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001

DIAGNOSTIC PERFORMANCE OF AN UNSUPERVISED ELECTRONIC STETHOSCOPE FOR COMMUNITY-ACQUIRED CHILDHOOD PNEUMONIA IN A PAEDIATRIC EMERGENCY DEPARTMENT: A FEASIBILITY CASE-CONTROL STUDY

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Introduction: With 1 million deaths, pneumonia is the leading cause of child mortality under five years of age worldwide. World Health Organization (WHO) developed a case management algorithm for the diagnosis of pneumonia. We primary aimed to characterise auscultation sounds by a digital stethoscope in acute lower respiratory infection and to develop an integrated algorithm to precise if these physiopathological patterns could be differentiated from normal clinical auscultation. We also compared automated lung sounds analysis with current WHO algorithm for consolidated bacterial pneumonia.

Methods: An electronic stethoscope was evaluated (Littman 3200) in an ongoing feasibility prospective case-control study at the paediatric emergency department of the Geneva University Hospital from January 2016 to December 2017. Inclusion criteria were clinical diagnosis of a chest infection, age <5 years and signed informed consent by legal representative. Two experts in respiratory paediatrics segmented inspiratory from expiratory time and labelled each segmentation. Chest sounds were first transformed into Mel-spectrograms, and patches were created for the machine learning process. A pipeline containing a Convolutional Neural Network and a Hidden Markov Model was used for inspiratory and expiratory event prediction. Inter-observer reliability of lung auscultation sounds was assessed by using Fleiss'Kappa. Association between manual and computer generated respiratory rates was performed using Pearson correlation coefficient. Local Ethics Committee approved this protocol.

Results: We recruited 60 cases, analysed 48 patients in a complete cases analysis, and 5 controls, with 424 recordings segmented and analysed. Mean age was 31 months, with 32 males and 16 female patients. Mean time procedure was 5.2 minutes [4.1–6.3]. We diagnosed 21 bacterial pneumonia, 5 viral pneumonia, and 22 bronchiolitis. For bacterial pneumonia, inter-observer reliability among the experts was good, with a Fleiss' Kappa of 0.89 [0.77–0.92], CI 95%. This method significantly differentiates consolidated bacterial pneumonia from other patterns of auscultation, with an Area Under Curve of 0.89 [0.79–0.99], significantly higher than the WHO algorithm specificity.

Conclusion: The digital stethoscope provides a good diagnostic performance. This method is more objective and can be more readily standardized than subjective auscultation. It could become an unsupervised diagnostic tool for paediatric pneumonia diagnostic in low-resource settings.

Disclosure of Interest: None declared

002

THE SIMPLE 10-ITEM PARC TOOL PREDICTS CHILDHOOD ASTHMA IN AN EXTERNAL VALIDATION COHORT

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Introduction: External validation of prediction models is important for assessing applicability in populations other than the development population. The Predicting Asthma Risk in Children (PARC) tool, developed in the Leicestershire Respiratory Cohort (LRC), uses information on preschool respiratory symptoms to predict asthma at school age. We performed an external validation of PARC using the Avon Longitudinal Study of Parents and Children (ALSPAC).

Methods: We defined inclusion criteria, prediction score items and outcomes in ALSPAC to match those used in LRC. The prediction score was calculated as the sum of score-points from each scoring item. We assessed discrimination of PARC by calculating sensitivity, specificity, predictive values, likelihood ratios, receiver operator curves and area under the curve (AUC). Overall performance was assessed using Brier score and Nagelkerke's R-squared. Sensitivity analyses were performed by altering the definitions of inclusion, scoring items and outcome.

Results: The ALSPAC validation population included 2690 children with preschool respiratory symptoms of which 373 (14%) had asthma at school age. Discriminative performance of PARC was equally good in ALSPAC (AUC = 0.77, Brier score 0.13) as in LRC (0.78, 0.22). The score cut-off of 4 showed the best sensitivity (69%) and specificity (76%). Changes to inclusion criteria, scoring items or outcome definitions barely altered the prediction performance.

Conclusion: PARC performed equally well in the validation cohort as in the development cohort. PARC is a valid tool for predicting asthma in birth cohorts and its use in clinical practice is ready to be tested.

Funding: SNF32003B_162820

Disclosure of Interest: None declared

PRESENTATIONS NACHWUCHS

003

AQP1 INFLUENCES CELL WATER PERMEABILITY AND MOBILITY IN NEUROBLASTOMA CELLS

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Introduction: Cell water permeability has been implicated in mechanisms of tumor cell migration and agility. Aquaporin 1 (AQP1) is a water channel that is up regulated in several solid tumors. The aim of this study was to elucidate how AQP1 expression influences functional behaviour of neuroblastoma cells.

Methods: Cell permeability of neuroblastoma cells was visualized using confocal microscopy and quantified using stopped flow analysis. For assessment of AQP1 dependent cell motility AQP1 knockdown cells were compared to AQP1 positive cells.

Results: AQP1 expression significantly increases cell water permeability as measured by stopped flow analysis. This effect can be visualized by confocal microscopy. There are significant changes in cell motility depending on the level of AQP1 expression and AQP1 localization.

Conclusion: Neuroblastoma cell permeability and motility is influenced by the varying the levels of AQP1 expression thus modifying their ability to move. The potential of tumor cell to adapt their behavior by changing AQP1 expression might significantly contribute to tumor cell migration and the development of metastases.

Plaschke Prize: I wish to apply

I have read and understand application rules: Yes

Disclosure of Interest: None declared

004

PERIOPERATIVE COMPLICATIONS AFTER KASAI HEPATOPORTOENTEROSTOMY IN THE SWISS NATIONAL BILIARY ATRESIA REGISTRY

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Introduction: Kasai hepatoportoenterostomy (HPE) is the first line treatment for patients with biliary atresia (BA), and outcomes are mainly expressed as overall survival and survival with native liver (SNL, i.e. time until liver transplantation or death). To date, studies have focused on surgical technique- and disease-related complications, but little is known about the impact of perioperative surgical complications.

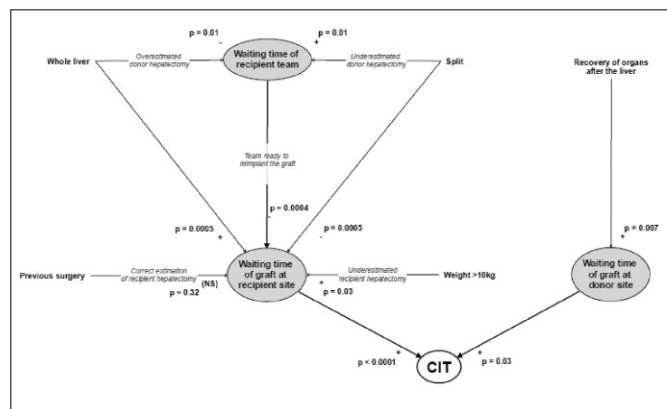
Methods: Patients enrolled in the Swiss National Biliary Atresia Registry who underwent HPE were studied. The following data were analyzed: patient characteristics, perioperative complications and SNL. Perioperative complications were defined as complications arising during HPE and up to 30 days postoperatively, and were stratified in 1) surgical and 2) medical complications. The Spearman rank correlation coefficient was used to determine the association between (i) patient characteristics (gestational age at birth, age and weight at HPE, syndromic BA) and complications, and (ii) complications and SNL. SNL was analyzed using Kaplan-Meier plots.

Results: 87 BA patients underwent HPE in Switzerland between 1994 and 2017. 62 patients were included in the analysis (mean gestational age at birth 38+/-2 weeks, at HPE: mean age 62+/-21 days, mean weight 4504+/-974 g). 26/62 patients (42%) had one or more complications: 19/62 (31%) had suspected or proven cholangitis; 6/62 (10%) had surgical perioperative complications, while 24/62 (39%) had medical perioperative complications. Lower gestational age at birth correlated with the occurrence of more overall complications ($p = 0.02$). Age and weight at HPE were not significantly correlated ($p = 0.77$ and $p = 0.98$, respectively), as was the syndromic form of BA ($p = 0.41$). There was also no correlation between overall complications and SNL ($p = 0.55$). There was no significant difference in SNL between patients with and without perioperative complications.

NaCHwuchs Prize: I wish to apply

I have read and understand application rules: No

Disclosure of Interest: None declared



Conclusion: CIT depends on waiting times during the entire recovery and transplantation process. These waiting times, especially of the recipient team or the graft at the recipient site, largely depend on the estimation of the lengths of hepatectomies in donors and recipients. An accurate estimation of length of hepatectomy – considering graft type, recipient's previous surgery, and recovery of the other organs after the liver by a different surgical team - allows a decrease in CIT and, consequently may improve the short- and long-term outcomes of patients after pediatric LT.

NaCHwuchs Prize: I wish to apply

Disclosure of Interest: None declared

006

IMPACT OF PSEUDOMONAS AERUGINOSA AND RESISTANT ESCHERICHIA COLI ON THE RATE OF INFECTIOUS COMPLICATIONS IN PEDIATRIC COMPLICATED APPENDICITIS

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Introduction: Antibiotic choice for complicated appendicitis should address local antibiotic resistance patterns. As our local data showed less than 15% resistance of *Escherichia coli* (*E. coli*) to co-amoxicillin, we opted for this antibiotic in 2013. The increasing recovery of *Pseudomonas aeruginosa* (*Pa*) thereafter challenged this choice. We aim to describe the bacteriology of peritoneal swabs of complicated appendicitis in our pediatric patients and determine the risk of infectious complications.

Methods: We designed a retrospective cohort study. All children (<18 years old) having surgery for a complicated appendicitis between January 1, 2010 and December 31, 2016 were available for inclusion if they had a peritoneal swab culture. Microbiological results are presented descriptively and we performed univariate analyses for potential determinants of infectious complications. All variables with a P-value <0.05 were then included in a multivariate logistic regression model, for which adjusted odds ratio (OR) and 95% confidence interval (CI) were calculated.

Results: One hundred and thirty-three patients were treated for a perforated appendicitis and had cultures of peritoneal fluid. Median age was 9.5 years (IQR 5.7–12.4) and there were 53 girls (40%). *Escherichia coli* was isolated in 94 patients (71%) and it was resistant to co-amoxicillin in 14% of cases. *Pa* was found in 31 patients (23%). In a multivariate analysis, only resistant *E. coli* significantly predicted infectious complications (OR 4.7, CI 95% 1.4–16.6, $p = 0.015$). However, in patients infected with *Pa*, the absence of adequate antibiotic coverage increased the risk of wound or intra-abdominal abscess.

Conclusion: We confirm the increased risk of postoperative complications in the presence of resistant *E. coli*. Adaptation of antibiotic choice should be done accordingly. More data are needed to justify the systematic coverage of *Pa* in children with ruptured appendicitis.

NaCHwuchs Prize: I wish to apply

Disclosure of Interest: None declared

005

LOGISTIC COORDINATION IN PEDIATRIC LIVER TRANSPLANTATION: CRITERIA FOR OPTIMIZATION

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Introduction: Logistic coordination of the recovery and transplantation process aims to reduce to a minimum the graft's cold ischemia time (CIT) to avoid deleterious short- and long-term effects for recipients. To date, studies of causality between the coordination process and CIT are scarce. This work aims to analyze factors of the logistic coordination process influencing CIT in pediatric liver transplantation (LT) and to evaluate the efficacy of this process in our center.

Methods: We conducted a retrospective analysis of pediatric LT (0–18 years) realized between 01/2006 and 12/2015 in the Swiss Center for Liver Diseases in Children in Geneva. A student t-test or the Chi square test was used in the descriptive analysis to compare categorized waiting times, length of surgeries and CIT according to the explanatory variables. Univariate and multivariate linear and logistic regressions were applied for the association between these variables to determine the most relevant factors.

Results: The analysis included 61 patients; graft types were 24 whole LT (liver transplants) and 37 split LT. Length of hepatectomies in donors was significantly increased for split LT ($p < 0.0001$), and length of hepatectomies in recipients was longer in case of previous surgery ($p = 0.06$). In our center, the recipient team experienced a longer inactive waiting time for the organ when the graft was a split ($p = 0.01$). The graft waited longer at the recipient site in case of i) whole LT ($p = 0.0005$), and ii) increased recipient weight ($p = 0.03$). The waiting time of the graft at the donor site was doubled in case of recovery of solid organs after the liver, done by the same surgical team ($p = 0.007$). CIT was significantly increased by the waiting time of the graft i) at donor site ($p = 0.007$), and ii) at recipient site ($p < 0.0001$). The length of surgery in the recipient is essentially increased by the graft type ($p = 0.01$).

007

VENTRICULOPERITONEAL SHUNT VERSUS ENDOSCOPIC THIRD VENTRICULOSTOMY IN CHILDREN LESS THAN ONE YEAR: A SINGLE CENTER REPORT

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Introduction: Endoscopic third ventriculostomy (ETV) is established as a treatment option in children with hydrocephalus. Age and etiology are considered to be main factors influencing success of ETV in children. In literature ETV seems to be associated with less revisions compared to VP-shunting. However the success rate in children less than 1 year of age is worse than in older children, as they are considered to be high risk patients.

Methods: In a matched-pair analysis we retrospectively compared two groups children (n = 20/group). Children <1 year of age to a maximum of 11 kg undergoing primary ETV or VP-shunting were included in the study. In all patients etiology was a different type of obstructive hydrocephalus. In VP-shunted patients a programmable (Medos Hakim®) shunt valve was implanted. During ETV procedure a Rickham Reservoir was applied for possible emergency drainage. Primary endpoints were time to failure (1), number of revisions (2) and patency (3) within a period of 18 months of observation. Failure was defined as the need for shunt revision, placement of a new shunt or re-ventriculostomy.

Results: In our patients obstructive etiologies most frequent were aqueductal stenosis following intraventricular hemorrhage (IVH) or due to Chiari malformations. The percentage of preterm babies was 35%. Mean age of VP-shunted children was 66 days, 9/20 (45%) children did not require a second procedure within time of observation. In those patients requiring secondary surgery mean time to revision was 99 days. Children after primary VP-shunting undergoing secondary surgery obtained 2.5 revisions in average. In the group of children treated primarily with ETV mean age was 92 days. 7/20 (35%) children did not require a second surgical intervention. For patients with secondary surgery time to revision was slightly shorter compared to VP-shunting with being 83 days in average, while the average number of revisions was 2.

Conclusion: In our patient population ETV and VP-shunting showed similar results after 18 months, however 35% of children after ETV could remain free of a shunt valve.

NaCHwuchs Prize: I wish to apply
Disclosure of Interest: None declared

008

FINGER AND HAND INJURIES IN A PEDIATRIC EMERGENCY DEPARTMENT – A 17-MONTH REVIEW FROM A SWISS TERTIARY HOSPITAL

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Introduction: Hand and finger injuries are common in the pediatric emergency department (PED). Demographics and injury patterns have been described previously but scarce data exist on treatment type and outcome. In our institution, most fractures and extensor tendon injuries are treated by pediatric surgery and more complex injuries including palm of the hand lesions are under hand surgical care. We aimed to evaluate differences in outcome depending on the treatment provided by different subspecialties.

Methods: All children presenting to the PED with hand/finger injuries or infections from November 2014 – March 2016 were included in this retrospective study. Polytrauma, injuries proximal to the distal radius and systemic infections were excluded. Demographics (age, sex), date/type of injury, treatment modality in the PED, subspecialty referral and subsequent treatment details were retrieved from the electronic hospital database. For patients admitted to the ward, length of hospital stay, definitive treatment details including antibiotics were collected and compared between departments. A written feedback form on clinical and functional outcome was developed and collected from the parents.

Results: Over a 17-month period, 932 children with hand/finger injuries were admitted to the PED, most commonly with hand contusion (25.5%), fracture (20.8%) and superficial laceration (14.9%). Admission to the ward occurred in 87 cases (9.3%) following multiple hand/finger injuries (19.4%), infections (18.4%), finger fractures (12.6%) and fingertip amputations (11.5%). In this subgroup 51 children were treated by pediatric (58.6%) and 37 by hand surgeons (41.4%). Mean duration of hospital stay was 3.37 days (range, 1–23 days). Antibiotics were administered to 57 patients (65.5%) with a

mean duration of 6.96 days (range, 1–21 days). The feedback form was returned in 75.8% of cases and reflected a very good overall satisfaction.

Conclusion: Our data are similar to previous studies regarding patient age and sex and type and distribution of injuries in different age groups. This study demonstrates an overall very good parental satisfaction with treatment. A dedicated subspecialty might be desirable for the treatment of pediatric hand injuries. Furthermore, the aptitude and limits of treatment by PEM physicians should be defined. Based on our results, we suggest the implementation of a referral pathway to improve and expedite treatment of more complex injuries.

NaCHwuchs Prize: I wish to apply
Disclosure of Interest: None declared

009

COURSE OF ACUTE NONSPECIFIC MESENTERIC LYMPHADENITIS: SINGLE CENTER EXPERIENCE

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Introduction: Reports dealing with acute nonspecific mesenteric lymphadenitis, a well-recognized and self-limiting cause of abdominal pain in childhood, do not address the total duration of symptoms. However, a time for recovery ≤ 4 weeks is commonly assumed. We aimed at investigating the course of acute nonspecific mesenteric lymphadenitis in childhood.

Methods: A retrospective charts' review was made of the patients aged ≤ 16 years in whom the diagnosis of acute nonspecific mesenteric lymphadenitis was established between 2011 and 2015 at the Pediatric Emergency Unit, Ospedale San Giovanni, Bellinzona. The records of the Pediatric Emergency Unit, those of the referring family doctors and the results of a structured telephone interview with each family were used. The diagnosis was made in children with a) acute-onset abdominal pain; b) an abdominal ultrasound demonstrating in the right lower quadrant small-bowel mesentery or ventral to the psoas a cluster of ≥ 3 enlarged lymph nodes with a short-axis diameter of ≥ 5 mm and of ≥ 8 mm in at least one; and c) no evidence for further possible causes of abdominal pain.

Results: Forty-four patients (25 girls and 19 boys) aged 2.5 to 16, median 8.2, years were included. Abdominal pain was cramping in 22, continuous in 9 and both cramping and continuous in 9 cases. A bimodal distribution in duration of symptoms was observed: symptoms persisted for ≤ 2 weeks in 22 patients and 3 to 10 weeks in 22. Clinical and laboratory characteristics were similar in children with symptoms persisting two weeks or less and in those with symptoms persisting 3-10 weeks.

Conclusion: In pediatric patients affected by acute nonspecific mesenteric lymphadenitis, it is advantageous to think of the time span for recovery in terms of ≥ 4 weeks.

Publication: Benetti C, et al. Course of acute nonspecific mesenteric lymphadenitis: single-center experience. Eur J Pediatr. 2018;177:243–6.

NaCHwuchs Prize: I wish to apply
Disclosure of Interest: None declared

011

ANALYSIS OF BLOOD TRANSFUSION PRACTICES IN NEWBORNS IN SWITZERLAND

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Introduction: Switzerland does not have a nationwide guidelines about blood transfusion practices in newborns. Therefore, the aims of this study were to gather the transfusion practices and the internal recommendations of all Swiss neonatology clinics and laboratories, to compare them with international recommendations and to find out whether there is a need to develop a valid nationwide recommendations.

Methods: All Swiss neonatology clinics level III und II according to the CANU classification and their laboratories were consulted from April to November 2016 by means of two different online surveys concerning their blood transfusion management. Neonatology clinics were kindly asked to submit their internal guidelines of transfusion thresholds. The blood products are provided in Switzerland in close collaboration with the regional blood donor services, so they were also asked to participate.

Results: Out of the 34 total neonatology clinics in Switzerland, 29 carry out blood transfusions in newborns. A total of 26 of these 29 clinics participated in the survey. A total of 26 of these 29 clinics participated in the survey (participation rate 90%). All 12 clinics having internal guidelines submitted these to the comparison. Out of a total of 25 laboratories, 18 took part in the survey (participation rate of 72%). The result of the survey revealed a considerable diversity of different transfusion practices, ranging from the preparation to the administration of blood products (quantity, time, conservation, control policy etc.). The clinic internal blood transfusion thresholds were also completely different. Despite existing recommendations of blood donation SRK Switzerland, most laboratories and blood donor services demonstrated a different management of the blood transfusions provision in newborns.

Conclusion: Regarding blood transfusion strategies, the present study thus clearly shows high diversity in transfusion practice. To our opinion, it would make sense to develop a common recommendation for the handling of blood transfusions in newborns in Switzerland. A nationwide guideline would be a valuable aid for all health professionals in the daily management of blood transfusions and an opportunity to positively influence neonatal care. Furthermore, Switzerland could keep pace with other countries and position itself in an international context.

Disclosure of Interest: None declared

012

HUMAN PAPILLOMAVIRUS (HPV) VACCINE IN SWITZERLAND: THE REASONS WHY SOME YOUNG WOMEN DO NOT GET VACCINATED

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Introduction: To assess the HPV vaccination rate among young women and their reasons for not being vaccinated.

Methods: A total of 2619 females aged 24–27 years taking part in the 2017 Swiss national study on sexual health answered a question on their HPV vaccination status: 1032 (39.4%) were vaccinated (YES group), 1011 (38.6%) were not (NO group) and 576 (22.0%) did not know (DNK group). Participants in the DNK group were not included in the comparisons. YES and NO groups were compared on socio-demographic, family, education, health and sexuality data. Significant ($p < .05$) variables at the bivariate level were included in a logistic regression analysis. Results are given as adjusted odd ratios (aOR) with 95% CI.

Results: At the bivariate level, females in the NO group were significantly more likely to be Swiss-born, to have two Swiss parents, to live in a rural area, to evaluate their family socio-economic status when they were aged 15 as below average and to report no lifetime sexual partner. No significant difference was found for education, perceived puberty onset, STI diagnosis, and average age at first sexual experiences. At the multivariate level, NO participants were more likely to have Swiss parents (aOR: 1.56 [1.27–1.91]) and to report a below average family socio-economic status (1.29 [1.04–1.62]), and less likely to report one (0.56 [0.36–0.87]), two-three (0.49 [0.32–0.75]) and four or more (0.59 [0.39–0.90]) lifetime sexual partners. Reasons given for non-vaccination were: refusal (40.9%), unaware of the vaccination (26.2%), not in the age range for insurance reimbursement (9.5%), discouraged by family or friends (10.7%), already infected with HPV (2.2%) and other reasons (10.5%). Reasons for refusal were: overall opposition to vaccination (37.2%), HPV type-restricted protection (34.8%), afraid of side effects (14.0%) and other reasons (14.0%).

Conclusion: Rates of vaccinated and non-vaccinated women are almost identical. The first reason for not being vaccinated against HPV is the refusal to be vaccinated, mainly reported as an overall opposition to vaccination. The current trend to be opposed to vaccines should not prevent the clinical discussion on HPV. Discussion should also be conducted with females without sexual activity, as the vaccine is more effective before sexual debut. Finally, the non-negligible rate of females who do not know if they were vaccinated (one in 5) should be considered to ensure that enough information is given at the time of administration.

Disclosure of Interest: None declared

DISEASE EVOLUTION IN SYSTEMIC-ONSET JUVENILE IDIOPATHIC ARTHRITIS: PRELIMINARY DATA FROM JIRCOHORTE

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Introduction: Systemic onset juvenile idiopathic arthritis (SoJIA) is a potentially severe disease with both systemic and joint inflammation; different evolutive forms were described (monophasic, polyphasic or persistent), but the outcome is hardly predictable at diagnosis. This study aims to identify early predictors of disease evolution within the SoJIA population enrolled in the Juvenile Inflammatory Rheumatism cohort (JIRcohorste), an international prospective cohort study.

Methods: 152 SoJIA patients with a minimum of two-year follow-up were enrolled. 5 patients were excluded due to missing data on diagnostic visit. Demographics and clinical data were collected (retrospectively if diagnosis < 2015 and prospectively if diagnosis ≥ 2015) and described for 147 patients. At diagnosis, median age and disease duration was 5.3 years and 1.9 months, respectively, male to female ratio was 1:1.2. We present the preliminary results in 30 patients with complete data for disease activity and medication use.

Results: Corresponding to their evolution, patients were classified in the monophasic ($n = 10$; MONO), polyphasic ($n = 14$; POLY) or persistent group ($n = 6$; PER). Females were predominant in the MONO and POLY group with 80% and 79%, respectively. No female patient was in the PER group. At diagnosis, all patients in the PER and POLY group had arthritis; only 80% did in the MONO group. Hepatomegaly and splenomegaly was present in 30% and 50% of MONO patients versus 17% and 0% of PER patients, respectively. Joint count at 6 months was ≥ 5 joints for 83% of the PER population compared to 30% for the MONO. Four patients in the MONO group did not have any biologics, whereas all patients from the other groups had one or more (POLY: 7 patients had one, 5 patients had two and 2 patients had three different biologics). Disease Modifying Anti-Rheumatic Drugs (DMARDs) were used in 4 patients of the MONO, 13 of the POLY and 5 of the PER group. Median duration of corticoid-use for the MONO group and the POLY group is 0.3 years and 2.68 years, respectively.

Conclusion: Arthritis seems to be more prevalent in the POLY and PER groups whereas systemic presentation in the MONO group. Polyarticular arthritis at 6 months is suggesting persistent disease evolution. Biologics, DMARDs and corticoids are more often and longer used in the POLY and PER groups correlating with a more severe disease course. Data will be completed during 2018 for all 147 patients and deep phenotypic analysis will be performed.

Disclosure of Interest: None declared

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THE SWISS PAEDIATRIC SURVEILLANCE UNIT SPSU CONTRIBUTES TO CHANGE IN PUBLIC HEALTH POLICY AND PRACTICE

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Introduction: The Swiss Paediatric Surveillance Unit SPSU is a research network to study rare paediatric diseases or rare complications of more frequent diseases in hospitalized children. All 33 paediatric clinics of Switzerland are members and contribute to active case finding on roughly 1.3 Mio. children under the age of 16. SPSU has been established by the Swiss Paediatric Association and the Swiss Federal Office of Public Health in 1995 and is member of the International Network of Paediatric Surveillance units INoPSU.

Methods: The SPSU makes available to principal investigators (PI) a nationwide active reporting system, which means that clinics proactively search for matches of patients who meet the inclusion criteria for running studies. Typically, 6-8 studies are run in parallel. While particular emphasis is placed on infectious diseases, studies on other relevant topics are also accepted. Cases are centrally registered at the SPSU administration office, which in turn notifies the PI who then collect more detailed data by a standardized questionnaire for each case child. The return rate is 100% for case notifications and >90% for questionnaires. Data analysis is done by the PI and results are collectively published in an annual report. In addition results are regularly published in peer review journals. The SPSU also contributes to international studies performed within INoPSU who specifically promotes joint studies run in several countries.

Results: Studies performed by SPSU have been useful for i. evaluating public health interventions such as vaccination against Pertussis that lead to an international consensus on booster vaccinations, ii. elucidating knowledge on epidemiological determinants of severe infections such as HUS in patients infected with EHEC iii. evaluating change in screening policies such as Toxoplasmosis screening in pregnant women, which lead to the abandon in several countries and iv. guidance in paediatric clinical practice such as introducing an additional dose of Vitamin K in infants to prevent late bleeding in all countries who opt for oral Vitamin K. **Conclusion:** SPSU is a unique highly reliable research network for the study of rare paediatric conditions. Data generated by SPSU are generalizable to the total of 1.3 Mio children in Switzerland and have proven to impact clinical practice and Public Health policy not only in Switzerland but also worldwide.

I have read and understand application rules: No

Disclosure of Interest: None declared

HIGH VACCINE COVERAGE RATES ARE NOT ENOUGH: VACCINATION DELAY AND RISK FOR VACCINE-PREVENTABLE DISEASES IN PEDIATRIC PATIENTS WITH RHEUMATIC DISEASES WITH AND WITHOUT IMMUNOSUPPRESSIVE THERAPY

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Introduction: Pediatric patients with rheumatic diseases (PedRD) are more susceptible to invasive infectious diseases, due to their underlying disease, high disease activity and immunosuppressive therapy (IT). In Switzerland, specially indicated (SI) vaccinations for PedRD patients against pneumococcus, varicella zoster, hepatitis B, influenza and human papilloma virus should be administered as early as possible. On-time and complete coverage reduces the risk of vaccine-preventable infections and its associated morbidity. Information about on-time vaccination is scarce, especially in PedRD patients. Therefore this study assess relationship of vaccine coverage and on-time vaccination in PedRD patients in Switzerland with and without IT.

Methods: Multicenter retrospective prevalence study based on the Juvenile Inflammatory Rheumatism (JIR) cohort, an international registry for rheumatic diseases. Patients treated in Swiss pediatric rheumatology centers (Basel, Geneva, Lausanne, Lucerne, Zurich) were included. Vaccine coverage and timeliness of administered vaccines were assessed for routine and for SI vaccinations according to the Swiss Federal Office of Public Health.

Results: 239 patients were included. 93 (39%) received IT during a median observation period of 2.7 years (IQR 1.1–5.4). Median age was 6.7 years (IQR 2.9–10.3) at diagnosis, 10.9 years (IQR 7–14.3) at reference date (12th April 2016). At diagnosis, 185 patients (77%) had complete routine vaccine coverage; 76 (32%) fulfilled Swiss on-time

immunization criteria. At reference date, the routine vaccine coverage rate decreased to 74% with on-time vaccination achieved in 29% of PedRD patients without IT and in 23% with IT, respectively. Completeness of SI vaccinations was documented in 19 PedRD patients (13%) without IT and 6 patients (6.5%) with IT. On-time vaccination for SI vaccination was fulfilled in 7 patients without IT; no patient with IS was vaccinated on-time. Only 23 (9.6%) patients showed complete and thereof 4 (1.6%) on-time vaccination status for routine and SI vaccinations.

Conclusion: Despite moderate to high routine vaccination coverage in Switzerland, vaccinations particularly recommended for PedRD patients to prevent from complications, are incompletely administered. Moreover, the high frequency of delayed vaccination implicates a potential increase of vaccine-preventable disease. Close surveillance of vaccination status improves standard of care for this susceptible population.

NaChwuchs Prize: I wish to apply

Disclosure of Interest: T. Welzel: None declared, T. Zumbunn: None declared, J. Bonhoeffer: None declared, E. Cannizzaro Schneider: None declared, M. Hofer Shareholder of: AbbVie, Novartis, Consultant for: Novartis, D. Kaiser: None declared, V. Hentgen: None declared, A. Woerner: None declared

016

ECHINACEA AND VITAMIN C FOR THE PREVENTION OF RESPIRATORY TRACT INFECTIONS IN CHILDREN 4 – 12 YEARS: A RANDOMIZED, BLIND AND CONTROLLED CLINICAL STUDY

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Introduction: Viral respiratory tract infections (RTIs) including colds and flu are the most frequent illnesses worldwide. *Pneumonia, sinusitis or otitis media* may result and are the primary reason for antibiotic prescriptions. Children are especially susceptible to infections and every third develops RTI complications triggering a tremendous need for antibiotics [1].

Methods: This study investigated *Echinacea* for the prevention of RTIs and cold days in children aged 4 to 12 years. An alcoholic extract of *Echinacea purpurea* (Echinaforce® Junior Tablets [EFJ], 1200 mg) or 150 mg vitamin C [VC] were applied under randomized and blind conditions. 2 × 2 months of prevention were separated by a 1-week treatment break and three study visits were carried out. Parents recorded respiratory symptoms using a validated scoring method (e-diary). At acute RTI, they took a sample of nasal secretion for microbial detection (Allplex RT-PCR).

Results: Overall, 429 cold days occurred in children with *Echinacea* (N = 103) in comparison to 602 days in children with vitamin C (N = 98; p < 0.001). *Echinacea* significantly prevented cold episodes with an odds ratio of OR = 0.51 [95% CI, 0.31–0.84] and p = 0.008. Four children (3.9%) with *Echinacea* required antibiotic treatment in comparison to 11 children with vitamin C (11.2%), yielding a statistically significant reduction of 65.2% (p < 0.05). A total of 31 and 111 days with antibiotics were registered, respectively (p < 0.0001). The reduction in antibiotics was associated with a marked prevention of bacterial superinfections (e.g. *angina tonsillaris*) and RTI complications (*pneumonia* or *otitis media*), where 9.7% and 20.4% of children experienced 10 and 20 events with EFJ and VC, respectively (p < 0.05). A significant reduction of Influenza (3 vs. 20 detections, p < 0.05) and membranous virus infections (28 vs 46 detections, p < 0.05) were found with *Echinacea*. Finally, 89.8% of parents noticed a (significantly) improved resistance status in their children after 4 months EFJ (p < 0.05). Over 4.1 months prevention 36.9% of children with EFJ experienced adverse events and 41.8% with VC. Three events were serious but not medically related to the treatment. No statistically significant or medically relevant difference was noticed between EFJ and VC.

Conclusion: The results provide evidence for the beneficial use of Echinaforce® Junior tablets in preventing RTI's (e.g. Influenza) and RTI complications, implicating a reduced need of antibiotics in paediatrics.

1 Taylor JA, et al. JAMA. 2003;290:2824–30.

Disclosure of Interest: M. Ogal: None declared, P. Klein: None declared, A. Suter Employee of: Bioforce AG, R. Schoop Employee of: Bioforce AG

017

THE IMPORTANCE OF WHOLE EXOME SEQUENCING AS A DIAGNOSTIC TOOL IN THE WORK UP OF NEURO-DEVELOPMENTAL DISORDERS IN A PEDIATRIC COHORT

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Introduction: In 2010, Whole Exome Sequencing (WES) has shown the most frequent genetic mechanism underlying intellectual disability is that of *de novo* dominant mutations. Thus, WES has rapidly been integrated in the diagnostic workup of intellectual disability and developmental delay, as well as of related conditions ranging from disorders of the autistic spectrum to more severe encephalopathies with or without epilepsy. Variants detected in a typical WES assay require careful evaluation and correlation with clinical findings in order to determine their pathogenicity.

Methods: We retrospectively studied a cohort of 64 pediatric patients, aged 0 to 17 years, who presented with a neuro-developmental disorder from January 2016 to October 2017. After appropriate genetic counselling and informed parental consent, a panel of approximately 1400 genes known to be involved in developmental delay, intellectual disability, autism and epilepsy ("TRDEV panel") was studied with WES. The variants found were first filtered according to epidemiological and biological criteria (frequency, type of mutation and expected effects), then subsequently discussed with physicians and correlated with clinical findings and literature data. When a mutation was considered as possibly pathogenic, it was verified by Sanger sequencing on DNA of the proband and, when available, of the parents to assess genetic segregation (bi-parental transmission or *de novo*).

Results: In the 28 patients for whom the WES analysis was completed, we identified likely pathogenic variants in 14 of them. In the remaining 36 patients with pending analysis, Sanger sequencing is currently ongoing in 12 of them, mainly to confirm potential pathogenic variants. Analysis is still ongoing in the last 24 patients. In this cohort, presence of epilepsy, abnormal MRI findings and microcephaly were correlated with a positive molecular finding.

Conclusion: The identification of pathogenic variants in pediatric patients with a neurodevelopmental phenotype in this cohort highlights the power of WES as a diagnostic instrument and the significant advance compared to earlier diagnostic methods. We underline the importance of a close collaboration between the laboratory, the expert geneticists and pediatric neurologists in the prioritization and interpretation of the mutations identified.

Disclosure of Interest: None declared

018

CORD BLOOD CYSTATIN C IS A PREDICTIVE MARKER FOR RENAL FUNCTION AT ONE YEAR IN NEONATES PRENATALLY DIAGNOSED WITH CONGENITAL KIDNEY ANOMALIES

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Introduction: Congenital anomalies of the kidney and urinary tract (CAKUT) occur at a frequency of 1 for 500 pregnancies and are the main cause leading to end-stage renal disease and transplantation during childhood. Cord blood Cystatin C (CysC) is a useful marker to evaluate kidney function at birth and reflects the severity of the kidney anomaly. The aims of the study were to evaluate if cord blood Cys C may predict renal function (RF) at one year and to identify determinants for decrease in renal function during this first year of life.

Methods: This single-centre, cohort study included all neonates prenatally diagnosed with CAKUT from January 2006 to December 2016 at the Geneva University. All neonates had cord blood Cystatin C drawn at birth (reference interval 1.5–2.6 mg/L). Among those, 131 had repeated measures of CysC during the first year and at one year. A Kaplan-Meier survival and log-rank test analysis was used to measure CysC normalisation at 1 year (reference interval 0.65–1.24 mg/L).

A multivariate analysis was used to identify risk factors for RF progression during the first year. Following factors were considered: gestational age, weight, size, gender, CAKUT diagnosis subgroups, bilateralism of the kidney anomaly, reflux, renal scars, intervention, urinary tract infection (UTI).

Results: Of the 131 children included, 69.5% were males. Mean CysC at cord blood was 2.0 mg/L (1.18–4.23 mg/L). Bilateral kidney anomaly was present in 44% neonates, 80% had pelvic dilatation. A cystography was done in 85% of the neonates, resulting in 21% vesico-ureteral reflux (VUR) and 12% bladder anomalies. Among the neonates who had a scintigraphy (56.5%), 50% presented a RF asymmetry and 24% renal scars. 27.5% patients had a UTI and 30% needed surgery. At one year, the percentage of patients with normalized CysC was 72.5% (95% CI 57.5 to 82.2). In the multivariate model, CAKUT subgroup consisting in bladder anomalies, kidney agenesis, ectopic or dysplastic ($p < 0.001$), the presence of reflux ($p = 0.003$) and increased cord blood CysC ($p = 0.004$) were significant risk factors to present an abnormal RF (non normalized Cystatin C value) at one year, while renal scars, intervention and UTI were not significant.

Conclusion: This study showed that Cord blood Cys C is a valuable marker in identifying neonates with a worse RF prognosis at one year. Determinant for abnormal RF at one year were the presence of VUR and CAKUT diagnosis.

Disclosure of Interest: None declared

019

DOES SERUM NEUTROPHIL GELATINASE-ASSOCIATED LIPOCALIN, BRAIN NATRIURETIC PEPTIDE, PARATHYROID HORMONE OR ALBUMIN IMPROVE THE PERFORMANCE OF THE PEDIATRIC GLOMERULAR FILTRATION RATE ESTIMATING FORMULAS?

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Introduction: Estimation of glomerular filtration rate (eGFR) is important in clinical practice. Most eGFR formulas used in children include serum creatinine (Cr) and/or cystatin C (CysC). This study aims to assess if the inclusion of additional biomarkers will improve the performance of eGFR formulas.

Methods: 116 children between 3 and 18 years of age with renal diseases were enrolled. Data for age, weight, height, inulin clearance (iGFR), CysC, Cr, Neutrophil gelatinase-associated lipocalin (NGAL), parathormone (PTH), and brain natriuretic peptide (BNP), were prospectively collected. These variables were added to the following formulas: revised Schwartz formula, combined Schwartz formula, Quadratic formula and combined Quadratic formula. We calculated the adjusted r-square (r^2) and tested the improvement in variance explained by means of the likelihood ratio test. We attempted to construct a parsimonious multivariate model predicting inulin clearance including all statistically significant predictors. The results are compared to the iGFR, which is the gold standard method for GFR measuring.

Results: The r^2 of the Schwartz, the combined Schwartz, the Quadratic, and the combined Quadratic formulas, for iGFR were 0.57, 0.67, 0.71 and 0.73, respectively. After including NGAL, the r^2 of the Schwartz, the combined Schwartz, the Quadratic, and the combined Quadratic formulas, for iGFR were 0.57, 0.67, 0.71 and 0.73, respectively, and did not improve significantly the formulas performance. Addition of BNP leads to better fit with an r^2 of the Schwartz, the combined Schwartz, the Quadratic, and the combined Quadratic formulas, for iGFR at 0.62, 0.69, 0.73, and 0.74, respectively. Neither PTH nor albumin added to these formulas did improve the prediction of GFR further.

Conclusion: The combined Quadratic formula has the best performance for estimating GFR. The addition of NGAL, BNP, PTH and albumin did not lead to a further improved prediction.

Disclosure of Interest: None declared

SNORING IN SWISS SCHOOLCHILDREN: PREVALENCE AND RISK FACTORS

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Introduction: Habitual snoring can lead to poor health and unfavorable cognitive and behavioral outcomes, but population based data on prevalence and risk factors are scarce. We aimed to describe the prevalence of self-reported snoring in a representative population-based sample of school-aged children, and to determine risk factors for habitual snoring.

Methods: As part of the Luftibus field study of respiratory symptoms and lung function, we obtained questionnaires from parents of 1353 children aged 6 to 16 years from a representative sample of schools from the canton of Zurich, Switzerland. We investigated associations between habitual snoring (snoring almost every night) and potential risk factors (age, sex, rhinitis, hay fever, body mass index (BMI), low maternal education (only elementary school), parental smoking, and adenotonsillectomy using logistic regression.

Results: 41% (551/1353) of children were reported to snore: 17% (233) snored only with colds, 19% (268) also without a cold and 5% (68) snored habitually. Five percent had undergone adenotonsillectomy. The prevalence of habitual snoring did not vary between boys and girls, nor by age. Six percent (82) reported disturbed sleep because of snoring. Among all children, 30% (402) had chronic rhinitis, 23% (311) hay fever, 29% (399) were exposed to parental smoking and 10% (130) of the mothers had a low education level. Habitual snoring was more common in children who had undergone an adenotonsillectomy (odds ratio 3.1, 95%CI 1.5–6.6, $p = 0.003$), but we found no relationship with age, sex, BMI, rhinitis, hay fever, low maternal education or parental smoking.

Conclusion: Habitual snoring is reported for 5% of schoolchildren. Risk factors, treatment options and long-term health outcomes will merit further investigations.

Funding: Lunge Zurich, Switzerland; SNF32003B_162820

Disclosure of Interest: None declared

O20

Conclusion: Reported symptoms like "Exercise induced wheeze," and "wheeze triggered by allergens" were important predictors of wheeze persistence into adolescence. None of the clinical measurements including BPT did improve prediction based on reported symptoms.

Funding: SNF32003B_162820

Reference: 1. Barben, et al. *Pediatr Pulmonol* 2011;46:842–8.

Disclosure of Interest: None declared

O22

EARLY POSTNATAL OVERFEEDING INDUCES TRANSIENT OVEREXPRESSION OF SENEESCENCE PROMOTING FACTORS IN THE MOUSE KIDNEY

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Introduction: Transient postnatal overfeeding is known to induce adverse renal effects in adulthood, as it is associated with arterial hypertension, glomerulosclerosis, proteinuria, and chronic kidney failure. However, the underlying molecular mechanisms are poorly understood. We aimed to study the intrarenal molecular mechanisms triggered by transient postnatal overfeeding.

Methods: We used a rodent animal model of postnatal overfeeding during the first 3 weeks of life by litter size reduction. Smaller litters result in overfeeding during the suckling period, as a smaller number of pups compete for mother's milk. Pups were sacrificed at 3 weeks (at the end of suckling period) and 7 months of life (mid adulthood). Inflammasome (NLRP3 and Il1), mTOR (P-AKT/AKT, P-S6K/S6K) and senescence (P16, P21, P53, P-Rb/Rb) activation pathways in the kidney were analysed by western blot.

Results: Transient postnatal overfeeding was significantly associated with an overexpression of senescence promoting factors P53 ($P = 0.001$), and P21 ($P = 0.005$), with a significant decrease in P-Rb/Rb ratio ($P = 0.02$) compared to control group at 3 weeks of age. Our results also showed a non-significant overexpression of P16 ($P = 0.07$) in overfed mice compared to control group at 3 weeks of age. At 7 months of life, expression of these factors was similar between early overfed and control animals. In addition, expression of the inflammasome pathway (NLRP3 and Il1) and of the mTOR pathway (P-AKT/AKT and P-S6K/S6K) were unchanged at 3 weeks and 7 months of life between early overfed and control animals.

Conclusion: We found that transient postnatal overfeeding was associated with an elevation of intrarenal senescence promoting factors at 3 weeks of age, which normalized in adulthood. However, early overexpression of these factors could contribute to worsened adult renal outcome, as they may induce irreversible renal damage, which decreases the renal reserve and contributes to further renal injury, although the primary pathological stimulus and the associated pathway activation are discontinued. In conclusion, early intrarenal activation of senescence pathway may contribute to CKD programming after transient postnatal overfeeding. Inflammasome and mTOR pathways did not seem to be involved in programming in this setting.

Disclosure of Interest: None declared

O23

SEQUENTIAL ANALYSIS IN NEONATAL RESEARCH – SYSTEMATIC REVIEW

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Introduction: As more new drugs are discovered, traditional trial designs come at their limits.

Methods: Ten years after the adoption of the European Paediatric Regulation, we performed a systematic review on the US National Library of Medicine and Excerpta Medica databases of sequential trials involving newborns.

Results: Out of 326 identified original articles, 21 trials were included. They enrolled 2832 patients, of whom 2099 were included in sequential data-analysis. They included a median of 48 (IQR 22–87) neonates per trial, whose median gestational age was 28.7 (IQR 27.9–30.9) weeks. 18 trials applied sequential methods to calculate

EXERCISE-INDUCED SYMPTOMS PREDICT PERSISTENCE OF WHEEZE INTO ADOLESCENCE IN A CLINICAL POPULATION

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Introduction: It is unclear if a detailed clinical investigation including bronchial provocation tests (BPT) helps to predict long-term outcome of schoolchildren investigated for asthma.

Aim: We determined which symptoms and clinical tests predict long-term prognosis in children seen in paediatric outpatient clinics for possible asthma.

Methods: We studied a random sample of children aged 6–16 years, referred to 2 Swiss pulmonary outpatient clinics with a history of wheeze, dyspnoea, or cough in 2007. The initial assessment included lung function testing, BPT by exercise, methacholine and mannitol, and skin-prick tests [1]. Respiratory symptoms were assessed with parental questionnaires at baseline and at follow-up 7 years later. Associations between baseline factors and wheeze at follow-up were estimated by logistic regression.

Results: 85 (77%) of 111 children completed both questionnaires, of whom 61 (72%) wheezed at baseline and 39 (46%) at follow up. After adjusting for sex and age, 5 factors predicted wheeze at adolescence. Among reported symptoms, these were hay fever (OR [95% CI] 2.5 [1.0–6.3]), wheeze triggered by pets (4.3 [1.2–15.5]), by pollen (3.9 [1.4–10.7]), and by exercise (3.1 [1.2–8.0]). Among clinical tests only a positive exercise test (3.5 [1.2–10.0]) predicted wheeze persisting into adolescence. However, this association was no longer significant when we adjusted for reported exercise-induced wheeze at baseline.

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sample size, while 3 used continual reassessment methods for dose-finding studies. In 16 studies reporting sufficient data, the sequential method allowed to non-significantly decrease the number of enrolled neonates by a median of 24 (31%) patients (IQR –4.75 to 136.5, $p = 0.067$) when compared to a traditional trial. When the number of neonates included in the analysis process was considered, the difference became significant: 35 (57%) patients (IQR 10 to 136.5, $p = 0.0033$).

Conclusion: Sequential trial designs have not been frequently used in newborn trials. They might potentially reduce the number of patients in drug trials, but this is not always the case.

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Disclosure of Interest: None declared

ORAL PRESENTATIONS SSP 1

024

A NOVEL DIGITAL HEALTH INTERVENTION IMPROVES PHYSICAL PERFORMANCE IN OBESE YOUTH

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Introduction: One in five adolescents in Switzerland is overweight or obese. Effectiveness of usual obesity care is limited due to low resources and poor adherence in this age group. Therefore, digital health interventions are promising, but achievement of long term health goals remains mostly unproven. The purpose of our study is to test a novel concept of obesity therapy for adolescents including a health application game.

Methods: A mobile chat App with game character was designed for smartphones, including two channels: one text-based healthcare chatbot (THCB) with pre-defined answer options and one for direct communication with health professionals (HP). A dashboard overview alerts HPs, when no activity is measured for >2 days. During the intensive phase of 24 weeks, the THCB coach encourages patients to achieve daily challenges like steps per day and relaxing breathing exercises in order to earn virtual rewards. The intervention group (IG) had four visits and treatment-as-usual group (C) had monthly visits on site over the first 6 months. Beside BMI, clinical and stress parameter, quality of life, mental health and physical performance (modified Eurofit-Test) are being assessed at start and after 6 and 12 months. Thirty one adolescents are participating in this ongoing 12-month randomized controlled study. We report first data on physical performance in 14 IG patients and 7 C patients after 6 months (T1). **Results:** Age (13.9 years, 11.6–19), mean BMI ($30 \pm 3.7 \text{ kg/m}^2$) and physical performance were similar in both groups at start. After 6 months almost 70% of IG patients continued using the App daily. Although BMI adjusted for age and sex did not change considerably at T1, physical performance improved significantly in 5 of 8 tests in the IG (agility, flexibility, strength) and only in 2 tests in C patients (agility, endurance).

