at day 7 but MRI anomalies persisted 2 months later.
Discussion: PRES is a clinical and radiological syndrome characterised by variable associations of seizure, consciousness impairment, headache, visual abnormalities, cortical blindness, vomiting and focal neurological signs, after exclusion of central nervous system (CNS) infection or malignancy, methotrexate encephalopathy, stroke and metabolic causes. MRI findings are most often symmetrical and posterior cerebral abnormalities. PRES in children is generally found as a complication of cancer treatment (leukaemia > solid tumors). Hypertension is a main condition for PRES. According to one of leading patho-physiologic hypothesis, hypertension, even mild, causes PRES by exceeding brain vascular autoregulation leading to disruption of blood-brain barrier and vasogenic edema. No particular chemotherapeutic agent has been incriminated in childhood PRES. Outcomes show that the neurologic deficits are reversible in 50 to 86% of cases.
Conclusions: PRES is a well known complication of childhood cancer treatment. It should be rapidly diagnosed when neurological symptoms and hypertension are present. MRI with diffusion sequences should be performed to confirm diagnosis. Prompt initiation of anticonvulsant and anti-hypertension drugs is important in the acute phase to avoid complications of vasogenic edema.

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Post-varicella angiopathy and stroke

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Introduction: Varicella zoster virus (VZV) is a highly neurotropic virus. After primary infection, it remains latent in the nerve cells and may reactivate many years later, resulting in various conditions affecting the central nervous system, such as vasculopathy and stroke. We report the case of an immunocompetent child who presented with cerebrovascular disease 7 months after chickenpox.
Case report: A previously healthy 3 y.o. girl was admitted for balance impairment, moderate right hemiparesis, right unilateral neglect and dystonia. Neurological symptoms evolved gradually starting from 10 days before admission. No fever, headache, consciousness or cognitive impairment were noted. The interval from varicella rash was 7 months. RMI showed moderated signal impairment in left globus pallidus, stenosis of the homolateral middle cerebral artery (MCA) and of the anterior cerebral artery (ACA). The magnetic resonance angiogram (MRA) confirmed the lesion with raising signal of the correspondent arteries. Serology shows positive VZV IgG antibodies (3827 UI/l) and negative IgM. Herpes simplex virus and Lyme antibodies were negative. The child received acetylsalicylic acid, 100 mg/day, with good clinical evolution. Most symptoms regressed within a month, but a mild right hand residual paresis. Clinical following with transcranial Doppler ultrasonography shows the persistence of blood flow acceleration of the left MCA and ACA, corresponding to residual stenosis of <50%, confirmed by MRI 6 months later.
Discussion: Arterial ischemic stroke during childhood is relatively rare. The etiologies are multifactorial and related to an arteriopathy in many cases. Post-varicella angiopathy is a transient cerebral arteriopathy, caused by productive viral infection in cerebral arteries. Ischemic stroke following varicella is rarely described, with an estimated incidence of 1/15.000 children.
Conclusion: This case highlights the need to consider chickenpox in the differential diagnosis of ischemic stroke in children who had VZV infection during previous 12 months. After exclusion of other diagnosis, antiaggregant drugs are recommended to prevent thrombus expansion and recurrent stroke, but there›s no consensus about therapy duration. Acyclovir and steroids should be considered. Neurologic outcome is usually good also if vascular anomalies can be long-lasting.

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Hydrocephalus in children: report of five consecutive cases and comprehensive review of the literature

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Introduction: Hydrocephalus can be progressive or spontaneously arrested. In arrested hydrocephalus (AH) the balance between production and absorption of the cerebrospinal fluid is restored, ventricular dilatation is no longer progressive, and the intracranial pressure returns to normal values. Patients are mostly asymptomatic, and no surgical treatment is necessary. We report the case of 5 patients diagnosed with AH.
Methods: Data of 5 children referred to our two institutions were prospectively collected over 2 years. They included sex, age, clinical features, brain magnetic resonance imaging (MRI), and follow-up.
Results: The mean age of the 5 patients at the first control was 2.6 years (SD 2.0 years; range 0.3–4.8 years), and mean follow-up time was 1.2 years. The main complaint at presentation was a macrocephaly. Mild motor disorders were present in four out of five cases. Typical symptoms and signs associated with raised ICP were absent in all 5 patients. MRI studies showed different combination of ventricular, and extra-axial subarachnoid space enlargement. No definitive cause for hydrocephalus was found in any of our patients. In each case we proposed an initial conservative approach. During the follow-up, we observed complete resolution of the motor disturbances in 2 patients and improvement in another. One patient remained asymptomatic throughout the follow-up. subsequent MRIs showed unchanged radiologic features. In one child we observed an aggravation of the motor disorders, and shift to a surgical approach was needed.
Discussion: Until now, no specific clinical or paraclinical features permitting the certain diagnosis of AH have been described. Making a distinction between arrested and progressive hydrocephalus is fundamental, because of the opposed appropriate management. If AH is assumed as a benign self-limited condition, progressive hydrocephalus is a potentially fatal state, which requires a prompt intervention. With this work we offer a review of the current literature available to date, and we propose our recommendations for the management of this particular condition.

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PCDH 19 female related infantile epilepsy: a case report

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Background: PCDH19 encodes protocadherin 19 and is located on chromosome Xq22. Abnormalities in this gene cause female-related epilepsy of variable severity. PCDH19-related female epilepsy (PCDH19-epilepsy) is a unique X-linked disease, characterized by early onset, fever sensitivity, seizure clusters of focal seizures, varying degree if intellectual disability and/or autistic traits.
Case report: We introduce a 14-year girl known for seizures, occurring in clusters and mostly fever-associated since she was 15 months. She usually presents loss of consciousness, right arm extroversion, salivation, cyanosis, chewing movements or extended tonic contraction, in a variable sequence. These events were investigated through electrophysiological analysis (EEG, polysomnography) and brain imaging. Several critical events were recorded, clinically stereotyped, coming from frontal cortex without signs of lateralization. Brain-RMN presented an uncertain small dysplastic area in the frontal cortex. Our patient experienced seizures during day or night, sometimes with unexpected falls and head traumas, one of these causing subdural hematoma with favourable outcome. At onset, treatment started with valproic acid, then phenobarbital was added, as the least was not fully effective. This double therapy allowed good control of symptoms, except during febrile status. After a long period without seizures, the patient presented clusters of crisis. Through a period of 5 months she was given several combination of anti-epileptic drugs (topiramate, levetiracetam, oxcarbazepine, perampanel) with poor control of symptoms. In the last hospital admission she seemed to be rather well responsive to clonazepam and lacosamide, but significant deterioration occurred in course of febrile conditions. This inadequate reaction to anti-epileptic drugs lead us searching for genetic bases. The patient was tested for several genetic mutation epilepsy-related, finding out the PCHD19 gene aberration.
Conclusion: The clinical spectrum associated with PCDH19 mutations can overlap that of Dravet syndrome (DS). However, DS-like

patients with PCDH19 mutations slightly differ on average from classical DS patients: age at onset is slightly older, status epilepticus and occurrence of myoclonic jerks are less frequent, and long-term outcome is better in PCDH19-positive patients[5]. In closing, we sustain PCDH19-molecular testing being performed in females with early onset epilepsy poorly responsive to farmacologic treatment.

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A case of glycine encephalopathy

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Case report: This girl was born by elective cesarean section at $39^{4}/_{7}$ weeks gestation after an uneventful pregnancy with a normal neonatal adaptation (Apgar 6/9/10). On the 2nd day of life she was transferred to our hospital because of muscular hypotonia, lethargy, feeding difficulties and desaturations. Family history was unremarkable. Apart from a bilateral pes equinovarus and slight retrognathia, there were no dysmorphic signs, muscle-tendon reflexes were normal. Sepsis work-up, blood gas analysis, serum glucose, lactate, ammonia and urinary ketones were normal. Cranial ultrasound showed agenesis of corpus callosum. Shortly after admission, the girl showed myoclonic seizures, which responded well to phenobarbital. An EEG on day 3 showed a discontinuous pattern without epileptiform signals. A high glycine concentration (631 umol/L) was detected in serum as well as in the cerebrospinal fluid (154 umol/L) with an elevated liquor-serum-ratio (0.24). The diagnosis of nonketotic hyperglycinemia was corroborated also by typical changes seen on cranial MRI and MR spectroscopy. Myoclonic seizures reoccurred on the 5th day of life, the EEG now showing a classical suppression-burst-pattern. Therapy with sodium benzoate, as well as supportive treatment with phenobarbital, levetiracetam and folinic acid was started, with clear reduction of the seizure activity and some degree of increased alertness.
Discussion: Glycine encephalopathy (= nonketotic hyperglycinemia) is a rare autosomal recessive disease, first-described in 1965 by Gerritsen and colleagues and in 1968 by Ziter and colleagues. It is characterized by lethargy, apnea and difficult-to-manage myoclonic seizures, typically appearing during the early neonatal period. The prognosis is poor, with most patients dying within months or a few years. However, survival up to 10–15 years of age has been described, although the survivors have severe mental retardation and intractable seizures. Glycine acts as an inhibitory neurotransmitter in the brainstem and spinal cord (explaining lethargy and hypotonia) and an excitatory neurotransmitter (NMDA-receptor) in the cerebral cortex (explaining hyperexcitability and seizures). Glycine encephalopathy is caused by mutations in genes encoding proteins in the mitochondrial glycine cleavage system (the only glycine degradation pathway) leading to glycine accumulation. Treatment encompasses reduction of glycine by sodium benzoate, as well as NMDA-antagonism using dextromethorphane or ketamine. Early myoclonic encephalopathy is often associated with metabolic disorders and, as demonstrated by our patient, there needs to be a high index of suspicion for a metabolic cause when confronted with a neonate presenting with seizures and signs of encephalopathy.

