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Abstracts

Annual Meeting Swiss Society of Paediatrics

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FREE COMMUNICATIONS FM 1 – FM 30

FM 1

Epidemiology of the Kawasaki disease in children in Switzerland

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Introduction: Kawasaki disease (KD) was first described in 1967 and is now the leading cause of acquired heart disease in children in developed countries. It is an acute febrile illness of unknown aetiology occurring predominantly in infants and young children and most commonly affecting coronary arteries. The epidemiology of KD is unknown in Switzerland, therefore we conducted a national study to investigate the demography, diagnosis and treatment of children with KD.

Material and methods: We worked with the Swiss Paediatric Surveillance Unit (SPSU) to take a census of the children hospitalised with the diagnosis of KD in Switzerland from March 2013 to February 2017, in a prospective manner. We defined complete KD by the AHA criteria: the presence of ≥ 5 days of fever and ≥ 4 of the 5 principal clinical features (cutaneous rash, cervical lymphadenopathy >1.5 cm diameter, conjunctivitis, changes of lips or oral/pharyngeal erythema and extremity changes). The cases with less than 4 clinical signs were considered incomplete. We included all children under 17 years of age.

Results: We included 175 patients, 105 (60%) were boys, with a median age of 38.2 months (standard deviation (SD): 31.0). The most frequent clinical sign was a rash (85.4%). The diagnosis was made 7.3 days (SD 4.1) after the first symptom was identified. The complete form of the disease was reported in 107 children (61.1%) and an abnormal echocardiography was found in 91 patients (52.3%). Most of the patients had the recommended treatment of intravenous immunoglobulin (IVIG) (174; 99.4%) and aspirin (172; 98.3%). A second dose of IVIG because of persisting fever was given in 39 cases (23.8%). Second line treatment with corticosteroids or infliximab was necessary in 29 children (16.6%) and 1 patient (0.6%) received rituximab as 3rd line treatment. One (0.6%) child died of KD.

The global incidence of KD in children under 16 years of age in Switzerland 3.05/100'000/year. For the children less than 5 years old, the incidence is 6.8/100'000/year.

Conclusion: The incidence of the disease in our cohort is in the range of other European countries (5-10/100'000/year in children less than 5 years old). The complete form of the disease was reported in 61.1% of the patients. More than half had an abnormal echocardiography during the acute phase. In most cases, IVIG and aspirin are appropriately before day 10 of fever.

FM 2

Immunization coverage against tetanus, measles, rubella and varicella among migrant children

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Background: Migrants are at higher risk of developing infectious diseases. Epidemics often start in refugee camps as consequence of poor hygiene and lower vaccination coverage. Tetanus, measles, rubella and varicella are four vaccine-preventable diseases. The aim of this study is to assess the immunity against these four diseases among migrant children in Geneva Children's Hospital and to identify factors that could be used to predict the usually unknown vaccinal status.

Methods: We retrospectively analyzed the serostatus for tetanus, measles, rubella and varicella of children aged 1 to 16 years of age using an enzyme-linked immunosorbent assay (ELISA) for tetanus, measles and rubella and an electrochemiluminescence assay for varicella.

Results: Among 507 children analyzed between January the 1st 2012 and December the 31st 2016, 98% were protected against tetanus. Teenagers had significantly higher negative results (4.2% vs 1.3%, $P = 0.048$). 93.1% were seropositive for measles and the seropositivity increased with age. 79.2% children had a positive result for rubella. African children had predominantly negative serologies (60% vs 20.8%, $P < 0.01$) while others were predominantly positive. 68.6% were immune against varicella with a significant increase of seropositivity with age.

Conclusion: A large proportion of children were immune against tetanus, measles, rubella and varicella. However, some subgroups of children were less protected at least against one of the four diseases analyzed. We therefore could establish recommendations depending on the origin and the age of the children on who to vaccinate first (without testing serologies) and verify seroresponse after and who to test first and vaccinate only when needed. Such recommendations avoid unnecessary and unpleasant testing, and/or vaccination, and may have an impact on health costs.

FM 3

CXCL13 as a diagnostic marker of neuroborreliosis in children, a retrospective case-control study

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Aim: Lyme neuroborreliosis (LNB) is a frequent manifestation of Lyme disease in children and its diagnosis, which requires proof of intrathecal antibody production, has limitations especially in early cases. The elevation of the chemokine CXCL13 in the cerebrospinal fluid (CSF) of adult patients with LNB has been demonstrated and suggested as a new diagnostic marker. Our aim was to evaluate the diagnostic value of CSF CXCL13 in children with suspected LNB and to determine a cut-off concentration of CSF CXCL13.

Methods: For this single-center retrospective case-control study we used a diagnostic-approved ELISA to measure CXCL13 concentrations in the CSF of 185 children with suspicion of LNB at presentation. Patients were classified into definite LNB (patients with a positive intrathecal antibody index as a gold standard), non-LNB (controls with other CNS affections, i.e. viral meningitis, inflammatory diseases, idiopathic facial palsy) and possible LNB. A receiver operating characteristic curve was generated by comparison of cases and controls.

Results: CXCL13 was significantly elevated in the CSF of 53 children with definite LNB (median 774.7 pg/ml) compared to 91 non-LNB patients which exhibited low concentrations of CXCL13 (median 4.5 pg/ml). A cut-off of 55 pg/ml resulted in a sensitivity of 96.7% and a specificity of 98.1% for the diagnosis of definite early LNB. The positive and negative predictive values of the test were respectively 94.6% and 98.9%. Elevated CSF CXCL13 levels were also detected in 3 cases of viral meningitis (enterovirus $n = 1$, varicella-zoster-virus $n = 2$) while other CNS affections such as idiopathic facial palsies did not lead to any CXCL13 elevation. In patients with possible LNB ($n = 41$) where a definitive diagnosis could not be obtained, CXCL13 ranged from 4.5 to 1'418.6 pg/ml (median 16.7 pg/ml) and 27% of these patients had elevated CXCL13 values.

Conclusion: We confirm that also in children CSF CXCL13 is highly elevated during early LNB. This marker is highly sensitive and specific and should help to differentiate LNB from other CNS affections in paediatric patients.

FM 4

Ongoing burden of Streptococcus pneumoniae sepsis in children after introduction of pneumococcal conjugate vaccines

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Background: Population-based studies assessing the impact of pneumococcal conjugate vaccines (PCV) on burden of pneumococcal sepsis in children are lacking. We aimed to assess this burden following introduction of PCV-13 in a nationwide cohort study.

Methods: The Swiss Pediatric Sepsis Study (09/2011-12/2015) prospectively recruited children <17 years of age with blood culture-proven sepsis due to *Streptococcus pneumoniae*, meeting criteria for systemic inflammatory response syndrome. Infection with vaccine serotype in children up to date with PCV immunization was defined as vaccine failure. Main outcomes were admission to pediatric intensive care unit (PICU) and length of hospital stay (LOS).

Results: Children with pneumococcal sepsis ($n = 117$) accounted for a crude incidence of 2.0 per 100,000 children (95% CI 1.7-2.4) and 25% of community-acquired sepsis episodes. Case fatality rate was 8%. 42 (36%) patients required PICU admission. Children with meningitis (29; 25%) were more often infected by serotypes not included in PCV (69% vs 31%; $p < 0.001$). 16 (26%) of 62 children up to date with PCV immunization presented with vaccine failure, including 11 infected with serotype 3. In multivariable analyses, children with meningitis (OR 6.8; 95% C.I. 2.4-19.3; $p < 0.001$), or infected with serotype 3 (OR 2.8; 95% C.I. 1.1-7.3; $p = 0.04$) were more often admitted to PICU. Children infected with serotype 3 had longer LOS (coefficient 0.2, 95% CI 0.1-1.1; $p = 0.01$).

Conclusions: The incidence of pneumococcal sepsis in children shortly after introduction of PCV-13 remained substantial. Meningitis mostly due to non-vaccine serotypes and disease caused by serotype 3 represented significant predictors of severity.

FM 5

Mycoplasma pneumoniae-induced mucocutaneous disease: a prospective longitudinal cohort study

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Objectives: To report the occurrence and clinical presentation of *Mycoplasma pneumoniae*-induced mucocutaneous disease in a prospective longitudinal cohort study of children with community-acquired pneumonia (CAP).

Methods: We investigated *M. pneumoniae*-induced mucocutaneous disease among 152 children enrolled during a prospective longitudinal CAP study from May 1, 2016, to April 30, 2017 at the University Children's Hospital Zurich. Infection with *M. pneumoniae* was diagnosed by polymerase chain reaction (PCR) in pharyngeal samples and confirmed with the measurement of peripheral blood immunoglobulin (Ig) M antibody-secreting cells (ASCs) by enzyme-linked immunospot (ELISpot) assay.

Results: Mucocutaneous eruptions developed in 10 (23%) cases of CAP positive for *M. pneumoniae* by PCR ($n = 44$), all of whom tested positive for specific IgM ASCs. *M. pneumoniae* PCR-negative CAP cases had skin manifestations in 3% ($p < 0.001$). The spectrum of *M. pneumoniae*-induced mucocutaneous disease included *M. pneumoniae*-induced rash and mucositis (MIRM; $n = 3/44$, 7%), urticaria ($n = 2$, 5%), and exanthematous skin eruptions ($n = 5$, 11%). Two cases had ocular involvement as sole mucosal manifestation (bilateral anterior uveitis and non-purulent conjunctivitis). Cases with *M. pneumoniae*-induced mucocutaneous disease had longer prodromal fever ($p = 0.02$) and higher CRP levels ($p = 0.04$) than cases with *M. pneumoniae* CAP without skin manifestations. They were also more likely to require oxygen ($p = 0.007$), hospitalization ($p = 0.01$), and to develop long-term sequelae ($p = 0.03$).

Conclusion: Mucocutaneous disease occurred in one out of four cases with *M. pneumoniae* CAP, significantly more frequent than in CAP of other etiology. *M. pneumoniae*-induced mucocutaneous disease was associated with increased systemic inflammation, morbidity, and higher risk of long-term sequelae.

FM 6

Frequency and indication of antipyretic use in infants hospitalized for RSV bronchiolitis

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Introduction: Acetaminophen and non-steroidal anti-inflammatory drugs (NSAIDs) are frequently used in infants to reduce fever and pain. Although they are usually well tolerated in childhood, any medication should only be used for a specific indication. In Switzerland, there is a lack of information on the frequency and indication of antipyretic use (APU) in inpatient infants with RSV bronchiolitis. The aim of this study was to examine whether the main reason for APU in these infants was fever.

Methods: 2-year retrospective single centre evaluation of data from all infants hospitalised for RSV-bronchiolitis in 2017 and 2018 in a tertiary non-university children hospital of Switzerland. Data on age, sex, underlying disease, vital signs and any treatment received including use of antipyretics and oxygen consumption was extracted from the electronic clinical information system. Infants with an underlying disease or need for intensive care were excluded. Fever was defined as documented body temperature $>38^{\circ}\text{C}$.

Results: During the observation period, 79 infants were included where of 51.9% were male with a median age of 2.5 months (range 0.5-12). Median hospital duration was 4 days (range 1-15) and oxygen was necessary for a median of 3 days (range 0-13). 60/79 (76%) children received a total of 381 antipyretic doses with a mean of 6.4 doses per infant (range 1-27, 95%-confidence interval 4.67-8.03). The most prevalent drug used was acetaminophen with 333/381 (87.4%), followed by ibuprofen with 48/381 (12.6%). For 19/79 (24%) infants no APU was documented during their hospital stay. Of all 60 infants with APU, 32 (53%) of all infants received their first APU while having fever. Only 10 infants (17%) received APU as a result of elevated body temperature only. 19/60 (32%) never had fever. A fixed dose of APU was administered to 10 infants, 5 infants had an additional indication for pain due to acute otitis media as a known complication of their RSV bronchiolitis. 8 of all infants received antibiotics.

Conclusion: Interestingly, the indication for APU remains unclear in the majority of children and only a minority received antipyretics as a result of elevated body temperature only. However, results from this small retrospective single centre evaluation do not allow to ascertain whether a specific antipyretic treatment was justified or not. To further clarify indications and identify possible areas of unnecessary APU, a prospective multicentre study is planned.

FM 7

Metabolic surgery in adolescence as part of a multi-professional therapy of obesity

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Introduction: Treatment of obesity in children and adolescents is an interdisciplinary challenge. Obesity has long-term physical and psychological consequences. The urgent need for therapy is beyond doubt. In Switzerland, multi-professional group programs (MGP) and multi-professional structured individual therapies (MSIT) are established for therapy. However results in general are moderate and unsustainable. Metabolic surgery is often not included as a treatment option.

Clinical setting: At the Lucerne Cantonal Hospital, a cooperation between the Children's Hospital, Lucerne Psychiatry and the Central Switzerland Obesity Center for multi-professional therapy in the treatment of children and adolescents has been established since September 2013. Various specialist disciplines pool their experience. Both MGP and MSIT are provided and surgery is indicated for selected cases when conservative measures over a minimum of 2 years have failed.

Results: From September 2013 to January 2019, 330 patients between the age of 10 and 18 years were assigned for therapy. By January 2019, 168 young patients with their families were active in the therapeutic program. Of them, 18 patients (9 female, 9 male) aged 14-17 years (mean 16.7 years) underwent surgery: Gastric bypass was performed twice and a sleeve resection in 16 patients. All operations were performed laparoscopically without any surgical complications. At time of surgery, average BMI was 46.1 kg/m² (range 37.4-60.3 kg/m²) with weight of 138 kg

(109–178 kg) and height of 1.7 m (1.63 – 1.85 m). Postoperatively, mean BMI and weight changed as follows: After 6 months: BMI 37.7 kg/m² and weight 100 kg, after 1 year: BMI 30.9 kg/m² and 86 kg, after 2 years: BMI 28.8 kg/m² and 86 kg, and after 3 years: BMI 28.2 kg/m² and 83 kg.

Conclusions: In specialized centers offering a conscientious selection with intensive preparation and follow-up, surgery can be carried out safely in obese adolescents as part of a multi-professional therapy regimen. Weight loss through surgery proved to be superior as compared to conservative therapy along with significant improvement of physical health and mental well-being.

FM 8

Humanitarian paediatrics: a race to reach those most in need

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Background: Since 1990, the world has taken significant actions to reduce child mortality from 12.6 million of children under 5 deaths per year to less than 5.4 million in 2017. However in humanitarian or conflict contexts the rates of reduction are much slower. The presence of conflict in a country correlates very closely with a higher rate of U5 mortality. Nonetheless, most pediatric global health protocols and policies have been developed for low-resource but relatively secure and politically stable settings. There are major characteristics of humanitarian contexts that need to be taken into account to allow tackling child mortality in a more efficient way.

Methods: A review of the current literature focused on Humanitarian Paediatrics was performed using Web of Science, PubMed, Scopus, and EMBASE. In addition, we analyzed neonatal and <5 year mortality rates to assess associations with violent conflict. Data were derived from estimates of the United Nations Inter-agency Group for Child Mortality Estimation (UN IGME) for 2017. Political and violence data were estimates from the World Bank Political Stability and Absence of Violence/Terrorism (PS/AV) indicator for 2017. The PS/AV is a scale 0-100 with countries <15 considered unstable and experiencing high rates of violence.

Results: We found only 122 articles were relevant to child health in humanitarian settings with less than 5 making a specific reference to humanitarian paediatrics. Neonatal and <5 mortality rates were highest for countries experiencing violent conflict. Of the 20 countries with the highest neonatal mortality rates, 18 had PS/AV figures of <15. In the Sub-Saharan region, countries with PS/AV figures <10 accounted for approximately 50% of total <5 deaths in the region. Literature review identified 3 general issues that distinguish pediatric care in humanitarian and conflict areas: 1) epidemiology of pediatric conditions, particularly acute malnutrition, measles, and trauma; 2) security, including the targeting of health workers and facilities; and 3) requirements of humanitarian principles, including neutrality and independence, which may prohibit access and close working relationships with host governments and other local actors.

Conclusion: There is a need for the development of a new field, humanitarian paediatrics, which can address both the growing contribution of and special requirements of providing child health care in humanitarian or conflict settings.

FM 9

A systematic literature review of challenges in health care delivery to migrants and refugees in high-income countries – the 3C model

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Background: Migrants and refugees have important health needs and face inequalities in their health status. Health care delivery to this patient group has become a challenging public health focus in high income countries. This paper summarizes current knowledge on health care delivery to migrants and refugees in high-income countries from multiple perspectives.

Methods: We performed a systematic literature review including primary source qualitative and quantitative studies between 2000 and 2017. Articles were excluded if the study setting was in low- or middle-income countries or focused on skilled migration. Quality assessment was done for qualitative and quantitative studies separately. Predefined variables were extracted in a standardized form. Authors were approached to provide missing information.

Results: Of 185 identified articles, 35 were included in the final analysis. We identified three main topics of challenges in health care delivery: communication, continuity of care and confidence. All but one study included at least one of the three main topics and in 21/35 (60%) all three topics were mentioned. We further developed the 3C model and elaborated the interrelatedness of the three topics. Additional topics identified showed that the specific regional context with legal, financial, geographical and cultural aspects is important and further influences the 3C model.

Conclusions: The 3C model gives a simple and comprehensive, patient-centered summary of key challenges in health care delivery for refugees and migrants. This concept is relevant to support clinicians in their day to day practice and in guiding stakeholders in priority setting for refugee and migrant health policies.

FM 10

Rare diseases and complex care of asylum-seeking children in a tertiary hospital in Switzerland

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Background: In the years 2016 and 2017 an estimated 20'000 children applied for asylum in Switzerland. Although there are several studies investigating the health status and needs of asylum-seeking children, no study has focused on the need of frequent and complex care in this population. The aim of this study is to assess the characteristics of the asylum-seeking children most frequently visiting a tertiary care hospital in Switzerland, detailing their underlying medical conditions and analyzing patterns of care provided.

Methods: Patients with asylum-seeking status receiving care at the University Children's Hospital Basel in Switzerland between January 2016 and December 2017 were identified using administrative and electronic health records. Only patients with frequent visits were included in this analysis defined in a first step as having ≥11 total visits or ≥1.5 visits per months. Of those patients with <7 days admissions and or <5 total visits were further excluded. Patient data including main diagnoses, nationality, age, gender, admission and discharge date, escape route, parental consanguinity and primary care physician was extracted.

Results: A total of 462 patients with 1816 visits were identified. Of those 19/462 (4%) fulfilled the inclusion and exclusion criteria with 811/1816 (45%) of the total visits. Patients age ranged from 0 to 16.7 (median 7.0) years. At total of 16/19 (84%) patients were in two age groups: those <2 and the others >12 years of age. In infants genetic diseases (5/8; 63%) and nutritional problems (6/8; 75%) were most common, whereas in adolescents orthopedic diseases (4/8; 50%) and mental health problems (4/8; 50%) were prevalent. A genetic disease was diagnosed in 6/19 (32%) patients. Of the total visits 34/811 (4%) were admissions, 66/811 (8%) were visits in the emergency Department and 320/811 (39%) were visits in the outpatient Departments. The haemato-oncological Department was the most frequently visited outpatient Department 123/320 (38%). Non-physician and physician visits were equally distributed 413/811 (51%) and 420/811 (52%) respectively. Exercise therapy visits were most frequent in the non-physician visits 197/413 (48%).

Conclusion: Asylum-seeking children most frequently visiting a Swiss tertiary health care facility suffered from rare and chronic diseases with a high proportion of genetic diseases. This patient group required multi-disciplinary and complex care resulting in many health care visits.

FM 11

The perspective of asylum-seeking caregivers on the quality of care provided by a Swiss paediatric hospital – a qualitative study

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Objectives: This study investigated the perspective of asylum-seeking caregivers on the quality of health care delivered to their children in a qualitative in-depth interview study. The health of asylum-seeking children is of key interest for health care providers, yet knowledge of the perspective of asylum-seeking caregivers when seeking health care is limited.

Setting: The study focused on one paediatric tertiary care hospital in Basel, Switzerland.

Participants: Interviews were carried out with thirteen asylum-seeking caregivers who had presented with their children at the paediatric tertiary care hospital. Nine female and four male caregivers from Tibet, Eritrea, Afghanistan, Syria, Iraq, Albania, and Macedonia were included. A diverse sample was chosen regarding cultural and social background, years of residence in Switzerland, and reasons for seeking care. A previously developed and pilot tested interview guide was used for semi structured in-depth interviews that took between 36 and 92 minutes. Data analysis and reporting was done according to consolidated criteria for reporting qualitative research (COREQ). The interviews were carried out until saturation was reached.

Results: The interviewees described a mismatch of personal competences and external challenges. Communication barriers and unfamiliarity with new health concepts were reported as challenges. These were aggravated by isolation and concerns about their child's health. The following factors were reported to strongly contribute to satisfaction of health care delivery: a respectful and trustful caregiver-provider relationship, the presence of interpreters and immediate availability of treatment.

Conclusions: A mismatch of personal competences and external challenges importantly influences the caregiver-provider relationship. To overcome this mismatch establishment of confidence was identified as a key factor. This can be achieved by availability of interpreter services, sufficient consultation time, and transcultural trainings for health care workers. Coordination between the family, the asylum and the medical system is additionally required to facilitate this process.

FM 12

Home visits to identify the role of lifestyle and stress in families with obese children: An explorative field study

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Background: Obesity is a frequent disease in paediatrics with lifelong consequences, but even with an evidence-based multiprofessional approach, therapeutic success is only moderate. Previous results from the national KIDSSTEP study suggest that lacking awareness of obesogenic environment at home and psychosocial stress may impede lifestyle changes. With an explorative field study using home visits, we aim to assess the impact of psychosocial stress and resources and whether the home setting promotes a healthy lifestyle.

Methods: At start of the 12-months multiprofessional obesity group program (MGP), two health professionals (HP) visited the patients' homes. They observed the home setting and conducted semi-structured interviews according to an evidence-based checklist, indicating supportive versus obesogenic environment in % of all families, and the Heidelberg Stress scale (HBS, 2012), evaluating e.g. the patient's and families' stress.

Results: Eighteen children fit the criteria to participate in a structured MGP (11 male, age at baseline 10.93±1.69, BMI at baseline = 28.05kg/m²±4.98, BMI-SDS at baseline = 2.87±0.72, means±SD). Mean BMI-SDS decreased significantly during MGP (-0.14, p <0.05). HBS showed elevated stress levels (mean±SD including all investigated areas 31.37±17.38, clinical cut-off 20 in reference sample for compensated stress levels) in patients as well as in the whole family. Concerning obesogenic environment, in 67% of the families food was constantly available, only 28% did planned grocery shopping and 22% of the children were engaged in chores such as meal preparation. Regarding physical activity, only 11% of the parents were active on a regular base and 33%

of the children participated in a sports club. 72% of the patients commuted to school by walking or cycling. Incidence of divorce or separation of the parents (33%) and the number of persons per household (4.1) were above average. However, the reduction of BMI-SDS was not significantly associated with favorable therapy conditions or family stress levels.

Conclusions: The home visits clearly show, that parents of obese children have a high psychosocial burden and limited resources. They cannot fulfil a positive role model function, especially regarding physical activity. This knowledge about the patient's lifestyle and home enables the HP to individualize counselling and strengthen the patient-HP relationship, which may increase the readiness of the families to start psychosocial therapies.

FM 13

Thalamocortical connectivity and neurodevelopmental outcome at 2 years in preterm infants

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Background: Disruption of the thalamo-cortical system is considered as a major component of preterm brain injury and thalamo-cortical connectivity has been shown to be associated with cognitive performance at 2 years of age.

Aim: To correlate the thalamo-cortical connectivity strength and thalamo-cortical fiber length with neurodevelopmental outcome at 2 years of age in preterm infants.

Methods: In 58 infants (mean (SD) gestational age at birth 29.75 (1.44) weeks and CGA at scanning of 41.1 (2.09) weeks) diffusion tensor imaging was performed. Thalamo-cortical tractography was performed to project connectivity strength of cortical voxels using ProbtrackX algorithm and to map mean fiber strength within the thalamus. Children were assessed using the Bayley Scales of Infant and Toddler III Assessment at a mean corrected age of 22 months (2.3). The model was corrected for GA, CGA and SES.

Results: Thalamo-cortical connectivity strength correlated significantly with MDI in the frontal lobe, predominantly in pre-motor, prefrontal, dorsolateral prefrontal and orbitofrontal regions. These with MDI correlating regions showed connectivity to the ventro-anterior, ventro-lateral, mediodorsal and centrolateral thalamic nuclei. Thalamo-cortical fiber length correlated with MDI in the ventro-anterior, ventro-lateral and centrolateral thalamic nuclei.

Conclusion and discussion: More mature and stronger thalamo-cortical connectivity, especially to the frontal lobe, correlated with better cognitive outcome at 2 years of age in children born preterm. These results reflect the importance of the ventrolateral, mediodorsal and ventro-anterior thalamic nuclei in cognitive functioning in children born preterm.

FM 14

Development of a motorized guardian for the rehabilitation of the hand in hemiplegic children

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Introduction: Hemiparesis is the partial loss of voluntary motor activity of one half of the body, which is the most frequent disease in infantile cerebral palsy. Infantile cerebral palsy is defined as a group of permanent disorders of movement and posture development, which cause a limitation of activity, attributing a permanent (non progressive) damage that occurs in the brain during brain development in the first years of life. An important therapeutic aspect of the hemiparesis is rehabilitation; at the level of the lower limb it is often possible to obtain good functionality, however this is not always the case for the upper plegic limb. The objective of the rehabilitation in this limb is to find a stimulator that allows the patient to recover as much as possible the functionality of the upper limb whilst also allowing a reorganization of the compensatory brain circuits.

Method: Develop a device (brace) that allows the patient to guide the movement of the hand and fingers, individually or in pairs, to exert a pre-

established sequence of movements that simulates the movements that are made with the hand in daily use.

Results: By working closely with a hemiplegic patient, it was possible to develop an active brace that allows the patient to perform rehabilitative movements at hand level.

Conclusion: Having a brace that allows these patients to have a better function at the level of the upper plegic limb, guarantees them a better quality of life.

Financing: Faulhaber Minimotor SA of Croglio has made the following electromechanical equipment available

FM 15

Recanalisation treatment in childhood and young adult stroke – comparison of safety and outcome

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Objective: Intravenous thrombolysis and endovascular therapy (IVT/EVT) are increasingly used in the pediatric arterial ischemic stroke (AIS) population. However, it remains unknown if these treatments are as safe and effective as in adult AIS patients. This study aimed to compare safety and outcome of IVT/EVT in children and young adults with AIS.

Methods: Retrospective study (01/2000–12/2017) of patients aged 1 month – 45 years treated with IVT/EVT for AIS. Clinical and radiological data of children (1 month – 16 years) were compared to young adults (>16 years – 45 years) by univariate analysis. Delayed treatment initiation was defined >4.5 hours and >6 hours after symptom onset for IVT and EVT respectively. Outcome was assessed using the mRS 3-6 months after AIS.

Results: 19 children (age 11 +/- 3.9 years, 6 (31.6%) females) and 175 adults (age 36 +/- 8.1 years, 85 (48.6%) females) were included. Overall, children showed different etiologies ($P = 0.001$) and lower recanalisation rates ($p = 0.019$), whereas initial pedNIHSS, bleeding complications, mortality and outcome did not differ between the two groups. Comparing IVT to EVT, children receiving IVT showed a higher initial pedNIHSS ($p = 0.003$), delayed treatment initiation ($p = 0.001$) and worse outcome ($p = 0.003$) compared to adults treated with IVT. No major differences were observed in children and adults treated with EVT.

Interpretation: Safety and outcome of recanalization treatment in children is comparable to findings observed in young adults. Delayed treatment initiation is a serious concern in the pediatric population treated with IVT and is probably responsible for the observed differences in outcome parameters.

FM 16

AutoPlay: smart games for evolutionary screening under 14 months

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Autism is a particularity of development that involves about 1 child per 100 births. In addition to alterations in social relations, the diagnosis speaks of narrow interests that manifest themselves in different ways. The scientific literature describes an atypical playful development in children with autism. Despite this fact, today this particularity is still little

taken into account in the early screening of children under 14 months of age. AutoPlay is a kit of 5 intelligent toys (1 doll, 3 elephant cubes, 1 teaspoon, 1 ball, 1 toy car) developed at SUPSI in synergy with the Neuropediatric Unit of the Department of Paediatrics and a paediatric centre in Mendrisio. The introduction of sensors inside each toy allows, thanks to the development of specific algorithms, to recognize and follow the evolution of the patterns of manipulation of the toys by the child aged between 9 months and 14 months. The AutoPlay project therefore intends to objectify these patterns of manipulation in order, and in the future, to objectify and measure the play trajectory of children with autism. Thanks to the BREF funding for social innovation, of the Gerbert Ruff Foundation and the financial support of SUPSI itself, the AutoPlay project is currently in its phase of stabilization of the methodology. That is to say, it is about the construction of a core of algorithms, clinically relevant, from the measurements made on about 15 children in two nurseries. The first algorithms are being elaborated and allow to encode and detect play patterns in a reliable and precise way.

FM 17

Sporadic acute benign calf myositis: systematic literature review

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Background: Acute benign calf myositis is an infection-associated syndrome of muscle pain. It presents with pain affecting the calves, recovers within a week and occurs either in epidemics or sporadically. Epidemic cases are usually associated with influenza virus type B. Little is known on the microorganisms that have been associated with sporadic cases.

Methods: In order to characterize the sporadic form and increase the knowledge of this condition, we systematically reviewed the literature reporting sporadic cases using the PRISMA strategy.

Results: We identified 72 reports, which included 451 patients (325 males and 126 females). Sporadic acute benign calf myositis affected subjects ≤ 18 years of age ($N = 450$; 99%), was preceded by a flu-like illness ($N = 411$; 91%) and presented with pain affecting only the calves for ≤ 10 days ($N = 441$; 99%). The creatine kinase level was always increased. An acute kidney injury was never reported. Microbiological studies identified an infectious trigger in 181 cases: influenza (type B more frequently than type A) or parainfluenza viruses ($N = 95$), Dengue virus ($N = 41$), Epstein-Barr virus ($N = 12$), Mycoplasma pneumoniae ($N = 7$), further microorganisms ($N = 26$). Recurrences of the myositis were observed in 13 cases. -Limitations: Two limitations of this work should be stated. First, available data do not allow documenting the prevalence of sporadic acute benign calf myositis. Second, since microbiologically uncharacterized cases are less likely to be published than cases caused by Dengue virus (publication bias), our data might overestimate the frequency of this microorganism as a cause of acute calf myositis.

Conclusions: Sporadic acute benign calf myositis characteristically follows a prodromal flu-like illness and affects subjects ≤ 18 years of age with a male-to-female ratio of 2:1. It presents with bilateral calf pain, elevated total creatine kinase level, is never complicated by myositis-associated acute kidney injury, recovers within ≤ 10 days and is often associated with mild leukopenia or thrombocytopenia. The information generated from this review may help physicians to become familiar with this condition.

References: Capoferri G, Milani GP, Ramelli GP, Ferrarini A, Bianchetti MG, Lava SAG. Sporadic acute benign calf myositis: systematic literature review. *Neuromuscul Disord* 2018; 28: 443-9.

FM 18

Preschool children with developmental delay: the standard of care evaluated

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Developmental delay (DD) with a prevalence of 15 percent of all children is one of the most frequent disorders in early childhood affecting annually thousands of individuals in Switzerland. Early identification of children with DD is critical to ensure appropriate therapeutic interventions, to sup-

port the families and finally to prevent chronic (i.e., life-long) health, educational and social consequences. It is widely accepted that early intervention programs are both ethically mandatory and cost-effective for the society on a long-term perspective. However, we note that there is a large paucity of information about supply, demand and effectiveness of services for children with DD in Switzerland. In the Canton of Zurich, there is a centrally organized registration of all children with DD in need of early interventions at the Unit of Special Needs Education (USNE), based at the University Children's Hospital Zurich and the Center for Social Pediatrics of the Kantonsspital Winterthur. We have collected and analyzed data from all children (age 0-4) admitted to the USNE in 2017 (n = 2033) and will present descriptive data on demographic and clinical features of the children (e.g. age at admittance, developmental profiles, reasons for early support, medical diagnoses, regional distribution, sociodemographic and language background, and quality of the supporting network), pathways of access, and patterns of service usage. Our analyses provide comprehensive insights into the structures and the utilization of health and educational care of children with DD in the Canton of Zurich and give an overview on the current practice with respect to early interventions in the Canton of Zurich. Furthermore, we report on the status of the establishment of a sustainable registry of services for DD children based on the USNE database.

FM 19

Impact of therapy on quality of life in patients treated for retinoblastoma

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Background: Treatment of retinoblastoma (Rb) includes a combination of local treatments, systemic chemotherapy with focal treatments (CT), external beam radiotherapy (EBR) and/or enucleation. Since a few years, intraarterial (IAC) and intra-vitreous administration of chemotherapy (IVC) are used as new conservative treatments. Limited data are available regarding the impact of treatments on the health-related quality of life (HRQoL) in cured patients. Our study assessed HRQoL in Rb survivors and compared the results between four different treatment modalities: enucleation, EBR, CT and IAC/IVC.

Methods: This is a population-based cross-sectional study. Questionnaires were sent out to all RB survivors (aged 3-20 years) who were entirely treated at our centre and who had a minimal follow-up of 3 years since end of treatment. The HRQoL was assessed using the KIDSCREEN-52 self-reported and parent proxy version and the total score and the scores of each dimension were compared between the four different treatment modalities.

Results: Seventy-eight Rb survivors and their parents participated. The overall total score was 43.10/50 (SD = 6.03). Rb survivors who received primary CT scored significantly higher than the other groups in the total QoL score and in the school environment dimension score. They scored significantly higher compared to those who were enucleated, in the dimension of psychological well-being, and scored higher than the EBR group in the dimension of self-perception and autonomy. The treatment groups did not differ in physical, moods and emotions, parent relations, social support and social acceptance domains. Factors associated with a lower score were: older age of the patient at study time, older age at first examination (pediatric oncology unit and Ophthalmic Hospital), self-reported questionnaires and unilaterality.

Conclusion: Our results show that the perceived HRQoL by Rb survivors was globally good and that the HRQoL differed according to the treatment received. Patients treated with EBR have, as expected, the worst HRQoL. To our surprise, systemic chemotherapy, although related to more general side effects, scored best in almost all domains, even when compared to the newer local chemotherapies (IAC/IVC). In order to confirm these data, patients who were not yet available will be included and socio-demographic factors such as parents' education, migration background and country of residence, added since they could also affect the QoL.

FM 20

In children with chemotherapy for cancer 39.0 °C ear temperature is a safe temperature limit defining fever. Results: of the randomized controlled multicenter trial SPOG 2015 FN Definition (NCT02324231)

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Background: Fever in severe neutropenia (FN) is the most frequent potentially lethal complication of chemotherapy for cancer. In children, FN mortality is <1% thanks to emergency treatment including empirical broad-spectrum antibiotics. The temperature limit defining fever (TLDF) used clinically, varies from 37.5°C to 39.0°C, reflecting the lack of evidence. As previously published, a high versus low TLDF can avoid FN diagnoses in patients spontaneously recovering without therapy. This study aimed to determine if a high TLDF of 39.0°C ear temperature is non-inferior to a low TLDF of 38.5°C regarding safety in pediatric oncology.

Methods: In this randomized controlled non-blinded multicenter study, pediatric patients with chemotherapy for cancer were randomized in monthly clusters to a TLDF of 39.0°C or 38.5°C. FN diagnosis and therapy below the randomized TLDF was allowed for clinical reasons. The primary outcome was the rate of FN with any safety relevant event (SRE) per chemotherapy exposure time (CET). SREs were death, admission to intensive care unit (ICU), severe sepsis and bacteremia.

Results: 6 of 9 sites of the Swiss Paediatric Oncology Group (SPOG) recruited patients of all diagnostic categories from April 2016 to August 2018. After the second of 3 planned interim analyses, the study stopped for success. 269 patients were randomized 2547 times during 195 years of CET. All were treated per protocol. An SRE was diagnosed in 72 (20%) of 360 FN episodes (death, 0; ICU admission, 16; severe sepsis, 22; bacteremia, 56). In 92 CET years randomized to 39.0°C, 151 FN episodes were diagnosed (rate, 1.64/year), including 51 (34%) below 39.0°C and 22 (15%) with SRE (rate, 0.24 per year). In 103 CET years randomized to 38.5°C, 209 FN episodes were diagnosed (2.03/year), including 28 (13%) below 38.5°C and 50 (24%) with SRE (rate, 0.49 per year). The mixed Poisson regression rate ratio of FN with SRE in 39.0°C versus 38.5°C was 0.57, with a 95% upper confidence limit of 0.72. The predefined non-inferiority margin was 1.33.

Conclusions: In pediatric patients with chemotherapy for cancer, the use of a high TLDF of 39.0°C versus 38.5°C ear temperature is both efficacious and safe. For Switzerland and comparable settings, 39.0°C can be recommended as new evidence-based standard TLDF. The responsible physician will decide to diagnose and treat FN below this TLDF if clinically indicated. In non-comparable settings, confirmatory trials are needed before clinical use.

FM 21

Neuroendocrine cell hyperplasia of infancy (NEHI), a childhood interstitial lung disease with good prognosis

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Introduction: Neuroendocrine cell hyperplasia of infancy (NEHI) is a unique sub-type of childhood interstitial lung diseases (chILD), first described in 2005. It is a clinical entity with typical history, clinical exam and computed tomography (CT) findings and a pathophysiology that is poorly understood. We present a case series of four patients with NEHI treated in our institution over the last four years.

Case series: All infants were referred to our clinic for further evaluation because of persistent signs of respiratory distress at the age of four, six, seven and thirteen months respectively. Pregnancy and labour had been unremarkable in all of them and they were born full term with an uneventful neonatal course. On clinical examination, tachypnea, bilateral retractions and diffuse crackles were found. Three infants presented with hypoxemia and failure to thrive. CT showed typical ground glass opacities in the paramediastinal lung areas, in the right middle lobe and lingua, as well as air trapping. Differential diagnoses such as pulmonary infection, immune deficiency and other congenital cardiopulmonary diseases were ruled out. Lung biopsy, which showed an increased percentage of neuroendocrine cells in the airways, was performed in one infant. Infants presenting with hypoxemia were supported with oxygen therapy. Nutritional support was necessary in one case. Three of our four patients were asymptomatic by the age of two years, which is in line with the good prognosis of the disease. The fourth patient who is currently under one year old has only been diagnosed recently and is still under oxygen therapy.

Discussion: The etiology, incidence and prevalence of NEHI remain uncertain. The literature suggests that it is a rare and sporadic disease. Our case series with identification of four patients in a single institution in the last few years suggests that NEHI is probably underdiagnosed and more frequent than has been thought so far.