Conclusion: Motivation to increase activity is enhanced by the daily encouragement through a virtual coach, the excitement built up by a persuasive game and the competition with peers. In contrast to high dropout rates usually observed in standard obesity care as well as in usage of health Apps, adolescents in the present setting showed a high compliance with gamified health issues and were able to improve their physical performance after 6 months.

Disclosure of Interest: None declared

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TEEN VIOLENCE: WHAT ABOUT GIRLS?

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Introduction: To determine the prevalence rate of violence among females and their characteristics.

Methods: Data were drawn from the baseline (2015) wave of the GenerationFRee study, a cohort study among 5179 students (47.1% females) aged 15–24 years in post-mandatory education in the canton

of Fribourg. A violence scale (VS) was created based on the performance of eight violent behaviors (physical harm towards an adult, carrying a weapon, using a weapon during a fight, snatching, vandalism, theft, dealing drugs, setting fire to something) in the last 12 months. Based on the VS, females were divided into 3 groups: no violent behavior (VS0; N = 2075; 85.1%), one behavior (VS1; N = 282; 11.6%); two or more behaviors (VS2; N = 82; 3.3%). Groups were compared on sociodemographic, family and academic data, emotional well-being, perceived puberty onset and substance use. Significant variables at the bivariate level were included in a multinomial regression controlling for age. Results are given as relative risk ratios (RRR) and 95%CI.

Results: Among violent females (N = 364; 14.9%) the most reported behavior was theft (58.3%: 51.8% among VS1 and 80.5% among VS2). At the bivariate level, compared to VS0, VS1 and VS2 were more likely to live outside the parents' home, to have non-Swiss born parents, to report being below average students, to report poor emotional well-being, to assess their socio-economic status as below average, to use substances and to perceive an out of the norm puberty timing. At the multivariate level, compared to VS0, VS1 and VS2 were more likely to live outside the parents' home (2.53 [1.44–4.43]; 2.74 [1.23–6.07]), to report being below average students (2.44 [1.37–4.36]; 2.88 [1.20–6.91]), to use cannabis (2.24 [1.55–3.23]; 6.27 [3.71–10.59]) and other drugs (2.29 [1.05–5.03]; 2.88 [1.23–6.77]). VS1 were more likely to perceive a delayed (1.73 [1.06–2.82]) or advanced (1.47 [1.05–2.07]) puberty onset and VS2 were more likely to misuse alcohol (2.58 [1.49–4.48]).

Conclusion: Although young men are overrepresented in statistics and public opinion and their behaviours are more visible, one in seven females in this study reported at least one violent behaviour in the last 12 months. According to the associations that we found, violent females are part of a vulnerable environment in terms of education, housing, substance use and emotional well-being. Health professionals dealing with young females should also screen and discuss violence with them.

Disclosure of Interest: None declared

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WHAT DO SCHOOL DOCTORS IN THE CANTON DE VAUD DO?

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Introduction: Issues related to pupil health, useful to address in school, have changed in nature in recent decades. Actually the school doctor collaborates with the school nurse in the assessment of the particular situations of the pupils in collaboration with the teachers, the family doctor and the other care providers, contributes to school vaccination and epidemic control measures, ensure that the school has the capacity to respond to medical emergencies, participate in prevention and health promotion activities and exceptionally performs a medical examination.

Methods: Since the school year 2015–2016, the Service de la santé publique du canton de Vaud has asked the Unité de promotion de la santé et de prévention en milieu scolaire (Unité PSPS) for a report on the activity of school doctors (except Lausanne and specialized institutions). For the analysis, we used the available timesheets for the year 2015–2016.

Results: The timesheets of 28 (35%) mandatory school doctors and 13 (85%) post-mandatory school doctors were analyzed. These 28 mandatory school doctors working in 26 schools (29'860 pupils) completed 1163.15 hours of activity. While their 13 postsecondary

colleagues working in 29 schools (24'979 pupils) completed 691.8 hours. The main activities are: – in the mandatory schools, the response to the special needs of pupils (30.8%), vaccination (20.7%) and community health projects (18.7%); – in the post-mandatory schools, the response to the special needs of pupils (39.1%), community health projects (27.3%) and support to institutions in response to medical emergencies and / or crisis situations. The pupils who benefited of the involvement of the school doctor had mainly social and/or family problems, psychological problems and/or learning disabilities. The school doctors evaluated the situations, identified solutions with the school nurses and other school staff, and liaised with health and social care professionals outside of schools.

Conclusion: Despite the removal of routine medical examinations, the school doctor continues to play an important role in schools. Psychosocial issues and learning disorders have replaced screening for somatic or communicable diseases. In addition, he actively contributes to the implementation of community health projects. Issues related to pupils' health, useful to address in school, have changed in nature in recent decades. School medicine should no longer focus on routine medical examinations, but must take into account the current health needs of pupils.

Disclosure of Interest: None declared

O27

TELEMONITORING VIA A MOBILE DEVICE APP REDUCES HBA1C IN TYPE 1 DIABETIC CHILDREN, WITHOUT INCREASING THE PREVALENCE OF HYPOGLYCEMIA

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Introduction: Prevention of type 1 diabetes (T1DM)-related complications is dependent on metabolic control of the disease. HbA1c values <7.5% (58.5 mmol/mol) are recommended in children. Unfortunately, these goals are met only by a minority of children and adolescents. *Webdia* is a patient-designed diabetes self-management app that was developed by the father of a diabetic child, in collaboration with our pediatric diabetology team. It helps children with T1DM to calculate insulin doses and evaluate the carbohydrate content of meals. In addition, all glucose values entered on the app become instantly available to the patient, his parents and the diabetology team.

Methods: 55 patients aged 10–18 years were included into this single center, randomized, double crossover study that took place in our unit. The intervention consisted of using *Webdia* during 3 months and to get a monthly feedback and adaptation of the treatment by the diabetology team. Main outcome was modification of HbA1c.

Secondary outcomes were the prevalence of patient-reported hypoglycemia and quality of life (QoL), as evaluated by the Diabetes Quality of Life for Youths questionnaire. Results were tested using 2-tailed paired samples T-tests.

Results: Of the 55 included patients, 33 completed the study, 9 dropped out and 13 were excluded due to insufficient use of the app (<4 times / week). Risk factors for poor use of the application were older age (mean 15.1 ± 2.4 vs. 13.3 ± 2.3 years in patients who completed the study, p = 0.024) and longer duration of T1DM (mean 86 ± 52 vs. 52 ± 35 months, p = 0.014). The program was well accepted by the users (46.4% rated the program as good and 39.3% as excellent). Three-months use of *Webdia*, combined with monthly feedback by the diabetes team lead to a reduction of HbA1c by 0.54%, as compared to the control group (p = 0.048) in patient with HbA1c values >8.0% (63.9 mmol/mol) at inclusion, without increasing the prevalence of hypoglycemia (8.52 ± 9.45 hypoglycemia during last two weeks of intervention, vs. 7.62 ± 6.37 during last two weeks of observation, p = 0.680). QoL scores were not modified by *Webdia* use.

Conclusion: Three months use of *Webdia*, resulted in a significant decrease in HbA1c, without increasing the prevalence of hypoglycemia in patients with initial HbA1c >8.0% (63.9 mmol/mol). Insufficient use of *Webdia* by adolescents points towards a difficult age group and suggests that the app may have to be adapted for these users.

Disclosure of Interest: None declared

BARRIERS ENCOUNTERED BY GENEVA PEDIATRICIANS FOR THE SCREENING OF ALCOHOL, TOBACCO AND CANNABIS USE BY ADOLESCENTS BETWEEN 11 AND 18 YEARS OLD

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Introduction: Alcohol, tobacco and cannabis use by adolescents continues to be an important health concern in Switzerland. This consumption has consequences on morbidity and mortality in all stages of life. Therefore, the prevention of the use of these substances is essential and pediatricians can play a role through screening and brief interventions. However, the literature shows many barriers limiting this detection. The purpose of this study was to identify the barriers encountered by pediatricians in their daily practice and the solutions to overcome them. We also asked the practitioners about their screening practices.

Methods: We contacted established pediatricians in Geneva and conducted a three steps study based on the Delphi expert consensus method, using questionnaires sent by post. The results of the previous step were added to each next step to help pediatricians to refine their answers and obtain a consensus concerning the barriers.

Results: Of 122 contacted pediatricians, 62 answered the first questionnaire, 34 the second and 25 the third one. More than 99% of the first questionnaire participants think that screening is part of their role. 78% of these participants talk about substance consumption with their patients at least once and 33% consider doing a systematic screening. Five barriers frequently encountered by Geneva pediatricians were identified and some solutions were proposed by them to overcome these obstacles. The barriers were the following: reason of consultation not related to substance use, lack of opportunity (few consultations during adolescence), lack of patient motivation to change behavior, presence of parents during the consultation, lack of treatment or uncertain efficiency.

Conclusion: Some barriers encountered by Geneva pediatricians are specific to their health context. Therefore, the solutions that these practitioners propose are essential because they are adapted to this context and represent useful tracks to help the development of interventions in order to improve prevention in this area.

Disclosure of Interest: None declared

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FROM TOBACCO-RELATED PRODUCTS TO SMOKING: RESULTS FROM A LONGITUDINAL STUDY

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Introduction: To assess longitudinal factors related to smoking among youths depending on smoking status at baseline.

Methods: Data were drawn from the 2 first waves (T0:2016-T1:2017) of the GenFree longitudinal study conducted in 11 post-mandatory schools in the Canton of Fribourg. Participants (N = 1606, mean age 16.2 at T0 [15–24], 45.8% females) were asked at T1 whether they smoked (G1, N = 571) or not (G2, N = 1035). Groups were compared on the use of tobacco-related products (TRP) (e-cigarettes, hookah, snus, and smokeless tobacco) in the past 12 months, use of other substances (alcohol, cannabis) and socio-demographic (age, gender, family structure, SES, academic track) variables at T0 separately by smoking status at T0. Groups were compared at the bivariate level and then at the multivariate level using a logistic regression analysis to assess the variables associated with being a smoker at T1 using non-smokers as reference category. Data are presented as odds ratios (OR) with 95% CI.

Results: Smokers at T0: At the bivariate level, smokers at T1 were significantly more likely to be apprentices and showed a clear trend (p = .052) to be less likely to live in an intact family. At the multivariate level, both variables remained significant: they were more likely to be apprentices (OR: 2.32 [1.28:4.22]) and to live in a non-intact family (1.75 [1.01:3.04]). **Non-smokers at T0:** At the bivariate level, those who became smokers at T1 were significantly more likely to have used TRPs (45.7% vs. 18.2%; p < .001), to have misused alcohol, and to have used cannabis at T0. In the multivariate analysis, having used TRPs (3.05 [1.92:4.85]) and cannabis (2.89 [1.41:5.90]) remained significant. Moreover, when analyses were repeated separately by product, they remained significant (E-cig: 2.46 [1.54:3.91]; Smokeless tobacco: 1.88 [1.12:3.15]; Snus: 2.05 [1.07:3.92]; Hookah: 2.68 [1.73:4.14]).

Conclusion: These longitudinal results show a key association between using TRPs and starting to smoke and seem to confirm the gateway effect of products such as hookah, snus, smokeless tobacco, and e-cigarettes. Thus, the potential harmful impact of these should not be underestimated. The association with cannabis is also important given that in Switzerland cannabis is mostly consumed in joints including tobacco. As an important percentage of non-smoking youths try different TRPs and act as an entry point to cigarette use, TRPs should be part of substance use screening among adolescents. Tobacco prevention should be broadened to include TRPs.

Disclosure of Interest: None declared

care, especially since this might be the only time the adolescent is seen by a health professional. Further research is necessary in order to suggest the best approach when dealing with adolescents and improve medical documentation.

Disclosure of Interest: None declared

ARE ADOLESCENTS WITH CHRONIC CONDITIONS MORE BULLIED OR CYBERBULLIED?

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Introduction: To assess whether bullying and cyberbullying are associated with having a chronic condition at adolescence.

Methods: Data were drawn from the 2017 wave of the GenerationFRee longitudinal study conducted in 11 post-mandatory schools in the Canton of Fribourg. Participants (N = 2208, mean age 17.7 [16–25], 46.3% females) were asked whether they had a chronic condition (CC) that limits their daily activities (CCLim, N = 54, 2.45%), that does not limit their daily activities (CCNonLim, N = 255, 11.55%), or no condition (NoCC, N = 1899, 86.01%). Groups were compared on having been bullied in the past 12 months defined as having been mocked or insulted, physically aggressed or brutalized, or excluded or prevented to participate; and having been cyberbullied in the past 12 months defined as having been mocked or insulted or excluded from a group on Internet, or that someone tried to steal their identity on internet and posted harmful facts. Groups were compared at the multivariate level using separate multinomial regressions analyses for bullying and cyberbullying using NoCC as the reference category, controlling for age, sex, and academic track. Data are presented as relative risk ratios (RRR) with 95% CI.

Results: Bullying prevalence was: 56.22% for CCLim, 54.18% for CCNonLim, and 47.17% for NoCC. Cyberbullying prevalence was: 21.42% for CCLim, 8.94% for CCNonLim, and 8.98% for NoCC. At the multivariate level, the CCNonLim group was significantly more likely than the NoCC group to be bullied (RRR: 1.34 [1.01:1.78]). The CCLim was significantly more likely than the NoCC group to be cyberbullied (2.83 [1.42:5.64]).

Conclusion: These results show additional vulnerability of adolescents suffering from a CC regarding cyber/bullying. For the CCNonLim, a hypothesis can be that they might be more easily bullied because of their CC although not severe enough to arouse pity that can be protective against bullying. On the contrary, for the CCLim, while peers might not dare bullying offline those who have a more intrusive CC-thus acting as a buffer against bullying-, they might get more easily cyberbullied as the limiting condition is not visible online. Given these results, adolescents with CCs need to be screened for bullying and cyberbullying even more than their healthy counterparts because their CC does not necessarily protect them from it, or can even become the cause of it. These youths need to be given tools early on to avoid being cyber/bullied or to know how to deal with it.

Disclosure of Interest: None declared

THE HEADLINE OF ADOLESCENT MEDICINE IN THE EMERGENCY ROOM

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Introduction: Adolescence is a time of multiple biological, cognitive and psychological changes. Youngsters nowadays are confronted with new experiences and risk taking behaviors. They usually seek healthcare in the pediatric emergency room. HEEADSSS (Home, Education, Eating, Activities, Drugs, Sexuality, Suicidality, Safety) is the acronym of a psychosocial screening interview instrument. The use of this tool is suggested for every consultation with a teenager, during individual interview, due to its potential to detect a patient at risk. The aim of this research is to monitor the use of HEEADSSS in our emergency department.

Methods: Retrospective study to examine the number of cases in which HEEADSSS had been used. Every adolescent between 15 and 18 years old who consulted to the emergency department of "Hopital de l'Enfance" during September 2017 was included and their medical files were reviewed.

Results: We included 156 patients (54% females). The main reasons for consultation were trauma/muscular pain (43.5%), pediatric emergency (abdominal pain included) (45.5%), surgical emergency (7%) and psychiatric reason (4%). Only 47 of 156 (29.5%) records included HEEADSSS documentation, all of which were incomplete, with only 4 of 47 (8.5%) files being considered as relatively complete. Female cases with HEEADSSS were more frequent (62%). Caregivers always used HEEADSSS when a patient consulted for a psychiatric issue (6/6 100%), less often for a pediatric emergency (43.5%) and rarely for trauma or surgical emergency (12% and 18%, respectively). Commonly, pediatricians collected information about Home (68%) followed by Sexuality (57%). Sexuality was more often addressed in females than males (70% and 30%, respectively). Information concerning Education (55%) and Drug consumption (49%) was also collected. However, only 16 files included information about Activities (34%) and 14 about Eating habits or Suicidality (30% each). Surprisingly, only one case included information about risk taking (2%).

Conclusion: We conclude that important information about adolescents' psychosocial status is insufficiently documented during emergency consultation. HEEADSSS could ameliorate preventive

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ORAL PRESENTATIONS SSP 2

HIGH COVERAGE OF HEPATITIS B VACCINATION AND LOW PREVALENCE OF CHRONIC HEPATITIS B IN MIGRANT CHILDREN DICTATE A NEW CATCH-UP VACCINATION STRATEGY.

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Introduction: Worldwide coverage of hepatitis B (HB) vaccination is increasing. This should be considered when determining the best strategy for catch-up HB vaccination in migrant children who rarely have written proof of past immunization. This study aimed to estimate vaccine protection and chronic HB prevalence in this population and to identify determinants of vaccine protection.

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Methods: New arriving migrant children followed at the Lausanne University Hospital were prospectively enrolled from October 2014 to July 2017. Children and adolescents aged 1 to 18 years were approached for inclusion if they had an unknown immunization status and accepted a dose of HB vaccine injection. HB surface antibody (anti-HBs) serology was performed after 4 to 6 weeks. Anti-HBs ≥ 100 IU/L (group 1) were considered consistent with a booster type antibody response. Patients with anti-HBs < 100 IU/L (group 2) received additional doses of vaccination accordingly, after exclusion of chronic HB in children with anti-HBs < 10 IU/L. Potential determinants of vaccine response were compared between groups.

Results: Two hundred children were available for analyses. Median age was 8.9 years (IQR 4.8–12.9) and 97 (49%) were girls. The majority came from the Oriental Mediterranean WHO region (124/200, 62%). One hundred and sixty-one children (81%) had a booster-type antibody response. Only 1 patient ($< 1\%$) had a chronic HB. Patients with anti-HBs ≥ 100 IU/L were significantly younger ($p < 0.001$), came from the Eastern Mediterranean region ($p = 0.037$) and from urban versus

rural areas ($p = 0.002$), were accompanied Minors ($p = 0.009$) and had no school attendance ($p = 0.023$). Younger age (OR per decreased year, 1.28; 95%CI, 1.05–1.57; $p = 0.017$) and coming from urban areas (OR yes versus no, 1.16; 95%CI, 1.01–1.33; $p = 0.043$) remained significantly associated with group1 in the multivariate analysis. **Conclusion:** A high proportion of children had a booster type response after a single dose of HB vaccine and only one had a chronic HB. These results suggest that HB pre-vaccination screening and full course HB immunisation catch-up would be most of the time not necessary. The younger age of patients with booster type response reflects the recent increase in HB vaccination worldwide. No single determinant could definitely predict seroresponse. Thus, post-vaccination serology remains necessary.

Disclosure of Interest: None declared

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IMPLEMENTATION OF NEWBORN SCREENING FOR SEVERE COMBINED IMMUNODEFICIENCY (SCID) AND SEVERE T CELL DEFICIENCY IN SWITZERLAND

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Introduction: Newborn screening (NBS) for severe immunodeficiencies will be implemented in Switzerland soon. Rationale to screen for severe combined immunodeficiency (SCID) and severe T cell deficiency is that these diseases are asymptomatic at birth and might be fatal within the first year of life, the confirmation of the diseases is easy, and early hematopoietic stem cell transplantation (HSCT) is a curative treatment. Quantification of TRECs (T-cell receptor excision circles) from dried blood spots (DBS) is a sensitive and specific screening test for SCID and severe T cell deficiency. TRECs are a reliable marker of the number of circulating naïve T cells recently emigrated from the thymus. TRECs are undetectable or very low in infants with SCID or severe T cell deficiency. Combination of TREC quantification with detection of kappa-deleting recombination excision circles (KREC) allows to better classify and predict severe defects of T and B cell development and is currently cheaper than TREC measurement alone. **Methods:** To prepare a proposal to the Swiss Health Ministry (BAG) to implement NBS for SCID and severe T cell deficiency in Switzerland, TREC copy numbers from DBS of the original newborn screening cards of 7 babies with confirmed SCID as well as 57 controls were quantified by PCR from 1.5 mm punch. Simultaneous amplification of beta-actin allowed to monitor the amplification of a sample. **Results:** 68 TREC measurements from 7 SCID patients showed a mean of 0.6 copies/µl blood with a range from 0–9 copies/µl, while 131 measurements from 57 controls showed on average 136 copies/µl with a range from 17–350 copies/µl. Thus TREC copy numbers of the 7 SCID patients separated well from controls and were all below the 2.5th percentile of 36 copies/µl suggested as cut-off by the manufacturer. Simultaneous measurement of TREC and KREC on prospective DBS and retrospective DBS from patients with confirmed SCID is currently ongoing. **Conclusion:** The TREC assay is a reliable assay, easily to be implemented into NBS programs. NBS for SCID and severe T cell deficiency is already recommended in the US and a few other countries. Since early HSCT before the occurrence of irreversible organ damage can provide cure for these patients, a proposal to the Swiss Health Ministry (BAG) regarding inclusion in the routine NBS screening program in Switzerland was recently approved and will be implemented nationwide in 2019.

Disclosure of Interest: None declared

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EVOLUTION OF PERCUTANEOUS ENDOSCOPIC GASTROSTOMY FROM 2007 TO 2016 IN CHILDREN AT A TERTIARY CENTER (CHUV)

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Introduction: The insertion of a percutaneous endoscopic gastrostomy (PEG) defines the apposition of the gastric wall to the abdominal wall by endoscopy, using a tube in order to have direct access to the stomach for food, liquid and drug administration. Gastrostomy is the ideal therapeutic approach for longterm enteral nutrition for children with chronic pathologies. PEG has been used

for many years at our institution but until today, no study has been performed assessing risks and benefits of this intervention. We therefore studied over a period of 10 years which patients benefit from the PEG, its complication rate, as well as its impact on weight gain. **Methods:** Monocentric retrospective study (Pediatric Gastroenterology Unit, CHUV, Lausanne) including all paediatric patients, who have benefited from a gastrostomy insertion between January 2007 and December 2016. Creation of a database including the following patients details: sex, age, diagnostic, indication(s), weight and influence on eating behaviour.

Results: Eighty-two patients underwent PEG insertion between 2007 and 2016. One patient was excluded due to lack of information. The median age was 7 years old (1 month 9 days – 17 years 8 months 22 days). There were 46 boys (57%) and 35 girls (43%). Neuromuscular pathologies (46%) were amongst the most frequent indications for PEG insertion due to insufficient caloric intake (85%). Weight gain one year after the intervention was significantly increased (z-score increase of 0.56; –2.41 to –1.85; $p < 0.002$). Ninety-one percent of orally fed patients before PEG insertion continued to do so. Seventy percent of patients without oral feeding before PEG insertion started oral intake probably due to increased appetite whilst on tube feeding.

Conclusion: PEG is an important mean to address nutritional needs of patients with chronic diseases, especially neuromuscular diseases. Regardless of underlying disease, PEG allows significant weight gain, without stopping per oral intake.

Disclosure of Interest: None declared

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ELECTROLYTE AND ACID-BASE ABNORMALITIES IN INFANTS WITH COMMUNITY-ACQUIRED ACUTE PYELONEPHRITIS: PROSPECTIVE CROSS-SECTIONAL STUDY

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Introduction: Retrospective case series suggest that abnormalities in fluid, electrolyte, and acid-base balance can occur among infants with a febrile urinary tract infection. Potentially inaccurate laboratory methods of sodium testing have often been used.

Methods: Between January 2009 and June 2016, we managed 80 previously healthy infants (52 males and 28 females) ≥ 4 weeks to ≤ 24 months of age with their first episode of acute pyelonephritis. Ionized sodium, ionized potassium and ionized chloride were determined by direct potentiometry, as recommended by the International Federation of Clinical Chemistry. Bicarbonate was calculated from pH and carbon dioxide pressure.

Results: Electrolyte or acid-base abnormalities were disclosed at presentation in 59 (74%) of the 80 infants: hyponatremia ($n = 54$), hypobicarbonatemia ($n = 18$), hyperkalemia ($n = 14$), hyperbicarbonatemia ($n = 6$), hypochloremia ($n = 3$), hypokalemia ($n = 3$), and hyperchloremia ($n = 1$). Patients with and without electrolyte or acid-base abnormalities did not differ with respect to age, sex distribution, and whole blood glucose. Blood tonicity was lower and poor fluid intake, frequent regurgitations or loose stools more common among infants with electrolyte or acid-base abnormalities.

Conclusion: This prospective study shows that electrolyte or acid-base abnormalities occur in approximately 3 quarters of infants with acute pyelonephritis. Among these, hyponatremia is the most frequent abnormality.

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Disclosure of Interest: None declared

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PEDIATRIC FEVER AND NEUTROPENIA WITH BACTEREMIA – PATHOGEN DISTRIBUTION AND ANTIBIOTIC EFFICACY OVER TIME IN A RETROSPECTIVE COHORT STUDY

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Introduction: Fever and neutropenia (FN) is a potentially life-threatening complication of chemotherapy in pediatric cancer patients. Current standard of care is emergency hospitalization and empirical initiation of broad-spectrum antibiotic therapy. This single center study evaluated epidemiology of bacteremia in children with FN over a 20-year time period.

Methods: We analyzed in retrospect FN episodes with bacteremia in pediatric cancer patients in a cohort from 1993 to 2012 in a single center not applying antibacterial prophylaxis beyond *Pneumocystis jirovecii* prophylaxis. The distribution of pathogens, efficacy of antibiotic therapy, and their trends over time were assessed. Efficacy of the standard empirical antibiotic therapy in our center (ceftriaxone plus amikacin) was compared to currently recommended antibiotic therapy regimens.

Results: From a total of 712 FN episodes reported, we assessed 136 FN episodes with bacteremia with 198 pathogens isolated in 102 patients. Gram-positive pathogens (127, 64%; 95% CI: 57%–71%) were more common than Gram-negative (71, 36%; 95% CI: 29%–43%). This proportion did not change over time ($p = 0.26$), but we observed a significant change in Gram-negative pathogen distribution ($p = 0.036$), mainly due to an increase of *Klebsiella* spp. Coagulase-negative staphylococci (64, 32%), viridans group streptococci (42, 21%), *E. coli* (33, 17%), *Klebsiella* spp. (10, 5%) and *P. aeruginosa* (nine, 5%) were the most common pathogens. No extended spectrum beta-lactamase-producing *Enterobacteriaceae* or methicillin-resistant *S. aureus* were isolated. Comparing the efficacy of empirical antibiotic therapy on isolate level for ceftriaxone plus amikacin (91%; 95% CI: 84% – 96%), cefepime (90%; 95% CI: 83% – 95%), meropenem (93%; 95% CI: 86% – 97%), and piperacillin/tazobactam (89%; 95% CI: 81% – 94%), respectively, showed no significant differences.

Conclusion: Over two decades, we reported a relative stable pathogen distribution. No significant trend in efficacy of standard empirical antibiotic therapy was found in our setting. Different recommended antibiotic regimens showed comparable in vitro efficacy, although piperacillin/tazobactam could not be recommended as a first line treatment in Bern, because of local resistances among *Enterobacteriaceae*.

Disclosure of Interest: None declared

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MICRO-RNA'S IN SICKLE CELL DISEASE

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Introduction: Sickle Cell Disease (SCD) is a monogenic blood disorder, which is seen more frequently in Switzerland due to migration. Due to a single nucleotide mutation in SCD the red blood cells (RBCs) take on a sickled shape, clog small blood vessels and lead to vaso-occlusion, organ ischemia and hemolytic anemia. Specifically recurrent pain crises, chest syndrome and strokes occur, compromising the patients quality of life and can even be life threatening. The symptoms can ameliorate in the presence of higher amounts of fetal hemoglobin (HbF). HbF can be elevated in RBCs of children and adults by pharmacological agents, such as hydroxyurea (HU). Most patients benefit from this treatment, however, the treatment responses are highly variable and the mechanism is not yet well understood. We hypothesize that altered micro-RNA expression might be one effect of HU. Micro-RNAs (miRNAs) are small non-coding ribonucleic acids which play a role in gene expression. MiRNAs control genes post-transcriptionally by binding to mRNAs with (partial) sequence complementarity. They hereby degrade or repress these mRNAs. miRNA profiles hence suggest different gene expression profiles in individuals.

Results: Previously, we have analysed the expression of 380 miRNAs in healthy individuals, SCD patients and SCD patients treated with HU. We observed clear differences in miRNA expression profiles between these groups. In *ex vivo* erythropoiesis experiments (cultures of erythropoietic cells) we propose to show the effects HU has on miRNA expression of both controls and of SCD patients with and without HU treatment. The results will be compared to the *in vivo* results already generated in the above mentioned patient groups. With the *ex vivo* erythropoiesis we will have developed a biological model in which we can perform further research on SCD and on HU treatment.

Conclusion: By understanding the regulation of erythropoiesis by miRNAs in patients and healthy individuals we could gain more insight into the mechanism of HU and HbF regulation, into other hematological disorders, such as thalassemia and ultimately identify new therapeutic targets.

Disclosure of Interest: None declared

TESTICULAR TISSUE CRYOPRESERVATION FOR FERTILITY PRESERVATION IN MALE PEDIATRIC CANCER PATIENTS: A PROMISING EXPERIMENTAL RESEARCH PROCEDURE

038

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Introduction: Major advances in the treatment of childhood cancer have resulted in a 5-years survival rate of more than 80%. Unfortunately, cancer treatments may adversely affect fertility. More than 40% of male cancer survivors will have compromised reproductive function. While sperm cryopreservation (SC) is an established method for fertility preservation (FP) in pubertal males, testicular tissue cryopreservation (TTCP), preserving spermatogonial stem cells (SSC), is the only available option for prepubertal boys. Although not yet available in humans, auto-transplantation of SSC or *in vitro* spermatogenesis may restore fertility in infertile cancer survivors.

Methods: Since 2015 an experimental research protocol offering TTCP to prepubertal boys and pubertal patients who failed to cryopreserve sperm is ongoing in HUG (Geneva), CHUV (Lausanne), and more recently in UKBB (Basel). The experimental procedure is offered only to patients scheduled to receive a highly gonadotoxic treatment, and after acceptance by a multidisciplinary team. The testicular biopsy, usually performed in combination with another procedure, is cryopreserved at the Laboratory d'Andrologie et Biologie de la reproduction (CHUV). In Suisse Romande, costs of the procedure are covered by the Association Zoé4life

Results: Between July 2015 and December 2017, TTCP was proposed to 14 patients (age: 2–17 years, mean 7.1 years) and was accepted by 11 families. An informed consent form was signed in all cases.

Patient	Age (years)	Pubertal status	Diagnosis	Chemotherapy prior to testicular cryopreservation	Highly gonadotoxic treatments planned	Testicular cryopreservation / complications	Number of testicular fragment collected
1	2 y	Prepubertal	High risk neuroblastoma	Yes	Bu-Mel (HSCT)	Yes / No	12
2	4 y	Prepubertal	Medulloblastoma	No	CCNU - Cisplatine	Yes / No	26
3	14 y	Pubertal	Medulloblastoma	No	CCNU - Cisplatine	Yes / No	55
4	2 y	Prepubertal	High risk neuroblastoma	Yes	Bu-Mel (HSCT)	Yes / No	16
5	7 y	Prepubertal	High risk neuroblastoma	Yes	Bu-Mel (HSCT)	Yes / No	26
6	2 y	Prepubertal	Relapsed acute myeloblastic leukemia	Yes	Bu-Cy (HSCT)	Yes / No	23
7	2 y	Prepubertal	High risk neuroblastoma	No	Bu-Mel (HSCT)	Yes / No	22
8	10 y	Prepubertal	Progressive Hodgkin disease	Yes	BEAM (HSCT)	Yes / No	19
9	3 y	Prepubertal	High risk neuroblastoma	Yes	Bu-Mel (HSCT)	Yes / No	18
10	17 y	Pubertal	Acute lymphoblastic leukemia, P1+	Yes	Flu-BU-Thiolepa (HSCT)	Yes / No	60

HSCT: Hematopoietic Stem Cell Transplantation

Table

Testicular fragments were collected and cryopreserved for 10 patients (age: 2–17 years, mean 6.3), in one case TTCP was not possible because of a prolonged aplasia. All patients except 3, underwent several courses of non sterilizing chemotherapy before TTCP. Post-surgery follow-up was uneventful in all patients. The average number of testicular slices cryopreserved was 27.7 per patient (12–60). In all cases, only one testis was biopsied and no tumoral cells were detected. One patient died of the disease during the follow-up period.

Conclusion: Cancer treatments can be highly gonadotoxic in male patients treated during childhood. For peripubertal or pubertal patients SC has to be proposed. Our results confirmed that TTCP is a safe experimental procedure which can be offered when sperm retrieval is not feasible. Since testicular tissue contains SSC from which spermatozoa are derived, future procedures may restore fertility in these patients.

Disclosure of Interest: None declared

039

CALORIC RESTRICTION AT ADULTHOOD REVERSES HEPATIC DYSFUNCTIONS INDUCED BY TRANSIENT POSTNATAL OVERFEEDING IN MICE

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Introduction: Increased rates of childhood obesity, leading to increased occurrence of metabolic syndrome, are observed worldwide. An altered nutritional environment during critical periods of development can lead to metabolic disorders later in life. The liver, involved in lipid/glucose homeostasis, is particularly vulnerable to nutritional programming during the perinatal period. Oxidative stress (OS) associated with stress-induced premature senescence (SIPS) have been involved in metabolic and liver dysfunctions. In this study, we investigated in a murine model whether transient postnatal overfeeding (OF) can lead thereafter to metabolic and hepatic disorders associated with OS and SIPS, and if a moderate caloric restriction (CR) at adulthood can reverse these dysfunctions.

Methods: C57BL/6 male pups were maintained, during the lactation period, in litters adjusted to 9 pups for normal feeding (NF) or reduced to 3 pups to induce transient postnatal OF. After weaning all mice had

free access to a standard diet. At 6 months of age, mice from NF and OF groups were randomly assigned to either the *ad libitum* (AL) diet or the caloric restriction diet (CR, daily food supply reduced by 20%) for one month. The following parameters were studied at 7 months of life: i) body weight; ii) markers of OS (reactive oxygen species, antioxidant defenses); iii) markers of SIPS (factors involved in cell cycle arrest (p21, p53, Acp53, p16, pRb/Rb), SIRT-1); iv) liver structure (histological analysis).

Results: At 6 months of life, OF vs. NF mice displayed an increased area under curve of blood glucose concentration after glucose challenge as well as a higher blood glucose concentration after insulin injection ($p < 0.05$). At 7 months, body weight of OF mice was 11.7% higher compared to NF animals. In liver from 7 months old OF vs. NF mice we observed: i) higher levels of superoxide anion, decreased catalase and superoxide dismutase expression ($p < 0.01$); ii) increased expression of p21, p53, Acp53 and p16, but decreased pRb/Rb and SIRT-1 expression ($p < 0.01$); iii) microvesicular steatosis and hepatic fibrosis. CR at adulthood decreased body weight, reversed OS, SIPS, and microvesicular steatosis, but did not reverse hepatic fibrosis.

Conclusion: A transient postnatal OF during the lactation period leads at adulthood to liver oxidative stress, SIPS and altered hepatic structure. A moderate CR at adulthood reverses these hepatic dysfunctions.

Disclosure of Interest: None declared

GONG SHOW

040

THE GANGLIONEUROMA: A RARE TUMOR OF THE POSTERIOR MEDIASTINUM

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Introduction: Ganglioneuroma is an uncommon benign tumor that arises from the sympathetic nervous system accounting for less than 1% of all soft-tissue neoplasms. We report a case of thoracic ganglioneuroma.

Methods: A 6-year-old girl with no pathological history. The clinical examination was strictly normal except for a persistent dry cough.

Results: Initial chest radiograph shows a large mass in the right side of the posterior mediastinum with no rib destruction. The chest CT scan showed right posterior mediastinal mass, well circumscribed and contained a mixed attenuation with calcifications. The surrounding structures do not appear affected by the mass. There was no mediastinal lymphadenopathy and the rest of the exam results were normal. The surgical exploration and histologic examination revealed a ganglioneuroma.

Conclusion: the ganglioneuroma is a rare neurogenic tumor. His discovery is often fortuitous during a radiological examination. His diagnosis is confirmed by histological examination. It is a benign tumor of good prognosis. The treatment is surgical.

NaCHwuchs Prize: I wish to apply
Disclosure of Interest: None declared

Results: Both patients presented at birth with a lower body IH, myelopathy, bony deformities and anorectal malformations. The male patient additionally had urogenital malformations. In both children, the IH was of the segmental type. Both were treated operatively for anorectal malformations, whereas the male patient was also treated for an intraspinal myelocystocele and lipomyelocele. Both patients are in regular follow-up at our institution.

Conclusion: Unusual findings in the pelvic area in association with an IH should prompt an evaluation for LUMBAR syndrome. This includes an MRI of the spine, abdomen and pelvis. The IH in this setting are usually of the segmental type and should not be mistaken for port-wine stains.

Disclosure of Interest: None declared

042

PERCUTANEOUS TREATMENT OF FRALEY SYNDROME IN A 2-YEAR-OLD BOY

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Introduction: Fraley's syndrome describes the dilatation of the superior renal calyx resulting from a compression of the upper pole infundibulum by intrarenal vessels. The current literature mostly reports open surgical approaches when treating this condition.

Methods: We present the case of a boy with prenatal diagnosis of an empty left renal fossa, a dilated upper part of the right kidney and a cardiac ventricular septal defect (closed by a patch at 1.5 years). The patient was asymptomatic with no urinary tract infection and normal renal function. Because of the progressing dilatation of the superior system up to 38 mm a cystoscopy with retrograde pyelogram was performed to better understand the anatomy and revealed the presence of a single kidney with a narrowing of the superior infundibulum and an ectatic upper calyx with delayed drainage (fig. 1). At the age of 2.5 years a percutaneous balloon dilatation of the stenotic upper pole infundibulum (Armada 4/20 mm balloon, 16 bars) was performed. An 8.5 French drainage catheter was placed for one week.

041

LUMBAR SYNDROME – YET ANOTHER ASSOCIATION WITH ANORECTAL MALFORMATIONS

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Introduction: Anorectal malformations have been associated with various syndrome complexes, such as VACTERL association and Currarino syndrome. Furthermore, they may occur in the setting of a segmental infantile hemangioma (IH) of the lower body, together with spinal and urogenital malformations – an association known as LUMBAR syndrome (Lower body IH and other skin defects, Urogenital anomalies or ulcerations, Myelopathy, Bony deformities, Anorectal malformations or arterial anomalies, and Renal anomalies). We present 2 cases with LUMBAR syndrome to highlight the features and management of this rare syndrome.

Methods: Review of two cases with LUMBAR syndrome treated at our institution.

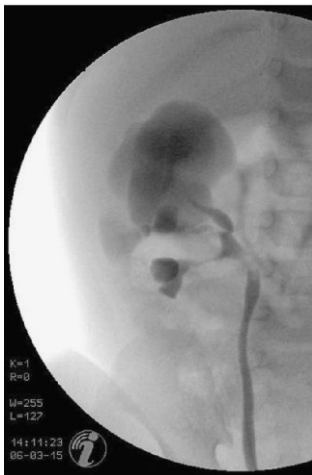


Figure 1

Results: Postinterventional ultrasonographic control showed an initial regression of the calyx dilatation, and this remained at one-year follow-up. Dynamic renal MAG3-scintigraphy one year after the percutaneous dilatation described no significant obstruction, with good drainage of the upper calix and a stable upper- and lower-pole function.

Conclusion: To the best of our knowledge this is the first description of a percutaneous dilatation-treatment of Fraley's Syndrome in a paediatric patient. This case also emphasizes the utility of a retrograde pyelography to diagnose Fraley's syndrome.

Disclosure of Interest: None declared

O43

EOSINOPHILIC CYSTITIS – A RARE CAUSE OF PAINLESS MACROSCOPIC HEMATURIA: CASE REPORT

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Introduction: Eosinophilic Cystitis is a very rare condition in pediatric population. Symptomatology is versatile and treatment protocols are non-standardized.

Methods: We report the case of a 12 year old boy, who presented with recurrent painless hematuria, without any additional symptoms concerning micturition.

Results: Patient history showed two urinary tract infections without fever. Ultrasound demonstrated a polypoid mass in the posterior wall of the bladder. A cystoscopy revealed extensive papillary tumors with solid parts in the trigone and the right and left bladder wall, respectively. The morphology of the mass seemed similar to a rhabdomyosarcoma of the bladder. Biopsies indicated an inflammatory lesion dominated by eosinophils (eosinophilic granulomatous cystitis). A MRI of the abdomen and pelvis showed a thickened and irregularly configurated bladder wall. An 8 week course of oral corticosteroids and antihistamins was initiated. A repeat cystoscopy two weeks after finishing therapy demonstrated resolution without any residual lesions. Biopsies showed only a few eosinophils. Macroscopic hematuria was not seen any more and the patient remained asymptomatic in the nine months follow up.

Conclusion: Eosinophilic Cystitis in the pediatric population is very rare. Therapy with oral corticosteroids and antihistamins in our case was successful. To date, less than 60 pediatric patients with this condition have been reported in the literature, with described treatment options ranging from spontaneous remission to partial cystectomies.

Disclosure of Interest: None declared

O44

UNUSUAL ISOLATED FALLOPIAN TUBE TORSION

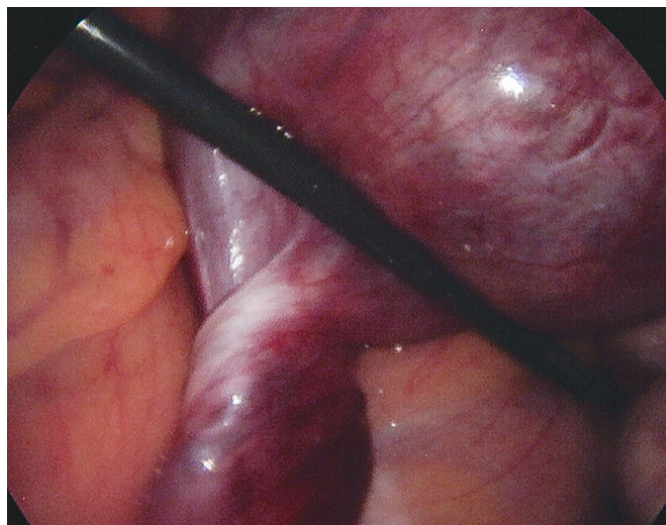
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Introduction: Isolated fallopian tube torsion (IFTT) without torsion of the adnexal structure is a rare surgical emergency. The incidence of this pathology is 1 in 1.5 million. We present the case of a teenager presenting with a large fallopian tube cyst causing IFTT and additional kinking of the tube over the meso of the sigmoid.

Methods: A 15 year old girl had previously been diagnosed with a suspected large ovarian cyst (8 cm) and started on combined oral contraceptive pill for 3 months with no significant improvement. She was referred to our pediatric emergency department by her gynecologist when she presented with acute severe pain in the left upper abdominal quadrant with vomiting and fever at 38.9°. She had usual menstruation the previous week with negative pregnancy test. The physical examination revealed tenderness and a palpable mass in the left subcostal area without guarding. The blood test showed a slight leucocytosis 13.9 (N: 4.5–13) and the ultrasound described an 9 cm ovarian cyst in the left upper abdomen with normal vascularization of the ovary and moderate ascites.

Results: An urgent laparoscopic exploration demonstrated a large cyst incorporated in the left fallopian tube. The tube was elongated with a 180 degrees twist (image 1) and folded over the meso of the sigmoid causing it to lie in the upper abdomen. Once the tube was freed from the mesosigmoid and after derotation of the fallopian tube, we were able to identify the left ovary which looked normal. The benign-looking cyst was drained, and we were able to simply peel off the cyst in its totality. Histopathology confirmed a simple tubal cyst.



Conclusion: Our report underlines the need to consider IFTT in the differential diagnosis of abdominal pain even if located in the upper abdominal quadrants. It also reminds us that paraadnexal and adnexal cysts can be found outside the pelvic area – especially in children. The combination of a fallopian torsion, with the cyst and fallopian tube folded over the mesosigmoid is a rare finding and, to the best of our knowledge, has not yet been described in the literature.

Disclosure of Interest: None declared

O45

PERISTALSIS FOR ETERNITY

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Introduction: Foregut duplication cysts are rare congenital malformations. They result from abnormal development of the embryonic foregut and are classified into three main types: bronchogenic, enteric, and neurenteric. Here, we present the case of an extremely rare neurenteric cyst with a striking "functionality".

Methods: Case Report: A 3-year-old healthy boy presented with a lump located over the spinal process of C5, present since birth. Imaging studies revealed a complex, partly cystic malformation residing in the posterior mediastinum and right thoracic cavity, involving also bony and neural structures. After observing remarkable growth, the lesion was resected by a multidisciplinary surgical team. The specimen exhibited a spontaneous, strong and long lasting peristalsis that was video-documented. Histology revealed a neurenteric cyst.

Conclusion: Cystic mediastinal masses extending into the spinal canal or associated with spinal malformations may be neurenteric cysts requiring a detailed interdisciplinary diagnostic workup as well as expert surgical therapy.

Disclosure of Interest: None declared

046

INTRAABDOMINAL LYMPHANGIOMA CAUSING OBSTRUCTIVE ILEUS IN A CHILD: REPORT ON A RARE CASE

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Introduction: Intrabdominal lymphangiomas are rare tumor-like lesions mostly found in the first decade of life. Literature indicates a fetal origin caused by scars of lymphatic tissue.

Methods: We report on a 3-year old boy who presented to the emergency department (ER) with a history of repetitive vomiting for 72 hours and abdominal discomfort without diarrhea or fever. Laboratory results on admission were not specific for inflammation. Physical examination revealed a non-tender, soft abdomen and regular bowel persistalsis. Due to biliary vomiting on day 2 an abdominal x-ray was done consistent with obstructive ileus. Laparotomy was indicated and a cystic lesion with 15 cm in diameter approximately 65 cm proximal to the cecal valve and originating from the mesentery was found. Resection of the cyst as well as a segmental intestinal resection with a consecutive end-to-end anastomosis was carried out.

Results: The patient recovered uneventfully and could be discharged on day 11. Histological examination revealed a serosa-coated lymphangioma without any signs of malignancy. Ultrasound after 3 and 6 months could rule out any recurrence of the lymphangioma.

Conclusion: Abdominal lymphangioma is an uncommon cause for acute abdomen in children. Complete surgical resection is effective and has to be considered as treatment of choice.

NaCHwuchs Prize: I wish to apply

Disclosure of Interest: None declared

047

PERINATAL BILATERAL TESTICULAR TORSION – A CASE REPORT

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Introduction: Perinatal testicular torsion (PTT) is a rare condition, especially if it presents bilaterally. It is defined as prenatal testicular torsion or within the first 30 days of life. Only 25% of them occur after birth. There are only few studies, mostly case reports. Larger studies include often patients with prenatal onset. Therefore different treatment strategies are described, but prognosis remains poor.

Methods: We present a newborn boy, who was transferred to our Department at an age of 6 hours. He was delivered at 40 weeks of gestation out of breech presentation. Shortly after birth, a painful discoloration and swelling of the scrotum was noticed. Clinical examination as well as an immediately performed ultrasound was highly suspicious for testicular torsion of the right side. We decided for emergency surgery and took the boy to the operating theatre.

Results: During operation we found a bilateral testicular torsion with dark coloration of both testicles. After packing for about 15 minutes, the right side seemed to recover slightly. The testicles were left in situ, and a bilateral orchidopexy was performed. After 2 days, ultrasound examination showed reperfusion of both testicles and the boy was discharged from hospital. Follow-up was done after 2 and 6 months, where we had normal clinical examination and sonographically persistent perfusion of both testicles. Endocrinological investigation during minipuberty showed normal hormonal values.

Conclusion: Bilateral perinatal testicular torsion represents a very rare condition. Immediate surgical exploration is recommended in case of postnatal onset, with exploration of the contralateral side and bilateral fixation. Concerning the risk of anorchia in bilateral cases, preservation of the testicles should be considered even in ischemic testicles without obvious recovery.

Disclosure of Interest: None declared

048

MANAGEMENT OPTIONS IN ACUTE ON CHRONIC HYDROCEPHALUS IN CHILDREN

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Introduction: Acute exacerbation of hydrocephalus is a relatively rare entity in children, yet can require urgent surgical intervention. We aim to discuss the surgical options for treating acute hydrocephalus.

Methods: We report on clinical presentation and management of a 6-year old boy who was admitted to our Emergency Department (ED) presenting with a history of vomiting and headache for three weeks and recently emerged ataxia. Cerebral MRI with contrast revealed obstructive hydrocephalus internus with enlargement of both lateral ventricles as well as the third and the fourth ventricle. Trans-ependymal CSF-diapedesis was found as representing a sign of a chronic increase in intracranial pressure; most likely resulting from a former bleeding in the fourth ventricle with subsequent septum formation (see MR scan). There was no evidence of an intracranial tumor. To decrease intracranial pressure an emergency surgery was performed while intracranial pressure (ICP) was 35 cm H₂O. As our treatment option of choice we placed a ventriculo-peritoneal (VP) shunt with an opening pressure of 16 cm H₂O after gradually reducing the ICP. Postoperative clinical course was uneventful during a follow up of 3 months.