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Subacute sclerosing panencephalitis: still present? A case report

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Subacute sclerosing panencephalitis (SSPE) is a devastating "slow virus" brain disease resulting from persistent measles virus infection of neurons and characterized by progressive, ultimately fatal, neurologic deterioration. SSPE is associated with the acquisition of measles before 2 years of age and onset of symptoms presents usually 2 to 10 years after infection. So far, no cure and consensus on its therapy has been found. Most therapeutic suggestions are based on clinical trials and single case reports. We describe the case of a 15 years old teenager who presented with acute onset of gait ataxia followed by myoclonic jerks and subtle cognitive decline. Definite diagnosis of SSPE was made by measuring elevated measles antibodies in cerebrospinal fluid (CSF), confirming specific intrathecal antibody production. According to the actual international literature and experience, we started a therapy with daily orally Isoprinosine (antiviral effect) in combination with an immunomodulatory therapy with Interferon beta (INF-b) subcutaneously three times a week. This regimen is reported to slow the progression of the disease and improve life expectancy in some patients. Unfortunately, after a few months of therapy, no significant clinical improvement was noted. Some data describe a benefit of local central nervous system instillation of Interferon alpha, with better outcome on the progression of disease compared to subcutaneously Interferon beta. We will try this mode of administration with the implantation of an intrathecal pump system. Further research is needed to better understand the pathophysiology of this disease and develop better therapeutic regimens. Measles and SSPE are preventable by maintenance of high rates of immunization in the population.

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Erythromelalgia: Doctor, please stop this pain!

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Introduction: Erythromelalgia (EM) is a rare, chronic, debilitating condition characterized by a triad of symptoms: redness, warmth and severe burning pain in the distal extremities. Despite progress in the understanding of the disease, the complete underlying pathophysiology remains unclear. Patients suffer from severe pain with substantial morbidity and mortality.
Case report: A 10 year-old-boy, in good health, presented with a 3-week history of new onset of symmetrical palms and soles pain. No evidence of recent infection besides a streptococcal pharyngitidis. The patient describes severe permanent stabbing pain with paroxysmal attacks causing major insomnia. Symptoms are somewhat relieved by regular friction of his hands or feet but mostly by immersion of the extremities in cold water. Family history did not reveal any rheumatic or neurologic diseases. Physical examination showed no sign of oedema or erythema, only slight local warmth and dry skin, no autonomic dysfunction. He had normal physical and neurological examination, except from hyperalgesia and allodynia in some parts of the palms and soles. WBC showed thrombocytosis 450G/l. Erythrocyte sedimentation rate returned normal. Large serological screening, ASLO, heavy metals screening, vitamin analyses, Autoimmune testings and Fabry disease were launched and returned all normal. EM was pointed out even in the absence of erythema (30% of overall cases) because of the pathognomonic feature of the pain amelioration with immersion in cold water. Due to notoriously difficult to treat pain, a multidisciplinary setting group was created. Genetic analyses, particularly SCN9A mutation were added. Doppler flow returned normal. Electromyography testing showed small fibre neuropathy, affecting neurovegetative components. Skin biopsy was done. The patient was given Aspirin for its antiaggregant effect. A transfer was made towards the intensive care unit for analgesia control. He received the listed medication with only partial response and with no negligible side effects: cetirizin, acetaminophen, NSAIDs, continuous morphin switched to fentanyl, associated to ketamin and clonidin, midazolam, Gabapentin switched to Lyrica, lidocain patch, lidocain IV switched to oral mexiletine, nozinan, chloral hydrate. After the skin biopsy, he received methyl-prednisolone and intravenous-immunoglobulin.
Conclusion: EM is a painful and debilitating disease. No single or combination therapy has proved consistently effective, suggesting individual polymorphism. Our patient failed different therapies including a combination with a mainstream medication, mexiletine.

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Postural orthostatic tachycardia syndrome (POTS), an important cause of chronic fatigue in children and adolescents not to be missed

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Introduction: Postural orthostatic tachycardia syndrome (POTS) is an autonomic dysfunction characterized by chronic fatigue, orthostatic intolerance and excessive postural tachycardia. Clinical symptoms are ill-defined: fatigue, headache, dizziness, sleep disturbances, altered mental quality, exercise intolerance, coldness in the extremeties may be observed. Current diagnostic criteria include a heart rate increase of minimum 40 beats per minute, within the first 10 minutes of standing, in the absence of orthostatic hypotension. Full recovery is possible with multi-faceted treatment. We present a patient who was recently diagnosed with that condition at our center.
Case report: This previously healthy 13 year-old girl was examined at our neurological clinic because of chronic headache, hypersomnia and a complaint of "brain fog". Her symptoms had started 6 weeks before after a febrile throat infection. She slept 18 hours a day and felt exhausted by any physical activity. Routine daily activities were no more possible. Her neurological exam was normal. Our initial evaluation and work-up allowed excluding mononucleosis,

hypothyroidism, common intoxications, cerebral tumor, and depression. Cerebral imaging and EEG were normal. A tilt-test showed a postural tachycardia with a sustained increase of a more than 40 beats/min while standing, the signature of POTS.
Conclusion: Adolescent fatigue is a common presenting complaint in teenagers. In many situations, clinical examination and work-up do not reveal any specific condition. Our case reports illustrates the fact that POTS should be included in the differential diagnosis of adolescents who present with chronic fatigue and other ill-defined symptoms, like headache, dizziness, sleep disturbances and altered cognition. To provide targeted treatment is capital to allow these patients to recover quickly. Therapeutic approaches consist of increased fluid and salt intake, regular physical exercise and the wearing of compression stockings. The addition of medications like Fludrocortisone or Beta-Blockers may be helpful if these initial measures fail.

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Pediatric spontaneous pneumomediastinum: literature review and meta-analysis

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Introduction: Described since 1939 in the adult population, spontaneous pneumomediastinum (SPM) is less known in children. Due to its symptomatology and a generally spontaneous benign evolution, it is probably an underestimated diagnosis. However, it has to be considered in the differential diagnosis of acute thoracic pain.
Methods: The incidence being low, we conducted a systematic literature review to identify the circumstances leading to a PMS, the most relevant signs and symptoms, investigations and treatment recommendations.
Results: Of 216 patients, 66.2% are boys and median ages range from 6.9 to 14 years old. The most frequent co-morbidity in children is asthma (22.2%) and the most common trigger factors are bronchospasm (49%), cough (45.6%), various respiratory tract infections, vomiting (10.3%) and foreign body aspiration (8.3%). It remains idiopathic in 33.3%. Relevant signs are chest pain (54.6%), neck pain and/or sore throat (53.3%) and dyspnea (41.2%). The most relevant sign is palpation of subcutaneous emphysema (66.4%). The classically described Hamman's crunch is only present in 11.6%. Chest X-ray provides the right diagnosis in 99.5% of the patients. Pneumothorax is associated in 11.6%. Most patients are hospitalised (88.3%), treatment is based on oxygen therapy, painkillers and rest. In some series, there can be up to 25.8% patients requiring intensive care and 5.5% drainage. Survival rate is 92.5% and long term follow-up shows normal X-rays after 4 days and no recurrence.
Conclusion: SPM is uncommon in children but must be considered in children with acute chest and/or neck pain. History taking, physical examination and standard Chest X-ray are most often diagnostic and there is rarely need for other investigation. Hospitalization is not always indispensable. Outcome is good and follow-up can be clinical, avoiding further X-rays.

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Children exposition to passive smoking: an innovative campaign

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Introduction: The amount of kids exposed to passive smoking and the quantity of smoke they are exposed to are difficult to measure. Only few data exist and there are not so many prevention or awareness project that are focused on this topic. The CIPRET-Valais (Information Center about prevention of tobacco addiction – Valais – Switzerland) has developed a prevention campaign called "Childhood without smoke" (2010-2014). The goal is to reduce the number of children exposed to smoke, also creating a new measuring tool.
Methods: The CIPRET is proposing a simple and effective measuring tool: MoNic Badge, developed by the University Institute of health at work. It enables to take nicotine which kids are exposed to. The amount of nicotine registrated from the badge is translated into the equivalent amount of cigarettes passively inhaled during the exposure period. Parents and professionals were also informed with posters in specific places (parking, shopping center) and with different pictures: baby's bottle full of cigarette butts and an ashtray made of Lego. The poster is asking the question: Is your child is a smoker? Small posters were also proposed in maternities. Nursery nurses have been trained to do short prevention briefings to encourage parents to stop their smoking habits.
Results: With the appropriate media coverage, the population in Valais was made aware of passive smoke at home. In 2013, in Valais, 95% of smokers are maintaining not to be smoking at home in presence of children or teenagers. Many badges were distributed. Then statistics are now available about children exposure to passive smoking.
Conclusion: Evaluations of the campaign are showing the necessity to also focus on the pregnant women. The new campaign is called "Free tobacco Pregnancy" and will start this spring 2015. The main objectives are to inform people about the risks tied to the use of tobacco during pregnancy and to reduce the number of smokers among pregnant women. This campaign will also enable to create reliable statistics, which is the lack of information to date.

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Status asthmaticus – not always what it looks like

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Introduction: Extramedullary leukemic tumor is a rare hematological neoplasm. It's also known as granulocytic sarcoma. It may precede, be simultaneous or appear after the diagnosis of leukemia, mostly acute myelogenous leukemia (AML) in children younger then 15 years old. The localization in the mediastinum without foregoing specific warning signs is very rare.
Case report: A previous healthy 7 year old girl with an unremarkable patient history suffered from acute dyspnea, cough and subfebrile temperatures for two weeks. The symptomatology was interpreted in the context of a viral infection and treated with orally betamethason and inhaled salbutamol. Despite the therapy her condition worsened progressively, she became increasingly dyspnoeic and was referred to the hospital in a status asthmaticus accompanied by an unusual inspiratory stridor and a swollen left arm. The blood gas analysis was unremarkable. Blood counts and CRP were within normal limits, fibrinogen was low, and D-Dimers extremely high. The chest x-ray revealed a broadened upper mediastinum with partial constriction of the trachea. A CT scan unveiled a huge mass in the upper and medium mediastinum compressing half of the tracheal lumen and the vena brachiocephalica. A biopsy of the lesion and a bone marrow aspirate / biopsy confirmed the diagnosis of an AML. Immediate chemotherapy according to the BFM-AML protocol and therapeutic heparinization was started. Her dyspnea and the signs venous obstruction responded within days.
Conclusion: Extramedullary leukemic tumors in AML are rare. Their presentation can be dominated by the anatomical structures affected, without signs or symptoms of a leukemia. If as in the presented case asthma symptoms are accompanied by inspiratory stridor or signs of venous congestion appropriate radiological investigations have to be initiated to rule out an obstructing mass. The combination with a superior vena cava syndrome strongly suggests a malignant pathology.