FM 22

Clinical symptoms do not reflect functional impairment in early cystic fibrosis lung disease

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Background: Lung impairment in cystic fibrosis (CF) can already occur within the first year of life. However, frequency and severity of overt respiratory symptoms in infants with CF are not known.

Methods: We included 50 infants with CF and 50 healthy matched controls from two prospective birth cohort studies. Respiratory symptoms and respiratory rate were documented weekly throughout infancy. Infants performed lung function measurements within the first weeks of life.

Results: The analyses included 4552 data points (2217 in CF). Respiratory symptoms (general, mild or severe) were not more frequent in infants with CF (OR:1.1; 95% CI: [0.76,1.59] p = 0.6). Early lung function and respiratory rate measurements were elevated in infants with CF compared to healthy controls, but not associated with respiratory symptoms.

Conclusions: We found no difference in respiratory symptoms between healthy and CF infants. This may indicate that early subclinical changes might not be captured by the clinical presentation of patients.

FM 23

Impact of breastfeeding on lung function, respiratory and allergic diseases in school children

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Background: A protective effect of breastfeeding on lung functional growth has been demonstrated in cohorts of children with asthma or risk for asthma; however it is unclear whether this effect is mediated by inflammation or by a direct effect on lung growth. We assessed the impact of breastfeeding on lung function and symptoms at the age of six years in an unselected, healthy birth cohort.

Methods: We prospectively studied 377 healthy term infants from the Bern-Basel Infant Lung Development (BILD) cohort from birth up to 6 years. Any breastfeeding was assessed weekly during the 1st year of life. Risk factors (e.g. smoking exposure, parental atopy, and education) were obtained using standardized questionnaires. The primary outcomes were lung function parameters measured at 6 years of age by spirometry (FEV1, FVC, FEV1/FVC, and FEF25-75) and body plethysmography (FRC, TLC and Reff) and FeNO. Secondary outcomes included ever wheeze, wheeze in the past 12 months, asthma, atopy, atopic dermatitis, allergic rhinitis, and allergic sensitization.

Results: We found no evidence that breastfeeding could influence any lung function parameters and FeNO. After adjustment for confounders we found no associations of breastfeeding with respiratory symptoms or sensitization. However, a protective effect of breastfeeding on atopic dermatitis was observed only in girls (odds ratio (OR) per each week of breastfeeding 0.96; 95% confidence interval (CI) 0.92-0.99).

Conclusion: This study suggests that breastfeeding has no effect on lung function in unselected healthy children with low risk for asthma, our findings do not support our hypothesis that breastfeeding might have a direct impact on lung function at 6 years.

FM 24

Oximetry as a criteria for hospitalization: is the credence given to oximetry reliable?

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Introduction: Bronchiolitis guidelines recommend that patients should be hospitalized and oxygen therapy initiated if SpO2 is <90%. However, the criteria for using oxygen therapy vary widely without evidence that oxygen saturation predicts disease progression and length of hospital stay. Furthermore, few is known about the natural history of desaturations in bronchiolitis. We aim to evaluate the desaturation rate and the time for desaturation in patients with normal SpO2 (>90%) upon arrival to the emergency Department (ED). A secondary aim was to identify the hospital readmission rate.

Methods: Prospective clinical trial in children aged <1 year admitted to the ED with bronchiolitis during two RSV seasons. Patients included in our study received the standard management, and they were provided with a portable oximeter to continuously record SpO2 for 36 hours after ED admission. Desaturation was defined as at least 1 documented O2 saturation less than 90% lasting 1 minute or more.

Results: Of 217 patients enrolled (112F:105M), 186 patients were hospitalized (18 patients in the intensive care) and 31 were discharged, 70% were positive for RSV. The initial clinical presentation of hospitalized patients was more severe with higher respiratory rates (P = 0.015) when compared to those discharged home. Despite that, SpO2 values at ED arrival did not differ between the two groups (P = 0.19). In hospitalized patients, 73% of desaturation were noted, and occurred earlier than in patients discharged home [3 hours (IQR 2-9) vs. 12 hours (IQR 8-21), P <0.001]. The majority of discharged patients (71%) also experienced desaturations at home, but only 27% were readmitted due to a worsening of their clinical status.

Conclusion: Infraclinical and self-resolving desaturations may occur in infants with bronchiolitis at home, but were not always accompanied by a worsening of the clinical status or need for hospitalization. The decision to discharge home or to hospitalize infants with bronchiolitis should be based maybe more on the clinical presentation than on the SpO2 values.

FM 25

Posttraumatic stress and health-related quality of life in parents of children with cardiac rhythm devices

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Background: Studies have shown a high prevalence of post-traumatic stress disorders (PTSD) among parents of children with life-threatening

diseases. Parents of children living with a pacemaker (PM) or implantable cardioverter defibrillator (ICD) face a variety of stressors related to their child's arrhythmia and medical treatment. However, it is yet unknown whether parents of children with cardiac rhythm device develop posttraumatic stress symptoms or even PTSD.

Methods: This cross-sectional investigation is part of a comprehensive single-center study of long-term medical and psychosocial outcomes in pediatric patients with PM and ICD. 69 patients (78%) were included in the study, with the participation of 69 mothers and 57 fathers. Parents completed the Posttraumatic Diagnostic Scale and Medical Outcomes Study Short Form-36 item questionnaire. Child's medical data was collected retrospectively from patients' hospital records.

Results: At assessment, the patients (39% females) were on average 11.2 years old and belonged mostly to the middle or upper SES class (96%). The predominant device type was PM in 56 cases (81%), while 13 cases (19%) had an ICD. The mean time since device implantation was 6.3 years (SD = 4.3, range 0.4-17.9), with 6 children (9%) who received their device within the last 12 months. Full heart-disease related PTSD based on the Diagnostic and Statistical Manual of Mental disorder, 4th edition (DSM-IV), was diagnosed in one mother and no father, while partial PTSD was diagnosed in 3 mothers (4%) and 2 fathers (4%). Parental HRQoL – especially regarding the mental health dimension – was affected in both parents. In both parents, PTSD symptom severity scores were significant predictors for parental mental health after controlling for child age at implantation, presence of other non-cardiac disease in the child, parental age, and presence of own chronic disease in a parent.

Conclusions: Thus, special attention should be given to parental PTSD symptoms in the clinical follow-up of PM and ICD patients as some parents might probably benefit from psychological support.

FM 26

Evolution of pediatric eosinophilic esophagitis over 10 years

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Aim: Eosinophilic esophagitis is a unique form of non-IgE-mediated food allergy characterised by esophageal eosinophilic infiltration. The aims of this study were to describe a cohort of children diagnosed and treated for EoE over the past decade, to describe the causative allergens identified by dietary elimination, medical and dietary treatments used by physicians.

Methods: Single-center retrospective study at the pediatric gastroenterology unit, University Hospital Lausanne from 2006 to 2016. Inclusion criteria: confirmed diagnosis by history, endoscopy, and presence of more than 15 eosinophils (HPF).

Results: In total, 22 patients diagnosed between 2006 and 2016 were analyzed, 77% boys. The time from symptoms to diagnosis was 3.5 years and age at diagnosis was 10 years. The most frequent indications for endoscopy were: dysphagia (77%), gastroesophageal reflux (55%), bolus impaction (27%), refusal food (36%) and abdominal pain (55%). These different symptoms occur with a median age of 3.5 years for gastroesophageal reflux, 8 years for abdominal pain as well as dysphagia, and 13 years for bolus impaction. Between 2006 and 2010 on average 0.6 case were diagnosed per year, versus 3.17 cases per year between 2011 and 2016. Food allergies were identified in 73%, aeroallergies, in 73%, respectively. Eczema was found in 40%, whereas 9% didn't have any known allergy. Positive family history for allergies was reported in 40%, eosinophilic esophagitis in 5%, respectively. Middle and distal third of esophagus was affected in 91%, proximal third in 68%, respectively. Endoscopy appeared macroscopically normal in 9% of patients. No patient showed failure-to-thrive. Development of functional abdominal pain, anxiety-depressive disorders and behavior disorders were observed in 27% of patients. Many had trouble sleeping. Topical steroids were used without elimination diets in 27%, both diet and steroids in 64% and esophageal dilation in 5%.

Conclusion: Eosinophilic esophagitis is a rare pathology but with an increase incidence. A multidisciplinary approach is essential with pediatricians, gastroenterologists, pathologists, allergists, dieticians and child psychiatrists.

FM 27

Derivation and validation of a clinical decision rule to identify children at risk for intussusception

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Background: Intussusception is the most common cause of intestinal occlusion in young children. It cannot be reliably ruled out with clinical examination and the diagnosis is usually made with ultrasound performed by a radiologist.

Objective: The primary objective of this study was to derive and validate a clinical decision tool to identify children at risk for intussusception who need an ultrasound among children younger than 5 years old.

Methods: This was a prospective cohort study performed in a tertiary care pediatric emergency Department. Participants were a convenience sample of children younger than 5 years old who necessitated an abdominal ultrasound for suspected intussusception according to the treating physician. The primary outcome was radiological confirmation of intussusception. A-priori, 24 independent variables were identified through literature review and experts consensus. Independent variables were collected prospectively before ultrasound using a standardized research clinical report form. Derivation of the clinical score was performed using multiple variable logistic regression among the participants after the recruitment of 50 cases of intussusception. Then, a second sample including at least 25 patients with an intussusception was used to measure predictive ability of the score using ROC curve (validation phase).

Results: A total of 238 children fulfilled the inclusion/exclusion criteria and were recruited during the derivation (n = 175; 50 cases) and validation (n = 63; 25 cases) phases. Using a multivariate model, a risk score was derived and includes 4 clinical criteria. This score stratifies patients at low, moderate or high risk. When applied to the validation study population the score showed moderate predictive ability with an area under the ROC curve of 0.63 (95%CI: 0.49 To 0.77).

Conclusion: We derived a clinical decision score to stratify the risk of intussusception among young children with clinical suspicion of intussusception. Using this score will improve resources utilisation for suspicion of intussusception.

FM 28

Prosecution for shaken baby syndrome (SBS) cases: association between time of medical diagnosis, filing of criminal charges and the judicial outcome.

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Introduction: Shaken baby syndrome (SBS) is a serious brain injury resulting from forcefully shaking an infant. Lesions are essentially cerebral, eventually associated with subdural hematomas and retinal hemorrhages. SBS is an act of physical child abuse liable to public prosecution. Once diagnosed, health care professionals in the canton de Vaud have a legal obligation to report suspected cases of SBS to the childhood protection service that may then file criminal charges. In some cases, physicians have directly filed criminal charges. The aim of this study was to assess whether judicial outcome is influenced by the quality of the medical data and/or the period of time between diagnosis and filing of criminal charges.

Methodology: The study, approved by the Ethical Committee CER-VD (n° 2017-00436), included infants diagnosed with SBS at the University hospital of the canton de Vaud (CHUV) between 2001 and 2016. Inclusion criteria were children less than 2 years old with certain medical diagnosis of SBS, for whom charges had been filed, criminal cases were closed and the full accessibility to the legal files had been obtained. The cohort was divided into two groups according to the judicial outcome: conviction or acquittal.

Results: Twenty-three infants were enrolled in the study. They presented suggestive clinical signs and symptoms of SBS (100%): subdural hematomas (100%), retinal hemorrhages (87%) and associated lesions (96%). Thirteen cases (57%) resulted in an acquittal and 10 in a conviction (43%). Time between diagnosis and filing of criminal charges was 29,2±67,7 days and 7,1±13,3 days for the acquittal and conviction group, respectively. This indicated that convictions were more likely when

charges were filed rapidly. Time until the filing of charges was short when that was done directly by physicians [acquittal group: 7.9±6.2 days (n = 9) / conviction group: 3±3 days (n = 9)] but was prolonged when it was done by the childhood protection service [acquittal group: 76.8±117.7 days (n = 4) / conviction group: 44 days (n = 1)].

Discussion: Medical characteristics of SBS cases diagnosed at the CHUV were similar to the available medical literature. The certainty of diagnoses was never questioned during legal proceedings, neither in the acquittal, nor in the conviction group. Convictions were more likely when criminal charges were filed more swiftly. Direct filing of criminal charges by physicians was associated with a reduction of this time span.

FM 29

New height references for children in Switzerland

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Background: In 2011, World Health Organization (WHO) growth curves replaced those of the first Zurich longitudinal study (Prader curve) in Switzerland. Meanwhile, various European studies showed that locally determined growth curves are clearly superior to the WHO curves and that, above all, the 3rd percentile leads to missed or delayed diagnoses.

Aim: To present height-for-age percentiles accurately reflecting today's growth of children in modern multicultural Switzerland.

Subjects and methods: Between 2017-2018, we collected 15,000 cross-sectional data sets of growth and weight from 34 pediatric practices as well as 4,800 measurements made at 20 participating schools. The prospective data was supplemented with 3 retrospective data sets from Swiss military recruits of 2013-2017, Swiss newborns registered from 2012 to 2016 and obligatory examinations in public schools of Zurich aged in 2017. The total sample comprised 30,033 boys and girls aged 0-20 years. Height reference curves were created using Cole's LMS method. Derived height percentiles were compared with those of the Prader curve, WHO data set and new reference data from neighboring countries.

Results: The first 5 years of growth are almost identical with the 55-year-old Prader reference data. Thereafter, children in Switzerland today are taller. The difference of 1 cm between 5 and 9 years in girls and 5 and 10 years in boys may possibly be attributed to a more prominent adrenarche. The main difference in height of 3.5 cm for girls at 11 years and 4 cm for boys at 13.5 years is the consequence of an earlier pubertal growth spurt. The small difference in adult height (1 cm) mirrors the declining secular trend. The comparison between our curves and those established by WHO shows that Swiss children, from the second year of birth until adulthood, are taller. In addition, Swiss boys and girls are both 3 cm taller from the age of 6 years and above. When reaching final height, Swiss boys and girls are 1.8 cm and 3.5 cm taller respectively, than the WHO population. The most important difference lies in the 3rd percentiles, which are about 4 cm below the 3rd percentile of our new growth references. Our new data are corroborated by recent growth curves from Germany and Austria. Over the entire age range, the comparison shows only small differences on the 50th as well as 3rd percentile.

Conclusions: This new height-for-age reference data should be used for assessing individual growth of children in Switzerland.

FM 30

TINU syndrome with severe ocular involvement: case report and literature review

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Background: Tubulointerstitial nephritis and uveitis (TINU) syndrome is characterized by interstitial nephritis associated with anterior, posterior or global uveitis. The latter can precede, follow or be concurrent with renal disease. Most patients are adolescents or young adults with a female predominance. Whereas prognosis of renal involvement is usually good, uveitis tends to relapse. First line therapy are topical and/or systemic steroids. We report a rare case of TINU with severe and atypical ocular involvement and the preliminary results of a systematic literature review.

Case report and literature review: A 13 year-old boy was addressed to our Department for diagnostic workout in the context of fatigue, abdominal pain, vomiting and increased creatinine level. Two weeks earlier he was diagnosed with right anterior uveitis treated with topical corticosteroids and complained of polyuria and polydipsia. At that time, a diabetes mellitus was excluded. Laboratory evaluation showed a moderate anemia (Hb 117 g/L), renal failure (creatinine 134 µmol/L), tubular proteinuria and an inflammatory reaction (ESR 84 mm/h). Renal biopsy showed an acute tubulointerstitial nephritis, suggesting the diagnosis of TINU syndrome. Renal impairment spontaneously improved. Despite local steroid therapy, uveitis progressed into severe bilateral panuveitis with bilateral papillary edema. Systemic steroid and azathioprine were needed. We planned a systematic literature review searching in PubMed, personal files and secondary references peer-reviewed articles concerning TINU, retaining for analysis case reports and case series only.

Results: The literature search retrieved 377 articles: 172 were excluded (not pertaining, duplicate, narrative review, language). A preliminary analysis of 97 cases shows a median age of 16 years with a female-to-male ratio of 1.5:1. In 20% of cases uveitis precedes, in 50% follows and in 28% is concomitant to nephritis. In most cases (82%) a bilateral uveitis is diagnosed. Complete renal recovery is documented in 78% of cases. Uveitis relapse occurs in 56% of cases, motivating systemic steroid therapy. Second line immunosuppressive drugs are needed in about 10% of cases, mainly for ocular indication.

Conclusion: TINU syndrome should be suspected in front of uveitis concomitant, preceded or followed by reduced renal function or signs of tubular dysfunction. Uveitis can be severe and steroid-dependant, sometimes needing long-term immunosuppressive therapy.

SWISSPEDNET SPN 1 – SPN 11

SPN 1

Insufficient planning time results in poor planning performance in children and adolescents born very pretermDisselhoff V^{1,2}, Schnider B^{1,2}, Latal B^{2,3}, Hagmann C^{1,2}, Wehrle F^{1,2,3}¹Department of Pediatric and Neonatal Intensive Care, University Children's Hospital Zurich, Switzerland; ²Children's Research Center, University Children's Hospital Zurich, Switzerland; ³Child Development Center, University Children's Hospital Zurich, Switzerland**Background and aims:** Many children and adolescents born very preterm (VPT) show planning deficits, particularly in more complex tasks. The current study aimed to investigate whether insufficient planning time underlies poor planning performance in highly demanding tasks in VPT children and adolescents.**Methods:** 41 VPT participants aged 10 to 16 years with normal cognitive abilities and 42 term-born (TB) peers completed a computerized planning task with increasing levels of difficulty (Stockings of Cambridge, SOC). Individual changes in error rates from the lowest to the highest task-difficulty level were correlated with respective changes in planning time, using Pearson's correlation. Moreover, differences between the baseline planning time and the planning time after having made an error were compared between groups using repeated measures ANOVA.**Results:** Across all participants, higher error rate changes across increasing task-difficulty levels were related to lower changes in planning time ($r = -.325$, $p = .003$). VPT participants did not increase their planning time after having made an error (interaction birth status x time-point: $F = 4.857$, $p = .032$).**Conclusions:** Across both groups, participants, who don't increase their planning time as the task becomes more difficult, make more planning errors. Unlike their term-born peers, very preterm individuals further fail to adapt their planning time after having made an error. This impaired adaptation may underlie VPT participants' problems in complex planning tasks and could serve as a target for interventional approaches.

SPN 2

Executive function abilities predict behavioral problems at school-age in children and adolescents born very pretermSchnider B^{1,3}, Disselhoff V^{1,3}, Latal B^{2,3}, Hagmann C^{1,3}, Wehrle F^{1,2,3}¹Department of Neonatology and Pediatric Intensive Care, University Children's Hospital Zurich, Switzerland; ²Child Development Center, University Children's Hospital Zurich, Switzerland; ³Children's Research Center, University Children's Hospital Zurich, Switzerland**Background and aims:** Children and adolescents born very preterm are at an increased risk to develop executive function deficits and to suffer from social, emotional and attentional problems. This study aimed to investigate whether deficits in executive functions underlie behavioral problems in everyday-life.**Methods:** A group of 38 children and adolescents born very preterm with normal intellectual abilities and 41 term-born peers were assessed at a mean age of 12.9 (± 1.8) years with a comprehensive battery of executive function tests. A composite score was calculated to reflect overall executive function abilities. To assess behavioral problems, parents completed the Strength and Difficulties Questionnaire (SDQ). The total problem score reflects problems across four dimensions (emotional-/conduct-/peer problems, hyperactivity/inattention).**Results:** Birth status (preterm vs. term birth) significantly predicted the SDQ total problem score ($\beta = -0.33$, $p = .045$, 95% CI [-0.65, -0.01]) after taking into account sex, age, socio-economic status and IQ (adjusted $R^2 = .12$, $p = .014$). Adding executive functions as a predictor to the model increased the explained variance from 12% to 22% ($p < .001$). While executive functions significantly predicted the SDQ total problem score ($\beta = -0.43$, $p = .002$, 95% CI [-0.69, -0.16]), the effect of birth status diminished to non-significant.**Conclusions:** Increased behavioral problems in children and adolescents born very preterm may be explained by poorer executive function abilities rather than birth status per se. Consequently, executive function training should be considered as an intervention to reduce behavioral problems in the preterm population.

SPN 3

Diagnosing asthma accurately in school-aged children suspected to have asthmade Jong CCM¹, Pedersen ESL¹, Goutaki M¹, Trachsel D², Barben J³, Kuehni CE¹¹Institute of Social and Preventive Medicine, University of Bern, Bern, Switzerland; ²Paediatric Respiratory Medicine, Children's University Hospital of Basel, Basel, Switzerland; ³Paediatric Respiratory Medicine, Children's Hospital of Eastern Switzerland, St. Gallen, Switzerland**Introduction:** Several diagnostic tests are used in children suspected for asthma, but the accuracy of these tests to diagnose asthma is unclear.**Aim:** We aimed to assess the diagnostic accuracy of reported respiratory symptoms and clinical tests to diagnose asthma in school-aged children under investigation for asthma.**Methods:** We studied cross-sectionally children aged 6-16 years referred to 2 pulmonary outpatient clinics with suspicion of asthma. All children underwent clinical evaluation including spirometry, fractional exhaled nitric oxide (FeNO), skin prick tests, and bronchial provocation tests (BPT) by exercise, methacholine, and mannitol. Symptoms were reported in a parental questionnaire. Asthma was diagnosed by the physicians based on medical history, clinical examination, and all clinical test results. We calculated sensitivity, specificity, positive and negative predictive value and area under the curve to assess the diagnostic accuracy of symptoms and clinical tests.**Results:** Of the 111 participants, 68 (61%) were diagnosed with asthma. The combined sensitivity and specificity to diagnose asthma was highest for wheeze without colds ((sensitivity/specificity) 0.76/0.65), wheeze (0.87/0.51), wheeze triggered by pollen (0.59/0.79) and more than 3 attacks of wheeze (0.49/0.88). Of the diagnostic tests, the area under the curve was highest for FeNO (0.81) and BPT by exercise (0.79) and lowest for FEV1 (0.52) and FEV1/FVC (0.58).**Conclusion:** This study, which needs validation in a larger sample, suggests that specific questions about triggers and severity of wheeze, FeNO and BPT by exercise are more useful to diagnose asthma in school-aged children than spirometry, skin prick tests and other BPTs.

SPN 4

Factors influencing quality of life in parents of children with an inborn error of metabolismDimitrova Nevena¹, Dumont Lisa¹, Camenzind Lucia¹, Urben Sébastien¹, Müller-Nix Carole¹, Wüthrich Valentine¹, Aboulkheir Rachel², Ballhausen Diana²¹Service de Psychiatrie de l'Enfant et de l'Adolescent, Centre Hospitalier Universitaire Vaudois; ²Unité Pédiatrique des Maladies Métaboliques, Service de Pédiatrie, Centre Hospitalier Universitaire Vaudois

Inborn errors of metabolism (IEM) often require medical monitoring along with a restrictive diet and/or drug treatment. Such constraints likely decrease the quality of life (QoL) in affected patients and their families. However, this topic remains largely unexamined. In this study, we examined the factors that influence QoL in parents of children with IEM.

We assessed self-reported QoL in 87 parents (64% mothers; both parents responded for 10 children) of 67 children (Mage = 7.5 years; range 0-17 years; 39% girls) using an adaptation of the Phenylketonuria Quality of Life questionnaire. Additionally, parents responded to questions about mental health (Hospital Anxiety and Depression Scale), stress related to their child's IEM (Pediatric Inventory for Parents), coping strategies (Cognitive Emotion Regulation Questionnaire) along with questions about their child's mental health (Strengths and Difficulties Questionnaire). Treating physicians evaluated the severity of the IEM (adaptation from Interned). Correlations revealed that parental QoL was linked to the severity of child's IEM disease ($p = .005$), to parental depression ($p < .001$) and anxiety ($p < .001$), as well as to all aspects of illness-related stress reported by parents (i.e., communication, emotional functioning, role function, and medical care; $ps < .001$). Interestingly, parents of pediatric IEM patients showing higher QoL difficulties reported higher maladaptive coping strategies, such as self-blaming for the experience ($p = .028$) and thoughts that explicitly emphasize the difficulty of their experience ($p = .015$).

Taken together, findings reveal that parental QoL is related both to objective factors pertaining to the child's medical condition and to subjective, psychological factors, intrinsic to parents. Special care should be addressed to identify psychologically vulnerable parents already at IEM diagnosis in order to offer preventive interventions, such as supplying adaptive strategies to cope with difficulties related to parenting an IEM-diagnosed child.

SPN 5

Diagnosis of *Mycoplasma pneumoniae* pneumonia with measurement of specific antibody-secreting cells

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Background: *Mycoplasma pneumoniae* is a frequent cause of community-acquired pneumonia (CAP) in children. Current diagnostic tests for *M. pneumoniae* infection, including polymerase chain reaction (PCR) and serology, are unreliable in differentiating infected patients and carriers suffering from CAP caused by other pathogens. We investigated the measurement of specific antibody-secreting cells (ASCs) by enzyme-linked immunospot (ELISpot) assay as new diagnostic test for *M. pneumoniae* CAP.

Methods: Longitudinal observational study of 152 children with CAP and 156 healthy controls enrolled from May 2016–April 2017; thereof, 63 CAP patients and 21 controls were included based on the availability of fresh (isolated ≤ 4 h) peripheral blood mononuclear cells for an *M. pneumoniae*-specific IgM ASC ELISpot assay, as well as PCR on pharyngeal specimens and IgM serology.

Results: *M. pneumoniae* DNA was detected by PCR in 32 (51%) CAP patients and IgM ASCs in 29 (46%) patients ($p = 0.72$). The three patients who were PCR-positive but IgM ASC-negative were diagnosed with another pathogen. *M. pneumoniae* DNA was detected in 10 (48%) controls, all of whom tested negative for IgM ASCs ($p = 0.0002$). A positive IgM serology was also found in 5 (15%) IgM ASC-negative CAP patients and 5 (24%) controls. IgM ASC detection re-classified 15% ($n = 13/84$) of PCR-positive ($p = 0.0009$) and 12% ($n = 10/84$) of IgM-seropositive study participants ($p = 0.004$).

Conclusions: The measurement of specific IgM ASCs by ELISpot improves the diagnosis of *M. pneumoniae* CAP. Extending this method to other pathogens may pave the way for timely and reliably determining disease etiology in childhood CAP.

SPN 6

Assessing neonatal visual maturity in children with congenital heart disease – a pilot study

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Background: Children with congenital heart disease (CHD) are at increased risk for neurodevelopmental impairment. Early detection of children at risk is pivotal to provide early therapies and tailored follow up programs. In preterm infants, the assessment of neonatal visual maturity is a strong predictor of neurodevelopmental outcome at 1 year of age and reflects white matter maturation and integrity. The aim of our study was to assess the feasibility of applying this neonatal visual assessment in the population of CHD newborns.

Methods: Neonates with complex CHD were recruited at the Neonatal ward of the University Children's Hospital Zurich and underwent neonatal visual assessment according to Ricci et al. (Ricci et al., 2008) before and after cardiopulmonary bypass surgery or catheter intervention. Healthy control subjects were recruited at well-baby wards. The 9-item neonatal visual test battery assesses a wide range of visual functions. Scoring was performed by scoring each item outside the 90th centile (reference data of neonates at 72 hours of life) as abnormal (score 1) with total scores ranging from 0-9 and higher scores indicating worse neonatal visual maturity. Total scores were compared between CHD infants and healthy controls.

Results: Twenty neonates with CHD and 12 controls were enrolled. Preoperative assessment was obtained in 5 and postoperative in 17 neonates. Preoperative examination could not be performed because the infant was too fussy ($n = 11$) or because of organizational reasons ($n = 4$). Reasons for missing postoperative assessments were delay of surgery beyond the neonatal period ($n = 3$). Median [IQR] age at preoperative and postoperative assessment was 14 [10, 14] and 25 [21, 32] days in CHD neonates and 17 [5, 21] days in controls. For CHD infants, preoperative median score was 1 (range 0-3) and postoperative 0 (0-9). In controls, median score was 0.5 (range 0-3) with no difference to CHD infants ($p = 0.59$ preoperative, $p = 0.73$ postoperative).

Conclusion: Preoperative visual assessment of CHD infants is limited while postoperative assessment is feasible. The majority of postoperatively examined CHD infants had normal neonatal visual assessment. A larger sample and normative values for older infants is necessary to determine the predictive value of a visual assessment for neurodevelopmental outcome in the CHD population.

SPN 7

Pitfalls of using IQ short forms in neurodevelopmental disorders: A validation study in patients with congenital heart disease

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Background and aims: IQ is widely used to profile cognitive disabilities in populations with neurodevelopmental deficits. Short forms of IQ assessments (S-IQ) are time-efficient and highly predictive for the estimation of the full IQ (F-IQ) in typically-developing children and adolescents and thus often used in a research setting. However, populations at risk for neurodevelopmental deficits may show a different distribution of IQ scores with a larger proportion of patients below average. So far, there is a lack of information on the agreement between S-IQ and F-IQ for these populations. Thus, the current study aims to investigate agreement between S-IQ and F-IQ in two samples of patients with congenital heart disease (CHD) and to test an approach to resolve potential disagreement.

Methods: The WISC-IV (German version) was applied in two independent samples of patients with CHD aged 9 to 16 years ($n_1 = 77$, $n_2 = 55$). F-IQ and a well-established four-subtest S-IQ version was calculated for each patient. First, the reliability of the short version was tested by correlating S-IQ and F-IQ. Second, we investigated whether the agreement between S-IQ and F-IQ was equal across the whole spectrum of IQ scores by correlating the size of the measurement error with the estimated true IQ (mean of S-IQ and F-IQ). Finally, we tested a method to adjust IQ scores to resolve potential disagreement between S-IQ and F-IQ.

Results: S-IQ ($M(SD)_1 = 96(17)$, $M(SD)_2 = 108(21)$) and F-IQ ($M(SD)_1 = 96(13)$, $M(SD)_2 = 104(17)$) correlated strongly, indicating a high reliability of the short version ($r_1 = 0.95$, $r_2 = 0.96$, both $p < 0.001$). The size of the measurement error significantly differed among the spectrum of IQ scores, indicating larger error for high and low scores (i.e. < 85 , > 115 ; $p < 0.001$, both samples). Adjusting the S-IQ scores of the second sample (n_2) with correction parameters from the first sample (n_1) eliminated the measurement errors in extreme cases ($p = 0.08$).

Conclusion: S-IQ assessments may be beneficial due to their time-efficiency and high reliability with F-IQ scores, particularly in a research setting. However, the current study shows that significant measurement errors may occur especially outside the normal range, namely, underestimation of low and overestimation of high IQ scores. This has to be considered, when patients with potential cognitive impairment are assessed. Importantly, this lack of agreement can be minimized by using an appropriate correction formula.

SPN 8

Asthma control in children referred to Swiss outpatient clinics.

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Background: Children with asthma should be adequately controllable with existing treatments. National data on asthma control are scant.

Aim: To describe asthma control and its predictors in children referred to Swiss paediatric respiratory outpatient clinics.

Methods: We used cross-sectional data from the Swiss Paediatric Airway Cohort (SPAC), a prospective clinical cohort study of all children aged 0-16 years seen in respiratory outpatient clinics in Basel, Bern, Luzern and Zurich, between August 2017 and July 2018. We collected information on diagnosis from the hospital letter and information on symptoms, treatment, socioeconomic and environmental factors from a parental questionnaire prior to the visit. We included children with diagnosed asthma. We used parent reported activity limitations, night-time symptoms, missed school days and frequent exacerbations, to classify asthma control into well (no symptoms), partly (1-2 symptoms present) or uncontrolled (3 or more present). Asthma control medication included inhaled corticosteroids (ICS) alone or combined with long acting-beta agonists (LABA), or leukotriene receptor antagonists (LTRA). We analysed associations of exposures with asthma control using ordinal logistic regression.

Results: We analysed data from 264 children (median age 10 years (IQR: 7-13), 35% female, study response rate: 53%). Asthma was well-controlled in 27 (10%), partly in 89 (34%) and uncontrolled in 148 (56%) children. Any use of asthma control medication was reported by 13 (48%) of the well, 64 (73%) of the partly and 113 (77%) of the uncontrolled children. Among the uncontrolled, 46 (41%) used ICS alone, 6 (5%) LTRA alone, 11 (10%) ICS + LTRA, 40 (35%) ICS + LABA and 10 (9%) ICS + LABA + LTRA. In the multivariable model, older age was independently associated with better asthma control (OR:0.92, 95% CI:0.85-0.99 for each year increase in age). Sex, parental education, region, nationality, current exposure to tobacco smoke, mould or humidity in the house and reported asthma control medication, were not associated with asthma control.

Conclusion: Asthma control is insufficient in most children referred to Swiss respiratory outpatient clinics, especially among younger children. Analysis of asthma treatment prescribed at the outpatient clinic and a follow-up survey 1 year later, will allow to assess whether asthma control improved after establishing a treatment regimen in a tertiary centre.

SPN 9

Association of time to antibiotics (TTA) and clinical outcomes in patients with fever and neutropenia during chemotherapy for cancer (FN), a systematic review

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Background: Fever in chemotherapy-induced neutropenia (FN) is the most frequent potentially lethal complication in patients with cancer. Prompt empiric broad-spectrum antibiotic therapy is the standard of care. The association of time to antibiotics (TTA) with clinical outcomes is not clear, however, and recommendations are based mainly on studies involving immunocompetent subjects with sepsis. We systematically reviewed the available data on the association between TTA and clinical outcomes in patients with FN.

Methods/design: The search covered 7 databases, reference lists were reviewed, forward citations searched and experts contacted. Studies were screened, and data extracted by one researcher and independently checked by a second. Confounding biases and study quality were assessed with the ROBINS-I tool. Safety (composite outcome: death, intensive care unit (ICU) admission, sepsis) and treatment adequacy (relapse of primary infection, persistence or recurrence of fever) were assessed as primary outcomes.

Results: Of 5389 titles and abstracts screened, 142 studies were retrieved and 9 studies, were included. All were observational studies, and all but one with at least moderate risk for bias, mostly due to baseline confounding. Predefined outcomes were reported variably. There was inconsistency among the findings regarding safety. Two studies at lower risk for bias and controlling for triage bias showed a possible association between longer TTA and impaired safety: specifically a lower 28-day mortality was found in patients with TTA ≤ 30 min (3.0%) vs. 31-60min (18.1%) (n = 307; HR, 1.18; 95% CI, 1.10 to 1.26); and less adverse events (mortality, ICU admission, fluid resuscitation) in patients with TTA ≤ 60 min (5.2%) vs. 61-120min (14.2%) (n = 1628; OR, 2.88; 95% CI, 1.70 to 4.89). Meta-analysis was feasible on 4 studies at moderate risk of bias, with 3 studies each reporting on death (OR 0.78, 95% CI 0.16 to 3.69) and on ICU admission (OR 1.43 95% CI 0.57 to 3.60). No study reported data on treatment adequacy. Triage bias, i.e., faster treatment of patients in reduced general condition or with known risk for poor outcome, was identified as a relevant confounding factor.

Conclusion: There seems to be an association between longer TTA and impaired safety. More precise knowledge about TTA effects on safety are important to optimize treatment guidelines for FN. Controlling for triage bias and other confounders is possible and necessary to gain further evidence.

SPN 10

Are there still sex-specific differences in asthma symptoms and treatment in Swiss school-aged children?

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Introduction: Studies done in the 1990s in Eastern Switzerland found that girls received less often treatment for asthma symptoms than boys despite similar symptoms. We aimed to describe the prevalence of wheeze and asthma medication use and to study treatment differences between boys and girls with wheeze using a recent survey.

Methods: The Luftibus study collected information on respiratory symptoms by parent completed questionnaires in unselected school-aged children from the canton of Zurich, 2013-16. We investigated whether wheeze prevalence and asthma inhaled medication differed between girls and boys using logistic regression.

Results: 3148 children participated in the survey, 1587 girls and 1561 boys, with median age of 12years (IQR 10-14, range 6-16). 253 (8%) children reported current wheeze and, from 6-9 to 13-16 year olds, the proportion decreased in boys from 11% to 8% while it increased from 5% to 9% in girls. 78 (2%) children had frequent wheeze (≥ 4 attacks in the past year). In the past year, among children with current wheeze, 129 (51%), 52% girls and 50% boys, used short-acting beta 2 agonists (SABA) and 94 (37%), 39% girls and 36% boys, were treated with inhaled corticosteroids (ICS). Among children with frequent wheeze only 45 (58%) and 27 (35%) used SABA and ICS respectively. Adjusting for frequency of attacks and number of asthma symptoms, there was no difference between boys and girls in the reported use of SABA (OR 0.95, 95% CI 0.54-1.7) and ICS (OR 0.93, 95% CI 0.53-1.7) overall. Among 6-9-year olds, boys tended to be less often treated with ICS than girls (OR 0.22, 95% CI 0.067-0.73).

Conclusions: A significant proportion of schoolchildren with current and frequent wheeze did not use inhaled asthma medication in the past year. We found no evidence that girls with asthma symptoms received less treatment than boys.

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SPN 11

Lung clearance index tracks progression of early lung disease from preschool to school age children with cystic fibrosis

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Background: Lung disease in children with cystic fibrosis (CF) starts early in life but often without clinical signs. Identifying lung disease in those silent years is crucial to prevent irreversible lung damage and preserve lung function. Lung clearance index (LCI) derived from multiple

breath washout (MBW) is a sensitive surrogate marker for tracking early lung disease in children with cystic fibrosis.

Objectives: This study aims to define whether LCI from clinical routine measurements can be used to track early lung disease from preschool to school age in children with cystic fibrosis.

Methods: Lung function data were collected from children with CF at preschool age (4-6 years) and subsequently at school age (6- 10 years) from routine outpatient clinics. Primary outcome was abnormal LCI derived from MBW and abnormal FEV1 from spirometry.

Measurements and main results: Twenty-eight clinically stable children with CF had lung function testing at both time points. Fourteen chil-

dren (42%) had an abnormal LCI at preschool age, with a positive predictive value (PPV) for abnormal LCI at school age of 86%. Negative predictive value for LCI was 58%. FEV1 was abnormal at preschool age in only two patients (7%) with a PPV of 100%. All children with an abnormal FEV1 at school age had already an abnormal LCI at preschool age. Negative predictive value for FEV1 was 86%, however the majority of children (23; 82%) still had normal FEV1 at school age, whereas LCI was elevated in 18 (64%).

Conclusion: Ability of LCI to predict abnormal lung function already at preschool age is excellent. LCI is superior to FEV1 in monitoring silent lung disease progression.

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P 1

Experience of VV ECMO in children with the Avalon® cannula

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Background: The Avalon® bicaval double lumen cannula (Avalon®) represents an innovative concept for veno-venous(VV)ECMO support in children. We report our experience with the use of this cannula for pediatric respiratory support.