Results: Patients with acute progressive hydrocephalus require urgent surgical intervention. There are 4 treatment options: 1) VP-shunt, 2) temporary external ventricular drain (EVD) with risk of infection and abrupt pressure decrease, 3) endoscopic third ventriculostomy (ETV) risking uncontrolled decrease of CSF and 4) posterior occipital craniotomy.

Conclusion: In children with onset of acute on chronic ICP elevation with clinical and radiological symptoms of liquor-diapedesis, narrowed extracranial space and neurological deterioration, a controlled reduction of ICP through a VP-shunt is feasible, safe and effective.

NaCHwuchs Prize: I wish to apply

Disclosure of Interest: None declared

049

LAPAROSCOPIC ADRENAL-SPARING RESECTION OF BILATERAL METACHRONOUS PHEOCHROMOCYTOMA IN A PATIENT WITH VON HIPPEL-LINDAU DISEASE

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Introduction: Pheochromocytomata occur in about 20% of patients with von Hippel-Lindau disease (VHL). As they can present bilaterally, adrenal-sparing surgery is mandatory to prevent patients from developing Addison disease. To our knowledge, successful adrenal-sparing resection of bilateral metachronous pheochromocytoma as well as of a recurrent tumour on one side has not been described so far.

Methods: We report the case of a 11 years old boy with VHL and bilateral metachronous pheochromocytoma. The patient showed no symptoms. During routine check-up, urine catecholamine levels were elevated and MRI revealed a tumour on the left side. In 02/2016, the patient underwent laparoscopic adrenal-sparing surgery with a complete enucleation as R0-resection of the pheochromocytoma. Preoperatively, the patient received a blockade with Inderal and Propranolol. In 06/2016, recurrence of the tumour was suspected when serum levels of catecholamine increased. MRI-evaluation revealed a contralateral tumour on the right side and therefore surgery was repeated in the same fashion. Again, complete tumour resection could be achieved. In the further course the patient was asymptomatic both clinically as well as for laboratory investigations. In 11/2016, routine follow-up investigation through MRI showed a recurrent tumour on the left side, however, urine and serum catecholamine levels remained within normal range. Therefore we proceeded with a closely monitored watch and wait for ten months. In 09/2017, a F18-DOPA-PET/CT revealed the tumour to be active metabolically, suggesting a recurrence of the pheochromocytoma. Again, laparoscopic adrenal-sparing surgery was performed. Histology of the resected tumour showed complete margins.

Results: Postoperatively, sonography showed regular perfusion of both kidneys. Initially, serum-levels of cortisol were low, but recovered spontaneously to normal after a few days.

Conclusion: In patients with pheochromocytoma, adrenalectomy is the Gold standard. As patients with VHL are at risk to develop a bilateral pheochromocytoma, which can occur metachronously, adrenal-sparing surgery, though technically challenging, is mandatory. It allows the possibility to maintain endogenous steroid production. However, with remaining adrenal gland tissue there is a risk of developing a recurring tumour. Laparoscopic adrenal-sparing resection of pheochromocytomata in paediatric patients is safe and provides curative therapy for patients with VHL.

Disclosure of Interest: None declared

050

A SIMPLE TREATMENT METHOD OF A PINGPONG-FRACTURE IN A NEWBORN

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Introduction: Depressed skull fractures in newborns are rare. They may develop prenatal or perinatal, when they are most often associated with instrument-assisted deliveries (i.e. forceps). They are also referred to as pingpong fractures whenever there is no complete interruption of the bone. Often the deformation of the head caused by depressed skull fractures seems to resolve spontaneously; but there are also cases where a poor cosmetic outcome persisted into adulthood. Traditionally these fractures are managed surgically under general anesthesia. We would like to present a case in which we used a simple method of a closed reduction of the fracture.

Methods: The newborn boy presented with a depressed skull fracture after being born via forceps-assisted C-section. The fracture was in the right parietal bone and the altered shape of the head could be easily palpated, but also seen from distance. Other than that the boy didn't show any abnormalities in the clinical examination. An ultrasound of the head confirmed the diagnosis of a pingpong fracture and showed no evidence of intracranial hemorrhage. We decided to perform a

closed reduction in applying a vacuum directly over the fracture. We used a pediatric CPR mask with a 50ml empty syringe connected to it. After inducing an analgosedation with ketamin we applied a negative pressure over the fractured skull for around 2 minutes. The fracture then showed itself to be completely reduced and the shape of the head looked harmonic.

Results: In this case the closed reduction using this simple device was very successful. It was a short and simple procedure for which the child didn't need to undergo general anesthesia. The post-reduction ultrasound done directly after the procedure showed a very small epidural or subdural hematoma limited to the location of the reduced fracture, which resolved spontaneously in one day. The boy never showed any clinical abnormalities and could be discharged the next day.

Conclusion: We would like to draw the attention to this rare form of skull fracture and its simple and safe treatment option with excellent cosmetic results. Medical staff working with newborn children should be aware of this option and know that it is best performed during the first 72 hours of life. Therefore a newborn baby with a depressed skull fracture should be transferred to a hospital where this procedure can be done during the first days of its life.

Disclosure of Interest: None declared

ORAL PRESENTATIONS SSCP 1

051

MATERNAL COMPLICATIONS AFTER OPEN FETAL MYELOMENINGOCELE REPAIR

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Introduction: Open fetal myelomeningocele (MMC) repair provides better outcomes than postnatal care. The present study wants to document type and incidence of maternal complications.

Methods: We analyzed the data of our first 40 fetal MMC repairs performed between 2010-2016. Using the Clavien-Dindo classification, maternal complications were subdivided into 5 severity levels: grade 1 is a minor complication not requiring any pharmacological treatment or surgical intervention; grade 2 mandates pharmacological treatment; grade 3 requires surgical intervention; grade 4 is a potentially life-threatening complication requiring IC/ICU management, and grade 5 is death.

Results: We observed no grade 5 complication. 5 (12.5%) women demonstrated grade 4 complications. One case of uterine rupture (2.5%) was a direct consequence of fetal surgery. A third-degree AV-block was clearly attributable to magnesium tocolysis (now abandoned). The other three grade 4 complications, including bilateral lung embolism, chorioamnionitis, and urosepsis, are not unequivocally caused by fetal surgery. 26 (65%) women showed minor complications (grade 1-3).

Conclusion: Severe maternal complications are rare, while minor complications are relatively frequent. Notably, the toll for the baby's better prognosis is courageously paid by the expecting mother, an "innocent bystander".

Disclosure of Interest: None declared

Methods: 8 children received an omphaloplasty in GOSH-technique in our institution between 07/2014 and 12/2017. The technique involves using two skin flaps at each side of the midline incision, one with subcutaneous pedicle and the other with intact cutaneous bridge. The first flap is rolled with its pedicle to create the internal part of the umbilicus and fixed it in the midline. Then the other flap is used to configure the external portion of the neo-umbilicus by rolling it around the first flap. Finally the midline incision is closed and reconstructed.

Results: In 6 children omphaloplasty was created during complex urologic reconstructive surgeries; 3 boys with bladder exstrophy, 2 boys with complex cloacal exstrophy and in one girl with Prune-Belly syndrome associated with cloacal anomaly. In the other two cases it was during reconstruction of abdominal wall; one boy with giant omphalocele and one boy after multiple surgeries of complicated gastroschisis. During follow up there were no complications of wound infection or necrosis, patients and their families were satisfied with the results of stable elastic and cosmetically acceptable neo-umbilicus.

Conclusion: Omphaloplasty in GOSH-technique has been developed to obtain a stable inverted neo-umbilicus in children with bladder exstrophy and other abdominal wall defects.

Disclosure of Interest: None declared

053

WOUND REPAIR MANAGEMENT: SUTURE VERSUS GLUE – DO WE HAVE A CLUE? PRELIMINARY RESULTS OF A PROSPECTIVE NON-RANDOMIZED CLINICAL TRIAL

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Introduction: Traumatic skin lacerations are a very common reason for Pediatric Emergency consultations. Despite these lacerations being a very common problem, evidence for the best treatment strategy is still pending. According to the Cochrane Database Analysis of 2009, tissue glue warrants an acceptable alternative to suture. The advances of glue are a shorter and less painful procedure. However, prior studies have shown a certain increased, but statistically not significant risk of complications, mostly wound dehiscences. In addition, a few studies have reported large discrepancies between the patients' body image and their scar rating by a medical professional. For this reason, The Children's University Hospital of Zurich launched a large multidisciplinary clinical trial, prospectively investigating the long-term outcome of 400 pediatric patients with skin lacerations in the facial area.

052

OMPHALOPLASTY IN GOSH-TECHNIQUE – OUR FIRST EXPERIENCE IN PATIENTS WITH BLADDER EXSTROPHY AND ABDOMINAL WALL DEFECTS

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Introduction: Body image perception is crucial for both males and females with abdominal wall defects, like the absence of a normal looking umbilicus with lower abdominal scars. We present our first experiences in omphaloplasty in GOSH-technique.



Figure 1 (The photograph is published with an informed consent from the parents.)

Methods: Every patient with a traumatic facial skin laceration, who presents at the Pediatric Emergency unit of the Children's University Hospital of Zurich and meets the inclusion criteria, is eligible. The first visit consists of the wound repair, using either tissue glue or suture. We schedule follow-up visits at our plastic surgery outpatient clinic 5–10 days after the accident (wound check-up), and 6–12 months postinjury (scar check-up). At every visit, we perform a clinical examination and a brief interview. Photo documentation is completed at all visits. Primary outcome is the cosmetic appearance, assessed by external plastic surgeons in England and Canada. Secondary outcomes are the occurrence of complications, cost-effectiveness, patient's satisfaction and children's quality of life, using standard assessment scales.

Results: To date, we enrolled 208 patients, of whom 131 with sutured (= group A) and 77 with glued (= group B) wounds. Patients demographics between the two groups were not significantly different. As expected, median treatment time was significantly longer for suture (15 min) than for glue (5 min). Rate of complications (as so far monitored at date of wound check-up) is low: 1.7% in group A, 1.3% in group B. Patient's satisfaction and quality of life was high in both groups without significant difference.

Conclusion: Wound repair using tissue glue is a good wound treatment option if compared with suture. Herewith, we would like to introduce our ongoing study, present the first results and give recommendation for optimizing the wound repair management of traumatic skin lacerations in children.

Disclosure of Interest: None declared

054

URACHAL REMNANTS – IS SURGERY INDICATED IN ANY CASE?

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Introduction: Urachal remnants vary in form and in symptoms and may even remain asymptomatic. We reviewed our experience with urachus remnants to discuss the diagnosis and the management.

Methods: We reviewed retrospectively all patients presenting with urachal remnants from January 2012 until December 2017 in our institution. The demographic data, symptoms, urachal forms, diagnostic modalities and management were evaluated.

Results: We identified 22 patients diagnosed with urachal remnants (UR). The mean age of the patients was 3.5 years (0–16 years). UR varied from urachal cysts (n = 12), patent urachus (n = 8), urachal sinus (n=1) to urachal diverticulum (n = 1). Eighteen patients (82%) presented with symptoms of which an acute infection was the most common symptom (n = 12) and clear umbilical drainage (n = 4). All UR were diagnosed by sonography. Surgery was performed in 14 patients (64%), including 4 patients (18%) whom were asymptomatic. In 4 patients UR were excised laparoscopically (29%), nine patients underwent an umbilical revision (64%) and one UR was excised during another surgery. In 8 patients (36%) the UR resolved spontaneously.

Conclusion: In a subset of patients UR are expected to resolve, especially after an acute infection or in the first year of life. We believe that UR can be observed and surgery is an option for recurrent symptoms. The malignant transformation in adulthood is to be determined in long-term follow-ups and needs to be discussed.

Disclosure of Interest: None declared

VASCULAR MALFORMATIONS IN PEDIATRIC HANDS: INTERDISCIPLINARY APPROACH AND SURGICAL CONSIDERATIONS

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Introduction: The evaluation of pediatric vascular malformations is optimally managed via an interdisciplinary approach, which involves dermatologists, radiologists, interventional radiologists and surgeons. A distinct clinical presentation as well as objective evaluations (such as ultrasound studies and MRI) not only aid in the diagnosis (hemangioma vs. vascular malformation) but also define the extent of the lesion (local or systemic). In general, treatment is only recommended in symptomatic patients, those with functional impairment, or cosmetic disfigurement. Conservative medical management is considered a first-line approach for the treatment of vascular malformations. However, interventional treatment options (such as sclerosis/embolization or interventional laser therapy) may be considered. Surgical intervention using open conventional surgical dissection and/or dissection with Sonopet Ultrasonic Aspirator are also definitive treatment modalities.

Methods: A retrospective chart review was undertaken utilizing the electronic medical records database at the University Children's Hospital Zurich. A search was conducted for all patients with a diagnosis of a vascular malformation of the hand who were surgically treated in the last 15 months. 6 patients met the aforementioned criteria and their data was included herein.

Results: Between November 2016 and November 2017, a total of 6 children with vascular malformations of the hand were surgically treated at our hospital. Of those, 3 (50%) were female and 3 (50%) were male. The mean age at time of surgery was 9 years (Range: 3 2/12 – 15 7/12 years). Indications for surgery were: pain (5/6 patients) with concomitant impaired hand function (50%). One patient's indication was based on appearance and to reduce psychological stigma (bullying). 5 of the malformations were classified by histopathology as venous malformations, and 1 as a microcystic lymphatic malformation. All procedures for venous malformations were done by conventional surgical dissection plus dissection with Sonopet. Postoperatively, no patients suffered from any serious adverse events related to the procedure. Most patients self-reported pain-free at follow-up (80%). All patients with preoperatively impaired function were found to have an improvement in function. The patient who suffered from social stigmatization reported an improvement in overall appearance.

Conclusion: Although these lesions may recur, surgical excision of vascular malformations usually reduces pain and improves function.

Disclosure of Interest: None declared

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KNOWLEDGE ASSESSMENT OF SPORTS-RELATED CONCUSSION IN CHILDREN AMONG HEALTHCARE PROVIDERS

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Introduction: The aim of this study is to assess the knowledge of paediatric concussion among pediatricians, pediatric surgeons and pediatric nurse practitioners.

Methods: a working group developed a 33-item anonymous survey incorporating four domains: concussion general knowledge, concussion diagnosis, concussion investigations and concussion management. The survey was distributed by e-mail to university (2) and regional (8) hospitals with a pediatric emergency level 1 primary care centre in the French speaking part of Switzerland. The target participants were pediatricians and pediatric surgeons with emergency department activities and pediatric nurse practitioners.

Results: The survey was fully completed by 277 healthcare providers. Of the respondents, 97% recognized symptoms truly related to acute concussion and 73.2% correctly identified general statements about concussion knowledge. However most did not identify symptoms not related to concussion ("red herring symptoms"). Respondents showed a good knowledge of return-to-play and return-to-learn protocols but both physicians and nurses demonstrated a lack of knowledge about the role of feedback from school and sporting staff in the post-acute period. Moreover knowledge about specific neuropsychological tests, such as SCAT, ImPACT or ACE was lacking and most participants requested unnecessary additional imaging (72% brain MRI; 70% CT scan; 45% skull X-ray).

Conclusion: While most primary care providers demonstrated sufficient knowledge regarding concussion, considerable gaps exist regarding symptoms, investigation and management. This study highlights the need for healthcare providers to improve knowledge

about established guidelines in order to enhance concussion management and prevention.

NaCHwuchs Prize: I wish to apply
Disclosure of Interest: None declared

ORAL PRESENTATIONS SSP 3

O58

DELAYED DECREASE IN SPO₂ VALUES IN INFANTS WITH BRONCHIOLITIS: EMERGENCY DEPARTMENT (ED) OBSERVATION PERIOD FOR SAFE DISCHARGE – PRELIMINARY RESULTS

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Introduction: Studies are lacking to evaluate the time necessary to a safe ED discharge without risk of desaturation in patients with bronchiolitis. In a retrospective study we showed that by using a threshold of 11 hours of ED observation period for patients ≥3 months old and of 25 hours for patients <3 months we were able to detect 95% of the patients with bronchiolitis at risk of delayed oxygen desaturation. We aimed to confirm these results with a prospective study

Methods: Prospective clinical trial in children aged <1 year admitted to the Pediatric ED with bronchiolitis during two RSV seasons. Patients included in our study received the standard management, except that they were provided with a portable oximeter to continuously record SpO₂ for 36 hours after admission

Graphic 1: Median time until desaturation according to age in both retrospective and prospective studies

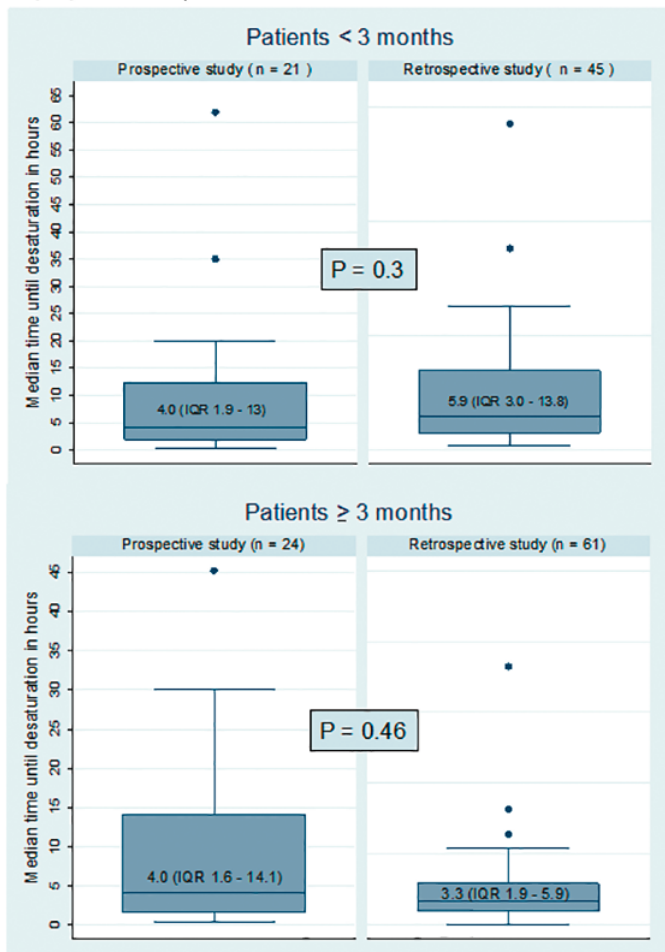


Figure 1

Table 1: Characteristics of study patients and outcome according to age.

	<3 months (n = 35)	≥3 months (n = 34)	P
Female sex %	54.3	44.1	0.398
RSV+ status, %	88.6	82.3	0.736
Duration of symptoms at presentation, median(IQR),days	4 (3–4)	4 (3–7)	0.224
ED observation time, median (IQR) hours	4.6 (3.1–6.8)	4.8 (2.9–7)	0.852
Respiratory Rate above normal range for age,% *	73.5	69.7	0.728
Moderate/severe retractions, %	58.8	76.5	0.120
SpO ₂ at arrival, mean (IQR)	97 (94–99)	97(94–98)	0.667
Time to desaturation, median (IQR) hours *	4 (1.9–12.4)	4(1.6–14.1)	0.918
Hospitalized %	97.1	91.2	0.289
Length of hospital stay, median (IQR), days	4.6 (2.7–6.1)	4.5 (3.7–6.9)	0.541
Required NGT, %	62.9	58.8	0.731
Required oxygen therapy,%	62.9	70.6	0.496

NGT nasogastric tube, IQR interquartile range, SpO₂ pulse oxygen saturation, RSV respiratory syncytial virus

* Normal values for age, groups: 0–1.9 months of age, 45 breaths/min; 2–5.9 months, 43 breaths/min; 6–11.9 months, 40 breaths/min.
 *n = 45

Results: Of 69 patients enrolled (34F:35M), 65 were hospitalized and 4 were discharged home with the oximeter. The characteristics of the patients, the initial presentation, and the outcomes were similar in patients < and ≥3 months (table 1). In 95% of patients with a delayed desaturation this decrease occurred within 35 hours for patients <3 months and within 30 hours for patients ≥3 months. Although these values were higher than those found in the retrospective study, the median time to desaturation was similar in both studies (graph 1).
Conclusion: Based on these preliminary results, the median time until desaturation for patients < and ≥ 3 months were similar from those found in our retrospective study. We will continue to include more patients to validate our findings

Disclosure of Interest: None declared

O59

IMPACT OF MATERNAL POSTPARTUM DEPRESSION AND POSTTRAUMATIC STRESS SYMPTOMS ON COGNITIVE FLEXIBILITY OF VERY PRETERM PRESCHOOLERS

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Introduction: An important proportion of mothers of preterm children (27%>40%) suffer from postpartum depression (PPD) or from posttraumatic stress disorder (PTSD, 26%>41%). Very preterm (VP) children (<32 gestational week (GW)) are at risk of problems in executive functioning and emotion regulation that is related to cognitive flexibility, which have been shown to predict professional and personal life trajectories. Research in healthy, full-term infants has shown that maternal mental health is associated with executive and emotional functioning of full-term children. However, research in VP infants is missing. This study aimed to investigate relationships between maternal PPD and PTSD symptoms and cognitive flexibility of VP preschoolers aged 3.5 years and to explore risk factors related to altered cognitive flexibility in VP children.

Methods: This is a study nested in a randomised controlled trial of an expressive writing intervention to prevent PTSD for a cohort of mothers of VP infants. The sample consisted of 55 dyads of mothers and their children born very prematurely ($M = 30.2.6$ GW, $SD = 2.21$ GW) and/or with a birth weight <1500 g ($M = 1120.37$, $SD = 318.97$). At 3 months corrected age, maternal PPD was assessed using the Edinburgh Postnatal Depression Scale (EPDS) and PTSD the Perinatal PTSD Questionnaire (PPQ). At 41 months ($M = 40.56$, $SD = 8.73$), flexibility index (FI) of the Behavior Rating Inventory of Executive Function – Preschool Version to evaluate preschooler’s cognitive flexibility was completed by mothers, and health-related information was retrieved from medical records.

Results: A significant association was shown between FI ($M = 48.09$, $SD = 8.04$) and EPDS ($M = 6.88$, $SD = 4.46$, $r = .26$, $p = .030$), whereas no significant correlation appeared with PPQ. EPDS, bronchopulmonary dysplasia (BPD), and Largo score predicted 22.1% of cognitive flexibility.

Conclusion: VP preschoolers’ emotion regulation was negatively correlated with maternal PPD symptoms, while no significant link emerged with PTSD symptoms. Medical risk factors, PPD, BPD and a low socioeconomic status predicted 22.1% of cognitive flexibility, which is linked to emotion regulation. Thus, it is important for specialists working with VP children to be aware of the increased vulnerability of children whose mothers are depressed. Early screening of maternal PPD would allow highlighting situations at increased risk of executive dysfunction and support families in the scaffolding of their VP child’s development.

Disclosure of Interest: None declared

O60

ADVERSE EVENTS AND ASSOCIATED FACTORS DURING INTRA-HOSPITAL NEONATAL TRANSPORTATION: A PROSPECTIVE OBSERVATIONAL STUDY

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Introduction: Hospitalized neonates often require intra-hospital transportation to perform diagnostic or therapeutic procedures that cannot be done at the bedside. A number of studies attest the risk of intra-hospital transportation of adults and children hospitalized in intensive care units, but literature concerning newborns is sparse. Our goals were to determine the incidence, the type and the severity of adverse events during intra-hospital transportation of newborns and to identify potential associated factors.

Methods: We conducted an observational prospective study from 1.6.2015 to 31.5.2016 in the neonatal intensive care unit (NICU) of the University Hospital of Lausanne, Switzerland. All newborns hospitalized in the NICU and undergoing intra-hospital transportation were included. Emergency admission transports from external hospitals and delivery room were not included.

Results: One hundred and thirty eight newborns of a median gestational age of 37 weeks (Q1–Q3 30–39 weeks) and of a median birth weight of 2470g (Q1–Q3 1296–3200 g) underwent 429 intra-hospital transports. Reasons for transport included 130 MRIs (30%), 98 surgeries (23%), 65 ultrasounds (15%), 42 endoscopies (10%), 20 CT scans (5%), and 74 other diagnostic procedures. One hundred and three adverse events occurred during 79 (18.4%) transports, including 24 (30%) desaturations, 22 (28%) agitations, and 20 (25%) episodes of hypothermia. No adverse event was moderate, severe or led to death. Factors associated with complicated transports included low gestational age and birthweight, underlying cardiovascular disease or symptoms requiring transport, the use of morphine, mechanical ventilation, return transports, time out of the NICU and transports from surgery and bronchoscopy rooms.

Conclusion: This study reveals that neonatal intrahospital transportations are at risk of frequent low severity critical events. This should raise awareness among transport staff, but also among staff attending diagnostic and therapeutic procedures outside the NICU. Absence of severe complications indicates that newborns can be safely transported within the hospital.

Disclosure of Interest: None declared

REPAIRED BUT NOT CURED: UNDERPERFORMANCE OF CHILDREN WITH CONGENITAL HEART DISEASE (CHD) AT CARDIOPULMONARY EXERCISE TESTING

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Introduction: Cardiopulmonary exercise testing (CPET) is an important clinical tool to assess the physical capacity of children and to observe cardiopulmonary adaptation to exercise. Several tests and protocols can be used, but there is currently no consensus and standardized protocols. The purpose of this study is: 1) to analyze the tests and protocols used and the percentage of tests having reached the maximum effort; 2) to compare the cardiopulmonary adaptation of children with and without cardiovascular intervention; and 3) to propose an algorithm based on our data and the literature to select the best protocol according to the clinical indication and patient status.

Methods: Patients under 18 years old who had a CPET at the pediatric cardiology clinic of the CHUV between 2013 and 2015 were included and grouped in two categories: children who had one or more cardiovascular interventions (INTERV group) and children who had no intervention (NO_INTERV group). P Values <0.05 are considered significant.

Results: Among the 632 CPET, 74% were performed on a treadmill with the Bruce protocol and 26% on a cycle ergometer with the Godfrey protocol. A maximum effort, defined as reaching 85% of the theoretical maximum heart rate (HR), was achieved in 83%, 85% and 77% of all CPET, of all Treadmill-Bruce protocols and of all Cyclergometer-Godfrey protocols, respectively. With the Treadmill-Bruce protocol, INTERV group has a significantly lower maximal HR (183.5 vs 197), maximal metabolic equivalent (13.85 vs 15.4), duration of the exercise (12.25 vs 13.06) and VO2max (42.7 vs 48.95); with the Cyclergometer-Godfrey protocol, INTERV group has a significantly lower maximum HR (178 vs 187), maximal metabolic equivalent (8.55 vs 10.7) and VO2max (36.6 vs 40.6). An algorithm to select the best test is provided (fig. 1).

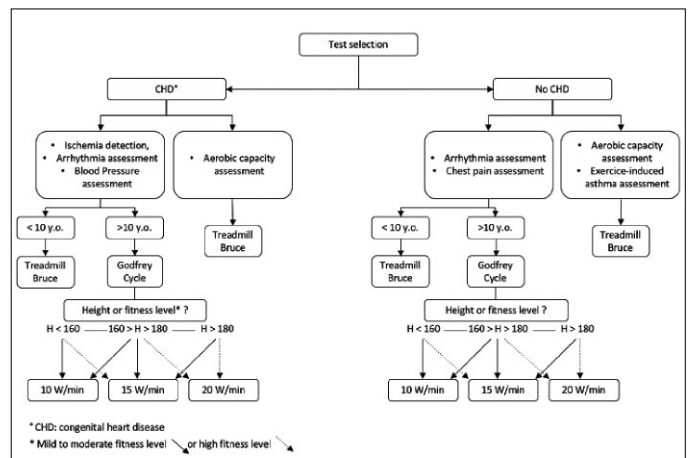


Figure 1

Matching the ergometer with the desired information.

Condition or question	Preferred ergometer	Rationale
Aortic stenosis/insufficiency (unrepaired/repaired) - Repaired transposition of great arteries - Coronary artery anomaly, (repaired/unrepaired)	Cycle	Ischaemia detection on ECG easier because there is less motion artifact
Repaired tetralogy of Fallot	Cycle	Arrhythmia assessment during exercise easier
Arrhythmia assessment/ Long QT syndrome	Cycle	Arrhythmia detection and measurement of QT interval on ECG easier
Coarctation of the aorta (unrepaired/repaired)	Cycle	Blood pressure assessment more accurate
(Functional) single ventricle	Cycle/treadmill	Depends upon query , ischaemia and arrhythmia or aerobic capacity assessment
Exercise-induced asthma/bronchospasm/chest pain	Treadmill	Running more likely to induce symptoms than cycling
Aerobic capacity	Treadmill	Higher VO2max with treadmill than with cycle
Assessment of rate responsive pacemakers	Treadmill	Better activation of accelerometer-based sensors

Figure 2

Conclusion: Our most commonly used protocol is the Treadmill-Bruce protocol, which allows to reach the maximal effort in a greater percentage of patients. However, the protocol that is likely to give the most useful information or to answer the questions of the referring physician should be selected: a cycle ergometry in most instances for children with CHD (figure 2). Our algorithm for test and protocol election should improve this aspect. We established that the INTERV group has a lower physical and aerobic capacity. Further studies should determine if this results from residual cardiac damage or a lower level of fitness due to the lack of training.

Disclosure of Interest: None declared

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SCREENING FOR OBSTRUCTIVE SLEEP APNEA IN CHILDREN

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Introduction: Obstructive sleep apnea-hypopnea syndrome (OSAS) affects about 2-4% children, especially between 2 to 8 years old. Symptoms for OSAS which may be reported are frequent snoring, observed apneas, mouth breathing and fragmented sleep. Untreated OSAS leads to significant neurocognitive and neurobehavioral dysfunctions, metabolic deregulations and cardiovascular dysfunctions. Many questionnaires have been developed to predict OSAS in children; however, none of them were satisfactory until now. We aimed to test the adequacy of a new questionnaire, called Obstructive Airway Child test (OACT), compared to home respiratory polygraphy (HRP) in a selected population.

Methods: This is a cross-sectional study held in our department. We recruited 45 subjects, aged between 5- to 16 years-old, with suspected OSAS. All of them responded to the OACT questionnaire, composed of 12 questions, and performed an HRP. Children with an Apnea Hypopnea Index (AHI) $\geq 5/h$ were diagnosed with moderate or severe OSAS.

Results: The sensitivity and specificity of the OACT questionnaire for moderate OSAS were not very good (71% and 34%, respectively). Therefore, we built a modified version (named Short Obstructive Apnea Pediatric Questionnaire or SOAP-Q) including the 5 most statistically significant items. This new SOAP-Q was more accurate with a sensitivity of 71.4% and specificity of 76.3%. The ROC curve improved from 0.750 to 0.835. The new questionnaire correctly diagnosed moderate OSAS in 71% of children

Conclusion: The OACT questionnaire is not reliable in our population. However, a modified version of the questionnaire showed better sensitivity and specificity, and correctly diagnosis 71% of children with moderate OSAS. A score compatible with moderate OSAS should urge the practitioner to refer for a HRP in order to start medical therapeutic management.

Disclosure of Interest: None declared

063

«SANTÉ ET MOUVEMENT» PHYSICAL RECONDITIONING PROGRAM FOR CHILDREN WITH CHRONIC CONDITIONS

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Introduction: Cardiovascular diseases (CVD) are the leading cause of morbidity and mortality in developing countries, in consequence of the formation of atherosclerosis lesions. Atherosclerosis is known to begin as early as in the first decade when risk factors, such as hypertension, dyslipidemia, diabetes, obesity, smoke, hereditary or lack of physical activity, are already present. Some children, especially those who are suffering from chronic conditions, cumulates several of those risk factors and are therefore at higher risk for premature CVD. It will concern an increasing number of future adults, as the prevalence of chronic conditions in children is constantly increasing. Between 1988 and 2006, their prevalence in children, aged from 8 to 14 years

old, has increased from 11.2 to 26.6% in the US. It concerned 12.5% of Swiss children in 2010. Chronic diseases share a common feature: most children are limited in their daily activities. Cardiorespiratory fitness (CRF) is dependant of the amount of physical activity and has been demonstrated to be closely linked to mortality. Deconditioning may play a major role in children with chronic diseases as they may lead relative sedentary lifestyles, in part imposed on them by physicians, teachers, and parents or by themselves.

Methods: Since June 2016, we implemented a physical reconditioning program for children with chronic conditions. Children in our program are suffering from congenital heart or pulmonary diseases or are under treatment for an oncologic condition. This program includes measures of global physical capacities, as well as pulmonary function and CRF at baseline and at the end of the program. The baseline data are used to propose an individually adapted physical activity program supervised by a trained physical activity teacher during 2 to 4 months.

Results: 63 children performed a cardiopulmonary exercise test. Among those children, 24 entered the program with 13 of them still ongoing. 26 additional children were seen by our physical activity teacher to receive advices for home and community based physical activities. Results will be presented in the following years.

Conclusion: Children with chronic condition are less active than healthy ones. With our physical reconditioning program we aim to improve their cardiorespiratory fitness and therefore their CVD risk factors; increase their confidence in their own capacities; help them to learn how to adapt their physical activity to their limitations and encourage those children to move more.

Disclosure of Interest: None declared

064

PERFORMANCE OF BLOOD PRESSURE MEASUREMENTS AT AN INITIAL SCREENING VISIT FOR THE DIAGNOSIS OF HYPERTENSION IN CHILDREN

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Introduction: Hypertension in children is defined as sustained elevated blood pressure (BP) over several visits. For the screening of hypertension, it is standard in clinical practice to obtain several BP readings at the initial visit if the first value is elevated. However, there is no recommendation on the minimum number of readings needed. We evaluated the performance of BP readings obtained at one initial screening visit to predict the diagnosis of hypertension in children.

Methods: In a school-based study conducted in Switzerland, BP was measured three times on up to three visits in 5207 children. Children were considered to have hypertension if the mean of the last two BP readings was elevated in all three visits. Sensitivity, specificity, negative predictive value (NPV), and positive predictive value (PPV) of elevated BP at the initial screening visit for the identification of hypertension were estimated using the 1st (R1), the 2nd (R2), and the 3rd BP readings (R3), as well as (R1+R2)/2 and (R1+R2+R3)/3. These performance indices were compared with (R2+R3)/2 used as the reference method. The ability of BP readings at the first visit to discriminate children with and without hypertension was further evaluated with receiver operating characteristic (ROC) curve analysis.

Results: The prevalence of systolic/diastolic hypertension was 2.2%. The greatest performance to identify children with systolic/diastolic hypertension was obtained with R2 (sensitivity: 97%; specificity: 88%; PPV: 15%; NPV: 100%) and the average of R2 and R3, the reference method (sensitivity: 100%; specificity: 90%; PPV: 18%; NPV: 100%). With R1 and the average of R1 and R2, specificity and PPV were lower. The AUC using R1, R2, (R1+R2)/2, and the reference method (R2+R3)/2 were high for the identification of hypertension (0.90, 0.93, 0.92 and 0.95, respectively).

Conclusion: Using solely the second BP reading at the initial screening visit to identify children at risk of hypertension had a similar performance compared to the reference method, that is, the average of the second and the third readings. Obtaining two BP readings at an initial screening visit may be sufficient to screen hypertension.

Disclosure of Interest: None declared

O65

EFFECTS OF PREMATURITY ON MATERNAL AND CHILD ATTACHMENT

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Introduction: Premature birth affects both parents and children, especially in their socio-emotional functioning (Treyvaud et al., 2012). Among the socio-emotional difficulties, premature children as well as their parents show more insecure attachment representations than full-term born children and their parents (Borghini et al., 2006). Nevertheless, studies haven't examined attachment quality in premature-born preadolescents, nor the long-term link between parent and child attachment. In this study, we investigate whether at 11 years, premature children have a less secure attachment than their full-term born peers and whether there is a link between maternal attachment during perinatal period and child attachment ten years later.

Methods: From the cohort described by Borghini et al. (2006), 31 premature children ($M_{AG} = 30$ weeks) and 20 full-term born children ($M_{AG} = 40$ weeks) as well as their mothers participated in the study. Attachment style (secure vs. insecure) was assessed in mothers at 18 months (corrected age) with the Working Model of the Child Interview and in children at 11 years of age with the Friends and Family Interview.

Results: At 11 years, premature children had more insecure attachment representations (81%) than full-term born children (55%; $\chi^2(1) = 3.85, p \leq .05$) – which is similar to the pattern found in mothers at 18 months (preterm infants: 77%, full-term born: 40%; $\chi^2(1) = 7.28, p = .007$; Borghini et al., 2006). Comparing maternal attachment at 18 months and child attachment at 11 years, we found a link in 65% of premature children and in 45% of the full-term born children. Importantly, this link is especially pronounced for insecure attachment representations: in 79% of the preterm sample both maternal and child attachment was insecure compared to 14% in the full-term sample (z -test = 3.2, $p = .0016$).

Conclusion: Results show that prematurity affects child attachment up to preadolescence. More specifically, maternal insecure attachment representations were also found in the child, especially in premature-born children. Taken together, our results suggest that attachment problems in mothers and in their premature children add up to the socio-emotional problems described in the literature. Considering the strong link between parental representations of insecure attachment and these of their premature children, it is important to intervene with parents in order to prevent attachment difficulties and, more generally, socio-emotional problems related to prematurity.

Disclosure of Interest: None declared

O66

STRESS REGULATION EXERCISES IMPROVE CRAVING AND DISORDERED EATING IN ADOLESCENTS: RESULTS OF AN INFORMATION-TECHNOLOGY (IT) SUPPORTED INTERVENTION*

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Introduction: Reduction of stress is a worthwhile approach to overcome eating problems and improve BMI in obese youth. The purpose of this study is to test the feasibility and effects of an IT supported biofeedback home training, to reduce stress, improve well-being and change eating habits.

Methods: In a prospective pilot study 8 overweight adolescents (BMI 27–42 kg/m², age 14–18 y.) performed home relaxation exercises, with individualized food stimuli twice a day. Additionally all subjects received a lifestyle therapy (nutrition counseling, physical activity training, and psychoeducation). BMI and acute stress levels measured by serum cortisol, skin conduction and Self-Assessment Manikin SAMS as well as chronic stress levels by Trier Stress Inventory TICS) were assessed at start (T0), and after 4, 7 and 10 weeks (T1, T2, T3 resp.). During the intervention all subjects are encouraged to practice their exercises and to report well-being (KIDSCREEN) via SMS coach.

Results: Response rate to SMS guided home exercises was 90.3%. From T0 to T3, BMI and BMI-SDS decreased significantly in the overall study group ($p = 0.055$ or $p = 0.023$); Of the stress parameters, the decreases in the TICS and SAMS were not statistically significant. During each biofeedback session, serum cortisol decreased significantly ($p = 0.031$). Eating habits improved significantly, both in emotional eating (FEV-Path scale) ($p = 0.047$) and in the desire for overeating/craving (FUN scale) ($p = 0.039$).

Conclusion: The extraordinary adherence of obese adolescents to SMS guided home exercises make this kind of health care a valid tool for weight management programs at distance in this critical age group. This pilot study shows that a treatment including four sessions of electrodermal biofeedback and daily SMS-assisted exercises with stimulus picture is well accepted by adolescents. Further studies have to show, how stress reduction and improvements in eating habits are related to the observed decrease in BMI-SDS. Ongoing research has to examine longterm effects of biofeedback therapy as well as the implementation in a clinical setting.

NaChwuchs Prize: I wish to apply
Disclosure of Interest: None declared

O67

OMEGA-3 FATTY ACIDS IN CHILD AND ADOLESCENT MENTAL HEALTH – MYTHS AND FACTS

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Introduction: A growing body of literature suggests that omega-3 fatty acids supports brain development from early age onwards. Randomized placebo-controlled trials (RCTs) have been performed in a range of child and adolescent psychiatric disorders ranging from pervasive developmental disorders, motor coordination disorders, dyslexia, attention deficit and hyperactivity disorders, major depressive disorders, at risk mental states for psychotic disorders, oppositional defiant disorders and others.

Methods: A qualitative review of the literature of randomized controlled omega-3 fatty acid supplementation trials was performed based on previous meta-analysis and recently published RCTs.

Results: The available data is very heterogenous and despite a range of high quality meta-analysis no clear conclusions could be drawn if omega-3 fatty acids will have a place in pediatric mental health care or not. Several reasons could be identified, such as the ratio between Eicosapentaenoic acid (EPA) and Docosahexaenoic acid (DHA), the total daily dose of Omega-3 fatty acids, the type of condition (e.g. depressive symptoms in MDD versus non-MDD), monotherapy versus augmentation therapy, and others.

Conclusion: There are currently three major areas of child mental health where several omega-3 fatty acid supplementation trials provide some evidence that omega-3 fatty acids may be superior over placebo: pediatric major depressive disorders, at risk mental state for psychosis and pediatric ADHD. Limitations of current RCTs will be presented and discussed.

Disclosure of Interest: None declared

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STRESS REGULATION EXERCISES IMPROVE CRAVING AND DISORDERED EATING IN ADOLESCENTS: RESULTS OF AN INFORMATION-TECHNOLOGY (IT) SUPPORTED INTERVENTION*

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¹Ostschweizer Kinderspital, St. Gallen, Switzerland

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Conclusion: The extraordinary adherence of obese adolescents to SMS guided home exercises make this kind of health care a valid tool for weight management programs at distance in this critical age group. This pilot study shows that a treatment including four sessions of electrodermal biofeedback and daily SMS-assisted exercises with stimulus picture is well accepted by adolescents. Further studies have to show, how stress reduction and improvements in eating habits are related to the observed decrease in BMI-SDS. Ongoing research has to examine longterm effects of biofeedback therapy as well as the implementation in a clinical setting.

NaCHwuchs Prize: I wish to apply
Disclosure of Interest: None declared

O69

GROWING UP ON AN AUTISTIC SPECTRUM DISORDER (ASD)

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Introduction: Autistic patients usually have reduced and/or delayed autonomy in their development. Yet there are many other patients who have a hard time leaving childhood and entering adulthood. Some "eternal adolescents" have reduced working capacities, anxiety disorders, suicidal behaviours, emotion regulation disorders or psychotic features. Could some of them actually be undiagnosed patients with autistic traits or Asperger's syndrome? What is actually Asperger's syndrome? After a journey through its – history – nosology – epidemiology – co-morbidities and differential diagnosis (ADHD, HIP, PTSD, BPD...) – review of neuroscientific research We will explain the actual hypothetic core symptoms and concepts of 1- sensitivity disorder 2- emotion regulation 3- theory of mind 4- central coherence. We will then end with the measures and treatments that can help in case of autistic traits or Asperger's syndrome. These themes will be illustrated by a few clinical stories and videos of consultations and it will be showed in which way core symptoms of ASD and treatments have an impact on autonomy and growing up.

Conclusion: In conclusion: – Yes, autistic traits and Asperger's syndrome are underdiagnosed and can result in many different symptoms, including struggles with growing up. – Yes, recognizing it and taking measures according to it is crucial for our patients and our society.

I have read and understand application rules: No
Disclosure of Interest: None declared

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SERUM ZINC LEVELS IN PATIENTS WITH EATING DISORDERS

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POSTERS

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NEW MUTATION IN GLYCOGEN STORAGE DISEASE TYPE VI

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Introduction: Eating disorders (ED) are associated with severe long-term medical and hormonal consequences. Evidence from animal studies, in particular involving rats has investigated many aspects of starvation induced by Zinc (Zn) deficiency. There is some evidence for Zn deficiency being intimately involved in anorexia nervosa (AN), with Zn deficiency if not acting as an initiating cause playing an accelerating or even exacerbating role thus deepening related pathology. Findings suggesting that Zn deficiency and resulting decreased serum levels are associated with the pathophysiology and malnutrition found in AN.

Methods: Serum Zn concentrations were obtained from healthy controls and from acutely ill and remitted young patients with AN and Bulimia nervosa (BN).

Results: Zn concentrations were higher in recovered subjects with AN when compared with acutely ill AN patients. Remitted and acutely ill patients with BN even had elevated serum Zn levels compared with controls.

Conclusion: In conclusion we found that Zn status is changed in patients with ED. Surprisingly zinc serum concentrations are elevated in recovered ED patients and not decreased in acutely ill patients. The reasons for that are unknown. This may due to a special kind of eating behavior and choice of food preferences in patients with ED. An association between Zn levels and different hormonal parameters is discussed.

Disclosure of Interest: None declared

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HEALTH-RELATED QUALITY OF LIFE AMONG ADOLESCENTS WITH ANOREXIA NERVOSA

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Introduction: Eating disorders are a major cause of disease in adolescents. These chronic diseases don't just impact physical-well being but also social and psychological life. The purpose of the present study is to examine the impact of anorexia nervosa on health-related quality of life (HRQOL).

Methods: The HRQOL in adolescents with anorexia has been measured with the KINDL[®] questionnaire. The control population (N = 52) was recruited at the Lausanne children hospital and the cases (N = 26) were recruited at the sub-division for adolescents at the Lausanne hospital. The six sub-scales in the KINDL[®] HRQOL correspond to the physical well-being, emotional well-being, self-esteem, family, friends and school. In addition to those six questions, a disease module is added for the cases group. Further more important variables in the file have been asked, sex, age, size and weight, social economic status, familial environment and school.

Results: Adolescents with anorexia disorder have a poorer quality of life (QOL) than the standard population (64.8 ± 11.54 versus 74.98 ± 7.84; p < 0.001). QOL is significantly lower for the following sub-scale: emotional well-being, self-esteem and friends. Lower weight is not associated to lower QOL.

Conclusion: Our finding demonstrates the association between anorexia and QOL. However, Body Mass Index wouldn't be associated with a lower quality of life. The disease itself would be responsible for this low QOL.

Disclosure of Interest: None declared

Methods: A twelve-months old girl presented with hepatomegaly and growth retardation. Biochemical analysis showed elevated liver transaminases, high cholestasis parameters, hypertriglyceridemia, ketosis and hyperlactatemia. The diurnal glucose profile revealed various episodes of hypoglycemia (during daytime as well as during nighttime). Assuming glycogen storage disease, molecular genetic testing was performed.

Results: Exome sequencing revealed a homozygous mutation in exon 1 of the *PYGL* gene (c.43A>G; p.(Ser15Gly)). Based on the confirmed glycogen storage disease type VI the girl was treated accordingly with frequent small meals as well as formula milk via gastrostomy tube (mainly during nights). Due to frequently recurrent hypoglycemic episodes and missing normalization of liver transaminases, cornstarch was added later. This regimen led to improvement of laboratory findings, hepatomegaly and growth and no further hypoglycemic episodes. The described c.43A>G p.(Ser15Gly) mutation in the *PYGL* gene is a novel mutation. However, similar mutations in this gene account for the glycogen storage disease type VI. In addition, this mutation affects a highly conserved residue and is predicted pathogenic in six in-silico-programs supporting its disease-causing role for glycogen storage disease type VI. Further, specific treatment improved most of the clinical and biochemical symptoms in this patient.

Conclusion: The described c.43A>G p.(Ser15Gly) mutation in the *PYGL* gene is novel; its presence in homozygosity indicates an earlier common ancestry in the supposedly unrelated parents. This mutation is the likely cause of glycogen storage disease type VI. Specific treatment, immediately initiated after the genetic diagnosis, improved the patient's situation. Multidisciplinary team approach and collaboration in the metabolic network were helpful in establishing timely diagnosis and adequate treatment.

Disclosure of Interest: None declared

P002

AN UNUSUAL EVOLUTION OF A FRIEDREICH ATAXIA-ASSOCIATED DIABETES MELLITUS

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Introduction: Friedreich ataxia (FRDA) is the most common hereditary ataxia. It is a progressive neurodegenerative disorder, caused by a genetic mutation inserting a GAA repeat expansion within intron 1 of the *FXN* gene. This leads to a lack of the protein frataxin and a mitochondrial dysfunction. FRDA patients have an increased risk of developing a non-autoimmun, mitochondrial form of diabetes mellitus. Both insulin resistance and deficiency play a role in the manifestation of diabetes in FRDA. The onset of diabetes is often acute. No remission is expected.