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Treating a rare cause of laryngotracheitis

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Case report: A 3-months old female infant was recurrently seen in the emergency department due to infect-associated severe episodes of inspiratory stridor. Skin examination revealed hemangiomas at the neck and behind the ear. Therefore, the girl underwent further investigations. On bronchoscopy we found multiple subglottic hemangiomas with a tracheal obstruction of >50%. Treatment was started with a beta-blocking agent (propranolol). At eight months follow up the patient was asymptomatic, laryngoscopy and MRI demonstrated an impressive regression of all lesions. Two years later, the child was presented again with episodes of stridor and cough during upper airways infections and a novel cutaneous hemangioma. At re-evaluation, endoscopy showed no tracheal obstruction. On MRI, no development of new vascular lesions could be seen.
Discussion: It is important to initiate further investigations in a child presenting with stridor and hemangiomas. Administration of propranolol was a successful treatment in our patient with obstructing haemangiomas of the airways. Both invasive surgical procedures and long-term treatment with oral corticosteroids could be avoided. In our young patient, sole treatment with propanolol showed favourable results at follow-up two years after cessation of treatment. Further

studies are needed to determine long-term effect, dosing strategy, treatment duration and side effects of propranolol as a first line treatment of hemangiomas. Furthermore efficacy of propranolol in children presenting with hemangioma beyond the first years of life needs to be investigated.

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Difficult start in life with cystic fibrosis

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Cystic fibrosis (CF) is a genetic disease due to a mutation in the cystic fibrosis transmembrane regulator (CFTR) gene resulting in a dysfunctional mucosal transport of chloride and production of dehydrated and viscous mucus in multiple organs (lungs, pancreas, liver and intestine). Patients suffer mainly from chronic lung disease due to recurrent bacterial infection and failure to thrive due to malabsorption. Different patients may present different degrees of symptoms. Here we present a patient with a complicated postpartal and neonatal course of the disease. Prenatal ultrasound already showed intestinal dilatation suggesting intestinal obstruction. Postnataly the patient presented with meconium ileus that resolved with conservative therapy. After exclusion of Hirschsprungs disease (rectal biopsy) cystic fibrosis was diagnosed by sweat testing (sweat chloride 110 mmol/L, Norm <30 mmol/L). However, newborn screening test showed a normal IRT (immunoreactive trypsinogen) value. Genetic testing revealed a homozygote deltaF508 mutation of the CFTR-gene on chromosome 7. Reduced stool elastase indicated pancreas insufficiency. Although adequate therapies have been initiated (inhalation of hypertonic saline, pancreas enzyme, salt and vitamine substitution) he presented early with increasing jaundice, recurrent respiratory infections, failure to thrive and recurrent intestinal discomfort due to reduced intestinal motility. The prolongated cholestatic jaundice and hepatomegaly could be explained by a inspited bile duct syndrome a known complication of neonatal CF. The origin of the associated splenomegaly with thrombocytopenia (hypersplenism) is still unclear after extensive testing of infectiological, hematological and metabolic differential diagnoses. Failure to thrive could be treated by introducing a novel, alkalinised pancreatic enzyme product and nasogastric feeding. Cholestasis was treated with ursodeoxycholic acid improving hepatic bile flow. Aged four and five months the patient aquired two viral airway infections (RSV positive and RSV negative, respectively), the latter complicated by atelectasis of the right upper lobe associated with pronounced respiratory distress. Despite chest physiotherapy and intensivated inhalational therapy the atelectasis persisted and brochoscopy was performed in suspicion of mucus plugging. Unfortunately a bleeding complication occurred during endoscopy requiring four days of intensive care. Under invasive ventilation the atelectasis resolved. The patient rapidly improved and could be discharged from the hospital. Despite introduction of CF newborn screening and the possibility of early initiation of CF-specific therapy CF is still associated with a significant burden of morbidity in some neonatal patients.

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Croup or not croup, that is the question

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Background: Croup is a common respiratory illness, caused by viruses characterized by a barking cough and various degrees of respiratory distress. Epidemiology of potentially life-threatening upper airway infections is changing actually and viral croup and epiglottitis have been eclipsed by bacterial tracheitis (BT) in children. BT is a bacterial complication of a viral croup which can result in a severe airway obstruction. Differential diagnosis between viral croup and BT remains challenging.
Case report: We describe a case of an 18-month-old previously healthy boy who was admitted to the pediatric emergency unit for a first episode of croup. His immunization status was up-to-date. The clinical picture was characterized by upper respiratory tract infection, low-grade fever since 3 days, coryza, barking cough, hoarse voice and intermittent stridor associated to a moderate respiratory distress. The patient was discharged due to clinical improvement 6 hours after administration of a single dose of dexamethasone and one dose of nebulized epinephrine. One day later the child was readmitted to the hospital due to a deterioration of respiratory symptoms and signs of acute severe respiratory distress. Clinical assessment showed SpO_2 92% on air, respiratory rate 40, temperature 37.4 °C, and pulse 132, respiratory retractions, nasal flaring and stridor at rest. As the administration of five doses of nebulized epinephrine and two doses of intravenous steroids resulted in an only transient improvement the patient was transferred to the pediatric intensive care unit. The patient needed to be intubated; during the procedure a subglottic swelling and thick/ mucupurulent exudates were observed. Invasive ventilation was necessary for three days long. Moreover, during the hospitalization he became febrile up to 39 °C. A diagnosis of BT was clinically evoked, confirmed by the presence of Staphylococcus aureus in endotracheal aspirations and successfully treated by appropriate antibiotics. The patient was discharged from intensive care 7 days later and has since made a full recovery.
Conclusion: BT remains an uncommon cause of upper airway obstruction but should be considered in children who present with acute life-threatening upper way infection or clinical worsening. The varied clinical presentation of BT regarding fever, respiratory symptoms, and general appearance could lead to a misdiagnosis of this clinical condition with a delay in timely optimal treatment and disease management. Our case emphasizes that there is currently a lack of clear guidelines to early differentiate severe croup and life-threatening BT.

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Is sweat-test still useful in cystic fibrosis neonatal screening era?

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Case-report: A 40 day-old girl was hospitalized for bronchiolitis. She was born in Switzerland on term, in 2013, weighing 3.4 kgs, from Albanese Macedonian non consanguineous parents. On her 17th day, she had an electrolytic and acid-base work-up for insufficient weight gaining, as well as control of her Guthrie test, all normal. When admitted for bronchiolitis with feeding difficulties, her weight was 3.8 kgs, with clinical hypotrophy. A respiratory distress with hard coughing rapidly appeared, with oxygen need. Microbiologic tests were negative, including respiratory viruses, Bordetella Pertussis, Mycoplasma and Chlamydophila Pneumoniae. The following 16 days were complicated by left atelectasia, needing for CPAP ventilation, and insufficient gain of weight (10g/day) despite enteral feeding. A sweat-test was made with Nanoduct device, showing 120 mmol/l of Cl. Fecal elastase was low. Her genetic status was homozygosity for F508del, confirming cystic fibrosis (CF). Result of her Guthrie was retrospectively obtained: Immunoreactive Trypsin (IRT) was 49 ng/ml, while screening threshold is 50.
Discussion: CF Neonatal screening with Guthrie IRT was introduced in Switzerland in 2011 with a pilot study, and definitely implemented in 2013, more than 30 years after the first national screening study in Australia and New Zealand. Enough evidence exists now, proving the lower CF morbidity in countries with this program. IRT threshold was percentile 99.2 of the screened population, corresponding to 50 ng/ml in Switzerland. The more usual 7 mutations were studied in children screened and in those with meconial ileus. Five CF children were missed up to march 2014, IRT being normal at screening. Eighteen mutations are now included in the genetic screening panel. False negative will persist, and CF diagnosis must be kept in mind when clinical arguments are present.
Conclusion: Unusual coughing or pneumonia, failure to thrive as well as meconial ileus remain solid clinical arguments to control sweat-test, even in CF screening era.