Methods: Retrospective analyses of all VV ECMO support using Avalon®cannula (2014-2018).

Results: Eighteen patients with a median age and weight of 3.6 years (0.1-13) and 19 kg (4.2–50) respectively, received respiratory support using Avalon®cannula. Respiratory failure occurred due to viral or bacterial pneumonia in 7 (including 3 oncologic patients), septicemia in 3, a neoplastic disease in 4, near drowning in 2 and post heart-/chest-surgery in one patient each. VV ECMO was mainly installed at bedside under echocardiographic guidance. In 3 patients, the initial veno-arterial (VA) ECMO was successfully converted to VV ECMO after a mean of 5 days. In one patient, an initial VV ECMO needed conversion to VA ECMO after 12 days of support; in one patient a second VV ECMO run was needed. Successful weaning was possible in 16 patients after median support of 6 days (2-32). Two patients died due to their underlying disease. Overall survival to discharge was 83% (n = 15). Median ventilation time after VV ECMO withdrawal was 3 days (1-32), median ICU stay 16 days (5 – 64). Three patients suffered 4 major complications (all haemorrhagic): 1 haemopericardium due to cardiac perforation (needing sternotomy); 2 explorative thoracotomies for bleeding after lung biopsy and 1 spontaneous haemopericardium due to therapeutic anticoagulation. Cannula repositioning (3 patients) and change of oxygenator (one patient) were other notable events. 4/16 patients (25%) experienced thrombosis of the internal jugular vein. At a median follow-up of 166 days (0-1400) overall survival was 78%.

Conclusion: VV ECMO with Avalon®cannula provides a safe and versatile respiratory support in paediatric patient population, with excellent survival in reversible lung pathologies. Surgical complications during placement, haemorrhagic complications due to anticoagulation or thrombotic complications of the oxygenator or the internal jugular vein may occur. Once survived to discharge, the long-term survival is good.

P 2

3D models to plan complex surgery in intrathoracic malformation

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Background: To diagnose complex intrathoracic malformation in newborns, it is important to have imaging of the heart, vessels, airways and bones simultaneously. In critically ill patients, a reasonable way to receive all the information is an angiographic CT-scan. Because new Dual-Source CT-scan is extremely rapid and theoretically possible without sedation as there are no movement artefacts. Today's scanner can record cardiac images without slowing down heart frequency. CT-data can be used to construct a 3 D model to plan advanced surgery.

Questions/methods: How detailed can the cardiovascular system be rebuilt in 3D and segmented in stereolithographic format? What is the effective x-ray dose needed for a good quality image? How much contrast is needed? We used this method in 10 cases of newborns with complex congenital cardiac malformation to analyze our questions.

Result: 3D reconstruction models from CT-scan data is perfect to illustrate complex cardiovascular malformation such as TAPVR (total anomalous pulmonary venous connection) or lung perfusion by MAPCAS (Major aortopulmonary collateral artery) as known in pulmonary atresia. It is possible to reconstruct airways in detail until 3rd generation of bronchia. Bones, airways and vessels can be illustrated in one model and segmented with different colors, which helps to a better understanding of the anatomy for the whole team, even if they are not familiar with CT-scan reading.

P 3

Kawasaki disease presenting with phlegmonous adenopathy

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Case report: A 4 y.o. girl was referred with a h/o cervical adenitis non-responding to 48h oral co-amoxicilline. She was febrile (40°C) for 2 days with worsening general state, vomiting and unilateral submandibular adenopathy. On admission, inflammatory markers were raised (CRP 204 mg/L, WBC 27 G/L), US showed a cervical adenitis with no abscess. She was hospitalized and no improvement was observed despite 2 days IV co-amoxicilline. Cervical CT confirmed a phlegmonous adenitis and revealed a retropharyngeal phlegmon without abscess. Based on persistent fever and rising inflammatory markers (CRP 549 mg/L, ESR >140 mm/h, hyperleukocytosis 33 G/L with left-shift), we suspected atypical Kawasaki disease (KD), as only 2 criteria out of 5 were present. Diagnosis was supported further by additional investigations revealing anemia (Hb 82 g/L), elevated ASAT (106 U/L), hypoalbuminemia (22 mg/L) and gallbladder hydrops on abdominal US. IV immunoglobulins (2g/kg) and Aspirin (100 mg/kg/d) were administered. The following days, the patient developed reticular rash, strawberry tongue, bilateral conjunctivitis and hands edema. Echocardiogram revealed coronary vessels hyperechogenicity without dilatation and slight mitral and tricuspid insufficiency compatible with KD. Outcome was favorable; patient was discharged 4 days after IVIG and control CRP was <50 mg/L a week later.

Discussion: KD diagnosis requires: high fever (>5 days) and 4/5 of major criteria (cheilitis/strawberry tongue; polymorphous rash; bilateral conjunctivitis; extremities erythema/edema/desquamation; unilateral cervical lymphadenopathy). Incomplete form requires fever and only 2 or 3/5 criteria, making the diagnosis trickier. Additional biological tests (raised CRP and ESR, anemia, elevated liver enzyme, hypoalbuminemia, hyponatremia, thrombocytosis, abnormal lipid levels, sterile pyuria and proteinuria) and involvement of coronary arteries on echocardiography can be helpful. At first, our patient had only 1 clinical criteria, but 4 laboratory criteria and gallbladder dilatation that motivated prompt IVIG and Aspirin administration. Clinical evolution and echocardiogram confirmed the diagnosis of KD, justifying pursuit of Aspirin and cardiac follow-up.

Conclusion: KD should be considered in children with febrile cervical adenitis resistant to antibiotics. IVIG can be administered even in the absence of all KD criteria, as early diagnosis and treatment are mandatory to prevent cardiac complications.

P 4

An unusual case of anomalous left coronary artery from the pulmonary artery (ALCAPA)

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Objective: ALCAPA is a rare congenital anomaly of the coronary arteries with an incidence of approximately 0,021% in live births to 0,2% in autopsy studies. Its hallmark is the development of intractable heart failure during infancy due to myocardial ischemia, however, up to 15 percent of the children with ALCAPA can remain asymptomatic until adulthood. Nevertheless, if undiagnosed those children are at high risk for sudden cardiac arrest (SCA) or sudden cardiac death (SCD) due to arrhythmias, especially during exercise.

Case: We present the case of a 2 3/12-year-old boy who was admitted to hospital because of a pulmonary infection. Chest radiograph showed mild cardiomegaly. Further work-up revealed discrete ST-segment depression in the 12-lead ECG and echocardiography strongly suggested ALCAPA with retrograde perfusion of the LCA and circumflex artery through multiple prominent coronary fistulas and diastolic flow into the pulmonary artery. The anterolateral papillary muscle showed increased echogenicity, hence myocardial ischemia was assumed. The boy was referred to a tertiary centre. After confirming the diagnosis by angiography, the LCA was reimplemented into the aorta, which he tolerated well. Postoperative he developed a moderate insufficiency of the mitral valve. He was placed on betablockers and ACE-inhibitor and left the hospital 12 days after the operation.

Conclusion: Having no harmful effects in fetal life, ALCAPA presents predominantly in infancy due to perfusion of the left ventricle with desaturated blood and shunting from the coronary arteries to the pulmonary artery because of the fall of the pulmonary artery pressure below systemic pressure after birth. ALCAPA may be mistaken for common paediatric conditions and may even remain asymptomatic but at a high risk for SCA or even SCD. Therefore, awareness of this condition is essential for prompt recognition to enable surgical intervention to improve the prognosis for these children.

P 5

One cardiac defect, very different evolutions

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Ventricular septal defect (VSD) is one of the most common congenital heart disease and has previously been estimated to affect approximately 0.5% of all newborns. It can exist in isolation or as integral components of other cardiac anomalies. The classification depends of the position in the interventricular septum and of the sizes.

Case description: We describe different possibilities to correlation between similar echocardiographic VSD and clinical outcomes. We report 3 patients (P) with diagnosis of a medium-sized (6-8mm) perimembranous ventricular septal defect (VSD). We compare the evolution during six months (m). All were newborns at the moment of the diagnosis and had a good clinical conditions and normal ECG. -P1: No symptoms; no heart failure; large left atrium at 1°m; no dilatation of ventricles; no pulmonary hyperflow; no pulmonary hypertension; started diuretics at 1°m and finished at 5°m. - P2: symptoms at 3°m; no heart failure; large left atrium at 3°m; no dilatation of ventricles; shows pulmonary hyperflow; no pulmonary hypertension; started diuretics at 3°m and did not stop. - P3: symptoms at 1°m; heart failure at 3°m; showed large left atrium and dilatation of ventricles; pulmonary hyperflow; pulmonary hypertension; started diuretics at 1°m and then needed captopril; the corrective surgery was performed at 5°m. - After six months of regular checking: P1 had a little VSD, without symptoms, normal growth and normalization of left atrial dimensions. We discontinued the diuretics. P2 had a medium VSD, without symptoms, normal growth and large left atrium. Continued with diuretics, did not need surgery.

P3 had a medium VSD with symptoms of cardiac failure, pulmonary hypertension, dilatation of left atrium and both ventricles, stagnation of growth, treatment with diuretics and captopril and we had to delivery to cardiac surgery.

Conclusion: Clinical manifestation depends on the size of the defect, and on the relationship between systemic and pulmonary vascular resistances. Symptoms include failure to thrive, along with the manifestations of the increase of flow to the lungs. Diagnosis can be made by physical examination, and is confirmed by echocardiography. Follow-up is very important to recognise the different evolution and decide at the right time which treatment is the most appropriate. In infants with large shunts and persistent symptoms despite treatment with drugs, surgical closure is requested.

P 6

Three cases of pneumomediastinum without pneumothorax: a rarity?

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Introduction: Pneumomediastinum (PM) is a condition in which air is present in the mediastinum. Usually occurs in young people without an apparent precipitating factor or disease. It's very uncommon in children: in people admitted to a hospital with chest pain and dyspnea, 1/368 has a SPM. Other symptoms include laboured breathing, subcutaneous emphysema. It is often recognized on auscultation by a "crunching" sound timed with the cardiac cycle (Hamman's crunch). The diagnosis can be confirmed via chest X-ray or CT scanning of the thorax. Conservative treatment consisted of bed rest, oxygen therapy, and analgesics. The mean hospital stay ranged between 3 and 10 days.

Cases: We have three cases of spontaneous PM without pneumothorax (PNT): CASE 1: Female, age 15. Chest pain at presentation during activity, neck pain and weakness as related symptoms, Ghent nosology score positive, Marfan symptoms and genetic tests negative. Initial chest x-ray: negative, no hospitalization. -CASE 2: Male, age 15. Chest pain

at presentation during activity, neck pain as related symptom. Ghent nosology score negative, Marfan symptoms negative. Initial chest x-ray: positive. Three days of hospitalization. -CASE 3: Male, age 16. Chest pain at presentation during resting time, subfebrile temperature as related symptom. Ghent nosology score negative, Marfan symptoms negative. Initial chest x-ray: positive. Two days of hospitalization.

Discussion: Our cases are negative for Marfan syndrome (MS), even case 1 with familiarity. Literature reported that MS is a risk factor, but a rare cause of PM. All the cases we had stayed in hospital for less time than the period expressed in literature data.

No one of our patients had PNX with PM. Many studies confirm our results, reporting that PNX is associated only in 11.6%. In one case the first interpretation of X-ray was negative. Only a second one and CT scan confirm the diagnosis. Bibliography shows the same data, confirming that the initial chest reading could miss the diagnosis.

Conclusions: PM is a rare condition in children, but rarely linked to PNX and potentially life threatening. We have to think about it everytime we have chest pain, dyspnea, neck pain, in adolescents, appearing during sports or physical activity. We also keep in mind that the initial X-ray could be difficult to read and we could miss the problem.

P 7

Feeding difficulties, failure to thrive and systolic murmur in a toddler: Think of vascular anomalies!

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Introduction: Kommerell diverticulum with left aberrant subclavian artery is a rare congenital vascular structural variation. It can be asymptomatic or symptomatic owing to mass effect. This very rare vascular anomaly, which is often diagnosed beyond the first year of age, presents with unspecific symptoms such as wheezing, dysphagia or failure to thrive.

Case: A 2-year-old boy was referred to our clinic because of feeding difficulties and failure to thrive. The clinical examination revealed an underweight and slightly meteoric abdomen. The laboratory tests were normal, especially no signs of a resorption disorder such as celiac disease were found. In connection with a 2/6 systolic murmur in cardiac auscultation, an echocardiography was performed which showed an aortic arch with suspected A. lusoria. A barium swallow showed an increase of the oesophageal narrowing at the level of aortic arch, being suggestive of extrinsic compression. MRI examination of the thorax/heart revealed a complete vascular ring, compatible with a right aortic arch. Further a Kommerell diverticulum and a ligamentum arteriosum with impression of the oesophagus in the middle third were described. These findings explained the feeding difficulties and failure to thrive. The patient's discomfort indicated operative repair of this condition. The boy was therefore transferred to the pediatric heart center. After the operation, which consisted of removing the Kommerell diverticulum, implantation of the left subclavian artery to the left carotid artery and severing the arterial ligament, the patient's eating behaviour normalized and the boy tolerated diet without dysphagia.

Conclusion: Diagnosis of vascular ring with Kommerell diverticulum is difficult and may be delayed, as it often presents with unspecific symptoms. In our case, feeding difficulties, dysphagia and failure to thrive prompted further investigations which finally led to the correct diagnosis.

P 8

Rebound of involuted infantile hemangioma following administration of selective β -2 adrenergic agonist

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Background/objectives: Since the discovery of propranolol in the treatment of infantile hemangioma (IH), there has been ongoing investigation on the mechanisms by which β adrenergic receptor-blockers regulate hemangioma cell proliferation. We report the case of an involuted IH following propranolol therapy that exhibited a full and rapid rebound during the intravenous administration of salbutamol, a selective β 2 adrenergic agonist, for an episode of severe obstructive bronchitis.

Case report: A 9-week old female was referred for the evaluation of a rapidly growing IH on her left shoulder. Physical examination revealed a focal hemangioma measuring 2.0cm x 2.0cm x 0.8cm. Propranolol was initiated and marked involution was achieved after 14-months of therapy. At the age of 26 months, she was admitted for an acute episode of severe obstructive bronchitis. She required rescue treatment with continuous intravenous (IV) salbutamol (2mcg/kg/min), IV-methylprednisolone (2mg/kg/day), ipratropiumbromide inhalation and was mechanically ventilated for 5 days. Within 24 hours a significant increase in redness and remarkable swelling of the IH were observed. These changes persisted during IV-salbutamol and methylprednisolone treatment and reduced somewhat during the following days after switching to salbutamol inhalation therapy. Re-evaluation of the IH 2 months later showed a flat lesion with residual redness.

Conclusions: We were intrigued to see a very rapid and marked swelling of the IH with the initiation of systemic β AR-agonistic treatment with salbutamol at the age of 26 months, despite concurrent systemic corticosteroid therapy. To the best of our knowledge, this is the first demonstration of IH regrowth triggered by a β 2-AR agonist. Yet, salbutamol is widely prescribed in young children for the treatment of acute bronchospasms. Apart from demonstrating the clinical relevance of β 2-agonist effects in IH, this case raises awareness of the potential proliferative response of IH to β 2 AR agonist treatment such as salbutamol.

P 9

An incomplete case of Kawasaki disease in a 10 month-old patient

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Kawasaki disease (KD) is a self-limiting systemic vasculitis most often occurring in children between 1 and 5 years of age and associated with a high risk of development of coronary artery aneurysms. The diagnosis depends on clinical manifestations. Incomplete or atypical cases are difficult to recognize, especially in infants. Case: A 10-month-old infant was admitted to our paediatric Department for high intermittent fever for 9 days, cough and runny nose. His past medical history was silent. Physical examination revealed a hyperaemic oropharynx with tonsillar hypertrophy, a mild perineal maculopapular exanthema, and an adenopathy of the right anterior neck-triangle. He was febrile (39.5°C), with normal heart and breath rates. A blood test was performed that revealed: Hb 104 g/L; PLT 593.000x10⁹/L; WBC 15.8x10⁹/L; ESR 60 mm/h and CRP 35 mg/L. Hepatic-renal function, albuminemia, urine dipstick, chest X-ray and abdominal ultrasound were normal. PCR for respiratory viruses was positive for Rhinovirus and Enterovirus. The persistency of high intermittent fever prompted us to consider other diagnosis in particular an incomplete form of KD. Echocardiography showed dilatation of the right coronary artery with border-line left coronary measures. At 10th day of fever a therapy with intravenous immunoglobulins (IVIG) in association with acetylsalicylic acid was started. Because of fever persistency a therapy with methylprednisolone was demanded with clinical response. The child was discharged at the 8th day of cure in good health condition. After 2 weeks echocardiography showed normalization of the aneurysms.

Discussion: The diagnosis of incomplete KD should be considered in any infant or child with prolonged unexplained fever, 2 or 3 of the principal clinical findings, and compatible laboratory or echocardiographic findings. IVIG and ASA are the mainstay of initial treatment; the role for additional primary therapy in selected patients is discussed. Nevertheless, approximately 10% to 20% of patients do not respond to initial treatment and additional therapies, such as corticosteroids or monoclonal antibodies, are needed.

Conclusions: KD is a potential life-threatening disease frequently presenting with sneaky signs and symptoms. A prompt diagnosis and treatment is capable in reducing the incidence of possible important co-morbidities such as coronary disease.

P 10

Challenges in the diagnosis of heart failure in a 3-month year-old during bronchiolitis epidemics

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Introduction: Heart failure is a rare condition in children which is usually caused by a congenital heart disease or a cardiomyopathy. Presenting

symptoms may be very non-specific, and can easily be mistaken for a respiratory disease, especially if presented concomitantly with signs of respiratory tract infection, such as fever, cough and rhinorrhea.

Case summary: We report the case of a 3-month old female infant with an unremarkable past medical history; she presented twice to the Pediatric Emergency Room with fever and cough in the two weeks prior to admission. It was during the Swiss winter season, coinciding with the annual epidemic of RSV (respiratory syncytial virus). A clinical diagnosis of bronchiolitis was made. Upon her third presentation, feeding and growth were suboptimal without manifest failure to thrive. Respiratory distress remained though the fever had resolved. She was hospitalised for supportive care, without any oxygen requirements on admission. The PCR testing for RSV was negative. Clinical worsening appeared on the second day of hospitalization, with decreased blood oxygen saturation, significant respiratory distress with central cyanosis. On cardio-pulmonary auscultation, bilateral end-expiratory wheezes and rales were noted without any heart murmurs or gallop. No edema or hepatosplenomegaly were found. A chest x-ray showed cardiomegaly and bilateral alveolar infiltrates. A working diagnosis of heart failure with worsening possibly superinfected bronchiolitis was considered. Given the respiratory and hemodynamic instability, the patient was transferred to the Pediatric Intensive Care Unit, at the Cantonal University Medical Center. Upon arrival, an echocardiography showed a left ventricle ejection fraction of 10% with dilated right and left ventricles. NT-proBNP value was measured at 160.000ng/L and troponins were low. The cardiac MRI did not reveal any evidence suggesting myocarditis. Intensive medical management was immediately started for heart failure including levosimendan treatment in the context of a suspected congenital dilated cardiomyopathy. The causal etiology remains as of yet undetermined.

Discussion: This case illustrates the challenges encountered in the diagnosis of heart failure in young infants. Performing a thorough history taking, assessing for the unusual and periodically reviewing the differential diagnosis in the non-improving child remain fundamental key points in order to prevent late diagnosis and poor outcome.

P 11

Herpes zoster meningitis in two immunocompetent pediatric patients without a rash

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Varicella zoster virus (VZV) reactivation is commonly related with shingles and less frequently with neurologic manifestations in mostly immunocompromised patients. We report two cases of previously healthy children with aseptic meningitis as a result of reactivated VZV without a rash.

Cases

A 12-y-old male presented with worsening frontal headache since 6 days with photophobia and vomiting. He had fever (38.7°) once on the 3rd day of symptoms. He denied history of neck stiffness, skin rash. Normal examination except lumbar pain on extreme neck flexion, without stiffness. His blood tests showed no anomaly. Lumbar puncture revealed 733 WBC/mm³ (92% lymphocytes), protein of 0.81g/l and glucose of 2.5mmol/l. PCR of the CSF tested positive for VZV and negative for other viruses. Gram stain and CSF cultures were negative. His mother revealed he had varicella at the age of 9 months. Treatment with intravenous acyclovir 1500mg/m²/day in 3 doses (13mg/kg/dose) for 14 days was initiated. His symptoms resolved 48h later and the patient was discharged at day 13 with no neurological sequelae. – A 11-y-old male was admitted with a 5-day history of severe headache, tiredness and a 2-day history of abdominal pain and vomiting (once per day in the morning). There was no history of fever, photophobia, phonophobia, neck stiffness, skin rash. Normal examination except lumbar pain on extreme neck flexion, without stiffness. Cerebral MRI was normal. His blood tests showed no anomaly. Lumbar puncture revealed 613 WBC/mm³ (lymphocytes 94%), protein of 0.65g/l and glucose of 2.8mmol/l. PCR of the CSF tested positive for VZV and negative for other viruses. Gram stain and CSF cultures were negative. Varicella infection during childhood was reported by his parents. Treatment with intravenous acyclovir 20mg/kg 3 times a day was initiated. Doses were adapted after 48h because of transient renal toxicity. The patient was discharged on day 12 with no neurological sequelae and normal renal function.

Headache is a frequent reason for consultation in emergency room. Aseptic meningitis due to VZV reactivation is unknown and rarely described in pediatric patients even if the use of PCR during the last decades has increased it's early diagnosis. We highlight the possibility of

this diagnosis in front of persistent headache even without «red flags». In the immunocompetent pediatric population the outcome is good with acyclovir treatment even started after 5 days of symptoms.

P 12

F. tularensis: an emerging disease in Switzerland?

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Francisella tularensis is a gram negative coccobacillus, which is a well known cause of zoonosis of the northern hemisphere. It is transmitted to humans by direct contact with infected animals (rabbits, rodents, domestic cats and dogs), tick bite, ingestion of contaminated water and meat or inhalation of contaminated aerosols. Person to person transmission has not been reported. Tularemia represents a large variety of syndromes; the most common presentation is the ulceroglandular syndrome.

We present a case of a 9 years old Swiss girl with no past medical history except a Lyme disease in July 2018 treated by 10 days of Amoxicillin. She lives in a farm with horses, cats and turtles and she went in Turkey in August 2018. She reported no recent tick bite. The 23rd of October, she developed fever for 4 days and sacral pain. A few days after fever resolution, a sacral erythematous plaque was noted. November the 6th, she consulted for inguinal bilateral lymph nodes enlargement with persisting sacral lesion. Amoxicillin po was prescribed without improvement. The 13th, the sacral lesion begun to erode with low grade fever. Amoxicillin was switched for Amoxicillin+clavulanic acid iv without clinical success. A pelvic MRI showed no sign of bone involvement but inguinal and pelvic lymph nodes enlargement without abscess. EBV, CMV, HIV, Bartonella sp, Brucella sp, Toxoplasmosis sp serology was negative. Interferon-Gamma Release Assays came also negative. An excisional biopsy of an inguinal lymph node was performed on the 23rd. The serology for *F. tularensis* was added on the serum and Ciprofloxacin was started. *F. tularensis* IgM and IgG came back positive as well as a PCR on the lymph node confirming the diagnosis of tularemia (culture remained negative). The evolution after 10 days of ciprofloxacin was excellent. As often, the source of infection could not be determined but might be environmental. We suspected the sacral lesion to be the bacterial entry point even if atypical (non exposed skin). In Switzerland, since 2004, it is mandatory to report the *F. tularensis* infection (in humans and animals). The incidence has been rising fourfold between 2010 and 2017 with a disparity in cantonal distribution.

In conclusion, Tularemia is emerging in Switzerland. This diagnosis should be systematically discussed in case of lymphadenopathies after a tick bite or in case of adenitis not responding to betalactamin treatment.

P 13

A healthy boy with a unilateral parapharyngeal abscess

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Introduction: Parapharyngeal abscess is a deep neck space infection which involves the space that extends from the skull base, to the greater cornu of the hyoid bone occupying the space between the muscles of mastication and deglutition and which includes the internal maxillary artery, the ascending pharyngeal artery, the pterygoid venous plexus and lymph nodes. This report describes a case of a parapharyngeal abscess secondary to an upper airway infection.

Methods: A 4-year-old boy, resident of the Broye District in the Canton of Vaud, presented to our ER three times in six days after the beginning of symptoms due to a possible viral infection. Within 72 hours of the early symptoms, this child developed conjunctival purulent discharge with unilateral left palpebral oedema, fever, neck pain with neck stiffness, early left acute otitis media but no meningitis signs. Despite symptomatic and local treatment, fever followed along with worsening neck pain. Significant neck pain extending to the thoracic region with sweating and high fever temperature were noted. He was then hospitalised for clinical surveillance. Cefuroxime IV was instated and a brain/spine MRI was realised which revealed an extended left prevertebral abscess. A transfer to the university hospital of Lausanne was initiated where an immediate tonsillectomy and a surgical simple and intra-oral drainage were realised twice during his hospitalisation due to the persistence of neck stiffness. Antibiotics (Amoxicillin-clavulanic acid firstly IV and then per os) were administered for a total of 15 days as clinical improvement was gradually observed. Further investigations (repeat of an MRI, blood tests and US) were performed during his hospital stay and his postsurgery follow-up as

well. Full recovery was thereafter noted with no sequelae found on his ENT follow up (>15 days)

Results: Parapharyngeal abscesses arise secondary to oropharyngeal infections that spread either by direct continuity or by lymphatic drainage. The most common causes include Group A Streptococcus, *S. aureus* and anaerobic bacteria. It occurs predominantly to male (2:1) with seventy percent of the patients are younger than six years of age.

Conclusion: Based on good clinical outcomes and low incidence of complications, the present case report suggests that good patient history taking, antibiotic therapy complemented with a timely surgical treatment, is a valid treatment option in refractory parapharyngeal abscesses.

P 14

Rat bite fever – arthritis caused by *Streptobacillus moniliformis* in two brothers

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Introduction: Rat bite fever (RBF) is a rare bacterial zoonosis which is characterized by arthritis, fever and a rash. RBF is due to infection with *Streptobacillus moniliformis* (*S. moniliformis*) and rarely with *Spirillum minus*. RBF is usually transferred by domestic rats.

Case report: A 3 year old boy was admitted to our hospital due to fever and limited movement of his right arm. The physical examination showed a painful wrist swelling and a crusted rat bite wound on the dorsal hand. Laboratory analysis showed elevated erythrocyte sedimentation rate (ESR) and C-reactive protein (CRP). Despite no signs for joint effusion in the ultrasound examination, septic arthritis was suspected and an empiric antimicrobial treatment with amoxicillin/clavulanic acid was initiated. 1 day later, a painful knee swelling appeared. Sonographically minimal joint effusion was detectable in the recessus suprapatellaris. The diagnosis of septic arthritis of the wrist was confirmed by MRI. The history of the rat bite raised a high index of suspicion that *S. moniliformis* could be the causative pathogen. This hypothesis was reinforced when his 8 year old brother presented with fever and painful right wrist swelling three weeks later. Diagnostic tools (ESR, CRP, MRI) showed resembling results and antimicrobial treatment with flucloxacillin was initiated. Both brothers showed fast clinical response after established antibiotic treatment. The saliva of 2 of the 5 pet rats was tested positive for *S. moniliformis* in a veterinary laboratory. We performed a blood serology for *S. moniliformis* in the younger brother and detected an increasing Mean Fluorescence Intensity (MFI) from 552 to >4000 6 weeks after infection.

Discussion: 1 of 10 bites of a rat colonized with *S. moniliformis* results in an infection in humans, with a mortality rate of up to 13% if untreated. Having contact to contaminated body fluids is sufficient to cause an infection. Possible complications are fulminant sepsis, meningitis, hepatitis, pneumonia, myocarditis, pericarditis, osteomyelitis and arthritis. *S. moniliformis* is difficult to culture and joint fluids remain often sterile. *S. moniliformis* Single Plex Serology analysis is performed in animal samples but is not an accredited test method in humans.

Conclusion: If a patient keeping domestic rats presents with fever and arthritis, RBF should be considered as differential diagnosis.

P 15

Dilemma: enterohaemorrhagic *E. coli* associated haemolytic uremic syndrome and *E. coli* associated pyelonephritis in a 5-week-old boy

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Introduction: *E. coli* is part of the normal colonic flora, but may cause infections such as diarrhea, urinary tract infection (UTI) or haemolytic uremic syndrome (HUS) depending on the pathogenic strain. While UTI is common, infection with Enterohaemorrhagic *E. coli* (EHEC) and HUS occurs in 2–3/100,000 children <5 years of age in the US and Europe.

Case report: A 5-week-old male infant presented to our emergency Department with fever, watery diarrhea and vomiting for the last 4 days. He was in a reduced general condition and irritable with delayed capillary refill (3–4 seconds), tachycardia (184/min.) and signs of moderate dehydration. Investigations showed inflammation (Lc 17.8 G/L, CRP 156 mg/L) and severe metabolic acidosis (pH 6.99, HCO₃- 11 mmol/L, Lactate 6.7 mmol/L). For a working diagnosis of gastroenteritis with moderate dehydration and possible sepsis with compensated shock, parental fluids and antimicrobials (Amoxicillin and Amikacin) were started and he was transferred to intensive care. Initial stool PCR was negative for Salmonella,

Shigella, Campylobacter, Yersinia. Viral PCR was negative for Adenovirus and Rotavirus. The urine culture grew *E. coli* 10⁶ (non-EHEC), whereas blood culture was negative. Concomitant pyelonephritis was suspected and antimicrobial treatment adjusted according to the antibiogram. After 2 days, renal insufficiency (creatinine 67 µmol/l), hemolysis (Hb 42 g/l, LDH 4626 U/l) and thrombocytopenia (Tc 46 G/l) occurred, consistent with HUS. Complement C3/4, ADAMTS-13, homocystein and renal ultrasound were normal. Stool PCR examination was extended for pathogenic *E. coli* strains revealing EHEC. On conservative therapy including 3 blood transfusions, the patient rapidly recovered and was discharged after 10 days without any signs of renal impairment. The further follow-up has been uneventful.

Conclusion: EHEC related HUS in neonates and very young infants is rare. Watery diarrhea may be a sign of sepsis or viral enteritis. EHEC, however, may also cause this symptom. Bloody diarrhea is not an obligatory sign for diarrhea-positive HUS, thus misleading the clinician. The common stool PCR for bacteria misses pathogenic EHEC, if not specifically ordered. Antimicrobial treatment of EHEC associated enteritis is not recommended as it may increase expression of Shiga toxin and severity of HUS. We however, suspected pyelonephritis as second, coincidental infection and continued antimicrobial treatment.

P16

Accuracy of ICD-10 coding for sepsis and organ dysfunctions in children with blood culture-proven sepsis in Switzerland, results from the prospective Swiss Pediatric Sepsis Study

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Background: The recent resolution on sepsis by the World Health Organization urges member states to better characterize the burden of sepsis. Sepsis coding based on the International Statistical Classification of Diseases and Related Health Problems (ICD-10) is widely used, however, there is a lack of paediatric data assessing accuracy of ICD-10 sepsis coding. In addition, the recent change in sepsis definitions implies a need for reliable coding of organ dysfunctions. We analysed accuracy of sepsis diagnosis based on ICD-10 codes in a prospective cohort of children with sepsis.

Methods: We performed a multicentre, prospective cohort study at ten paediatric hospitals in Switzerland from 01.09.2011 to 31.12.2015, recruiting children younger than 17 years with blood culture-proven sepsis. For this analyses, we excluded prematurely born neonates and infants less than 7 days old at sepsis onset. We used 2005 consensus definitions – based on prospectively collected clinical and laboratory data – as the reference standard to define sepsis and organ dysfunctions. ICD-10 codes were extracted from mandatory official hospital discharge data related to the sepsis episodes at 8 study sites.

Results: Of 679 episodes with blood culture-proven sepsis recruited in the participating hospitals, 455 (67%) were classified as sepsis by ICD-10 codes. 305 (45%) had at least one organ dysfunction according to 2005 consensus criteria and 249 (37%) according to ICD-10 codes; the accuracy of ICD-10 codes for organ dysfunction was 69%, sensitivity 56%, and specificity 79%. In 163 (24%) episodes multi-organ dysfunction syndrome (MODS) was present as per 2005 consensus criteria and in 98 (14%) with ICD-10 codes; the accuracy of ICD-10 codes for MODS was 64%, sensitivity 39%, and specificity 93%. Sensitivity of ICD-10 codes was especially poor for central nervous system (6%), hepatic (7%), respiratory (26%), and hematological dysfunction (30%), and only fair for renal (45%) and cardiac dysfunction (48%).

Conclusion: Compared to the reference standard, ICD-10 codes underestimated incidence of sepsis, organ dysfunctions, and MODS and did not reliably depict organ dysfunctions in children with blood culture-proven sepsis.

P 17

A case of neonatal acute parotitis in a 23-day-old baby

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Introduction: Salivary gland infections are rare in neonates; they most commonly involve the parotid glands. Neonatal suppurative parotitis classically presents with unilateral parotid swelling and variable degrees of local signs of inflammation. *Staphylococcus aureus* has been reported to be the most frequent pathogen causing neonatal suppurative parotitis, but both Gram-positive and Gram-negative microorganisms may be involved. Treatment usually consist of intravenous antibiotic therapy; rarely surgical incision is needed.

Case report: A 23-day-old full-term, breast-fed, female neonate presented to our hospital because of left preauricular swelling and erythema. She had a 1-day history of irritability and restlessness. She had been born by normal vaginal delivery after an uneventful pregnancy with a birthweight of 2550g (P3) and a head circumference of 32cm (P2). Because of the microcephaly a work-up for cytomegalovirus infection had been made previously, which revealed that she suffered from congenital cytomegalovirus infection. On examination, the infant was very irritable and crying. Her rectal temperature was 37.9°C and she had an indurated and warm swelling over the left parotid gland. Laboratory tests revealed raised inflammatory markers with a hemoglobin of 14.5g/dl, white blood cells 27.7 10⁹/l, and CRP 21,2mg/l. Ultrasound examination of the left parotid and pre-auricular region showed an enlarged parotid gland with a hypodense area of 31mm x 17mm. Color Doppler ultrasonography revealed an increased vascularity and a broadened Stensen's duct. Based on clinical presentation and ultrasound findings, she was diagnosed with neonatal suppurative parotitis. She was empirically started on intravenous amoxicillin/clavulanic acid. A swab of the pus was collected for microbiological analysis. Because of a worsening general condition, we transferred the girl to the University Children's Hospital in Bern. A full septical workup revealed methicillin sensitive *S. aureus* bacteremia. The child completely recovered with conservative therapy and has been well on follow-up examinations.

Conclusions: Although acute suppurative parotitis is rare it should be suspected in case of neonatal facial swelling. The diagnosis is clinical and supported with the use of ultrasonography. Treatment with parenteral antibiotics is usually effective and surgical intervention only rarely needed. The prognosis seems to be excellent with rare complications and recurrence is uncommon.

P 18

A bitter raspberry

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Background: Yaws, also known as *Frambesia tropica* (deriving from "framboise", french name for raspberry), is an endemic, non-venereal treponematosis caused by the bacterium *Treponema pallidum* subspecies *pertenuis*. It is spread by skin-to-skin contact and usually occurs in tropical regions and overcrowded communities, mostly affecting children under 15 years of age with limited access to basic amenities, such as water and sanitation, as well as health care.

Case report: We report a case of a 16-year-old boy who presented with maculopapular and pustular lesions over the back of his foot and lateral malleolus, appeared during the last 3 months prior to admission. Originally from Guinea, the patient travelled during the last year through Mali, Burkina Faso, Niger and Libia. No other symptoms were reported. For further evaluation a skin biopsy was collected. The histological result demonstrated epidermal hyperplasia and papillomatosis, with focal spongiosis, neutrophils accumulate in the epidermis and a dense dermal infiltrate of plasma cells, confirming the diagnosis of Yaws. The patient was treated with a single dose of Benzylpenicillin IV. After 3 weeks he assisted to a complete healing of the lesions.

Conclusion: Clinical manifestations of Yaws occur in three distinct stages. A primary phase characterized by a single "raspberry like" lesion (or mother yaw), appearing approximately 3 to 4 weeks after infection at the site of inoculation; a secondary phase which consists in widespread dissemination of smaller skin lesions containing high numbers of treponemes; a tertiary phase which occurs in approximately 10% of untreated persons about 5 to 10 years after the initial infection. Tertiary yaws is characterized by destructive cutaneous lesions accompanied by bone and joint deformities. The recommended treatment is a single intramuscular dose of benzathine benzylpenicillin in a dosage of 1.2 million units for patients older than 10 years of age, or 6 million units for patients younger than 10 years of age.

P 19

Fever and shoulder pain: non always arthritis

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Background: Liver abscesses (LA) in children are infrequent in industrialized countries. The vast majority is represented by pyogenic LA and *Staphylococcus aureus* is the most common causing pathogen worldwide. Ultrasonography and computed tomography are widely used as diagnostic techniques. The gold standard of treatment of LA in children is still discussed.

Case description: We report the case of a previous healthy 8-year-old boy who presented to our emergency department with persistent fever for 6 days despite empirical oral antibiotic therapy and a newly presented right shoulder pain. Laboratory findings revealed high C-reactive protein and neutrophilic leukocytosis. Clinical examination was normal. To better evaluate the shoulder pain, a shoulder ultrasound and a Chest X-ray were done, both no pathological findings. The abdominal ultrasound showed instead an hepatic mass, confirmed at the MRI as a well demarcated, mostly cystic mass of 7.5 cm. Nevertheless transaminase levels were only few increased. Serologies for *Echinococcus* were negative and serum levels of α fetoprotein and β HCG were in range of normality. In order to exclude a liver cancer, a liver biopsy was performed, showing an hepatic abscess. The culture grew positive for *S. Intermedius* and *Aggregatibacter aphrophilus* as causative pathogen. No immunological deficits were found. A conservative treatment with intravenous antibiotic therapy was decided but because of failure, a percutaneous drainage was also necessary. Complete recovery was observed after 6 weeks and the follow up demonstrated a total resolution at the US after 3 months.

Conclusion: LA clinical presentation can be misleading and they could also develop without any predisposing risk factor. The intravenous antibiotic treatment is crucial for the management, but depending on the size of LA a percutaneous drainage or even a surgical approach may be necessary, so that an interdisciplinary approach is essential for the management of these patients.