Methods: Case description: A 17-year-old adolescent boy with Friedreich ataxia, wheelchair-bound presented with polydipsia and polyuria since two weeks, with a blood glucose level of 31 mmol/l and an HbA1c of 10.7% without diabetic ketoacidosis and negative Type 1 Diabetes specific autoantibodies. An augmented insulin pump therapy, combining an insulin pump and continuous glucose monitoring, was introduced due to the patient's inability to inject insulin with a pen and to use a conventional glucose monitoring device. Insulin requirement at the beginning was 0.65 U/kg body weight per day. In the following months insulin requirement decreased quickly. At that time the mother introduced an oral treatment with vitamin B1 and kurkuma. Five months after the onset of diabetes, insulin therapy could be stopped for more than 6 months. Due to higher blood glucose levels and an increasing HbA1c, insulin therapy was restarted thereafter with a low insulin requirement of 0.2 U/kg body weight per day and a measurable C-peptide level more than one year after the onset.

Results: –

Conclusion: Discussion: FRDA is associated with a higher prevalence of diabetes mellitus. The onset of diabetes is often acute, ketoacidosis has been reported as the presenting symptom of diabetes in FRDA. No remission is expected. This seems to be the first report of a Friedreich ataxia-associated Diabetes mellitus with a complete remission for more than 6 months. It remains unclear whether the introduction of vitamin B1 and kurkuma influenced the insulin requirement. There exists some data about a favourable response to thiamine in motor symptoms in FRDA patients but not concerning the influence on glucose metabolism. Further research is required to determine the influence of vitamin B1 and kurkuma on glucose metabolism in patients with Friedreich ataxia-associated diabetes mellitus.

Disclosure of Interest: None declared

QUALITY OF LIFE IN PEDIATRIC PATIENTS WITH INBORN ERRORS OF METABOLISM AND THEIR PARENTS

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Introduction: Inborn errors of metabolism (IEM) are a group of rare genetic diseases caused by enzyme deficiencies that lead to intoxication, pathological storage and/or energy failure. Because of heightened risks for severe non-reversible physical and/or psychological sequelae, IEM require systematic medical monitoring along with a restrictive diet and/or drug treatment in a majority of these disorders. Such constraints likely decrease (health-related) quality of life and, more generally, psychosocial functioning of patients with an IEM and their families. However, this topic remains largely unexamined; the few existing studies focused on a given type of disorders (e.g., galactosemia, Bosch et al., 2004; phenylketonuria, Landolt et al., 2002; Thimm et al., 2013) and their findings remain inconclusive. In this study, we aimed to assess the quality of life of pediatric patients with an IEM and of their parents. Furthermore, we asked whether there might be protective and/or risk factors that influence quality of life.

Methods: A group of pediatric patients aged 0 to 18 years (ongoing data collection) along with their parents was asked to complete a survey. Quality of life was assessed with the Phenylketonuria Quality of Life questionnaire (Bosch et al., 2015; Regnault et al., 2015), which targets practical, social and emotional life, diet and/or taking dietary supplements. Additionally, we examined child psychopathology with the Strengths and Difficulties Questionnaire (Ravens-Sieberer et al., 2008) along with parental anxiety/depression with the Hospital Anxiety and Depression Scale (Razavi et al., 1989). In order to account for protective and risk factors, we assessed parental health-related stress with the Pediatric Inventory for Parents (Streisand et al., 2001) and coping strategies with the Cognitive Emotion Regulation Questionnaire (Jermann et al., 2006). Importantly, to control for heterogeneity in IEM, physicians evaluated the severity of the disorder by filling out the biological domain scale from the Intermed evaluation of health service needs (Guex, 2001).

Conclusion: This ongoing study is expected to provide a comprehensive overview of how IEM affect the quality of life of pediatric patients and their parents. Moreover, this study will allow us to develop a finer understanding of the protective as well as risk factors influencing quality of life. We expect to present data from at least 50 patients.

Disclosure of Interest: None declared

P003

VULVAR EPITHELIAL INCLUSION CYST AS A COMPLICATION OF FEMALE GENITAL MUTILATION

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Introduction: Female genital mutilation (FGM) refers to the procedures that intentionally alter or cause injury to the female genital organs for non-medical reasons. The procedure has no health benefits for girls and women and is a violation of human rights. There are more than 200 million girls and women alive today who have been subjected to FGM. The procedure is performed in at least 30 countries mainly in western and northeastern Africa, the Middle East and Asia. In some of these countries up to 98% of women in reproductive age are affected. FGM is mostly carried out on young girls between infancy and age 15. Even though religious motifs are often used as a pretext, there are no religious scripts prescribing the practice. The reasons why FGM is performed vary over time and from one region to another and are sociocultural in nature. FGM is classified into 4 major types depending upon severity and method. The procedure has a broad range of immediate and long-term complications and interferes with the natural function of women's and girl's bodies. One of the well-known long-term complications is the evolution of a vulvar epithelial inclusion cyst occurring with an incidence of up to 5% depending on the type of FGM.

Methods: Case Report: An 11-year old girl from Eritrea was referred to our pediatric hospital for a gynecological consultation. She had a progressive mass in her vulvar area since 7 years. This mass disturbed the girl increasingly while urinating and was sensitive to pressure. The girl was a refugee from Eritrea and had been in Switzerland only for one month. Her father told us that the girl had been subjected to FGM as a toddler. On clinical examination there was a fluctuant mass of 6x4 cm at the apex of the vulva. It was painful on pressure. The clitoris and the labia minora had been removed but there was no infibulation. The findings were consistent with Type II FGM (WHO-Classification). The

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ultrasound exam showed a homogenous mass without perfusion. The suspected diagnosis was a vulvar epithelial inclusion cyst. According to the literature the mass was surgically removed and the suspected diagnosis was histologically confirmed. The girl could be discharged from the hospital the day after.

Conclusion: With increasing migration from countries where FGM is practiced more women and girls with FGM will present to our medical institutions. Therefore sensitization for FGM, its complications and possible preventive measures are necessary.

Disclosure of Interest: None declared

P005

BANANA FIBERS MASQUERADING AS WORMS IN THE STOOL OF A 13-MONTH-OLD BOY

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Introduction: When it comes to interpret filamentous objects in babies' stools, pediatricians generally tend to think of worms, but this is not always the case.

Methods: A previously healthy 13-month-old boy was presented to the emergency department with the complaints that he passed long black worms in the stool starting the prior day. On examination, the boy was playful, afebrile and with normal vital signs. His abdomen was non-tender and not distended. A stool examination for ova and parasite was negative.

Results: On taking a detailed dietary history, it was found that the "worms" appeared in the stool after feeding him several bananas. The family was reassured about the nature of the "worms".

Conclusion: The first description of banana fibers that mimic worms in stool was made more than 100 years ago (Ward HB. Precision in the determination of human parasites. JAMA 1903;16:703-9). Surprisingly, modern descriptions are rather uncommon. Misdiagnosing this phenomenon with intestinal helminthiasis might result in redundant investigations and unnecessary management, which can cause apprehension in the parents. Remarkably, existing textbooks do not refer to it.

Disclosure of Interest: None declared

P006

"SAME SAME BUT DIFFERENT" – TWO CASES OF PSEUDOHYPOPARATHYROIDISM

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Introduction: Pseudohypoparathyroidism is a group of rare disorders with heterogeneous genetic causes. It is characterised by end-organ resistance of parathyroid hormone (iPTH). Affected children present with hypocalcemia, hyperphosphatemia and elevated levels of iPTH. A vitamin D deficiency has to be excluded. Some forms of pseudohypoparathyroidism present with Albright's hereditary osteodystrophy, a clinical entity with heterogeneous clinical findings such as brachydactyly, round face, short stature and subcutaneous ossifications. Additional features in patients with pseudohypoparathyroidism type 1a may include obesity, intellectual impairment and resistance to several other hormones with G-protein coupled receptors like TSH, GHRH and calcitonin.

Methods: We present the clinical cases of two toddlers treated at the University Children's Hospital Bern who presented with obesity and subclinical hypothyroidism.

Results: Two toddlers were referred to the University Children's Hospital Bern for further investigations. Both children had obesity as well as subclinical hypothyroidism and laboratory results showed in both children elevated iPTH with normal/low calcium levels in the absence of significant vitamin D deficiency. In addition, the girl showed signs of Albright's osteodystrophy and showed a mutation in the GNAS-gene that had been previously described in pseudohypoparathyroidism type 1a. Her mother also had severe brachydactyly dig IV and V with inadequate high iPTH for the normal calcium level and showed the mutation. The boy, in contrast, showed no signs of Albright's osteodystrophy. The results of the genetic testing of the boy are pending.

Conclusion: We present two cases of a very rare and heterogeneous disease treated in our outpatient clinic with very similar clinical presentation. We think that it is important to consider even rare diseases in the diagnostic work-up of patients with common problems.

Disclosure of Interest: None declared

P007

A SYSTEMATIC REVIEW OF THE PREVALENCE OF OVERWEIGHT AND OBESITY IN SCHOOL-AGED CHILDREN IN LOWER-INCOME COUNTRIES

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Introduction: Children in lower-income countries increasingly face the co-existence of under and over nutrition, also called the double burden. The aim of our study was to provide a global view of the prevalence of overweight and obesity in children between 6 and 12 years old in low and lower-middle income countries, and the related extent of the double burden, in order to inform the development of prevention programs in these countries.

Methods: We followed the PRISMA guidelines to undertake a systematic review of the literature. The PUBMED and EMBASE databases (1st January 2006 to 31st December 2016) as well as the WHO and UNICEF websites were searched for references on the prevalence of overweight and obesity in children aged between 6 and 12 years old in low and lower-middle-income countries (as defined in the World Bank List of Economies, 2016). We also searched the reference lists of the selected articles. We summarized the prevalence data for overweight, obesity and undernutrition by country and by gender, and mapped out these data to identify countries with similar prevalence profiles.

Results: 21 articles met the inclusion criteria, providing prevalence data for 12 of 83 eligible low or lower-middle income countries. The prevalence of overweight and obesity in school-aged children was the highest in Nepal (25.9%), and the lowest in Vietnam (0.03%). We identified three distinct country profiles: (1) prevalence of overweight exceeding that of undernutrition (2) similar prevalence of overweight and undernutrition (3) prevalence of undernutrition exceeding that of overweight.

Conclusion: There were little publically available data on the prevalence of overweight and obesity in lower-income countries. The identification of three distinct country profiles in relation to the double burden could inform obesity prevention programs and favor synergies between countries with similar profiles.

Disclosure of Interest: None declared

P008

ACHIEVEMENT OF FERTILITY BY TREATMENT OF HYPOGONADOTROPIC HYPOGONADISM WITH RECOMBINANT FSH AND HCG IN AN ADOLESCENT

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Introduction: Congenital hypogonadotropic hypogonadism (CHH) results from absent secretion or action of gonadotropin-releasing hormone (GnRH). It is characterized by incomplete or absent puberty by the age of 18 years and can be associated with anosmia and various developmental abnormalities. CHH has a strong male predominance of 3-5 to 1 and results from mutation of more than 30 different genes that are transmitted in a X chromosome-linked recessive, autosomal recessive and dominant mode of inheritance. Standard treatment during childhood consists of puberty induction by testosterone or estradiol, thereby permitting pubertal changes and psychological development. Since this treatment does not result in activation of the resting gonads, it does not permit to induce testicular growth, nor induction of fertility. Classically, treatment with gonadotrophins is proposed during adulthood, when fertility is desired by the patient.

Methods: We report the case of an 11-year-old boy who presented at our outpatient clinic for evaluation of a micropenis. CHH was diagnosed and puberty was induced by a treatment of recombinant follicle stimulating hormone (rFSH) and recombinant human chorionic gonadotropin (hCG).

Results: Patient history revealed anosmia and physical examination confirmed a micropenis and absent pubertal development. CHH was diagnosed on the basis of a cerebral MRI that showed absent olfactory bulbs and of hormonal measurements showing low levels of gonadotrophins, testosterone and Inhibin B. Genetic analysis of 21 known CHH genes by next generation sequencing showed no

abnormalities. To induce testicular growth and fertility, instead of a treatment with testosterone, a treatment with rFSH was begun at the age of 13 years, at a dose of 37.5 IU sc, three times per week. The dose was increased every month to reach 150 IU. At that timepoint, hCG was added at a dose of 400 IU sc, twice per week. Two years after initiation of the treatment and at a final dose of 225 IU FSH and 400 IU hCG, a sperm analysis turned out to be satisfactory and permitted preservation of several specimens. Development of the external genital organs was normal, testes reached a volume of 9.5 ml, height reached family target at the age of 15 years, thanks to a satisfactory growth spurt during treatment.

Conclusion: Pubertal induction with the above-mentioned protocol is feasible without side effects and leads to satisfactory results in terms of pubertal changes, and above all permits to achieve fertility.

Disclosure of Interest: None declared

TRANSITION: FROM THE FIRST SWALLOWING TO THE FIRST ADULT MOLAR (ORAL PRESENTATION)

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Introduction: The milk dental occlusal balance, witness and relay in the general postural balance development: Daily concern for pediatricians in growth monitoring. Pediatric orthodontics doesn't really exist by lack of the link with medical part, first with pediatricians, but this gap is the field where I am concerned and searching – practicing from more than 40 years. The result of this experience is that we can understand now the reason for failure or relapse of 50% at least of regular orthodontic treatments: it is because of the lack of knowledge, of regard and of adapted therapeutic strategies for dysfunctions and minor neuromotor dyscoordinations during early sensorimotor steps (in particular within the early coordination between nose breathing, swallowing maturation and postural balance deficiencies.)

Methods: Starting with fundamental neuro-embryologic data from the –80's, we can present a new reading with the tools of neuroscience, across the complexity of the implementation of neuromotor orofacial loops. They also allow to explore their very tight synchronization with sensorimotor implementation of the determinants for crânio-cervical posture. In parallel, from my study of the early sensorimotor assessment of Bullinger (UNIL-2014), for 3 years, I am pursuing personal clinical research by collecting and comparing pré- néo- and post-natal data for sensorimotor development with tridimensional crânio-facial dysmorphia. (more than 150 cases until now).

Results: By this integrative and straightforward reading of the early and close coordination between the implementation of the first three-dimensional determinants of occlusal balance with those of postural balance, we can propose 3 daily practical tools for pediatricians: 1. A new and better understanding of the reinforcing auto-reverse interlinkages between nasal breathing – swallowing – milk dental occlusion – head neck posture and general balanced growth. 2. Daily tools to relate very early signs of occlusal imbalance in milk dentition with early postural control problems. 3. Relevancy of new early therapeutic strategies in pediatric orthodontics.

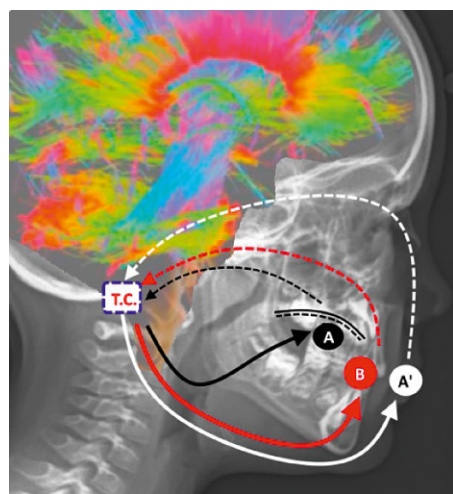


Figure 1

Conclusion: Today we can pretend that 50% around of severe orthodontic disbalances are related to early neonatal and sensorimotor troubles and to latter general posture or breathing troubles (OSA). Thank you for giving me this occasion to start to bridge the gap between dentistry and medicine.

Disclosure of Interest: None declared

P010

DIENCEPHALIC SYNDROME: A RARE BUT POTENTIALLY FATAL CAUSE OF FAILURE TO THRIVE

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Introduction: Diencephalic syndrome is a rare cause of failure to thrive and is associated with hypothalamic / optochiasmatic brain tumors. The clinical findings may be rather unspecific thus not immediately suggesting a tumor diagnosis.

Methods: Case report.

Results: Clinical case: A 5 month-old boy presented with conjugated nystagmus and failure to thrive since his 3rd month of life. Patient history revealed uneventful full term pregnancy and normal development until the age of 3 month when the parents noticed unusual eye movements, weight stagnation, vomiting and reduced appetite. Neuropediatric and ophthalmologic consultations were without signs of impaired visual acuity and compatible with congenital nystagmus. Vomiting ceased spontaneously and appetite normalized but failure to thrive persisted despite good appetite and hypercaloric diet by that time. Clinical work-up did not reveal any signs of malabsorption or maldigestion and extended examinations ruled out hepatopathy, nephropathy, cardiopathy, food intolerance, thyroid dysfunction and pathological intestinal passage. Upon admission the boy was in good general condition. Physical examination was normal except for conjugated nystagmus and cachexia (5.41 kg, <P3). Length and head circumference were within normal range and psychomotor milestones had been reached appropriately. Diencephalic syndrome was suspected and cerebral imaging was performed revealing a tumor in the optochiasmatic region. An open microsurgical biopsy confirmed a pilomyxoid astrocytoma. Endocrine work-up ruled out hormonal pituitary deficiency. Ophthalmologic re-evaluation showed impaired visual acuity. As total resection of the tumor was not feasible, chemotherapy Vincristine/Carboplatin) was administered. In addition, a percutaneous endoscopic gastrostomy tube was placed to ensure sufficient nutrition. Short term follow-up seems to be favorable with weight gain of 400 g within 2 months and well tolerated therapy. Evaluation by cerebral imaging is pending.

Conclusion: Diencephalic syndrome is a very rare cause of failure to thrive. Underlying pathophysiology is unclear. Clinical findings may be unspecific such as vomiting, hyperalertness, hyperkinesia and often nystagmus. The discrepancy between failure to gain weight despite adequate caloric intake and normal height and head circumference is characteristic. The presence of nystagmus or other symptoms of pre-genicular visual dysfunction in a child with failure to thrive should prompt timely cerebral imaging.

Disclosure of Interest: None declared

P011

NOCTURNAL SWEATING AND TACHYCARDIA AS A FIRST SIGN OF GLYCOGEN STORAGE DISEASE TYPE 1 (GSD-1)

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Introduction: Glycogen storage disease type 1 (GSD-1) is a rare inherited disorder of carbohydrate metabolism with an autosomal recessive pattern. Underlying is a defect of the enzyme glucose-6-phosphatase, which is playing a major role in regulating blood sugar levels. Therapy of GSD-1 consists of a strict diet with regular carbohydrate intake to prevent hypoglycemia. Hepatomegaly, lacticidosis and/ or hypoglycemic seizures in early infancy are often the first symptoms. Hypoglycemia in fasting periods may result in sympathoadrenal activation symptoms like sweating and tachycardia; and, unrecognized in coma and death.

Methods: Case report: We report on a 2-month old boy who was referred to our emergency department due to incidental finding of a self-limiting tachycardia (up to 200 bpm) during regular 2-month check-up.

Results: Pregnancy and birth were uneventful. After birth, he was admitted to our neonatal ward due to asymptomatic hypoglycemia (min. 0.9 mmol/l). I.v.-Glucose (max. of 4.4 mg/kg/min) was administered for 5 days. Other than that medical history was uneventful. Feeding intervals ranged between 2 (day) – 12 hours (night), weight gain was along the 10th percentile. On enquiry, the parents reported increased sweating -mainly nocturnal- for the last 2–3 weeks. On clinical examination hepatomegaly was found, heart rate range was around 150/min. Abdominal ultrasound confirmed hepatomegaly without distension of the hepatic veins. Echocardiography and electrocardiography were normal. Due to apparent nocturnal sweating on our ward, especially after prolonged fasting periods, investigations to rule out inborn metabolic disorders were undertaken. Blood testing revealed postprandial normoglycemia with slightly elevated lactate, and after 4 hours of fasting asymptomatic hypoglycemia (1.2 mmol/l) and hyperlactacidemia (max. 7.1 mmol/l). Further testing showed hypertriglyceridemia (5.67 mmol/l in later controls max. 18.4 mmol/l), normal uric acid but elevated metabolites from the citric-acid-cycle and elevated serum biotinidase. Liver biopsy confirmed enzymatically a glycogen storage disorder type 1a and adequate feeding was started.

Conclusion: Tachycardia and sweating in infants are often seen in pediatric cardiology but can also be caused by hypoglycemia. In severe or recurrent hypoglycemia, inborn metabolic disorders must be suspected and thorough workup has to be performed.

Disclosure of Interest: None declared

P012

FAILURE TO THRIVE IN A NEWBORN WITH CBLC DEFICIENT METHYLMALONIC ACIDURIA AND HOMOCYSTINURIA

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Introduction: The Cobalamin C deficiency, which is characterized by increased levels of homocystein and methylmalonic acid, is a very rare disease, but the most common inherited disorder of the cobalamin metabolism. Patients can present from the prenatal period to late adulthood with very unspecific symptoms.

Results: We report about a boy born at 38 + 2/7 age of gestation by caesarean section because of intrauterine growth restriction. A few weeks after birth he presented with failure to thrive, feeding difficulties, recurrent vomiting, lethargy and muscular hypotonia. Biochemical evaluation showed a normal blood count and a metabolic acidosis with high anion gap and elevated lactate without improvement after rehydration. The urine status showed a microscopic hematuria, without biochemical evidence for hemolytic uremic syndrome. Further investigation including sonography of the abdomen and skull as well as an echocardiography showed normal results. Metabolic testing revealed a low plasma methionine, increased plasma homocysteine and methylmalonic acid, and a significantly elevated propionylcarnitine, leading to a strong suspicion of a disorder in the cobalamin synthesis pathway. After admission a therapy with intramuscular hydroxycobalamine and intravenous carnitine was immediately started, which resulted in improvement of feeding, weight and biochemical markers. Genetic testing confirmed the diagnosis of CblC deficiency by compound heterozygosity for 2 formerly described mutations in the *MMACHC* gene (c.271dup and c.435_436del). Ophthalmological examination showed a mild maculopathy and a low vision therapy was initiated. Therapy with intramuscular hydroxycobalamine and oral carnitine was continued. After two weeks the patient was discharged in good general condition.

Conclusion: Measuring plasma homocysteine is recommended in every child presenting with feeding difficulties and failure to thrive in combination with unexplainable neurological, hematological or cardiac symptoms, visual impairment, atypical hemolytic uremic syndrome or vascular events. In contrast to the late-onset CblC disease, the early-onset form is described with poor long-term outcome. An immediate treatment significantly decreases mortality.

Disclosure of Interest: None declared

HUTCHINSON-GILFORD PROGERIA SYNDROME: A RARE CASE REPORT

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Introduction: Hutchinson-Gilford Progeria Syndrome (HGPS) is a rare genetic disorder characterized by premature aging involving the skin, bones, heart, and blood vessels. This is due to a dominant mutation of the Lamine A gene (LMNA). This interesting case is reported for its rarity.

Methods: An 11-month-old male infant from a no consanguineous marriage. We notice the death of his younger brother of an unknown cause. Hospitalized several times for global stunting with undernutrition.

Results: The clinical examination revealed an abnormal, indurated, sclerodermal skin associated with a flexum in the lower limbs. The scalp was covered with sparse hair with increased visibility of the subcutaneous veins. The facies was dysmorphic (bulging forehead, prominent eyes, small ears and micrognathia). He had a major stunting-weight loss associated with normal psychomotor development. Radiologically, there was an appearance of a piriform thorax with a diastasis of sutures in the skull. The serum lipid profile showed a decrease in the HDL level.

Conclusion: Progeria is a rare genetic disease. The cutaneous anomalies are constant and must allow the diagnosis of the disease and the early detection of atherosclerosis. Interest of genetic counseling. The antenatal diagnosis is feasible.

Disclosure of Interest: None declared

P013

CONGENITAL HYPERINSULINISM: ABOUT A DIAZOXIDE SENSITIVE FORM

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Introduction: Congenital hyperinsulinism (CH) is a rare disease. It is caused by an inappropriate hypersecretion of insulin responsible for severe and refractory hypoglycaemia. It is a therapeutic emergency because of the risk of neurological sequelae. It is a heterogeneous pathology in clinical, genetic, histological and treatment response.

Methods: A 7-month-old male infant, born of a no-consanguineous marriage, with no notable pathological history, admitted for the treatment of a pyretic seizures.

Results: A blood glucose test objectified a rate of 0.3 g/l. Monitoring of the glycemic cycle has shown profound hypoglycaemia of anarchic and rebellious schedules despite a continuous parenteral carbohydrate intake. Insulinemia concomitant to hypoglycaemia was elevated to 23 IU/m. Injection of glucagon with hypoglycemia resulted in a wide and prolonged response. The somatic examination revealed a delay of psychomotor acquisitions. Abdominal M.R.I revealed a discrete hypodensity of the pancreas head.

Conclusion: CH is the most common cause of severe hypoglycemia in infants. The prognosis remains reserved because of the risk of neurological sequelae. The interest to distinguish between the focal and the diffuse form will determine the surgical modalities and genetic counseling.

Disclosure of Interest: None declared

P014

EARLY EFFECTS OF ANTI-TNF PERFUSION ON BODY COMPOSITION AS DETERMINED BY BIOELECTRICAL IMPEDANCE ANALYSIS IN PEDIATRIC CROHN DISEASE

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Introduction: Tumor necrosis factor alpha (TNF) is a proinflammatory cytokine responsible for the majority of the manifestations of Crohn's disease (CD). This study aims at investigating early effects of anti-TNF perfusion on body composition as determined by bioelectrical impedance analysis (BIA)

Methods: This was a prospective study involving children (<18 years of age) followed for CM treated by infliximab in the pediatrics unit at the CHU of Amiens. The parameters determined by BIA prior to and 2 h after the perfusion of infliximab were: lean body mass (LBM), body fat (BF), body cell mass (BCM), total body water (TBW), intracellular fluid (ICF), and extracellular fluid (ECF).

P015

Results: Ten children were included, of whom four were girls, with an average age of 12.4 years, and an average BMI of 15.1 ± 2.9 kg/m². The body composition before and after the perfusion of infliximab was: 33% vs. 25% ($p = 0.03$) for BF in relation to weight and 47% vs. 53% ($p = 0.04$) for the ECF in relation to the TBW, respectively. Prior to the perfusion, the BCM (kg) and the ECF (l) were negatively correlated with markers of inflammation (CRP/ESR). After the perfusion: the BF (kg) and the LBM (kg) in relation to weight were positively correlated, while the BCM in relation to weight and the ECF in relation to the TBW were negatively correlated.

Conclusion: Our study indicates that early changes occur in the body composition of children treated with infliximab. These results nonetheless require confirmation in order to be of clinical significance.

NaCHwuchs Prize: I wish to apply

Plaschke Prize: I wish to apply

I have read and understand application rules: Yes

Disclosure of Interest: None declared

P016

COW'S MILK PROTEIN ALLERGY ASSOCIATED WITH PROTEIN-LOSING OR EXUDATIVE ENTEROPATHY CAN LEAD TO SEVERE HYPOGAMMAGLOBULINEMIA MIMICKING IMMUNODEFICIENCY

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Introduction: Protein-losing enteropathy (PLE) is a rare disorder with protein's loss in the digestive tract that can result in edema and hypoproteinemia. Immunoglobulins (Ig) are part of the lost proteins. We report 3 cases of PLE caused by cow's milk protein (CMP) allergy associated with severe hypogammaglobulinemia (hypolg).

Methods: A 5-months-old boy breastfed presented with failure to thrive, liquid stools, postprandial vomiting and general edema for 12 days. Severe hypolg without any infection was diagnosed. A primary immunodeficiency (PID) was suspected and the lymphocyte typisation was abnormal with low B and T cells. A digestive pan-endoscopy (PE) showed an active focal inflammation of the duodenal and caecal mucosa confirmed at histology. 10 days of parenteral nutrition allowed clinical improvement without recurrence under partially hydrolyzed formula milk diet. A 7-weeks old girl breastfed but occasionally receiving infant's formula complements presented with bloody diarrhea and postprandial vomiting since 3 weeks. Biopsies collected during a macroscopically normal PE showed severe inflammation and eosinophilic infiltration (EI), with massive glands destruction. Immunological explorations revealed severe hypolg. Amino-acid based formula allowed recovery of the digestive lesions. A 27-months-old boy presented with palpebral edema since 1 year of age, recent diffuse edema, and numerous upper airways infections. The child was fed with diversified diet and 1 liter of cow's milk per day. Immunologic exploration revealed a hypolg. Biopsies collected from a macroscopically normal PE showed a severe EI. Video-capsule endoscopy revealed actively bleeding linear ulcerations alternating with pale zones throughout the small bowel, suggesting villous atrophy. CMP eviction diet was applied with resolution of symptoms.

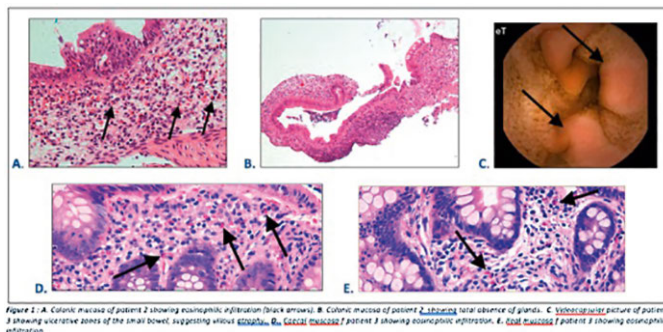


Figure 1

Results: We report 3 cases of infants with severe CMP allergy and associated PLE mimicking a PID at onset but whose symptoms resolved under CMP eviction diet and did not reappear during challenge test proposed 6–12 months after diagnosis. Because of the profound hypolg, Ig replacement therapy (IgRT) was administered before endoscopy. Ig levels normalized spontaneously after several

months, without infectious complications. The clinical severity and the patients young age required endoscopic procedures. We are showing characteristic endoscopic and histologic pictures.

Conclusion: These cases highlight that severe CMP allergy can mimic a PID and that IgRT should be reserved to specific cases.

Disclosure of Interest: None declared

P017

SYMPTOMATIC VITAMIN C DEFICIENCY IN A TWO YEAR OLD BOY

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Introduction: We present the case of a 2-year old boy with symptomatic Vitamin C-deficiency due to very selective alimentation habits.

Methods: The two year old boy was admitted to the University Children's Hospital Zürich with a history of 2 weeks of leg pain and refusal of walking, without any trauma. In the blood test we found elevated C-reactive protein and an even higher blood sedimentation rate. A meningitis and a fracture of the legs have been excluded. Clinically he was having tenderness on palpation on the thoracic spine. Our suspected diagnosis of a spondylodiscitis has not been confirmed by MRI. Instead, there were several bright intervals, suspicion of oedema, on the metaphysis of the proximal and distal femur, the proximal and distal humerus and the proximal and distal tibiae. Although the symmetry was not typical for any infectious or oncological disease, we did a bone marrow puncture, which was without any pathological findings. The clinical condition worsened every day, finally the patient was just lying in bed, without moving at all. Clinically we also found a gingivitis, a hyperkeratosis pilaris and scruffy hair. In his medical history he was having very selective eating habits, which concerns the parents since a long time. Since the age of 1 year he only eats noodles, special cream-cheese and baby-biscuits and only drinks water. So the patient had no intake of Vitamin C since about 1 year. In the literature we found reports about children with special eating habits and Vitamin C-deficiency, having exactly the same symptoms as our patient has. The changes in the MRI were the same, the gingivitis, the hyperkeratosis pilaris and the scruffy hair also were described. We highly suspected a symptomatic Vitamin C-deficiency and started a treatment with high-dosed Ascorbinacid (300 mg) per day for 7 days via nasogastric tube, then reduced to a maintenance dose of 100 mg per day.

Results: After 4-5 days of high-dose therapy with Ascorbinacid, the boy started to sit again, than tried to stand up. Before discharge, almost two weeks later, he could walk, holding his parents hands, without any sign of pain. Besides, the boy was brightening up, started to smile more, was more awake and wasn't anxiously all the time. Only his alimentation behaviour couldn't be changed so fast.

Conclusion: Vitamin C-deficiency is very rare nowadays but can have fatal effect on the children and their parents.

Disclosure of Interest: None declared

P018

IS NTCP DEFICIENCY A CAUSE OF FAILURE TO THRIVE?

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Introduction: Very little is known about deficiencies of bile acid transporters on the basolateral side of hepatocytes. In 2015 Vaz et al. published a first case of SLC10A1 deficiency causing hypercholanemia with normal bilirubin and autotaxin levels. Clinically their index patient presented with failure to thrive, without pruritus or jaundice (Vaz et al. 2015). Deng and Liu et al. added to the initial description with another case and a small series (Deng et al. 2016, Liu et al. 2017). However, full spectrum of clinical presentation of mutations in SLC10A1 is not known. Liver histology of patients with sodium-taurocholate cotransporting polypeptide (NTCP) deficiency has not yet been reported and the genotype-phenotype correlation is not known. We present another patient with a NTCP deficiency caused by a novel loss of function mutation in SLC10A1.

Methods: Case report

Results: Retrospective chart review of a female born in 2002 presenting with feeding disorder and failure to thrive leading to the discovery of liver disease of unknown etiology characterized by persistently elevated serum bile acids. A first liver biopsy is performed at a very young age. Ursodeoxycholic acid treatment is introduced and the liver panel is monitored. Clinically she is well. At age 14, a second biopsy is performed. Finally, genetic analysis is performed. The liver panel including ASAT, ALAT, gamma-GT, total bilirubin and direct bilirubin is normal. Bile acids are chronically elevated, up to 70 fold. Autotaxin level is normal. Liver biopsy at 2 years of age shows unspecific degenerative hepatopathy with focal steatosis. The second liver biopsy at 14 years of age, is unremarkable. Genetic analysis reveals a homozygote 5bp deletion in the *SLC10A1* gene leading to a frameshift with alteration of the first stop codon predicted to lead to a complete loss of function

Conclusion: We describe a new case of hypercholanemia due to a novel mutation of the *SLC10A1* gene coding for NTCP. The identification of this mutation leading to a complete loss of function confirms the major role of NTCP in hepatic bile acid uptake. Though the full spectrum of clinical presentation of NTCP deficiency is not known, we could hypothesize that there is a link with failure to thrive. We suggest taking NTCP deficiency into consideration when confronted with a patient presenting failure to thrive, unexplained anicteric cholestasis or when confronted with a liver biopsy showing mild steatosis of unclear etiology.

Disclosure of Interest: None declared

YOUTHS WITH CHRONIC CONDITIONS SHOW NO DIRECT ASSOCIATION WITH RISK BEHAVIORS

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Introduction: To compare risk behaviors between youths living with a chronic condition (CC) and their healthy peers, controlling for the severity of the condition.

Methods: Data were drawn from the baseline wave of the GenerationFRee study in 2015. Students aged 15–24 years in post-mandatory education participated in a web-based self-administered anonymous questionnaire assessing lifestyles. The sample (N = 5179) was divided into CC youths without limitations (N = 536; 10.4%), youths with limitations (N = 114; 2.2%), and a control group (N = 4'529; 87.4%). Groups were compared on internalizing (perceived health status, vision of their future, emotional wellbeing) and externalizing behaviors (substance use [tobacco, alcohol, cannabis, other drugs], gambling, excessive internet use, disordered eating, violent and antisocial acts) controlling for potential confounders. Statistical analyses were carried through a structural equation modeling. Results are given as unstandardized coefficients.

Results: Overall, CC youths showed an association with internalizing behaviors (coefficient: 0.80) but not with externalizing ones. The connection with externalizing behaviors was indirect via the internalizing ones (0.32). Perceived health status' coefficient being fixed as 1, the coefficient between CC youths and latent internalizing variables showed a significantly higher likelihood to report poor emotional wellbeing (coefficient: -1.64) and a negative vision of their future (-2.03). CC youths reporting psychological issues were more likely to adopt every externalizing behavior, ranging from 0.09 for disordered eating to 1.55 for the use of illegal drugs. Analyzing separately youths with CC limiting daily activities and those without limitations, the results were very similar, but the association with internalizing behaviors was much higher for those with limitations (2.21 vs. 0.44).

Conclusion: Our results show that there is no direct link between suffering from a CC and adopting risk behaviors, but that the association is indirect through internalizing factors, mainly a negative view of their future. Health professionals should address emotional wellbeing and perception of the future rather than focus exclusively on the effects of risk behaviors on specific diseases. The results highlight the need for these youths, especially those with limiting conditions, to have a life plan including educational and vocational goals instead of reducing it to a plan centered on their condition.

Disclosure of Interest: None declared

P019

HYPERTHYROIDISM IN AN ADOLESCENT TREATED WITH AMIODARONE

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Introduction: We report the case of a 13 year-old female who presented with severe hyperthyroidism secondary to amiodarone treatment.

Methods: This patient has been diagnosed during childhood with Prinzmetal's angina, a rare disease at this age, in which various (stressful) situations induce coronary vasospasm. In our patient, vasospasms were often complicated by sustained ventricular tachycardia, sometimes leading to cardiac arrest (CA), reason why amiodarone treatment (150 mg/day) was started, as it also plays a protective role on coronary vasospasms. After 2 years of treatment without any recurrence, she was hospitalized following a new episode of loss of consciousness from presumed CA. In the work-up, a severe hyperthyroidism with an undetectable TSH level and a very high free T4 level (98.8 pmol/L, normal values 12–22 pmol/L) was detected. Thyroglobulin was also increased, reflecting a possible iodine deficiency. Thyroid auto-antibodies were negative and the ultrasound was normal. Of note, at initiation of amiodarone, the thyroid function was normal. Thyroid hormone levels had never been monitored since amiodarone treatment was started because the patient had a history of CA secondary to blood sampling.

Results: The literature reports that amiodarone-induced dysthyroidism develops in 20% of adult patients. Hypothyroidism is the most frequent side effect. However, amiodarone-induced hyperthyroidism (AIH) can be found in patients with underlying thyroid pathology and in iodine-deficient regions (Type 1), or can be secondary to destruction of thyroid tissue caused by an amiodarone-induced inflammation (Type 2). Type 1 AIH responds to antithyroid agents (ATA) while type 2 AIH are treated with steroids. Treatment of AIH is difficult because amiodarone is often the only way to manage the patient's arrhythmia and cannot be suspended. Furthermore, discontinuation of amiodarone treatment might not resolve the thyrotoxicosis since it is highly lipophilic and has a long elimination half-life. For our patient, the type of AIH was uncertain and Carbimazole treatment was started with only little effect: steroids were then added. At this time, efficacy is still unsatisfactory. Cardiac alternative treatment to amiodarone are discussed, but surgical thyroidectomy may become a solution.

Conclusion: Adverse effects of amiodarone treatment, such as hyperthyroidism, are rare but can be severe, leading sometimes to surgical thyroidectomy. Monitoring must be performed carefully during treatment.

Disclosure of Interest: None declared

P020

A PUZZLING CASE OF PROTEIN-LOSING ENTEROPATHY OR PROTEIN-LOSING ENTEROPATHY DUE TO SEVERE IRON DEFICIENCY ANEMIA?

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Introduction: Protein-losing enteropathy (PLE), presenting mainly with edema due to hypoproteinemia, is a rare complication of a broad variety of disorders and might be due to a circulatory problem (venous or lymphatic congestion) or disruption of the normal structure of gastrointestinal mucosa.

Methods: Case report: A 15-month-old boy presented with swollen eyelids for 3 weeks and several days of fatigue, intermittent diarrhea and poor appetite. Past medical history revealed an uncomplicated pyelonephritis with 3 months and multiple episodes of otitis media. Physical examination was unremarkable besides bilateral periorbital edema. Laboratory studies showed hypoalbuminemia, hypogammaglobulinemia and severe iron deficiency anemia. There was no renal protein loss and elevated fecal α 1-antitrypsin confirmed suspected enteral loss. Iron deficiency was considered secondary to a presumed enteral pathology and substituted orally. Enteropathogenic *E. coli* found in stool motivated an ineffective trial of azithromycin therapy. Chest x-ray and abdominal ultrasound were normal. Tc-99 scintigraphy didn't identify pathologic protein loss. Upper and lower endoscopy showed duodenitis with villous atrophy but without crypt

P021

hyperplasia or increased intraepithelial lymphocytes. Autoimmune enteropathy or common variable immunodeficiency was suspected and a treatment with prednisolone was initiated, along with a diet rich in medium chain fatty acids. The patient showed a very rapid and complete recovery. Extended laboratory studies didn't confirm immune deficiency nor identify anti-enterocyte antibodies. Considering the paucity of symptoms upon presentation, no long-term immunosuppression was started. Prednisolone therapy was tapered after 2 and stopped after 8 weeks. On follow-up the patient had no complaints and laboratory values normalized. One year after initial diagnosis upper endoscopy was normal.

Conclusion: PLE should be considered in every patient with hypoproteinemia after exclusion of malnutrition, defective synthesis or proteinuria. Finding the underlying disease can be challenging. Here, despite thorough work up, the cause remains unclear. Iron deficiency might have played a more important role than initially thought. Literature research revealed several cases – mostly reported in the 1980s – with PLE due to iron deficiency anemia, which improved solely under iron supplementation. The underlying pathophysiologic mechanism is not known. Nevertheless, it should be considered as a possible cause of PLE.

Disclosure of Interest: None declared

P022

NEVUS SEBACEOUS, CLINICAL COURSE AND COMPLICATIONS

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Introduction: Neonates may have rashes of all shapes at birth. Among them, it's important to discriminate skin lesion with potential systemic issue. Nevus sebaceous (NS) is a rare type of birthmark with broad range of abnormalities that may affect other organ systems.

Methods: Case presentation The patient was born at 40 weeks of gestation, of an uneventful pregnancy and a normal antepartum ultrasound morphology. At the first examination, we found an isolated left temporal skin lesion, described as a linear, orange, cerebriform birthmark of 5x1 cm, along lines of Blaschko. There were no other anomalies. Family history revealed no skin abnormalities or congenital defects. This atypical presentation required expertise of pediatric dermatologist, who diagnosed a NB, with potential associated ocular and cerebral abnormalities. Neurologic examination and cerebral ultrasound were normal. Ophthalmologic consultation has been planned. Skin biopsy will be done to confirm exact diagnosis at 6 months. Excision of the lesion was recommended after one year, because of cosmetic issue and risk of malignant transformation.

Results: Discussion: NB is a rare benign hamartoma of the skin, characterized by hyperplasia of the epidermis, immature hair follicles, sebaceous and apocrine glands. It is apparent at birth and presents as a well-defined, thin, orange, linear plaque, located on the scalp. In older children, lesions tend to be more elevated and verrucous. NB syndrome is defined by the association with cerebral, ocular or skeletal defects. NB occurs in 0.3% of newborns. Epithelial neoplasms may arise including basal cell carcinoma. Diagnosis is usually based upon the clinical presentation. Biopsy may be required if diagnosis is uncertain or to confirm the type of lesion. For patients with a suspected NB syndrome, a neurologic and ophthalmologic examination should be performed. Additional evaluations include neuroimaging studies and skeletal radiography if there are skeletal disorders. Treatment is full-thickness excision. Decision to excise should be made based upon age, extension and location of the lesion and the patient's concern about cosmetic appearance and risk of malignancy.

Conclusion: It's important to identify NB to rule out cerebral, ocular and skeletal abnormalities. Excision was recommended but observation may be reasonable as the risk of malignant transformation seems to be lower than believed. Among the differential diagnosis there is aplasia cutis if lesion appears thin and glabrous.

Disclosure of Interest: None declared

MANAGEMENT OF COMBINED CENTRAL DIABETES INSIPIDUS, SIADH AND CEREBRAL SALT-WASTING SYNDROME AFTER NEUROSURGERY IN INTENSIVE CARE

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Introduction: Central diabetes insipidus (CDI) results from vasopressin (AVP) deficiency and leads to inability to concentrate urine, resulting in polyuria and polydipsia. It can occur secondary to processes affecting the hypothalamus or the posterior pituitary gland. Salt and water balance disturbances are frequent complications of neurosurgical interventions, occurring in 83% of patients with intra- or parasellar tumors and can follow a triphasic course. An initial transient CDI can be followed by an oliguric phase due to the syndrome of inappropriate anti-diuretic hormone secretion (SIADH), which can then be followed by a permanent CDI. In addition, the clinical picture can be complicated by cerebral salt wasting (CSW) that occurs in 4% of the children, due to release of natriuretic peptide.

Methods: We present the case of a three-year-old boy who developed a triphasic course of salt and water disturbances concurrently with CSW after neurosurgical removal of a suprasellar pilocytic astrocytoma. We describe the challenging post-operative fluid and electrolyte management.

Results: On day 1 after surgery, our patient developed a maximal diuresis of 14.6 cc/kg/h, a hypernatremia of 158 mmol/L and increased plasma osmolality of 320 mOsm/kg, compatible with CDI. Treatment with desmopressin, an analogue of AVP, was begun and water losses were replaced hourly. On day 3, diuresis suddenly decreased to 1.4 cc/kg/h, natremia dropped to 132 mmol/L and plasma osmolality to 271 mOsm/kg, in absence of desmopressin, permitting diagnosis of SIADH and necessitating restriction of fluids. Desmopressin was suspended. On day 5, diuresis increased again up to 19 cc/kg/h, fluid replacements had to be increased again and desmopressin was reintroduced. Hourly evaluation of water balance and natremia was needed to maintain a euvolemic state and to control natremia. On day 6, despite an equilibrated hydro-urinary balance, the patient developed hyponatremia up to 124 mmol/L with concomitant urinary Na up to 185 mmol/L and CSW was suspected requiring NaCl substitution up to 38 meq/kg/day.

Conclusion: Recognition and proper management of CDI, SIADH and CSW is mandatory to prevent life-threatening electrolyte disturbances. Thus, close monitoring of water balance, natremia and natriuresis in intensive care is important after neurosurgery. Indeed, diuresis and natriuresis can change abruptly, necessitating continuous adjustment of water and NaCl replacement as well as desmopressin administration.

Disclosure of Interest: None declared

P024

P025

A CONGENITAL HAIRLESS LESION OF THE SCALP

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Introduction: In 1998, Rudolph Happle first introduced the term Nevus psiloliparus (Greek *psilo* = hairless, *liparus* = fatty) for a distinct type of mesodermal nevus of the scalp.

Methods: (Case report): A full-term female neonate was referred on second day of life to paediatric dermatology for a soft, hairless swelling on the right parietal-temporal scalp. Pregnancy, labour and family history were unremarkable. On examination, the 25 x 20 x 5 mm oval, soft, skin-coloured lesion presented a smooth surface and absence of mature hair follicles on dermoscopy. A complete physical work-up including cardiology and ophthalmology was unremarkable. Sonography revealed a subcutaneous 5 mm deep lipomatous mass. Cerebro-spinal duplex-sonography at age 2 weeks was normal. Follow-up at age 2 months showed normal development and head circumference. Close paediatric follow-up is planned, counselling for later plastic surgery will take place at age one year.



Figure 1 (The photographs are published with an informed consent from the parents.)



Figure 2

Results: The characteristics of the scalp lesion led to the diagnosis of a nevus psiloliparus with no obvious signs of encephalocraniocutaneous lipomatosis (ECCL). ECCL is a neurocutaneous syndrome including a variety of skin, eye and cerebro-spinal anomalies such as choristomas, colobomas, intracranial and spinal lipomas, hydrocephalus and porencephalic cysts. Considering the poor correlation of symptoms with imaging, transcranial and spinal sonography was considered an appropriate diagnostic approach in early infancy to investigate the possible presence of major cerebral and spinal anomalies, avoiding general anaesthesia for MRI and profiting from the ideal sonographic conditions at this age. MRI may be considered in later follow-up or in case of symptoms. Up to now, most cases of nevus psiloliparus (approx. 70) reported in the literature were those in the context of ECCL, only few being non-syndromic. Our case is most likely a further example of the non-syndromic form. In line with several authors, we suspect that the non-syndromic form is highly underreported. Nevus psiloliparus can present unexpectably with only limited size. Misdiagnoses, e.g., as aplasia cutis, lipoma, nevus sebaceous or smooth muscle hamartoma may be common.

Conclusion: Clinicians should include nevus psiloliparus in their differential diagnosis for hairless congenital scalp lesions. Although nevus psiloliparus is considered a hallmark for ECCL, the non-syndromic form is very likely underreported due to unfamiliarity with, and misdiagnosis of, this rare skin condition.

Disclosure of Interest: None declared

PNEUMOCOCCAL FATALE INFECTION DUE TO STREPTOCOCCUS PNEUMONIAE SEROTYPE 23B IN A 2-MONTH-OLD INFANT: CASE REPORT.

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Introduction: Swiss infant vaccination includes protection against pneumococcal infections since 2010 with a vaccine, Prevenar[®] (PCV13) which contains 13 serotypes of *Streptococcus pneumoniae*. Increasing numbers of serotypes in vaccine helps to cover virulent germs and to improve protection of the children. However, it leads to the emergence of new serotypes that can infect infants because of their immunological immaturity. We report the case of a boy, not vaccinated and who was infected by an invasive pneumococcus of serotypes 23B.