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High flow nasal cannula (HFNC) therapy for children with brobchiolitis: an observational study

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Bronchiolitis is a common lower respiratory tract infection but effective therapy is lacking. The traditional treatment consists of supportive therapy with fluids, supplemental O_2 and an adequate respiratory support. The conventional oxygen delivery is a dry, not heated gas, at 100% concentration via low-flow nasal cannula or face mask. An emerging method to support breathing is high flow therapy HFNC. The device allows to modify the concentration of oxygen and the temperature of the gas. The physiological effects of high flows include pharyngeal dead space washout, decreased nasopharyngeal resistance leading to improved pulmonary compliance and decreased work of breathing. A positive end expiratory pressure (PEEP) effect may be generated. HFNC is a less invasive than conventional nasal

CPAP, with a lesser incidence of pneumothorax. This kind of respiratory support increases the CO_2 wash-out, improves the mucociliary clearance and gives a minimal and not constant PEEP. Since September 2014 we have treated 7 patients with moderate/severe bronchiolitis with HFNC. The clinical data are still lacking for a statistical analysis, but our impression is positive. We didn't transfer any patient to a pediatric intensive care unit, and no one of the patients had pneumothorax or other side effects. HFNC appear to be feasible and well tolerated. For these reasons we have started a prospective study to better evaluate the real efficacy and the possible side effects of HFNC in pediatric patients with moderate/severe bronchiolitis. Inclusion criteria: pediatric patients with bronchiolitis and respiratory acidosis (pH <7.35); CO_2 increase (>6 kPa); hypoxemia and moderate to severe respiratory distress despite standard flow oxygen; mild/severe respiratory distress; recurrent apnea. Starting therapy: we start the HFNC system at 1 L per kg per minute and we go up to 2 L per kg based on work of breathing. We adjust oxygen fraction via blender to maintain target oxygen saturation (94–98%). Weaning of high flow nasal cannula therapy when clinical condition is improving as indicated by: decreased work of breathing; normal or improved respiratory rate. Once child is stable and in FiO_2 of 40%, standard nasal cannula oxygen can be implemented. Actually only one randomized and some observational studies point out the positive effects of HFNC in infants with bronchiolitis; they provide some indication that HNFC is feasible and well tolerated. So this device could be a new inexpensive and well tolerated treatment for moderate to severe bronchiolitis.

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Changes in breathing pattern upon pure oxygen during multiple breath washout in children

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Background: Nitrogen multiple breath washout (N_2MBW) with 100% oxygen (O_2) is frequently used as diagnostic tool for abnormal lung function. However, it has been shown that the use of pure O_2 can influence breathing pattern in infants. It is unknown whether O_2 influences breathing pattern in older children. We systematically compared normal breathing pattern (ambient air) and possible changes due to 100% O_2 administration.
Methods: We performed one tidal breathing and two N_2MBW measurements in former preterm children and healthy controls using Exhalyzer D (Spiroware, Eco Medics) according to ERS/ATS consensus. The first 10 breaths of tidal breathing measurements were excluded for analysis. We compared tidal volume (VT) and coefficient of variation of V_T (CV_{VT}) between tidal breathing in room air vs. O_2-exposure during N_2MBW by paired t-test. Sensitivity analysis was performed by comparing the effect of different numbers of breaths (8, 10, and 12).
Results: We measured 40 children (24 = healthy; 16 = former preterm) aged mean (sd) 6.8 (0.9) years. There was no difference in VT or in CV_{VT} between tidal breathing in room air compared to breathing during the first or second N_2MBW. These findings were consistent in both preterm and term children and confirmed by sensitivity analysis. There was no difference in the lung clearance index (LCI) between the first and the second O_2-exposure.
Conclusion: We did not find any difference in VT after administration of 100% O_2 in healthy and former preterm children. CV_{VT} did not differ between measurements but was generally high in this age group (mean: 25.8%). The known effect of 100% O_2 in infancy seems to be outgrown at early school age.

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Ability of lung clearance index to track changes in a routine clinical CF setting

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Background: Lung clearance index (LCI) has shown to be a sensitive outcome parameter in children with cystic fibrosis (CF) for specific treatment interventions. Whether this holds true for a non-study setting is unclear.
Aims: We assessed the sensitivity of LCI to track lung function decline as well as improvement in children with CF in clinical routine without specific interventions.
Methods: We retrospectively analyzed multiple-breath washout measured in a clinical routine setting between 2011 and 2014 in 44 children with ≥3 test sessions at least one month apart. We selected children with a LCI deterioration of >10% on two consecutive visits within 12 months and assessed how many of those showed a subsequent LCI improvement of >10% within 12 months. LCI was compared to spirometry (FEV1).
Results: Out of 44 children (mean (SD) age 11.4 (2.8) years), 31 children showed a LCI deterioration of >10% from mean (SD) z-scores of 5.4 (3.9) to 10.4 (4.7) compared to –1.0 (1.2) to –2.0 (1.1) FEV1 z-scores. Out of those 31 children 16 showed a subsequent LCI improvement of >10% to 4.5 (3.3) z-scores, compared to a FEV1 improvement to –0.8 (1.1) z-scores. 10 out of 16 children with >10% LCI improvement showed a parallel FEV1 improvement of >10%. A cut-off of >20% LCI change as inclusion criteria confirmed the higher sensitivity of LCI compared to FEV1.
Conclusions: Not only in interventional trials but also in clinical routine measurement of children with CF, LCI seems to be more sensitive than FEV1 to assess lung function changes; both for deterioration and improvement. LCI and spirometry provide complementary functional information in children with CF.

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Abnormal lung function in infants with cystic fibrosis shortly after birth

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It is unknown at what age small airway disease starts to evolve in children with cystic fibrosis (CF). We assessed ventilation heterogeneity shortly after birth in infants with CF diagnosed by newborn screening. We performed multiple-breath washout (MBW) measurements in 43 infants with CF (median (95%-CI) age 7.3 (6.5–8.0) weeks) and in 46 weight-matched healthy controls (age 5.2 (5.0–5.3) weeks). We assessed lung clearance index (LCI), functional residual capacity (FRC) and tidal breathing parameters. LCI was elevated in infants with CF (mean difference (95% CI) 0.9 (0.3 to 1.5), p = 0.0025), as was FRC (mean difference (95% CI) 17 (9 to 25) mL, p <0.0001). In 21 out of 43 infants with CF either FRC or LCI were above the upper limit of normality, seven out of these showed elevated values for both FRC and LCI. Tidal breathing parameters were all normal in CF infants. Almost half of CF infants show increased ventilation inhomogeneity and hyperinflation shortly after birth. This highlights the usefulness of non-invasive lung function measurement as outcome marker and the importance of early therapy in infants with CF.

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Sleep onset transition: insights from functional connectivity analyses

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Introduction: Despite the advances in our understanding of sleep in recent years, many questions regarding the process of falling asleep remain unresolved. Principally, we aim to answer the basic question: are there characteristic brain oscillations in different frequency bands as hallmarks of the sleep onset process? If so, can we describe them analytically, by using mathematical methods? For this purpose, we determined functional connectivity between cortical brain areas, which may be determinant assessing neuroelectrical changes underlying the sleep onset process. In a broad sense, functional connectivity refers to correlations or statistical dependences between neuronal activity of different brain regions.
Methods: EEG data (27 derivations) collected in 8 healthy young adults during baseline and recovery nights (40 h of sustained wakefulness) was analyzed with 3 different methods: mutual information, normalized Shannon entropy and phase-locking index. We aim to detect relevant non-linear changes of EEG brain activity, by comparing functional connectivity variables ("synchronization") before and after sleep onset (defined as first occurrence of stage 2).

Results: We have observed an increase of synchronization (i.e., phase-locking and amplitude correlations) in low frequency bands up to approx. 15 Hz (in particular between frontal and occipital areas) preceding the sleep onset transition. The three functional connectivity methods revealed similar results. The changes are particularly significant for the high delta and theta bands. In contrast, after the transition, synchronization in the sigma band (associated with spindles) reaches significance. A gradual decay of the number of significant connections between pairs of derivations in the course of sleep onset has been discovered, especially in theta band. Increased sleep pressure resulted in faster or steeper changes. Moreover, the results were to some degree depending on the reference applied.
Conclusion: We think our results could relate the hypothesized breakdown of effective connectivity explaining the fading of consciousness during sleep onset (Massimini et al., 2008). Using functional connectivity methods to address neuronal network properties may reveal new insights.

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The impact of infectious diseases on sleeping behaviour in preterm infants

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Hypothesis: One of the main reasons for preterm birth is perinatal infection. Preterm infants are very vulnerable and therefore susceptible to contagious diseases. Infection may have an influence on the wake-sleep regulation and the length of the sleep phases. The hypotheses of this study were that the sleep duration is longer and that sleep phases are altered in infected compared to non-infected preterms.
Methods: 40 preterm infants (20 girls, 20 boys, less than 32 weeks gestational age [mean: 29/1, range: 24/0 to 37/3] and/or less than 1500 g birth weight [mean: 1119 g, range: 420 g to 1650 g]) were video-recorded for an average duration of three hours on several days within the first week after birth and once every following week until they left the incubator. The 277 resulting recordings were scored every ten seconds differentiating between wakefulness (W), active sleep (AS) and quiet sleep (QS). The total time of each state and the mean duration of a sleep cycle were calculated for every measurement. We performed multilevel linear regression analysis to estimate the association of infection and sleep architecture. To comprehend a recording infected we considered proven or highly suspected infections of the infant and for all the first-week recording also signs for maternal infection and the main reason for preterm-birth. This made 63 infected and 214 non-infected measurements.
Results: There was a positive association between infection and the duration of sleep cycles ($p = 0.004$), the time asleep ($p < 0.001$) and the time in quiet sleep ($p = 0.025$). Furthermore male participants showed a trend ($p = 0.08$) towards being less awake and spending more time in active sleep ($p = 0.001$) than their female counterparts.
Conclusion: In very premature babies, the infectious state significantly affects sleep-wake regulation and the duration of sleep phases. Infected preterm infants generally sleep more, spend more time in quiet sleep and therefore have longer sleeping cycles.

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Role of the family doctor in the care of sleep disorders in young people

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Introduction: There are few studies on the prevalence of sleep disorders in young people: they show that sleep disorders are common in this age group. We found no data regarding young people consulting family doctors. It is important to identify sleep disorders since they can be both a cause and a consequence of other health problems. Family doctors (general internists and pediatricians) are the ideal actors of the health care system to address sleep disorders in this population since most young people see a family doctor at least once a year. The goals of this study were to identify the type and frequency of sleep disorders in young people consulting a family doctor, the perception of sleep disorders by the patients, and the methods used by family doctors to investigate sleep disorders.
Methods: Cross-sectional study involving family doctors and their patients in practices in the Canton of Geneva, Switzerland. The doctors were invited to fill out an online survey on sleep disorders occurring in their patients between 15 and 24 years old. Young people between the ages of 15 and 24 years old consulting these doctors for any motive were invited to complete a confidential survey regarding their sleeping habits and their expectations towards their family doctor regarding possible sleep disorders. These surveys were collected over one month between November 2014 and February 2015.
Results: 10 family doctors and 33 patients between 15 and 24 were included in the study. The results are being analysed and will be available in June 2015.
Conclusions: This study gives us a first look into the expectations of young people towards their family doctor regarding sleep disorders and the answers family doctors can offer them in response to their needs.