P 20

Tularaemia – When to suspect? How to treat? Should tularaemia still be considered a rare disease in the southeast of Switzerland?

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Introduction: Tularaemia is considered a rare zoonotic disease. It is caused by the gram-negative rod *Francisella tularensis*. BAG data until 2016 report a national incidence of as low as 0.43/100'000/year. However, in the last 3 years incidence has risen to 4-fold. Timely diagnosis and antibiotic treatment with gentamicin or ciprofloxacin allow for a favourable course in the majority of cases. Here, we describe the clinical course of three cases of tularaemia diagnosed at our clinic within the last year, and we review current literature on tularaemia.

Case series: We report on 2 cases of glandular and 1 case of ulceroglandular tularaemia. All 3 patients presented with unspecific lymph node enlargement. Patients' age was 4 to 5 2/12 years. Mean time interval from empiric to targeted antibiotic treatment was 20 days (7 to 28). Mean duration of targeted antibiotic treatment was 47 days (14 to 77). One patient had recurrent tularaemia. Finally, all patients proceeded to surgery.

Discussion: Diagnosis may be delayed due to initially unspecific clinical signs. In addition, specific antibodies are not detectable before the second week of disease. Causes for the rise in incidence remain unclear.

Apart from an actual increase in patient numbers there might be heightened physicians' awareness.

Take home message: Tularaemia is still a rare disease in south-eastern Switzerland despite its recently rising incidence. Diagnosis is based on clinical suspicion. When targeted antibiotic therapy is delayed or when complications occur surgical intervention is indicated.

P 21

Metabolic control in children and adolescents with type 1 diabetes followed at the University Children's Hospital of Bern

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Background: Good glycemic control prevents long-term complications of microvascular and macrovascular diseases in type 1 diabetes (T1DM). We aimed to investigate whether our patients had A1c values <7.5% as recommended by ISPAD and how therapy modality, duration of diabetes and pubertal status affected the metabolic control of our patients. We also set out to compare our quality of care with our results of 2008 and with other published data.

Methods: In 2017/18, we enrolled all patients with T1DM who were followed by the outpatient clinic of the University Children's Hospital Bern over a period of 6 months in an observational cross-sectional study. Each patient was assessed once during the observational period, including demographic and clinical data (sex, age, diabetes duration, pubertal status, insulin treatment modality, use of continuous glucose monitoring (CGM), A1c levels).

Results: 160 patients participated in the study, 41% (n = 82) were boys and 49% (n = 72) were girls. Patients had a mean age (SD) at time of visit of 11.4 (3.4) years (range 2-17 years) and a mean duration (SD) of diabetes of 4.8 (3.5) years (range 1-16 years). Most patients, 63% (n = 100) received functional insulin treatment, 29% (n = 47) used insulin pump and 8% (n = 13) injected insulin on multiple times per day (twice-daily/three-dose). CGM devices were used by 43% (n = 68) of patients in their diabetes management. Mean A1c was 8% and 71% had HbA1c >7.5%. Compared to results from our hospital from 2008, A1c was slightly higher (8% vs 7.6%), but more patients had diabetes for >2 years (80% vs 47%). Patients with T1DM duration >2 years had more often HbA1c levels above 7.5 % than patients with duration <2 years (p <0.001). A1c values were significantly lower (p <0.05) in patients using CGM devices (7.8% vs 8%). A1c values did not differ between prepubertal and pubertal patients or insulin treatment modality (functional insulin treatment, insulin pump or multiple daily injections).

Conclusion: The overall glycemic control was poorer 2017/2018 than in our study from 2008. This may be due to the higher percent of patients with diabetes duration >2 years, thus with more patients out of the remission phase. Patients wearing CGM devices performed better. Unfortunately, our patients in Bern did not reach the target A1c set by ISPAD, similar to results to other diabetes centres in Europe and the United States. This highlights the importance of regular consultations and extended use of CGM.

P 22

Successful treatment with enzyme replacement therapy in a girl with severe infantile Hypophosphatasia

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Background: Infantile Hypophosphatasia (HPP) is an inborn error of metabolism characterized by low serum alkaline phosphatase activity caused by loss-of-function mutations within the ALPL-gene encoding the tissue nonspecific isoenzyme of ALP (TNSALP). TNSALP controls skeletal and dental mineralization by hydrolyzing inorganic pyrophosphate, a potent inhibitor of bone mineralization. Patients develop substantial skeletal disease, failure to thrive, and sometimes vitamin B6-dependent seizures before 6 months of age. Without treatment, HPP results in 50–100% mortality, typically from respiratory complications.

Presenting Problem: We present a 3 months old girl with infantile HPP caused by 2 heterozygous mutations in the ALPL gene. At the age of six weeks she presented with a lack of weight gain because of vomiting and respiratory insufficiency. Clinical investigations showed rhizomelia of the upper arms and femora, short stature, broad nose bridge, high forehead, a bulged fontanelle and muscular hypotension. A single cerebral seizure terminated spontaneously. Laboratory examinations revealed a very low

serum ALP activity and a high urinary excretion of phosphoethanolamine. Radiographic findings include hypomineralization with cup-shaped distensions of the metaphysis and irregular zones of ossification.

Clinical Management: Starting enzyme replacement treatment 2 mg/kg s.c. every other day was associated by a supportive therapy with oxygen, enteral nutrition through nasogastric tube, physiotherapy and supplementation of calcium, pyridoxine and analgetics. As a result of therapy x-rays showed an increase of bone mineralization. Stabilization of the chest wall led to a normal breathing pattern without need of oxygen support after 8 weeks. After improvement of vomiting tube-feeding could be weaned after 4 weeks with good weight gain. Muscular strength and neurological function improved also.

Discussion: Infantile hypophosphatasia is extremely rare and may be life threatening. Our case demonstrates that treatment with the recombinant enzyme therapy led to an improvement in muscular hypotonia, neurological problems and skeletal mineralization and therefore, respiratory function, growth and weight normalised.

P 23

Vomiting without diarrhoea – think well!

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Case: An 8-year old child known for cartilage hair hypoplasia diagnosed at 5-mo of life and treated with allogeneic unrelated hematopoietic stem cell transplantation (HSCT) at 11-months of age. The unrelated HLA 8/10-HLA-matched unrelated cord blood transplantation had been performed as consequence of severe combined immunodeficiency. At the time of presentation, he had been off immunosuppressives for more than 7 years with good immune reconstitution with 100% donor chimerism and no hints for chronic graft-versus-host-disease (GvH). He was hospitalized for iterative vomiting, dehydration (10%) fever and worsening condition. Blood tests revealed hyponatremia (Na 124 mmol/L), ketosis and raised CRP (121 mg/L). Blood glucose and potassium were normal. IV rehydration was started and Na normalised. Stool cultures were negative. Cortisol (C 325 nmol/L) and urinary Na excretion (uNa <60 mmol/L) were interpreted as normal. After discharge intermittent vomiting persisted in an otherwise well looking child. Mother reported salt cravings with tiredness, excessive night sweating and secondary enuresis but he kept normal daily activity. Fluid intake was 1L/day. Moderately bronzed skin has been noticed for several months. Repeated investigations showed constantly hypoNa (134 mmol/L) whereas K, blood pressures were normal. C at 8h (188 mmol/l) followed by Synacthen® test (peak C 117 mmol/L) confirmed diagnosis of adrenal insufficiency (AI). High serum ACTH (1693 pmol/L [N <10.3]) supported adrenal origin and explained bronzed skin. Increased anti-21-OH (1280 U/L) confirmed autoimmune (Ai) origin of AI. A substitutive gluco-/mineralocorticoid treatment (CT) was started leading to resolution.

Clinical lessons and conclusion: Chronic fatigue and vomiting may be suggestive of AI onset. The presence of abnormal skin pigmentation in Ai-AI can be a helpful guide to the diagnosis. Basal C may be misleading and ACTH test is, therefore, mandatory to confirm diagnosis. HyperK, hypotonia, metabolic acidosis and hypoG are not necessarily present before decompensation. Secondary AI after HSCT has been described but specially after long term CT or GvH reaction. In this case, the interval between HSCT and AI without other pituitary deficiency would have been unusual. Late Ai manifestation after HSCT is suspected. Thyroiditis after HSCT has been reported but at our knowledge Ai-AI has never been described.

P 24

Diabetic ketoacidosis in adolescent with MCAD deficiency

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A 13-year-old Teenager with MCAD deficiency presented to the paediatric emergency Department with two days of fatigue, mild frontal headache and diffuse mild abdominal pain. The patient had a normal examination apart from mild pharyngitis. Due to the risk of hypoglycemia in patients with MCAD, a capillary blood gas analysis was done. The capillary blood gas showed the classic picture of a diabetic ketoacidosis with a metabolic acidosis (pH 7.229, pCO₂ 2.58kPa, HCO₃ 8.1 mmol/l, Anion Gap 27.1 mol/l, Lactate 1.2 mmol/l) and a high glucose-level of 26 mmol/l.

A marked ketonuria as well as glucosuria was shown in the urine test strip, thus confirming the diagnosis of diabetes mellitus. The patient showed a severe ketoacidosis despite his MCAD deficiency. On reviewing the patient's history, he disclosed several weeks of polyuria and polydipsia with weight loss of 9 Kilograms in the last two months. His MCAD was well managed and he did not require carnitine substitution within the last two years. After liaising with the tertiary center intravenous access was obtained, and fluids started. The patient was transferred for further management and work up and care to our tertiary center.

In this case report, the metabolic pathways of MCAD and Diabetic Ketoacidosis, and treatment approaches are discussed.

P 25

Evaluation of bedside sonography performed by emergency physicians to detect intussusception in children in the emergency department

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Background: Intussusception is difficult to exclude with clinical examination and the final diagnosis is usually made with ultrasound performed by a radiologist. Previous studies suggested that emergency physicians highly experienced in point-of-care Ultrasound (POCUS) have similar performance to formal ultrasound to identify intussusception in children.

Objective: The aim of our study was to evaluate the ability of emergency physicians with various levels of POCUS experience to detect intussusception with POCUS among children visiting a pediatric emergency Department (PED).

Design/methods: This was a sub-analysis of a prospective cohort study conducted in a tertiary care PED. The study population was a convenience sample of children younger than 5 years old who necessitated an abdominal ultrasound for suspected intussusception according to the treating physician. Participating physicians were experienced in pediatric emergency medicine but had various levels of POCUS experience. They received a 1-hour didactical and practical training session on intussusception ultrasound. To be included in the sub-study, participants had to be seen by a physician who received the POCUS training. All POCUS were performed by the treating physician before further radiological evaluation following initial physical exam. Final outcome was determined by radiological final evaluation performed by a pediatric radiologist. The primary analysis was a simple proportion for the sensitivity and specificity of POCUS to identify intussusception.

Results: During the study period, a total of 238 children were recruited. Among them, 131 were evaluated by POCUS by an emergency physician, of which 45 (34%) had an intussusception. Twenty-four physicians performed between 1 and 25 POCUS for this study. The POCUS identified 39/45 intussusception for a sensitivity of 0.87 (95%CI: 0.74-0.94). A normal or inconclusive POCUS was reported for 83 of the 86 controls for a specificity of 0.97 (95%CI: 0.90-0.99). When excluding the 28 patients for whom the physician reported an inconclusive exam, the sensitivity improved to 0.98 (95%CI: 0.97-1.00) and the specificity to 0.94 (95%CI: 0.85-0.99).

Conclusion: This study demonstrated an excellent sensitivity and specificity of POCUS for intussusception in children when performed by multiple emergency physicians with variable POCUS experience.

P 26

How useful is a complete urinary tract ultrasound in orchiepididymitis?

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Introduction and objectives: Orchiepididymitis (OE) is a frequent cause of paediatric emergency Department (PED) attendance in boys presenting with acute scrotum. The aetiology of OE is still under debate. Many studies describe cases of OE secondary to genitourinary malformations, leading some teams to investigate each episode with a complete urinary tract ultrasound or voiding cystography in order to detect an underlying anomaly. In our tertiary centre, all suspected OE are investigated with white blood cell count, C-reactive protein, dip sticks, urine culture and a complete ultrasound (testicular, urinary tract and kidney).

The aim of this study was to analyse the number and type of urinary tract malformations detected by ultrasound following a first episode of OE and the frequency of associated urinary tract infection.

Methods: After approval from the local ethics committee, we retrospectively reviewed the electronic records of all boys under 16 years of age, presenting to our PED with an acute scrotum, between Jan 2011 and Dec 2017, and included all patients with a first episode of echographically confirmed OE and no previously known urinary tract malformation. Patients with incomplete radiological data were excluded. We extracted demographics, presenting features of OE, laboratory results and outcome. **Results:** 495 boys attended PED for acute scrotum, of which 119 had a radiologically confirmed first episode of OE and 99 patients were included in our study with a complete urinary tract ultrasound. In our population, complete urinary tract ultrasound did not detect any genitourinary malformation. Mean age at presentation was 9.7 years (SD 3.9) with a 3-peak incidence of OE: 10-13 years, 4-5 years and during infancy. On examination: pain was present in 98.0%, scrotal erythema and oedema in 66.7% and 65.3% respectively. The cremasteric reflex was described as absent in 27.5% of cases. Urinary cultures showed urinary tract infections in 3/99 patients (3.1%). There was only one recurrence of OE.

Conclusions: Over a 6-year period, in a country with a high availability of antenatal follow up, no new urinary tract malformation was found on routine ultrasound in patients under 16 presenting with a first episode of EO.

P 27

What works to defeat childhood obesity? Introduction of an inter-professional formation of paediatricians and other health care workers in Switzerland*

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Background: Childhood obesity represents a major burden for the Swiss health system, as more than 60'000 obese children need continuous care. Due to beneficial effects of multiprofessional group therapy (MGP) shown by the KIDSSSTEP project, treatment is reimbursed by Swiss health insurances. An interprofessional formation for paediatricians and other health care workers (HCW) was established to ensure an adequate multimodal treatment of obese children within MGPs and paediatric obesity reference centres. First results of evaluation are presented here.

Methods: The multimodal formation on medical, psychological, nutritional and physical activity, bariatric surgical therapy for obese children included basic science, various educational concepts as well as practical exercises, e.g. role-playing, motivational interviewing. After 3 days of course, 1 day of work shadowing in a MGP centre, a report and 1 final review day, participants were accredited as obesity therapists by several professional associations (AKJ, SGP, PhysioSwiss, SVDE, SMOB). Trainees rated practical relevance, scientific quality and meeting the expectations by a five point Likert scale; further remarks and cost-effectiveness were analysed.

Results: Out of 22 HCW (7 physicians, 7 nutritionists, 1 physiotherapist, 5 psychologists, 1 nurse, 1 social education worker), 18 graduated. A good to excellent rating was given in 86% for meeting the expectations, 95% for quality of the lectures and 92% for the benefit in practice. The interprofessional exchange of expert knowledge, the insight in the multimorbidity of children with obesity and the practical experience with humorous artists' or experiential therapy were mostly appreciated. The course fees of 1550 SFr. including certification will cover all costs if 21 trainees participate.

Conclusions: The new interprofessional formation based on multidisciplinary experts' knowledge and practical exercises was implemented to the participants' high satisfaction and is cost effective. There is now a unique nationwide Swiss paediatric therapy network, with professional formation linked to certification procedures, but in practice, difficulties arise, because costs of interprofessional exchange of HCWs, e.g. nutritionists and physiotherapists, are not reimbursed by health insurances. Evaluation by databases in paediatric obesity reference centres should assess, whether care for complex cases of obesity can be improved by comprehensive HCWs' education.

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P 28

Brain-lung-thyroid syndrome due to a new NKX2-1 mutation

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Introduction: Autosomal dominant thyroid transcription factor 1 (NKX2-1/TTF-1) mutations are a potential cause of the brain-lung-thyroid syndrome (OMIM_600635). The NK2 homeobox 1 (NKX2-1) gene is located on the long arm of chromosome 14q13.1-q21.1, which encodes the thyroid transcription factor 1 (TTF-1). TTF-1 is expressed during early embryonic development, especially of the thyroid, lungs, hypothalamus and basal ganglia. The phenotypic spectrum of human TTF-1 mutations is highly variable, even within the same family, ranging from isolated primary hypothyroidism to associated severe pulmonary disease and hereditary chorea with motor delay.

Objectives: We describe the clinical and genetic characteristics of a 3-year-old boy with a novel mutation of the NKX2-1 gene.

Methods: Genetic analysis of the NKX2-1 gene was performed in a 3-year-old child, which presented at birth with respiratory distress, pneumothorax, and a pathologic neonatal screening test for congenital hypothyroidism (TSH: 260 mU/L, normal level: ≤ 15 mU/L). Thyroid gland agenesis as well as severe muscular hypotonia were diagnosed. In the following years, he developed an asthma and neurological disorders with choreoathetosis and ataxia.

Results: Genetic analysis revealed that the child was heterozygous for a thymine to guanine change at nucleotide position 527 of the TTF-1 gene (c.527T>G), resulting in a substitution of leucine with arginine at amino acid position 176 within the homeodomain (c.527T>G; p.Leu176Arg) and rated as likely pathogenic (ACMG guidelines for interpretation of sequence variants; class 4).

Conclusions: A novel mutation of the NKX2.1 gene has been identified which likely affects TTF-1 expression. Although uncommon, the triad of primary hypothyroidism associated with neurological and respiratory disorders is extremely suggestive for a NKX2-1 gene variation and should prompt genetic workup. This case report highlights the importance of molecular medicine for solving complex clinical cases and enable genetic counseling for affected families.

P 29

Scurvy: The 21st century reappearance of a 19th century disease

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Background: Scurvy results from a deficiency of ascorbic acid. This disease first appeared in children during the 19th century with the emergence of new dietary habits; in particular, heating milk that lead to a loss of ascorbic acid. Even though Scurvy has become a rare condition in western countries, many cases are still reported in paediatric patients, especially in those who lack proper nutrition due to neurological or psychiatric illnesses. Symptoms include bleeding and swollen gums, loosening of teeth, bone abnormalities, arthralgia, delayed wound healing, anaemia, petechiae and purpura. The radiological bone lesions are mainly irregularities of long bones metaphyses.

Case report: We report the case of a five-year-old boy who presented with arthralgia and limb deformation (genua valga). The patient was investigated for vitamin deficiencies to exclude rickets. The radiologic investigations revealed metaphyseal signs compatible with Scurvy. During the hospitalisation the patient was observed to have abnormal eating patterns and the Scurvy was attributed to malnutrition.

Conclusion: Although the occurrence of Scurvy is rare, it remains essential to detect this disease in children at risk of developing vitamin deficiencies. Without targeted treatment, the complications of Scurvy can be serious and potentially fatal.

P 30

Bilateral hip luxation leading to hypophosphatasiaRovina L.¹, Steigert M.¹, Dierauer S.¹, Strozzi S.¹¹Departement of Pediatrics, Kantonsspital Graubünden, Chur

Introduction: Hypophosphatasia (HPP) is a rare inherited metabolic disorder due to deficiency of the tissue- nonspecific alkaline phosphatase (TNAP), caused by mutations in the ALPL Gen encoding for TNAP. The enzyme is mainly expressed in the bone. The severity of the disease depends on the type of mutation and age of appearance. The main clinical signs (rickets, fractures, premature tooth loss) are related to defective bone and dental mineralisation. In severe forms respiratory problems, seizures and muscle weakness occur. Recently, Enzyme replacement therapy with asfotase alfa (Strensiq[®]) has been approved for patients with severe forms of HPP. We present a case with severe infantile hypophosphatasia with bilateral hip luxation as the leading sign, where diagnose was genetically confirmed.

Case report: First child of unrelated healthy parents, born by caesarean section at full term after uneventful pregnancy. Neonatal course included irritable crying, generalized muscular hypotonia and severe bilateral hip luxation. Transient respiratory insufficiency with oxygen requirement and transient feeding problems occurred. Initial chest and pelvis X- ray were normal. Infant developed increasing muscle weakness with poor head control and delayed motor development. Unsuccessfully conservative hip luxation therapy led to surgical intervention. The surgeon was surprised by the structure of the femoral head. Biochemical investigations with elevated urinary level of phosphoethanolamine leading to suspicion of HPP. Diagnosis was confirmed by reduced ALP activity, elevated pyridoxal 5'- phosphate level and genetic testing, revealing a heterozygote mutation in the ALPL gene. Only then the chest X- ray showed a mild thoracic hypoplasia with rickets signs. Due to the age of the child and the severe neurological complication of HPP we initiated treatment with asfotase alfa (Strensiq[®]).

Conclusion: HPP is a rare disease with initial often unspecific presentation. In our patient bilateral congenital hip luxation was the leading sign. Making the diagnosis is important since a therapy exists. In a newborn with bilateral hip luxation and muscle weakness consider HPP.

P 31

Health information seeking behaviour of Swiss German parentsJuvault Sibylle¹, Jaks Rebecca¹, Robin Dominik¹, Baumann Isabel¹, Julia Dratva^{1,2}¹ZHAW Department Gesundheit, Institut für Gesundheitswissenschaften, Winterthur CH; ²Universität Basel, Basel CH

Background: Little is known on parental use of different information resources when it comes to children's health in Switzerland, and even less regarding digital media. Since digital natives are becoming parents and the potential of digital counselling is considered high, we investigated how, when and for which reasons parents utilize digital media in the context of child health.

Methods: A random sample of parents with children age 0-2.5 yrs. in the canton of Zürich (N = 2573) received an online and paper questionnaire on use of digital media in the context of child health. Qualitative data was collected in a parental focus group and in semi-structured interviews with pediatricians. Descriptive and multivariate regression analyses were performed.

Results: Questionnaires were filled out by 677 mothers and 89 fathers (N = 769, participation rate 31%). Parents' mean age was 35.5 yrs. (sd. 4.0) and 38 yrs. (sd. 6.1), respectively; mean age of youngest child was 14.7 months (sd. 7.1) and number of children 1.6, (sd. 0.7). Most parents used digital media for seeking information on their child's health and development (91%) and when the child is sick (79%), but continue to use traditional resources, personal contacts and print media. Main reason for digital use was the 24/7 availability (79%) Parents searched digital information on nutrition (25%), child development (22%), regulatory disturbance (19%), general information on acute health problem (54%), and alternative treatment options (42%), as well as prior (45.5%) and after (27%) the last visit to their paediatrician. There were no significant differences by sex, age or educational status, albeit an observed higher use of social networks by lower (P = 0.04) as compared to higher educated parents. Parents feel insecure regarding digital child health information, but rarely verify the received information, and voiced their wish for guidance. The quantitative findings are supported by qualitative data.

Conclusions: Parents frequently use digital media to inform themselves on child health related topics. They differentiate when and whom they trust, depending on the health context. Topics searched for range from

child nutrition to treatment options. While parents are aware of strengths and limitations of digital information, they voice insecurity. Guiding and enabling parents to use digital media competently may impact positively on parental child health literacy and, ultimately, child health.

P 32

Previsit psychosocial screening tools for adolescents and young adults: a systematic scoping reviewGlasner J^{1,2}, Ambresin A-E¹, Baltag V²¹University Hospital Lausanne (CHUV), Interdisciplinary Division for adolescent health (DISA); ²World Health Organization (WHO), Department of Maternal, Newborn, Child and Adolescent health (MNCAH)

Background: Adolescent and young adult patients performing a psychosocial screening during well-visits is a well-established practice. Several tools are used to detect typical psychosocial issues experienced by adolescents and young adults such as mental health problems, substance use and abuse or bullying. One of the most famous one is probably the HEADSS assessment. Tools have been developed around the world to identify these issues prior to the actual consultation. These pre-visit screening tools can have various formats (paper questionnaire, application, website) and can be used at various points in time before the encounter with the health care provider. There is a lack in the literature of a recent review on the existing tools used to perform this pre-visit screening.

Objective: We aim to review all existing tools in performing previsit psychosocial screening of adolescents and young adults in the 21st century, and to describe the characteristics of those tools.

Methods: This research is a joint effort between the University Hospital of Lausanne, Switzerland (CHUV) and the World Health Organization (WHO) Headquarters. This collaboration allows us to have a very clinical approach and at the same time a global health perspective. We follow the "Guidance for conducting systematic scoping reviews" developed by the Joanna Briggs Collaboration (<https://www.ncbi.nlm.nih.gov/pubmed/26134548>). We conducted a literature search in Embase, PubMed, CINAHL, PsychINFO, Cochrane, Web of Science, Proquest, SciELO and Global Index Medicus using a standardized search strategy including key concepts such as adolescents and young adults, psychosocial screening/assessment and previsit. In addition, we conducted bibliography mining to identify additional sources. Finally, we included sources of grey literature identified through contacting WHO Departments, research groups and international associations working on this topic. Sources included in the final analysis were identified in 2 steps, first by title and abstract screening, then by screening the full text. We are currently doing the data extraction that includes parameters of interest such as the context of the screening, format of the tool, and covered psychosocial domains.

Results: The work is still in progress. Based on preliminary results, we are expecting to display around twenty tools using various modalities such as paper and smartphone apps. The final results and discussion will be available in the coming months.

P 33

Early-onset psychosis in an adolescent girl with 22q11.2 Deletion Syndrome: A case reportGaspari Laura¹, Queirolo Sara¹, Zambotti Marco², Aldini Alessia², Ferrucci Elisabetta¹¹Istituto Pediatrico della Svizzera Italiana, Clinica dell'Età Evolutiva, Ospedale Regionale di Lugano, Lugano, Svizzera; ²Servizio Medico Psicologico, Organizzazione Sociopsichiatrica Cantonale, Ticino, Svizzera

Introduction: DiGeorge Syndrome (DGS) is a disorder caused by 22q11.2 deletion and is the 2nd most common genetic cause of development delay after Down syndrome, with a prevalence of 1/4000 live birth. Carriers of the deletion have a markedly increased risk for neurodevelopmental brain disorders, including cognitive and behavioural problems in early adolescence. The features of DGS may vary widely, even among members of the same family. DGS may be frequently misdiagnosed with consequent inappropriate intervention that may increase morbidity.

Case report: We describe a female patient with a diagnosis of complex congenital cyanotic heart disease at birth, with typical features of DGS, where chromosome analysis revealed a 22q11.2 deletion. During growing up the patient presented delayed developmental milestones and moderate mental retardation (IQ level = 49) necessitating a special school. At the age of 12, due to social closure and unjustified fears, she

underwent a psychological treatment. At 16, she developed contamination obsessions with weight loss, intrusive obsessive catastrophic thoughts, auditory and visual hallucinations, persecutory delusions and disturbed sleep. A treatment with Olanzapine 2.5 mg/day was introduced, only partially effective on disturbed sleep. No improvement was noticed on positive symptomatology. A cardiological check was performed, pointing out a prolonged corrected QT interval (cQT = 483 msec). Thus a therapy with Aripiprazole 10 mg/day was started, since it presents a lower long-term effect on metabolic parameters and less impact on cQT prolongation. The patient achieved an improvement in positive symptoms, disturbed sleep and weight loss, but on the other hand, she presented some extrapyramidal drug side effects such as bradykinesia, rigidity, and dystonia. The drug dose was thus progressively decreased to 5mg/day, with extrapyramidal and cardiological side effects reduction (cQT = 450 msec). Nevertheless, she continued to report auditory and visual hallucinations.

Discussion: Practical guidelines for managing patients with 22q11.2 deletion, include screening for psychiatric illness during early adolescence, when psychotic decompensation may emerge. This case underlines the needs for paediatricians to be aware of any behavioural manifestation in DGS. Early diagnosis provides in fact the best opportunity for influencing the course of illness and optimizing outcomes.

P 34

Psychotropic drugs without borders: an analysis of off-label use according to three national authorities in a pediatric service of an university hospital

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Context: Adult patients can benefit from a greater choice of drugs compared to pediatric patients. The significant use of off-label drugs in the latter population raises questions of efficacy and safety when prescribing psychotropic drugs, whose marketing authorizations (MAs) can be very different from one country to another.

Aim: The aim of these studies was to characterize the prescription of psychotropic drugs in a pediatric service of a university hospital and to analyse the proportion of off-label use according to the different age, indication or dosage recommendations approved for several countries (Switzerland, France and USA).

Method: We conducted two studies: the first, a retrospective study (RS) carried out over a period of 7 months, aimed to include all hospitalized patients with at least one psychotropic prescription. The 2nd was a prospective study (PS) conducted over the remaining 5 months, whose aim was to include the same type of patients in the same pediatric ward. In both cases, the data collected were demographic data, medical data available and medication data. Off-label prescriptions have been identified through a comparison with the MAs published in the selected countries.

Results: A total of 74 patients were included in the RS and 33 patients in the PS. The average age was 13 ± 3 years and 14 ± 2 years respectively and the proportion of girls was 47% and 62%. Suicidal ideation and suicide attempts were the leading cause of hospitalization (33% of patients in both studies) followed by temper tantrum (12% and 14%). Of the 176 psychotropic prescriptions collected in the RS and the 88 prescriptions in the PS, 70% and 71% respectively were off-label according to the swiss MA, mainly concerning hydroxyzine, levomepromazine and aripiprazole. Compared to the french AMM (n = 174 and n = 84), 61% and 67% were considered off-label and compared to the american MA (n = 124 and n = 72), off-label prescriptions were only 56% and 51%.

Conclusion: Off-label prescription of psychotropic drugs is a common practice in pediatric services. However, several American MAs allow a prescription of these drugs from an early age and a harmonization with european MAs could lead to a better and evidence-based use of psychotropics in children and adolescents.

P 35

Gender differences in the management of child psychiatric patients hospitalized in a general pediatric ward: a retrospective cohort of 661 patients

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Introduction: The complexity of psychosomatic pathologies in children and adolescents requires an approach that integrates different fields of medicine, including pediatrics and child psychiatry. But who are these young people concerned about mental illness? Especially those who are hospitalized after a visit to the emergency Department?

The purpose of this study was to evaluate gender differences in this care context; does gender influence the care process? Does the projection of caregivers ("more anxious girls", "less depressed boys") have an impact on care decisions?

Methods: The discharge letters of 661 patients hospitalized from the emergency Department of the HEL (35,000 visits per year, several thousand for psychosomatic problems, of which 300 will be hospitalized) were examined. The data analyzed (separately for girls and boys) included: socio-demographic data, management data, reason for hospitalization (diagnosis) and referral at the end of the hospitalization.

Results, Discussion: Significant differences were observed in terms of socio-demographic characteristics (older girl, girl living more often in blended families or in educational home ("foyer éducatif"), boy more in specialized education and/or out-of-school); chief complaints (girl consulting for self-aggressive behaviour, eating difficulties) and definite psychiatric diagnoses (fewer girls with anxiety disorders, more girls with depressive disorder and eating behaviour); care (less urgent for girls) and aftercare (more girls referred to outpatient consultations and more boys to day care centres or hospitalized). These results are interpreted using three explanatory models, namely the biological model, the social-development model and the socio-constructivist theory.

Conclusion: This study aims to contribute to the evolution of the acceptance of mental illness, the disappearance of stereotypes and the unidirectional attributions of certain diseases to a particular gender. These data make it possible to contribute to better health education for the psychosomatic problems of caregivers.

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A retrospective analysis of the liaison between the Servizio Medico Psicologico of Locarno and the pediatric ward of the Hospital of Locarno "La Carità"

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Introduction: Since 2016, the pediatric ward of the hospital of Locarno has collaborated with the Servizio Medico Psicologico of the city with the main purpose of jointly treating patients (i.e. teenagers under the age of 16) who are affected by primary or secondary psychiatric problematics within hospitalized stationary environments.

Methods: For this analysis, several medical records of patients, who were treated with implemented pedopsychiatric support, were analyzed. Each hospitalization was reviewed for various aspects such as: the admission diagnosis, the duration of the hospitalization, possible comorbidities, the diagnosis on discharge and the follow up after the recovery. The analysis has taken into consideration patients who were hospitalized between the 01.01.2016 and the 31.12.2018.

Results: Overall, 41 hospitalizations have been analyzed, which included 38 patients (some patients had repeated hospitalizations). The mean age of the patients was circa 14 years and 5 months and the male-female ratio was of 1 to 2. The average hospitalization duration lasted about 12 days (the shortest hospitalization lasted a day, while the longest 47 days), with a total placement of beds over a 593 days-span framework. The principal disorders, which determined hospitalization, were eating disorders (circa 25%), followed by adjustment disorders, anxiety disorders, disorders due to psychoactive substance use, onset psychotic disorders, and others.

Conclusions: The interaction between the pedo-psychiatric and the pediatric teams allowed for an effective treatment and recovery of 38 young patients in a stationary hospitalized environment. At the time of discharge, nearly all the patients were put under the responsibility of either the psychiatric services for minors or the ambulatory specialists of the region. In 7 cases, patients benefitted from placements in suitable facilities or with foster families. According to the preliminary data that has emerged from the study, it is possible to affirm that a pedopsychiatric connection allows a prompt and intensive intervention of care for the minor so as to significantly reduce the evolutionary risk of critical situations.

The continuous controls that followed the post discharge period confirms the initial indications about hospitalization's functionality, as well as highlighting the importance of a stable and long-term based intervention of care of the patients.

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Adolescents and parents' perspective regarding confidentiality in healthcare: a clinical study

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Introduction: Confidentiality is an integral part of adolescent-oriented standards of care, which is also referred to as youth friendly health care. Physicians find themselves at the crossroad between the confidentiality wishes of the adolescents and their parents' desire to know the ongoing status of the medical care of their child. There is limited literature regarding the experience of confidential care amongst adolescents and their parents. There seems to be a gap between parent theoretical views on confidential care vs perception of confidentiality when confronted to clinical situation with their own child. The aim of this study is to explore in depth the representations and perceptions of adolescents and their parents regarding confidentiality in a clinical context.

Method: Qualitative study conducted at the interdisciplinary division for adolescent health (DISA) at Lausanne university hospital (CHUV). Nine semi-structured interviews of 1h-1h30 were conducted with four adolescent-parent dyads and one adolescent alone. Four out of the five teens were girls and all the parents were the biological mothers. Data were analyzed using a "Grounded Theory" methodology.

Results: A central phenomenon emerged from the analyses: "the dynamic adjustment towards autonomisation". Three other major themes were articulated around this central phenomenon: "the relationship to health care professionals", "the influence of prior experience with the health care system" and "the perception of the impact of confidentiality on health outcomes".

Discussion: Our study helped identify that parental ambivalence in regards to confidentiality largely reflects the difficulty towards adolescents' autonomisation in the context of their medical care, with adolescents seeking autonomy and responsibility on one hand and parents in a process of letting go and handing over responsibility on the other; an extremely delicate task for many. This double transition towards adolescent empowerment is marked by a dynamic adjustment of parental and adolescent experiences, which depends mostly on the triangular composition of the adolescent-physician-parent relationship. For the physician, managing trust, balancing the role of both parties and communication are all necessary issues to promote a positive experience of confidential care.

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Darbepoetin alfa: experience in young infants with renal failure

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Background: Anemia treatment in infants with advanced or chronic kidney disease (CKD) represents an important challenge to nephrologists. The use of darbepoetin alfa, a novel erythropoiesis stimulating agent, has largely replaced recombinant human erythropoietin in older children and in adults with CKD. However, studies reporting the use of darbepoetin alfa in infants below 1 year of age are rare.

Case presentation: We report the data of three infants with advanced stage kidney failure, aged 1, 4, and 7 months, who were treated with darbepoetin alfa and followed for 18–41 months. Hemoglobin levels increased in all three patients, reaching the target levels of 10.7–12 g/dl by 11, 19, and 22 weeks respectively, without any documented adverse effects. Patients younger than 1 year of age required a larger darbepoetin alfa dosage (ranged from 1.2 to 2.9 µg/kg per month) as compared to older children. A review of the literature found only three studies using darbepoetin alfa successfully in such young infants, with similar dosage and clinical success.

Conclusion: In these three patients with advanced kidney disease, darbepoetin alfa was effective in correcting anemia with no observed side effects. It reinforces its potential use in very young patients with advanced CKD.

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TINU syndrome – A link between nephrology and ophthalmology

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We present the case of a 15 year old girl with a quite rare disease, that links nephrology and ophthalmology. She was sent to the ophthalmologist because of a burning red eye. The diagnosis uveitis anterior was made and a local therapy with steroid containing eye drops was initiated. The girl also had an anaemia and the involved haematologist sent her to our Department of nephrology because of an elevated creatinine. At first presentation she showed a moderate renal insufficiency and a tubulopathy. Associated with the history of long lasting systemic symptoms as weight loss, fatigue and back pain we suspected the diagnosis of a TINU (tubulointerstitial nephritis and uveitis) syndrome. A renal biopsy confirmed a tubulointerstitial nephritis, consistent with our suspected diagnosis. The patient was treated with a gradually reduced dose of corticosteroids (prednisolone) and recovered within weeks. The TINU is a rare disease, most often seen in teenaged girls. The pathogenic mechanisms are poorly understood. To prevent misunderstanding of those patients and to treat the disease efficiently it is important to make the diagnosis as early as possible. The prognosis is good, relapses are possible and there are some patients who have persistent mild renal insufficiency.

P 40

A case of acute lobar nephronia

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Introduction: Acute lobar nephronia (ALN) is a localized non-liquefactive inflammatory renal bacterial infection. It is considered to be a mid-point in the spectrum of upper urinary tract infection between uncomplicated pyelonephritis and intrarenal abscess. Lacking specific symptoms and laboratory findings, the disease is probably underdiagnosed.

Case Summary: We describe a 5-year-old boy presenting to the ER with periumbilical and right flank pain, fever, shivering, vomiting, poor alimentary intake and fatigue for 3 days. Past medical history was positive for acute pyelonephritis at age 1 and 4. The abdominal ultrasound showed no sign of appendicitis and a normal urinary tract. Laboratory showed a massive inflammatory syndrome (leucocytes 19.7G/l, CRP 625mg/l) but no leucocyturia, nor nitrite in urine. Blood culture were taken and Ceftriaxone iv was initiated. Abdominal CT-scan showed multiple hypodense lesions in the right kidney, without sign of kidney abscess, fulfilling the diagnosis for acute lobar nephronia. Urine culture was positive for *Enterococcus faecalis* 10E5, blood culture remains negative. The patient responded to Amoxicilline iv treatment with no more fever after 3 days, allowing the switch to an oral treatment for 2 more weeks. The voiding cystourethrogram (VCUG) showed no sign of vesicoureteral reflux nor urethral valves.