Methods: A 2-month-old healthy boy was transferred in cardiopulmonary arrest. He was suffering from an upper airways viral infection, which became complicated into bronchiolitis 3 days before admission. Since the morning, food intake was reduced. Parents reported tiredness and noticed that the baby was no more arousable. An ambulance was called and when medical staff arrived, the boy was in cardiopulmonary arrest. Cardiopulmonary resuscitation (CPR) was started. After 45 minutes, decision to stop the reanimation was taken. An autopsy was practiced and specimens showed a panlobular necrotizing pneumonia. *Streptococcus pneumoniae* 23B was detected in pulmonary, cardiac and tympanic tissues. Respiratory syncytial virus (RSV) RNA was retrieved in lung tissues. Severity of the infection motivated a search for a primary immune deficiency which was not found. We concluded that our patient was suffering from a bad evolution of a RSV pneumonia with superinfection by pneumococcus.

Results: The death was unexplained and could have been secondary to a worsening undetected respiratory insufficiency or to an acute apnea due to RSV pneumonia. However, the autopsy revealed an invasive pneumococcal infection with a rare 23B serotype which is not included in PCV13. The emergence of new strains could be explained by the selection of rare strains uncovered by PCV13. Moreover, this baby was at a vulnerable age and has probably not been protected by maternal antibodies probably because his mother herself has not been infected or colonized in infancy by this rare strain.

Conclusion: In conclusion, it is important to emphasize, first, the importance of autopsy to maximise the chance to identify the real cause of an unexplained infant death and secondly the importance of the surveillance of the emergence of new strains which could be selected after the implementation of a new vaccination.

Disclosure of Interest: None declared

KINGELLA KINGAE ENDOCARDITIS: A SLOW PROGRESSION BUT A LIFE-THREATENING CONDITION

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Introduction: We report the case of a child with a cardiogenic shock following *Kingella kingae* (Kk) endocarditis.

Methods: A previously healthy 6-year-old girl presented with a hypotensive cardiogenic shock. Her parents related fever for 8 days with initially reported oral ulcerations, progressive malaise and anorexia. On arrival, she showed respiratory failure, tachycardia (150/min), poor peripheral perfusion, low blood pressure (73/45 mm Hg), and hepatomegaly. Chest X-Ray confirmed an enlarged cardiac silhouette with increased vascular markings. Inflammatory markers were elevated (C-reactive protein 143 mg/L, procalcitonin 4 µg/L) and metabolic acidosis was present (lactates 4.8 mmol/L). Echocardiography revealed heart failure with 18% ejection fraction, moderate mitral and severe aortic insufficiency and an abscess on a bicuspid aortic valve. Hemodynamics worsened rapidly with a cardiac arrest after intubation. Extracorporeal membrane oxygenation was started followed by cardiac surgery to treat the major aortic regurgitation and to remove the abscess. Presence of Kk was identified in blood, nasopharyngeal and pericardial fluid cultures.

Results: This case illustrates a cardiogenic shock due to Kk endocarditis in a child with no underlying cardiac anomaly except a bicuspid aortic valve. This congenital anomaly affects 1–2% of the population and represents a risk factor for infectious endocarditis (IE). Although IE is a rare affection in children, it is a significant cause of morbidity and mortality. The most common responsible organisms are *Streptococci viridans* and *Staphylococcus aureus*. However, Gram-negative species endocarditis (HACEK group) like Kk may rarely be encountered in pediatric population, but they are known to cause severe complications. Kk is commonly found in the oropharyngeal flora of children under 4 years with 10% carrier state and is transmitted through close contact. The majority of Kk infections (mainly osteoarticular manifestations) is usually diagnosed in children less than 4 years old. They commonly manifest with subtle clinical symptoms and mild biological inflammation.

Conclusion: Kk endocarditis typically presents with subacute nonspecific symptoms before the patient manifests with a life-threatening condition. The emergency pediatrician should adequately recognize impending cardiogenic shock. Rare pathogens such as Kk may be responsible for severe endocarditis in patients with no or even mild congenital heart disease.

Disclosure of Interest: None declared

P026

P028

USE OF NASAL BUBBLE CPAP IN NEWBORN: REPORT FROM A TERTIARY TEACHING PEDIATRIC HOSPITAL IN DAKAR, SÉNÉGAL

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Introduction: Neonatal mortality accounts for 40% of all under 5 deaths. Preterm, infections, asphyxia are the leading cause of morbidity and mortality and the main clinical manifestation is respiratory failure. Nasal bubble continuous airway pressure (bCPAP) is preferred and recommended by WHO in developing countries for easy applicability and low cost. Because it is unexplored in Senegal, we sought to evaluate its utility in very ill and/or hypoxemic newborns.

Methods: Retrospective evaluation of all newborns receiving bCPAP during one year (2016) in the Pediatric Unit of a teaching and referral hospital in Dakar, Senegal. Pressurized oxygen from a central source was delivered through a flow meter to the patient via nasal prongs.

Results: 76 newborns (<1 month of age), 40 boys et 36 girls: 54% (n = 41) full term babies, 24% (n = 18) premature babies between 28 and 32 weeks of gestational age, 22% (n = 17) >32 weeks. Duration of hospitalization: <3 d 26% (n = 20), 3–7 d 25% (n = 19) >7d 49% (n = 38). Causes of admission: Respiratory distress 53% (n = 40), neurologic (convulsions, fits) distress 39.5% (n = 30), severe bacterial infection 7.5% (n = 6). Cyanosis was present before initiating bCPAP in 85.5% (n = 65) and SpO₂ measured in 91% (n = 65): SpO₂ was <60% in 12, >60% and <90% in 47 and >90 in 6. Final diagnosis (>100%) were: Respiratory infection 77% (n = 55), Meningitis 22% (n = 17), Amniotic fluid aspiration 53% (n = 41), Hypoxic ischemic encephalopathy 35% (n = 27), Hyaline membrane disease 14% (n = 11). Clinical evolution was globally quickly effective with decrease of respiratory distress; SpO₂ dropped >90 after initiating CPAP for 90% (n = 69) of patients. Total bCPAP duration: <24 h 23% (n = 17), >24 h 77% (n = 59). 40 babies survived with complete recovery, 4 with neurological sequelae; 30 babies died (severe septic choc, brain damage or refractory hypoxia). Failure and complication with the bCPAP device were negligent.

Conclusion: Nasal bCPAP is safe and effective in newborn in Senegal where respiratory support is mandatory. A prospective trial with definite inclusion criteria must be conducted and bCPAP progressively implemented in pediatric units in Senegal. Its utility must also be considered in older children with respiratory disease (bronchiolitis, pneumonia...).

Disclosure of Interest: None declared

P029

PREGNANCY AND DELIVERY DURING ADOLESCENCE: A PUBLIC HEALTH PROBLEM

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Introduction: Adolescents' pregnancy and delivery is considered as a public health problem in the world. In this descriptive and prospective study, authors provide Moroccan data about the adolescents' mothers as well as their pregnancy evolution, the delivery mode and the characteristics of the newborns.

Methods: This is a descriptive and prospective study conducted during the period between January 2015 and December 31st, 2015. This study involved 308 teenagers' mothers who attended the consultation at the medicine and neonatal reanimation unit at the Children's Hospital of Rabat.

Results: The percentage of teenagers' delivery is 1.8%. The median age of mothers is 17 years old and 6 months, 6% of whom were multigravida and 14% presented an intercurrent disease. The percentage of premature delivery is 12%. Only 18.2% of the teenagers had a C-section delivery. A percentage of 10.5% of the newborns had a low birth weight. Fifteen newborns have been hospitalized in the intensive care unit. Congenital malformation has been found among 1.3% of the newborns.

Conclusion: Adolescents' pregnancy and delivery in Morocco does not seem to be free of risks. A prevention policy and a good prenatal monitoring have to be applied to prevent adverse outcomes of early pregnancy.

Disclosure of Interest: None declared

STRIDOR IN NEWBORNS: NOT ALWAYS A LARYNGOMALACIA

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Introduction: Case report: a female infant born at 31 weeks of gestation was admitted to our department at birth for prematurity. Fetal ultrasounds were unremarkable. The pregnancy was complicated due to maternal pre-eclampsia. A caesarean section was performed because of increasing symptoms of pre-eclampsia in the mother. Birthweight was 1490 g, Apgar 6/7/7 and umbilical arterial pH 7.32. At birth, the symptoms of acute respiratory distress syndrome were present due a combination of surfactant deficiency and wet lung. The initial respiratory support was nasal CPAP which then later on was reduced to High Flow nasal cannula system. After a period of respiratory improvement, the baby developed a continuous inspiratory and intermittently expiratory stridor with no additional respiratory sounds, requiring persistent respiratory support in the form of a High Flow. The stridor persisted and it worsened during feedings. A change of position of the baby didn't show any improvement. Further diagnostics were performed in search of the etiology and therefor an echocardiography was done, which revealed a double aortic arch. The baby was then referred to a cardiac centre for interdisciplinary treatment.

Conclusion: Conclusion: a chronic stridor is a common clinical finding in newborns and infants. The phase of the stridor corresponds to the anatomical level of airway obstruction. The most frequent cause of stridor is a laryngomalacia, which, in most of cases, presents exclusively with inspiratory stridor and worsens in a supine position. A laryngomalacia does not require respiratory support and further diagnostics, since it resolves spontaneously over time. When a stridor is biphasic and requires respiratory support, further diagnostics are mandatory.

Disclosure of Interest: None declared

P030

OPTIMIZING RESIDENT EXPOSURE TO RESUSCITATIONS: A QUALITY IMPROVEMENT PROJECT

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Introduction: Involvement in resuscitations is a key aspect of pediatric medical education. Such exposure is often random and unequally divided among the residents, which may lead to discrepancy in knowledge acquisition. Using a Plan-Do-Study-Act (PDSA) quality improvement methodology, we created a simple Emergency Department (ED) tool to even out the exposure while theorizing that by extension, this would optimize resident training.

Methods: Using the administrative database of a tertiary care center pediatric ED with an annual visit of 29,000 patients treating children 0 to 16 years of age, resident exposure to resuscitation cases was analyzed. A pre-intervention anonymous survey was sent to the residents rotating through the pediatric ED. The survey inquired about the perception of the residents to resuscitation case exposure, as well as their opinion about the utility of our project. Resuscitation cases were defined as one with possible alteration of the Pediatric Advanced Life Support (PALS) assessment triangle. A simple paper solution was then developed which kept track of the resuscitation cases seen by the residents, and placed in a central location in the ED. At each shift change, the resident and the attending physician team reviewed the table, to decide which resident would be first mobilized for a resuscitation case that day. Post intervention data was analyzed.

Results: Pre-intervention analysis of the database revealed a variance of 6.9. All eleven residents rotating in the ED responded to the initial survey. Resident perception of their involvement in resuscitation cases was inaccurate 40% of the time (95% CI 0.22–0.63). Their opinion regarding the utility of the project was favorable in 90% of the cases. Post intervention analysis showed a variance of 1.7.

Conclusion: In our cohort, resident perception of their exposure to resuscitation cases is inaccurate. Residents agree that evening out the unequal exposures to resuscitation cases is a worthy effort. A simple tool such as a table placed centrally and regularly referred to has led to substantial narrowing of the inequality. Further analysis is necessary to confirm the sustainability of our results and their impact on residency training.

Disclosure of Interest: None declared

P031

P032

COMA DUE TO SPONTANEOUS INTRACEREBRAL HAEMORRHAGE IN CHILDREN: POOR AWARENESS OF INTRACRANIAL HYPERTENSION AND POOR EMERGENCY NEUROLOGICAL LIFE SUPPORT

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Introduction: The Paediatric Intensive Care Unit physician team of our hospital has noticed that intracranial hypertension and herniation signs and symptoms were not recognised by medical emergency teams in children with spontaneous intracranial haemorrhage (SICH). Furthermore, recent guidelines for empiric emergency clinical evaluation and treatment of intracranial hypertension and haemorrhage were propagated by the "Emergency Neurological Life Support" (ENLS).

Methods: In our retrospective quality management study we focussed on children, 1 month to 16 years old, with spontaneous intracranial haemorrhage and an initial Glasgow coma scale ≤ 8 , who were admitted to the paediatric intensive care unit of our hospital between 1993 and 2015.

Results: In order to evaluate the awareness of the medical emergency team within our study, we retrospectively searched during the entire pre- and intrahospital emergency care of each patient for symptoms or signs of intracranial hypertension and herniation and for the items listed in the Pre-intubation Neurological Assessment Checklist of ENLS. We also searched for working diagnosis and documented awareness for intracranial hypertension and herniation. We compared treatments given to children suffering from coma and SICH with such state of the art paediatric ENLS. We evaluated the completeness of the emergency treatments given to each single child consisting of the sequential treatment approach to intracranial hypertension and herniation as bridging therapy until appropriate imaging and surgical treatment be performed.

Conclusion: In summary, our study showed that in 14 children presenting with acute coma due to spontaneous intracranial haemorrhage all patients had additional symptoms and signs of intracranial hypertension and herniation, including abnormal pupillary findings in 57% of them. All were intubated and ventilated, but none had documented treatment targets for ventilation and blood pressure, no child with pathological pupillary findings had forced hyperventilation, only one patient received single drug osmotic therapy and none had multiple drug osmotic therapy prior to imaging and final diagnosis. These findings confirmed that children with SICH presenting with acute coma received insufficient baseline stabilisation and minimal empiric management of intracranial hypertension and herniation when compared with therapeutic manoeuvres proposed by the Emergency Neurological Life Support.

Disclosure of Interest: None declared

P033

NEONATAL PERTUSSIS WITH LEUKEMOID REACTION: HYDROXYUREA AS AN ALTERNATIVE?

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Introduction: A 73-days-old infant of 34 1/7 weeks' gestation, vaccinated following the Swiss vaccine recommendations, was hospitalized with respiratory distress caused by Respiratory Syncytial Virus and secondary co-infection with *Bordetella pertussis* (BP) treated with clarithromycin for 7 days. Initially, non-invasive ventilation was needed. However, due to hypoxemia and persistent hypercapnia a transfer on day 5 to Pediatric Intensive Care Unit was needed for invasive ventilation. The White Blood Cell (WBC) count showed a leukemoid reaction with a highest WBC of 70 G/l at day 6. Echocardiography highlighted pulmonary arterial hypertension without cardiac failure. Therefore, there was no indication for leukapheresis nor exchange transfusion. As the respiratory situation did not improve, we decided to give an off-label hydroxyurea (Litalir[®]) treatment with increasing doses from 10 to 30 mg/kg/day. We noticed a concomitant decrease of leukocytosis to 34 G/l after 5 days of treatment. Simultaneously, we observed a respiratory improvement with extubation at day 14 and discharge at day 24.

Methods: Case-report and literature review

Results: Since more than 100 years, an association between the leukemoid reaction induced by BP toxin and a fatal evolution has been proven. The risk is greatly increased by WBC over 100 G/l, and a negative impact on the survival is associated with leukocytosis over 50 G/l. Currently, there is no treatment for severe whooping cough;

no ventilation strategy has proved superior for resolving hypoxia; many pulmonary vasodilators did not impact survival; and extracorporeal oxygenation is associated with a high mortality. Consequently, it is essential to treat the leukemoid reaction as the origin of fatal complications (right heart or respiratory failure, intracranial hemorrhage, and cerebral hypoxia). The effect of hydroxyurea on hyperleukocytosis of acute myeloid leukemia is known and permits a decrease of 50 to 80% of the leucocyte count in 24–48h. Borrowing from the usage in oncology, we decided to introduce an off-label treatment at a lower dosage to decrease the leukemoid reaction and avoid complications. This strategy permitted a favorable evolution without side effects and without resorting to more invasive and risky therapeutic measures.

Conclusion: The usage of off-label hydroxyurea in this case of leukocytosis due to BP infection is an effective and safe non-invasive alternative in the absence of criteria for invasive treatment. This indication deserves to be further studied.

Disclosure of Interest: None declared

P034

SUBOPTIMAL SEQUENTIAL ORAL CHLORAL HYDRATE AND INTRAVENOUS MIDAZOLAM RESCUE SEDATION PROTOCOL FOR PAEDIATRIC MRI'S

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Introduction: Chloral hydrate (CH) sedation is routinely used to allow motion free magnetic resonance imaging (MRI) in children. Sedation success is usually close to 90%. A sedation procedure for paediatric (0–16 y) MRI using oral CH (75 mg/kg, respectively 25 mg/kg if not asleep after 30–45 minutes, maximal cumulative dose 2000 mg) followed by one or two iv midazolam rescue doses (0.1 mg/kg, maximum 2 mg each) has been in use in our institution since 2010. We determined the sedation success rate and compared it with literature reports.

Methods: In a prospective case analysis, we studied the whole procedure, from iv line placement to end of images acquisition, to determine the global success rate – defined according to images quality or sleep induction failure. Critical steps and side effects in the sedation procedure as well as efficient use of MRI room were analyzed.

Results: From 01.01.2014 to 31.12.2015, 293 paediatric MRI's were realised, of which 60 under sedation. 58 patients, median age 2.2 years (range 0.1–9.7) were analysed. 30/58 (52%) children slept through the whole procedure. 32/58 (55%) procedures led to perfect sedations/images. Sedation quality allowed to answer clinical questions in 47/58 (81%) patients (clinical success). 10/58 (17%) procedures failed to be realized within the booked imaging time-slot. 7/11 failures were associated with arousal during the child's transfer from his/her bed to MR tube.

Conclusion: Our sedation procedure with oral CH followed by iv midazolam rescue for paediatric MRI showed a clinical success rate of 81%, which compares poorly with literature reports. The critical sedation step was the transfer from the patient's bed to the MR tube. Sedation procedure adaptations with other sedatives and/or a MR compatible restraining mattress are potential optimizations to be investigated.

Disclosure of Interest: None declared

P035

LEUCOCORNEA IN A NEWBORN: ATHALAMY AS A RARE CAUSE

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Introduction: Athalamy is a dysgenetic pathology of eye's anterior segment, with total or subtotal disappearance of the eye's anterior chamber. We present a case of a newborn with leucocornea overlying an athalamy.

Methods: Case presentation: a term newborn with normal maternal serologies was born after a pregnancy marked by a non-requiring gestational diabetes mellitus. At birth, a right leucocornea was noticed associated with severe microphthalmia on this side only. Further investigations showed a severe dysgenesis of the anterior eye

segment, kerato-irido-lenticular synechias and an ocular hypotension (4 mm Hg). An ultrasound revealed a retinal detachment, possibly caused by uveal effusion secondary to the severe microphthalmia. The process didn't seem to be recent. Left eye was normal. The visual prognosis being null, an eye prosthesis will be placed to allow a good orbital growth and avoid a facial asymmetry. Genetic investigations are on the way to try to find the cause of the microphthalmia, probable source of all the findings.

Results: Discussion: athalamia is caused by the absence or insufficient amount of aqueous humor. Mainly of post-traumatic or post-surgical origin with aqueous humor leakage, congenital cases are also known. Eye's anterior segment's malformation are the consequences of developmental and maturational anomalies. Different phenotypes are seen, depending on the timing of insult in fetal life. In our patient, the null visual prognosis is related to the posterior ocular involvement. Differential diagnosis of eye microphthalmia and anterior segment malformation is relatively wide, but the present case's phenotype could correspond to a severe Peter's anomaly, with mutation of PAX6 gene. Other genes can however be involved, as PITX2 and CYP1B1. This anomaly can be associated to craniofacial anomalies, cardiac defects, pulmonary, genital, urinary or central nervous system malformations and hearing defects.

Conclusion: A neonatal leucocoria can have many origins: glaucoma, keratocone or trauma (forceps) and anterior chamber dysgenesis. Neonatal athalamia is a rare cause. The possibility of a Peter's anomaly must lead to investigations in search for associated malformations.

Disclosure of Interest: None declared

P036

CHRONIC RECURRENT URTICARIA WITH LONG LASTING LESIONS: DON'T FORGET TO CHECK FOR BEDBUGS!

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Introduction: Chronic urticaria is defined by the presence of pruritic hives lasting over 6 weeks. It can be characterized by persistent or intermittent lesions. Its differential diagnosis is large and an underlying allergy, in particular to food, is often suspected by parents. In particular in patients with recurrent symptoms, papular urticaria should be considered and a causing insect actively sought out.

Methods: We present the case of an 11 years-old healthy, non-atopic boy who was addressed to our allergy consultation for a two months evolution of mild to moderate pruritic lesions, particularly on the upper and lower extremities. The single lesions lasted 2–3 weeks without leaving scars and new lesions were almost daily observed. No other symptoms, in particular fever, weight loss or arthritis were reported. At physical examination, the boy was in good general conditions with 2–5 cm large, hyperemic hives with a central hardened papulae on arms, legs and back. The rest of physical examination was normal.

Results: Based on clinical history and characteristics of skin lesions, papular urticaria was suspected and parents were asked to actively search for an insect infestation in the child's bed or on animals to which the boy had regularly contact. A bedbug infestation was in fact found on the mattress and after disinfestation and symptomatic treatment, the lesions rapidly disappeared and never reoccurred.

Conclusion: Papular urticaria is a pruritic hypersensitivity reaction due to insect bites, in particular mosquitoes, fleas and bedbugs leading to recurrent papular pruritic lesions. It is more often observed in young children between 2 and 10 years of age. Diagnosis is made clinically but is often delayed as usually only one child is affected which doesn't lead the family to suspect an infestation. Although mosquitoes are the most common insect causing papular urticaria, a bedbug infestation should be actively sought out. Although clinical manifestations are aspecific, the presence of a central papule, bite marks, grouped lesions in exposed areas of the body are typically observed. Bedbugs (*Cimex lectularius* and *Cimex hemipterus*) are obligate, blood-feeding insects that typically infest rather older building and beds. Although primarily affecting less developed countries, bedbug are increasingly observed in Europe and their presence not necessary correlated with a poor hygienic status.

Disclosure of Interest: None declared

CHEST PAIN IN CHILDREN

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Introduction: Non-traumatic chest pain is a common symptom in children and adolescents. It is one of the most frequent reasons for referral to the emergency department and to the pediatric cardiologist. An accurate history and physical examination are usually sufficient to determine the etiology and identify patients who require additional work-up or acute intervention. The management of pediatric patients who present with chest pain can lead to extensive investigations, which are costly and unnecessary in most cases. Pediatric chest pain also, has important functional consequences because it may result in restriction of activities and school absences.

Methods: The aims of this work is to review the available literature from recent years regarding non-traumatic chest pain in children, as well as to presentation a treatment protocol for pediatric patients presenting with chest pain.

Conclusion: Non-exertional chest pain in children and adolescents, in the absence of intercurrent illness or abnormal findings on physical examination is not due to cardiac pathology in the majority of cases. The most common cause of chest pain in children is idiopathic chest pain. An ECG or laboratory testing is necessary only in a small proportion of patients. Echocardiography or an exercise stress test are indicated only in a few cases. Primary care physicians should be aware of the differential diagnosis of chest pain in children. They should make an effort to rule out an organic cause, reassure the child and his family about the benign nature of the complaint, and look for red flag symptoms.

Disclosure of Interest: None declared

P037

P038

RECURRENT PERICARDIAL EFFUSIONS – AN UNEXPECTED CAUSE

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Introduction: Pericarditis is a rare cause of precordial pain in children and is most commonly due to acute viral pericarditis. However, in children after cardiac surgery, suspicion for post-pericardiotomy syndrome should be raised. We report here a case of recurrent pericardial effusions after pacemaker implantation, refractory to conventional therapy.

Methods: Case presentation: An 8 year-old boy presented 3 weeks after epicardial pacemaker implantation for congenital complete atrio-ventricular bloc to cardiology clinic. On the day before he was diagnosed with scarlet fever. Cardiac wise he was asymptomatic. Surprisingly, echocardiography revealed a large pericardial effusion. He underwent pericardial drainage and was started on Ibuprofen 10 mg/kg three times a day for a 4 weeks trial with a good result. One month after ibuprofen was stopped, he presented to A&E with symptoms of fever, chest pain, shortness of breath and rhinitis. Echocardiography showed again a moderate pericardial effusion and the patient was started on prednisone with a slow taper. However, 2 months later, he presented a third relapse of pericardial effusion with typical symptoms of pericarditis. Colchicine was added to his prednisone treatment.

Results: The boy could finally be weaned from all therapy and is now off symptoms since 8 months.

Conclusion: Post-pericardiotomy syndrome is an important cause of recurrent pericarditis in children and can appear in 2% of children after pacemaker implantation. In children, treatment strategies for recurrent pericarditis are not standardized and there are no clinical trials to guide therapy. However, this case report suggests that colchicine added to steroids gives good results in this patient population.

Disclosure of Interest: None declared

P039

PATHOLOGIC FRACTURE REVEALED SEVERE GENETIC LIPODYSTROPHY

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Introduction: Lipodystrophies are a group of disorders, acquired or genetic, characterised by localised or generalised loss of fatty tissue. Lamin dysfunction on *LMNA* gene mutations are a rare genetic cause of partial lipodystrophy. Described complications are diabetes mellitus, hypertriglyceridemia, hepatic steatosis, and premature osteoporosis.

Methods: The Male patient was born after an uneventful pregnancy. Parents were healthy and non-consanguineous. A pathological fracture of the right femur was diagnosed at 17 y.o. He had mental retardation, progeria, skin dyscheratosis, short stature [154 cm (< P3)], scoliosis and articular contractures. RX showed osteoporosis. Further analysis displayed hypovitaminosis D and K, increased GOT and GPT, hypercholesterolemia and hypertriglyceridemia; abdominal ultrasound showed hepatomegaly and liver steatosis, no splenomegaly. ECG showed infero-lateral Q waves. The echocardiography revealed left ventricular dilatation with severe systolic dysfunction (EF 25%). On cardiac magnetic resonance imaging a transmural late gadolinium enhancement in the inferolateral wall was found. The coronary angiography showed normal coronary arteries.

Results: In suspect of a syndromic lipodystrophy, exome sequencing was performed: de novo *LMNA* gene c.29T>C (p.T10I) mutation was found.

Conclusion: The pathologic fracture of the femur discovered a syndromic form of lipodystrophy. Genetic analysis revealed a point mutation on the lamin gene, *LMNA*. Up to now, only 3 patients with the same mutation have been described [Csoka et al.2004; Mory et al. 2008; Hussain et al. 2017], none of them presented pathologic fractures nor dilated cardiomyopathy. A detailed cardiologic evaluation should be performed at diagnosis of lipodystrophy because sudden death or heart insufficiency are important causes of mortality in these patients, as our case report demonstrates.

Disclosure of Interest: None declared

P040

UNILATERAL ABSENCE OF PULMONARY ARTERY IN MICRODELETION 22.Q11.2

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Introduction: Unilateral absence of pulmonary artery (UAPA) is a rare congenital cardiovascular anomaly caused by malformation of the 6th aortic arch during embryogenesis. UAPA may occur isolated (30%) or in association with other cardiovascular anomalies. The prevalence of isolated UAPA varies from 1 in 200,000 to 1 in 300,000. The diagnosis is often delayed into adult age due to possible initial asymptomatic presentation. Pulmonary hypertension, which develops early in the disease process, is seen in up to 25% of patients at first presentation and it's a poor prognostic factor.

Methods: We report the case of a 1-day-old fullterm newborn with breastfeeding difficulties. At physical examination we found a symmetrical hypoplastic newborn with facial dysmorphism consisting of low set ears, microstomia with limited mouth opening, microretrognathia without palatal abnormalities, periorbital fullness, prominent nose with hypoplastic nares. The remaining clinical examination resulted normal. In particular the sucking reflex was adequate, allowing a valid bottle-feeding alimentation. The breastfeeding difficulties were attributed to his anatomical features.

Results: Searching for associated malformations we performed an abdominal and cerebral ultrasonography and a chest x-ray, which did not show significant alterations. By echocardiography, the left pulmonary artery could not be visualized, an arterial duct was not found and a right-sided aortic arch was documented. Considering the facial dysmorphism in association with this type of cardiac malformation we now suspected a Microdeletion 22.q11.2. We started a therapy with prostaglandin E1, assuming the presence of an atypical arterial duct with duct-dependent left lung circulation. A fast track CT-Scan and a diagnostic cardiac catheterization were performed, both confirmed an agenesis of the left pulmonary artery, the left lung was perfused by collateral vessels arising from the aorta. We stopped the therapy with prostaglandin E1 and the arterial duct closed spontaneously. Genetic testing confirmed a microdeletion 22q11.2.

Conclusion: A rare congenital cardiac malformation, normally diagnosed at a mean age of 14 ± 2 years, was identified during the first days of life as a result of further diagnostic investigations because of suspected genetic syndrome. The early diagnosis allowed us to ensure a correct cardiac follow up in order to promptly treat possibly complications such as pulmonary hypertension.

Disclosure of Interest: None declared

P041

WHEN NEONATAL HISTORY FOLLOWED BY FAILURE TO THRIVE LEADS TO A RARE DIAGNOSIS

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Introduction: Childhood interstitial lung disease (chILD) is a heterogeneous group of rare chronic respiratory disorders, which interfere with gas exchange and growth. Chronic pneumonitis of infancy (CPI) is an entity specific to infancy, with a typical histological pattern of thickened of alveolar septae, enlarged airspaces, pneumocytes hyperplasia, focal intraalveolar macrophages and proteinaceous material. CPI can be associated with surfactant dysfunction disorders. Clinical presentation is non specific; diagnostic approach is based on clinical history, imaging studies, laboratory and genetic testing, the final step being an open lung biopsy.

Methods: Case report

Results: This late preterm boy was born by vaginal delivery at 35 2/7 weeks of a twin gestation. Adaptation was initially good but he developed respiratory distress syndrome (RDS) complicated with a right pneumothorax and pulmonary arterial hypertension. The respiratory evolution was unusual, the baby needing intensive and prolonged respiratory support. He also presented repetitive vomiting and feeding problems without a conclusive diagnosis. He was finally discharged home on day of life 46 where the first weeks were marked by fast breathing and coughing during feeds and sleep, general discomfort and frequent vomiting. At 3 months, he was hospitalized for failure to thrive, tachypnea and significant desaturations. Presenting history and chest X-ray were highly suggestive of chILD, confirmed by chest CT imaging showing diffuse ground glass opacities. Clinical improvement and proper weight gain occurred within days on oxygen therapy and nutritional optimization, awaiting further testing. Echocardiography, bronchoscopy with bronchoalveolar lavage, complete blood count, immunoglobulin levels, metabolic testing and genetic analysis for surfactant protein mutations or for brain-lung thyroid syndrome didn't reveal any abnormalities. Finally, a surgical lung biopsy at about 6 months led to the diagnosis of CPI. Oral hydroxychloroquine was started at 8 mg/kg/day for immunomodulation. The boy is now 21 months old, still on treatment and oxygen therapy but thriving and developing well.

Conclusion: ChILD should be suspected in any infant with chronic nonspecific respiratory symptoms and failure to thrive, taking into account neonatal history. Diagnosis is step wise, treatment and management should involve multidisciplinary care.

Disclosure of Interest: None declared

P042

A PALE AND POORLY BREATHING INFANT

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Introduction: A 3-m.o. infant presented with cough and dyspnea since 2 days, decreased oral intake and oliguria without fever. She was pale, uncomfortable, with perioral cyanosis during breastfeeding, tachycardia (169 bpm), tachypnea (70/min) and no cardiac murmur. Femoral pulses were weak, blood pressure normal (95/60 mm Hg). Liver was enlarged (3 cm from costal margin). Blood tests showed lactic acidosis (pH 7.30, lactate 3.8 mmol/l). Cardiomegaly on X-ray (index 0.65). In suspicion of cardiogenic shock, she received amine support and was intubated. Echocardiography revealed hypokinetic dilated left ventricle (LV) with severe systolic dysfunction (Left Ventricular Ejection Fraction-LVEF 15%) and possible abnormal origin

of the left coronary artery from the pulmonary trunk (ALCAPA). Angiography confirmed the diagnosis. MRI demonstrated LV sub-endocardial ischaemic lesions. After stabilisation, the left coronary artery was reimplanted to aorta under bypass. LV dysfunction was managed with 8 days ECMO and 6 days Left Ventricular Assist Device (LVAD). Systolic function improved with LVEF 60% at 1.5 months.

Discussion: Cardiogenic shock is challenging in paediatric emergencies, especially in infants. Hepatomegaly is an alarming sign. It requires cautious management of fluid resuscitation and amine support. Cardiomegaly suggests LV dilatation secondary to myocardial dysfunction or congenital heart defects (CHD). Plasma Troponin levels and echocardiography guide the diagnosis. Angiography and MRI can be useful. Etiologic workup includes search for metabolic, genetic, toxic and inflammatory causes. ALCAPA is a rare CHD (0.4%), usually revealed after few months of life, when pulmonary, subsequently coronary pressure decreases, leading to ischemic heart failure of variable intensity. Discomfort, pallor and sweating (angina pectoris) while feeding are typical clinical signs of this anomaly in infants. ECG shows myocardial ischemia. Ultrasound reveals left ventricular dysfunction and dilatation as well as abnormal origin of the left coronary artery. ALCAPA is confirmed by angiography and MRI. Treatment is surgical anastomosis of left coronary artery to aorta, prognosis is good if achieved early.

Conclusion: Infant respiratory distress can be of cardiac origin. ALCAPA is the most frequent cause of myocardial ischemia in infancy. Its clinical picture can be cardiogenic shock.

Disclosure of Interest: None declared

P043

PENILE INVOLVEMENT AS A RARELY REPORTED GENITOURINARY MANIFESTATION OF HENOC-SCHÖNLEIN PURPURA

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Introduction: Henoch-Schönlein purpura (HSP) is the most common immune-mediated systemic vasculitis in children, affecting the small vessels of the skin, the gastrointestinal tract, the kidneys, the joints and, rarely, the lungs and the central nervous system. Characterized by a purpuric rash, most commonly involving the buttocks and legs, HSP also frequently manifests with arthralgia, joint swelling, and gastrointestinal symptoms such as abdominal pain and hematochezia. However, genitourinary manifestations have seldom been reported.

Case report: We report the case of a previously healthy 5-year-old boy who presented with a painful edema and purple rash of the penis, without any previous trauma nor dysuria. He also complained of bilateral foot and ankle pain, with petechiae and purpuric rash on the legs. His right hand was swollen, but examination was otherwise normal. He had suffered from an acute obstructive bronchitis a week earlier. Urinalysis and blood tests including full blood count, coagulation studies and kidney function were normal. We diagnosed HSP with penile involvement, a rarely reported genitourinary manifestation of the disease.



Figure 1

Discussion: Genitourinary manifestations of HSP include acute scrotum, epididymo-orchitis, ureteritis, hematoma of the bladder wall, urethritis, hemorrhagic spermatic cord, priapism, and thrombosis of the spermatic veins. In the literature, a 13–22% >rate of scrotal edema is reported in boys suffering from HSP, sometimes as the only initial presentation of the disease. However, involvement of the glans,

foreskin and penile shaft has rarely been reported. Penile manifestations of HSP include thrombosis, priapism and cutaneous purpuric lesions. These notable complications result from the deposit of IgA complexes in the small vessels of an end organ with a complex microvascular architecture. Although rare cases may progress to end-stage renal disease, HSP usually has an excellent long-term outcome, and relies mainly on symptomatic treatment. Genitourinary manifestations do not appear to confer poor prognosis. There is no reliable recommendation concerning the indication for steroid treatment in case of genital involvement.

Conclusion: Extrarenal genitourinary manifestations of HSP mostly manifest as acute scrotum. Penile involvement has rarely been reported. No correlation is reported between genitourinary involvement and renal complications or poor prognosis of the disease. The indication for steroid treatment in such cases remains controversial.

Disclosure of Interest: None declared

P044

IDIOPATHIC PULMONARY HEMOSIDEROSIS: A RARE CAUSE OF IRON-DEFICIENCY ANEMIA

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Introduction: Idiopathic pulmonary hemosiderosis (IPH) is a chronic, rare disorder confined to the lung, which is commonly characterized by the triad of recurrent hemoptysis, diffuse parenchyma infiltrates on chest radiography, and iron-deficiency anemia, subsequently confirmed by the finding of hemosiderin-laden macrophages (HLMs) by bronchoalveolar lavage.

Methods: A 3-year-old child from a no-consanguineous marriage was followed by an iron deficiency anemia of unlabelled etiology. Hospitalized for the management of concomitant acute anemia with acute respiratory distress syndrome.

Results: The chest X-ray showed macronodular bilateral opacities, predominant in the para-hilar regions. The biological evaluation showed regenerative hyposidemic anemia. The analysis of bronchoalveolar lavage revealed the presence of HLMs. Cardiac echocardiography, chest CT scan, renal and haemostatic evaluation, and autoimmune and allergy evaluation excluded a cause secondary to intra-alveolar haemorrhage.

Conclusion: IPH is a serious and exceptional disease whose pathogenesis, prevalence and prognosis remain poorly understood. This is a diagnosis of exclusion. The evolution is progressively towards a chronic respiratory insufficiency interrupted by acute exacerbations. The Management must be early and adequate to prevent the occurrence of fatal complications.

Disclosure of Interest: None declared

P045

PRIMARY DISTAL RENAL TUBULAR ACIDOSIS: A CASE REPORT

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Introduction: The primary distal renal tubular acidosis (dRTA) is a rare genetic disorder. It is secondary to defective excretion of H⁺ by the cells of the collecting duct. dRTA is inherited, whether autosomal dominant or autosomal recessive.

Methods: A 20-month-old female infant from a second-degree consanguineous marriage. The death of the younger brother of an unknown cause is noted. The family survey revealed a notion of renal lithiasis in both paternal cousins.

Results: The clinical examination revealed signs of rickets associated with overall stunting. Biologically there is hypokalemia, metabolic acidosis, alkaline urinary pH with a urinary density of 1004 and hypercalciuria with a high Nordin index. An urine acidification test with ammonium chloride was performed, the urinary pH was always higher than 5.5, thus confirming the diagnosis. The renal ultrasound revealed bilateral medullary nephrocalcinosis.

Conclusion: dRTA is a rare genetic disorder with autosomal recessive or dominant inheritance. Once the diagnosis is made, the treatment is often simple based on the alcahanization. It must be correctly taken and continuously. The prognosis is related to renal failure.

Disclosure of Interest: None declared

P046

BIRTH ANOMALY OF THE PULMONARY ARTERY: AN EXCEPTIONAL PATHOLOGY

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Introduction: Birth defects of the pulmonary vessels (BDPV) often have clinical implications and are important to know and recognize. They have a significant mortality. This case is illustrated for its rarity.

Methods: A 40-day-old male infant with no pathological history. Hospitalized for respiratory distress with refusal of feedings.

Otherwise there is a good psychomotor development.

Results: The clinical examination revealed signs of cardiac decompensation (tachycardia, polypnea and hepatomegaly). The rest of the physical examination was normal. The chest X-ray showed cardiomegaly with large vascular hiles. The Echocardiography showed pulmonary arterial hypertension (PAH). The chest CT angiography showed a birth anomaly of the right pulmonary artery of the ascending aorta. The rest of the malformation assessment was without abnormalities.

Conclusion: Birth defects of the pulmonary vessels is an exceptional pathology. The chest CT angiography has become an essential examination for finding an etiology. The treatment is medical-surgical.

Disclosure of Interest: None declared

P047

TRANSIENT ARTERIAL HYPERTENSION INDUCED BY GONADOTROPIN-RELEASING HORMONE AGONIST TREATMENT FOR PRECOCIOUS PUBERTY

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Introduction: Gonadotropin-releasing hormone agonists (GnRH agonist) are used as a safe and effective treatment of the precocious puberty. Triptorelin is one of the long lasting GnRH agonist, which suppresses reversibly the pituitary-gonadal axis.

Methods: We hereby report the case of a 10-year-old girl with precocious central puberty, who, during her treatment with triptorelin, developed an asymptomatic stage II hypertension. Initial workup showed no renal, thyroid or electrolytes abnormalities. The renal ultrasound showed no parenchymal disease and no renal artery stenosis. Echocardiography and ocular funduscopy were normal. Hypertension (stage II) was confirmed with ambulatory blood pressure monitoring (ABPM). Literature review showed a few cases of hypertension secondary to GnRH agonists, improving with endocrine treatment cessation. Therefore antihypertensive treatment was not started immediately in our patient.

Results: Indeed, after completion of her treatment with triptorelin, we observed a complete normalization of her blood pressure, without any medication, and this was confirmed with ABPM.

Conclusion: This young presented with hypertension during treatment with GnRH agonist. The complete normalization of her blood pressure after stopping triptorelin, together with a negative workup strongly speaks for a causal effect of her endocrine treatment. The pathophysiological mechanisms in this setting may be explained by estrogen depletion.

Disclosure of Interest: None declared

P048

HEME-PIGMENT INDUCED ACUTE KIDNEY INJURY

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Introduction: Acute kidney injury (AKI) requiring renal replacement therapy (RRT) in children without preexisting conditions is rare. In outpatients post-infectious glomerulonephritis, hemolytic uremic syndrome (HUS), volume depletion and drug toxicity are common causes of AKI; with exception of HUS, they rarely need RRT.

Case presentation: A 3-year old, previously healthy girl, presented with vomiting, dark urine and jaundice after a one-week history of febrile respiratory tract infection. Laboratory tests revealed hemolytic anemia (Hb 99 g/l, LDH 2237 U/l, bilirubine 112 µmol/l, direct antiglobuline test (DAT) ++) without fragmentocytes and normal

platelet count (251 G/l). Serum creatinine was 103 µmol/l (eGFR 42 ml/min/1.73 m²) and urea 15.0 mmol/l. At presentation, urinalysis showed active sediment with red blood cells (12500/µl), protein/creatinine 634 g/mol, albumin/creatinine 131 g/mol. Despite subtle treatment with fluid management and frusemide, the patient became anuric within 12 hours of admission. In absence of history with gastrointestinal complaints and normal platelet count as well as a positive DAT, hemolytic anemia with heme-pigment nephropathy was suspected. Renal biopsy showed signs of tubular damage without features of thrombotic microangiopathy. Additional laboratory tests revealed no evidence of infectious cause (Cytomegalo-, Epstein-Barr and parvovirus B19, mycoplasma, antistreptolysin-O) and C3-complement was normal. Further investigation finally revealed a positive Donath Landsteiner Test leading to the diagnosis of Donath-Landsteiner hemolytic anemia. Peritoneal dialysis was initiated due to persistent anuric renal failure (eGFR 13 ml/min/1.73 m²). By day 5 of dialysis renal function improved and returned to normal by day 14. At latest follow up renal function was normal and no recurrence of hemolytic anemia occurred so far.

Conclusion: Solid data about prevalence and incidence of AKI in pediatric out-patients is lacking mainly because of inconsistent definition and probably underreporting. Differential diagnosis in pediatric AKI is broad, including prerenal, intrinsic (vascular, glomerular and tubulo-interstitial) and post-renal causes. Careful work-up is crucial in order to rapidly offer adequate treatment. Heme-pigment nephropathy is a very rare cause of AKI, in which tubular obstruction, direct proximal tubular epithelial cell injury and vasoconstriction can lead to renal failure.

Disclosure of Interest: None declared

P049

CARDIAC FIBROMA AS AN UNUSUAL CASE OF CHEST PAIN IN A CHILD

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Introduction: Less than 5% of chest pain in children is cardiac in origin, yet this complaint still represents one of the main reasons children are referred to pediatric cardiologists. A systematic approach including a pertinent history, thorough clinical examination and ECG can identify those children who require further testing. Red flags for cardiac causes of chest pain include, among others, the occurrence on exertion, the association with palpitations and abnormal findings on physical examination or ECG.

Methods: We present a case of a patient with a cardiac fibroma that caused recurrent chest pain and palpitations.

Results: A 9-year-old boy presented with a one-month history of recurrent chest pain associated with palpitations, while playing ice hockey. He consulted the emergency department of a peripheral hospital while having a novel episode, and was found to have a heart rate of 250/min. He was hemodynamically stable, and the physical examination did not reveal any other abnormal findings. A 12-lead-ECG showed wide-QRS tachycardia, which resolved spontaneously, and the patient was transferred to our institution for further evaluation. A transthoracic echocardiogram showed a left ventricular mass at the apex and a cardiac MRI confirmed the diagnosis of a cardiac fibroma. The patient underwent surgical excision of the tumor without complications. Antiarrhythmic therapy with amiodarone was started at the moment of diagnosis, and maintained for 3 months after surgery. It was discontinued after an electrophysiological study demonstrated no inducible ventricular tachycardia. The patient has remained asymptomatic at a 3-year follow-up with normal left ventricular systolic function.

Conclusion: Primary cardiac tumors in children are rare and most often benign. Fibromas are well-defined solitary masses of fibroblasts that do not regress and can invade the cavity space, causing ventricular outflow tract obstruction. They are strongly associated with dysrhythmias and usually require surgical excision. If the tumor is completely resected, patients will have disease-free survival and remain asymptomatic.

Disclosure of Interest: None declared

P050

DIFFERENTIATING CEREBRAL MALARIA FROM BACTERIAL MENINGITIS IN RESOURCE-LIMITED COUNTRIES: A LITERARY REVIEW

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Introduction: Due to the similarity of their clinical presentation, differentiating cerebral malaria (CM) from bacterial meningitis (BM) is challenging, particularly in malaria-endemic regions. The aim of this study was to review the literature in order to identify potential symptoms, signs and routine biological features that could guide caretakers in discriminating CM from BM in rural settings of developing countries, thus reducing the burden of both diseases.

Methods: A title and abstract search was conducted using multiple Internet databases, reviewing the available literature on diagnostic approaches for CM and/or BM in developing countries.

Results: 23 articles were selected. The majority of studies were carried in sub-saharian Africa. In malaria-endemic countries, co-infections with severe malaria and invasive bacterial infections are not uncommon (6.4%). Impaired consciousness, prolonged seizures, prostration and meningeal irritation signs were not useful in differentiating CM from BM. Malarial retinopathy was the only distinctive sign that could potentially discriminate between CM and BM, although not systematically present in CM. A leukocyte count of >100/uL in cerebrospinal fluid (CSF) was highly suggestive of BM. In the two studies where CSF leukocyte counts of >10/uL were found in CM children without evidence of BM, neither specified if all patients had undergone bacterial cultures or antigen testing, nor if antibiotherapy was used before lumbar puncture was performed.

Conclusion: Retinal examination by trained caretakers may be useful in differentiating CM from BM in malaria-endemic regions. An elevated leukocyte count in CSF is the most useful laboratory testing to discriminate between both diseases particularly when the leukocyte count is >100/uL in the presence of negative malarial tests. If the leukocyte count is low, repeating the lumbar puncture after 24 hours while continuing empirical therapy for both diseases may be useful.

Disclosure of Interest: None declared

P051

PNEUMOCYSTIS JIROVECI PNEUMONIA IN A 4-MONTH-OLD INFANT: THE ICEBERG TIP.

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Introduction: *Pneumocystis jirovecii* pneumonia (PJP) is an opportunistic infection occurring only in patients with underlying diseases. In children, the main associated conditions are infection with the human immunodeficiency virus (HIV), oncological diseases and immunosuppressive or immunomodulatory drugs. Otherwise, primary immunodeficiency should be actively searched. We report the case of a 4-month-old infant who developed PJP, revealing a severe combined immunodeficiency disorder (SCID).

Methods: A previously healthy 4-month-old infant was admitted to the pediatric intensive care unit for an acute respiratory distress syndrome, preceded by progressive fatigue over the past few days and decreased food intake, without any infectious symptoms. Chest CT revealed a homogeneous bilateral ground-glass infiltrate and *Pneumocystis jirovecii* was found in endotracheal secretions. She was treated with trimethoprim-sulfamethoxazole and corticosteroids with a good clinical course. The finding of an opportunistic infection should trigger the search of an associated medical condition.

Results: Our patient was found to have a T-B-NK+ SCID with a double mutation on the *RAG2* gene that encodes a protein involved in the initiation of V(D)J recombination of T and B cell receptors. Protective measures were introduced, including a negative pressure room, monthly infusions of immunoglobulins, prophylactic antibiotics such as fluconazole and trimethoprim-sulfamethoxazole and passive immunization with palivizumab. After two months of hospitalization, she received a myeloablative chemotherapy followed by hematopoietic stem cell transplantation from an HLA-matched cord blood. The clinical course was marked by failure of the transplant after 4 weeks. A second stem cell transplant of maternal origin was successfully performed.

Conclusion: *Pneumocystis jirovecii* pneumonia in a previously healthy child involves the active search of a primary immunodeficiency. In case of an underlying severe combined immunodeficiency disorder, aggressive, early management, including hematopoietic stem cell transplantation, is the standard of care.