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Sleep wave analysis: a toolbox for the detection and analysis of waves in sleep

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Over the past two decades, the focus of much sleep research has shifted from the temporal dynamics of sleep to the spatio-temporal aspects with the advance of high-density electroencephalography (EEG) and novel experimental paradigms. The most common approach has been to use frequency based analysis to detect “slow wave power”. However, this is generally just a proxy measure of various properties of the individual slow waves found in deeper stages of sleep, and the accurate detection and exploration of these individual waves would provide much greater insight and specificity into the local mechanisms under investigation. Here we present a user-friendly, open-source, toolbox designed for the analysis of high-density EEG recordings in sleep. The toolbox›s main aim is to provide the user with a fast and accurate tool to detect the various waves found in these recordings, such as individual slow waves, sleep spindles and saw-tooth waves. Although default settings for all measurement paradigms are given, the user is able to freely change the parameters and explore the results using a wide range of visualisation options available. We examine the typical workflow from the preparation of raw data recordings, to publication ready measures, including how the toolbox can be used in the scoring sleep stages, as well as the available parameter settings and their expected effects on a sample of 11 control participant›s sleep recordings. In the analysis parameter settings such as choice of EEG reference, amplitude threshold, and channel correlation are examined for their effect on several outcome measures such as basic measures of mean amplitude and wave density to more detailed measures of wave globality, and traveling properties of the individually detected waves. The principle conclusions are that even minor changes in parameter settings can have large subsequential effects on the number and properties of the waves ultimately detected, and as such there is a need to standardise the detection methods, and provide clear justifications for any chosen settings. The toolbox is easy to use, is consistently updated with new features, visualisation options and algorithm optimisations, as well as being designed specifically with potential collaborations in mind. The toolbox can be freely downloaded from https://github.com/Mensen/swa-matlab.

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Resting State Networks in Subjects under Low and High Sleep Pressure

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Introduction: Recently, studies investigating resting state networks (RSN) have increased and typically identified 10 RSN, with a focus on the default mode network (DMN). Sleep studies reported sleep stage dependency of DMN connectivity and sleep pressure dependent changes of intra-network connectivity. Few studies examined RSN with ASL (absolute measure of cerebral blood flow; CBF). Thus, we analysed RSN of combined BOLD/CBF data, from subjects under low and high sleep pressure.
Methods: Eyes open data from 18 healthy adults recorded after ~8–9 h of wakefulness (low sleep pressure) and 18 healthy male adults after ~18.5 h of prolonged wakefulness (high sleep pressure) was analysed. BOLD resting state was measured (2x2 min eyes open), followed approx. 1 h later by ASL (5 min eyes open). Data were realigned and CBF images quantified. BOLD and CBF images were co-registered to individuals' anatomical T1 images, normalized, and smoothed. RSN analysis of combined BOLD/CBF data was performed with the GIFT toolbox (20 independent components; ICs), functional connectivity (FC) with the MANCOVAN toolbox.
Results: Eighteen ICs were identified that highly correlated with RSN ICs previously described in [1]. Spatial maps did not differ between groups. However, the basal ganglia network was less strongly connected with a visual IC under normal sleep pressure ($p < 0.01$, FDR corrected).
Conclusion: Increased sleep pressure seems to change the FC between networks rather than to alter their spatial extent (which is to be expected in subjects of similar age/health). As the basal ganglia are involved in sleep-wake regulation [2], the increase in FC between the basal ganglia network and the visual IC in subjects with higher sleep pressure might reflect their struggle to maintain wakefulness.

1 Allen et al. Front. Syst Neurosci. 2011.
2 Lazarus et al. Curr Opin Neurobiol. 2013.

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Sleep regulation: effect of sleep restriction and extension

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Introduction: Mechanisms of sleep homeostasis regulate sleep intensity: if sleep is restricted, then sleep intensity increases, and in case of excess sleep, sleep intensity is decreased. However, some studies reported only an initial increase in slow-wave activity (SWA) during chronic sleep restriction, and then SWA values are maintained at or even below baseline levels. Our goal was to investigate sleep regulation in humans undergoing sleep restriction and extension.
Methods: Data of 25 subjects with polysomnographic recordings of two sessions (restriction and extension condition) were analyzed. Each session consisted of 9 nights: baseline night (8 hours of sleep), 7 condition nights (6 or 10 hours of sleep), and a recovery night (12 hours of sleep) after ~40 hours of sleep deprivation. Power density spectra and slow-wave activity (SWA; power in 0.75–4.5 Hz range) were calculated. Slow-wave energy (SWE) reflects the total dissipation of sleep pressure during sleep, and was determined as cumulative sum of SWA in 30-min bins across the night.
Results: SWE in the 1st extension night was 6 % higher than in baseline, reached baseline levels in the 3rd and 4th night of extension and decreased to 92 % of baseline levels thereafter. SWE in the first restriction night was at 92 % of baseline levels, reached baseline levels in the 3rd and 4th night and decreased below baseline in restriction nights 5–7. Sleep deprivation resulted in a similar increase of SWE in both restriction and extension conditions.
Conclusion: Sleep deprivation led to an equal homeostatic response after both sleep extension and restriction. The observed time course during extension and restriction needs further investigations. However, inter-individual differences were large and not all individuals may show the same response to extension and restriction.

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Memory cueing during sleep modifies the interpretation of ambiguous scenes

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The individual tendency to interpret the numerous situations in life that are ambiguous depends on our previous experiences with similar situations. A negative interpretation bias is linked to affective disorders with these often starting to emerge early in life. Here, children and adolescents learnt associations between ambiguous situations and positive or negative words defining the situations' outcome. We examined whether cueing these associations during post-learning sleep can modulate the interpretation of new ambiguous situations that are similar in content to the previously seen ambiguous situations. Cued, as compared to un-cued, picture-word associations were remembered better the next morning, and successful cueing was coincided by pronounced oscillatory theta power over central and parietal regions. Cued, but not un-cued, positive associations led to a more positive interpretation of similar ambiguous situations. Cueing during sleep might strengthen positive interpretations of everyday-life situations, with implications for intervention development of inventions for affective disorders.

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Local slow wave deprivation during sleep by auditory time-looked stimulation based on real time slow wave detection

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Sleep slow waves (EEG frequency 1–4.25 Hz), the hallmarks of deep sleep in the EEG, are causally involved in sleep recovery processes important for cortical functioning. A recent study shows that slow wave sleep can be manipulated by auditory stimuli, which are applied time-locked to the ongoing slow waves. Our goal was to locally deprive slow waves, by playing tones precisely time-locked to the sleep slow waves of one selected brain region. During 2 counterbalanced nights, high density (128 electrodes) sleep EEG was recorded in 7 healthy young men (24.5 ± 0.7 years). During one night (stimulation night), automatic time-locked (i.e., in-phase) auditory stimulation was performed to locally deprive slow waves. Slow waves were detected in real time based on the signal of the electrode P4 (right parietal cortex), and tones (50 ms duration, ~50 dB) were applied during a specific phase of the parietal oscillation to interfere with the ongoing sleep slow waves. With this approach we aimed to disturb sleep slow waves selectively over the right parietal cortex without interfering with global structure of sleep. No difference in sleep architecture was found between the two experimental nights. Under both conditions subject slept well with high sleep efficiency (stimulation night: ~89.1%; sham night: ~91.2%). When comparing the 2 nights, slow wave activity (SWA, power in the slow wave frequency range) was reduced under stimulation by ~16.2 ± 11.5% in a local cluster of 9 electrodes over the right parietal cortex ($p = 0.019$; SWA stimulation: 31.6 ± 9.9 μV^2/Hz, baseline: 37.2 ± 9.8 μV^2/Hz). However, global SWA (mean over all electrode) did not differ between the two nights ($p = 0.35$). Our results show that deep sleep can be manipulated locally by the application of time-locked tones without disturbing the global structure of sleep. Such real-time in-phase manipulation provides a straight-forward tool to interfere with ongoing brain oscillation of distinct brain regions during sleep. Whether such a local disturbance of slow wave sleep also impacts the brain on a functional level needs to be examined in the future.

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The density of sleep spindles is reduced in early onset schizophrenia patients

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Introduction: Schizophrenia is a complex neuropsychiatric disorder with a typical onset during late adolescence. Abnormal brain connectivity, specifically, thalamocortical deficits have been reported in adult schizophrenia. Sleep spindles, thalamocortically generated phasic oscillations between 12–15 Hz during non-rapid eye movement (NREM) sleep, reflects the integrity of the thalamocortical system. Adult patients with schizophrenia show a decrease in sleep spindles. Furthermore, these spindle deficits are associated with a greater severity of positive symptoms. The focus of our study was on investigating sleep spindles in adolescents at an early stage of the disease by means of high density electroencephalogram (EEG) and relating alterations in sleep spindles to symptom severity.
Methods: All-night high-density EEG was recorded in 9 patients (16.1 ± 0.5y) and 9 controls (16.2 ± 0.5y). Study inclusion for the patients required a diagnosis of schizophrenia, schizophreniform disorder or a brief psychotic disorder according to DSM-IV. Actual symptom severity was assessed using the Positive and Negative Syndrome Scale (PANSS). Sleep stages were visually scored. Automatic sleep spindle detection was performed. We focussed on the first hour of artefact free NREM sleep.
Results: Similarly to adult patients, our early onset schizophrenia patients showed a global reduction of spindle density. Electrodes over centro-parietal and temporal brain areas reached significance. Furthermore we found a negative association between spindle density and severity of positive symptoms ($r = –0.75$, $p = 0.02$). No such association was found for negative symptoms.
Conclusion: Our findings indicate that sleep spindle deficits can already be detected in early onset schizophrenia, are associated with positive symptoms and may therefore be an electrophysiological marker of the disorder.