Conclusion: Acute lobar nephronia should be suspected in patient with fever, rapid deterioration of clinical condition, flank/abdominal pain and vomiting. CT-scan remains the gold standard for the diagnosis of ANL but abdominal ultrasound with Doppler is taking a growing place. The treatment recommended is mainly a 2 to 3-week antibiotic treatment, depending if the ALN is considered as simple or complicated (based on CT-scan lesion and clinical evolution). Further radiological examination are mandatory as urinary tract abnormalities are found in nearly half of the patients with ALN and are considered as the major risk factor. A kidney scintigraphy is recommended in the follow up as residual lesion, such as cyst or scarring of renal parenchyma could remain.

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Myelin oligodendrocyte glycoprotein antibody-associated neuromyelitis spectrum disorders

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It has just recently become evident that the myelin oligodendrocyte glycoprotein antibody (MOG-Ab) positive neuromyelitis spectrum disorders are a distinct group of autoimmune disease in the pediatric population. Therefore, MOG-Ab-associated neuromyelitis spectrum disorders and

their treatment are of special interest in modern neuropaediatrics. Here we report a case study of a ten-year-old male patient with acute disseminated encephalomyelitis at onset, followed by relapsing optic neuritis. The patient was initially diagnosed with acute disseminated encephalomyelitis (ADEM) in June 2017 which was treated with intravenous methylprednisolone for two four-day-cycles due to a relapse after the first cycle. Bilateral optic neuritis was first diagnosed in July 2018 with acute impairment of visual acuity in both eyes. Treatment was with intravenous methylprednisolone and after lack of improvement of visual acuity changed to plasmapheresis for eight days in total. After five months without symptoms our patient relapsed twice in January 2019, this time with severe visual impairment in the right eye and eye movement pain. The patient was treated with methylprednisolone for five days but did not show satisfying visual improvement. Detailed antibody diagnostics demonstrated positive MOG-Abs. MOG-Abs have been consistently attributed to acquired demyelinating syndromes and increased probability of disease relapse. However, a guideline for therapeutic strategy has not yet been established. A clinical study in children by Yael Hachohen et al. (2018) found treatment with regular intravenous immunoglobulins most effective in annual relapse rates (ARRs) and Expanded Disability Status Scale (EDSS) compared to disease-modifying drugs i.e. rituximab. The study conveys an important insight in the state of the art of treatment of neuromyelitis spectrum disorders and the importance of a correct identification of MOG-Ab-associated optic neuritis in order to minimize probability of disease relapse. In our patient, therapy of the suspected demyelinating disease was then started with corticosteroids for three months and intravenous immunoglobulins once a month. The promising therapeutic approach and the disease relapse development of the patient will be closely examined in ophthalmological and neurological follow-up visits.

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A rare complication of Influenza B

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Case report: 8 y.o. boy, known for chronic inflammatory demyelinating polyneuropathy without recurrence since 2014, was admitted for sudden choreic movements, right then left painful camptocormia and inability to walk after prodromal illness (fever, rhinorrhea and odynophagia). Neurologic exam revealed legs strength at M4 and arms at M5, abolished tendon reflexes (both preexisting), trunk hypotonia, absent abdominal skin reflexes and widened base of support. During first 24 hours, he was confused, somnolent with fluctuating state of consciousness. He also had fever, erythematous throat and sub-mandibular adenopathy. The diagnosis of encephalitis was made. Nasopharyngeal swab: Influenza B positive. Lumbar puncture: White cells (3 cells/uL), proteins (410mg/L). IgG oligoclonal bands signing blood-brain barrier alteration, negative Influenza PCR. Metabolic work-up (homocysteine, ceruloplasmin, amino acids in blood, urine and CSF, organic acids in urine, acylcarnitine, Mn, Mg), autoimmune and inflammatory work-up (TPOAb, TgAb, ANA, anti-dsDNA, ASLO, anti-DNaseB, anti-NMDA, GABAB, AMPA, VGKC) were normal. Brain MRI: on T2 and diffusion sequences, bilateral and symmetrical cytotoxic edema of globus pallidus, splenium and knee of corpus callosum and bulb. Spinal cord MRI: normal. EEG: slow delta activity. Metabolic, autoimmune encephalitis and Sydenham chorea were excluded. Diagnosis of acute necrotizing encephalitis secondary to Influenza B was retained. On acute phase, he received methylprednisolone 1g/1.73 m² for 5 days (tapering over 4 weeks). Clinical course was slowly favorable, with gradual recovery in mobilization using physio- and occupational therapy.

Discussion: Influenza infection rarely evolves to acute inflammatory encephalomyelitis leading to substantial mortality and neurologic morbidity. Flu-like prodrome is often present 3 days before neurologic symptoms. Influenza is rarely found in CSF. EEG and MRI reveal signs suggestive of encephalitis. Complications include Reye syndrome, acute disseminated encephalomyelitis, Guillain-Barré syndrome and acute necrotizing encephalopathy. Oseltamivir is infrequently given, with no evidence in prevention of severe complications. High-dosed corticosteroids are recommended, hypothermia is still debated.

Conclusion: Influenza is rare but well-known cause of acute necrotizing encephalopathy. Physiopathology is unknown, it is considered as a parainfectious form of encephalitis. Treatment is based on high dose steroids.

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Ouch! My hair and head hurt!

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Background and importance: Headaches are a common reason of consultation among the pediatric population. Diagnosis is mainly based on history and physical examination. Neuralgias are less common in children, making them more difficult to recognize and treat. Occipital neuralgia is characterized by a uni- or bilateral pain, often starting in the upper neck or base of the skull, over the distribution of the occipital nerve. The pain is described as jabbing, throbbing, stabbing, shooting or electric. Hair or scalp pain in the affected region is a common complain. The etiology is unclear and it is usually considered idiopathic. The Tinel's sign, consisting in numbness or tingling elicited with percussion over the occipital condyle, is usually positive.

Clinical presentation: An 8-year-old boy has a 1-week history of daily debilitating left parieto-temporo-occipital headache without photo- or phonophobia. The pain is constant and described as shooting with some spikes of intensity during the day, and inconstantly also during the night. On physical exam, he has allodynia of the left-sided scalp with a decreased sensitive and thermal stimuli discrimination capacity. We moreover noticed a left paravertebral muscular contraction. He had a positive Tinel's sign.

Conclusion: Even if rare in children, occipital neuralgia is part of the differential diagnosis of headaches. A unilateral intense headache associated with allodynia of the scalp, hair and trigger points at the emergence of the greater occipital nerve, suggests an occipital neuralgia. Work-up and management are discussed.

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Doctor, my hands and feet are "on fire"

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Case report: A 10 y.o. boy presented with paroxysmal burning pain in his palms and sensation of electric discharges in his fingers, causing insomnia. Symptoms started during a Streptococcal pharyngitis. Symptoms were relieved with ice pack or cold-water application. Usual analgesic drugs were ineffective. Physical examination was normal, without local redness or heat. Small fiber neuropathy (SFN) was suspected.

Initial laboratory tests (blood cell count, ESR, CRP, Lyme serology, ANA, ANCA, RF) and complementary blood tests for porphyria and Fabry disease were negative. Genetic analyses of the genes TRPA1, SCN9A, SCN10A and SCN11A revealed no mutations. Sudoscan[®] was abnormal, with total loss of sudation on his hands and feet. Skin biopsy, on day 11 from onset was normal, probably because of the early identification. Post-infectious SFN was suspected. At the 10th days of evolution IV immunoglobulin for 5 days was initiated in association with neuropathic pain killers (gabapentine, clonazepam), sodic channel blockers (carbamazepine and mexiletine) and sedative drugs (chlorpromazine) with complete symptoms relief 4 weeks later. He was discharged on mexiletine for 2 more months and stays asymptomatic after 9 months.

Discussion: SFN is a rare disorder in pediatric population affecting the small, little or non-myelinated nerve fibers conducting pain and temperature. Clinical signs: burning pain in the extremities and local signs of dysautonomia. Etiology is variable, from primary (genetic) to secondary (mostly disimmune/toxic) origine. Diagnosis is confirmed by pathologic skin biopsy showing loss of small nerve fibers. Sudoscan[®] can be useful tool to evaluate small nerve fiber function in children. Underlying disease must be excluded in order to apply the causative therapy. Symptomatic treatment consists of sodium channel blockers, drugs against neuropathic pain and ice pack application. Early diagnosis is important in order to avoid complications (depression due to chronic pain, ulcers, necrosis or amputations due to aggressive cooling).

Conclusion: SFN is characterized by burning pain of the distal extremities (heat and redness can also occur). Diagnosis is based on clinical presentation. Sudoscan[®] and skin biopsy are essential for evaluation of small nerve fibers. Symptomatic treatment consists on neuro-modulating agents. Immunoglobulin treatment should be considered in case of post infectious erythromelalgia.

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Unusual presentation of myasthenia gravis with facial and bulbar symptomsEnzmann Cornelia^{1,2}, Hafner Patricia^{1,5}, Wörner Andreas⁴, Klein Andrea^{1,2,3}¹Department of Pediatric Neurology, Children's University Hospital, UKBB, Basel; ²Department of Pediatric Neurology, Children's University Hospital, Inselspital Bern; ³Department of Pediatric Neurology, Children's University Hospital, CHUV, Lausanne; ⁴Department of Pediatric Rheumatology, Children's University Hospital, UKBB, Basel; ⁵Division of Neurology, Medical University Clinic, Kantonsspital Baselland, Bruderholz**Background:** Juvenile myasthenia gravis is a rare auto-immune disease with antibodies against the neuromuscular junction. Clinical features include ocular symptoms (with ptosis, ophthalmoplegia, diplopia), bulbar symptoms (dysphagia, dysphonia) and generalized symptoms with exercise intolerance and fluctuating weakness.**Case report:** We report an unusual presentation of juvenile myasthenia gravis in a 17-year old girl with predominant facial weakness and bulbar symptoms. At the beginning pain in the mandibula and stiffness in facial muscles when talking or chewing, with slurred speech and problems swallowing occurred. Additional muscle pain and fluctuating skin changes first led to a rheumatologic examination. Muscle MRI did not reveal inflammatory changes, CK was normal. She reported generalised fatigue, tiredness with increased sleep time, exhaustion in motor activities and cramp-like pain in the hand during writing. She stopped doing sports. In the neurological examination, we found reduced facial expression, without ptosis and normal eye movements, unremarkable tongue motility and a very mild proximal weakness. Simpson test was negative and arm abduction was symmetrical after tiring the contralateral side. Uncertainty about the somatic origin of the symptoms occurred due to ill described history of fluctuating pain and skin changes and because the girl did not seem to be worried ("la belle indifférence"). Because of the fluctuating symptoms and history of fatigability we referred her to electrophysiological testing, that led to the diagnosis: stimulated single fibre EMG showed markedly increased jitter indicating neuromuscular transmission disorder. Subsequently tested Anti-Acetylcholine receptor antibodies were positive. Mestinon led to a rapid amelioration, immunosuppressive treatment was started with glucocorticoids and azathioprine; thymectomy was performed. Clinical symptoms gradually improved and remission was achieved within three months.**Conclusions:** Myasthenia gravis can present with a variety of symptoms. Especially the lack of the typical ocular symptoms can lead to a delay of diagnosis. Therefore, timely referral of patients with fluctuating symptoms to electrophysiological diagnostics is strongly recommended. Stimulated single fibre EMG can be done even in youngest children without sedation, does not need active collaboration and is usually tolerated well.

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A series of children with fibrous dysplasia and review of the literatureThomi A.¹, Guzman R.², Cartolari R.³, Cristallo Lacalamita M³, Ramelli GP¹¹Institute of Pediatrics EOC, Neuropediatrics Unit, Ospedale San Giovanni, Bellinzona, Switzerland; ²Department of Pediatric Neurosurgery UKBB, University Children's Hospital Basel, Switzerland; ³Department of Radiology EOC, Ospedale San Giovanni, Bellinzona, Switzerland**Introduction:** Fibrous dysplasia (FD) is a rare skeletal disorder, characterized by replacement of normal bone with fibrous tissue. The etiology is probably located in a somatic mutation of the GNAS1-gene on chromosome 20. FD is classified in three subtypes: monostotic, polyostotic, and the McCune-Albright syndrome. The abnormal growth of fibrous tissue can cause a large spectrum of symptoms including pain, pathological fractures, swelling and deformity of the affected areas. In some cases, the development of the disease remains asymptomatic. We focused on FD of the skull, studying three new cases and introduced them to those already quoted in the literature, comparing the medical data of the patients with the information described in the literature.**Case report:** We reported on three patients: a 13-year old girl, a 4-year-old boy and a 25 year old adult. The girl reported a monostotic FD of sphenoidal bone with compression of the optic nerve, which caused impaired vision. The two males presented the McCune-Albright syndrome; a disease characterized by coexistence of polyostotic FD, endocrinopathies and Café au lait spots. This typical triad, with a severe endocrine disorder, affected the boy. However the adult patient only reported skin

hyperpigmentation and polyostotic lesions, which lead to a massive headache.

Discussion: As already reported in the literature, the clinical presentation of FD is various and often unspecific. The common symptoms in our study are: pain, impaired vision, tiredness and nasal obstruction. The compression of the optic nerve is reported in the two younger patients. The management of this lesion is controversial. Many studies don't advise a prophylactic decompression as primary surgery, because blindness remains a possible complication of this operation. Instead, the continuous follow up of the patients with neuro-ophthalmological examination is important. The symptoms, especially by patients in pre-pubertal age, are often self-limiting. The McCune-Albright syndrome is presented by the male patients. Here, growth hormone excess is a possible endocrinological disorder and his suppression with dopamine-agonist is of prognostic importance. In our report, headache remains a severe symptom despite the medical and surgical treatment.

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MERS – Mild encephalopathy/encephalitis with reversible lesion of the splenium due to 3 potential pathogensDrexler I.¹, Nava E.¹, Bernhard-Stirmemann S.¹, Boxheimer L.², Scarcia-Sallenbach S.¹¹Department of Paediatrics, Cantonal Hospital Aarau, Switzerland; ²Department of Neuroradiology, Cantonal Hospital Aarau, Switzerland**Introduction:** Children presenting with focal neurological deficits and encephalopathy frequently receive MRI scans. Alterations are often unspecific and do not explain symptoms. In the acute stage of MERS (mild encephalopathy/encephalitis with reversible lesion of the splenium), transient hyperintense signals in the splenium of the corpus callosum can be seen as a characteristic neuro-radiological sign.**Case report:** A 10 year old boy presented with a history of fever since 5 days, coughing and bifrontal headaches. Clinical examination revealed reduced general condition, a gait and limb ataxia and a syndrome of inappropriate antidiuretic hormone secretion (SIADH). Analysis of the cerebral spinal fluid (CSF) was normal. Serologies in the blood were positive for Human-Herpes-Virus-6 (HHV-6)-IgM/-IgG and B. burgdorferi-IgM. Cranial MRI revealed a hyperintense signal in the splenium of the corpus callosum without other MRI abnormalities. Suspecting MERS due to a viral infection, he was treated symptomatically. Within 8 days, his condition improved significantly. Only slight residual gait and limb ataxia were seen at discharge. 9 days later, he represented with fever, fatigue, headaches and vomiting, but no new focal neurological signs, whereas his general condition was worse. CSF analysis showed mononuclear pleocytosis. Despite the absence of intrathecal B. burgdorferi antibodies, an antibiotic treatment with ceftriaxone was initiated due to unspecific intrathecal IgM synthesis and positive B. burgdorferi-IgM antibodies in the serum. Further serological analysis revealed positive tick borne encephalitis (TBE)-IgM and -IgG, intrathecal TBE-PCR was positive. His clinical condition improved rapidly and a cranial MRI 5 weeks later was normal.**Discussion:** We describe the characteristic radiologic finding of hyperintense signal in the splenium, the posterior part of the corpus callosum, which – in combination with an infection – leads to an encephalitis called MERS. Typical neurological signs are ataxia, confusion, headache, vomiting and slurred speech, often combined with a SIADH. Several infectious pathogens have been described as potential triggers. In our case, the responsible pathogen could not be clearly elucidated, but all three may have contributed to the prolonged course.**Conclusion:** In MERS with neuroradiologically isolated hyperintense signal in the splenium of the corpus callosum, the clinical course is usually favourable without sequelae.

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From psychiatric symptoms to a life-threatening disease: two cases of NMDA-receptor-encephalitisCornaz-Buros Sandrine¹, Schmidt Jasmin², Budäus Sonja³, Horn Michael⁴, Slavova Nedelina⁵, Kohler Barbara¹, Bigi Sandra¹¹Department of Pediatrics, Division of Child Neurology, University Children's Hospital, University of Bern, Switzerland; ²Department of Pediatrics, University Children's Hospital, University of Bern, Switzerland; ³Department of Pediatrics, Division of Intensive care, University Children's Hospital, University of Bern, Switzerland; ⁴University Institute of Clinical Chemistry, University hospital and University of Bern, Switzerland; ⁵Department of Neuroradiology, University Hospital and University of Bern, Bern, Switzerland

Background: Anti-N-methyl-d-aspartate receptor (NMDAR) encephalitis is a life-threatening condition characterized by an autoantibody directed against the central NMDA receptor. Targeted treatment approaches are available and the time point of treatment initiation influences outcome. We report on symptoms and management of 2 cases of anti-NMDAR encephalitis.

Case reports: A 12-year-old boy presented with a 2-week history of behavioral changes, sleep disturbance and episodes of mutism followed by aggressive outbursts without focal neurological deficit. An MRI of the brain showed swelling and FLAIR hyperintensity of the amygdalae; the lumbar puncture revealed a lymphocytic pleocytosis of 56 cells along with positive titers of anti-NMDAR antibodies (CSF 1:40; serum 1:20). The patient required ICU admission due to aggressivity and severe dyskinesia. High dose methylprednisolone, plasma exchange and administration of IVIg led to improvement of symptoms and the patient started an intensive neurorehabilitation program.

A 7-year-old girl presented with a 1-week history of progressive sleep disturbance, behavioral changes, aggressivity and single episodes of focal seizures. EEG showed general slowing; brain MRI showed a subtle swelling and FLAIR hyperintensity of the hippocampi. Behavioral changes deteriorated over time, followed by progressive fluctuation of the level of consciousness (LOC). A lumbar puncture revealed a lymphocytic pleocytosis of 43 cells, followed by an empiric antiviral therapy. The patient required ICU admission due to fluctuating LOC, severe dyskinesia and agitation. Anti-NMDAR antibodies were positive in CSF (1:80), negative in serum. Acute treatment with high dose methylprednisolone, plasma exchange and IVIg was initiated. After deterioration, the patient required treatment escalation with rituximab, leading to rapid improvement of symptoms.

Discussion and conclusion: New onset of fluctuating behavioural and psychiatric symptoms are characteristic clinical features of anti-NMDAR encephalitis. Diagnosis requires the assessment of anti-NMDAR antibodies in paired serum/CSF samples. As illustrated in case 2, testing of anti-NMDAR antibodies in serum only is insufficient. Untreated, evolution into intractable seizures, disabling movement disorders, severe autonomic dysregulation and coma is known. Therefore, the recognition of distinct clinical patterns is important in order to allow for timely diagnosis and initiation of immunosuppressive treatment

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Diagnosics and treatment of ADHD in Switzerland: A physician perspective on practice and challenges

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Background: Attention deficit/hyperactivity disorder (ADHD) is a major public health problem with numerous negative outcomes for the affected individuals and with a high burden to families and society. Although a multimodal therapeutic approach (i.e., pharmacological treatment as well as non-pharmaceutical treatment options) is considered the gold standard, the observed increasing methylphenidate (MPH) treatment could correspond to a reduced use of other treatments options. This study therefore aims to provide insights into the current practice of pediatricians as well as perceived challenges during the process of diagnosis and treatment.

Methods: An online survey on diagnostic and therapy procedures, personal attitudes, and perceived challenges regarding ADHD was sent to the members of the Swiss Society for Pediatrics (without reminder). With 151 questionnaires that were analyzed, response rate was low (9.3 %).

Results: Pediatricians reported the exchange with parents and children as well as the burden of the children to be central when selecting a therapy. On average they arrange three meetings with parents, of which two take place in presence of the child. They report including information from several sources before arriving at a diagnosis. Pharmacological therapy was most frequently prescribed, followed by psychotherapy, and occupational therapy. Challenges mentioned were: the subjective character of the diagnosis and frequent co-morbidities, limited resources for case management, limited availability of child and youth psychiatric and psychotherapeutic treatment, and unfavorable public attitudes toward medication.

Conclusions: Participating pediatricians considered a multimodal approach when treating ADHD and showed a high involvement of family and child in the choice of therapy. However, they highlight the potential

of improving the cooperation with other specialists, such as teachers and school social workers, and of improving the availability of psychotherapy and information on ADHD.

Main messages: Pediatricians are in close exchange with parents and children and consider several (non-) pharmacological ADHD treatment options. Challenges comprise the interprofessional cooperation and coordination as well as the availability of psychotherapy and information on ADHD.

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A stridor can hide a glioma

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Introduction: Brain tumors are the most frequent solid cancer in children. Diffuse intrinsic pontine glioma (DIPG) is a rare, inoperable and fatal children tumor. Most diagnosis occurs between 5 and 7 years of age and the prognosis is better if the child is under 3 or is more than 10 years old, has less extension beyond the pons or only few symptoms at diagnosis. Until this day, no curative treatment exists. Radiotherapy is the gold standard but it's a palliative treatment. The clinical presentation is very miscellaneous and can be manifested by ataxia, hemiparesis, cranial nerve palsy, mostly strabismus and diplopia, or symptoms of intracranial hypertension. This case describes a glioma of the brainstem highlighted by a stridor.

Case description: A 21-month-old child, born by IVF (In vitro fertilization), hospitalized at the Hôpital Intercantonal de la Broye, in Payerne, in early November 2018 for respiratory distress who presented a stridor during the second day of hospitalization. The stridor had been present for about 2 months, was positional, appeared especially during sleep and was variable in intensity. Several outpatient treatments, such as Ventolin, Betnesol and Axotide had been initiated by his pediatrician, with only moderate and transient efficiency for corticoids. A nasofibroscopy without narcosis had been performed and was described as "normal". During hospitalization, a followup with the CHUV's (Centre Hospitalier Universitaire Vaudois) pediatric airway team was organized. The child had an endoscopy under general anesthesia, which has shown bilateral vocal cord paralysis. In this context, a brain MRI was performed and highlighted an infiltrating glioma of the brainstem.

Discussion: A stridor can have multiple causes, the most common before the age of 3 years being laryngomalacia and paralysis of the vocal cords. When physicians discover a vocal cord paralysis it is essential to look for post-intubation or surgical trauma, as well as for an expansive mass at the cerebral level. Shortly after the diagnosis, the patient developed a dysphagia. He is now followed by the CHUV. He is already undergoing chemotherapy and receiving multidisciplinary care but his prognosis is reserved.

Conclusion: This case shows the importance of establishing a differential diagnosis and not being limited by a single "normal" exam. Multidisciplinary care and cooperation between hospitals is also essential, and this case shows it.

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Cranio-cervical artery dissection in childhood – Results: from the Swiss Neuro-paediatric Stroke Registry and a systematic review of the literature

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Background: Cranio-cervical artery dissection (CCAD) in children has mainly been described in stroke studies. However, the incidence of CCAD has not been reported, and little is known about CCAD presenting with non-stroke symptoms. The goal of this study was to gain a comprehensive overview on CCAD in childhood including and beyond its presentation with a stroke.

Methods: Population based observational study, the Swiss Neuropaediatric Stroke Study (SNPSR) and a systematic review of the literature.

Results: Out of 203 publications, we extracted 382 pediatric CCAD cases, 12 patients with CCAD were registered in the SNPSR. Of the total of 394 patients, 120 were female. Thirty-four were neonates; the remainder had a median age of 9 (IQR 6-14) years. Trauma in the month prior to CCAD was described in 195 (33 severe, 125 mild, severity unknown in the remainder). Presenting signs and symptoms were mostly due to ischemia (320 cerebral, 1 spinal), followed by intracranial hemorrhage (36), and local mass effect (12). CCAD was localized in the anterior circulation in 215, in the posterior circulation in 172 and in both in 7. The most commonly involved vessels were the internal carotid artery (161) and the vertebral artery (132). Localization was extracranial in 50%, intracranial in 45% and both intra- and extracranial in 5%. Forty-three children died. Survivors had a good outcome defined by independence in activity of daily living in 66%, with recurrence of CCAD described in 7 only.

Conclusion: CCAD has been described throughout all pediatric age groups including neonates, with preceding trauma being the single most important risk factor. Presentation is largely due to cerebral ischemia or hemorrhage, exceptionally due to a local mass effect only. Unlike in adults, intracranial is almost as frequent as extracranial localization. The case fatality rate is high, but two-thirds of survivors have a good outcome, with CCAD recurrence being very rare.

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Cerebral palsy register of Southern Switzerland

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Background: Infantile cerebral palsy (CP) is the most common cause of motor disability in the pediatric age. Currently in Switzerland there are around 3000 children holding a diagnosis of CP, while another 12000 are now adults. The first Swiss data, sent in 2015 to the Surveillance of Cerebral Palsy in Europe (SCPE), came from the Canton of St. Gallen. The aim of this study was to create a new register, collecting data from the southern part of Switzerland, gathering also data regarding MRI scans.

Methods: All patients matching the SCPE inclusion criteria, born in the Canton of Ticino and in the southern part of Canton of Grisons between January 1, 2000 and December 31, 2012 were included. A retrospective data collecting based on the medical background and anamnestic data provided by the family was carried out, and a standardized report was completed. Evaluation of MRI scans was performed. Data were analyzed using descriptive statistics.

Results: 44 reports of children with CP were examined (23 boys, 21 girls; age 10.2±4.5 yrs). Prevalence of CP was 1.4%. Average birth weight was 2623 gr (450-4670), 10 children (22.7%) were admitted to an Intensive Care Unit at birth, in 8 cases (18.2%) mechanical ventilation was necessary. 12 patients (27.3%) showed active epilepsy. 24 MRI scans were retrieved (54.5% of the sample): 7 scans (29%) were normal, brain malformations were diagnosed in 3 cases (12.5%).

Conclusions: The retrospective data retrieved in this study are basically in line with those of the registers of the Canton of St. Gallen and SCPE, and will be entered into the Swiss CP registry located at the Institute of Social and Preventive Medicine of the University of Bern (created in 2017). The collected data will be used to better anticipate therapeutic, educational and clinician needs in the future.

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Posterior arterial ischemic stroke in childhood: Clinical features and neuroimaging characteristics

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Background and purpose: Literature regarding the clinical manifestation and neuroradiological findings of pediatric posterior circulation arterial ischemic stroke (PCAIS) is scarce. This study aims to describe epidemiological features, clinical characteristics and neuroimaging data of pediatric PCAIS in Switzerland using the populations based Swiss Neuropaediatric Stroke Registry (SNPSR).

Methods: Children aged 1 month to 16 years presenting with a purely PCAIS between 2000 and 2016 were included. Epidemiology, clinical manifestation, stroke aetiology and neuroradiological features were summarized using descriptive statistics. Stroke severity was assessed using the Pediatric NIHSS. Correlation analysis was performed using Spearman correlation coefficient.

Results: Forty-three children with PCAIS were included (27 boys (62.8%), median (IQR) age 7.9y (5-11.7)). The incidence of PCAIS in Switzerland was 0.183/100 000 and represented 16 % of all childhood AIS. Most patients presented with unspecific neurological complaints such as headache (58.1%) and nausea/vomiting (46.5%). The most frequent clinical manifestations were ataxia (58.1%) and motoric/sensory hemisindrome (53.5%/51.2%). Unilateral focal cerebral arteriopathy (FCA) was the most common aetiology (11 children, 25.6%). The majority of infarcts were located in the cerebellum (46.5%) and thalamus (39.5%). A shorter diagnostic delay correlated with more severe stroke symptoms at presentation ($\rho = -.0365$, $p = 0.016$).

Conclusions: Pediatric PCAIS is due to FCA in one quarter of the patients in our cohort. The frequently reported unspecific clinical symptoms, especially when associated with mild neurological findings, carry the risk for a delayed stroke diagnosis. A high index of suspicion and increased awareness are required for a timely diagnosis and treatment initiation.

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Survey on the screening of early symptoms of autism-spectrum-disorder during well-child visits in Zurich pediatric practices

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We conducted a survey among Zurich pediatric practices to determine how frequently early symptoms of Autism-Spectrum-Disorder (ASD) are considered during regular well-child visits and whether using the SGP checkup-lists would raise this frequency. Fifty-four percent of the contacted pediatricians responded. Of those, 50% reported using the SGP checklists. There was a strong tendency that those using the checklists assess for early ASD-symptoms more regularly than those who didn't. 29% of the answers showed a statistical significant difference with $p \leq 0.05$ between the two groups. Furthermore, only 27% of the pediatricians used the M-CHAT-R questionnaire as an additional screening tool. This survey showed that ASD screening in pediatric well-child visits could be improved, and that the use of the SGP checklists might be a useful tool to screen for abnormalities in social development.

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Relapsing cerebral vasculitis as a complication of pneumococcal meningoenzephalitis

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Introduction: Streptococcus pneumoniae is a common cause of bacterial meningitis in childhood. An overwhelming systemic inflammatory response can lead to vasculitis, brain edema and secondary ischemia. We report a girl with relapsing cerebral vasculopathy after pneumococcal meningoenzephalitis with fatal outcome.

Case report: A 10 years old girl presented with high fever, cough, vomiting without food intake and raised inflammatory markers. Chest X-ray showed consolidations and further clinical symptoms suggested meningoenzephalitis. Cranial CT-Scan showed a Chiari-1-malformation and enlarged ventricles; therefore, lumbar puncture (LP) was postponed. Despite initial treatment with ceftriaxone she went comatose and developed unilateral oculomotor nerve palsy. Cranial MRI (cMRI) revealed multifocal areas of restricted diffusion including the brainstem suggesting cerebral vasculopathy. Despite additional high dose steroids the girl's condition further deteriorated with supranuclear hemiparesis and bladder dysfunction. cMRI showed new small ischemic lesions in the brain and spinal cord. Intravenous immunoglobulins (IVIg) were given. As the patient's condition improved, LP was performed (53 cells/ μ l, 59% polynuclear). PCR-analysis of CSF confirmed pneumococcal infection. The patient

had been fully vaccinated including PCV-13. Ceftriaxone was given for 21 days. After a steady phase of slow improvement, steroids were tapered. 4 weeks later, the patient suddenly deteriorated again, became comatose and quadriplegic. LP showed pleocytosis (1082 cells/ μ l, 95% polynuclear). Microbiological investigations were negative. Repeated cMRIs showed new widespread ischemic lesions in thalamus, brain stem and cerebellum and stenosis secondary to inflammation of the middle cerebral artery that progressed despite resuming high-dose methylprednisolone and IVIG. The girl slowly recovered over the next week but presented in a locked-in syndrome requiring tracheostomy and PEG-insertion. 10 weeks after initial admission, she was transferred to a rehabilitation center, where she rapidly developed another relapse with additional brain stem dysfunction and central respiratory failure. Parents consented to withdrawal of intensive care and the patient died.

Conclusion: Pneumococcal meningitis can be complicated by cerebral vasculitis. As the vasculitis can have a relapsing and progressive course, anticipation of this complication and careful monitoring of anti-inflammatory therapy is recommended.

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Anti-NMDA-receptor-encephalitis in a 17 year old girl

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A 17 year old girl was assigned to our emergency Department with increasing psychiatric and neurologic symptoms from a psychiatric clinic. She was treated with neuroleptics and benzodiazepines since four weeks because of panic attacks and progressive delusions. Working diagnosis was catatonic schizophrenia. Despite the therapy, her condition worsened, she developed incontinence, catatonia, tremor, mutism, increased sweating and dysphagia. Malignant neuroleptic syndrome was suspected, neuroleptics were stopped and Biperiden was started. Even though the neuroleptic therapy was stopped, her condition aggravated. Therefore, further diagnostic was initiated. Cerebral MRI was normal. The EEG indicated a generalized slow activity but no epileptiform discharges. A lumbar puncture showed mild elevation of proteins without pleocytosis. Infectious diseases were ruled out. Testing for anti-central-nervous-system-antibodies was positive for Anti-NMDA-Receptor-antibodies in liquor and serum. Thereby diagnosis of autoimmune encephalitis was established and an immunosuppressive therapy with intravenous immunoglobulins and methylprednisolone for five days was started. A maintenance therapy with prednisone was established and immunoglobulins were injected monthly. Additionally an anticonvulsive therapy with levetiracetam was started due to a two-time status epilepticus. Early rehabilitation was initiated. Recovery started after a few days and after almost one month she was transferred to a neurorehabilitation centre.

Anti-NMDA-Receptor-encephalitis is a relatively new disease which was first discovered in 2007. The exact incidence and prevalence is not known yet. However with the increasing number of case reports it seems underdiagnosed. It is a multistage illness with typically psychiatric symptoms (anxiety, delusions, insomnia, fear, mania) in the beginning and rapid progress with catatonic symptoms, movement disorders, seizures and autonomic instability. In children seizures and movement disorders may appear as first symptoms. It affects predominately young female adults and can be associated with tumors, especially teratoma. First line treatment includes intravenous immunoglobulins, steroids, plasmapheresis and tumor resection. If treatment is started early, prognosis is good. As it is still a "young" disease, many aspects are not well understood and further studies are needed.

P 57

Raised transaminases: not always liver disease

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Case presentation: A 6-year-old Afghan boy just arrived in Switzerland was referred for suspected tuberculosis (this suspicion was subsequently not confirmed). The laboratory evaluation of the child, who was anicteric and without liver and spleen enlargement, disclosed relevantly raised transaminases: aspartate aminotransferase 294 U/L (reference: <40), alanine aminotransferase 390 U/L (reference: <40). Alkaline phosphatase, γ glutamyl transpeptidase, bilirubin and prothrombin time were normal. The subsequent work-up (hepatitis A, B, C and E, Epstein-Barr virus, cytomegalovirus, HIV and renal copper excretion) was noncontributory. In retrospect, examination of the child disclosed a waddling gait,

lumbar lordosis and calf enlargement. Furthermore, he used his hands to push on his legs to stand up (Gowers sign). Very high creatine kinase levels (13265 U/L; normal: <149) and, subsequently, molecular biology studies confirmed the diagnosis of Duchenne dystrophy.

Discussion – take home message: Transaminases are highly concentrated also in muscle cells. Consequently, muscle diseases such as Duchenne or Becker dystrophy (and strenuous exercise) may lead to hypertransaminasemia. Textbooks and reviews recommend that, when confronted with elevated aminotransferases, it is mandatory to search for signs such as waddling gait, lumbar lordosis, calf enlargement and Gowers sign. Furthermore, in the context of hypertransaminasemia without jaundiced or hepatosplenomegaly, the first consideration is to ensure that the enzymes are of liver origin by checking the creatine kinase. It behooves us to recall this frequently disregarded recommendation. In our opinion, finally, pediatricians might consider obtaining creatine kinase in all children requiring determination of transaminases (thereby avoiding unnecessary evaluation and diagnostic delays).

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Heterozygous GNPTAB c3515A>G missense mutation in a patient with mucopolipidosis II: a case report

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Background: Mucopolipidosis II (I-cell disease) is a slowly progressive inborn lysosomal storage disorder. It presents with a perinatal clinical onset with typical facial dysmorphism and affection of multiple organs, causing contractures in large joints, skin thickening, thoracic deformity, cardiac involvement with thickening and insufficiency of the mitral valve and finally respiratory insufficiency because of progressive narrow airways and gradual stiffening of the thoracic cage. The mental and motor development in patients with Mucopolipidosis II is delayed. No causal treatment is available as yet and life expectancy is limited with a fatal outcome generally in early childhood. Mucopolipidosis II affects the GNPTAB gene on chromosome 12q23 and is inherited in an autosomal recessive manner. Up to now several dozen pathogenic variants are known. The mutation leads to improper targeting of lysosomal acid hydrolases to lysosomes causing a higher activity of nearly all lysosomal hydrolases in plasma and other body fluids. The resulting excessive urinary excretion of oligosaccharides can be an important diagnostic sign.

Case presentation: We report the clinical, biochemical and genetic diagnosis of mucopolipidosis II in a boy who presented at the age of 24 days with multiple joint contractures, sucking weakness, facial dysmorphism, gingival hypertrophy and hypotonia of the trunk. In the following months a retardation of the psychomotor development, hypersomnia, scoliosis and spasticity of the limbs became evident. In the age of 19 months a cardiological examination showed a prolapse with a mild insufficiency of the mitral valve. Biochemical tests showed highly increased levels of several acid hydrolases (α Iduronidase, β Glucuronidase and β Hexosaminidase A + B) in the serum as well as reduced levels of GalNAc-6-sulfatase in the leucocytes with normal levels of β Galactosidase in the serum. Genetic analysis of 50 genes involved in lysosomal disorders showed a heterozygous variant of unknown significance in the GNPTAB gene as a c.3515A>G missense mutation. This mutation has not been known before as a molecular pattern of a lysosomal disorder.

Conclusion: This case shows a previously undiscovered mutation leading to a clinical picture of Mucopolipidosis II, contributing to the genotype-phenotype spectrum of this disease.

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Cutting or not cutting – The management of paediatric cerebral cavernous malformation

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Background: Cerebral cavernous malformations (CCM) consist of thin-walled, dilated capillaries. CCM may occur sporadically, after radiotherapy or due to an underlying genetic mutation and can present with seizures, headache, neurological deficits or as incidental finding on MRI. We report on symptoms and management of three patients with CCM.

Case reports: A 14-year-old boy presented with new onset of marked right sided hemiparesis including palsy of the right N.hypoglossus and leftsided N.abducens. In suspicion of acute stroke, MRI was performed revealing multiple supra- and infratentorial CCM. A large pontine CCM showed signs of acute haemorrhage. Neurosurgical resection of the pontine CCM was performed, followed by an extensive rehabilitation program. The family history of CCM was positive, genetic testing is pending. An 8-year-old boy presented with sudden, unilateral headache, intractable vomiting over 2 days, intermittent hypaesthesia of the left hand and left sided central facial nerve palsy. The MRI revealed a large right sided supratentorial CCM with signs of acute haemorrhage and oedema. Neurosurgical resection of the CCM led to complete remission of symptoms. A 17-year-old boy known for a history of cranial irradiation due to an embryonal carcinoma in the pineal gland underwent routine MRI as part of his oncological follow up. The MRI showed a growing number of asymptomatic supra- and infratentorial CCM. No intervention was performed.

Discussion and conclusion: CCMs are an important differential diagnosis of intracranial haemorrhage in children. MRI is the diagnostic gold standard, showing the characteristic "mulberry-like" lesions. Immediate MRI is important to rule out CCM mimics such as arterial ischemic stroke, where time point of diagnosis directly influences treatment decisions. Multiple CCMs prompt further genetic investigations. Management of paediatric CCM remains a topic of debate. Surgical resection is recommended for symptomatic CCM in easily accessible areas. For symptomatic CCM in highly eloquent areas, surgery should be considered, especially if the risk of rebleeding is deemed very high. High-resolution anatomical and functional imaging, precise neuronavigation and intraoperative electrophysiological monitoring are mandatory to avoid postoperative neurological impairment. CCMs without symptoms need regular follow-up and education of the patient.