Disclosure of Interest: None declared

SEVERE PERTUSSIS IN A NEONATE – ANTICIPATE AND TREAT HYPERLEUKOCYTOSIS EARLY

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Introduction: Hyperleukocytosis, caused by pertussis toxin, is rare but life-threatening. In young infants this may lead to increased viscosity and vascular occlusion of vital organs, especially the lungs. Leukocyte depletion via exchange blood transfusion is an established treatment of hyperleukocytosis. The most effective prevention of pertussis in infants less than 3 months is immunization of women during second or third trimester in every pregnancy.

Methods: A 24 day old female neonate presented with a one-week history of afebrile rhinitis, paroxysmal cough and “turning dark” as reported by her mother. Both parents had a cough for 4 weeks. She appeared unwell with tachypnoea, paroxysmal coughing and apnoeic spells.

Results: Pertussis was suspected and Azithromycin started. PCR from nasopharyngeal swab was positive. Day two the patient developed severe respiratory distress requiring oxygen. On Xray right upper lobe atelectasis. Laboratory: hyperleukocytosis ($52.5 \times 10^9/L$; $25.7 \times 10^9/L$ lymphocytes), hypercapnia (pCO₂ 7.79 kPa) and normal CRP (<5 mg/L). Further increase of leukocytes to $63.5 \times 10^9/L$ 12 hours later. Exchange transfusion was performed after Intubation and ventilation. Significant leukocyte decrease to $22.5 \times 10^9/L$ was achieved by the next day. Gradual improvement but oxygen supplementation for total of three weeks was necessary. The mother had not been immunized against pertussis during pregnancy.

Conclusion: Patients with severe pertussis and hyperleukocytosis can benefit from early blood exchange transfusion to prevent respiratory failure. Repeated full blood count analyses in infants with pertussis help to detect hyperleukocytosis and anticipate the dynamics. Pertussis is not an exclusive childhood disease. It should always be part of the differential in adults with prolonged coughing. Recent intervention studies have shown that booster vaccination during pregnancy is key to protect young infants, the most vulnerable population. Gynaecologists should always take this intervention into their routine care bundle when consulting pregnant women.

Disclosure of Interest: None declared

P052

TENOSYNOVIAL GIANT CELL TUMOR IN A PATIENT WITH NOONAN SYNDROME: WHEN SURGERY IS NOT AN OPTION

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Introduction: Noonan syndrome (NS) is a genetic disease caused by gain of function variants in genes in the Ras-MAPK pathway. Tenosynovial giant cell tumor (TGCT) is a rare benign proliferative disorder of the synovial membrane of unknown origin. Surgery is the mainstay of treatment in patients with TGCT, but local failure is frequent and relapses up to 50%. Patients who are not operable or where operation may be morbid, can be treated with tyrosine kinase inhibitors (TKI), as imatinib, which has showed tumor regression or stable disease in most patients [1, 2]. NS has been associated with other multiple giant cell lesions, in particular with the giant cell granulomas of the mandible, or cherubism (3). To date, only few cases of TGCT have been described in association with NS, and any of these has been treated with TKI (3–5). We report a case of a patient affected by NS who has been diagnosed with multiple TGCT.

Methods: Case report.

Results: A 14-year-old boy affected by NS with SOS1 mutation and cherubism presented with invalidating multiple tenosynovitis in both ankles. He was initially diagnosed with juvenile idiopathic arthritis (JIA) and treated with intra-articular steroid injections, sulfasalazine and etanercept. Unfortunately, none of these treatments was effective; furthermore, wrists and knees synovitis progressed. Because of this atypical evolution, a multiple TGCT was suspected and a synovial biopsy performed, showing villous synovitis, giant cells and previous hemorrhage. The diagnosis of TGCT was then confirmed. A multidisciplinary meeting including pediatric rheumatologists, orthopedic surgeon and oncologist was done in order to discuss about

P053

systemic treatments, as surgery could not be undertaken (multiple localizations, nonhealing surgical wounds and lymphedema after the biopsy, high risk of relapse). It was decided to prescribe a TKI, in particular the imatinib. The patient's health insurance was contacted in order to take charge of the treatment and the process is still ongoing. **Conclusion:** The TGCT should be considered as alternative diagnosis in patients with JIA who do not respond to conventional treatment. This is particularly true in patients affected by NS, where an association with multiple giant cell lesions has been reported. Our clinical case shows the importance of the multidisciplinary collaboration in complex situations, where the specific evaluation of every expert allows taking decisions even in rare or not described cases.

NaCHwuchs Prize: I wish to apply
Disclosure of Interest: None declared

P054

ANALYSIS OF THE COMMUNICATION STRATEGY AND THE INFORMATION GIVEN TO THE PATIENT IN PEDIATRIC ALLERGOLOGY

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Introduction: Communication and learning are the cornerstones of the management of allergic diseases. Our research aimed to identify and analyze the information needs of parents of patients with allergic diseases.

Methods: The study is based on the thematic analysis of transcripts of semi-structured interviews conducted with fifteen parents of allergic patient consulting at the Hospital Children's Clinic. The interviews were conducted with the help of a questionnaire aimed at assessing satisfaction with the parents' communication and information needs. The result of this field study, the content and operation of a consultation summary program was defined and presented as a decision tree. The work was done within the frame of a Master's degree in medicine.

Results: Parents are satisfied with the communication with their doctor, who adequately meets their expectations with sufficient and understandable information. Understanding the decisions of care such as the use of treatments, the means of prevention as well as the prognosis of the child's disease are the most requested information, which are also sought in parallel on the internet.

Ignorance of the reliability criteria of websites is a limiting factor. All parents would welcome receiving a summary of the consultation. **Conclusion:** Two obstacles limit the good transmission of information: understanding medical language and forgetting information. Lack of identification of reliable websites hinders access to complete documentation. Results presented here are modeling the ideal summary of the content of the consultation to parents and their children. Further objectives will be to reinforce the communication techniques, to help the patients in their research of information and to develop a summary of the consultation. In practice, this summary will be a reminder and will serve as a relay to ensure continuity of care.

Disclosure of Interest: None declared

P055

A STIFF NECK AND FEVER – ALWAYS THINK OF A RETROPHARYNGEAL ABSCESS

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Introduction: A retropharyngeal abscess is typically seen in children between 2 and 4 years of age and usually preceded by an upper respiratory tract infection. It is commonly caused by a polymicrobial infection (Strept. pyogenes, Staph. aureus & respiratory anaerobes). Clinical findings may include fever, dysphagia, drooling, torticollis, respiratory distress or trismus. The diagnostic test of choice is a computed tomography (CT) with intravenous contrast. Nevertheless, airway management has to be the first priority. Children without signs of airway compromise and only small abscesses <2.5 cm² can receive empiric treatment with broad-spectrum antibiotics for 24–48 hours. Surgical drainage is suggested for patients with large abscesses or with failure to respond to parenteral antibiotics. The complications of retropharyngeal abscesses are rare, but potentially fatal (Lemierre's syndrome, jugular vein thrombosis, mediastinitis...).

Methods: Case report: A 17-month-old girl was sent to our hospital with suspected meningitis because of fever, rhinitis, vomiting and

inability of fluid intake. Clinical examination showed a bad general condition with GCS 11, tachycardia, prolonged capillary refill, enlarged tonsils and swollen cervical lymph nodes on the left side with torticollis. Cerebrospinal puncture showed 2 mononuclear cells and blood test revealed massively elevated inflammation parameters. Sonographically, an abscess formation was detected within the enlarged lymph nodes. Therefore, an empiric antibiotic therapy with amoxicillin/clavunate was initiated. However, after 2 days there was no improvement of the torticollis but progressive drooling and inspiratory stridor. A CT scan revealed a retropharyngeal abscess (3.1 × 1.3 × 1.6 cm). After the abscess incision, the antibiotic treatment was continued orally for 10 days with a rapid clinical improvement.

Results: –

Conclusion: – Neck stiffness and bad general condition may have other causes than meningitis. – With the red flags toddler, torticollis and fever, always think of a retropharyngeal abscess as first differential diagnosis. – A superficial abscess formation does not exclude other reasons for torticollis. – Continued fever and lack of improvement despite treatment should lead to further evaluation for deep neck infection with a CT scan. – Empiric therapy of a retropharyngeal abscess includes broad-spectrum antibiotics. In case of a large abscess or lack of response to empiric antibiotic therapy, a surgical drainage is necessary.

Disclosure of Interest: None declared

P056

RARE IMMUNE COMPLICATION AFTER MENINGOCOCCAL SEPTIC CHOC

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Introduction: Invasive meningococcal disease can be devastating in children and potentially fatal. In Switzerland, 40 cases of meningococcal infections were reported in 2015, with a mortality rate around 8%. Although 6–11% of meningococcal infections are followed by immune complex associated complications (ICAC), it has rarely been described in the literature, and is poorly recognized by physicians.

Methods: Case report: An 11-year-old girl was hospitalized for septic choc caused by severe meningococcal type C infection. As she was recovering under ceftriaxone treatment, she developed on day 4 recurrence of fever, new erythematous non-purpuric rash, ankle monoarthritis, along with a raise in inflammatory parameters. After exclusion of life-threatening complications, this association suggested an ICAC. We completed the investigations with a cardiac ultrasound, which showed asymptomatic pericardial effusion. Anti-inflammatory treatment with ibuprofen (30 mg/kg/day) was initiated but clinical response was incomplete. On day 8, a 5-day course of prednisone (50 mg/day) was initiated, resulting in complete arthritic relief.

Results: Discussion: ICAC is a poorly recognized subacute complication of meningococemia. It is caused by the deposit of immune complexes and can present as arthritis, vasculitis, pericarditis, pleuritis, episcleritis, and rarely nephritis. It usually starts on day 4–10 following onset of infection, with recurrence of fever and secondary raise in inflammatory markers. Arthritis is the most common presentation (involving knees>ankles>elbows) and does not always require treatment. Vasculitic rash is frequent, and sometimes misdiagnosed as a penicillin allergy. Pericardial effusion usually requires steroid treatment to prevent cardiac tamponade. Our patient showed several criteria of ICAC, including pericardial effusion. Incomplete response after 2 days of ibuprofen motivated the introduction of prednisone with full recovery.

Conclusion: ICAC should be suspected as a subacute complication of meningococemia when a recovering patient presents a secondary fever and/or increase in inflammatory parameters, after ruling out ongoing sepsis, new infection or other cause of systemic inflammation. A careful attention must then be paid to identify ICAC symptoms such as arthritis, vasculitis, pericarditis, episcleritis and pleuritis.

Disclosure of Interest: None declared

P057

SHINGLES WITHOUT CHICKENPOX. A CASE REPORT

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Introduction: Herpes Zoster (HZ); is uncommon in children and rare in infants. In immunocompetent children; the cases have been attributed to 1 of 3 primary exposures: intrauterine exposure to the Varicella-Zoster-Virus (VZV), exposure due to vaccination and postnatal exposure to wild-type VZV.

Methods: We report a case of an immunocompetent child of 32 months old who developed HZ with secondary infection to *S. aureus*. No history for chickenpox, but exposure to VZV pre-partum and during the neonatal period was found.

Results: A 32 month-old girl, presented with a 5-day history of vesicular rash that started at the right elbow. No other symptoms. First evaluation seemed as a primary herpes infection with a bacterial co-infection (mother had a lip blister days before). Swabs were sent for bacterial and viral identification. Initial treatment with Co-amoxicillin and Acyclovir was started for 24 hours until reception of the results: Herpes Simplex Virus (HSV) 1 and 2 Negatives; *S. aureus penicillin resistant*. Antiviral therapy is stopped. The spread continued distally and unilaterally thru the right forearm, hand and 3th 4th and 5th fingers. Pain within the lesions increased. At 5th day, dermatological advice was demanded and the diagnosis established was Zoster impetiginized. She was admitted for pain control and follow up. Advice from the Pediatrics infectiologist was undertaken. VZV Polymerase chain reaction (PCR) was highly positive. History of exposure to Varicella intrauterine and post-partum was reported. She was breastfed, mother has a positive history for chickenpox. No clinical signs of Varicella developed after exposure. The lesions followed the dermatomes: Cervical 7 (C7); Cervical 8 (C8); and Thoracic 1 (T1). Pain was controlled with paracetamol, itchiness with histamine H1 antagonist and *Staphylococcus aureus* infection with Co-Amoxicillin 50 mg/kg/dose 3 times a day for 7 days. The patient was discharged and vesicular involution required up to a month to heal. An advice from the immunologist was demanded and a follow up and investigation for an immunosuppression was scheduled.



Figure 1

Conclusion: Zoster Infections are not frequent in toddlers, but is not excluded. A medical history with exposure needs to be clarified in every patient with suspected lesions for Shingles, and keep it in mind as a differential diagnosis. Also An immunologic status of the patient with Zoster, should be investigated in children younger than 10 years old.

Disclosure of Interest: None declared

P058

DOCTOR, MY DAUGHTER IS CONSTIPATED AND SHE DOESN'T WANT TO SIT ANYMORE...

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Introduction: Case 1: 1 y.o. girl had 3 days of abdominal cramps, diagnosed as constipation and treated with enema. 4 days later abdominal pain increased and she refused to sit. She was discharged with laxatives. 2 weeks later she would not sit, crawl or stand, her general condition was poor. Spine and pelvis were painless, neurological examination was normal. Work-up revealed slight inflammation (ESR 28 mm/h, CRP 5 mg/l, thrombocytes 403 G/l). MRI showed L2–L3 spondylodiscitis (SD). Throat swab was positive for *Kingella kingae* (Kk), blood culture was negative. Screenings for Tuberculosis (Tb), Brucellosis, Strepto A were negative. PCR on vertebral biopsy was negative for Tb and Kk. She had completely recovered after 7 days of IV Co-amoxicillin followed by oral treatment (7 weeks) while waiting for biopsy results. Case 2: 2 y.o girl consulted after 3 days of pain in thigh, limp and coryza. Laboratory results were unremarkable, X-rays were normal. She was discharged with painkillers. 4 days later she presented with increasing pain and refusal to walk and sit. She had fever, impaired general state and constipation

for the last 5 days. She had pain on sacrum and pelvis compression and on lumbar flexion. Lower limbs and neurological exam was normal. Work-up revealed CRP 39 mg/l, ESR 60 mm/h and thrombocytes 352 G/l. MRI showed acute sacral osteomyelitis. Blood culture was negative but Kk was positive in throat swab. She was treated with 14 days IV followed by 14 days oral Co-amoxicillin. She had totally recovered at discharge.

Results:

Discussion: Vertebral osteoarticular infections (VOAI) are rare in children (incidence 1/32500 for SD, 2/100000 for vertebral osteomyelitis). In toddlers younger than 4 y.o. the main organism responsible for osteoarticular infections is Kk, which often colonizes superior airways and may cause haematogenous infection. Clinical presentation is often unspecific, delaying diagnosis. Precocious diagnosis of VOA prevents growth anomalies and joint mobility impairment. Early detection of VOA is crucial to avoid neurological complications. In toddlers limping or refusing to walk, crawl or sit without history of trauma, spine and pelvis should be carefully examined. Constipation is often associated. Diagnosis is confirmed by MRI and throat swab PCR for Kk. Due to Kk's low virulence, biopsy cultures are often negative.

Conclusion: It is mandatory to consider Kk VOA in children who refuse to walk, sit or crawl. Kk VOA diagnosis is made with throat swab PCR test and MRI.

Disclosure of Interest: None declared

P059

A SEPTIC INFANT FROM INFECTED MOTHER

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Introduction: A previous healthy 6 weeks-old Eritrean boy was referred for one-day fever (38°C), ill-appearance, and grunting, coughing for few days. He was term born in Switzerland, BW 3250 g. Mother received ceftriaxone a week before for Méti-S *Staphylococcus aureus* (MSSA) breast abscess. She maintained breastfeeding. At admission, he was hypotonic, pale and mottled, with respiratory distress. Temperature was 38.2°C, cardiac and respiratory rate were 200 and 76/min respectively, BP 109/52 mm Hg, pulse oximetry 95% on ambient air, refill time was >4 seconds. Blood gas and CSF were normal, CRP 202 mg/l, WBC 26.7 G/l (27% band form, 41% neutrophils) and platelets 823 G/l. X ray revealed left inferior lobar pneumonia. Blood culture grew for MSSA. Echocardiography was normal. After fluid resuscitation, he received two weeks antibiotherapy (ceftriaxone, switched to flucloxacillin on day 5). Breast milk culture was negative and breastfeeding was pursued. Radiologic follow-up identified left large bullae on day 4, and later pneumatocele, which spontaneously resolved in 5 months.

Methods:

Discussion: *Staphylococcus aureus* (*S. aureus*) is a common human pathogen, responsible for soft tissue, bone and joint, endocardia or pulmonary infections. They are nosocomial or community-acquired. In infants pneumonia and/or clinical sepsis are the most presenting picture. *S. aureus* is a classical aetiology of pneumatocele, which is a thin-walled gas-filled cavity within lung parenchyma. Pneumatocele can complicate infants' and young children's pneumonia. It usually regresses spontaneously in weeks or months, and rarely require surgical intervention or drainage. Main complication is pleural rupture inducing tension pneumothorax. Pathogenesis remains unclear: inflammation could form a check-valve obstruction, inducing air trapping. Majority occurs in infants probably because Kohn's pores (allowing collateral ventilation) are not complete.

Conclusion: *S. aureus* invasive disease remains a cause of necrotising and bullous pneumonia in infants. Feeding mother with breast abscess can transmit the bacteria. Echocardiography must be performed. Pneumatocele is a rare complication of pneumonia in infants and children, most often with spontaneous recovery.

Disclosure of Interest: None declared

P060

CHICKENPOX: A BENIGN CHILDHOOD DISEASE?

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Introduction: Central nervous system (CNS) vasculitis is an inflammatory brain disease defined by the presence of neurological or psychiatric manifestations in previously healthy children. In contrast with the primary forms, an underlying disease can be identified in secondary forms. Infections are the most frequent underlying cause and Chickenpox among these plays the main role.

Methods: Case report

Results: We report a 6-year-old girl with a progressive left sided hemiparesis with involuntary and uncoordinated movements of the ipsilateral leg and arm as well as a mild aphasia since 1 week. Further clinical examination including blood pressure was normal. There was no history of trauma. She reported a mild manifestation of varicella zoster virus (VZV) infection 3 months prior to this event. In the past she had been healthy and vaccinated according to the Swiss national vaccination plan. Cerebral MRI demonstrated a stenosis of the M1 segment in the right middle cerebral artery with perivascular enhancement, consistent with vasculitis. This finding could also be visualized in the transcranial doppler ultrasonography. An autoimmune etiology was ruled out. The cerebrospinal fluid PCR was positive for VZV. VZV serology showed positive IgG and IgM, consistent with the recent VZV infection. A therapy was initiated with high-dose iv Methyl-Prednisolone 30 mg/kg/day for 5 days, iv Acyclovir 60 mg/kg/day for 10 days and oral Acetylsalicylic acid 5 mg/kg/day. The treatment was continued orally after discharge with clinically complete recovery in few weeks. In the follow up after 1 month a new vascular lesion of the right anterior cerebral artery without clinical manifestation was found. Therefore a second course of iv corticosteroids was administered for 3 days. Thereafter the patient remained asymptomatic and a progressive regression of the radiological findings was observed. As a consequence the steroid therapy was progressively reduced until they its interruption.

Conclusion: VZV infection complications, such as bacterial skin and soft tissue infections, are well known. However, CNS complications and particularly VZV associated vasculitis is rare but carries a high morbidity. Therapy of these cases is difficult as evidence for optimal care is scarce. Universal VZV vaccination in early childhood could contribute to reduce this complication.

Disclosure of Interest: None declared

P061

GRISCELLI SYNDROME TYPE II: KNOWING HOW TO THINK ABOUT IT

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Introduction: Griscelli syndrome 2 (GS2) is a rare autosomal recessive disorder characterized by partial albinism with variable immunodeficiency. It is due to a mutation of the Rab27A gene located on chromosome 15q21.

Methods: A 9-month-old male infant from a no consanguineous marriage. One infant died in his siblings. Hospitalized for recurrent bronchopulmonary infections.

Results: The clinical examination revealed a particular morphotype associating gray-silver hair, depigmented eyebrows and a clear complexion. In addition, there is good growth and weight with normal psychomotor development. Biological assessment, blood smear, and baseline immune status were normal. The Examination of the capillary structure under the light microscope showed the presence of large pigmentary clumps in the hair sheath of irregular and scarce distribution.

Conclusion: GS2 is a rare genetic disease. The diagnosis of certainty can be done by molecular biology. It allows genetic counseling and allows to program a bone marrow transplant, the only cure. The antenatal diagnosis is feasible.

Disclosure of Interest: None declared

P062

AN OBSTRUCTIVE BRONCHITIS LIKE NO OTHER: A CASE OF INVASIVE GROUP A STREPTOCOCCAL DISEASE

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Introduction: The worldwide incidence of invasive group A streptococcal (iGAS) disease has been increasing over the last two decades. Co-infection of GAS with viral pathogens is common and may delay the diagnosis. We describe such a case.

Methods: Case report

Results: A healthy 3-year-old girl presented to the emergency department for upper respiratory symptoms with a day of fever and dyspnea. Examination showed signs of respiratory insufficiency with

oxygen saturation of 82%. Auscultation revealed diffuse crackles. An urticarial rash was noted. A chest X-ray revealed a possible retro-cardiac opacity. Laboratory analysis showed elevated inflammatory parameters with a leucocytosis and a left shift. A blood culture was drawn. Treatment with inhaled salbutamol improved the respiratory status. Rapid testing for respiratory syncytial virus (RSV) was positive. She was hospitalized for oxygen therapy with a working diagnosis of obstructive bronchitis with an elevated inflammatory syndrome. Ten hours after hospitalization, she was transferred to the intensive care unit (ICU) for altered mental status with signs of shock and later developed multi-organ failure. Throat rapid testing was positive for GAS, which also grew in blood cultures. She was started on vasopressors and parenteral antibiotics with a diagnosis of toxic shock syndrome. She fully recovered after 30 days. Because of the severity of the episode and a prior episode of pneumonia, an immunological workup searching for a primary immunodeficiency was performed and returned normal.

Conclusion: GAS bacteremia is a rare disease, arising in 2–3/100,000 patients/year. Its association with GAS pharyngitis is also rare, occurring in 0.3% of patients. Epidemiological studies support the idea that a preceding viral infection increases the risk of iGAS disease, by inducing changes in bacterial adherence and reducing cell mediated clearance. It is well described as a co-infection of Varicella Zoster, as well as influenza virus. High fever and elevated inflammatory markers are typical but non-specific findings. A scarlatiniform rash followed by desquamation is often noted. This case report reminds us of the importance of keeping the differential diagnosis open despite an initial presentation compatible with a respiratory viral illness, especially during the winter season when the prevalence of such illnesses is high. Prompt recognition and treatment of iGAS is essential in preventing morbidity and mortality.

Disclosure of Interest: None declared

P063

A RARE BUT FEARED PATHOGEN IN A CASE OF NEONATAL MENINGITIS

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Introduction: *Neisseria meningitidis* is responsible for severe meningitis and sepsis, especially in young children and adolescents. In Switzerland the incidence of invasive meningococcal disease is higher in infants compared to adolescents (8.2 vs. 1.5 per 100'000). This group has also the highest mortality. Neonatal *N. meningitidis* associated meningitis (NMM) is a rarity. Acquisition is thought to be via the maternal genital tract or postnatally from other caretakers who are carriers.

Case Report: A 21 day old male term infant was assessed at our pediatric emergency department presenting with fever and poor feeding since the last 12 hours. Pregnancy and perinatal period were unremarkable. GBS screen was negative. He was febrile (38.9 °C), in a reduced general condition and irritable. On examination normal skin, normal capillary refill time, no respiratory distress and normotensive. The anterior fontanelle was full but not bulging. Laboratory investigations showed a normal full blood count, CRP 12 mg/L and Procalcitonin 2.53 µg/L. The urinalysis was normal. Cerebrospinal fluid (CSF) was taken before starting empiric antibiotics (Amoxicillin and Amikacin) and showed a white cell count of 49/µL (90% polynuclear, 10% mononuclear), glucose 3.8 mmol/L (CSF/serum glucose ratio 0.59), protein 0.61 g/L. We commenced regular iv. hydration. After 4 hours we had the CSF PCR result: positive for *N. meningitidis*. Subsequently treatment was changed to Ceftriaxone for a total of 10 days. The CSF and blood cultures grew *N. meningitidis* (serotype not yet specified, sensitive for Ceftriaxone). Chemoprophylaxis was administered to the family members. The further course was uneventful, without any signs of neurological or audiological impairment at time of discharge. Follow-up is planned.

Conclusion: The presentation of NMM differs from that in children and adolescents. Neonates may only present with recent fever and poor feeding. Inflammatory markers may be normal or just slightly elevated. Procalcitonin may give an early clue to an invasive bacterial infection. CSF may be normal or show just a mild pleocytosis. This again supports the obligation of performing a full septic work-up in this age group and start empiric antimicrobial therapy early to prevent sepsis and rapid deterioration.

Disclosure of Interest: None declared

P064

A PRE-TRAGIAL ACTINOMYCOSISM. Rohr^{1,*}, N. Wagner¹, K. Posfay-Barbe¹¹Maladies infectieuses pédiatriques, HUG, GENEVE, Switzerland

Introduction: The genus *Actinomyces* is a group of gram positive bacilli, part of the commensal microbiota of oral, gastro-intestinal and genito-urinary tracts. Despite their low virulence, some species are increasingly associated with infections and especially in the Ear, Nose and Throat (ENT) area.

Results: We present a case of a young, previously healthy Mongolian adolescent girl. Her medical history is notable for a left pre-tragial cyst operated in Mongolia in 2006, with an unremarkable evolution. Seven years later, after moving to Switzerland, she reported a purulent discharge in front of her right ear and was treated with oral amoxicillin/clavulanic acid and topical fusidic acid for 15 days for an abscess. Initial microbiological samples were positive for methicillin sensitive *Staphylococcus aureus* and *Actinomyces* sp. Due to the poor outcome, a preauricular and tragus fistula was clinically suspected and later confirmed during the surgical management of July 2013. The intraoperative microbiological samples were negative. The patient was lost of follow-up for 2 years, when she reported again a preauricular discharge. After a new surgical excision of the abscess, antibiotic treatment was started (amoxicillin/clavulanic acid). The diagnosis of actinomycosis was confirmed with positive samples for *Actinomyces turensis*. Antibiotic treatment was narrowed to amoxicillin and was prescribed for long-term treatment but without significant improvement. Two new surgeries were performed with extensive debridement several months later followed by a total parotidectomy with radical resection of the skin and cartilage in April 2017. Bacterial sensitivity remained unchanged since the beginning of the treatment. Observance was reported as good until September 2017, i. e. 5 months after the last surgical treatment. However, treatment was stopped by the patient in September 2017 against medical advice, in a context of suicidal ideation. Since then, the patient is clinically stable and all (superficial) cultures are negative 5 months of discontinuation.

Conclusion: Despite the commensality of *Actinomyces* sp. in the ENT microbiota, any positive *Actinomyces* specimen taken from a site with a chronic wound should suggest actinomycosis. Usually, a combination of (very) long term antibiotics and surgery are necessary to cure ENT actinomycosis. However, the optimal duration of the antibiotics is unknown.

NaChwuchs Prize: I wish to apply

Plaschke Prize: I wish to apply

I have read and understand application rules: Yes

Disclosure of Interest: None declared

P065

DIAGNOSIS OF SHWACHMAN-DIAMOND-SYNDROME IN A 3-YEAR OLD GIRL PRESENTING WITH NECROTISING SOFT TISSUE INFECTIONH. Schmid^{1,*}, J. Bonhoeffer¹, A. Kämpfen¹, N. Ritz¹¹University of Basel Children's Hospital, Basel, Switzerland

Introduction: Repeated or severe infections in a patient should lead to assessment of possible immunodeficiency. One rare cause, especially in children, which we present in the following case report, is Shwachman-Diamond-Syndrome (SDS). This autosomal recessive multisystem disease with an estimated incidence of 1:80'000 is predominantly characterised by bone marrow dysfunction and exocrine pancreatic insufficiency.

Results: Case report: A 3-year old girl presented to the emergency department with a history of fever for 4 days and skin lesions of the nose and left mamilla, first showing as maculopapular lesions 3 days earlier. Both lesions had necrotising centres of 1 cm in diameter on the dorsum of the nose and 2 mm on the mamilla respectively and each an extensive surrounding erythema. There was no previous trauma. Complete blood count showed leucocytosis of 15.27 G/l (10.5% banded, 37% segmented neutrophils with absolute count 7.25 G/l, 29.5% lymphocytes, 23% monocytes) and C-reactive protein was 172 mg/l. Ultrasound and a subsequent computed tomography of the head confirmed an abscess of the left mamilla and extensive perinasal and periorbital cellulitis. Surgical removal of the nasal necrosis was performed by plastic surgeons. Empiric broad-spectrum antibiotic therapy with ceftazidim (50 mg/kg TID) and clindamycin (15 mg/kg TID) was initiated. *Pseudomonas aeruginosa* was identified from the necrotic material and antibiotic treatment changed to oral ciprofloxacin (15 mg/kg BID). Blood cultures remained sterile. The skin defect on the nose was closed by glabella flap 10 days after admission and the patient was discharged one day later. Total antibiotic treatment duration was 3 weeks. The patient had

a history of several and severe bacterial infections (necrotising pneumonia, cervical lymphadenitis, recurring otitis media) and known enzyme-substituted exocrine pancreatic insufficiency (confirmed anew by low fecal elastase and sonographic signs of lipomatosis) as well as a positive family history of the latter in the older brother. Further signs and symptoms including growth <3rd percentile and intermittent moderate neutropenia (minimal 0.73 G/l) prompted genetic confirmation of SDS.

Conclusion: Associated neutropenia in SDS may lead to severe and rapidly progressing infections. Early disease identification is required for optimal disease management and prevention and appropriate handling of high risk situations such as infections with *P. aeruginosa*.

Disclosure of Interest: None declared

P066

CERVICAL ADENITIS HIDING LIVER ABSCESS: THINK CHRONIC GRANULOMATOUS DISEASE A CASE REPORTA. Schneider^{1,*}, E. Ranza², K. Posfay-Barbe³, A. Diana¹, M. Laurent⁴, A.-L. Rougemont-Pidoux⁵, M. Hauri-Hohl⁶, J. Reichenbach⁶, G. Blanchard Rohner¹

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Introduction: Cervical adenitis in children has many possible etiologies ranging from benign systemic viral infections to severe immunological or oncological diseases. When confronted with persisting or complicated cervical adenitis, chronic granulomatous disease (CGD), a rare primary immunodeficiency, should be suspected. We present a case of CGD diagnosed in this context.

Methods: case report

Results: A 19-month-old boy presented with persisting fever, despite cervical adenitis treated with intravenous antibiotics and surgical drainage. Abdomen ultrasound was therefore performed in the course of infectiological workup and revealed multiple liver nodules. CGD was confirmed by absence of reactive oxygen species production by dihydrorhodamine test. Genetic analysis revealed a non-sense hemizygous pathogenic variant c.676C>T:p.(Arg226*) in the *CYBB* gene, encoding the gp91phox subunit of the phagocyte NADPH oxidase on the X chromosome. The boy was initially treated 4 weeks with intravenous Cefepime, Clarithromycin, Caspofungin and Teicoplanin, and then switched to oral treatment using Trimethoprim-Sulfamethoxazole and Voriconazole, followed by prophylactic doses of Itraconazole and Trimethoprim-Sulfamethoxazole. Liver biopsy showed a focal fibro-inflammatory process but no granulomas, suggestive of an infectious process. Intravenous methylprednisolone (1 mg/kg/d) for 2 weeks was followed by oral prednisone with a tapering schedule, allowing complete resolution after 5 months. 10 months after initial diagnosis the patient remains in stable condition and is scheduled for allogeneic hematopoietic stem cell transplantation.

Conclusion: In conclusion, we present a case of early diagnosis of CGD clinically manifesting with refractory cervical adenitis, while liver abscess was still clinically silent.

Disclosure of Interest: None declared

P067

A CASE OF VARICELLA ZOSTER VIRUS MENINGITIS IN A IMMUNOCOMPETENT CHILDC. Sonnessa^{1,*}, P. Luggen¹, P. Zimmermann¹, S. Fluri¹

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Introduction: Acute infection with varicella zoster virus (VZV) is typically a self-limiting childhood illness with few complications. However, following an acute infection, VZV becomes latent in cranial nerves and dorsal root ganglia and may reactivate years later, potentially causing neurological complications.

Results: (Case presentation): A 13-year-old boy presented to our hospital because of severe headache. A few days prior to this, the mother had noticed a maculopapular rash on the boy's abdomen corresponding to two dermatomes, which was diagnosed as Herpes Zoster and treated with oral valacyclovir, at a lower than usually recommended dose of 2 × 100 mg per day. During the following days, the headache increased and became unresponsive to analgesic

therapy. A cerebrospinal fluid (CSF) examination showed a pleocytosis with a white blood cell count of $188 \times 10^6/L$ (94.5% lymphocytes and 2% atypical cells), a red blood cell count of $172 \times 10^6/L$, an increased protein concentration of 720 mg/L, and a glucose level of 2.8 mmol/L (blood glucose level 5.2 mmol/l). VZV DNA was detected in CSF by means of polymerase chain reaction (PCR), whereas PCR analyses for herpes simplex virus 1 and 2, enterovirus, tick-borne encephalitis virus and the bacterial culture remained negative. Serologies for *Borrelia burgdorferi* and *Treponema pallidum* in blood were unremarkable. An immunological screening, including testing for HIV, was normal. The valacyclovir dose was increased and treatment was continued for a total of 14 days. On follow-up at the end of the examination, the boy was asymptomatic and had a normal neurologic examination. Neuropsychological testing 6 months later showed mild deficits in memory functions.

Conclusion: Neurological complications due to VZV reactivation can develop in immunocompetent children and adolescents. Rare complications of lumbosacral herpes zoster include neurogenic bladder dysfunction or ileus with intestinal obstruction or transverse myelitis with possible persistent paralysis or sensory deficits. Hence, prompt diagnosis and initiation of therapy are crucial.

Disclosure of Interest: None declared

P068

TWO CASES OF ACUTE CARDIORESPIRATORY ARREST FOLLOWING HUMAN BOCAVIRUS INFECTION

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Introduction: Human bocavirus (HBoV) is a Parvoviridae first isolated in 2005 and found since then in about 10–25% of respiratory illnesses in young children 6m–3y of age. Few cases of severe invasive disease have been reported sporadically. No correlation between viral load and severity of infection could be established.

Results: We present two cases of acute cardiorespiratory arrest following human bocavirus infection. N° 1: A 30 mo girl born at 30 weeks of gestation without other comorbidities. She initially presented with 3 days of fever and a diagnostic of otitis and obstructive bronchitis, treated with co-amoxicillin and salbutamol inhalations. On day 3 she presented a temporary improvement except one episode of vomiting. Later that day she was unwell and presented a sudden loss of consciousness. Health care services present 15 minutes later found her in cardiorespiratory arrest that did not respond to CPR performed for one hour. HBoV was the only respiratory virus that could be detected by PCR with a high viral DNA load (Ct 15). It was also found postmortem in all pulmonary lobes as well as blood, liver, kidney and CSF swabs. N° 2: A 27 mo girl without medical priors apart from an episode of obstructive bronchitis 2 weeks earlier with 48h of hospitalization for oxygen therapy. She again presented typical signs of obstructive bronchitis for two days with respiratory distress for which she received salbutamol inhalations. At the arrival to the emergency department she had acute respiratory acidosis with a normal chest x-ray. She received a treatment of IV magnesium, continuous salbutamol inhalation and 2 mg/kg prednisone with subsequent improvement of clinical signs. About 4 hours later, while in the ICU, she presented an episode of vomiting triggering a laryngospasm followed by hypoxemia and acute cardiorespiratory arrest. She survived thanks to immediate CPR but had an absence of pulse for approximately 5–10 minutes. A rapid antigen test in the ER was positive for influenza B. A PCR panel for respiratory viruses performed 6 days later was negative for influenza but positive for HBoV with a high viral DNA load (Ct 23.7) HBoV DNA could also be detected in a blood sample on day 7. **Conclusion:** Future investigations are needed to clarify the role of HBoV in such severe respiratory infections. Are there susceptible patients for invasive HBoV infection? Are there any particularly aggressive strains? Should HBoV be screened in severe respiratory illnesses? Further studies are warranted.

Disclosure of Interest: None declared

P069

AN UNUSUAL ABDOMINAL TUMOR IN A YOUNG MALE ADOLESCENT

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Introduction: A child with an abdominal mass represents a frequent challenge in terms of differential diagnosis in pediatrics / pediatric oncology. Here, we report on a rare etiology of an adolescent with this clinical finding.

Methods: We present the case of a 15 year-old male patient, who presented to our pediatric emergency department with a history of abdominal discomfort over a couple of months, worsening over the last two weeks. The main clinical findings were constipation, loss of appetite, weight loss and nocturnal sweating. He had no other particular past medical history. At the clinical exam, he had high arterial pressure, the abdominal exam was overall painful and an abdominal mass was easily palpable in the peri-umbilical region.

Results: The radiological imaging (PET-CT) confirmed the presence of a voluminous intra-abdominal mass of $11 \times 10 \times 08$ cm with multiple infra- and supra-diaphragmatic adenopathies. An important compression of the left ureter was seen, causing a left renal impairment. Transcutaneous imaging-guided biopsy led to the diagnostic of a localized desmoplastic small round cell tumor (DSRCT), further confirmed by molecular pathology techniques. This aggressive sarcoma has a male predominance, and is often seen in adolescents or young adults. A pelvic or abdominal primary localization is typical. The management consensus of the DSRCT includes a multimodal treatment with neo-adjuvant chemotherapy, surgery, hyperthermic intraperitoneal chemotherapy (HIPEC), and radiotherapy, but the outcome remains poor. In this case, surgery took place 8 months after initial diagnosis, after 7 cycles of Ewing-like chemotherapy. Over 160 tumors (diameter of 2 mm- >5 cm) were found during the laparotomy. At the end of surgery, HIPEC (cisplatin) was performed. HIPEC is usually used for peritoneal cancerous diseases and is a promising treatment in disseminated diseases with multiple diffuse tumors like DSRCT. As soon as his clinical condition allowed it, he underwent 20 days of abdominopelvic radiotherapy for a total dose of 30 Gy. To complete the treatment, three additional cycles of chemotherapy are planned.

Conclusion: DSRCT is a rare tumor, affecting predominantly adolescents and young adults. In this challenging disease, multi-disciplinary approaches are needed including aggressive cytoreductive surgery. New data regarding genetic aspects for targeted therapies are currently being investigated also in our center.

NaCHwuchs Prize: I wish to apply

Plaschke Prize: I wish to apply

I have read and understand application rules: Yes

Disclosure of Interest: None declared

P070

CASE REPORT: HAPPY BLUE BABY

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Introduction: Methemoglobinemia, is a disorder characterized by the presence of high methemoglobin levels in the blood. Methemoglobin is an oxidized form of hemoglobin, which has an increased affinity for the oxygen and reduced ability to release oxygen to tissues.

Methods: We report a case of a 6 months-old baby who arrived at the pediatrics emergency department presenting bluish discoloration of the skin around the mouth, hands and foot, that began few hours earlier with no other symptoms.

Results: Female patient 6 months old. Born at full term vaginal delivery of healthy well controlled pregnancy. Aequat growth and development since birth. Breastfed until 2 months old and then bottle feeding with infant formula. At age of 5 months old, solids were introduced (fruits and vegetables) with good compliance. Food was made 24 hours before, and refrigerated until consumption. The day of the symptoms appeared, she ate a bowl of carrots mixed with potatoes and a smashed peach. At the clinical examination, she was in good condition with no respiratory distress, vital signs were normal (Heart rate 156/min; Respiratory rate 28/min. Oxygen saturation 95% in room air). When performing the clinical status, we observed the bluish discoloration around the mouth, face and hands. The episode was really quick and the baby was even smiling. No heart murmur or respiratory abnormal sounds were found. A Blood gas test was demanded with specification of methemoglobin concentration, resulting in 15.5% (reference values <1.2%). Blood gas controls were performed 3 and 12 hours after, showing a reduction of methemoglobin concentration; 7.3% and 1.5% respectively. An advice of the toxicology reference center (TOX ZENTRUM) was demanded. Values of methemoglobin below 30%, does not required treatment (methylene blue). The infant was admitted for further controls and laboratory testing until regularization of methemoglobin values. She was

discharged after normalization of methemoglobin levels. We explained the need to use water with low nitrate concentrations (<10 mg/L) and be careful with the preparation and management of vegetables (carrots, spinach and other long leaf green vegetables).

Conclusion: Methemoglobinemia is an acquired disorder that can be easily misdiagnosed. The most frequent cause in children is upon ingestion of food contaminated with water or drinking water with high nitrates levels, also dehydration, sepsis, topical anesthetics, and incorrect storage of homemade foods, as supposed in our case.

Disclosure of Interest: None declared

P071

A HEMIPARESIS: COULD IT HIDE A SEVERE VITAMIN B12 DEFICIENCY?

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Introduction: Vitamin B12 deficiency in children is rare. It accounts for only 1.4% of all adult and children anemias. Diagnosis is difficult, as symptoms are unspecific and depend of the severity of the case. Vitamin B12 has a role in DNA synthesis and neuronal myelination. Deficiency generally causes isolated anemia or other cytopenias as well as variable neurologic abnormalities.

Methods: A previously healthy 11 year-old girl, was admitted for an episode of blurry vision, diplopia and temporary loss of consciousness. For the past month she has been feeling tired, nauseous and presenting episodic headaches and paleness. When admitted, she was pale, conscious and had tachycardia. She had right facial paralysis and left hemiparesis. Laboratory results showed pancytopenia, with a non regenerative anemia (Hb: 33 g/l). Iron level was normal. A cerebral MRI showed asymmetrical cerebral vascularization and absence of thrombosis. Neurologic symptoms disappeared during the observation, we could then reasonably determine she was experiencing a hemiplegic migraine. She received blood transfusions. Differential diagnoses for the pancytopenia like infectious (EBV, CMV, Parvovirus B19, HSV, viral hepatitis, sepsis), oncologic (leukemia) and hemolytic (thalassemia) were ruled out. Further exams revealed a severe deficiency in vitamin B12 at undetectable levels with normal folates level. The bone marrow aspiration histology was compatible with megaloblastic anemia. The gastroscopy showed an atrophic chronic gastritis and micro-nodular hyperplasia, the colonoscopy was normal. We determined the diagnosis to be autoimmune atrophic gastritis, despite negative auto-antibodies (anti-intrinsic factor and anti-parietal cells). She received vitamin B12 IM injections and her blood parameters normalized. She never displayed any other neurological symptoms.

Results: Clinical presentation of our patient with the vitamin B12 deficiency was atypical. We had two problematics at the same time: symptoms which suggested a stroke or a vascular thrombosis that we could rule out with a normal cerebral MRI and spontaneous symptom resolution. The second emergency was the severe anemia with pancytopenia, for which she received blood transfusion and vitamin B12.

Conclusion: In this case, we were faced with concurrent emergencies: a hemisindrome which could be of ischemic origin and a profound anemia which finally turned out to be associated with a migraine and a vitamin B12 deficiency.

Disclosure of Interest: None declared

P072

RECURRENT POST-TONSILLECTOMY HEMORRHAGE: UNUSUAL HEMATOLOGICAL DISORDER

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Introduction: Tonsillectomy is a common pediatric procedure. Prevalence of postoperative hemorrhage is variable: 2% up to 15% depending on series. Coagulopathies are reported in up to 4.5% cases, with von Willebrand (vW) disease as the most common.

Case report: A 10 y.o. boy, previously healthy, was hospitalized for 5 days febrile left cervical lymphadenopathy treated by paracetamol and NSAIDs. CT-scan showed left cervical abscess with adjacent cellulitis and massive tonsillar hypertrophy. He underwent abscess drainage and bilateral tonsillectomy. At H36 he presented with tonsillar hemorrhage requiring surgical revision. Hemorrhage relapsed 2 days

later, with a total of 7 bouts, 5 of which required surgical revision. He received platelets, fresh frozen plasma (FFP) and packed RBC transfusions as well as tranexamic acid and DDAVP. Laboratory investigations, before any factor substitution, revealed normal platelets count, PT, aPTT and fibrinogen, as well as vW factor. Markedly low factor XIII (FXIII) activity at 7% was found. After administration of a single dose of 40 IU/kg plasma-derived (pd) FXIII (Fibrogammin®) i.v., bleeding stopped without recurrence. FXIII activity gradually normalized (110%) at 6 weeks, confirming transient character of factor XIII deficiency. Furthermore, the presence of a FXIII inhibitor was excluded by a normal mixing study. Antiphospholipid antibodies were negative.

Discussion: FXIII is essential to stabilize fibrin-clot by covalent linkage of fibrin. If insufficient or absent, bleeding may occur due to instable clot formation. FXIII deficiency (FXIIID) should be suspected in infants with intracranial bleeding, prolonged bleeding and/or poor healing of the umbilical stump and/or delayed bleeding after surgery or trauma. Severe congenital FXIIID (<1%) is very rare (1:2'000'000 births), whereas partial congenital deficiency and/or acquired deficiency may be more frequent but likely underreported. Acquired FXIIID may result from impaired synthesis (liver failure) or increased consumption (surgery, sepsis, leukemia, Henoch-Schönlein, inflammatory bowel disease, stroke, disseminated intravascular coagulation). FXIII replacement in form of FFP or pdFXIII may be necessary in the presence of bleeding and/or poor healing.

Conclusion: Although rare, FXIIID (congenital or acquired) should be considered in the setting of recurrent postoperative hemorrhage, even in the absence of an evocative history and/or normal coagulation tests.

Disclosure of Interest: None declared

P073

BAHOP: FROM LOCAL TO NATIONAL – THE NEW GERMLINE DNA BIOBANK FOR CHILDREN WITH BLOOD DISORDERS AND CANCER IN SWITZERLAND

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Introduction: Childhood cancers are rare diseases: approximately 300 children and adolescents are diagnosed with cancer every year in Switzerland. Genetic variations and specific germline mutations are associated with cancer development, individual response to treatment, and long-term complications. Establishing a biorepository dedicated to the collection of biological materials, related clinical and genetic data from childhood cancer patients, is essential for validation of treatment strategies and identification of biomarkers. Advances in the knowledge of these inherited genetic factors can help modifying therapies in order to decrease short and long-term side effects as well as increase survival.

Methods: The Pediatric Oncology and Hematology Unit of the University Hospital of Geneva established a biorepository in 2016: the BaHOP (Biobank in Pediatric Hematology and Oncology). BaHOP collects clinical data and germline DNA of all patients treated in the Unit. For patients undergoing stem cell transplant, also RNA, urine, stem cells, plasma and serum are stored. BaHOP governance is structured as follow: Steering Committee, Scientific Committee and Advisory Board for the overall management; operational management shared between laboratory, data and sample access and quality management for operational processes.

Results: Within BaHOP we have already collected over 5'250 single aliquots distributed as follow: 3'500 aliquots of plasma and serum, 1500 aliquots of DNA, 250 of RNA. Clinical data is centrally stored in our Laboratory Information Management System. In 2017, we extended our biorepository to all Swiss childhood cancer survivors with BISKIDS (Biobank In Switzerland for germline DNA collection in KIDS with blood disorders and cancer) and linked it to the Swiss Childhood Cancer Registry in order to correlate genetic inheritance to clinical phenotype. A pilot project (the GECCOS study) will analyze in 1000 children the genotype-phenotype association in long-term pulmonary toxicity.

Conclusion: Conducting high throughput genetic and genomic research requires high quality biospecimens to unravel causal factors of childhood cancers. BaHOP will assist studies on cancer predispositions, targeted therapies, and treatment-related toxicity in keeping precious biological materials and clinical data available for later use. This makes the biobank essential for supporting projects exploring evaluation and validation of treatment strategies and biomarkers.

Disclosure of Interest: None declared

P074

CARBONIC ANHYDRASE II DEFICIENCY: A RARE AUTOSOMAL RECESSIVE DISORDER OF OSTEOPETROSIS

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Introduction: Osteopetrosis is a rare disease. About 50 cases have been described related to carbonic anhydrase type II (CAII) deficiency. It is a congenital metabolic disease that combines, in addition to the classic signs of osteopetrosis, renal tubular acidosis, brain calcifications, variable mental retardation, short stature, dental occlusion disorders and hearing impairment. It is due to mutations on the CA2 gene (8q21.2) which encodes CA II.