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Global field synchronization during sleep and waking

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Introduction: Sleep is characterized by a reduction of responsiveness and a loss of consciousness, which have been attributed to a breakdown of functional connectivity between brain regions. A method to estimate functional connectivity of brain processes is global field synchronization (GFS; Koenig et al., 2001). The GFS is a frequency-dependent measure of the global synchronicity of multi-channel EEG data. Our aim was to explore and extend the hypothesis of functional disconnection during sleep by comparing GFS spectra of different vigilance states (i.e., NREM and REM sleep and waking).
Methods: The analysis was performed on an existing data set of eight healthy male subjects (21–25 years). Twenty-seven EEG derivations were recorded during a baseline night, a recovery night after 40 h of sustained wakefulness and at 3-h intervals during the 40 h of wakefulness. Sleep stages were scored according to standard criteria. For consecutive 4-s epochs, GFS spectra were computed based on the complex spectrum (FFT) yielding a complex value for each frequency bin and derivation.
Results: Compared to NREM sleep, REM sleep showed generally larger GFS values in all frequencies except in the spindle frequency range, where NREM sleep showed a peak in GFS, both for baseline as well as for the recovery sleep. Sleep deprivation did not affect GFS spectra in REM and NREM sleep. The GFS values of the wake EEG were generally lower compared to REM and NREM sleep except for the alpha frequency band. The GFS alpha peak decreased as a function of time awake and was more prominent with eyes closed than with eyes open.
Conclusion: Our somewhat surprising finding of higher synchrony during REM sleep challenges the view of REM sleep as a desynchronized brain state and may provide insight into the function of REM sleep.

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Genetic variation in dopaminergic signaling and the consequences of sleep loss in humans

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Introduction: The distinct roles of dopaminergic neurotransmission in sleep–wake regulation are not conclusively established. Here we investigated the possible influences of functional genetic variations in the genes encoding the dopamine transporter (*DAT1*; rs6277) and the dopamine D_2 receptor (*DRD2*; rs28363170) on the effects of sleep loss in healthy volunteers.
Methods: Psychomotor vigilance task (PVT, n = 64) performance, waking EEG (n = 80) and subjective sleepiness (SSS, n = 80) were quantified at 3-hour intervals during a 40-hour sleep deprivation protocol. *DAT1* (variable-number-tandem-repeat polymorphism: 9R/9R, 9R/10R and 10R/10R allele carriers) and DRD2 genotypes (C/C, C/T or T/T allele carriers) were retrospectively determined. The effects of genotype, test-session (time of day), time-on-task (in quintiles) and day (before and after sleep loss) were analyzed by mixed-model ANOVAs.
Results: Prolonged wakefulness was associated with increased subjective sleepiness, enhanced EEG theta/alpha ratio, greater number of PVT lapses, reduced PVT speed (1/median reaction times) and decreased global alertness (variance in PVT speed) ('day' $p_{all} < 0.0001$). Both, *DAT1* and *DRD2* genotypes modulated the impact of sleep deprivation on sleepiness, PVT lapses, PVT response speed, and the EEG theta/alpha ratio ('day' x 'genotypes', $p_{all} < 0.02$). By contrast, global alertness revealed no genotype dependent modulation ('day' x 'genotypes', $p > 0.9$).
Conclusions: The findings are consistent with an inverted U-shaped relationship between genetically determined differences in dopaminergic signaling and the individual response to sleep loss. Because both *DAT1* and *DRD2* are mainly expressed in the striatum, the data suggest that striatal dopaminergic neurotransmission may be an important predictor of the response to prolonged wakefulness.

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Effects of DAT1 genotype and tolcapone on wrist activity during sleep deprivation

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Background: Cerebral dopamine (DA) plays a role in regulating rest-activity patterns. The DA active transporter (DAT) regulates DA levels in the striatum. The gene encoding DAT (referred to as *DAT1*) exhibits a functional polymorphism that has previously been associated with variable response to sleep deprivation (SD) and different wrist activity levels. The catechol-O-methyltransferase inhibitor, tolcapone, reduces DA metabolism in the prefrontal cortex. We here investigated the effects of SD and tolcapone on wrist actigraphy in healthy young men stratified by *DAT1* genotype.
Methods: Wrist activity was recorded in 12 homozygous carriers of a 10-repeat (10R/10R) allele of the 3'-variable-number-tandem-repeat polymorphism of *DAT1* and 14 9-repeat (9R: 10R/9R + 9R/9R genotypes) allele carriers completing two SD sessions of 40 hours of extended wakefulness. Either tolcapone (2 x 100 mg) or placebo was administered in randomized, double-blind, cross-over fashion after 11 and 23 h of wakefulness. Activity averaged over 15-minute bins, as well as over baseline and sleep-deprivation days, was analyzed with mixed-model repeated measure ANOVAs.
Results: The two genotype groups did not differ with respect to age, body-mass-index and daytime sleepiness. While SD and *DAT1* genotype did not affect wrist activity, a main effect of treatment was observed, such that activity was higher with tolcapone than with placebo. Analysis by 15-minute bins further revealed a *"treatment" × "genotype"* interaction, such that there was an effect of treatment in the 9R-allele carriers but not in 10R/10R allele homozygotes.

Discussion: Our controlled laboratory study revealed no significant differences in wrist activity between *DAT1* genotypes. The results of the pharmacogenetic intervention with tolcapone may indicate that increased DA neurotransmission increases wrist activity levels. The actimetric examination of motor activity in recovery sleep after SD will help to further elucidate the influences of *DAT1* genotype and tolcapone on sleep-wake quality.

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Non-visual impact of light on neurobehavioral performance and circadian physiology depends on inter-individual differences

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Introduction: A polymorphism in the clock gene *PER3* has been shown to be related to neurobehavioral and endocrinological responses to sleep loss and light exposure. More specifically, homozygous carriers of the long allele repeat (*PER3$^{5/5}$*) are assumed to be more vulnerable to the detrimental effects of sleep loss and are more sensitive to blue-enriched light when applied in the evening than *PER3$^{4/4}$* carriers. In this study we investigated whether an extended light exposure of moderately bright light at 250 lux for 40 hours can counteract a potential vulnerability to sleep loss in study volunteers carrying the PER35/5 when compared to *PER3$^{4/4}$* carriers.
Methods: Eight (6 men/ 2 women) homozygous long allele carriers (*PER3$^{5/5}$*: mean ± SE: 25.37 ± 1.10y) and eight (6 men/ 2 women) homozygous short allele carriers (PER34/4: mean ± SE: 23.62 ± 0.42 y) underwent 40-hours of extended wakefulness once under dim light (DL: 8 lux), and once under either white light (WL: 250 lux) or blue-enriched white light (BL: 250 lux) exposure. Subjective sleepiness and well-being was assessed hourly along with melatonin and cortisol assays. A cognitive test battery was performed every 2.5-h.
Results: Melatonin was significantly suppressed under both light conditions and in both genotypes, but with a significantly more pronounced effect under BL in the *PER3$^{5/5}$*. However, moderately bright light during 40 hours of sustained wakefulness induced a significant alerting response, better well-being and a decrease in cortisol levels only in the short allele carriers (*PER3$^{4/4}$*) under both light conditions whereas no effect was found under both light in the *PER3$^{5/5}$*. In contrast, the WL significantly decreased cognitive performance in the *PER3$^{5/5}$* while it remained constant in *PER3$^{4/4}$*.
Conclusion: We have evidence for a clock gene genotype-related modulation of the non-visual response to light at 250 lux under extended wakefulness. However, a long term light exposure was not sufficient to counteract the assumed vulnerability to sleep loss from the *PER3$^{5/5}$* carriers. In fact, we have evidence that such extended light exposure duration rather enhanced the effect in the less vulnerable genotype (*PER3$^{4/4}$*).

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Altered Topography of Sleep Spindle Activity in Very Preterm Children and Adolescents

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Background: The majority of very preterm infants survive without any severe neurodevelopmental impairments, however, up to 50% of all very preterm children suffer from subtle cognitive deficits and learning problems. Sleep spindles, thalamo-cortically generated phasic oscillations between 12 and 15 Hz during non-rapid eye movement (NREM) sleep, reflect the integrity of the thalamocortical system and have been reported to be associated with measures of IQ and learning. The current study aimed to investigate potential alterations in the topographical distribution of sleep spindle activity in very preterm children and adolescents.
Methods: Thirty-eight very preterm participants (29.5 ± 2.1 [M ± SD] weeks of gestation) and forty-three healthy term-born peers were assessed at a mean age of 13.1 ± 1.9 years. All-night high-density sleep EEG (128 electrodes) was recorded. Power maps were calculated based on the average spindle activity of the first hour of NREM sleep.
Results: Sleep efficiency was high in both groups (approximately 90%). The duration and architecture of sleep were not significantly different between the groups ($p > .40$). In a widespred temporal/parietal cluster of 23 electrodes, very preterm participants exhibited significantly less spindle activity than their term-born peers (decrease of 10.9 ± 2.4%, $p < .001$). Also, in a smaller cluster of five frontal electrodes, they exhibited significantly more spindle activity (increase of 14.3 ± 0.7%, $p = .02$).
Conclusion: The altered topographical distribution of sleep spindle activity in very preterm children may reflect alterations in thalamocortical connectivity and may be associated with the poorer general cognitive abilities and learning problems often described in this population. Interestingly, the widespread reduction in spindle activity in very preterm children was observed over brain areas involved in spatial information processing, language and calculation, cognitive domains which are frequently impaired in these children. The local increase over frontal brain areas may result from increased effort in frontal lobe functioning (i.e., executive functions) in very preterm children as increased spindle activity has been reported after learning. In the future, high-density sleep EEG may serve as a tool to better understand neuronal mechanisms underlying cognitive impairments in very preterm children and adolescents.