P 60

Acute ptosis with contralateral hemiataxia: the rare occurrence of Claude syndrome related to a midbrain stroke in a young child

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Case report: A 21-month-old boy was admitted in our Department following the sudden onset of gait disturbance. On examination, an isolated right ptosis associated with a left hemiataxia were observed, prompting a brain MRI. DWI sequences reveal an acute ischemic lesion at the level of the right midbrain involving the cerebral peduncle and the paramedian area, explaining the clinical manifestations. Extensive immunological and infectious etiological work up was negative. No cerebral vessels abnormalities were detected. Low dose Aspirin was started. The patient is discharged on day 4 with physiotherapy at home.

Discussion: The combination of unilateral ptosis and contralateral ataxia was highly suspicious of a rare midbrain stroke syndrome, named Claude syndrome. In its classical and complete form, it consists of a third cranial nerve deficit, with a contralateral ataxia. In our case, the paramedian lesion probably affected solely the fasciculus of the third nerve giving the fibers innervating the ipsilateral levator palpebrae muscle as well as rostral fibers of the superior cerebellar peduncle and the red nucleus, resulting in a contralateral ataxia. To our knowledge, this has never been reported in a child where midbrain stroke are exceptional.

Conclusion: As in adults where it has been mostly reported, specific brainstem stroke syndrome characterised by unilateral and contralateral neurological signs are typical findings. Such clinical pictures being extremely rare in children, delay in diagnosis in posterior circulation pediatric stroke is common and might impact early management.

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Mycoplasma pneumoniae-induced nonsexually acquired genital ulceration (Lipschütz ulcers)

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Background: Diverse mucocutaneous disorders are associated with the respiratory pathogen *Mycoplasma pneumoniae*. Genital involvement has been reported during *M. pneumoniae* infection.

Methods: Case report and literature review.

Results: We report a girl with severe genital ulcerations in the context of *M. pneumoniae* infection, and present a literature review of previously published cases.

Conclusions: *M. pneumoniae* infection needs to be recognized and considered in the differential diagnosis of genital ulcerations in nonsexually active girls.

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Labyrinthitis associated with acute otitis media – a diagnosis not always obvious

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Introduction: The labyrinthitis is a rare but serious complication of acute otitis media. A rapid treatment is necessary to reduce the risk of profound hearing loss and persistent vestibular dysfunction.

Case report: We report the case of a 5 year old boy who presented a left acute otitis media treated by ibuprofene and, forty-eight hours later, by antibiotics. One day after starting antibiotic therapy, the patient complains of nausea, vomiting and dizziness triggered by movements. He gets to emergency where a viral gastroenteritis is suspected. The symptoms gradually increase and five days later, the child refuses to walk reason why the parents bring him back to the emergency Department. The examination of cranial nerves shows a right-beating horizontal nystagmus, partially inhibited by fixation. The whispered voice test is pathologic at left. The audiogram tonal confirms a left conductive and sensorineural hearing loss. The video impulse test, which allows to test the vestibulo-ocular reflex, confirms a left vestibular deficit. It's clear now, the child presents a left acute otitis media complicated by labyrinthitis. A tympanocentesis is performed urgently and he receives an intravenous antibiotic treatment. He feels better after the treatment but the child complains of persistent vertigo and left hearing loss few weeks later.

Discussion: The labyrinthitis results of the spread of infection from the middle ear to the inner ear (cochlear and vestibular apparatus). It can cause conductive and sensorineural hearing loss as well as vestibular dysfunction. Bacterial labyrinthitis may occur by direct bacterial invasion or through passage of bacterial toxins and inflammatory mediators, such as cytokines, enzymes and complement, into the inner ear. Symptoms include nausea, vomiting, tinnitus, hearing loss, vertigo and ataxia. Symptoms such as hearing loss are usually transient but may persist if otitis is not treated promptly. Otologic surgery may be required to drain the middle ear in addition to antibiotics.

Conclusion: A labyrinthitis is a serious complication of acute otitis media. The lack of knowledge of this condition can lead to a delay in the diagnosis and a risk of significant sequelae. Treatment is an emergency to preserve inner ear function and to stop the spread of infection. A multidisciplinary care otorhinolaryngologist and pediatrician is essential.

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Chlamydia-induced rash and mucositis (CIRM) – a less common differential diagnosis in a febrile child with mucocutaneous symptoms

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Background: A febrile child with mucocutaneous symptoms often represents a diagnostic challenge. Prompt evaluation of whether this common combination of symptoms is benign or corresponds to a more serious illness is crucial.

Case report: A previously healthy 5-year-old boy was admitted to our hospital after 7 days of fever, despite the intake of oral broad-spectrum antibiotics. He presented febrile and in a slightly reduced general condition with bilateral conjunctival injection, cracked erythematous lips, a buccal enanthem, a mild cervical bilateral adenopathy and bilateral pulmonary crackles and wheezes. He did not have any skin findings. Laboratory workup showed a leucocytosis of 17 G/l, a thrombocytopenia of

130 G/L, a C-reactive protein concentration of 142 mg/l. Chest radiograph showed discrete hilar infiltrates. A polymerase chain reaction (PCR) of a nasal swab was positive for Chlamydia pneumoniae and negative for Mycoplasma pneumoniae. The boy was treated with supportive therapy and intravenous clarithromycin. On the third day of hospitalisation he had become afebrile, his general condition and mucosal symptoms had improved and the laboratory findings had normalised. He could be persuaded to take oral azithromycin, which was continued for another three days. At a 2-week follow-up he showed complete resolution of symptoms.

Discussion: In a febrile child with mucocutaneous symptoms, life-threatening illnesses, such as meningococcal septicaemia, staphylococcal or streptococcal toxic shock syndrome and Kawasaki syndrome must be excluded. The combination of high fever, stomatitis and conjunctivitis can also correspond to a mild form of Steven-Johnson syndrome. The presentation with affection of two or more mucosal sites but absent skin findings was historically known as Fuchs syndrome. The syndrome is typically associated with Mycoplasma pneumoniae infection. In our case, however, the syndrome was associated with Chlamydia pneumoniae infection. This association has, to date, only been described on rare occasions as Chlamydia-induced rash and mucositis (CIRM).

Conclusion: CIRM is a rare diagnosis in febrile child with mucocutaneous symptoms, but absent skin findings. Since the pathogenesis, treatment and prognosis is different to erythema multiforme or Steven-Johnson syndrome they should be seen as a different entity. It is currently unclear how much benefit antibiotics or steroids bring in this immune-mediated disease. The syndrome has excellent prognosis.

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Marshall syndrome (PFAPA). Case series

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Background: Marshall Syndrome (PFAPA) is a recurrent condition including fever episodes, pharyngitis, cervical lymphadenopathy and aphthous stomatitis. Aims. The authors characterized retrospectively PFAPA patients regarding diagnosis and evolution. The authors also evaluated the inflammatory status between illness exacerbations in order to identify sensitive biological markers for disease activity and diagnosis improvement.

Material and methods: There were included 22 patients that fulfilled PFAPA-inclusion criteria during 3 year-period (Jan. 2012 – Jan. 2015). Inclusion criteria: 1.the patients that accomplish the PFAPA-criteria; 2.children under 10 years of age; 3.patients between exacerbations. Exclusion criteria: PFAPA-patients during fever episodes. The authors analyzed age-onset of clinical manifestations, duration between exacerbations and also between onset and diagnosis confirmation. Inflammation background was tested 10 days after previous fever episode. During attacks all patients received one dose Prednison, the fever subsidence was noticed in all cases 4-6 hours later. The authors tested randomly inflammation markers for 6 patients with PFAPA between episodes: procalcitonin (to exclude systemic inflammatory response), C reactive protein (CRP), tumor necrosis factor (TNF), interleukins 8 (IL-8) and 10 (IL-10). Used laboratory methods were: enzyme-linked immunosorbent assay (TNF), enzyme immunoassay (IL-6), chemiluminescent immunoassay (IL-8) and semiquantitative procalcitonin-test. For statistical analysis One sample T-Test was used.

Results: The authors included 22 PFAPA cases and revealed: mean age of symptoms onset was 44.09 months; period between onset disease and diagnosis was 34.91 months, suggesting a low index of PFAPA suspicion; period between fever episodes was 52.5 days (range 20-139 days); the CRP mean value was 19.72 mg/l (range 2.4 to 95, normal <6), meaning a significant statistical increasing (p value 0.005). Mention: mean CRP-value during fever attacks was 62.7 mg/l. The TNF was high (mean level 11.26 pg/ml, range 9.3-14.5, normal <8.1), representing also a significant increased value (p value 0.013). The authors noticed normal range for interleukins IL-8/ IL-10.

Conclusions: 1.PFAPA diagnosis is late, so awareness programs are useful in order to improve diagnosis. 2.IL-8, IL-10 are not useful to appreciate PFAPA evolution; 3.CRP / TNF remain sensitive markers for disease activity in PFAPA patients, even out of fever attacks.

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A (not so) uncommon case of shock

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Background: Food Protein-Induced Enterocolitis Syndrome (FPIES) is a delayed non-IgE cell-mediated food allergy. FPIES reactions typically present with repetitive vomiting 1 to 4h after ingestion of the trigger food, often associated with bloody or watery diarrhea. Severe reactions could lead to dehydration, lethargy and shock. In infants cow's milk or soy proteins most commonly cause FPIES allergic reactions. Despite the potential seriousness of reactions, awareness of FPIES is low leading to mis- or delayed diagnoses. We report the case of an infant who presented a severe FPIES reaction to cow milk proteins.

Case report: A 3-month-old infant was brought by ambulance to our emergency Department following repetitive vomiting episodes at home one hour after assumption of a cow milk based formula, accompanied by muscle hypotonia, hyporeactivity, pallor and tachycardia. During the preceding weeks parents has changed different kinds of formula milk because of discomfort and constipation. At the emergency the child presented with a low state of conscience, pale, with tachycardia and arterial hypotonia. Despite the absence of fever and a laboratory work up showing normal inflammatory parameters, a septic shock of unknown origin was suspected and a treatment with intravenous antibiotics and fluid resuscitation was started with a rapid improvement of the clinical conditions within one hour. The radiological and laboratory work up was normal. Because the rapid clinical improvement after fluid resuscitation, the onset of diarrhoea during hospitalization and having ruled out other causes of shock, we hypothesized a severe FPIES reaction to cow milk proteins and started an amino acid-based formula with normalization of the clinical condition. After full resolution of the symptoms and having ruled out an IgE mediated reaction to cow milk proteins by prick-to-prick skin tests, we preceded two weeks later with an oral food challenge with formula milk, which confirmed the diagnosis of FPIES. The child is actually doing well with a cow milk free diet and a reintroduction of the cow milk is planned after his first birthday.

Conclusions: FPIES could present in infants with severe reactions characterized by repetitive vomiting 1 to 4h after ingestion of cow milk- or soy-based formula with or without diarrhea and can lead to critical clinical conditions leading to shock. A rigorous collection of the clinical history can avoid delaying the diagnosis of an FPIES.

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A novel SCID presenting with Omenn features and velopalatal defect

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Severe combined immunodeficiencies (SCIDs) are a group of fatal disorders of the infant immune system if remaining misdiagnosed and consequently untreated during the first two years of life. Many gene defects are involved in SCIDs but some patients present with a phenotype of SCID with corresponding pathogenic genetic variant remaining to be unraveled. We report the case of a 2 month-old female infant who presented with progressive dry erythrodermia since day seven of life. She was born after 36 5/7 weeks of gestation of non-consanguineous parents. After birth, she was diagnosed with a velopalatal defect. Two months later, she was admitted to the hospital for a bronchiolitis due to a Picornavirus infection. The clinical examination pointed out a non-bullous erythrodermia with fine desquamation of the scalp and trunk, hyperkeratotic lesions of the soles of the feet, alopecia, diarrhea, mild hepatosplenomegaly and bilateral axillar adenopathies. The initial laboratory tests were only relevant for hyper eosinophilia (6.5 G/L) and elevated total serum IgE level (2300 KUI/L). Face to these clinical and biological Omenn syndrome features, an extended immunological workup was done. It showed the absence of CD4/CD8 naive T cells including early thymic CD4 T cell emigrants and an oligoclonal distribution of T-cell receptor on CD4/CD8 mature T cells. This was consistent with an Omenn presentation of a T-B+NK+ SCID disease. The thymus was ab-

sent on thoracic ultrasound. While no maternal lymphocytes were detected in the peripheral blood, the pathology of the skin biopsy was consistent with a "maternal graft-versus-host-disease". Cytogenetics (karyotype and CGH-array) has excluded a 22q11 deletion syndrome. The patient was then explored further by new generation exome sequencing. Targeted bioinformatic analysis of a panel of 479 genes involved in immunological and auto-inflammatory disorders failed to demonstrate any pathogenic variant explaining the SCID. The patient received a match sibling allogeneic stem cell transplantation and is now well 4 months post transplantation. In conclusion, this patient illustrates a new case of SCID with velopalatal defect and with no pathogenic genetic variant identified yet. For now, it remains unclear whether the patient presents with a primary absence of thymus or if she suffers from a thymic aplasia secondary to the SCID disease. Additional genetic explorations are ongoing in order to elucidate the genetical background of this disorder.

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A rare cause of recurrent pancreatitis

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Case: A 12 years old indian boy presented to our hospital with severe abdominal pain and vomiting. Amylase level was 212 U/l, lipase 3178 U/l. The ultrasound and the MRI showed an oedematous pancreas with pseudocysts. At the age of 8 years, an explorative laparotomy had been performed in another hospital because of similar symptoms and revealed a necrotizing pancreatitis. Interestingly, the mother associated the first episode with the consumption of sesame and the current episode with peanuts, which were usually avoided by the boy. After ruling out common causes of pancreatitis the patient history made us consider an allergy-induced pancreatitis, an hypothesis further supported by a blood eosinophilia (1.42 G/l, 25%), the elevated total (1522 kU/l, n <200) and peanut specific IgE (42 kUA/l, n <0.35) despite the normal tryptase (<1 µg/l, n <11.4). In vitro testing confirmed sensitization to sesame, peanut (rAra h 1, 2, and 6 positive), cashew and pecan nut with a high risk for severe anaphylaxis and persistence of the peanut allergy. The abdominal symptoms abated under analgetic therapy and the pseudocysts disappeared after a few months. Under dietetic restriction (peanut and sesame) no further episodes occurred.

Background: Food-allergy induced pancreatitis is very rare. Since 1990 eleven children have been described (age 1-17 years, mean 6.5 years). The allergens were mainly egg and fish. Three children had a recurrent pancreatitis. Only one boy had several food antigens as triggers. The vast majority of cases was found in Japan. In our patient, a cross-reacting allergy between sesame and peanut was hypothesized, as the IgE-binding epitope of the peanut allergen Ara h 1 has 80% homology with the sesame allergen Ses i 3, both vicillin-like globulins. However, in our patient only nSes 1 was positive, therefore two different allergens must have been triggers for the recurrent pancreatitis. Food-allergy induced pancreatitis could emerge from eosinophilic gastroenteritis with swelling and obstruction of the ampulla of Vater, leading to reflux of bile into the pancreas and causing pancreatitis. However, the often described slender pancreatic duct may point to potential additional mechanisms.

Conclusion: This case shows the importance of considering allergy as cause of idiopathic pancreatitis, especially in Asian children. By avoiding the culprit food allergens recurrent episodes can be prevented.

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A case report with a combined immediate and delayed hypersensitivity to amoxicillin

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Background: Drug reactions can be categorized in immediate- and delayed-type reactions which are based upon the timing of the appearance of symptoms and distinguished them in IgE-mediated and non-IgE-mediated [1]. An overestimation of suspected antibiotic allergy has been demonstrated in multiple studies [2-3]. It is important to assess subjects with histories of hypersensitivity reactions to antibiotics, as up to 90% of tested children are able to tolerate antibiotic [4-5]. Skin tests, in vitro-specific IgE testing and immunological tests are useful for distinguishing atopic from non-atopic persons.

Case description: We report a case of a 13-year-old girl presented with an immediate and non-immediate reaction after taking amoxicillin/clavulanic acid due to a suspected sinusitis. She experienced an itch all over

her body without rash two days after starting the antibiotic therapy. After 5 days she developed a maculopapular exanthema which did not improve after oral treatment with antihistamines and cortisone; she stopped taking antibiotics. In the same evening she introduced herself at the emergency room because of feeling dizzy, dyspnea and subfebrile temperature up to 38°C. In the further course she developed hypotension and increasing dyspnea with need for an intensive care monitoring. She stabilizes herself after treatment with antihistamines, cortisone, intravenous volume therapy and adrenaline inhalation. The rash was resolved after a few days. Skin prick testing results were slightly positive (2mm) to amoxicillin with appropriate control. No specific immunoglobulin E antibodies were found for penicillins. The basophil activation test (BAT) showed an increased value for CD 63 (14,7%) in response to amoxicillin, suggestive for an immediate-type reaction. The lymphocyte transformation test (LTT) results were positive for penicillin G and amoxicillin/clavulanic acid, indicating a delayed-type reaction to these antibiotics.

Conclusion: The data of this case demonstrate an amoxicillin allergy with evidence of both a delayed and immediate hypersensitivity reaction with good correlation between the clinical history and the immunological results (BAT, LTT). As far as we know no case report with a penicillin allergy with a combined immediate and non-immediate reaction has been described so far.

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Linear pruritic rash, arthralgia and fever in a child: a quiz case for diagnosis

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History: An eight-year-old boy presented with a three-week history of a fluctuating pruritic rash, arthralgia and intermittent fever of up to 39°C. The mother reported that he had eczema since infancy, but recently different pruritic and persistent lesions had appeared.

Presentation: The patient presented in reduced general status with an elevated temperature of 37.9°C. Clinical examination revealed salmon colored macules and urticarial papules predominantly on the extensor surfaces of the hands. Additionally, he showed excoriated plaques with linear distribution on the back and the extensor surfaces of the extremities. He had generalized lymphadenopathy, as well as arthralgia and swelling of both upper ankle joints and the left elbow. -Investigations: Blood tests showed leukocytosis (20.75 G/l), elevated C-reactive Protein (CRP) (73mg/l), elevated erythrocyte sedimentation rate (ESR) (93mm/h), and high ferritin (3450mg/l). Antinuclear antibodies (ANA) and rheumatoid factors (RF) were negative. An infectious etiology was ruled out. -Diagnosis: Systemic juvenile idiopathic arthritis (sJIA), with concurrent classical and non-classical cutaneous presentation

Discussion: sJIA is a rare condition, which typically occurs in preschool aged children. Common clinical features consist of symmetrical arthritis, a salmon-pink, evanescent, maculopapular rash, spiking fevers, lymphadenopathy, a sore throat, as well as splenomegaly and liver dysfunction. Blood tests usually show leukocytosis, high ferritin and negative RF and ANA screening. Mainstay of treatment are systemic corticosteroids and NSAIDs. Other than the typical skin findings mentioned above, our patient additionally presented "persistent pruritic plaques". They mark one of several non-classical presentations of sJIA reported in the literature and are typically located on the back and on the extensor surfaces of the extremities, often with a linear configuration, sometimes with scales or crusts. Awareness of this clinical sign is important, since it might imply persistent disease activity in children with sJIA and thus might be a sign for the need for more aggressive treatment. In our patient, treatment with NSAID and systemic steroids resulted in prompt improvement of the maculopapular rash, fever and arthritis as well as in a decrease of inflammatory laboratory markers. However, the persistent pruritic plaques improved only slowly. Reduction of medication led to a rapid flare-up of the evanescent rash.

P 70

A rare cause of facial asymmetry

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Background: Progressive Hemifacial Atrophy (PHA) is an acquired, typically unilateral, facial distortion of unknown etiology. We report the case of a young girl with this condition.

Case: A previously healthy 12-year-old girl was addressed to our Department for chronic articular pain, which lasted a few months and had resolved 30 days prior to the visit, with an otherwise uneventful personal history. At clinical examination, we found a left hemifacial hypotrophy with a retraction of the left superior lip, mucosal hyperpigmentation, left-sided tongue hypotrophy, skin hypopigmentation of the left forehead with ipsilateral presence of white hair and eyelashes. The rest of the clinical exam was normal. A differential diagnosis between linear scleroderma en coup de sabre (ECDS) versus progressive hemifacial atrophy (PHA) was suggested. Complete blood count, liver and renal function tests, ESR and CRP were normal. Antinuclear autoantibodies, anti-ds-DNA and rheumatoid factor were negative. Brain MRI confirmed facial asymmetry, with a right relative exophthalmos, hypotrophy of adipose tissue and the left masseter muscle. The left cerebral hemisphere was slightly hypotrophic with a focal T2- and FLAIR hyperintense signal in the left temporal subcortical white matter.

Discussion: ECDS and PHA are variants of linear scleroderma. PHA is a very severe variant, probably a phenotype for more than one entity. PHA refers to hemifacial atrophy that affects subcutaneous tissue, muscle, and osseocartilaginous structures with minimal involvement of the skin. ECDS manifests as a linear depression located on the frontoparietal scalp or paramedian forehead. Involved skin is hyperpigmented, shiny, firm, and displays alopecia. While the two may coexist in the same patient, clinical features are typically used to distinguish PHA from ECDS. Seizures and headaches, for example, are the most common neurologic symptoms in patients with PHA. After initial presentation, the disorder is usually slowly progressive but self-limited. Standard treatment in progressive diseases are immunosuppressive agents (steroids and methotrexate). In our case, clinical and imaging features were suggestive of PHA.

Conclusion: PHA is a rare and potentially severe form of linear scleroderma that may be associated with neurologic involvement. Initial presentation may be slowly progressive making the diagnosis difficult. Immunosuppressive agents are the mainstay of the therapy.

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Atypical hematological manifestation of celiac disease

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Case report: A 2-year-old boy presented with a 6-week h/o recurrent loose stools and loss of appetite. Faeces were bulky, foul smelling, and occurred 4-5 times/day. He had lost 10% BW in a month weighted 12.7 kg (P25-50) with height of 95 cm (P>90). He looked pale with sunken eyes but not dehydrated and afebrile. He had no adenopathy, his abdominal exam was normal without hepatosplenomegaly. Interestingly, the initial laboratory workup revealed a bicytopenia with severe regenerative normocytic anemia (Hb 63 g/L, MCV 79 fl, reticulocyte 1%), leukopenia (WBC 3.9x10⁹/L) with moderate neutropenia (ANC 0.5x10⁹/L), otherwise normal platelet count (227x10⁹/L) and absence of abnormal cells on peripheral smear. Hemolysis workup, ferritinemia, plasma vitamins B9 and B12, liver and renal function tests were normal. Hemoglobin electrophoresis was unremarkable. Tests for EBV, CMV and parvovirus B19 were negative. Abdominal US was normal. Due to unexplained bicytopenia bone marrow aspirate and biopsy was performed revealing hypocellular marrow with decreased erythroid and myeloid precursors, and augmented megakaryopoiesis compatible with aplastic anemia (AA). Screening for malabsorption revealed elevated anti-transglutaminase (1330 IU/ml; N <20 IU/ml) and anti-gliadin antibodies (IgA 304 IU/ml and IgG 524 IU/ml; N <20 IU/ml). Endoscopy with duodenal biopsy confirmed pathologic features of fully developed celiac disease (CD). Patient received packed red cell transfusion and was started on gluten-free diet. Follow-up at one month showed an asymptomatic child and resolution of anemia with normalized bone marrow.

Discussion: CD has gastrointestinal and extra-intestinal features. Association between AA and CD is very rare (4 published pediatric cases). The physiopathological mechanism is likely a systemic chronic immune-mediated process triggered after gluten protein ingestion in genetically susceptible individuals. Some patients, mostly adults, have been managed with immune suppressive drugs followed by hematopoietic stem

cell transplantation. Our patient is the second pediatric case who responds to RBC transfusion and total gluten-free diet alone. This is probably consequent to shorter duration in exposure to immune processes.

Conclusion: Screening for CD should be performed in children with unexplained hematological abnormalities such as AA with or without gastrointestinal symptoms. In the presence of CD a total gluten-free diet may be sufficient to resolve AA.

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A rare complication of endoscopic procedure

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Endoscopy in children is a safe procedure. Intramural duodenal hematoma (IDH) is a rare complication with an incidence of 1/1250 endoscopies [1]. We report the case of an eight month old girl, known for a severe combined immunodeficiency, who underwent a bone marrow transplant (BMT) at five months of age. Since birth, the child has presented oral intolerance with poor feeding. Enteral nutrition was quickly started but not well tolerated, with gastro-esophageal reflux. After a severe episode of acute respiratory distress syndrome, only trophic feeding on a nasogastric tube was achieved. Abdominal ultrasounds did not show any signs of graft-versus-host disease, further confirmed by an upper GI endoscopy with duodenal biopsies. During the same procedure a naso-jejunal tube was placed. Two days after the endoscopy, the patient experienced discomfort, abdominal distension, vomiting, of progressive worsening. The blood tests revealed a pancreatitis (lipase of 1722 U/L; N: 13-60 U/L) and cholestasis (gamma-GT of 1090 U/l; N: 5-101 U/L; bilirubin of 66 umol/l; N: 0-10 umol/l). The CT scan revealed an IDH from the inferior duodenal flexure to D4, with digestive occlusion and dilatation of the pancreatico-biliary system. Pre-endoscopic work up showed platelets, and PT, PTT and fibrinogen in the normal range. She was managed conservatively and recovery occurred spontaneously with removal of the naso-jejunal tube.

Because of the IDH, the presence of some cutaneous-mucous bleeding and an unknown coagulation disorder in the family (her mother), a coagulation workup with Von Willebrand and XIII factor's activity was done. We found a decreased factor XIII activity (46.5% then 48.7%; N: 75-130%) suggesting a heterozygous FXIII deficiency. IDH is uncommon after endoscopic procedures in children without hematologic disease but it is frequently observed in children after BMT[2]. Heterozygous FXIII deficiency may be associated with a tendency to bleed, generally after a traumatic injury or a procedure, and rarely, severe bleeding. Endoscopists should be particularly careful during the duodenal biopsy procedure in children after BMT in presence of personal or familial bleeding history. Before any surgical procedure, extensive coagulation check-up is indicated in patients after BMT with highlighted risk factors.

[1] S. Hameed et al. 2014 Apr;44(4):392-7, *Pediatr Radiol*

[2] A. Sierra et al. 2016 Dec;63(6):627-632, *J Pediatr Gastroenterol Nutr*.

P 73

Two new mutations causing Congenital Glucose-Galactose Malabsorption – A Case report

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Background: Congenital glucose-galactose malabsorption (CGGM) is a rare autosomal recessive disorder characterized by life-threatening osmotic diarrhoea in infants. Mutations in the SLC5A1 gene, encoding the sodium-glucose co-transporter cause this disease. The corresponding clinical presentation is watery diarrhoea and hypernatremic dehydration. When stopping the intake of sugars, the symptoms abruptly cease.

Case presentation: In this case report we present a 4 day old newborn boy, who suffered from watery intractable diarrhoea and significant weight loss of up to 20% of birth weight. He was breast fed, had a normal drinking pattern and no other gastrointestinal or neurological symptoms. Blood tests showed a hypertonic dehydration with an impressively high hypernatremia and hyperkalemia. Rehydration with ringerlactate and glucose infusions together with an elemental diet showed no improvement of his diarrhoea and electrolyte shift. The abdominal sonography demonstrated liquid filled intestinal loops suggestive for malabsorption

or enteritis. No signs of infection or inflammation could be found in laboratory tests. A stool reducing substance examination tested positive for glucose. This clinical presentation of persistent watery diarrhoea and deterioration of the patient's general condition raised the suspicion for a glucose-galactose malabsorption. On his seventh day of life, we decided to change the nutrition to a carbohydrate free product. The watery diarrhoea stopped within a couple of hours and electrolytes normalized within the next day. The boy started to thrive well and could be discharged from the hospital on day 25. Follow up consultations were performed in our outpatient clinic. Up to now the 6-month old patient thrives properly receiving a carbohydrate free diet, poultry, fish and black salsify. The next challenge will be establishing further supplementary food. Genetic analysis showed two mutations in the SLC5A1 gene, supporting the diagnosis of CGGM. Although mutations in the SLC5A1 gene are responsible for CGGM, our mutations have not yet been described.

Conclusion: This case reveals a typical presentation of a very rare disease. Early detection of the diagnosis can prevent life-threatening symptoms such as intractable diarrhoea and electrolyte imbalance. An early changing of the oral nutrition to a carbohydrate free diet is essential to cease symptoms. The significance of the newly detected mutations in the SLC5A1 gene needs further investigation.

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An uncommon case of cholestasis in infancy

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Introduction: Cholestasis results from impairment in the excretion of bile, which may be due to mechanical obstruction of bile flow or impairment of excretion of bile components into the bile canaliculus. Cholestasis warrants prompt diagnosis to exclude conditions needing an urgent therapy, like biliary atresia. Cholelithiasis is relatively rare in children, but it is found more often in patients with predisposing disorders. We present an unusual and insidious case of symptomatic cholelithiasis with onset in infancy.

Case report: A previously healthy 3-months-old boy presented to the emergency Department with a two days lasting history of acholic stool and increasing agitation. On the physical examination, the boy was in good conditions, afebrile and with normal vital signs. He presented slightly icteric sclera, his abdomen was not tender. Laboratory tests showed a cholestasis with a conjugated hyperbilirubinemia with elevated gamma-GT and transaminases with normal liver function. The abdominal ultrasound showed an extrahepatic and intrahepatic biliary dilatation with presence of biliary sludge in the choledocus and gallbladder. A conservative treatment with ursodeoxycholic acid initially failed with worsening of the cholestasis and detection of a gallstone in the distal choledocus. A MRCP confirmed the diagnosis of a choledocolithiasis who resolved spontaneously after a few days, with normalization of the liver function tests and the biliary dilatation. An etiologic work up has been done but an underlying for cholelithiasis predisposing disease hasn't been found yet.

Discussion: Cholelithiasis is uncommon in children and more rare in newborn and infants who are often asymptomatic. If symptomatic it can lead to a cholestatic jaundice with acholic stool, abdominal pain complicated by pancreatitis, fever and sepsis. In symptomatic infants, gallstones are usually located in the common bile duct (CBD). 30-40% of cholelithiasis are idiopathic, while in 60-70% an underlying cause can be identified: hemolytic diseases (20%-30%), parenteral nutrition, ileal disease, congenital anatomical anomalies, metabolic or genetic. Spontaneous resolution of gallstones is frequent in infants and hence a period of observation is recommended even for choledocolithiasis. Radiologic-interventional procedures or surgery are rarely needed.

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Comparison of the health care use of asylum-seeking patients and local patients in a tertiary pediatric hospital in Switzerland

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Background: In 2017, a record number of 68,5 million persons and approximately 35,5 Million children were forcibly displaced worldwide. Asylum-seeking children have important health needs and may face inequalities in access to health care. Recent information about the use of health care services by asylum-seeking children is scarce. The aim of this study was to compare the use of health care services by asylum-seeking and non-asylum-seeking children.

Methods: Patients were identified using administrative and medical electronic health records included from 1st January 2016 to 31st December 2017. To ensure only recently arrived asylum-seeking patients were part of the analysis, patients with records prior to Jan 2015 were excluded.

Results: In 202'316 visits by 55'789 patients, 1674 (1%) of visits were created by 439 (1%) asylum-seeking patients and 200'642 (99%) visits by 55'350 (99%) non-asylum seeking patients. Median age of asylum seeking patients was 13 (IQR 3-16) years, coming from Eritrea (14%), Afghanistan (13%) and Syria (9%). Median age of non-asylum seeking patients was 7 years (IQR 2-12) coming from Switzerland (64%), Germany (7%), Turkey and Italy (4% each). The emergency Department was most frequently visited by both, but significantly more often by non-asylum seeking patients (64'315/200'642, 32% versus 317/1674, 19%). The proportion of patients within the groups with >15 visits was comparable, created nearly 50% of the visits in the asylum-seeking group versus 25% in the group of non-asylum seeking patients.

Conclusion: The study showed that visits from asylum-seeking children represented a minor group with in general lower user rates than their local peers. Quick demographic changes in nationalities and age of asylum-seeking children lead to changing health needs requiring a quickly adjustable health care provision. Further studies are needed focusing on asylum-seeking children frequently using tertiary facilities.

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Syrian crisis: an innovative response

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Introduction: The Swiss government started a resettlement program, in collaboration with the United Nation High Commission on Refugees (UNHCR) to facilitate the arrival of particularly vulnerable refugees from Syria. A universally new healthcare program was specially created for this population in the canton of Vaud: A family consultation. Its aims are to provide necessary healthcare and prevention, explain the health system and to see the family as a whole, with crucial links between its members.

Methods: A pediatrician, a general practitioner, an interpreter and a nurse see the family altogether and then separately. A synthesis is made at the end of each consultation with the medical staff and the family. The family is seen again after two months. They are finally directed to a private general practitioner and a pediatrician.

Results: Between July 2016 and December 2018, 249 persons have been seen for a first medical evaluation, including 111 adults and 138 children. The staff had subjective feelings of a better effectiveness compared to the regular system, a better communication between adults and children medical staff and a significant gain of time and financial savings for the health system and the families (less appointments and all their costs).

Conclusions: The interprofessional model of a consultation for Syrian migrant families is innovative and effective. It could be implemented in other cantons receiving Syrian refugees and could be extended to different refugee populations.

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OASIs: Holidays for surgeons

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Introduction: Acute scrotum in childhood encompasses numerous differential diagnoses. Among them, the Acute Idiopathic Scrotal Oedema (AISO, OASI in french), despite its serious clinical presentation is self limited and does not require special intervention.

Case presentation: A 5-year-old boy with a previous history of recurrent para-infectious urticaria presented to the emergency room with a new episode of urticaria, fever, and a red, painful and swollen scrotum

appeared the day of the consultation. Clinical examination displayed urticarial lesions on the back, face and lower limb. The scrotum was bilaterally swollen, purple and painful. The remaining of the physical exam was within normal limits. Considering the possibility of a Fournier's gangrene, an infectious screening was made and came back normal. Scrotal ultrasound showed an important thickening of scrotal wall, with diffuse hypervascularisation and subcutaneous oedema ascending from the scrotum to the inguinal regions. The diagnosis of Acute Idiopathic Scrotal Oedema (AISO) was made.

Discussion: AISO is an acute scrotal condition characterized by moderately painful oedema and erythema of the scrotum, most often unilateral, but commonly spreading to the other side or to the penis, groin and perineum. It typically occurs in children aged 3-12 years but is also encountered in adults. It represents 2.5–30% of scrotal emergencies in patients younger than 20 years. This wide interval reflects the fact that this pathology is commonly underdiagnosed. Some causes have been suggested, allergic origin being the most favored one, but evidence is lacking. No treatment is necessary. Recurrence rate is between 21-51%. AISO is a clinical diagnosis, confirmed with ultrasound, which will typically show thickening, oedema and marked hypervascularity of scrotal walls (building the Fountain Sign in traverse view), homogene and normal testes with normal and symmetrical doppler flow, reactive hydrocele and often enlarged and hyper vascular inguinal lymph nodes.

Conclusion: AISO shares some clinical features with other more serious acute scrotal pathologies. Although clinical presentation is impressive, the disease is benign and self limited. Clinical and ultrasonographic signs are highly discriminant, better than what is encountered in testicular torsion. Nevertheless, in unclear situations, surgical exploration might be warranted.

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Constipation – just a symptom or a hint towards a broad differential diagnosis?

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Introduction: Paediatric constipation contributes to 3% of all referrals to paediatric practice and up to 25% to paediatric gastroenterologists. We want to highlight three cases with constipation as leading symptom. They all received laxatives as first-line therapy.

Clinical cases: Case 1. A previously healthy 20-month-old boy repeatedly presented to our hospital after a history of progressive recurrent abdominal pain and constipation over a period of 4 to 5 weeks. Initially he was seen 2 weeks after onset of symptoms. At that time, due to history, clinical and radiologic findings and response to enema and daily intake of osmotic laxative, severe functional constipation was postulated. At the second visit he presented with progressive abdominal and back pain, and refusal to walk. Inflammatory markers (CRP, ESR) were elevated and MRI confirmed spondylodiscitis of the lumbar spine. –Case 2. A previously healthy 10-month-old boy was admitted to paediatric gastroenterology with a history of constipation since the age of 2-3 weeks without failure to thrive. Despite almost daily administration of suppositories and/or enema, constipation persisted with further aggravation after introduction of solid foods from the age of 5 months. Close clinical examination including triggering of the anal reflex revealed anal atresia with perineal fistula. No further associated malformations were detected. Posterior sagittal anorectoplasty was performed. Since then the boy has regular bowel movements. –Case 3. A previously healthy 11-month-old boy was seen in our hospital with persistent painful constipation for 2 weeks despite rectal and oral stool regulation. History revealed that he had also refused to sit and stand for 2 days. Abdominal X-ray showed massive coprostitis. Meticulous clinical examination revealed asymmetrical patellar tendon reflex. MRI showed an intraspinal transdural tumor ranging from Th 4-8 with massive compression of the spinal cord. The patient underwent surgery and chemotherapy.

Conclusion: Peak incidence of functional constipation occurs around the time of toilet training. Diagnostic criteria include a duration of a ≥ 1 month in a child of ≥ 4 years of age corresponding to Rome IV criteria. Before starting laxatives, a thorough history and clinical examination of children – especially under the age of 4 years – is mandatory to rule out organic causes.

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A rare cause of abdominal occlusion

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Introduction: Primary intestinal bands are rare cause of intestinal obstruction in children. Their etiology and incidence are unknown. We present a case of a boy with a diagnosis of primary intestinal band, followed by a review of the literature.

Case: 4 y.o. boy presented with sudden abdominal pain with nausea and vomiting. There was no history of diarrhea, constipation, hematemesis, hematochezia, prior abdominal surgery or trauma. After a short period of improvement, abdominal pains increased and vomiting reappeared. He was admitted for clinical observation. He was afebrile with no peritoneal signs, bowel sounds were reduced. Work-up didn't reveal inflammation, urine analysis was normal. No signs of intussusception on the abdominal US, appendix was not visualized, there was small free fluid collection in the right iliac fossa. As symptomatology persisted, few hours later a 2nd abdominal US was performed, showing increased fluid collection with dilated intestinal loops, thicker wall of the ileum, and decreased peristalsis in the right iliac fossa. Abdominal X-ray followed by an abdominal CT were performed with a diagnosis of intestinal obstruction without anatomic recognized causes (Meckel, appendicitis, tumor etc.). Laparoscopic exploratory surgery was first performed, followed by a localized laparotomy. A lateral jejuno-jejunal band entrapping an intestinal loop was found and resected. There was no Meckel's diverticulum. Histopathological exam showed focal necrosis of the small intestine with a structure corresponding to a congenital primary band. Clinical follow-up was uneventful.