Methods: A 3-year-old male child from a consanguineous marriage. The diagnosis is revealed by a weight-loss delay with facial dysmorphism associated with signs of bone marrow failure.

Results: The osteomedullary biopsy revealed a hypoplastic marrow with rare irregular calcified thin bone lamellae delimiting fibrous medullary spaces. The skeleton showed diffuse hyperdensity characteristic of the skull and limbs. The cerebral CT scan showed intracerebral calcification frontal and parietal. Renal ultrasound is suggestive of bilateral medullary nephrocalcinosis whose biochemical exploration has revealed distal tubular acidosis.

Conclusion: The diagnosis of autosomal recessive osteopetrosis due to carbonic anhydrase II deficiency is based on radiography of the skeleton with the presence of tubular acidosis and intracranial calcifications. It is confirmed by the molecular genetic test. Prenatal diagnosis is feasible. The treatment is symptomatic.

Disclosure of Interest: None declared

P075

CASE REPORT: SUBCLINICAL FIRST PRESENTATION OF ACUTE LYMPHOBLASTIC LEUKEMIA COINCIDING WITH AUTOIMMUNE NEUTROPENIA

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Introduction: Clinical presentation of acute lymphoblastic leukemia (ALL) can in rare cases be preceded by aplastic anemia. We report the case of a 1.5 year old boy, in whom diagnosis of ALL coincided with autoimmune neutropenia (AIN).

Methods: Case Report

Results: Clinical Case: The boy presented with acute onset of *E. coli* septicemia, agranulocytosis and anemia. Rapid clinical and erythropoietic recovery was observed upon initiation of antibiotic therapy but neutropenia persisted. A broad diagnostic workup for neutropenia was inconclusive except for anti-granulocytic antibodies (AGA). An initial bone marrow (BM) examination showed arrested myelopoiesis, and absent blasts. However, <5% of immature lymphoblast-like cells were detected by flow cytometry (FCM) and trephine BM-biopsy. However, the etiology of the alterations could not be clearly attributed and prompted subsequent BM-examinations. A second and third BM-analysis showed neutrophil recovery, but also first appearance of blasts on the BM-smear and a progressive increase of the lymphoblast-like population in FCM (10% → 20%). Cytogenetic analysis revealed high-hyperdiploidy and clonal IgH-rearrangement, confirming diagnosis of common ALL. The boy responded well to chemotherapy according to AIEOP-ALL 2009, was treated within the medium risk therapy group and is currently receiving maintenance therapy. In concordance with the AIN diagnosis, high absolute neutrophil counts (ANC) were found after initiation of immunosuppressive induction chemotherapy. After treatment blocks, neutropenia persisted despite rapid recovery of remaining lineages. Administration of a single dose of granulocyte stimulating factor resulted in a prompt increase of ANC.

Conclusion: In this case diagnosis of ALL was difficult to establish, due to the initial scarcity and non-specific alterations of the malignant clone. The specific detection of this evolving leukemic cell population was only possible through modern diagnostic tools, i.e. FCM and cytogenetic analysis. The finding of AIN posed an additional pitfall potentially leading to misdiagnosis. A small fraction of childhood ALL manifests as transient aplastic anemia prior to overt leukemia while preleukemic clones are already present. It has been suggested that alteration of immune surveillance plays a role in this leukemic transformation. It can be speculated that deficient immune surveillance likewise prompted emergence of AGA secreting B-cells in this patient.

Disclosure of Interest: None declared

P076

PERSISTENT HEMATOMA OF THE SKULL IN A 2-YEAR OLD: WHY PERFORM AN X-RAY

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Case Report: A 2-year-old girl presents at the pediatric emergency room. Her parents noticed a bump on her head which developed over the past 7 days. The bump is soft and painless. The girl is in good health without fever. Sonography of the skull shows a parietal defect with subgaleal fluid collection. Osteolysis is confirmed by X-ray. MRI shows parietal osteolysis and a non-infiltrative mass, partially hemorrhagic, with impression of the brain. Langerhans cell histiocytosis (LCH) is suspected and histopathologically confirmed by resection and evacuation of the hematoma. Clinically no further lesions are detected, therefore unifocal single system LCH is assumed. A watch & wait strategy is followed. Two months later the disease shows progression with a growing primary lesion and occurrence of three new skull lesions.

Background: LCH is a rare histiocytic disorder with inflammatory aspects and partly malign behavior. Aetiology remains unclear. LCH can affect all ages, but mostly occurs in infants. There is a broad disease spectrum. In 55% of the LCH patients one organ system is involved, 45% have multisystem disease. The majority shows bone involvement, most frequently the skull. Lesions can be asymptomatic or painful and can lead to fractures. Single bone lesions of the skull, formerly called eosinophilic granuloma, show a typically "punched-out" appearance in X-ray. Depending on the localization, there is a risk of CNS involvement. In this case or in multifocal bone or multisystem disease, systemic therapy is indicated with anti-inflammatory and chemotherapeutic drugs. "Risk organs" associated with worse prognosis are the lympho-hematopoietic system, liver and lung. Unifocal bone disease has to be confirmed by biopsy. Therapy is curettage, with or without injection of steroids followed by a watch & wait strategy. MRI or bone X-rays to exclude further lesions has to be discussed. Minimal diagnostic workup is thorough clinical examination including otoscopy, search for skin lesions, lymphadenopathy and organ involvement and basic laboratory investigations.

Conclusion: – In children presenting with soft tissue swelling of the skull, one should think of LCH, as this is a very common location. – The radiological finding is a typical "punched-out" lesion with a soft tissue mass. – Diagnosis has to be confirmed by biopsy. Prognosis is excellent with minimal surgery. – Follow up is mandatory, as in few cases unifocal bone LCH progresses or recurs and systemic therapy is needed.

Disclosure of Interest: None declared

P077

DISSEMINATED ISOLATED INTRACRANIAL JUVENILE XANTHOGRANULOMA

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Introduction: Juvenile Xanthogranuloma (JXG) is typically a benign, self-limiting histiocytic disorder of early childhood, predominantly manifesting as cutaneous lesions. Only about 4% develop systemic JXG. The commonest extracutaneous sites involve a solitary mass in the subcutis or deeper soft tissues, followed by liver, spleen, CNS and lungs. We present an atypical case with an isolated presentation of a disseminated, aggressive phenotype of an intracranial JXG. There are only 4 previous case reports about disseminated intracranial JXG without systemic disease; all of them show a relatively aggressive clinical picture.

Methods: Case Report: The 11 months old male infant was referred to us with a prolonged history of vomiting, severe hypotonia and developmental delay. An MRI demonstrated a large posterior fossa tumour with disseminated disease. He underwent partial resection. The lesion was morphologically and immunophenotypically consistent with JXG, BRAF negative. There was absence of systemic disease. He was treated in the high risk/ refractory LCH stratum according to LCH IV. He received Cladribine and Cytarabine concurrent with triple intrathecal chemotherapy. Subsequent MRI showed reduction in disease burden. Due to treatment related seizures, he was switched to a second induction therapy as per LCH high risk, group III with Vinblastine and Prednisolone, followed by maintenance chemotherapy. Treatment was stopped on the basis of toxicity and failure of further response.

Conclusion: There are several case descriptions of aggressive systemic JXG with activating MAPK pathway mutations, in keeping with the revised classification of histiocytoses (2016), where extracutaneous, disseminated JXG with MAPK mutations or ALK translocations have been included within Langerhans "L"-group together with Langerhans cell histiocytosis (LCH) and Erdheim Chester disease (ECD). Both, LCH and ECD have clonal mutations involving genes of the MAPK pathway in >80% of cases. This highlights the possibility that structural alterations involving kinases known to activate the MAPK pathway (not only BRAF) may be recurrent in L-type histiocytic neoplasms. The possibility of therapy with BRAF-, MEK- or even ERK inhibitors highlights the importance of identifying specific MAPK pathway alterations as part of diagnostic workup for patients with disseminated, especially intracranial JXG to consider aggressive and potentially targeted therapies with suitable inhibitors.

Disclosure of Interest: None declared

P078

ATAXIA AND SPEECH LOSS: A COMPLICATED COURSE OF "ATYPICAL" ROLANDIC EPILEPSY

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Introduction: Children with childhood epilepsy with centro-temporal spikes – shorter said rolandic epilepsy (RE) – can have a complicated course with motor, language or global regression associated with intense activation of epileptiform discharges during sleep, sometimes becoming continuous (continuous spike-waves during sleep (CSWS)). Focal rolandic seizures are not always in the forefront.

Results: Case report: 5.5 year-old girl, presented at age 28 months with subacute ataxia. Cerebral MRI and lumbar puncture were normal. She improved spontaneously but a few months later myoclonic jerks (limbs, perioral) were reported. An EEG revealed sleep – activated multifocal biphasic spikes, compatible with a an "atypical" RE. She improved with clobazam but at age 3.5 years, she had recurrent episodes of motor difficulties, speech and activity arrests. Neuropsychological evaluation revealed short attention span, oromotor and speech impairment. Comprehension and non-verbal skills were preserved. At age 4.5 years, she started to drool, refused eating solid food and stopped speaking. She also had frequent probably epileptic falls. The EEG showed multifocal activity and CSWS. I-v methylprednisolone-pulse-therapy was started (500 mg/d for 3 days, 1x/month) that had to be interrupted after 3 months, without significant effect. She improved clinically on high dose levetiracetam, but CSWS are still present. A *de novo* pathogenic variant in *GRIN2A* was found.

Conclusion: This child had an epileptic regression that mainly affected motor, speech and oromotor skills, related to CSWS. At presentation cerebellitis or a posterior fossa tumor was suspected, but epileptic "pseudo-ataxia" due to frequent myoclonic jerks was finally diagnosed. Oromotor and speech regression in the context of RE is known as "acquired epileptic opercular syndrome" and pathogenic variants in *GRIN2A* have been reported in families with an autosomal dominant pattern and sporadic cases.

Disclosure of Interest: None declared

P079

MULTIPLE CAFÉ AU LAIT SPOTS IN A CHILD: WORRYING DIAGNOSIS OF NEUROFIBROMATOSIS TYPE 1 OR THE LESS WORRYING LEGIUS SYNDROME?

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Introduction: Referral of children with multiple café au lait macules (CALMs) is a common scenario in the neuropediatric outpatient department. The reason is to evaluate a suspected diagnosis of neurofibromatosis type 1. When confirmed, these children have to be regularly followed up by their neuropediatricians, especially due to the

high risk of developing tumors in the central nervous system. This causes a lot of concern to the parents. A few of these patients however have the less common Legius syndrome (LS) (OMIM 611431). It is important to distinguish between these two syndromes which present very similarly but have extremely different prognoses. We present the case of such a child in order to increase the awareness of LS in children with CALMs.

Methods: Case report: This girl was referred by her general pediatrician due to CALMs present since birth. At the time of presentation the child was 12 months old, had 13 spots that were larger than 0.5 cm. The ophthalmological examination was normal. The clinical criteria for NF1 were not fulfilled. The mother was extremely worried and decided to ask for a genetic testing.

Results: Genetic analysis showed a pathogenic heterozygous mutation in the SPRED1 gene which is associated with LS. It is a dominantly (familial or *de novo*) inherited disease characterized by a mild neurofibromatosis-like phenotype; other manifestations of NF1 are absent, in particular no cutaneous neurofibromas, no optic pathway gliomas and no iris hamartomas. More than 200 cases have been reported.

Conclusion: Pediatricians should be aware that in children with CALMs the diagnosis of LS is a possibility. Identifying this condition is important because, unlike NF1, it is not associated with the occurrence of CNS tumors.

Literature: Stevenson D., Viskochil D., Mao R: Legius Syndrome: Gene Reviews-NCBI Bookshelf; January 15, 2015. Bernier A., Larbrisseau A., Perreault S.: CALMS and NF1: A review of the literature: *Pediatric Neurol* 2016 Jul;60:24–29.

Disclosure of Interest: None declared

P080

TRANSITORY EPISODES OF HEADACHES, CONFUSIONAL STATE AND LOW COGNITIVE EFFICIENCY: A CASE REPORT OF LATE-DIAGNOSED ATYPICAL HYPERPHENYLALANINEMIA

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Introduction: The authors present a case of neurological impairment in mild hyperphenylalaninemia (HPA) responsive to tetrahydrobiopterin (BH4), not detected in newborn screening and usually considered as benign.

Methods: Followed for mild intellectual impairment and severe developmental language deficit, a 13-year-old boy presented with recurrent daylong episodes of behavioral confusion, headache and anorexia. Non-convulsive status epilepticus was excluded by EEGs. A metabolic work-up showed a slightly elevated phenylalanine (Phe) plasma level (231 mmol/l, N 45–80).

Results: His Phe level was not elevated in newborn screening. Apart from an elevated Phe excretion in urine, metabolic investigations in blood and urine did not reveal any other abnormalities. We suspected a deficit in BH4 metabolism. DHPR activity and urinary pterins were normal. CSF analyses showed significantly elevated Phe (72 mmol/l, N 5–15), slightly elevated Tyr (19 mmol/l, N 5–17) and slightly diminished 5HIAA (63 nmol/l, N 74–163). Genetic analysis of the *PAH* gene revealed compound heterozygosity for two known missense mutations (Arg176Leu and Val388Met). A treatment with KUVAN[®] 800 mg/d (= 10 mg/kg/d) was started. A CSF control after two months of treatment showed less elevated Phe (49 mmol/l, N 5–15), normalized Tyr and neurotransmitters. The clinical situation of the patient improved slowly. After one year of treatment his non-verbal IQ, that had been stable over several years, improved from 71 to 81 (Wechsler Intelligence Scale) and emotional regulation and monitoring skills normalized (BRIEF questionnaire). Confusional states never reappeared during the 18 months follow-up.

Conclusion: In children with episodic confusional states, metabolic screening should be part of the clinical work-up. Mild hyperphenylalaninemia usually does not need to be treated and does not cause neurodevelopment impairment. However, there are other case reports with high Phe concentrations in the CSF pointing to rare subgroup of patients who might benefit from treatment.

Disclosure of Interest: None declared

P081

WHAT'S BEHIND THIS HAIRY TAIL

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Introduction: Diastematomyelia is a form of split cord malformation, a rare congenital condition resulting from failure of spinal development. It represents 4% of the larger group of closed spinal dysraphism. It occurs in 0.5 to 8/1000 live births and affects mainly females. It consists of a separation of the spinal cord into two parts, by an osseous or cartilaginous spur in the spinal canal.

Methods: We reported a case of a full term female newborn, born by vaginal delivery, after normal prenatal follow-up, with no perinatal complications. A routine examination, showed a hypertrichosis along the thoracic lumbar spine, a bulge of skin on the higher part of the back with a normal neurological and orthopedic examination. The lumbar ultrasound suggested diastematomyelia. A diagnostic MRI was performed that revealed a type 1 diastematomyelia from T3 to T5, a fibrolipoma of terminal filum and the fusion of spinal processes from L5 to S2. An exhaustive investigation revealed no associated abnormalities. The familial history reported no other cases of neural tube defects.



Figure 1

Results: Our patient is a typical case of diastematomyelia with a neonatal diagnostic. Most patients are diagnosed in the neonatal period and more rarely in childhood or adulthood. Two types of diastematomyelia are described. In type 1, each hemicord is situated in a separate dural tube, whereas in type 2, both hemicords are situated in the same dural tube. Cutaneous lesions should raise suspicion for closed spinal dysraphisms, especially a patch of hypertrichosis, one of the most common cutaneous sign. Our patient had normal neurologic examination but symptoms can appear later in life. Neurological disorders like autonomic and sphincteric dysfunctions or sensorimotor deficits in the lower limbs are common and can be stable or progressing, mainly due to tethering and/or syrinx within the cord. Multidisciplinary follow-up is mandatory, including neurosurgeons, urologists and pediatricians. Limbs as well as spinal deformities may appear late and warrant an orthopedic follow-up as well. No risk factor were identified in this case. The etiology is not clearly established but seems to be multifactorial including maternal diabetes, nutritional factors such as acid folic deficiency, maternal exposure to drugs and genetic factors.

Conclusion: Patch of hypertrichosis or other cutaneous anomalies should raise suspicion for closed spinal dysraphism. Early diagnosis triggers appropriate diagnostic workup and follow-up.

Disclosure of Interest: None declared

P082

PINEAL CYSTS: IS FOLLOW UP NECESSARY? A CASE REPORT AND LITERATURE REVIEW

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Introduction: Pineal cysts (PCs) are a common incidental finding in children, with an increased prevalence in girls and in older patients. Overall, PC represent 2–4% of incidental MRI findings in children. PCs are usually asymptomatic, even though a broad range of query associated nonspecific symptoms such as headaches (common finding) have been reported.

Methods: We present a 12-year-old girl who presented with recurrent tension-type headaches since three years. Personal history was otherwise uneventful, neurological examination was normal. Family history was negative for migraines or other neurological disorders. Due to the prolonged history of headaches the pediatrician ordered a neuroimaging which showed a pineal cyst with septations, measuring 9 × 7 mm. There was no nodularity and normal contrast behavior. The girl was then referred with the questions whether the PC is clinically relevant and whether a follow-up MRI is necessary.

Results: Considering the normal neurological examination in combination with the small size and normal contrast behavior of the PC no further follow up imaging was recommended. However, we informed the girl regarding handling and therapy of tension headaches. The diagnosis of PCs is usually established by MRI with defined radiological criteria to distinguish benign PCs from tumors of this area. The diameters of the symptomatic cysts are usually more than 20 mm. PC above 10–12 mm in diameter may need follow-up imaging, as a cystic pineocytoma may appear similar. The contrast material may diffuse from the enhancing rim into the fluid center of the pineal cyst on delayed imaging (60–90 minutes after gadolinium injection), producing an increasingly homogeneous enhancement pattern, suggestive of solid neoplasm. Therefore, it is important to take into account the time elapsed after contrast administration in such unusual cases. MRI follow-up is necessary when the imaging appearance of PCs overlaps with the appearance of pineal neoplasms including nodular enhancement, hemorrhage, or large size compressing severely the tectum or aqueductus Sylvii.

Conclusion: PCs in children are mostly benign, incidental findings, for which follow-up is not required, in the absence of unusual radiological characteristics or related clinical symptoms.

Disclosure of Interest: None declared

P083

DOOSE SYNDROME IN A 2 Y.O. BOY WITH FAVORABLE CLINICAL COURSE

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Introduction: A 2 y.o. boy, with known febrile seizures a month ago, presented with afebrile paroxysmal episodes of atony with head nodding followed by generalized hypertonia with upward rolling of the eyes and tonic vibrating seizures of the limbs. A few hours later, he developed 7 myoclonic seizures of the upper limbs with the eyes fixed, lasting for 2 seconds. Doose syndrome (DS) was presumed. The electroencephalogram (EEG) was compatible with this diagnosis. Cerebral magnetic resonance imaging (MRI) showed a right millimetric parietal dysplastic lesion. Despite treatment with valproic acid, myoclonic seizures with absences in the early morning hours continued and diazepam was added. 15 days later, exacerbations of the convulsions led to the addition of levetiracetam. Clinical course was favorable without relapse. The development remained normal for his age. At 9 months, EEG was normal. Responding to parental demand, we started tapering the antiepileptic medications, with positive course.

Results: Discussion: Myoclonic-atonic epilepsy (MAE) or DS, constitutes 1–2% of childhood-onset epilepsies. It is regarded as having a genetic etiology, with a positive family history of seizures in first, second, or third-degree relatives in about one third of cases. Variable semiological seizures including atonic fits and myoclonia suggest MAE. The first seizure usually occurs between 7 months and 6 years, but principally between 2 and 4 years. Males are mostly concerned, except for the first year of life, when the incidence is equal. The EEG, that may be initially normal, demonstrates brief bursts of 2 to 5Hz spikes, waves, polyspikes and wave complexes, as well as parietal thetas. The MRI is usually normal. DS is remarkably difficult to treat and the medications mostly proposed are ethosuximide and valproic acid. Ketogenic diet is also reported beneficial for patients with failure of two or more medications and leads to a reduction in seizures in more than the 50%. Two third of patients achieve seizure remission and have normal cognition. The remaining third may have severe cognitive or intellectual disability. Despite the difficulty to treat, our patient responded well to treatment with an early tapering of the medications.

Conclusion: In a child with variable semiological seizures including atonic fits and myoclonia a clinician should think about MAE. Prognosis is variable and independent of the severity of the epilepsy. Close developmental follow-up is mandatory.

Disclosure of Interest: None declared

P084

HEADACHES AND BRIEF NEUROLOGICAL DEFECTS IN A 7 Y.O. BOY... WHAT WOULD YOU DO?

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Introduction: A 7 y.o boy suffered from abrupt frontal headaches for 3 days. He quickly felt his right leg weak, causing a fall. He complained of transitory blind gap in right visual field. He remained asymptomatic for 2 days. Because of recurrence of headaches, he visited his pediatrician. He was then asymptomatic. He had no infectious symptoms or head/neck trauma. Family history revealed a post-operative leg thrombosis. Neurological status was normal. His pediatrician programmed an ambulatory MRI. It revealed subacute infarction of the left temporal lobe and of the anterior portion of the left occipito-temporal gyrus, with wall hematoma (P2 segment) of the PCA. MR angiography of the precerebral arteries was negative for carotid or vertebral artery dissection. Angiography confirmed occlusion of the left PCA with an important collateral flow, but no sign of vasculitis. It makes the diagnosis of spontaneous dissection of P2 segment of the left PCA causing infarction of the temporal territory. Inflammatory and thrombophilia work up were normal. Ophthalmologic exam was unremarkable. He was treated with anti-platelet drug (acetylsalicylic acid 100 mg daily). Control MRI one month later showed recanalization of the P2 segment of the left PCA with persistent P3–P4 occlusion, disappearance of wall hematoma and the absence of new ischemic infarcts.

Results: Discussion: Annual incidence of cerebral ischemic stroke in children is of 3 to 8/100 000. It must be ruled out in case of acute neurological symptoms, even transient. Boys are more concerned than girls (60% vs 40%), mainly in pre-scholar children and adolescents. Most complains are headaches, focal neurological symptoms, visual field defect and vertigo. Extra-cranial dissection is responsible for 7–20% cerebral stroke. Risk factors (infection, vasculopathy, cardiac, hematologic or metabolic diseases, drugs, trauma) are present in 20–50% of patients. Gold standard investigation is cerebral MRI completed by cerebral/neck angio-MRI (“black blood” wall study) and angiography. In the present case, infarction was due to acute dissection of the left PCA, which is a rare entity. Anti-platelets is the treatment of choice in case of asymptomatic patient. Anticoagulants should be considered in case of recurrence or radiological extension.

Conclusion: Ischemic stroke must be ruled out in pediatric population, even in cases of transient neurological defects. Investigations and management require specialized and multidisciplinary approach.

Disclosure of Interest: None declared

P085

NEONATAL SUBPIAL HEMORRHAGE: A FORGOTTEN FORM OF PERINATAL INTRACRANIAL BLEEDING?

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Introduction: Subpial hemorrhage is a rare cause of cerebral bleeding in preterm and term newborns. Its prognosis is usually favorable.

Methods: Case report: A 36-weeks newborn boy was investigated during the second day of life, because of generalized hypotonia and cephalhematoma. He was born by vaginal delivery with vacuum, with a confirmed prenatal diagnosis of aortic coarctation. A Pierre-Robin sequence was found at delivery. A large hyperechogenic lesion in the right temporo-parietal region was found on ultrasound imaging. Brain MRI showed a typical subpial hematoma with an ischemo-hemorrhagic component of the adjacent cortex. Despite hematological work-up, a definitive cause of the bleeding could not be identified.

Results: Discussion: Data concerning subpial hemorrhage in newborns are sparse. Typical intracranial hemorrhage related to the birth process are most often subdural or subarachnoid. Subpial hemorrhage occurs in the virtual space between the superficial brain parenchyma and pia mater meninges, probably consecutive to stretching and secondary rupture of the perforating vessels. It occurs most frequently in the anterior-inferior temporal lobe, which is especially vulnerable due to this immature venous configuration and anatomical position at the sutural confluence. Risk factors of subpial hemorrhages in term neonates include asphyxia, blood dyscrasia

or congenital vascular malformations. Clinical presentation is non-specific. Neonates can manifest with hypotonia, apnea and seizures, within 36–48 h after birth. Extracranial soft-tissue swelling can be associated, consisted in edema in the subcutaneous fat or cephalohematoma.

Conclusion: Hypotonia, seizure and apnea in an otherwise healthy neonate need to be investigate by cerebral imaging (ultrasound and MRI). Subpial hemorrhage is uncommon and its etiology remains often unclear. Prognosis is usually favorable but long-term developmental follow-up is recommended.

Disclosure of Interest: None declared

P086

THE EVER CHANGING PICTURE OF INFLUENZA VIRUS TYPE B INFECTION – CASE REPORTS WITH MILD ENCEPHALOPATHY AND REVERSIBLE SPLENIAL LESION (MERS) AND UNUSUALLY MARKED WHITE MATTER LESIONS

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Introduction: Influenza virus infection in children can be associated with a variety of neurological problems, some related to the infection itself others of parainfectious aetiology. Most frequently described are encephalitis/meningitis but also parainfectious mild encephalopathy with reversible splenial lesions (MERS). Here we report two children with influenza virus type B infection accompanied by MERS and unusually marked white matter (WM) problems.

Results: Case 1: A previously healthy 11-year-old boy complained about a few day history of upper respiratory tract infection (URTI) and 2 episodes of fluctuant bilateral weakness with dysarthria/dysphasia. Clinically bilateral facial palsy, bilateral limb weakness and decreased tendon jerks were noted. Additionally bilateral cavus foot was apparent. A nasopharyngeal swab antigen test detected influenza B, cerebrospinal fluid (CSF) was normal incl. negative PCR of influenza B. Diffusion- and T2-weighted cerebral imaging revealed marked bilateral posterior WM and callosal abnormalities. Spinal MRI was normal. EEG showed bilateral posterior slowing, no epileptic discharges. Without specific treatment, he recovered within days, but presented persistent areflexia. On insistent family history Charcot-Marie-Tooth-Disease (CMTX1) was known in the mother. Subsequent nerve conduction studies in the boy proved decreased velocity motor and sensory nerves. **Case 2:** In a previously healthy 13-year-old boy, symptoms of speech disturbance and decreased level of consciousness within the context of URTI evolved over 2 days. At presentation Glasgow Coma Scale was 11. Dysphasia, right sided hemiparesis and bilateral increased tendon jerks were noted. Influenza B in nasopharyngeal swab was positive, CSF incl. PCR normal. Cerebral MRI revealed meningeal enhancement, marked bilateral fronto-parietally predominant WM and callosal abnormalities. EEG showed diffuse slowing, no epileptic discharges. He was not specifically treated and fully recovered within days.

Conclusion: Both boys presented with neurological symptoms and MERS coincident with influenza B infection. The additional WM abnormalities were more pronounced than usually seen. In case 1 they might be related to concurrent CMTX. However, case 2 suffered even more pronounced WM problems without CMTX. Follow-up will show whether WM problems in case 1 only had worsened due to CMTX and whether in case 2 they had been induced by influenza B. Both patients showed a benign clinical course.

Disclosure of Interest: None declared

P087

“MOM, I’VE GOT A CLOCK IN MY MOUTH”: AN UNUSUAL PRESENTATION OF PALATAL TREMOR

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Introduction: Case report: A healthy 5y old boy came in the emergency department presenting a short (<12h) history of “hearing a noise” in his mouth. This clicking sound, audible by other persons with opened mouth only, was associated with regular rhythmic palatal contractions at frequency of 70 cycles per minute. There was no other associated general or Ear/Nose/Throat symptom, especially no tinnitus. This noise could be stopped by voluntary (speech, voice sounds) or involuntary (nausea reflex) contraction of the tensor veli muscle. It also disappeared during sleep. His neurological and developmental history was unremarkable, as was the entire

neurological examination. Family history showed a ganglioglioma of the right cerebellar peduncle by his 2y old younger brother (by whom main complain leading to diagnosis were gaze abnormalities). Because of the familial context and the possibility of truncal anomalies, a cerebral MRI was performed. The imaging results fortunately showed to be normal.

Methods: Review of literature.

Results: Palatal tremors – a rare entity in children – are comprised of two subtypes: the more frequent *essential type* (primary) and the *symptomatic type* (secondary to underlying lesion) – extremely rare in the paediatric population. Clinical presentation of essential palatal tremor, which is a benign entity, is usually an objective tinnitus heard with a stethoscope over the patient's ear. The aetiology of palatal tremor is yet uncertain, but in some cases a genetic predisposition has been proposed and psychological/psychiatric conditions can explain a few cases. The diagnosis is clinical; therefore a thorough clinical and neurological examination is mandatory, especially in order to rule out other cranial nerve abnormalities or other general signs suggestive of an underlying disorder.

Conclusion: This case report describes an unusual presentation ("clicking mouth") of a rare condition, the essential palatal tremor. In spite of its benign character and in the absence of strong recommendation, cerebral imaging seems reasonable to rule out any underlying brain lesion.

Disclosure of Interest: None declared

P088

ACUTE ROTAVIRAL ENCEPHALITIS WITH CORPUS CALLOSUM SIGNAL ABNORMALITY ON MRI IN A PREVIOUSLY HEALTHY CHILD

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Introduction: Rotavirus is a common cause of gastroenteritis in childhood, associated with central nervous system involvement in 2–5% of patients. We present a case of rotavirus encephalitis with cerebellar signs and symptoms.

Methods: A 3-year-old caucasian girl, not immunized against rotavirus, was admitted for 4 episodes of generalized tonic-clonic convulsions after a 3-day history of fever, vomiting and diarrhea. Intranasal midazolam allowed resolution of seizures. On day 4, she presented a recurrence of seizures. Between the seizures, she was lethargic, without meningism nor focal neurological sign. The seizures stopped after a loading dose of phenytoin, followed by maintenance doses. Her consciousness level improved, but we noted the presence of mutism, muscular hypotonia and trunk ataxia. On day 11, she presented recurrence of seizures. The phenytoin level being subtherapeutic, she received a new loading dose. On day 13, she began to utter a few words and to walk with ataxia. Last seizure was on day 14. On day 28, she had normal motor and cognitive functions, though remaining dysmetric. Rotavirus was detected in stool sample. Brain CT and cerebrospinal fluid analysis were normal. On day 5, brain MRI revealed high T2 signal intensity in the splenium and genu of the corpus callosum. EEG on day 28 was normal, and phenytoin treatment was discontinued. This clinical presentation strongly suggests an acute encephalitis associated with rotavirus infection.

Results: Acute rotaviral encephalitis is described as following a self-limited course without sequelae with characteristic clinical features: seizures, disturbance of consciousness followed by mutism. Splenial lesion on MRI is typical in acute phase. However, our case appears to be the first with prolonged seizures. Cerebellar mutism with subsequent dysarthria following a disturbance of consciousness is the most interesting clinical finding. The pathophysiology of CNS involvement remains unclear. Either a direct mechanism or an indirect action through a rotavirus nonstructural protein 4 acting as neurotransmitter have been proposed. In our case, PCR or antigens of rotavirus in the CSF were not tested, but the absence of pleocytosis makes the mechanism of direct invasion less likely.

Conclusion: CNS involvement of rotavirus has mainly been reported in Asian children. However, the pathophysiology of neurological manifestations deserves further assessment and the impact of rotavirus vaccines on extra-intestinal manifestations is yet to be demonstrated.

Disclosure of Interest: None declared

AFEBRILE CONVULSION WITH MILD GASTROENTERITIS (CWG)

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Introduction: Convulsion in the sequence of mild gastroenteritis is an entity characterized by seizures which occur in otherwise healthy children, usually in the absence of fever and in the presence of Rotavirus confirmed acute gastroenteritis (AGE), a very common yet clinically significant disease in the paediatric population. In Asian infants, this condition is designated by the name of convulsions with mild gastroenteritis (CwG).

Methods: We report two cases of non febrile seizures in the context of proven Rotavirus AGE, which occurred in two 20-month old children – one male and the other female. The children had no particularly relevant medical history; they presented seizures in clusters after an episode of gastroenteritis, with favourable clinical evolution.

Results: In both cases, the lumbar punctures showed no abnormal findings for cell counts, biochemistry and bacterial cultures. For the length of the study there weren't any recurrent seizures and there were only provided supportive measures. The CwG syndrome is a distinct entity characterized by the following criteria: (1) it occurs in previously healthy 6-month to 3-year-old children who present afebrile convulsions, (2) it is associated with mild gastroenteritis, (3) seizures tend to occur in a clustered manner, (4) interictal EEG shows no paroxysmal discharge, and (5) laboratory examinations are normal, including normal cerebrospinal fluid, serum electrolytes and glycemia. This syndrome is a self-limited disease with a short course and a good prognosis. In fact, electroencephalography, lumbar punctures, and radiological examinations are usually not useful because they tend to be normal in these patients. Prolonged seizures, on the other hand, which are usually not common, require antiepileptic treatments in the acute phase. As rotavirus is the most common causing agent of CwG among the diarrheal viruses, one could hypothesize that this virus may play a role in the pathogenesis of CwG.

Conclusion: Knowing the characteristics of CwG in children with Rotavirus gastroenteritis is essential once that the management of this condition can reasonably be conservative, avoiding useless hospitalisation, unnecessary long-term anticonvulsant therapy (as the drugs have potential side effects) and extensive neurological complementary exams, which are of low yield.

Disclosure of Interest: None declared

P090

COGNITIVE REGRESSION IN A SOMALIAN ADOLESCENT CAUSED BY SUBACUTE SCLEROSING PANENCEPHALITIS: THE TRAGIC BURDEN OF ONGOING MEASLES EPIDEMICS IN DEVELOPING COUNTRIES

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Introduction: A previously healthy 12 y-o Somalian boy presented with a 3 months history of subacute cognitive regression, repetitive falls (20/day), behavioral changes and inability to meet school requirements. An EEG and brain MRI performed soon after symptoms onset were normal. Birth and family history were non contributive; parents were non-related. The boy arrived at age 9 in Switzerland and received all recommended vaccinations (no prior measles immunization). On physical examination, he was disorientated with incoherent speech, poor attention span and short-term memory, global apraxia, stereotypical movements, motor hyperkinesia and bilateral palmar grasping. Overnight video-EEG revealed recurrent myoclonic seizures corresponding to periodic discharges with loss of physiological architecture. Cerebrospinal fluid examination revealed oligoclonal bands. Intrathecal synthesis of measles antibodies confirmed diagnosis of Subacute Sclerosing Panencephalitis (SSPE).

Results: Adolescent-onset cognitive regression deserves thorough evaluation. While neurometabolic disorders are leading causes of early onset neurodegenerative disorders, infectious etiologies such as SSPE must be considered, not only countries where measles remains endemic but also in Europe where occasional measles outbreaks are ongoing. Migrant children affected by measles before catch-up immunization are certainly at risk. SSPE is a chronic encephalitis secondary to measles infection causing diffuse progressive involvement of central nervous system appearing on average 6 years after measles' infection and with a fatal issue at 2 years in most cases. Evidence for treatment is low with at best stabilization or temporary improvement in less than half of patients. Therapeutic options

mainly studied are combinations of antivirals (ribavirin) and immunomodulators (interferon, inosiplex). Intraventricular route is associated with less adverse effects. Such treatment was considered not suitable due to parental beliefs that illness was due to sorcery, and advanced stage of disease at time of diagnosis. Parents declined treatment including antiepileptic drugs. Patient died 6 months later.

Conclusion: SSPE must be considered in the differential of childhood and adolescent cognitive decline, even with normal EEG and MRI at an early stage. Diagnosis is confirmed by the finding of intrathecal synthesis of measles antibodies. Measles immunization can prevent measles and its severe complications including SSPE, whose prognosis remains poor.

Disclosure of Interest: None declared

P091

PRES: ACUTE BLINDNESS SECONDARY TO TONSILLITIS – A CASE REPORT

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Introduction: Tonsillitis due to streptococcus pyogenes (GAS) infection is a common bacterial infection with quick recovery. Acute post-infectious glomerulonephritis (GN) is a complication of GAS-tonsillitis, often accompanied by arterial hypertension. Acute hypertension can lead to altered perfusion of the brain causing severe neurological symptoms.

Methods: Case report.

Results: A 12 year old boy presented with unilateral painful torticollis after mild trauma without signs of infection. He was discharged with analgesia. The next day he presented with painful swallowing, trismus and fever. Retropharyngeal abscess was suggested and confirmed with CT of the neck. After surgical incision and antibiotic treatment, the boy recovered quickly. Nine days later, he was referred with visual loss and flashing and lighting sensations, severe headache, swelling of the lower limbs and repeated focal and secondary generalized seizures. Pupillary reflex and bulbar motor function were normal, but visual fixation was not possible, suggesting cortical blindness. Blood pressure was highly elevated (164/145 mm Hg). Cranial MRI showed cortical and subcortical hyperintensity in parietal, parieto-occipital and frontal areas and smooth temporo-polar cortical swelling, compatible with PRES (posterior reversible encephalopathy syndrome) secondary to hypertensive crisis. The findings of elevated plasma creatinine (64 mmol/l), low serum C3, elevated ASLO-titer and proteinuria/haematuria were consistent with post-infectious acute GN. Blood pressure normalized with triple therapy (Sodiumnitroprusside, Furosemide and Amlodipine) and neurological symptoms resolved within the first day. Follow up after 3 months showed a boy in good clinical condition: Vision, neurological examination, blood pressure and repeated cranial MRI were normal. Pathogenesis of PRES is unclear. In cases with acute hypertension, disordered cerebral autoregulation and endothelial dysfunction lead to excessive cerebral blood flow, fluid extravasation and brain edema causing neurological symptoms, typically headache, visual disorders, seizures and decreased level of consciousness.

Conclusion: PRES is a rare complication of hypertensive crisis secondary to various reasons, e.g. acute GN or immunosuppressive medication. PRES is characterized by neuroimaging findings of reversible vasogenic subcortical edema without infarction. PRES requires timely neuroimaging by cranial MRI and early antihypertensive therapy. The course is often favorable with full recovery.

Disclosure of Interest: None declared

P092

LYME DISEASE-RELATED PSEUDOTUMOR CEREBRI IN CHILDREN – A SMALL CASE SERIES

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Introduction: Lyme disease is the most common tick-borne infection and endemic in most parts of the northern hemisphere including Switzerland. The main manifestations of neuroborreliosis in children are facial palsy and aseptic meningitis. An isolated intracranial hypertension caused by *Borrelia burgdorferi* is a very rare but serious manifestation. Its recognition is important since the treatment and outcome may differ from the standard care of patients affected by idiopathic intracranial hypertension (pseudotumor cerebri).

Methods: Case report

Results: Two girls at the age of 10 resp. 15 years presented within nine days on our emergency department with signs of intracranial hypertension. *Girl A* presented with chronic headache since five weeks, sober vomiting, wake-up at night and intermittent double vision without clinical signs of an obvious infection. A tick bite two months ago without erythema migrans was remembered. Neurological examination showed papilledema left more than right. Further clinical examination and the neuroimaging by cranial MRI (including venography) were without any pathological findings. Lumbar puncture revealed an elevated intracranial pressure of 43 cmH₂O, cell count was 135/ul and the serology was positive for *Borrelia burgdorferi*. Treatment with parenteral ceftriaxone and oral acetazolamide was started. After two weeks of therapy all symptoms had completely resolved and after two months ophthalmologic examination was unremarkable. *Girl B* presented with frontal headache since one week increasing during night, wake-up at night and recurrent sober vomiting. There were no fever, no signs of an infection or tick bite remembered. She was in reduced general condition and showed a unilateral peripheral facial palsy (House-Brackman Score 4). The neurological examination showed a marginal unilateral papilledema, cranial MRI (with gadolinium-contrast) was without any pathological finding. On lumbar puncture, the intracranial pressure was increased to 34 cmH₂O, cell count was 273/ul and the serology was positive for *Borrelia burgdorferi*. An intravenous therapy with ceftriaxone for two weeks was performed and acetazolamide was given additionally. After two months, only a minor asymmetry in cheek inflation was observed and ophthalmological follow-up was unremarkable.

Conclusion: Intracranial hypertension due to Lyme disease is a rare but possible entity and has to be considered in children presenting with signs of increased intracranial pressure.

Disclosure of Interest: None declared

P093

SWISS PEDIATRICIAN SURVEY ON COMPLEMENTARY MEDICINE

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Introduction: Complementary medicine (CM) summarizes all diagnostic, therapeutic and preventive methods that are used in conjunction with those of conventional medicine. The blending of complementary and conventional strategies to include all appropriate therapies in a patient-centered fashion and with a focus on interprofessional collaboration is described as integrative medicine. This integrative approach is in particular characteristic for the practice of CM by medical doctors in Switzerland. The present study was carried out to evaluate the attitude towards, the training in and the offer of CM among pediatricians in Switzerland.

Methods: National survey using a structured, self-reporting, anonymous online questionnaire. Its final version was provided in the three major national languages German, French and Italian and consisted of 19 items. The board of the Swiss Society of Paediatrics (SSP) approved the study and gave the opportunity to use their member list for the survey. Representativity of the sample of participating pediatricians was assessed using the statistics of the Swiss Medical Association FMH.

Results: 1890 members of the SSP were approached and 640 (34%) responded to the survey. Two thirds were female (65%), aged 35–55 years (65%), had a training as pediatric generalist (68%) and worked in a practice (63%). Among respondents 23% have attended trainings in CM, with phytotherapy and homeopathy being the most frequent. 16% offer CM to their patients and 8% have an official CM certificate, which is required for the reimbursement from the basic health insurance. 63% are interested in courses and trainings on CM issues. CM use was reported by more than half of the responding pediatricians for themselves (58%) or their immediate families (51%). 71% of respondents supported the establishment of an official Swiss working group for integrative pediatrics.

Conclusion: In a representative sample of pediatricians in Switzerland, the attitude towards CM is positive. With respect to the strong demand for CM, the offer of CM by pediatricians is rather low. Among the various reasons insufficient knowledge and institutional barriers deserve special attention. The great interest of pediatricians in CM training as well as their willingness to participate in CM research offer key elements for the future development of complementary and integrative medicine for children in Switzerland.

Disclosure of Interest: None declared

P094

NEONATAL BLISTERS

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Introduction: A term born male neonate presented at birth with blisters on the hands, lower back and palate. First infant; parents originated from the same village in Vietnam. Diagnosis of junctional epidermolysis bullosa (JEB) was suspected. Histopathological examination of skin biopsy confirmed sub-epidermal blistering. Epitope mapping with dedicated panel for congenital bullous diseases revealed complete absence of laminin V expression. Next generation sequencing of gene-panel revealed homozygous duplication c.76dup, p.(Cys26Leufs*28) in LAMB3, expected to result in premature stop-codon and protein lacking all its functional domains. This mutation has never been reported before. Evolution was marked by gradual appearance of new blisters, predominantly in areas of friction, trunk, mouth and nose. Management included topical antibiotics and non-adherent wet dressings. Pain relief was achieved by oral morphine. Main complications were respiratory distress and failure to thrive secondary to internal mucocutaneous blistering. He was discharged with regular follow-up by palliative care team and died 2 months later.

Results: Discussion: Epidermolysis bullosa is a genetically determined skin condition characterised by trauma-induced blister formation. It is classified into 3 major groups: simplex, junctional (JEB) and dystrophic. JEB are autosomal recessive genodermatosis and are split into Herlitz (JEB-H) and non-Herlitz subtype, JEB-H being the most severe form. Among the 3 diagnostic tools used to diagnose congenital mendelian bullous disorder – histopathology, epitope-mapping, germline mutation screen in proband – the latter has become gold standard. In JEB, majority of mutations occur in LAMB3, although syndromic JEB may be linked to mutations in ITG4 (associated with pyloric atresia) or plectin (associated with myopathy). To date, no clear genotype-phenotype correlation exists in JEB-H. In majority of cases, the disease is lethal in the first year of life; aerodigestive mucosa is most affected. Management of patients with JEB is supportive: trauma prevention, wound care, nutritional and infection control.

Conclusion: Neonatal blisters are suggestive of EB. Complications of JEB-H are severe and mostly lethal in infancy. Management is mainly supportive. Recently, a sustained positive answer to corrected keratinocyte stem cells transplantation was obtained in EBJ-H patient, paving the way for further hope in this dramatic disease.

Disclosure of Interest: None declared

CHANGE OF OUTPATIENT CARE OF CHILDREN AND ADOLESCENTS IN SWITZERLAND BETWEEN 2007 AND 2015

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Introduction: Primary care paediatricians [PCP], general practitioners [GP] and paediatric hospitals provide primary health care of children and adolescents in Switzerland. However, changes of primary health care over the past years have not been assessed in detail, although major system modifications have occurred (such as the introduction of DRG or adaptations of the tariff system). This master thesis examines the changes of outpatient care between 2007 and 2015.

Methods: This retrospective, descriptive study includes five databases. The work performance of PCP and GP was monitored by the data pool of *NewIndex AG* with exclusion of some cantons because of missing data. Outpatient hospital data was purchased from *SASIS AG*. Detailed outpatient consultation information for hospitals in the canton of Zurich was obtained by letter. Additional data was used from the *FMH* and *BFS*.

Results: Between 2008 and 2015, the number of PCP in Switzerland increased by 29.9% (GP only 12.2%). The female-male ratio grew by 57.0% from 0.89 in 2008 to 1.39 in 2015 while the number of male PCP remained constant. In the same period, the average working time of PCP decreased by 11.8% (GP 4.8%). Consequently, the real growth of PCP care was 12.2% (GP 7.1%) which outnumbers the subpopulation growth (3.3%). In 2007, children and adolescents consulted PCP and GP equally. Between 2015 and 2007, the total number of PCP consultations increased by 6.8% (in contrast to GP -29.9%). Thus, the average age when more GP were treating children in primary care than PCP increased from age 7.5 to 11 years [in other words, PCP see

more and more older children, fig.]. In 2014, outpatient hospital care demonstrated a high diversity between the cantons from 2.16 (Vaud) to 0.17 (Appenzell Inner-Rhodes) consultations per child and year. The same diversity was found for the growth of outpatient hospital care between 2007 and 2014: 1.7% (Geneva) to 163.1% (Neuchâtel). The number of outpatient consultations increased from 1% in the 0–5 to 12.2% in the 11–15 age group. In the canton of Zurich, non-emergency consultations in hospital increased by 73.2% and emergency consultations by 141.2% in 2015 compared to 2010.

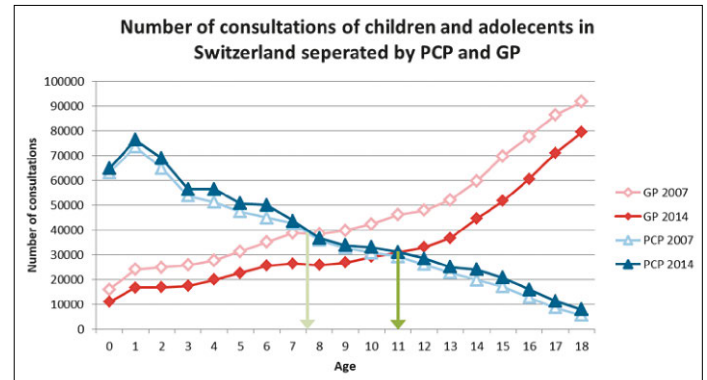


Figure 1

Conclusion: Outpatient care is undergoing major changes. Because of an increasing lack of GP in pediatric primary care, more and older patients are seen by PCP. Out- and inpatient care in hospitals and PCP care influence each other and should be seen as united. The development of this trend offers opportunities for pediatric primary care.

NaCHwuchs Prize: I wish to apply

Disclosure of Interest: None declared

P096

PIPADES.CH – A RESOURCE FOR PARENTS AND PROFESSIONALS TO PREVENT INJURIES IN PRESCHOOL CHILDREN

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Introduction: A secure environment and simple actions can reduce the risk of injuries in children, especially in preschool years. Parents and professionals need an easy access to reliable information.

Methods: In 2015, the Service de la santé publique of Canton de Vaud transferred the injury prevention program called Pipades to the CRIPE (Centre de référence des infirmières petite enfance). An assessment of the content of existing leaflets and website, completed with a literature review, led to a revision of the program with the following objectives: – provide reliable and up-to-date information about injury prevention that is relevant to families and to professionals; – increase the ways of accessing the information; – contribute to health professionals training on the topic.