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Lifestyle considerations in type 2 diabetes risk: sleep quality, sleep timing and habitual caffeine consumption

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Background: Controlled laboratory experiments and large-scale epidemiological studies suggest that short, low-quality sleep, or circadian misalignment, may increase the risk of developing type-2 diabetes (T2D). For caffeine, there is a paradox in the literature: long-term high coffee consumption may reduce T2D risk; yet, laboratory experiments in healthy and T2D individuals indicate that caffeine impairs glucose homeostasis. Caffeine also negatively affects sleep and can be used as a model of insomnia. We aimed to investigate potential roles for these factors in T2D risk.
Methods: In a case-control field study, 134 T2D patients (63% male; aged 64.1 ± 9.7 years) and 230 non-diabetic (ND) participants (37% male; 63.8 ± 9.9 years) of European descent completed questionnaires regarding demographic and health information; subjective sleep quality (PSQI); sleep timing and chronotype (MCTQ); the Epworth Sleepiness Scale (ESS); and habitual caffeine consumption. Participants gave two samples of saliva for 1) quantification of caffeine in saliva by HPLC and 2) genotyping for SNPs related to caffeine metabolism and sleep.
Results: Descriptive and non-parametric statistics suggest that self-reported subjective sleep quality, sleep duration, chronotype and social jetlag ($p_{all}>0.05$) are similar between T2Ds and NDs. During the day, T2Ds report napping for longer and feeling sleepier ($p_{all}<0.003$). T2Ds report consuming more caffeine than NDs (347 vs. 261mg per day); this higher caffeine intake stems from 82mg more coffee-based caffeine ($p_{all}<0.001$), equivalent to an extra 1–2 cups per day.
Conclusions: To draw firm conclusions from the questionnaire data, further analysis is required. For example, transformation of data and completion of parametric statistics that control for demographic differences between the T2Ds and NDs.

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Effect of alveolar hypoventilation on nighttime blood glucose in obese patients with obstructive sleep apnoea

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Background: Obstructive sleep apnea (OSA) has been associated with alterations in glucose metabolism in obese individuals. However, it is not known whether nighttime alveolar hypoventilation (aHyp) in addition to OSA has an additive impairing effect on glucose metabolism.

Methods: We performed a case-control study in 98 patients with OSA and a body mass index (BMI) >30 kg/m². Bicabonate >27 mmol/l in capillary blood gas analysis was chosen to indicate aHyp. All subjects underwent polysomnography and we determined nighttime blood glucose (mg/dl) by capillary blood gas analysis at 10.00 PM, 01.00 AM and 05.00 AM. Differences in blood glucose during nighttime between subjects with and without aHyp were assessed by calculating the area under the curve (AUC) from blood glucose measurements.
Results: 22 of the 98 patients with moderate to severe OSA had aHyp. Patient characteristics of the two groups are shown in the table. Blood glucose levels were significantly higher in patients with additional aHyp compared to patients with OSA only (OSA: AUC 789.0 (694.0/902.2); aHyp: AUC 902.5 (811.6/1056.6); p = 0.001). In a multivariate linear regression model, aHyp (B = 201.360; 95%CI: 27.25/375.46; p = 0.024) was independently associated with increased nighttime blood glucose levels corrected for possible confounders such as duration of REM-sleep, glucose lowering medication, BMI, heart rate during sleep, arousal index and mean oxygen saturation.
Conclusions: Our findings suggest that aHyp may be an additive risk factor for impaired glycemic control in obese patients with moderate to severe OSA. Controlled interventional trials are needed to clarify the clinical significance of aHyp on glucose control.

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Atrial premature beats during the Mueller manoeuvre are a predictor for arrhythmias

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Introduction: Obstructive sleep apnoea (OSA) is associated with arrhythmias such as premature atrial beats (PABs) which can lead to paroxysmal atrial fibrillation (PAF) and sudden cardiac death. The Mueller manoeuvre (MM) generates negative intrathoracic pressures similar to the ones observed during an obstructive apnoea. We tested the hypothesis whether PABs induced during a MM are associated with PABs recorded during a 24h-ECG and thus has the potential to be an indicator for the vulnerability of the heart.
Methods: 12-lead-electrocardiograms were recorded continuously in 40 patients with paroxysmal atrial fibrillation during a MM. All patients underwent nocturnal respiratory polygraphic measurements and 24h-ECG. Associations were determined using linear regression adjusted for potential confounders.
Results: According to the sleep study, 50% of patients (age: 60 ± 10 years, 5 females) had OSA (apnoea hypopnoea index ≥5). Multivariable regression analysis showed that APBs during the MM were associated with the number of PABs in the 24h-ECG (p <0.001) independent of age, sex and body mass index. Obstructive apnoeas during the sleep study were more often observed in patients with PABs during the MM when compared with patients without PABs during the MM (p = 0.013) independent of age, sex and body mass index.
Conclusion: PABs during the Mueller Manoeuvre seem to be a predictor for the spontaneous occurrence of PABs during night- and daytime. Patients with OSA seem to be especially vulnerable to cardiac arrhythmias such as PABs.

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Effects of age, sleep deprivation and caffeine on random number generation

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Introduction: The random number generation task (RNG) requires the allocation of central executive resources and is a powerful tool to assess the connection between working memory and brain executive functions. Here we investigated whether advanced age and sleep deprivation, both known to impair executive functions, impact on RNG performance.
Methods: 193 healthy volunteers (age range: 17–92 years) were asked to produce at the pace of 1 Hz a sequence of 66 digits in a random fashion (response alternatives: 1–6). Deviations from randomness were quantified by established RNG variables (redundancy, RNG index, null-score quotient, adjacency, phi2 index) [1]. In a separate group of 11 older men (age range: 62–75 years), the effects of one night without sleep and 2 x 200 mg caffeine were evaluated in double-blind, placebo-controlled, cross-over manner.
Results: The phi2 index, reflecting repetition tendency within a sequence of two digits, gradually worsened with increasing age (p <0.02). Other RNG variables were not affected by age. By contrast, sleep deprivation increased counting tendency ("adjacency"). This consequence of sleep loss was attenuated by caffeine when compared to placebo (F = 9.5, p <0.003).
Conclusions: Advancing age and sleep deprivation differently affect RNG performance. The data demonstrate that the age-related decline in phi2 index does not reflect reduced vigilance, which increases counting tendency on the RNG.

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Identification of microsleep episodes

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Introduction: Microsleep episodes are brief episodes of sleep lasting up to 15 s and often individuals fail to respond to sensory stimuli. In the EEG, microsleep episodes are characterized by a change in oscillatory activity. In healthy subjects, microsleep episodes often occur under non stimulating conditions as a consequence of sleep deprivation or insufficient sleep, and in patients due to disease related excessive daytime sleepiness. Microsleep episodes may become dangerous due to the lapse in consciousness when occurring in situations that demand high attention such as driving or working, and subjective perception may be inadequate in some conditions. Thus, identifying early objective signs of sleep and microsleep episodes are of considerable clinical relevance. In order to diagnose the occurrence of microsleep episodes, the maintenance of wakefulness test (MWT) and the driving simulator test are often used.
Methods: The polysomnographic data and videos were recorded from participants performing MWT (sitting in a room with reduced light) or from a driving simulator test with the condition of driving on a highway road at night. In both tests, participants were instructed to stay awake. We analyzed data of 7 participants, each of them showing 1-9 microsleep episodes. Microsleep was defined here visually by experienced scorers as a sleep episode with duration of 3 to 15 s while the eyes were closed. We calculated spectrograms using an autoregressive model of order 16 of overlapping 1-s epochs moving through the data in order to visualize oscillatory activity. In addition, we quantified eye movements with the ratio between delta activity of the EOG and delta activity of the EEG of different derivations.
Results: Preliminary analysis revealed that microsleep episodes were mostly associated with a lack of eye movements, and often occurred after brief episodes of alpha activity.
Conclusion: Spectrograms proved to be useful to inspect the data and identify potential episodes of microsleep. Our aim for future work is to develop a method for semi-automatic detection of microsleep episodes, focusing on the data from the driving simulator.

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The eye closing behaviour in patients with Nonorganic Hypersomnia differs from the one in patients with Idiopathic Hypersomnia or Narcolepsy – a proof of concept study

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Objectives: Differential diagnosis of hypersomnia, particularly between Nonorganic and Idiopathic Hypersomnia, still remains a challenge. We hypothesise that the eye closing behaviour in the maintenance of wakefulness test (MWT) could be an additional diagnostic tool in the armamentarium of sleepiness assessment. The aim of this proof of concept study was to determine diagnosis specific parameters derived from the eye closing behaviour.
Methods: Up until now, four patients in each of the following groups have been included: Nonorganic Hypersomnia (NOH), Idiopathic

Hypersomnia (IH), Narcolepsy with and without cataplexy (N), and healthy sleep deprived controls (H). Based on face videography, MWT's were analysed from "lights off" to the first sleep fragment, defined by theta dominance in the electroencephalography and eye closure (>80%) lasting >3 seconds. The following parameters were compared between the four groups: total time of periods where eyes were closed, the latency to the first eye closure, and length of the eye closure period preceding the first sleep fragment. Eye blinks were discarded. Due to the small number of patients, no statistical analysis has been conducted.
Results: Even though the latency to the first sleep fragment is similar in patients with NOH (19 ± 4.6) and IH (20.3 ± 1.6 min), the latency to the first eye closure is much shorter in patients with NOH (5.2 ± 4.3 min) than in IH (9 ± 4.4 min), and the mean total time of periods where eyes were closed is longer (NOH = 170 ± 46 sec, IH = 92 ± 55). Based on the latency to the first sleep fragment, N (8.1 ± 3.1) and H (5.1 ± 3.2) could already be distinguished from patients with NOH and IH.
Conclusion: Patients with NOH close their eyes relatively early but stay awake longer with closed eyes, whereas patients with IH close their eyes later and then fall asleep more rapidly. The current preliminary data supports our hypothesis that a difference in eye closing behaviour between patients with NOH and IH in the MWT could be diagnostically useful. However, more patients need to be included, and further analysis must be conducted.