Discussion: Clinical presentation of primary intestinal bands is unspecific, often with clinical picture of intestinal obstruction. Abdominal X-ray or CT confirm clinical findings but can't find the diagnosis. Surgical exploration is mandatory. Laparoscopic exploration can help to determine diagnosis and guide the type of laparotomy. Due to intestinal loops dilatation, the treatment cannot be realized only by laparoscopy.

Conclusion: In the absence of prior trauma or surgery, it is mandatory to consider abdominal pain and vomiting as intestinal obstruction. Congenital abdominal band could be the cause and should be kept in mind. Radiological exams must be repeated if symptoms persist. Only few cases have been described. An additional literature review chart will be presented, summarizing these rare published cases.

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The spectrum of diseases and management in asylum-seeking versus non-asylum-seeking inpatients in a tertiary care paediatric hospital in Switzerland

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Background: The number of displaced children has reached an unprecedented size. In the last decade an increasing number of children arrived in Europe and local health care systems are challenged to adapt to their health needs and problems. The aim of this study was to systematically capture and compare the epidemiology and spectrum of disease of asylum-seeking and non-asylum-seeking children requiring hospital admission.

Methods: This is a retrospective cross-sectional study including health data from recently arrived asylum-seeking and non-asylum-seeking children admitted to a tertiary care hospital in Switzerland between Jan 2016 and Dec 2017. Asylum-seeking children were identified using the asylum status which is routinely documented at our institution since Jan 2016. Data was collected retrospectively using electronic administrative and medical health records.

Results: A total of 11'974 inpatient records were identified, of which 149 were from asylum-seeking and 11'645 from non-asylum-seeking children. The median age was 4 years in both groups (IQR 0-13 for asylum-seeking and 0-11 for non-asylum-seeking children), with 61% male sex in the asylum-seeking and 56% in the non-asylum-seeking children. The most frequent countries of origin of the asylum-seeking children were Eritrea (21.4%), Afghanistan (18.9%) and Syria (15.4%). Diseases of the respiratory system were the most frequent reason for admission in both groups. Non-asylum-seeking children were more frequently admitted for

injuries and poisoning. Asylum-seeking children were more frequently admitted for tropical infectious/parasitic diseases and mental disorders.

Conclusion: No difference in the age distribution and a high prevalence of respiratory system diseases was detected in both patient groups. Rare infections and mental health disorders are important diseases in asylum-seeking children and require special attention and training of staff working with pediatric asylum-seekers.

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Hemolytic-uremic syndrome after *Escherichia coli* urinary tract infection in humans: systematic review of the literature

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Background: Hemolytic anemia with red cell fragmentation, thrombocytopenia and acute kidney injury characterize hemolytic uremic syndromes. Many cases are linked to an intestinal infection caused by a shigatoxin-positive *Escherichia coli* (mostly of the "big seven group": O26, O45, O103, O111, O121, O145 and especially O157). Approximately 40 years ago, hemolytic-uremic syndrome was first linked to a symptomatic *Escherichia coli* urinary tract infection.

Methods: In order to characterize the form hemolytic-uremic syndrome that is associated with a *Escherichia coli* urinary tract infection, we conducted a systematic review of the literature using the PRISMA strategy.

Results: For the final analysis, we retained 23 original reports published since 1979. Five unselected pediatric case series addressed the possible occurrence of hemolytic-uremic syndrome after an acute symptomatic *E. coli* urinary tract infection among 266 cases and found the mentioned association in 8 (3.0%) cases. We also found 28 individual cases (17 females and 11 males) of hemolytic-uremic syndrome preceded by a symptomatic *Escherichia coli* urinary tract infection: 16 children aged from 2 days to 6.0 years and 12 adults aged from 22 to 75 years. Testing for shigatoxin, performed in 19 cases, was positive in 15 cases. *E. coli* serotyping was performed in 18 cases: testing for serotype O157, O103 and O145 was positive in one, one and two cases, respectively, while testing for serotype O26, O45, O111 and O121 was always negative. A urinary tract abnormality was present in 12 (43%) cases.

Limitations: This review exclusively integrates data from single case reports or very small case series that were sometimes poorly documented.

Conclusions: Symptomatic *Escherichia coli* urinary tract infections is a rare but established cause of hemolytic-uremic syndrome both in children and adults. The germs that trigger this form of hemolytic-uremic syndrome are usually shigatoxin-positive but do not belong to the "big seven group".

References: Lavrek D, Lava SAG, Milani GP, Simonetti GD, Bianchetti MG, Giannini O. Hemolytic-uremic syndrome after *Escherichia coli* urinary tract infection in humans: systematic review of the literature. *J Nephrol* 2018; 31: 919-24.

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Pyuria and microbiology in acute bacterial focal nephritis: systematic review

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Background: Acute focal bacterial nephritis is an uncommon but severe localized renal infection not containing drainable pus. It is considered as at the midpoint in the spectrum between pyelonephritis and intrarenal abscess. Presentation and imaging findings of acute focal bacterial nephritis have been extensively addressed. The aim of this review was to assess the prevalence of cases without pyuria or bacteriuria and the spectrum of microorganisms underlying this condition.

Methods: We conducted a systematic review of the literature in the National Library of Medicine and Excerpta Medica databases using the PRISMA strategy.

Results: For the final analysis, we retained 54 reports published between 1981 and 2018 describing 251 patients (159 females and 92 males) affected by focal bacterial nephritis, who have been specifically investigated with respect to urinalysis and standard bacterial cultures. They were 177 subjects ≤ 20 and 74 > 20 years of age. Pyuria and bacteriuria were absent in 33 cases, while pyuria was not associated with bacteriuria in 5 further cases. The vast majority of culture-positive cases

were caused by Enterobacteriaceae (slightly less than 80%) and *Pseudomonas* species (approximately 10%). Enterococcus species and *Staphylococcus aureus* were isolated in slightly more than 10% of cases.

Conclusions: 1. Acute focal bacterial nephritis may present without pyuria and bacteriuria. The rather common occurrence of both leucocyte- and culture-negativity suggests that this entity, like renal abscess, sometimes does not communicate with the urinary collecting system.

2. The study also points out that Enterobacteriaceae and *Pseudomonas* species are the predominant pathogens, followed by enterococci and staphylococci. The initial management consists of broad-spectrum antimicrobials with high tissue penetration, active against the mentioned microorganisms. Available literature suggests a total of three weeks of initial intravenous and subsequent oral antimicrobial therapy.

References: Janett S, Milani GP, Faré PB, Renzi S, Giannini O, Bianchetti MG, Lava SAG. Pyuria and microbiology in acute bacterial focal nephritis: systematic review. *Minerva Med*: accepted for publication.

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Visceral serositis in acute Epstein-Barr virus infectious mononucleosis: review of the literature

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Background: In healthy subjects, acute symptomatic Epstein-Barr virus infectious mononucleosis is a benign self-limited febrile disease that presents with sore throat, swollen lymph nodes or splenomegaly. In this condition, pericardial serositis has been reported in cases complicated by myocarditis, peritoneal serositis in cases complicated by pancreatitis, acalculous cholecystitis or appendicitis, and pleural serositis in cases complicated by pneumonia. Otherwise unexplained pericardial, peritoneal or pleural serositis are further uncommon complications of Epstein-Barr virus infectious mononucleosis that have not been incorporated in a comprehensive analysis.

Methods: We performed a systematic review of the literature in the US National Library of Medicine and Excerpta Medica databases and selected reports published after 1959. We used the PRISMA strategy.

Results: We found 22 patients aged from 1.2 to 80, median 19 years of age (13 males and 9 female subjects) affected by acute Epstein-Barr virus infectious mononucleosis associated with otherwise unexplained pericardial (N = 9), peritoneal (N = 15) or pleural (N = 15) serositis.

Limitations: The main limitation of the review results from the very small number of published cases, which were sometimes scantily documented.

Conclusions: Although the vast majority of patients who present with lymphadenopathy and serositis suffer from noninfectious diseases such as malignancy or inflammatory conditions, this analysis points out that otherwise unexplained visceral serositis can occasionally develop also in Epstein-Barr virus infectious mononucleosis.

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Venous thrombosis in a community-acquired pneumonia

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Background: The incidence of pediatric venous thromboembolism (VTE) is significantly lower than in adulthood and occurs primarily in hospitalized children. Pediatric VTEs are mainly secondary, with the presence of a central venous catheter (CVC) being the most common risk factor. Other major risk factors are infection, malignancy, chronic inflammatory conditions and inherited hypercoagulable state. Duplex ultrasound (US) is the first diagnostic tool and management depends on the etiology. Short and long-term sequelae are frequent and can be associated with significant morbidity.

Case description: We report the case of a previously healthy 3-year-old child with a 5-day history of fever, cough, fatigue and progressive respiratory distress. A diagnosis of left pneumonia with a small apical effusion was made and intravenous (IV) Cefuroxime was started. She also received IV Albumin and Furosemide for signs of capillary leak syndrome. After a week, pleural US showed a larger effusion and a chest tube was inserted. On the next day she developed oedema of the left arm and Duplex US performed to assess the presence of VTE was normal. Due to persistent painful oedema with appearance of collateral veins the Duplex US was repeated and revealed a thrombosis of the left internal jugular vein (IJV) and left subclavian vein. Subsequently, subcutaneous

therapeutic anticoagulation with Enoxaparin was started. Computed tomography angiography carried out after new fever spikes and respiratory deterioration also showed the presence of thrombosis in the left brachiocephalic vein and a worsening of pleuropneumonia. Antibiotic therapy was changed to IV Meropenem and Vancomycin, and the latter discontinued after broad-range PCR of the pleural effusion were positive for *Streptococcus pneumoniae*. The patient was discharged after 1 month with oral Co-Amoxiclav and initially 3 months of therapeutic anticoagulation. At 2-month follow-up she had no complaints, however, partial obstruction of the left IJV persisted. 5 months later only slight signs of post-thrombotic syndrome and cutaneous collateral veins were present.

Conclusion: VTE is uncommon among children especially in the absence of a CVC. VTE can occur in any venous system but in non-CVC-related VTEs upper extremity thrombosis is very rare and diagnosis can be difficult especially in complex cases. Clinical suspicion needs further and appropriate investigations to ensure prompt treatment to minimize acute and chronic complications.

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A souvenir with different faces and a long-lasting taste

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Background: Melioidosis, caused by *Burkholderia pseudomallei*, is endemic in north Australia and Southeast Asia. Contamination occurs mainly via percutaneous inoculation, inhalation, aspiration, and occasionally by ingestion. Particularly in children it may present as skin abscess, pneumonia, parotitis or osteomyelitis.

Case Presentation Summary: A 2-year-old previously well Swiss boy, was seen for evaluation of fever for 10 days after family vacation (December) in Thailand visiting cities and staying at a beach resort. He had coryza, diarrhoea but was otherwise well in himself. On his arm he had a papule which according to his mother might have been from an insect bite. Search for Malaria, Dengue, Chikungunya, Q-fever, Tularemia were negative. Repeated blood cultures and bacterial and viral stool examination remained negative. 10 days later the papule progressed to an abscess, cough persisted. Aspirate was positive for *B. pseudomallei*. Whole-body MRI revealed a pulmonary abscess but no further organ involvement. Ceftazidime was started. After 2 days fever defervesced. After 3.5 weeks iv. Ceftazidime, the eradication phase with oral trimethoprim-sulfamethoxazole was started and continued for 6 months.

Discussion / Learning Points: This is a rare paediatric case of imported Melioidosis from Thailand manifesting as cutaneous and pulmonary abscesses. We postulate percutaneous inoculation facilitated by the skin breach from an insect bite. Melioidosis may have a wide range of clinical manifestations, and severity varies from an acute fulminant septic illness to a chronic infection. It should be part of a differential diagnosis in returning travellers from South-East Asia presenting with an acute or chronic febrile illness particularly with an abscess and/or pneumonia. Antimicrobial treatment consists of an intravenous intensive phase followed by a prolonged eradication phase.

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Bacterial pyomyositis in children: Diagnosis, treatment and outcome in 7 cases

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Background: We searched our database for all pediatric patients admitted to the Kantonsspital Aarau between 2014-18 which were diagnosed with septic arthritis, osteomyelitis, myositis and pyomyositis. 110 cases were identified in total of which we excluded all those with arthritis, osteomyelitis, viral myositis and CRMO (chronic recurrent multifocal osteomyelitis) from further analysis.

Results: 7 cases of pyomyositis were identified, median age was 3y (range 1-11), 4 female and 3 male. Only 1 patient had a history of trauma (falling on its back), in 5 cases a preceding fever episode was described. Delay from starting symptoms to diagnosis was a few days. All patients except one suffered from pain in their hip girdle, 5 were limping, 1 refused walking. Sedimentation rate (ESR, range 31-131 mm/h) and CRP (range 39-160 mg/l) were elevated in all cases, positive blood/abscess cultures occurred in 4 patients (3 *Staphylococcus aureus*, 1 Group A *Streptococcus*). 6 patients underwent MRI for definitive diagnosis, 1 had ultrasound of left M. infraspinatus only. 6 patients showed abscess formation or in-

flammation in hip muscles (M. psoas, gluteus, iliacus, obturatorius, piriformis), 3 required surgical drainage. In 3 cases MRI revealed inflammation in adjacent joints or bones. Initial antibiotic treatment was started intravenously with Amoxicillin/Clavulanic acid in 3 patients, Clindamycin in 2 and Flucloxacillin or Ceftriaxon in one. Rapid clinical amelioration and reduction of ESR/CRP was observed in all but two cases. Mean time of antibiotic treatment was 14 days, therapy was switched to oral antibiotics after 3 to 5 days. No relapses were reported.

Conclusion: Over the last five years we observed bacterial pyomyositis becoming more frequent, MRI imaging seems to contribute here. Referring literature is still scarce. Especially in young children, diagnosis is challenging because of atypical signs and symptoms mimicking osteomyelitis. MRI leads to final diagnosis although ultrasound may be useful. Early start of antibiotic treatment seems to reduce need for surgical intervention due to abscess formation. Switching intravenous therapy to oral antibiotics quite early was possible without impairment of the outcome. Concomitant inflammation of adjacent joints or bones can be seen in MRI without clear signs of acute bacterial arthritis or osteomyelitis and completely resolves after treatment.

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An uncommon (Panton-) Valentine's day: when lungs date MSSA

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Introduction: Methicillin-sensitive *S. aureus* (MSSA) infection is common in children. We report a case of multiple disseminated pulmonary abscesses, due to MSSA infection, positive for Panton-Valentine leucocidin (PVL) toxin, in a previously healthy patient.

Case: A 6 y.o boy presented to the emergency Department with a 5 day history of fever, chest pain, and no cough. Extensive physical examination showed only localized reproducible chest pain on palpation. Chest X-ray was normal and C-reactive protein (CRP) was 10 mg/l. Because of his origin and family history, sickle cell disease was suspected and empiric intravenous (IV) ceftriaxone was started. 48 hours follow-up showed persistent fever. Blood cultures were positive for MSSA and CRP went to 102 mg/l, leading to patient hospitalization. Suspecting osteomyelitis, a chest CT-scan was performed and revealed multiple round intraparenchymatous and subpleural consolidations, centered on vessels. After 10 days of IV flucloxacillin, total body PET-CT showed regressing condensations with presence of excavations, without other abnormality. Extensive workup was negative, including screening for tuberculosis, endocarditis or immune deficiency. Sickle cell disease was excluded by genetic testing. Finally, PVL toxin was identified on blood cultures. The patient completed a 14 days course of IV flucloxacillin, followed by oral clindamycin for 6 weeks. A decontamination treatment was recommended for the whole family. 6 months follow up showed disappearance of lesions on CT scan.

Discussion: Most often associated with Methicillin-resistant *S. aureus* in USA and MSSA in Europe, PVL exotoxin leads to leukocyte destruction and tissue necrosis. Diagnosis can be made by immunochromatography or specific PCR. PVL is a virulence factor for skin and soft-tissues infection, sepsis syndrome, musculoskeletal diseases and necrotizing pneumonia. When associated with pneumonia, clinical findings may include hemoptysis, leukopenia, tachypnea, tachycardia, and often evolve to acute respiratory distress syndrome. PVL-positive skin and soft-tissues infections increase the risk to require surgery compared to PVL-negative. Test for PVL toxin should be considered for invasive or recurrent disease, and in case of positivity, decontamination must be proposed.

Conclusion: In our case, the clinical presentation was surprisingly atypical with mild symptoms compared to the radiographic imaging and fortunately our patient had a favorable clinical outcome.

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Persistent torticollis

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Case report: 15 y.o. healthy boy was referred for 7-day torticollis and elevated CRP (137mg/l) following initial fever and sore throat, he had been afebrile for 5 days. Tonsils were erythematous with painful and stiff neck. Injected CT scan showed bilateral atlanto-occipital (AOA) and atlanto-axial arthritis (AAA) with epidural collection and infiltration of soft tissues. CT guided articular puncture was performed and patient treated with IV co-amoxicillin. *Streptococcus agalactiae* grew from throat and articular fluid cultures. Blood culture was negative. Echocardiography excluded endocarditis. MRI showed no osteomyelitis. Co-amoxicillin was switched to ceftriaxone (10 days) followed by oral clindamycin (8.5 weeks). Clinical and radiological evolution at 6 weeks was good. Control MRI at 12 weeks showed arthritis improvement. Clinical recovery was complete at 5 months.

Discussion: To our knowledge, this is the first case of *Streptococcus agalactiae* cervical arthritis encountered in paediatric population. Septic AOA and AAA are extremely rare in children. Only one case of septic AOA was described in an 8-m.o. infant as a complication of *S. pneumoniae* otitis and 4 cases of septic AAA in children 11 to 14 months (no articular puncture, negative blood culture). In these cases CT scan were normal and MRI led to diagnosis. IV antibiotics alone permitted full recovery. Increasing number of *S. agalactiae* septic arthritis are described in adults. They affect same joints as typical septic arthritis, but they are often polyarticular and associated with cellulitis. Risk factors are immunocompromised patients, debilitating conditions and invasive manoeuvres. In paediatric population, *S. galactiae* arthritis are mainly described in neonates and are responsive to betalactam. In our case we speculate that physiopathology could be direct propagation of *S. agalactiae* from throat infection through soft tissue inflammation.

Conclusion: Painful and persistent torticollis, even afebrile, should be investigated by cervical MRI. Septic AOA and AAA must be excluded. *S. agalactiae* is not only involved in neonatal sepsis but can be responsible for septic arthritis in older children.

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Enteroviruses: A cause of symptomatic congenital infection

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Clinical case: A male infant, born by uneventful spontaneous vaginal delivery at 38 4/7 weeks, with a birth weight of 3350g. The antenatal history was unremarkable except for colonization with Group B streptococci and rupture of membranes had occurred 8 hours before delivery. At 74 hours of life, irritability, high fever (max 40.4°C), diarrhea and a cutaneous rash were reported, while the mother had developed diarrhea, a cutaneous rash and fever 24 hours earlier. He was admitted for IV antibiotic therapy. Infectious workup revealed a neutrophil left shift and an elevated C-reactive protein (CRP) of 28 mg/L. Work up 48 hours after initiation of antibiotics, CRP increased up to 46 mg/L. Blood and urine cultures were sterile. Lumbar puncture was performed, and cerebrospinal fluid (CSF) showed no pleocytosis, culture was negative and PCR was positive for enterovirus (EV). Antibiotic treatment was terminated after 72h. On day 3 of illness, desaturations without apnea were observed requiring oxygen therapy for 2 days. Chest radiograph, EEG, cerebral IRM and cardiac, hepatic and renal work up were normal. Evolution was favorable and the newborn was discharged home at day 9. The diagnosis neonatal EV encephalitis was retained.

Background: EV infections are common, causing 10-15 million symptomatic infections per year in USA, one third of these are infants under the age of one. Most often, these infections cause mild upper respiratory tract infections, self-limiting gastroenteritis, or are asymptomatic. Rarely, EV infections give rise to severe illness causing sepsis, hepatitis, coagulopathy, myocarditis, pneumonitis and meningoencephalitis. Though not well described in the field of neonatology, studies suggest that EV causes 3% of suspected systemic infections in neonates. Severe illness is more common when vertically transmitted or when symptomatic during the first 8 days of life. Clinical and laboratory follow up are recommended, particularly hepatic function and hemostasis.

Conclusion: This case underlines the importance of considering the diagnosis of EV infection in a newborn with febrile illness. Newborns are at higher risk for developing complications particularly in the first week of life.

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Atypical presentation of atypical bacteria

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Case report: 10 y.o. girl had 2 weeks h/o cough, runny nose and fever. Family physician suspected pneumonia and initiated oral amoxicillin with no improvement 3 days later. She was then referred for hospitalization. Examination revealed right lung hypoventilation with dullness on percussion. Work-up showed mild inflammation (CRP 45mg/L) without leucocytosis. Chest X-Ray revealed right-sided basal infiltrates difficult to distinguish from pleural effusion, leading to thoracic-CT which confirmed a basal opacity without necrosis, discrete effusion and hilar lymphadenopathy. Treatment was switched to ceftriaxone. Consequent to persistence of fever and lack of clinical improvement screening for atypical germs and *Mycoplasma pneumoniae* was performed. Yield was positive for *Mycoplasma pneumoniae*. We changed antibiotherapy to azithromycin, leading to patient's quick clinical status improvement.

Discussion: *Streptococcus pneumoniae* is the most frequent bacteria responsible for community-acquired pneumonias in children. But large coverage with PCV 13 vaccination has led to emergence of other bacteria. *Mycoplasma pneumoniae* is an intracellular bacteria that causes infection in humans, and is responsible for up to 40% of community-acquired pneumonias in children over 5 years of age. Radiological features frequently show extensive patchy opacities and thickened bronchial shadow. *M. pneumoniae* can also produce extensive infiltration and solid changes as observed in our case. Pleural effusions can be seen. Empyema is rare complication in *M. pneumoniae* pneumonia. The microbiological diagnosis is done by PCR.

Conclusion: Persisting symptoms in a well-treated child with suspected pneumonia, must lead to search for atypical germs regardless of a localised opacification on the chest X-ray. Pleural effusion can complicate pneumonia due to *Mycoplasma pneumoniae*. Macrolid antibiotics are the treatment of choice.

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Epidemiology and characteristics of fractures secondary to child abuse

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Introduction: Child physical abuse (CPA) may result in fractures, the most common lesion after bruising. Infants under one year are at higher risk. The aim of this study was to describe characteristics of CPA cases with fractures.

Methodology: The study, approved by the Ethical Committee CER-VD (n° 2016-02157), included children aged 0 to 18 years with fractures secondary to CPA diagnosed at the University Hospital of the Canton de Vaud (CHUV) between 01.09.2007 and 31.08.2017. Patients were stratified into 3 age groups (<1 year, 1 to 5 years and >5 years).

Results: 41 patients initially presented 84 fractures. This number rose up to 106 after further assessments (22 additional fractures). The most frequent fractures involved long bones (51 out of 106, 48%). There were 39 rib fractures, representing 37% of all fractures. They accounted for nearly half of those found in patients younger than one year (32/77 fractures, 42%), 25% in the 1 to 5 y.o. group (5/20 fractures) and 22% in the older group (2/7 fractures). There were 12 skull fractures (11% of all fractures). The last 4 fractures (4%) affected the tooth, nose, clavicle and hand. Associated non-bony lesions were present in 21 of the 41 children (51%). A limb's functional impotence was found at admission in 16 cases (39%), 6 children (15%) had a suspicion of CPA and 19 (46%) other reasons not related to CPA.

Discussion: Most children (61%) with fractures secondary to CPA were less than one y.o. One child out of two suffered from associated non-bony lesions, the most common being bruises, followed by typical lesions diagnosed in Abusive Head Trauma (intracerebral lesions with or without retinal hemorrhages). The reason for a medical consultation in abused children with fracture was not related to any kind of trauma in near 50% of cases.

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Recurrent arterial thrombosis in children: A diagnostic challenge

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Background: Arterial thrombosis is rare in children. The most frequent etiology is iatrogenic. A recent systematic review assessed the cumulative incidence of thrombosis at 24% for indwelling arterial catheters. However, other causes exist such as congenital disorders. In addition, there exist differences in the physiopathology between thrombosis in adults and children requiring an adaptation of treatment.

Case presentation: A 6-month old girl, without a significant past medical history, presented with a viral infection (RSV) in a state of shock to our hospital in Geneva. During hemodynamic stabilization, bilateral femoral arterial catheters were placed. Following the appearance of an asymmetry in the palpation of the femoral pulses, an angio-CT was performed and showed an aortobiliac thrombosis. A percutaneous thrombectomy was performed and intravenous unfractionated heparin was administered for 10 days. The patient was switched to low molecular weight heparin and a therapeutic level was achieved. Three days later, the patient presented with a mottled left leg. An echography and angio-MRI revealed a new thrombosis in the left iliac artery. The patient was again switched to intravenous unfractionated heparin with complete repermeabilisation after 10 days. In the familial history, we note a history of miscarriages in the mother and maternal grandmother. Due to familial history and recurrence of thrombosis, a thrombophilia screening was performed and the patient was found to have a positive lupus anticoagulant (LA). The mother was not positive for the LA. Iatrogenic thrombosis resulting from femoral artery catheterization in a critically ill infant with concomitant infection was retained as the principal diagnosis. The clinical relevance of LA as a prothrombotic risk factor in this context needs to be confirmed by the persistence of the antibody in follow-up blood work. In fact, transient LA positivity is frequently observed in children with infectious diseases or other stressors.

Conclusions: This case reveals the difficulty in the management of arterial thrombosis in the pediatric patient. Though clinical guidelines were applied for the treatment in this patient, these guidelines are not adapted to the pathophysiology in children. A treatment more suited to these mechanisms could be more effective. Studies investigating the prevention and treatment of arterial thrombosis in children are needed to improve the management of patients affected with this pathology.

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Continuous vocational training in child abuse and neglect: Improving safety for young patients.

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As paediatricians, we are daily potentially faced with various forms of abuse and neglect perpetrated against children. It is thus critical that every healthcare provider, who daily manages children, – paediatricians, emergency doctors, paediatric and emergency nurses – is well aware of the essence of the mistreatment and of its extremely complex implications. Healthcare providers should be trained with specific knowledge for early recognition of situations of potential abuse and neglect, as well as with tools to appropriately react to suspicious and dangerous situations. We all know that, as people who dedicate our whole life to “cure” children, situations of abuse and mistreatment are stranger to our comprehension and for this reason we instinctively tend to reject and mentally deny their existence. In order to face this situations, at the “Children’s and adolescents’ development care unit” of the Italian Switzerland Paediatric Institute, it is active a group aimed at the wellbeing and protection against childhood and adolescence mistreatment (GIMI). Working as a team, with different paediatric specialists, enables to actively prevent the “unconscious reflex of denial” and allows to be really protective towards children at risk. It is thus a priority to guarantee an accurate knowledge and culture of the problem through various forms of teaching supports – case reports, discussions, learning of practical skills – in order to assure a comprehensive and global understanding of the topic. Key points of

this continuous learning should include: Regular re-definition of the role of the paediatricians and of the “wellbeing and protection group” in the diagnosis and the management of child abuse and neglect; Collective discussion of case reports; Implementation of practical laboratories with radiologists, paediatric gynaecologists and paediatric psychiatrists; Improvement of the interaction with the authorities (police, attorneys, judges).

In conclusion, a continuous training of the paediatric healthcare providers will facilitate the identification and the management of various forms of child abuse and neglect during daily activities. This will promote a prompt and effective intervention in case of concern for the health and safety of children.

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Life experiences of Senegalese children suffering from cardiopathy and their families cared for by Terre des Hommes in Senegal and Switzerland

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Background: The high prevalence of congenital and acquired cardiopathies in sub-Saharan Africa's led to the development of a health program by Terre des hommes aiming to transfer children suffering from cardiopathy in European countries, including Switzerland, to receive specialized care. Given the high complexity of these procedures, our study aimed at determining (1) how Senegalese children suffering from cardiopathy and their families experience the disease and care before, during and after a stay with Terre des hommes in Switzerland and (2) how the professionals taking care of them perceive their experiences?

Methods: We used a qualitative methodology to collect data in two places: The National children hospital Albert Royer in Senegal and in the House of the children of Terre des hommes (Massongex) in Switzerland. Altogether, 31 semi-structured interviews were conducted: 3 with Senegalese children during their stay in Switzerland, 7 with children in Senegal after having been cared for in Switzerland, 15 with their family members in Senegal, 1 with a medical doctor in Senegal and 5 with professionals from Terre des hommes caring for the children in Switzerland. All interviews were transcribed verbatim, anonymised, and analysed with an inductive method.

Results: This study put forward the life experiences of these children and their families and the perspective of the professionals caring for them such as: The children's physical pain causing their families important emotional pain; the children's daily limitations and social exclusion caused by the disease especially by preventing them from playing with other children and going to school; the emotional pain due to the separation experienced by the children being transferred to Switzerland and their families staying in Senegal; the complex adaptation of the children when they arrive in Switzerland; the emotional difficulties experienced by the children when they have to leave Switzerland to return to Senegal; and finally fears that children and families go through, such as fear of pain, death, and surgery as well as hopes of healing and acquiring an ordinary life.

Conclusions: This study puts forward that these Senegalese children and their families experience many difficulties linked to their illness, transfer and stay in Switzerland. Given the importance of the pain caused by the separation between the children and their families, improvement of the health program needs to be considered in the future.

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Needs and perceptions of fathers with a newborn hospitalized in a neonatal unit: a descriptive study

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Background: Transition to parenthood can significantly affect parents' wellbeing, couple relationship, and attachment to the newborn child. All of these changes can cause stress and anxiety. When this is coupled with a newborn's hospitalization, the risk of parental stress is even greater and can lead to psychological, physical and social problems in the parents' lives. The transition to parenthood varies for each person,

depending on parental needs and perceptions of parenting, but it involves as much as for both, the mother and the father. However, compared to mothers, little is known on fathers' needs and experiences.

The aim of this descriptive study is to identify the needs and perceptions of fathers who have a newborn hospitalized in a neonatal unit.

Methods: Fathers with at least one newborn hospitalized for a minimum of 4 days and a maximum of 15 days in the neonatal unit of a tertiary referral hospital in Western Switzerland were eligible to participate in the study. Inclusion criteria were to be over the age of 18 years and be able to speak, read and write in French. Recruitment occurred between September 2018 and January 2019. Consenting completed two translated questionnaires: the "Neonatal Intensive Care Unit: Parental Belief Scale (NICU: PBS)" and the "short NICU Family Needs Inventory (NFNI)". Data were analyzed using descriptive statistics.

Results: Data collection has just ended, thus only preliminary results of socio-demographics are presented here. A total of 70 fathers completed the questionnaires (participation rate of 85%). The majority were aged between 30 and 39 years (62%, n = 44). Most fathers were Swiss (44%, n = 31) and married (60%, n = 42). The majority worked full-time (81%, n = 57). Nearly half of the fathers were on parental leave (42%, n = 29). For more than half of the participants (57%, n = 40) the hospitalized child was their first child. Hospitalized neonates were full-term (49%, n = 34), preterm (37%, n = 26) and extremely preterm (14%, n = 10). The complete results will be presented at the congress.

Conclusion: In the transition to parenthood, paternal involvement is an essential factor, which so far has received little attention in research. Results of our study will provide new data on how fathers perceive their experience in the NICU and related needs. They will constitute a solid base to inform how to best support them in the Swiss context.

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Perimorten caesarean section on an ambulance: case report

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Introduction: The authors describe a rare case of Perimortem Caesarean Section, performed on an ambulance in an external hospital emergency call, with survival of the fetus in the immediate future and a year from the event (without neurological deficiencies). This fact is analyzed both from a legal (according to the Italian law) and ethical-deontological aspect. In particular: the behaviour of a non specialist in obstetrics in a non-hospital environment, the role of international guidelines (conceived for the intra-hospital environment) and the value given to the evaluation of the chances of the mother's survival are analyzed.

Methods: Case report.

Results: A few cases of perimortem caesarean section executed in an extra-hospital environment are described in literature and most of the international guidelines are conceived for an intra-hospital approach (protected environment, technical equipment, different specialists). International guidelines for the extra-hospital emergency exist, as, for example, the ATLS[®], according to which the perimortem caesarean section is indicated in all the cases of cardiovascular arrest of the mother, whatever the cause might be, with the only exception of hypovolaemia. From the analysis of the available retrospective studies in literature, the cases in which the cut executed in a 'hostile' environment has determined the survival of the fetus without neurological sequence caused by hypoxic damage are relatively rare.

Conclusion: In the presented case the initial approach of the rescue team has been focusing on the mother's reanimation attempt only, following the indications of the known aphorism 'save the mother, save the fetus' and therefore trying to evaluate and correct the possible reversible causes of PEA. After having correctly evaluated the situation, the team acted believing that the best possibility of success in the attempt to in saving the fetus' life would have been represented by the attempt to save the mother's life. Gained the awareness of the inefficiency of the manoeuvres and of the likely fatal outcome for the mother (that presented fixed non-reacting mydriasis, possible rhinorrhoea, swollen jugular, cape cyanosis), the doctor decided to extract the fetus in the extreme attempt to save it and to eliminate an important physiopathological obstacle for the cardiac venous return, allowing in this way an extreme ultimate chance of success also in reanimating the mother.

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A systematic review of clinical practice guidelines for acute procedural pain on neonates

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Introduction: The evolution of neonatal intensive care is associated with improved survival of the neonates. During hospitalization in NICU (Neonatal Intensive Care Unit), neonates are exposed to many painful procedures within a stressful environment. Inadequately treated pain can lead to short and long-term complications from hyperalgesia to alterations of the pain experience. To date, many evidence-based guidelines are available. However, the quality of these guidelines and their clinical application remains unclear. Additionally, systematic prevention of procedural pain is rarely implemented in NICU and pain remains undertreated.

Objectives – aims: Determine the quality of the existing guidelines on the management of procedural pain in neonates and identify the most relevant evidence-based interventions to prevent procedural pain in neonates.

Methods: In total, four electronic databases (Embase, Pubmed, CINAHAL, Joanna Briggs Institute Database) and grey literature published between 2007 and 2018 were searched using keywords such as procedural pain and neonatal care. The methodological quality was analysed using AGREE II (Appraisal of Guidelines for Research and Evaluation).

Results: A total of 1154 records were identified from electronic databases and other sources. After screening for eligibility, 17 guidelines were included in this review. Among these, 11 were identified to be high quality guidelines. Besides the usual recommendations for pharmacological and non-pharmacological treatments, inclusion of parents, improving interprofessional collaboration and considering the setting were identified as important elements.

Conclusion: The results of this review show that there is a need to improve the methodological quality of the guidelines for procedural pain in newborns. The set of recommendations for procedural pain prevention needs to involve not only pharmacological and non-pharmacological pain treatment but also parents and interprofessional collaboration. Hence, these elements need to be included in the development of a complex intervention to prevent procedural pain in neonates.

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Development of an intervention to improve the management of painful procedures in neonates

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Introduction: The evolution of neonatal intensive care has increased survival of prematurely born infants. During hospitalization, neonates are exposed to a stressful environment and a high number of painful procedures. If pain is not treated adequately, short and long term complications may develop. Despite evidence about neonatal pain and available guidelines, procedural pain remains undertreated. This gap between research and practice is mostly due to limited implementation of evidence-based knowledge and time constraints.

Objectives – aims: The aim of this study is to develop a complex inter-professional intervention to improve the management of procedural pain in neonates.

Methods: This intervention is based on Craig's "Social Communication Model of Pain" and on a systematic review of evidence-based guidelines. The Craig's model provides an inclusive framework for organizing and considering biological, psychological, social determinants of pain and the context. This model takes into account the neonate and all those who care for him/her (parents and health care professionals). The guideline review allows for extraction of evidence-based recommendations. To include important stakeholders, we conducted a 2-round electronic Delphi

process. The Delphi panel included pain experts, healthcare providers of the participating unit and parents of neonates.

Results: The complex interprofessional intervention called NEODOL, is developed and designed for three groups: health care professionals, parents and neonates. A total of 16 panellists participated in the Delphi process. At the end of the Delphi process, the panellists endorsed the NEODOL intervention as important as well as feasible.

Conclusion: Complex interprofessional interventions need theoretical bases, careful development and integration of stakeholders to provide a comprehensive approach. Conducting this Delphi study was key to include important stakeholders in the development of the NEODOL intervention. Thus, an important step toward testing this intervention in a pre-post study at the neonatology unit of a cantonal hospital in Switzerland was made.

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Youngest patient treated with tisagenlecleucel for infant B-ALL

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Case report: We present the case of the first pediatric Swiss patient treated with tisagenlecleucel (Kymriah®) outside a study, and the youngest in the world receiving this specific type of CAR-T. Our 15 month old patient was diagnosed with an infant precursor B-acute lymphoblastic leukemia (infant B-ALL) in January 2018 at 5 months old. At diagnosis hyperleucocytosis was seen and CNS and testis were negative. Cytogenetic analysis showed MLL-rearrangement. He was treated according to Interfant 06 protocol and was prednisone poor responder. Molecular residual disease (MRD) was negative ($<10e-5$) after induction therapy. CSF relapse occurred during consolidation chemotherapy, treated with intrathecal chemotherapy before a 8/8 umbilical cord allogeneic stem cell transplantation (SCT) in May 2018. 2 months after SCT, disease relapsed with 40% blasts in bone marrow. CSF and testis were negative. MRD before CAR-T was 4.7 10e-3. Leukapheresis was performed 3 times in October 2018 due to age-related difficulties. CAR-T cells therapy was administered in Frankfurt with the HUG pediatric team in December 2018 (2x10e6 transduced Tcells/kg). At Day 14, B cells aplasia was noted and no significant toxicity. Bone marrow at Day 28 was MRD negative with 25% of CAR-T cells found in peripheral blood. The patient is now under IVIG therapy and asymptomatic.