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Impact of acute sleep deprivation and chronic sleep restriction on slow wave activity and vigilance

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Introduction: Sleep loss as during acute sleep deprivation (aSD) and chronic sleep restriction (cSR) results in impaired vigilance. Increased slow wave activity (SWA) during recovery sleep after prolonged wakefulness is a well-established marker of sleep pressure. We aimed at directly comparing the effects of aSD and cSR on vigilance, subjective sleepiness and SWA.
Methods: We investigated 6 male subjects undergoing 40 hours of aSD and 7 nights of cSR (5h instead of 8h sleep/night). We compared first hour SWA (power density in the 1.25–4.5 Hz range) of the electrode showing maximal change in recovery nights after aSD and cSR relative to a baseline night (chosen from 128 electrodes). We examined vigilance at 3 pm before the respective nights, using the psychomotor vigilance task (PVT), i.e. speed and number of lapses (reaction times >500 ms; transformed: $\sqrt{x}+\sqrt{(x+1)}$), and subjective sleepiness using the Stanford Sleepiness Scale.
Results: The SWA increase after aSD (+102.0 ± 30.2%, mean ± SD) was markedly higher ($p < 0.01$) than after cSR (+37.0 ± 9.8%). Subjective sleepiness was increased after aSD (+1.7 ± 1.2; $p < 0.05$) and cSR (+2.0 ± 1.3; $p < 0.05$) to a comparable amount ($p = 0.58$). In the PVT, speed was reduced to equal values ($p = 0.95$) after aSD ($-0.6 \pm 0.4s^{-1}$; $p < 0.05$) and cSR ($-0.4 \pm 0.3s^{-1}$; $p < 0.05$). An increase in lapses was seen after aSD (+2.2 ± 1.6; $p < 0.05$), but not after cSR (+0.8 ± 1.6; $p = 0.29$), with a trend for more lapses after aSD ($p = 0.08$).
Conclusions: In contrast to a higher impact of aSD on SWA and lapses in vigilance, we found comparable effects of aSD and cSR on psychomotor speed and subjective sleepiness. Further analysis in this ongoing study, i.e. topographical distribution of SWA changes, inter-individual differences, or changes during the course of cSR might reveal novel insights in the underlying mechanisms of these seemingly discrepant findings.

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Reduced Risk Aversion Following Chronic Sleep Restriction but Not Acute Sleep Deprivation

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Introduction: Acute sleep deprivation (aSD) has been reported to alter decision making. Although chronic sleep restriction (cSR) mirrors everyday life more accurately than aSD, few studies examined the effects of cSR on decision making behavior. Such sleep restriction particularly affects people in positions with vast responsibilities including managers, scientists, and politicians, where risky decisions could have far-reaching consequences. Our controlled, crossover, within-subject study investigates the influence of both aSD and cSR on decision making under risk.
Methods: We studied the decision making behavior under risk in ten healthy males (mean ± SD: 22.9 years ± 2.6 years). Their risk preferences were assessed (afternoon and evening) prior to and after cSR (reduction of sleep duration from 8h per night to 5h per night over a period of one week) as well as prior to and after aSD (one night of total sleep deprivation). Participants performed a risk task, adapted from Levy and colleagues (2010), allowing the analysis of the model-free risk aversion ratio. A repeated measures ANOVA (within-subject factors: *intervention*: aSD and cSR, *time-point*: before and after intervention, *daytime*: afternoon and evening) and paired samples t-tests were performed with $p < 0.05$ considered significant.
Results: Preliminary results from the repeated measures ANOVA yielded no significant main effects (*time-point:* $F(1,9) = 3.97$, $p = 0.08$, *intervention*: $F(1,9) = 1.10$, $p = 0.32$, *daytime:* $F(1,9) = 0.24$, $p = 0.64$). However, a significant interaction effect was observed between the factors *time-point* and *intervention* (*time-point x intervention:* $F(1,9) = 5.99$, $p < 0.05$). A subsequent paired t-test of the difference values (proportional change of aSD and cSR measurements compared to the baselines) revealed a significant difference between the afternoon risk aversion ratio change during cSR (mean ± SD: –32.0% ± 22.6%) compared to aSD (9.6% ± 45.3%, $p < 0.05$). Meanwhile, no such significant effect was observed for the evening sessions ($p = 0.09$).
Conclusion: Preliminary results from this ongoing study suggest a decrease in risk aversive behavior following cSR, while no change in the risk aversion ratio was noted after aSD. Therefore, cSR and aSD might have distinct effects on risk aversion. These findings are a first step towards understanding risk taking behavior under cSR.

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Psychological well-being and optimism are related to morning cortisol secretion and objective measures of sleep in school-age children

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Positive psychological characteristics including psychological well-being and optimism are important factors for physical and mental health. The aim of the present study was to test the relationship of morning cortisol secretion and objective measures of sleep with psychological well-being and optimism in school-age children. A total of 113 normally developing children (age range: 6–10 years) took part in the study; of those 58 were born very premature and 55 were born at term. Objective sleep and morning cortisol secretion (at awakening, and at 10, 20, and 30 minutes later) were assessed. Sleep-EEG is the gold standard for sleep assessment and provides objective measures of sleep duration, sleep continuity (i.e. sleep efficiency), and sleep architecture (sleep stage 1, sleep stage 2, slow wave sleep (SWS), and rapid eye-movement sleep (REM-sleep)). Mothers and fathers reported their children›s psychological well-being and dispositional optimism. As possible confounders analyses controlled child age, gender, and prematurity status. Psychological well-being was positively associated with slow-wave sleep (SWS; $\beta = .30$, $t = 2.95$, $P = 0.004$) and negatively with REM-sleep ($\beta = -.31$, $t = 2.93$, $P = 0.004$). Dispositional optimism was positively related with SWS ($\beta = .20$, $t = 2.02$, $P = 0.046$) and negatively with basal cortisol levels at awakening ($\beta = -.21$, $t = 2.23$, $P = 0.03$). The findings inform that lower levels of morning cortisol secretion, less REM-sleep, and more SWS are associated with positive psychological characteristics in normally developing children during middle childhood. Among the sleep stages SWS is generally considered as the most restorative sleep stage. While inferences regarding causal relations are not possible due to the correlative nature of the study, the findings inform on a correlational level regarding possible psycho-biological underpinnings of the development of positive personality characteristics in childhood.

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Assessing the role of cerebral autoregulation during intrathoracic pressure changes by near infrared spectroscopy (NIRS)

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Introduction: Changes in intrathoracic pressure in obstructive sleep apnea have strong intermittent effects on oxygenation of hemoglobin and left ventricular stroke volume. While compensatory systemic mechanisms are well studied the role of cerebral autoregulation remains essentially unknown. We aimed to compare hemodynamic response in muscle and brain assessed by NIRS during Valsalva- and Müller- maneuvers (VM respectively MM) and thereby describing the role of cerebral autoregulation.
Methods: In five healthy adults cerebral and muscular NIRS and esophageal pressure (EP) were continuously measured during normal breathing, two VM (EP +15/+30 cmH_2O) and two MM (EP -15/-30 cmH_2O). We compared NIRS-signal regarding relative changes in oxygen saturation [SO_2] and total hemoglobin [tHb] and in particular the area under the pulse-wave-curve of tHb which is thought to reflect left ventricular stroke volume.
Results: In both VM and more pronounced in MM a decrease in SO_2 and a subsequent increase with event resumption was seen while tHb increased during VM and decreased during MM. During the latter phase in both sustained MM and VM the area under the pulse-wave-curve in tHb was reduced. Relative decrease of SO_2 was attenuated in brain compared to muscle indicating compensatory cerebral autoregulation in response to intrathoracic pressure changes.
Conclusion: Cerebral and muscular hemodynamic changes can be monitored non-invasively by NIRS. Comparison of the muscular and cerebral NIRS-signal is a promising tool to describe how cerebral autoregulation can cope with the challenge of intrathoracic pressure changes.

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Impact of electronic device use on sleep in adolescents in Geneva

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Introduction: Sleep plays a critical role for cognitive and affective functions. Recent research suggests that experience-dependent synaptic changes do not only occur during adult sleep but also serve brain maturation in childhood and adolescence. Based on these observations, we hypothesized that the expansion of electronic media use might cause a state of chronic sleep deprivation in adolescents. Specifically, increased arousal and the blue-light emitted by the screens would delay sleep onset.
Methods: We thus assessed the use of electronic devices and measured the sleep habits of 413 Swiss adolescents between 12 and 21 years old using actigraphy, daily diary, and melatonin profile. Second, we evaluated the impact of a simple educative recommendation on sleep, by asking adolescents to stop using electronic devices after 9 pm (N = 276).
Results: Preliminary results revealed insufficient sleep in adolescents, with about one hour less than recommended (9h), particularly in older participants. Adolescents also spent 1h30 using media after 9 pm. Social network activities was the most used media and significantly delayed sleep onset ($r = 0.37$; $p < 0.001$) while homework do not have an impact on bed time ($r = 0.02$; $p = 0.52$). For those who stopped using media after 9 pm, media use decreased by 53 min (±51 min) and total sleep time increased by 15 min (±38 min). Reduction of media use in the evening significantly correlated with earlier sleep onset ($r = 0.38$; $p < 0.001$).
Conclusion: These findings demonstrate that the emergence of new technologies profoundly modifies sleep habits and induces a chronic sleep deprivation in adolescents. A simple recommendation (no media use after 9 pm) can improve their sleep.

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