Discussion: B-ALL relapse after SCT remains a leading cause of pediatric cancer deaths. Tisagenlecleucel is a new treatment based on genetically-modified T cells with chimeric antigen receptors (CAR) targeting CD-19 and was accepted by Swissmedic in 2018. Early studies found unprecedented results in relapsed B-ALL with initial remission rates as high as 70 to 90%. Durable remissions up to 24 months were observed. Long term remission is directly linked to duration of CAR-T cells in peripheral blood. Treatment failure seems linked to early loss of CAR-T cells or CD19-negative relapses. CAR-T cells pass into the CSF and were able to treat CSF relapse in some studies. Complications consist of cytokine release syndrome (80% of patients) ranging from mild to severe, neurotoxicity and B cell aplasia. Most of them resolve in less than a month with supportive treatment.

Conclusion: CAR-T cells therapy is a promising treatment for relapsed B-ALL. Ongoing studies are focusing on better duration of CAR-T cells and finding new targets for CD19 negative disease. This therapy could become part of standard care for ALL-B in Switzerland.

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Perinatal central cyanosis with a clinically favourable outcome: HbM-Iwate

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Background: The HbM-Iwate occurs rarely; mainly in the Iwate region of Japan, and a few cases are reported in central Europe, India, South America. A mutation in the alpha haemoglobin (Hb) causes the haemoglobin group to oxidise and form MetHb. It is responsible for cyanosis and low percutaneous oxygen saturation values. The mutant alpha chain of HbM-Iwate structurally interacts with and lowers the Hb oxygen affinity of the beta globin, which in turn allows oxygen to dissociate from the Hb more readily. Hence no severe oxygen insufficiency exists.

Case report: A female term neonate was born in a peripheral hospital. The pregnancy was without complications, no intake of medications or other drugs. Spontaneous birth, Apgar 9/9/9, umbilical artery pH 7.21, birth weight 3810 g. In the first hour of life, central cyanosis was observed. Percutaneous saturation was at 60-70%, and remained around 65% with oxygen supplementation for the next 5 days. The arterial blood gas analyses showed a constant saturation of 78-87% and a composition of 10% MetHb and SulfHb. An echocardiography and chest x-ray were performed without pathological findings. The haemoglobin analysis in HPLC showed no abnormalities, oxygen affinity for HbF was lowered, mass spectroscopy indicated an abnormal alpha globin, and the sequencing of alpha globin gene showed a heterozygous mutation at the position alpha 1 CD87 (C->T). In literature this mutation is described as the HbM-Iwate variant. The mutation was not found in the child's parents. Clinically the child continued to display a central cyanosis in the first 5 years of life, with no other pathological findings and a normal childhood development.

Conclusion: This case illustrates that in presentation with congenital central cyanosis life threatening causes have to be ruled out first (cardiological, pulmonary, dangerous MetHb induced by enzyme deficiencies, drugs or toxins). If by further testing, the HbM-Iwate variant is diagnosed, it has a favourable prognosis, with mainly cosmetic issues of cyanosis. However, it is important to document the HbM-Iwate in the patient history, especially for future anaesthesia procedures.

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Streptococcus pyogenes endocarditis with rupture of mitral valve chordae tendineae following varicella associated necrotising fasciitis – case report and review of the literature

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Background: Varicella-zoster virus (VZV) may cause serious and potentially lethal complications such as Group A Streptococcus (GAS) associated necrotising fasciitis. GAS is rarely described as a cause of infective endocarditis (IE).

Case presentation summary: A 5-year-old previously healthy boy presented with varicella and painful livid discoloration on the left buttock on day 3 of illness. Inflammatory markers were elevated. Cefuroxime and Clindamycin i.v. were started. Blood cultures grew *S. pyogenes*. CT suggested fasciitis of the gluteal muscle and urgent surgical debridement was performed confirming necrotising fasciitis. Two further debridements were necessary and vacuum assisted closure was applied. On day 5 of hospitalisation respiratory distress and a systolic murmur were noted. Echocardiography revealed mitral valve prolapse with regurgitation. The child deteriorated further and echocardiography 2 days later showed progressive prolapse of the mitral valve, assuming rupture of the chordae tendineae. Cardiac surgery confirmed IE, the mitral valve was reconstructed and neo-chordae were implanted. No growth of other pathogens was noted. Treatment was adjusted to Amoxicillin and continued for four weeks. He survived.

Learning points / discussion: IE is rare in childhood, especially in children without congenital or valvular heart disease. GAS associated IE as a complication of VZV and fasciitis has rarely been described in children. In the past 80 years only 15 cases of IE caused by GAS in children were reported. Acute deterioration secondary to rupture of mitral valve chordae tendineae, as described in our case, has not been reported in the literature yet. Serious complications like these could be prevented by an universal varicella childhood immunization programme which unfortunately is currently not in place in Switzerland.

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Aplasia cutis congenita associated with fetus papyraceus

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Introduction: Aplasia cutis congenita (ACC) is characterized by localized or widespread absence of skin at birth. It most commonly presents

as an isolated lesion on the scalp, however ACC is a heterogeneous condition that may be due to various mechanisms.

Case report: This girl presented with congenital bilateral linear pseudo-membranous skin defects involving the abdomen, trunk, upper arms and thighs in a striking symmetrical H-shape. The initial twin pregnancy had been complicated by the spontaneous loss of twin B at 14 weeks of gestation. In view of the characteristic presentation and history the diagnosis of ACC associated with fetus papyraceus (ACC-FP) was made. The baby was admitted to our neonatal care unit and antiseptic wound care was implemented with the use of polymer foam dressings (Polymem non adhesive®), with dressing changes every other day. We observed rapid wound healing and the patients was discharged after 22 days. Full closure of the skin defects was achieved after 38 days with marked scarring.

Discussion: The striking presentation of this neonate with ACC in a symmetrical linear H-shape on the trunk and extremities is pathognomonic for the association with fetus papyraceus. It is important to recognize this entity to exclude the suspicion of epidermolysis bullosa, perinatal injuries and ACC in genetic syndromes. The pathophysiology of ACC-FP remains poorly understood. The main hypothesis proposes the event of acute hypovolemia in the surviving fetus due to vasodilatation in the dying fetus leading to ischemia in the water-shed areas of the skin. Extracutaneous organ involvement such as gut atresia, pulmonary and central nervous system anomalies has been described, however this seems to be rare. Conservative wound care instead of an aggressive surgical approach is indicated, usually leading to epithelialization within a few weeks.

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Be aware of smartphone distraction after birth! – Another case of sudden unexpected postnatal collapse associated with parental smartphone use

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Introduction: Sudden unexpected postnatal collapse (SUPC) of a previously healthy newborn is a rare but frequently catastrophic event in the delivery room. Only recently, parental distraction by smartphones has been identified as an additional risk factor for this condition.

Methods: We report a case of SUPC with subsequent moderate hypoxic-ischemic encephalopathy (HIE) occurring in the context of smartphone use by the parents and review similar cases from the literature.

Results: Around 60 minutes after birth, the midwife found a previously healthy neonate in cardiorespiratory arrest in an asphyxiating position on his primiparous mother, while both parents were occupied with their respective smartphones. Pregnancy, vaginal delivery at 40+1 gestational weeks, adaptation and initial examination had been physiologic. The boy had been put on the mother for skin-to-skin care and initiation of breastfeeding according to current standards of care. Immediately initiated resuscitation with mechanical ventilation and chest compression reestablished spontaneous cardiorespiratory function. He was transferred to the NICU for therapeutic hypothermia for neonatal asphyxia (initial pH 6.82, pCO₂ 15kPa, BE -16 mmol/l, lactate 11.3 mmol/l) and HIE (Thompson score max. 11, self-limiting cerebral convulsions). MRI on day of life 9 as well as neurologic examination before discharge were normal. At 7 months of age he showed a favorable neurodevelopmental evolution. Up to now, only 4 cases of SUPC associated with maternal/parental smartphone use have been reported in the literature with similar conditions and outcomes.

Discussion: Smartphones are the most popular electronic device used ubiquitously today. Midwives are reporting an emerging trend of constant smartphone use around and after delivery even during the vulnerable phase of perinatal transition and early bonding. Distraction by smartphones is a well-known risk factor for traffic accidents, making it highly plausible to be a risk factor for SUPC as well. This seems particularly important in the context of unexperienced parents and while the baby is in prone position on or near the mother's breast.

Conclusion: Health care professionals working in the delivery room and maternity services should be aware of this easily preventable condition and provide appropriate surveillance and information for parents.

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A newborn with dextrocardia, coarctation of the aorta and ear malformation without prenatal diagnosis – A case report

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Despite the progress of the antenatal ultrasound screening, there still are malformations which are discovered after the delivery. These newborns and their family need a particular attention.

We report the case of a term newborn delivered via c-section who needed neonatal resuscitation with aspiration. The chest x-ray was performed because of the persistent heavy oral secretion. It showed a dextrocardia. The first neonatal examination brought out an ear malformation and weak femoral pulses but without a differential in blood pressure between the upper and the lower extremities. The newborn was hemodynamically stable. It was decided to transfer the newborn in an intensive neonatal care unit for further investigations. A coarctation of the aorta has been identified and surgically corrected. No other abnormalities were discovered. Genetic screening will be performed in february 2019 but until now no syndrome has been diagnosed.

With this case report we would like to summarize the management of a newborn with multiple malformations and draw attention to the malformations discovered in the postnatal period. Finally we would like to do a literature review of this association of malformations.

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Abdominal mass in a female full-term newborn

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Background: Abdominal masses and cysts are a common finding in prenatal ultrasound scans in fetuses affecting approximately 1:1000 pregnancies. The differential diagnosis of an intra-abdominal mass includes gastrointestinal tract disorders, genitourinary tract disorders and miscellaneous disorders. The correct prenatal diagnosis can be difficult.

Case report: We present the case of a female full-term newborn with an abdominal mass, which was first detected by prenatal ultrasound at 33 2/7 weeks of gestation. The mass measured 49 x 23 x 48 mm and was located in the right lower abdomen. As it had both solid and fluid components and no peristalsis and because of its location, the finding was interpreted as an intestinal obstruction at the level of the cecum. The mass was unchanged in ultrasonography done at 38 3/7 weeks. The newborn was born by cesarian section at 39 weeks of gestation and was vigorous with Apgar scores of 8/9/10. The physical examination was normal and the baby passed urine and meconium within the first 24 hours of life. An abdominal ultrasound of the newborn showed that the mass originated from the right ovary. On the 4th day of life the newborn was operated and an ovarian torsion with an enlarged and necrotic right ovary was found and an oophorectomy was performed. The diagnosis was confirmed on pathology and there were no signs of malignancy. The baby was discharged home on the 6th day of life.

Discussion: Despite advances in prenatal ultrasonography, the origin of an abdominal mass can be difficult to differentiate, particularly in the presence of complex masses with fluid, septated and solid components. An ovarian mass in a newborn is most commonly a physiologic cyst resulting from maternal hormonal stimulation in utero. An ovarian cyst increases the risk of ovarian torsion occurring, it is however uncommon for it to happen in the prenatal or perinatal period.

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Supernumerary nostril: a case report

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Introduction: Supernumerary nostril is a rare congenital malformation of the nose. It can be bilateral or unilateral, and can be associated to other congenital malformations. The treatment is surgery. A review of the literature revealed 50 published cases, the majority of which are reported in Asian regions. To our knowledge, this is the first published case in Switzerland.

Case report: Male sex neonate born at term from elective caesarean delivery for breech presentation. Pregnancy was achieved by donor oocyte in vitro fertilization (IVF-ET). Maternal serologic tests were normal. Maternal age was 46 years. On physical examination a right accessory nostril connected to the right nostril was immediately evident. The supernumerary nostril was located above the right nostril. Normal ipsilateral nostril was a little smaller and depressed compared to the contralateral one, but the size of the alar base and projection were similar to the left nostril. The internal nasal cavity structure was normal, as well as the bony structure of the face. The supernumerary nostril was round shaped. It was lined by intact skin and had an external diameter of 3 mm. Squeezing the accessory nostril revealed the spillage of about 1-2 ml of a gelatinous yellowish material. The internal diameter of the accessory opening was 1 mm and ended blindly at 1-1,5 cm depth. No alar cartilage was detected at palpation. There was no flaring or expansive effect of crying on the supernumerary nostril. No communication between the supernumerary nostril and the underlying ipsilateral nasal cavity was detected with nasal endoscopy and with methylene blue injection. Postnatal echocardiography, abdominal and cerebral US were negative for other associated malformations.

Conclusions: The clustering of the majority of reported cases of supernumerary nostril in Asian countries raises the suspicion about a possible co-causative role of genetic and environmental factors on the development of the condition. We cannot exclude that the advanced maternal age and the oocyte manipulation, may have played a role in the development of the anatomical anomaly in this case. No consensus has been reached on the best time to perform the surgical procedure. It is although recognised that early intervention has a better impact on social and psychosocial development and functional reconstruction.

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A mysterious case of severe intrauterine growth restriction

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Trisomy 9 mosaicism (T9M) is a rare chromosomal abnormality with less than 100 cases described in the literature. Because of an extremely variable phenotypic expression, it is probably quite often undiagnosed.

We report the case of a premature girl born at 30 1/7 GA by emergency cesarean section due to a pathological CTG. The pregnancy was marked by a severe and early intrauterine growth restriction (IUGR) and an oligoamnios. The triple test showed a 1/30 risk of trisomy 21, but the non-invasive prenatal screening was normal. She had a good APGAR score and was hospitalized for her prematurity and low weight (705g). She was diagnosed with severe harmonious IUGR, which was first thought to be of vascular origin, because of a hypotrophic and calcified placenta. She developed amongst others a pulmonary immaturity and a subependymal hemorrhage with cerebral cysts. We progressively noticed a narrow skull shape, hyperplasia of the upper lip brake, submucosal cleft palate, hypoplasia of the right thumb, hemihypoplasia of the right leg, global hypotonia, poor wakefulness and severe orality disorders. Familial history of consanguinity or genetic conditions was negative. The array-CGH showed T9M, confirmed by constitutional karyotype, with a 10% rate of mosaicism. We ran a complete check-up showing a patent ductus arteriosus and an open oval foramen. The polysomnography revealed an obstructive sleep apnea syndrome. The pH-metry revealed a gastroesophageal reflux without acidity. On the abdominal US we discovered a renal asymmetry to the detriment of the right kidney. Her cerebral MRI showed a ventricular dilatation and no T1 hypersignal of the posterior pituitary gland nor stalk evoking a pituitary stalk interruption syndrome. Hormonal investigations revealed a peripheral hypothyroidism. We diagnosed an alternating divergent strabismus, a pathological right acoustic otoemission and a congenital dysplasia and instability of the hip. She was operated at 2 months of age for an uncomplicated inguinal hernia. At term she was still completely fed by NG tube. T9M is associated with a multitude of pathological conditions described in our case. Physicians should look carefully for physical abnormalities evoking a genetic syndrome if faced with a severe harmonious IUGR, even if a maternal or vascular cause is already suspected. Knowing the diagnosis helps the patient to have the best medical care and follow-up and a new case report could raise awareness of this condition.

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When the clinical exam reassures

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Case report: A term newborn girl presented with vomiting at D2 of life. Pregnancy was marked by diet-treated GDM. Delivery was physiological, liquor was meconium-stained. Initial clinical exam (H15 of life) was normal. Child had already passed meconium and urine, and breastfed well. She looked tired the following day, refusing breast. She was supplemented with formula, and had glairy, yellowish regurgitations, becoming bilious vomiting at H40. She was pale with reduced spontaneous movements but responded to stimulation. Abdomen was flat, non tender, with normal sounds. Blood gases revealed slight lactic acidosis (lactates at 4 mmol/l). Abdominal X-ray showed normal gastric, intestinal and colic air distribution, but no air in the rectal zone. Abdominal US showed decreased intestinal motility and target sign in the pelvis with no sign of volvulus. Baby was transferred to tertiary centre: clinical evaluation, repeat US (mesenteric vessels not visualized) and enema did not allow to make a diagnosis. Within hours, she developed shock and massive rectorrhagia. Emergency laparotomy revealed volvulus on malrotation. The child died of refractory septic shock with extended intestinal necrosis on D17.

Discussion: Bilious vomiting in newborn must evoke intestinal obstruction. Differential diagnoses are duodenal atresia, malrotation/volvulus, small bowel atresia, meconium ileus, Hirschsprung's disease, meconium plug syndrome, anorectal abnormality. Volvulus is a time-critical diagnosis, intestinal twist causes vascular obstruction that can lead to necrosis within few hours. Usual clinical scenario is bilious vomiting in first 2-3 days of life in a term newborn. GI bleeding, abdominal tenderness and severe systemic disturbances may be associated. Abdominal X-ray can be abnormal but specificity is poor. Although abdominal US may show whirlpool sign (high sensitivity and specificity), visualization of mesenteric vessels and its interpretation can be difficult. Immediate laparotomy is indicated in case of malrotation or if volvulus is suspected. Clinicians should consider volvulus in presence of bilious vomiting associated with behavior change (notable by mother and nurses) and feeds refusal, even with reassuring abdominal examination, until proved otherwise.

Conclusion: Volvulus is a surgical emergency to exclude in newborn with bilious vomiting. As taught for decades it must be suspected despite normal abdominal examination, especially if clinical history is suggestive.

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Conjugated hyperbilirubinemia: think CMV

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Cytomegalovirus (CMV) is the leading cause of congenital viral infection: it occurs in 0.3–2% of all newborns. Fetal transmission can occur in every period of the pregnancy. Symptomatic congenital infection most frequently causes intrauterine growth restriction, microcephaly, hepatosplenomegaly, petechiae, jaundice, chorioretinitis and thrombocytopenia. 90–95% of infected newborns are asymptomatic at birth but 5–15% of them will develop late sequelae like hearing loss, visual impairment and motor or cognitive deficit. We report an unusual presentation of CMV congenital infection by a baby girl born at 40+1 weeks without any perinatal and neonatal warning signs. Her anthropometric parameters were in the norm, in particular no microcephaly or growth restriction. Because of an icterus praecox, she was hospitalized at 13 hours of life to be subjected to phototherapy. The phototherapy was initially interpreted in the context of an incompatibility B0 with positive direct Coombs. Clinically the patient presents jaundice and splenomegaly. After 12 hours of phototherapy, because of the presence of high conjugated bilirubin (max 98 umol/L), the phototherapy was interrupted. An other sign of cholestasis was an elevated GGT (max 385 U/L). As possible causes we could exclude a biliary atresia and some of the most common metabolic disorders. Laboratory finding with positive CMV-PCR in urine lead to the diagnosis of congenital CMV infection. None of the possible sequelae were founded (normal cerebral ultrasound, normal ocular examination as well as otoacoustic emission). In agreement with the Guideline of the European Society for Paediatric Infectious Diseases (ESPID, December 2017) there was no evidence to start a treatment for the risk reduction to develop sensorineural hearing loss. Indeed, no studies have clearly addressed treatment in the group of "mild" disease (defined as 1 or 2 at most clinical insignificant or transient findings such as conjugated hyperbilirubinemia and splenomegaly). Considered the persistence of cholestasis, therapy

with Fat-Soluble Vitamins was required. Neurodevelopmental, audiologic and ophthalmic follow-up are required.

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The importance of an accurate eye examination in newborns, 2 case reports

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Case 1: A term born boy, 3180g, APGAR 9/9/10, delivered by 1° cesarean section with normal postnatal course was found to have very wide pupils, undetectable iris bilaterally and normal red reflexes. Family history revealed maternal aniridia and cataract, however prenatal genetic counseling was not desired. Ophthalmologists confirmed the diagnosis of bilateral aniridia. In order to screen for WAGR-syndrome a renal sonography was performed and showed no signs of Wilms tumor. Further ophthalmologic controls were organized.

Case 2: A preterm girl, 33 3/7 weeks of gestational age, 1590g, APGAR 6/9/9 with IUGR of placental insufficiency was delivered by 1° cesarean section for preeclampsia. She was admitted to the neonatologic Department with respiratory distress syndrome (wet lung), whereof she recovered rapidly. During weekly examination a unilateral lens opacity with no other pathologic findings was discovered and controlled by ophthalmologists. Bilateral cataract was diagnosed and mix eyedrops twice daily were implemented. A cataract extraction operation was organized for around the calculated birth date. Family history showed no ophthalmologic diseases, TORCH serologies, amino acids in urine and Guthrie test were normal, excluding congenital infections, galactosemia and metabolic disorders. Thus, the etiology was assumed to be due to prematurity and IUGR.

We would like to emphasize the importance of a proper eye exam in newborns. Pay attention to the red reflex, an equal pupil size and light reactivity, retinal/subconjunctival hemorrhages, conjunctivae, lid edema (mind: lid eversion, postpone the examination if necessary), fix&track. Possible pathologic findings include hyphema, mostly birth mode related, conjunctivitis, which can be physiologic, chemically induced, infectious e.g. gonorrheal, chlamydial or HSV. Furthermore, keep in mind that lens opacity, leukocoria/white reflex and ocular tissue defects such as eyelid margin defects, aniridia and iris/retina defects require an ophthalmologic referral. Lens opacity refers to congenital cataract, often associated with TORCH virus infection and many others. Leukocoria can indicate retinoblastoma. Ocular tissue defects/coloboma can be hereditary, sporadic or syndrome-associated (CHARGE, Klinefelter, Trisomia 13, DiGeorge, M. Hirschsprung, Treacher Collins Syndrome amongst others).

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Post-traumatic synechia vulvae in adolescence: a case report

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Background: Synechia vulvae or labial fusion is sealing of the labia minora in midline. It's common in pre-pubertal girls especially during infancy and usually resolves spontaneously post puberty. Treatment, if required, includes topical oestrogen or betamethasone cream, surgery is rarely required. Post-pubertal labial fusions are very rare and only few cases have been published.

Case report: A girl post-puberty presented in our outpatient clinic for posttraumatic synechia vulvae. The only peculiarity found in medical history was a small clitoris injury two years prior to presentation. Surgery was needed to resolve the adhesions.

Conclusion: Labial fusion in adolescents and women is very rare. Local injuries, even minor and several years back, are the main risk factor. Treatment in these cases nearly always involves surgery.

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Subcutaneous tissue swelling and prolonged oedema: an unexpected reaction after irrigation with Octenidini dihydrochloridum (Octenisept®)

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Objectives: In wound management, the prevention and treatment of tissue infections are ordinary practices. Penetrating hand injuries are particularly common in children and may involve a span of structures such as skin, tendons, nerves, blood vessels, bones and joints. Octenidini dihydrochloridum (Octenisept®) is a widely used antiseptic agent for disinfection of acute or chronic wounds. It has a broad spectrum of antiseptic efficacy and has become an antiseptic of first choice in many hospitals. This paper illustrates a case report of prolonged oedema and tissue swelling after irrigation of deep wounds with Octenidini dihydrochloridum.

Methods: We present the case of a 7 years old child with a dog bite on his right hand that was flushed with Octenidini dihydrochloridum and NaCl, treated with oral antibiotic and supported by a splint. Despite this treatment, the hand was swollen, red and indurated with a consequent functional limitation. No other symptoms were registered. An intravenous antibiotics treatment was initiated and an orthopedic gilet was placed. An ultrasound was performed and a massive soft tissue oedema with cellulite was highlighted, without evidence of abscess or remaining foreign material. The microbiological investigation of the wound smear did not show any significant results. The blood test did not provide evidence of any significant inflammation parameters and the patient remained without fever during the period.

Results: The boy was regularly seen in our outpatient clinic after the initial presentation, swelling and induration were still present, although started to decline in following 4 months with a complete resolution in 6 months.

Conclusion: Aseptic, painful subcutaneous tissue swelling and oedema was observed after wound lavage with Octenidini dihydrochloridum (Octenisept®). The underlying pathobiological mechanism remains unclear. Hence, we recommend not to use Octenidini dihydrochloridum in any deep or complex wounds.

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Post traumatic thoracic pain

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Case: 14 y.o. boy, arrived at emergency Department after a ski accident with frontal shock against tree trunk. He had left thoracic pain and hemoptysis. His mobile phone (on the left side of his chest) was destroyed. When recovered, he was hemodynamically stable with reduced left chest expansion, crackles and muffled heart sounds. Chest X Ray (CXR) revealed mediastinum widening and pulmonary contusion. Thoracic CT scan showed aortic dissection associated to hemopericardium, hemo-mediastinum, bilateral hemothorax and pneumothorax, pulmonary laceration and multiple undisplaced ribs fracture but no flail chest. Initial management involved fluid resuscitation, beta blockade and transfer to tertiary center for urgent endovascular repair.

Discussion: Great vessels and mediastinal injuries are far less frequently encountered in the paediatric population than in adults. Mechanism of aortic injury is sudden deceleration (road accident or fall). Diagnosis of aortic injury in child can be difficult. Clinical presentation is unspecific. Eighty percent of children who sustain a thoracic aortic tear have significant associated injuries. Only half of these have external evidence of thoracic injury. Findings on CXR may include left apical cap, pulmonary contusion, mediastinal widening, shift of the trachea to the right, downward depression of left main stem bronchus and indistinct aorta. Normal CXR is highly predictive of no injury and is often the first imaging modality to identify patients who require a CT examination. Multi-Slice CT is the gold standard to evaluate chest injuries. Early surgical intervention is indicated in majority of cases. Endovascular approach is becoming the first choice treatment for thoracic aortic injuries in children. Child, who survives until diagnosis, will be alive at discharge in more than 70% of cases.

Conclusion: Although pediatric aortic injuries are rare, it is important to consider aortic rupture after a high kinetic trauma. Early recognition influences the prognostic. Thoracic CT scan remains gold standard diagnostic tool.

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When persistent barking cough in early infancy is not only due TO laryngomalacia

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Background: Vascular rings (VR) are anomalies of the aortic arch and the most common are the double aortic arch and the right aortic arch with aberrant left subclavian artery and left ductus arteriosus/ligamentum. The VR might compress the tracheobronchial tree and the esophagus, causing symptoms such as stridor, respiratory infection/distress, wheezing, cough or dysphagia, feeding difficulty and vomiting. The diagnosis is mainly posed with CT or MR-angiography and the only definitive treatment is surgery.

Case description: We report the case of a 6 months old baby-girl, born healthy, who by the age of two months was hospitalized for the first time for respiratory issues, first interpreted as obstructive bronchitis. In the following months, the patient manifested persistent inspiratory stridor with barking cough and was repeatedly hospitalized due to multiple airway infections. Since the barking cough was persistent even in the infection-free intervals, this was first interpreted in the contest of a laryngomalacia. At the age of 6 months, the patient was newly hospitalized for the umpteenth airway infection, associated with important alimentary difficulties and stagnation of growth. In this context, an echocardiography was performed, and showed the presence of a right aortic arch without bifurcation of the brachiocephalic trunk with the suspect of VR with a right aortic arch and aberrant subclavian artery aortic button. A transit esophagogram and Angio-CT were performed and confirmed the diagnosis of VR. This was characterized by a dominant right aortic arch with origin of the right subclavian artery and of the right carotis communis artery, and by an atretic left aortic arch with origin of the left subclavian artery and the left carotid artery, without any bound with the descending aorta. The patient was transferred to a specialized university centre for surgery resection of ligament of ductus, left atretic aortic arch and diverticulum of Kommerell, with attachment of the subclavian left artery to the pleura. The patient is now free of any cardiac symptom, the chronic barking cough is persisting but no growing or nutritional problems are noticed.

Conclusion: VR are very rare but need to be considered as differential diagnosis in patients presenting symptoms that might be due to tracheal or esophageal compression. In symptomatic patients, surgery is the only definitive treatment, but the clinical outcome is excellent with resolutions of the symptoms.

P 115

Recurrent pneumonia in infancy: think congenital!

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Background: Pneumonia is a common cause of infection in children. However, recurrent pneumonia should be investigated for an underlying condition.

Description: We report the case of a boy born at term, after a normal pregnancy during which fetal ultrasounds (US) were reported as normal. The delivery was unremarkable but the child developed respiratory distress (RD) a few minutes after birth due to spontaneous right pneumothorax confirmed on chest X-ray (CXR). A chest drain was inserted and respiratory support was briefly needed; clinical and radiological evolution were excellent without any identified residual anomaly. During the following months, he presented recurrent episodes of febrile and obstructive RD at 4, 7 and 8 months, twice associated with a left basal consolidation on CXR, thus suggesting pneumonia and treated with antibiotics. Clinical recovery was fair between the episodes, with residual cough and failure to thrive. Familial history was not contributive. Neonatal cystic fibrosis screening was negative. Basic immunity screening test was normal. Initial injected chest computed tomography (CCT) showed a left lower lobe (LLL) consolidation with microcystic lesions in the periphery. There was no other parenchymal or airway anomaly, and no aberrant systemic vessel. Bronchoscopy was done at 9 months to rule out an airway anatomical defect while allowing the bacterial documentation of *Moraxella catarrhalis* 10⁴5/ml and *Haemophilus parahaemolyticus* 10⁴3/ml. Other microbiological investigations were negative. Finally, retrospective analyses of fetal US identified probable left lung abnormalities, strongly suggestive of a congenital malformation. Following two weeks of intravenous antibiotics, the CCT showed multiple cysts of varying size in the LLL. The child underwent a left lobectomy at 10 months. Lung histology confirmed

the diagnosis of congenital pulmonary airway malformation (CPAM) type "small cysts" (Stocker class II). There were signs of chronic inflammation but no associated malignancy.

Discussion: Nowadays, CPAM is increasingly diagnosed prenatally. However, at times the diagnosis is made postnatally. In particular, when facing patients with recurrent or chronic pneumonia, an underlying condition must be sought. Further radiological imaging should readily be done to identify such CPAM to avoid complications. Surgery is the therapy of choice for symptomatic and/or voluminous CPAM, but needs multidisciplinary discussion in other cases.

P 116

Unusual presentation of a cyanosis in a 3-year old child

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Background: Pulmonary arteriovenous malformation (PAVM), which is defined as the presence of an abnormal connection between the pulmonary artery and pulmonary vein, is rarely seen in children.

While the age at first presentation varies widely, the majority of patients are diagnosed within the first three decades of life. About 80–95% of PAVMs are associated with hereditary haemorrhagic telangiectasia (HHT). The clinical presentation depends on the magnitude of intra-pulmonary right-to-left shunting, which is related to the number and size of PAVMs. Patients may therefore be completely asymptomatic or present with cyanosis at rest or during exercise.

Case report: A 3 year-old boy presented in our emergency Department with mild cyanosis of the lips as well as fingertips and sometimes even toes while at rest, with exacerbation during exercise. The child had a mild cold and the mother reported that he never had oxygen problems in the first year of life. His oxygen saturation levels on room air ranged from 79 to 81%. An abnormal haemoglobin had been ruled out. A three-dimensional CT scan confirmed the diagnosis of a PAVM located primarily in the lower lobe of the right lung. Subsequently, a transcatheter device occlusion of almost all direct AV connections using 42 vascular coils was performed resulting in a normalization of the saturation. Genetic testing revealed no pathological mutation of an HHT (hereditary hemorrhagic telangiectasia, also known as Osler-Weber-Rendu disease) in the following genes: ACVRL1, ENG, GDF2, SMAD4.

Conclusion: The etiology of the PAVM remains unclear. One possibility is an underlying congenital malformation and the cyanosis has been triggered by a respiratory infection. The absence of recurrent epistaxis and multiple mucocutaneous telangiectasia, as well as the negative family history in our patient did not suggest the diagnosis of HHT. The normal MRI of the head as well as negative genetic analysis in the following genes – ACVRL1, ENG, GDF2, SMAD4 makes a diagnosis of HHT even more unlikely. Nevertheless, PAVM needs to be included in the differential diagnosis of unclear cyanosis in an otherwise healthy child.

P 117

When the magic lamp is not a fairytale

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Background: Accidental hydrocarbon ingestion may lead to aspiration resulting in chemical pneumonitis. We report the case of a previously healthy 2-year-old boy who accidentally ingested an unknown amount of lamp oil.

Description: The child was first brought to the emergency Department 45 minutes after ingestion. As he was totally asymptomatic, he was discharged home. He came back about 3 hours later due to high fever, tachycardia and respiratory distress with tachypnea, grunting and intercostal retractions. Auscultation was normal. A chest X-ray (CXR) demonstrated perihilar infiltrates, suggestive of aspiration pneumonitis. There was no inflammatory syndrome. He was then admitted for surveillance during 2 days and discharged without treatment given that the respiratory distress spontaneously improved. However, 2 days later, the fever recurred, tachypnea worsened and the auscultation revealed a new right basal hypoventilation, suggestive of bacterial superinfection. Despite starting oral antibiotics (amoxicilline/clavulanic acid), the child remained febrile with worsening respiratory distress and apathy, requiring a second admission. Blood analyses demonstrated a high inflammatory syndrome

(CRP 322mg/l, Leucocytes 9.0 G/l) and the CXR showed worsening diffuse infiltrates, without pleural effusion. Treatment with oxygen supply and intra-venous antibiotics was initiated, leading to clinical improvement within 4 days. Additional CXR done at day 10 and 2 months later showed progressive regression of the infiltrates and no pneumatocele.

Discussion: Pulmonary symptoms are the most frequent manifestations of accidental hydrocarbon ingestion. Increased volatility, low viscosity and decreased surface tension of the ingested substance increase the risk of aspiration and lung injury. Furthermore, spontaneous or induced emesis and gastric lavage increase the risk of aspiration pneumonitis. Hydrocarbons cause surfactant destruction, tissue necrosis, alveolar and interstitial edema and inflammation, all leading to decreased pulmonary compliance, increased work of breathing and hypoxemia. Aspiration pneumonitis occurs and evolves over the first 6-8 hour following ingestion; thus, patients should systematically be observed for that minimal period after hydrocarbon ingestion. Potential complications in non-severe pneumonitis include bacterial superinfection and pneumatoceles that become visible after 6-10 days when pulmonary infiltrates resolve.

P 118

Simplified quality control criteria for the multiple breath washout technique

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Background: Multiple breath washout (MBW) is increasingly used in the clinical assessment of patients with cystic fibrosis (CF). However, guidelines for real-time quality control of MBW measurements are lacking.

Objectives: We aimed to develop simple quality control criteria for prospective assessment of nitrogen MBW measurements and to validate these criteria through retrospective analysis.

Methods: A system was developed to grade MBW quality whereby A, B, and C grades indicate acceptable trials, D grade indicates questionable trials with unstable breathing pattern, and F grade denotes trials that are technically not acceptable. We retrospectively assessed 52 clinically obtained MBW test occasions (134 trials) from children in two centres (Bern: patients with CF; Hannover: patients with CF after lung transplantation) using our simplified quality control criteria. We assessed success rate of MBW, comparison of MBW outcomes (lung clearance index (LCI)) reported before and after quality control, and interrater agreement of three experienced reviewers (KR, JC, BF).

Results: While all test occasions were accepted in clinics only 69% of test occasions were accepted after quality control. From the 134 trials, 68% were acceptable (A-C grade), 19% were questionable (D grade), and 13% were rejected (F grade). There was a significant difference in LCI between accepted (median LCI 8.9 (6.4-17.8)) and rejected test occasions (13 (7.3-19) $p = 0.01$). Interrater agreement between reviewers was good (85%, $\kappa = 0.6$).

Conclusion: Not performing quality control of clinical MBW data may result in overestimation of LCI. We recommend prospective MBW quality control in the clinical setting.

P 119

Unusual presentation of a lipoma in childhood

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Background: Lipomas are common findings of the skin. However, they can solely present in the chest. Intrathoracic lipomas are very rare in children, and most are asymptomatic for a long time. They are usually discovered radiologically by chance as large shadow on a chest X-ray. A CT scan of the chest can highly suggest a lipoma due to the well defined hypodense homogeneous mass. However, the definitive diagnosis

is made histologically. These tumors are benign but can grow to a very large size.

Case report: A 5 year old healthy boy presented in our emergency room due to a history of productive cough, rhinitis and daily high fever for a few days. The initial examination by the pediatrician showed attenuated breath sounds on the left side and a blood count with mild elevated inflammatory markers. The boy was then sent to the children's hospital, where he presented a slightly reduced general condition with normal respiratory rate and oxygen saturation. The chest X-ray showed a large shadow on the left side. Suspecting a pneumonia, he was put on Amoxicillin and was discharged after 3 days. Three weeks later, he was seen in the pulmonology outpatient clinic, where the boy showed a persistent pathologic X-ray, but his lung functions were within the normal range. As a consequence, a CT scan was performed which revealed a large intrathoracic homogenic mass suggesting a lipoma. The large tumor could easily be removed by the thoracic surgeon, and the histopathology could confirm the lipoma. A clinical and radiographic follow up after 4 weeks showed normal findings.

Conclusion: In clinical daily routine, symptoms such as cough and fever with additional shadows on the chest X-ray often suggest the diagnosis pneumonia. However, if antibiotic treatment does not improve the clinical and/or pathological findings, more diagnostic investigations are needed to exclude other rare diseases, which can cause a large shadow on a chest X-ray.

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Children with monolateral periorbital swelling: not always a simple diagnosis

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Background: The clinical evaluation of a child with monolateral periorbital swelling must be carefully undertaken, as it might at times hide complex diagnoses. Although viral/allergic conditions are the most common etiological causes of periorbital swelling, there are some warning signs and symptoms that should not be underestimated. We present a case of periorbital cellulitis, which actually concealed a malignant neoplastic disease.

Case: R., a 9-years-old-boy, was admitted to our paediatric Department because of burning and itching sensation and secretions from the right eye, unsuccessfully treated with decongestant eye drops. Due to persistence of symptoms, he was evaluated by an ophthalmologist who recommended a local therapy without benefit. R. was in good health condition, with non-contributory familiar history, no allergies and in compliance with the vaccination schedule. A following paediatric medical evaluation showed chemosis, proptosis and referred diplopia. The patient was hospitalized and blood tests were performed, showing mild alteration (Hb 134 g/L, WBC 6,9 G/L, Neutrophils 4,69 G/L, Lymphocytes 1,31 G/L, CRP <1 mg/L). A broad-spectrum antibiotic therapy was started after blood culture collection. The evaluation by an otolaryngologist did not show evidence of sinusopathy. The MRI was suspicious for right extraconical endo-orbital formation with local mass effect on extraocular muscles, without radiological sign of invasion of surrounding structures. Histological specimen on the bioptic sample was diagnostic for embryonal rhabdomyosarcoma.

Discussion: Approaching periorbital swelling means considering also rare diagnoses, especially when symptoms do not respond to first-line treatments. Orbital neoplasms differ in their appearance in the paediatric population. Although many of them are benign, they could lead to significant morbidity and mortality. Important warning signs are: proptosis, unilaterally reduced visual acuity, diplopia, orbital/periorbital pain. The diagnostic approach to these diseases is grounded on MRI, which is helpful in determining possible differential diagnoses. The gold standard remains the biopsy that allows to identify the final diagnosis.

Conclusions: approaching a periorbital swelling means considering the rarest diagnoses, especially in case of "red flag" signs or an unresponsiveness to first-line treatments.

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