Results: The new version of the website www.pipades.ch was launched in 2017. It is now easily read on smartphones, tablets and computers. Several sections can be searched with keywords. There was an average of 200 connections per month in 2017. We updated the Quick Reference Guide (“Aide-mémoire”; A4 leaflet) which explains the main risk of injuries at each developmental stage and gives simple advice to the parents. Available in 14 languages, it can be printed or ordered via the website. More than 22'000 leaflets were shipped in 2017 in Switzerland to pediatric practices, hospitals, visiting nurses, daycare centres, parents associations, educators, social services and to families. Between June 2016 and December 2017, the health visitors (infirmière petite enfance) of Canton de Vaud have discussed with parents of preschool children about injury prevention more than 15'000 times during home visits and group sessions.

Conclusion: Access to understandable, reliable and up-to-date information about risks of injuries and simple actions to prevent them is key for parents so they can secure the environment in which their children grow. Professionals use the leaflets and the website to support parents in their protective role.

Disclosure of Interest: None declared

P097

EVALUATION OF RISK FACTORS FOR FALLS IN YOUNG CHILDREN LESS THAN SIX MONTHS

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Introduction: Although many prevention strategies including education, training, creating safer environments, falls in children still a major cause of emergency room visits and admissions in pediatric hospitals.

Objectives: To investigate risk factors for falls in children aged <6 months and to propose effective policies to reduce this risk.

Methods: A retrospective study that included all patients aged <6 months who attended the emergency department of Children's hospital in Lausanne for falls between 2013 and 2017.

Results: A total of 372 patients had experienced falls, that is 0,23% of the emergencies attended in the study hospital. The most commonly affected part was the head (87,9%, N = 327). The most frequently mechanism was fall from parental bed (26,8%, N = 100). Falls from changing tables, parental and couches (56,7%, N = 211) were associated in 86% of cases with leaving the child alone without oversight. Fall from changing table (16,9%, N = 63) was the most frequently mechanism for children aged between 4 and 6 months (22,7%, N = 41) and was associated with hospitalisation in 92% of cases. The changing table is the factor mostly associated with injury risk (cerebral hemorrhage [N = 2] and cranial fracture [N = 2]). Parents of children falls from maxi cosi and stroller (23,6%, N = 88) did not tie them in 82 % of cases.

Conclusion: To decrease the occurrence of injuries caused by falls, strategies should target two axes: do not leave the child alone on the changing table or parental bed or couches and take care to tie the child in the maxi-cosi or the stroller. About the changing table we propose to use those with sides and ideally create protection systems with barriers.

Disclosure of Interest: None declared

P098

MENTAL HEALTH CARE NEEDS OF UNACCOMPANIED ASYLUM-SEEKING MINORS RESETTLED IN THE CANTON OF GENEVA: THE IMPORTANCE OF NETWORKING

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Introduction: Unaccompanied asylum-seeking minors (UASM) experience greater vulnerability both in terms of somatic and emotional status compared to other minor groups. Trauma experienced in the country of origin or during migration contributes to their altered social-emotional well-being which is further exacerbated by the relocation and the precarious social situation in the resettlement country. At their arrival in the canton of Geneva, all UASM are offered a global health assessment by a primary care physician in the Geneva University Adolescent Outpatient Clinic. If clinically indicated, they are further referred to a mental health professional whether at the same hospital or in collaboration with public outpatient mental health services. This study aimed to describe the mental health care needs of UASM.

Methods: Medical records of UASM who consulted at the Geneva University Adolescent Outpatient Clinic and at the Geneva Medical Pedagogical Institute were retrospectively investigated. We analyzed data concerning UASM (11–18 years) who arrived in the canton of Geneva in 2015–2016 and who were referred for psychological assessment. We summarized socio-demographic characteristics of the patients and we further analyzed data regarding the use of mental health services. We performed descriptive statistics using the IBM SPSS Statistics Program.

Results: Between 2015 and 2016, a total of 220 UASM arrived in the canton of Geneva (males 90%, mean age: 16.3). The great majority of them originated from Eritrea (40%) or Afghanistan (30%). Fifty-seven patients (25.9%) were referred to a mental health professional. The proportion of boys (26.8%) who needed an evaluation was higher compared to the respective proportion of girls (18%). Nearly one half originated from Afghanistan. Twenty-two percent were only once assessed whereas the rest 78% had regular follow-up. The median number of visits to the mental health practitioner was 7 (range: 1–79). Nearly one quarter (23%) visited at least once the emergency room for alteration of their emotional status. Four patients (93%) needed hospitalization.

Conclusion: UASM represent a high-risk population for mental health disorders and require specific mental care and treatment. Network collaboration between different services is crucial in order to facilitate UASM's access to mental health care services and to give them the opportunity to realize their full social-emotional potential.

Disclosure of Interest: None declared

P099

USING THE SYSTEMIC MODEL OF PARENTS AND CHILDREN INTERACTIONS TO CREATE AN ADAPTATIVE CONTEXT TO GO EASIER THROUGH TRANSITIONS

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Introduction: Over the years families have to stay stable facing in the same time internal changes caused by the membership development particularities and external pressures. All of these cause imbalances which require adaptation mechanisms to maintain the system cohesion. As systemic family therapist I have observed three qualities in the families where life is enjoyable. Those three are not present, or barely in families where one suffer for a long time. I have developed the systemic model of parents-children interactions based on these observations. (In French the "Modèle d'Observation Systémique des Interactions Parents-Enfants – MOSIPE[®]"). Use the initial PEB to remember it: sharing Pleasure, self-Estim, attention to Needs. (in French Needs = "Besoins"). Sharing Pleasure between the system members, gives as stability as flexibility and increases the desire to stay together. Having opportunities to receive self-Estim in these relations, gives more facilities to be different from one another (and equally to accept that the other one differs to ones), and to have different opinions, without feeling in danger. Self-Estim is good for the evolution of each system's member. Capacity to perceive and take into account the needs for other one gives evidences of the differentiation between the members of a system and develops there solidarity. These three qualities facilitate the family adaptability by giving flexibility to the system.

Methods: The MOSIPE: 2 types of use – To make the system interactions more flexible. – To estimate the adaptability of a system facing difficulties, in particular in term of child welfare.

Results: PEB gives an appreciation of the capacities to regulate the interactions. MOSIPE[®] is adapted to work with various types of families from various cultures, presenting various problems, with children of all ages from the birth through adolescence, and by professionals coming from educational environment or social sectors, or therapists. Thus, the use of this tool can be well integrated into multidisciplinary teams as well as multidisciplinary partnerships.

Conclusion: PEB it is a set of three fundamental regulators for systems crossing over existences crisis by limiting the risk of problem chronification. Using the MOSIPE is nothing more than organizing the reflection, the evaluation, the interventions, by following the PEB as the electrical wire.

This model described for families is usable to make the interactions more flexible within our teams and any human system.

Disclosure of Interest: None declared

P100

PERINATAL STRESS MODERATES THE LINK BETWEEN EARLY AND LATER EMOTIONAL SKILLS IN VERY PRETERM-BORN CHILDREN: AN 11-YEAR-LONG LONGITUDINAL STUDY

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Introduction: Very preterm (VPT) birth refers to an early stressful event putting children at heightened risk for emotional difficulties. However, there is an important individual variability, leaving unexplained why some VPT children do not develop emotional difficulties, while others develop such difficulties in the early years or later in life. In this study, we examined whether perinatal stress is a risk factor explaining heterogeneities in emotional problems in VPT children.

Methods: Thirty-six VPT children and 22 full-term born (FT) children participated in an 11 year-long study. Risk for perinatal stress was assessed at birth with the Perinatal Risk Inventory. Mothers reported children's emotional difficulties at 18 months of child age on the Symptom Checklist and at 11 years on the Child Behavior Checklist.

Results: Results indicated significant differences in emotional scores at 11 years not only between VPT and FT children but also between the low and high perinatal stress groups. More importantly, emotional scores at 18 months influenced variability in internalizing scores at 11 years *only* in VPT children with high perinatal stress.

Conclusion: Although prematurity affects the emotional abilities of preadolescents, the link between emotional skills in early and later childhood is moderated by the severity of perinatal stress. In particular, VPT children who are born with more complications, and as such experience a more stressful perinatal environment, are more likely to show emotional difficulties at preadolescence.

Disclosure of Interest: None declared

P101

YOUNG CARERS IN SWITZERLAND – WHY AND HOW SHOULD PHYSICIANS IDENTIFY THEM?

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Introduction: In Switzerland, the issue of young carers (young people under the age of 18) who take on significant or substantial caring tasks and levels of responsibility that would usually be associated with an adult – is still hardly recognised. Even physicians dealing with patients who are sometimes accompanied by young carers – especially during school hours – rarely identify these children and adolescents or are aware of the phenomenon. In our presentation, we show the need of raising awareness among health care professionals due to the high numbers of young carers in Switzerland and present red flags that will help doctors to identify young carers more easily.

Methods: An online survey of children in school grades 4 to 9 (mostly aged 10 to 15) was conducted in order to determine the prevalence of young carers in Switzerland using a 2-stage stratified sampling approach. The sample was allocated proportionally to the different language speaking areas of the country. Schools were sampled on a probability proportional to size basis.

Results: 4,082 respondents were drawn from 230 schools. 3,991 respondents were included in the analysis and of these 307 (7.7%) were identified as young carers. The population estimate of prevalence was 7.9%. These results suggest that there are around 38,400 young carers in school grades 4 to 9 in Switzerland. Extrapolating to the broader 9 to 16 age group gives a figure of almost 51,500. The Multidimensional Assessment of Caring Activities (MACA-18) questionnaire showed that 40% of the children identified as young carers carried out a 'high' or 'very high' level of care, and that this care was performed across a range of different activities, including, personal, emotional and domestic care. The mean age of young carers in this population of children was estimated to be 12.3 years (95% CI = 12.0–12.6) and the mean age at which they started their caregiving role was 9.0 (95% CI = 8.6–9.3).

Conclusion: Based on the survey results and earlier qualitative research in Switzerland, red flags for health care professionals will be highlighted in order to identify and provide support for young carers and their families.

NaCHwuchs Prize: I wish to apply

Disclosure of Interest: None declared

P102

PARENTS' UNDERSTANDING AND SATISFACTION OF MEDICAL INFORMATION WHEN ATTENDING THE «HÔPITAL DE L'ENFANCE DE LAUSANNE»

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Introduction: The objectives are to identify how medical information such as diagnosis, treatment(s) and reason(s) of hospitalization have been understood by parents when attending the «Hôpital de l'enfance de Lausanne»; to evaluate satisfaction regarding this information; and to assess how communication could be improved.

Methods: A qualitative study was conducted including semi-structured interviews with 23 French-speaking parents of children aged from 2 months to 5 years (median: 18 months). Interviews were audiotaped, transcribed, coded and analyzed in order to meet the objectives and to develop recommendations.

Results: Parents reported a good understanding of medical information. However they indicated some obstacles to understanding such as caregivers lacking to verify parents' understanding, parental stress. Many facilitators were also identified by parents such as simple language, clear explanations, availability of caregivers to answer questions. Overall satisfaction regarding medical information was

excellent, although parents gave some interesting ideas to improve communication, for example: using pictures or digital media, a more complete website or a walk-in day for parents. In fact this study showed 4 steps that are important for parents to correctly understand the medical information. First step is about the content of information. The language for instance was described as clear, with medical terminology but always given with explanation. Secondly the form was also important for a good understanding. Some parents preferred to receive oral and written information instead of oral information only. According to them it could improve their understanding. Thirdly the parents should be prepared to receive information. For example, parental stress should be first identified by paediatricians, then they should reassure them in order to finally be able to receive information. The fourth step is the assessment of understanding. The teach-back technique, for example, takes part of this last step. It could be used to verify and improve parents' understanding. In fact parents explain with their own words what they have understood, then the caregiver corrects and completes and the parents explain it again.

Conclusion: Parents' understanding was described as good and they were satisfied. Paediatricians should be aware of 4 essential aspects when communicating with parents: the content, the form, the parents' ability to receive information and the assessment of understanding.

Disclosure of Interest: None declared

P103

SUICIDE IN CHILDHOOD AND ADOLESCENCE: COMPARATIVE ANALYSIS OF RISK FACTOR IN THE SCIENTIFIC LITERATURE

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Introduction: In the World every thirty second a person commits suicide. In the last sixty years we observed a global increase in the number of suicide, but we can also see a decrease of young suicide in the beginning of 90's. The genre, the geographical area were we live, but there are also independent risk factor such as sexual orientation, the family and the community.

Methods: In the present study 98 articles focusing on juvenile suicide, published from 1993 to 2017, were analyzed. The authors focused on the comparative analysis of general risk factors, gender-related risk factors, individual history, sexual orientation and the controversial role of the family in the genesis of suicidal ideation; they check also the role of the community, belong to minor ethnic group and the role of "suicide contagious".

Results: Female teenager are incline to attempt suicide, instead of male teenager that committed suicide. Furthermore the risk increase in male adolescent if they are sexual attracted from other male and in female adolescent if there is an abuse of drugs substance. Most of the teenagers that committed suicide were depressed at the time of death and depression is one of the main predictors of suicidal ideation. The family plays an important role for the adolescent: family's problems can considered a risk of factor for adolescent suicide, but a good presence of a family can also be a protective factor from suicide. Communities, like religious, school, spiritual communities can also be considered protective factors which allow adolescents to confront with other adolescent or adults and to discuss with them their existential problems. It is important to considered television and internet a risk factor for suicide, in particular for the suicide contagious: WHO published a recommendation for mass media in order to avoid the "charm of suicide". The last, but not the least, risk factor that we can considerer is a minor group: rural communities and ethnic minorities have a suicide rate that is two to ten times higher than the average.

Conclusion: What brings to an high rate of suicides? There are two ideas: the first explains that suicidal behaviors are associated with personal pessimism, hopelessness and lack of control; the second explain the existence of discrepancy between the reported self-happiness level, the level of psychological stress and mental illness. In conclusion, suicide is a complicated phenomenon, result of the interaction of biological, psychological, social or environmental factors.

NaCHwuchs Prize: I wish to apply

Disclosure of Interest: None declared

PF1

HOLES IN BONES: MAYBE SIMPLE BUT GOOD TO KNOW (POST TRAUMATIC CYST-LIKE CORTICAL DEFECTS)

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Introduction: Small non-expanding cyst-like cortical defect after fracture, found either in routine follow-up examinations or as incidental finding following reinjury, is a rare occurrence in children. We present a 6-year-old girl who suffered from a distal left radius buckle fracture that was treated by 3 weeks cast immobilization without control X-Ray. At the age of almost 8 years old she presented a compound greenstick/buckle fracture of the same bone which was successfully treated by cast immobilization as shown by control X-Ray 5 weeks later. After seven months, she hurt again her left wrist and an X-Ray did not show a fracture but revealed two 4 mm round well-circumscribed lucent lesions without surrounding sclerosis close to the site of the previous fracture. A short-arm cast for analgesic purpose was worn for 13 days with a good clinical evolution. A control X-Ray 3 weeks later showed no changes in lesions and 6 months later revealed complete regression of cystic lesions.

Methods: Pubmed was searched with keywords such as: "post-traumatic/posttraumatic; cyst-like lesions/defects; fractures; bone; children". Case reports of expanding or destructive cyst-like lesion were excluded.

Results: We found 23 studies published between 1978 and 2016 on pediatric post-traumatic non-expanding bone cysts, for a total of 42 cases including ours. Median patient age is 8 years (range: 1,5–15). The most common localization is the distal radius (37/42, 88%) and they generally appear after a greenstick fracture (25/42, 60%). Median time between fracture and lesion discovery is 3 months (range: 1–45). Post-traumatic bone cysts are totally asymptomatic. They are usually seen within ossified layers of the new subperiosteal bone. They are often multiple, usually less than 10 mm of diameter and they disappear progressively. The etiology remains controversial. Their content is likely to be blood, fat or an admixture of both.

Conclusion: Small post-traumatic non-expanding cyst-like cortical defects are a known but rare condition compared with the frequency of fractures in children. Pediatricians need to be informed of these lesions in order to avoid unnecessary investigations and reassure patients and their families.

Disclosure of Interest: None declared

PF2

NEWBORN SCREENING FOR INHERITED METABOLIC DISORDERS, 45 YEARS IN FRANCE

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Introduction: Newborn screening is important for the early detection of many congenital genetic and metabolic disorders, aimed at the earliest possible recognition and management of affected newborns, to prevent the morbidity, mortality, and disabilities associated with an inherited metabolic disorder. It's began officially in France in 1972. It is organized by the French Association for Screening and Prevention of Infant Handicaps (AFDPHE) under the Ministry of Health. This comprehensive system includes; testing, education, follow up, diagnosis, treatment, management, and evaluation.

Methods: In France, 5 diseases are currently subject to newborn screening using biological tests on dried blood spots (Guthrie card): phenylketonuria in 1972, congenital hypothyroidism in 1978, sickle cell disease in 1985, congenital adrenal hyperplasia in 1995, cystic fibrosis in 2002 and MCAD deficiency in 2011.

Results: Spectacular results with a 100% coverage of newborns and full support of all patients by the regional health teams except MCAD deficiency. In France, For more than 45 years, 33,6 million newborns were screened. In 2016 this corresponded to: 1 / 20,826 for PKU, 1 / 2465 for CH, 1 / 23,981 for CAH, 1/525 for SCD in Overseas Departments, and 1/823 in metropolitan France and 1/6091 for CF. The average cost of multi-screening is 10.5 € per newborn

Conclusion: These data suggest that the newborn screening in France was effective and cost-efficient.

NaCHwuchs Prize: I wish to apply

Plaschke Prize: I wish to apply

I have read and understand application rules: Yes

Disclosure of Interest: None declared

PF3

TONGUE LACERATIONS IN CHILDREN – TO SUTURE OR NOT?

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Introduction: Tongue lacerations (TL) are common in children, occurring mostly from falls or sport injuries. The optimal treatment for TL is a challenge for pediatricians due to controversial recommendations and missing current guidelines. It remains unclear which TL benefit from suturing and for which spontaneous healing is a promising alternative. In recent years, the treatment of choice in our pediatric emergency department (ED) changed from generally suturing the wounds to advising secondary wound healing more frequently. The aim of this study was to analyze the treated TL at our ED in order to develop guidelines for an optimal management of TL in children.

Methods: This retrospective study was conducted to assess TL at the ED of the University Children's hospital in Zurich from January 2010 to August 2015. All families were contacted for informed consent and photo documentation of the healed tongue. Clinical records of all included patients were reviewed and different variables defined and analyzed.

Results: A total of 73 children with TL were included, 75.3% were boys and the mean age 4.0 ± 2.6 years. The mean size of the lacerations was 12.4 ± 8.3 mm with affected tongue borders in 51 cases (69.8%) and a through-and-through laceration in 23 patients (31.5%). A primary wound closure was performed in 12 children (16.4%); these wounds were larger compared to the secondary wound healing group (21.0 ± 10 mm versus 10.8 ± 6.8 mm), presented more frequently gaping wound edges with the tongue at rest (91.7% versus 32.8%) and showed more often through-and-through lacerations (91.7% versus 19.7%). The group with wound suturing needed longer to recover (14 days compared to 7 days) and had a higher rate of complications (25% versus 3.3%).

Conclusion: TL smaller than 2 cm without involving the tongue borders and not gaping with the tongue at rest do not require an intervention. The Zurich Tongue Scheme was developed to support clinicians which TL need suturing.

Disclosure of Interest: None declared

PF4

WHEN TO SUSPECT ETHYLENE GLYCOL INTOXICATION?

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Introduction: Ethylene glycol (EG) is a rare cause of intoxication that presents with distinctive hallmarks that practitioners should be aware of to avoid missing this potentially lethal affection.

Methods: Case report: A 16-year-old boy was admitted for an acutely altered level of consciousness. His mother reported progressive slurred speech and abnormal gait. The patient denied any ingestion of toxic products and had no alcoholic foetor. Temperature was normal, hemodynamics were stable. Neurological examination showed no focal anomaly but a diffuse weakness. Blood gas tests revealed metabolic acidosis (pH 7.27, pCO₂ 2.4 kPa, HCO₃⁻ 8 mmol/l) and elevated lactate (30 mmol/l). Further analyses ruled out any electrolyte disturbances, hepatic or renal failure, inflammatory syndrome or alcoholic intoxication. Routine toxicology screen was negative. Anion gap was elevated (36). We subsequently ordered EG dosage, which came back positive (1'345 mg/l). The antidote fomepizole was given 4 hours after admission. The patient developed progressive neurological impairment, with a drop in Glasgow Coma Score to 3 and Kussmaul breathing, with progression of his metabolic acidosis (pH 7.19, pCO₂ 1.7 kPa, HCO₃⁻ 4 mmol/l) and hyperkalemia (7.4 mmol/l). He was intubated and underwent a cerebral CT that ruled out any intracranial bleeding. He progressed to acute renal failure and required hemodialysis and haemodiafiltration for 7 days, with normalization of his renal function after 14 days.

Results: Discussion: EG is mainly found in antifreeze solutions. It is metabolized into oxalic and glycolic acid, which are responsible for its toxicity. Initial clinical manifestations mimic inebriation, are followed by

cardiopulmonary symptoms and eventually multi-organ failure, related to acute renal failure. Biological hallmarks evolve from an osmotic gap, to metabolic acidosis with elevated anion gap, and later, electrolytes imbalance in case of kidney failure and calcium oxalate crystals in the urine. Lactate may be falsely elevated. Treatment consists in prompt administration of the alcohol dehydrogenase antagonist fomepizole, which is the antidote of choice, hemodynamic support and hemodialysis. Complications are mainly related to kidney failure.

Conclusion: EG intoxication should be suspected in a patient with symptoms of alcoholic intoxication without alcoholic foetor, in the context of severe metabolic acidosis and possibly falsely elevated lactate. Fomepizole should be promptly administered when EG intoxication is suspected.

Disclosure of Interest: None declared

PF5

CONGENITAL BILATERAL PSEUDARTHROSIS OF THE CLAVICLE

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Introduction: Congenital pseudarthrosis of the clavicle is a very rare entity of unknown etiology with only 200 cases described worldwide. Pathogenesis is thought to be related to the embryology of the clavicle. It is a painless prominence in the anterior superior chest wall in the absence of trauma, noted soon after birth or during childhood, usually without functional impairment. It may be easily confused with the more common traumatic clavicular fracture, particularly in a situation where there was some degree of dystocia and mechanically assisted delivery.

Methods: We report a case of bilateral pseudoarthrosis of the clavicle diagnosed in the nursery soon after birth, in a term newborn presenting right shoulder's dystocia at delivery. At the first clinical examination it was only noted double loss of continuity of the clavicles with no other congenital deformities or abnormalities.

Results: A chest radiograph performed due to suspicion of bilateral clavicular fracture revealed findings consistent with bilateral pseudoarthrosis, in particular a prominent callus formation ("hypertrophic") around the clavicular non-unions (fig. 1). The newborn did not present functional impairment or signs of pain during manipulation. No other medical measures were provided at discharge; a surgical follow-up will happen when the child is 1 year old. In congenital pseudoarthrosis of the clavicle, the two primary ossification centers in the developing clavicle fail to unite during embryogenesis; clavicular fracture, on the other hand, is the most common bone injury with an overall incidence of 3–42% among the skeletal birth fractures. Discerning between a clavicular fracture and congenital pseudoarthrosis is not simple, as they have a similar radiological appearance; however, one should consider a diagnosis of pseudoarthrosis if the suspected clavicular fracture does not show evidence of healing on sequential radiographs. A fractured clavicle heals well without any sequelae, while pseudoarthrosis persists throughout life. Surgical treatment should be considered if the patient is symptomatic or presents dysfunction of the arm.

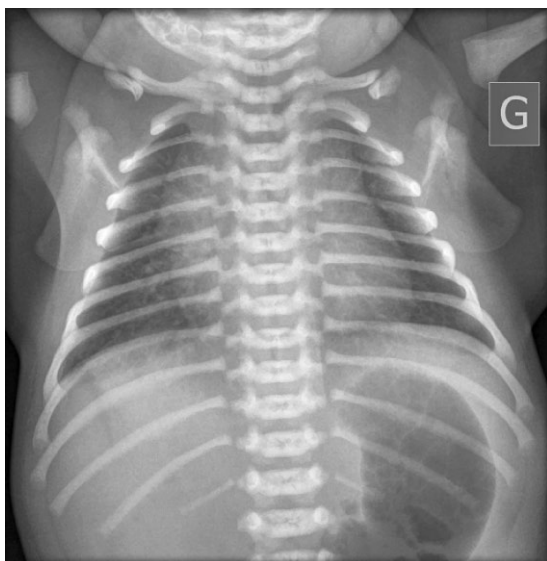


Figure 1

Conclusion: Recognition of this entity and its nontraumatic etiology is important since the workup and outcome are very different than in traumatic clavicular fracture. The management of pseudoarthrosis of the clavicle can be reasonably conservative, with operative repair delayed until early childhood.

Disclosure of Interest: None declared

PF6

HYDROMETROCOLPOS IN A CHILD WITH PERSISTANT ABDOMINAL PAIN AND NO BULGING HYMEANEAL MEMBRANE

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Introduction: Congenital imperforate hymen (IH) is the most common vaginal outflow obstruction in females, with a prevalence of 0.05 to 0.1% in girls and adolescents. Physical examination findings are not always classic. Here, we present such a case.

Methods: An 11-year-old girl consults the ED for persistent abdominal pain despite multiple consultations in the past months. She initially presented with epigastric pain and constipation, evolving as lower abdominal pain within the last week, characterized as cramping, with loss of appetite and polyuria. She is premenarchal and denies sexual activity, vaginal discharge or fever. She has tried several pain medications and laxatives with minimal effect. On physical examination, she is well appearing with normal vital signs. Her abdominal examination reveals mild tenderness elicited on deep palpation of the bilateral lower quadrants and suprapubic area, no obvious mass, no guarding or rebound tenderness. Her genitalia are a Tanner 4, with no protruding membrane. Laboratory shows no abnormalities. Pelvic ultrasonography reveals a hydrometrocolpos and MRI, performed to provide superior anatomic detail, shows no other urogenital abnormalities. After an uncomplicated hymenectomy and evacuation of the retained liquid, she is discharged without residual pain.

Results: IH is generally diagnosed during adolescence with primary amenorrhea, cyclic abdominal, back or pelvic pain in premenarchal females, associated with constipation and urinary retention due to the mass effect. Commonly, there is a discrepancy between advanced secondary sexual characteristics and amenorrhea, a translucent thin membrane inferior to the urethral meatus, appearing blue and bulging when performing Valsalva maneuver. Pelvic ultrasonography is indicated as the initial diagnostic test. MRI confirmation is imperative prior to any attempted surgical repair to exclude other genitourinary tract anomalies or other outflow obstructions such as a low-lying transverse vaginal septum. Surgery should be delayed until puberty to allow the hymen to become estrogenized and avoid scarring.

Conclusion: The differential diagnosis of persistent abdominal pain in an amenarchal female with developed secondary sexual characteristics should include IH. An external examination may not reveal a bulging membrane, and early ultrasound imaging may be necessary to coin the diagnosis, and to prevent delays in appropriate care.

Disclosure of Interest: None declared

PF7

AN INFREQUENT THOUGH CLASSICAL CAUSE OF RESPIRATORY DISTRESS IN PREMATURE NEWBORNS – A CHLAMYDIA TRACHOMATIS PNEUMONIA

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Introduction: *Chlamydia trachomatis* (*C. trachomatis*) is the most common cause of sexually transmitted genital infection in adults. Vertical infection to the newborn can cause conjunctivitis and pneumonia. *C. trachomatis* pneumonia typically presents with nasal obstruction, tachypnea and paroxysmal staccato cough. Its diagnosis can be very challenging as symptoms tend to be unspecific. Moreover, the incubation period varies widely, between 4 and 12 weeks.

Methods: We report on a girl born vaginally at 32 0/7 weeks postmenstrual age, due to early labor of unknown origin. The mother was 33 years old, G1/P0, without relevant history. The newborn necessitated CPAP at birth for less than 24 hours. At 3 weeks of age, she developed signs of respiratory distress and increasing apnea-bradycardia events. Paroxysmal cough was noted, as well as bilateral crackles at auscultation, however without upper respiratory symptoms.

Blood analysis showed peripheral eosinophilia. A bilateral interstitial thickening was noted on the chest radiography. The review of mother's records revealed a positive *C. trachomatis* vaginal test collected on the birth day. Treatment with azithromycin was immediately started and the diagnosis of *C. trachomatis* pneumonia confirmed by positive polymerase chain reaction of the infant's nasopharyngeal secretions.

Results: Nowadays, *C. trachomatis* is a rare cause of respiratory distress syndrome in the Neonatal Care Unit in Switzerland and is therefore often forgotten in the differential diagnosis. It is recommended to start treatment as soon as diagnosis is suspected but *C. trachomatis* pneumonia has unspecific symptoms, making it difficult to distinguish from other more common respiratory infections. Peripheral eosinophilia can help to suspect it. Of course, a history of untreated maternal *C. trachomatis* infection is very helpful but may not always be available.

Conclusion: *C. trachomatis* infection should be part of our differential diagnosis of neonatal respiratory distress syndrome as symptoms and radiological findings are unspecific. Some countries (as the United States) have implemented a systematic screening of all pregnant women on the first prenatal visit with a dramatic decrease in perinatal chlamydia infection. In Switzerland though, the screening for Chlamydia trachomatis infection in pregnancy is limited to women with risk factors or those presenting with premature labor of unknown origin.

Disclosure of Interest: None declared

PF8

ANEMIC NEW BORN: KEEP CALM AND THINK KLEIHauer, A CASE REPORT

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Introduction: Feto-maternal hemorrhage (FMH) is the circulation of blood cells from fetus to mother. FMH can occur at any time during pregnancy. Its clinical presentation depends on the quantity of fetal blood loss. Neonatal presentation varies from asymptomatic baby to stillbirth. Massive FMH is usually defined as more than 30 ml of fetal blood transfused (3/1000 pregnancies). Neonatal morbidity and mortality have been related to a transfusion of 80 ml or more (1/1000 pregnancies). Identified risk factors for developing massive FMH are: abdominal trauma, external obstetrical procedures, placenta abnormalities, preeclampsia, choriocarcinoma and monochorionic-monoamniotic twins. However, more than 80% of massive FMH remain unexplained. The Kleihauer-Betke test quantifies the fetal Hb (HbF) in the maternal blood and allows identifying and quantifying the degree of the FMH.

Methods: We report a massive FMH at the maternity of the hospital of Yverdon-les-Bains.

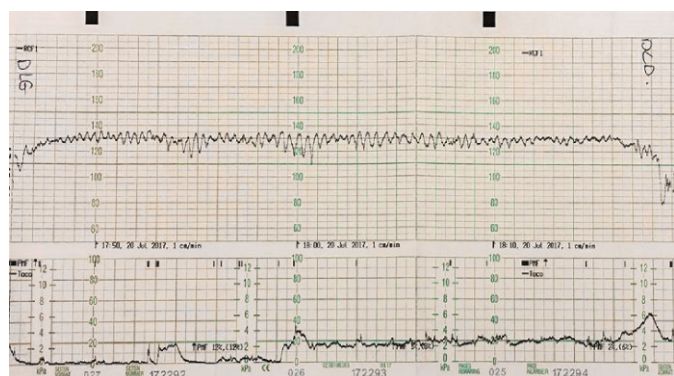


Figure 1

Results: A 28-years-old, nulliparous, A+ woman was seen at 38 gestational weeks (GW) for an ordinary obstetrical control. The pregnancy was normal except for fetal growth break at 36 GW and a maternal normochromic normocytic hyporegenerative anemia at the first trimester supplemented with iron. During this control, the mother reported decreased fetal movements for several days. A sinusoidal heart rate was observed which led to an emergency C-section (fig. 1). A 2830 g baby boy was delivered. He presented an extreme pallor, a hearth rate of > 100/ min, spontaneous respiration with oxygen support, hypotonicity and hyporeactivity. The APGAR score was 4/5/6 and the cord pH 7.17/7.22. Severe anemia was confirmed (cord Hb: 1.7

g/dl) and he received emergency transfusion of O Rh negative blood. He was transferred to the Neonatal Intensive Care Unit of Lausanne. Massive FMH was confirmed by the Kleihauer test that showed 10% of HbF, the equivalent of 450–500 ml of fetal blood (twice the total blood volume of the fetus). The new born presented a transient multiorgan dysfunction and was discharged after 13 days without apparent sequelae.

Conclusion: The occurrence of massive FMH in a physiological pregnancy, near to the term and without apparent reasons, remains a challenge for obstetricians and pediatricians. It is known that the physician's awareness improves the detection of FMH. We think therefore that better physician's awareness and a multidisciplinary management will help improve the management of massive FMH in peripheral pediatrics services.

Disclosure of Interest: None declared

PF9

MAXIMUM POST-NATAL SODIUM FLUCTUATION IN PRETERM NEONATES INCREASES WITH DECREASING GESTATIONAL AGE

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Introduction: Neonates experience loss of water and can develop hypernatremia during the first days of life. Hypernatremia seems to be more frequent in preterm neonates with low gestational age (GA) due to increased water loss. Hyponatremia in preterm neonates may also occur due renal salt wasting as a result of the inability of immature kidneys to produce adequate urine tonicity. Both hyponatremia and hypernatremia and in particular fluctuations of serum sodium are associated with increased risk of serious complications such as intracerebral hemorrhage. The purpose of this study is to identify factors that influence sodium fluctuations during the first 28 days of life in preterm neonates with GA <32 weeks.

Methods: This retrospective study included all preterm neonates with GA <32 weeks, born between 2007 and 2014 at the neonatal unit of the University Children's Hospital in Berne. We calculated the difference between sodium at baseline (measurement within first day of life) and observed maximum sodium and tested covariate effects on this measure of maximum post-natal sodium fluctuation. GA was categorized as <26, 26–27, 28–29, and ≥30 weeks of gestation, respectively. The current covariate analysis focused on the two key factors GA and gender. Other factors such as delivery mode, antenatal steroids, single or multiple birth will be investigated at a later time point.

Results: A total of 980 preterm neonates with a mean GA of 29.13 (± 2.1) weeks and a total of 21854 sodium measurements were eligible to be included in the study. Maximum observed sodium was observed at a mean post-natal age of 2.7 days. Sodium at baseline was similar across GA groups neonates (p >0.05), whereas maximum sodium fluctuation did significantly differ between GA groups (<26 vs. 26–28 weeks, p <0.05; 26–28 vs. 28–30 weeks p <0.01; 28–30 vs. >30 weeks p >0.05), indicating that maximum sodium fluctuation increases with decreasing GA. Neither sodium at baseline nor peak sodium (p >0.05) were different between male and female neonates.

Conclusion: Consistent with immature regulation of sodium and water balance maximum fluctuation increases in with decreasing gestational age.

Disclosure of Interest: None declared

PF10

PRENATAL COUNSELING AT THE LIMITS OF VIABILITY: WHICH FACTORS INFLUENCE CAREGIVERS IN A MULTIDISCIPLINARY AND INTERPROFESSIONAL APPROACH?

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Introduction: The Swiss Society of Neonatology has issued guidelines concerning the perinatal care of preterm infants born at the limit of viability for specialized professionals. A thorough prenatal interview with the parents-to-be must enable the decision about a reasonable care of the newborn, which has to be adapted from case to case. The interview is based on epidemiological data and ethics. More specifically, it rests on the survival possibility of the preterm infant with an acceptable quality of life, whose boundaries depend on personal factors. The goal is to describe the approach to prenatal counseling at the limits of viability for the professionals involved by exploring their

role, implication, and the scientific or personal references they adopt to inform and support parents.

Methods: The sample is represented by midwives, obstetricians and neonatologists of the CHUV Maternity unit. This research comprises two phases. The first consists of an online questionnaire which aims at delimiting the sample of participants and at identifying a subject matter. The resulting subject is then explored in the second phase which takes the form of a focus group.

Results: 33 of the 92 invited caregivers have completed the questionnaire and 5 of them have participated in the focus group. The role played by each professional group in the handling of situations presenting risk of premature childbirth is distinct and known to each participant of the counseling. However, the decision makers are not clearly identified. The participants of the prenatal counseling are well aware of the importance of the presence of each member, although it is sometimes difficult to reconcile the different positions. The caregivers' portrayal of the premature newborn's risk of death and sequela does not always correspond to current data, and there is evidence of a general tendency to overestimate risks, especially among midwives. The most important personal factors influencing the decision are their own conception of the quality of life as well as their individual experience. Nonetheless, the definition of the quality of life does not appear to be the same for each of the three groups.

Conclusion: In order to allow an enlightened decision, it is essential that caregivers provide correct information to parents in an appropriate way. Furthermore, to optimize the quality of prenatal counseling, it is crucial to improve the caregivers' preparation process and to assure that the perinatology team members proceed with unanimous consensus.

Disclosure of Interest: None declared

PF11

A HEALTHY BOY WITH AN UNUSUAL CONJUNCTIVITIS

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Introduction: Conjunctivitis remains one of the common complaints seen in a general pediatric practice. The etiology is generally viral and commonly seen in children below school age. This report describes an unusual case of oculoglandular infection (also known as Parinaud's oculoglandular syndrome) secondary to a tick born Tularemia infection.

Methods: A 4 year-old boy, resident of the Broye District in the Canton of Fribourg, presented to our ER department 5 days after a witnessed tick bite on his right upper eyelid. Within 48 hours, this otherwise healthy youngster developed palpebral erythema, pain and conjunctival purulent discharge. Despite local treatment, fever followed along with worsening erythema, chemosis and purulent discharge. Significant anterior and posterior cervical regional lymphadenopathies were noted. Amoxicillin-clavulanic acid IV was initiated and antibiotics administered for a total of 7 days as clinical improvement was gradually observed. However residual symptoms persisted beyond 10 days. Further investigations (CT imaging, serologies for tick born infections and cat scratch) were performed. On day 17 a positive antibody testing for Francisella Tularensis was reported as consistent with active infection (Laboratoires médicaux Dr Risch). IV Gentamycin treatment was initiated for 7 days, followed by Ciprofloxacin PO for an additional 3 days. Full recovery was thereafter noted; no sequelae were found on an ophthalmology follow up (>30 days).

Results: The causes of Parinaud's oculo-glandular syndrome include Bartonella Henselae, Herpes simplex infection and Francisella tularensis. The latter is the cause of a known though rare tick borne illness. The Swiss Federal Office of Public Health maintains an on-line registry of such tick borne illnesses and their geographic distribution throughout the 26 cantons. The clinicians were thus able to establish their differential diagnosis based on the child's place of residence and potential regional zoonosis as tracked by up-to-date Federal data.

Conclusion: Patient history taking remains one of the important cornerstones of medical practices especially in this age of increased human mobility. Our pediatric case, a rare case of tularemia, aims to emphasize the importance of a thorough history-taking, including a travel history. Data on local current infectious diseases that might be essential to the differential diagnostic process are readily accessible on-line and should be consulted.

Disclosure of Interest: None declared

DISTINCTIVE FEATURES OF CAMPYLOBACTER ENTERITIS MIMICKING ACUTE ABDOMINAL EMERGENCY: – A CASE-CONTROL STUDY

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Introduction: *Campylobacter* spp. is a frequent cause of gastroenteritis, presenting in some patients as acute abdominal emergency. Here we describe the distinctive clinical characteristics of these patients.

Methods: We designed a retrospective single-center case-control study. Children and adolescents under 18 years of age who had positive stool cultures with *Campylobacter* spp. during the period from June 1, 2008 and May 31, 2016 were identified from our database. Patients who had undergone abdominal radiographic investigation (ultrasound or computed tomography) or received a surgery consultation were included as "cases". All other patients were controls. Demographics, clinical characteristics and management were compared between cases and controls.

Results: 141 patients with *Campylobacter* spp. positive cultures were included in the analysis. 19 patients had received abdominal imaging or surgical consultation. Median age was 8 years (IQR 2–12.8 yr), and 60 female patients (43%) were included. The groups were matched for age and gender. Diarrhea was less frequent among cases (14/19, 74%) than controls (117/121, 97%) (p = 0.02). Cases reported a lower sense of general wellbeing than controls (8/18, 44% vs 8/108, 7%; p < 0.001). Localized pain (9/18, 50% vs 20/115, 17%; p = 0.002) and abdominal tenderness (3/18, 11% vs 0/111; p = 0.02) were more common among cases than controls. No patient underwent surgical intervention.

Conclusion: We identified a subset of patients with *Campylobacter* spp. gastroenteritis who present as acute abdominal emergency. The clinical presentation of such patients is characterized by decreased likelihood of diarrhea and increased frequency of localized abdominal pain and abdominal tenderness.

NaChwuchs Prize: I wish to apply
Disclosure of Interest: None declared

Table 1. Clinical and demographic characteristics of the study population

Characteristic	All subjects (n=141)	Controls (n=122)	Cases (n=19)	P-value
Gender, n female/total (%)	60/141 (43)	51/122 (42)	9/19 (47)	0.6
Origin, n Switzerland (%)	80/141 (57)	69/122 (57)	11/8 (58)	0.9
Age, median years (IQR)	8(2-12.8)	7.5 (3-12.8)	9.4 (2.8-3.6)	0.6
Comorbidities (%)	16/118 (14)	12/99 (12)	4/19 (21)	0.3
Duration of symptoms, median days (IQR)	3 (1-4)	3 (1-5)	3 (1-3)	0.3
Maximum temperature, mean days (SD)	39.2 (0.6)	39.2 (0.6)	39.2 (0.6)	0.6
Abdominal pain, n/total (%)	101/132 (77)	87/114 (76)	14/18 (78)	1
Localized pain, n/total (%)	29/133 (22)	20/115 (17)	9/18 (50)	0.002
Tenderness, n/total (%)	2/129 (2)	0/111	3/18 (11)	0.02
Rebound pain, n/total (%)	5/112 (4)	3/94 (3)	2/18 (11)	0.2
Diarrhea, n/total (%)	131/140 (94)	117/121 (97)	14/19 (74)	0.002
Vomiting, n/total (%)	59/118 (50)	52/104 (50)	7/14 (50)	1
Fever, n/total (%)	111/132 (84)	97/114 (85)	14/18 (78)	0.5
Loss of appetite, n/total (%)	62/72 (86)	53/63 (84)	9/9 (100)	0.3
Diminished general state, n/total (%)	16/126 (13)	8/108 (7)	8/18 (44)	<0.001
Dehydration, n/total (%)	12/141 (9)	10/122 (8)	2/19 (11)	0.7

Legend: IQR: interquartile range; SD: standard deviation

Table 1

PF13

IMMUNISATION AGAINST HEPATITIS A IN MIGRANT CHILDREN: THREE VACCINATION STRATEGIES, A RETROSPECTIVE STUDY

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Introduction: Hepatitis A is endemic in many countries. Swiss guidelines recommend vaccinating patients native from endemic areas who might temporarily return. In Geneva's Hospital, migrant children are screened and vaccinated if seronegative. Since Hepatitis A prevalence is decreasing worldwide, more children are seronegative at arrival, questioning the benefits of systematic serology. Other Swiss hospitals vaccinate systematically, regardless of serostatus. This study's aim is to assess migrant children's immunity according to origin and age, and the cost-effectiveness of different immunization strategies.

Methods: We retrospectively analysed 329 children's serostatus (aged 1 to 16) between 2012 and 2015, using ELFA (Enzyme-Linked Fluorescent Assay) method. Serology and vaccine costs were based on local prices. Groups were compared with chi-squared test and the age-seropositivity relationship was studied with linear regression.

Results: The predominant regions were the Eastern Mediterranean and European Regions with mostly negative serologies (71% and 83%) and the African Region with mostly positive serologies (79%). Immunity varied depending on birth country. Regardless of regions, seropositivity increased with age (P <0.001). The most cost-effective vaccination strategy was an individualized approach based on age and origin, reducing costs by 2% compared to systematic serology and by 17% compared to systematic vaccination.

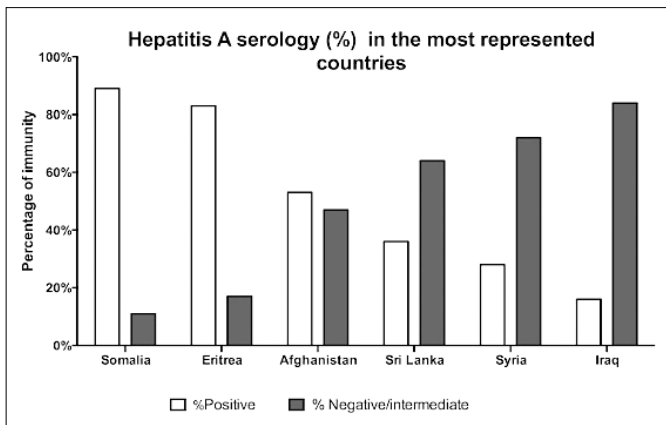


Figure 1

Conclusion: Many migrant children older than 5 years old are seronegative and at risk of clinical infection. New guidelines according to age and origin should be defined to reduce immunization costs. We recommend systematic vaccination for patients younger than 5 years old or native from low endemicity areas (<25% of seropositivity). For the others, we propose serology-based vaccination.

Disclosure of Interest: None declared

PF14

URINARY DIPSTICK: DIAGNOSTIC EFFICIENCY IN INFANTS AGED LESS OF 3 MONTHS OLD WITH UPPER URINARY TRACT INFECTION

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Introduction: Urinary tract infection (UTI) is one of the most common bacterial infection in infant of less than 3 months. Fever without source in infant is a frequent and serious symptom leading to consultation in Pediatric Emergency Department. Studies showed interest by using urine dipstick result combined with other lab test and clinical score to determine the necessity of an antibiotic treatment. Nevertheless, some studies showed a variable reliability of the urinary dipstick depending of child age, especially in infant. We aimed to defined the diagnosis efficiency of the urine dipstick in the infant aged less than 3 months in which a UTI is suspected.

Methods: We performed a retrospective monocentric study including children aged of 0 to 12 months old who underwent a urine culture after presenting with a fever without source between January and

December 2017. We first reported all infants who underwent urine culture analysed in our laboratory (Hôpital de l'Enfance, Lausanne). After what we selected those who presented with fever, and excluded bag urine samples. Then we compared the results of the urine culture to the urine dipstick results. We separated the patients into two groups sorted by age (0–3 months and 3–12 months) in order to analyze sensitivity and specificity of the urinalysis in young infants. Positive urine culture was defined as $\geq 100\,000$ CFUs/ml. Positive urine dipstick was defined as presence of leucocyte esterase and/or nitrites.

Results: 166 patients were eligible. The first group of patients (aged of 0–3 months old) consists of 59 cases and the second group (3–12 months old) consists of 107 cases. Prevalence of positive urine culture was 44% (31–57%IC95) for group 1 and 39% (30–49%IC95) for group 2. For group 1 sensitivity was 54% (35–75%IC95), specificity 100%. For group 2 sensitivity was 88% (78–98%IC95), specificity 97% (93–100%IC95).

Conclusion: Our results show a major difference in sensitivity in both groups, highlighting a strong variation of urine dipstick reliability with age. Therefore, if urine dipstick is positive for leucocyte esterase and/or nitrites in young infant consulting for fever without source, we recommend introducing antibiotic therapy adapted for upper urinary tract infection without delay. However, if urine dipstick is negative, UTI cannot be reasonably excluded and urine culture should be performed. Meanwhile, the decision of treating the patient should be based on reasonable evaluation of the clinical status and labs.

Disclosure of Interest: None declared

PF15

FULMINANT PROGRESSION OF A TOXIC SHOCK SYNDROME IN A PERIPHERAL PAEDIATRIC CLINIC – A CASE REPORT

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Introduction: Toxic Shock Syndrome (TSS) is an acute, toxin-mediated illness that is characterised by fever, hypotension, rash, and multi-organ involvement. In the early stages of the disease, signs and symptoms might resemble other common childhood illnesses rendering the diagnosis and management of TSS difficult, especially in peripheral paediatric clinics.

Methods: N/A

Results: A 9-month-old girl was presented in our emergency department with symptoms of an uncomplicated viral upper airway infection. In an unplanned visit two days later, the patient displayed similar symptoms with a slightly raised respiratory frequency. The chest X-ray was compatible with acute bronchitis. A follow-up visit was scheduled two days later. The following day, the patient returned with an obstructive breathing pattern, tachycardia and a progressive rash. After treatment with bronchodilators, intravenous (IV) antibiotics, low-flow oxygen, and a bolus of IV fluids, the patient stabilised and the rash did not progress further. Two large attacks of diarrhoea led to further administration of IV fluids. The patient deteriorated successively with higher needs of oxygen flow, physical agitation, tachypnoea, and extended recapillarisation time. The chest X-ray



Figure 1

showed a pleuropneumonia with massive effusion of the right thorax and mediastinal shift (fig. 1). Due to shortage of space in the nearby paediatric university clinic, the transport to another university clinic was organised. Rapidly, the patient became haemodynamically unstable with high demands of IV fluids and oxygen. After alarming the in-house resuscitation team, cardiopulmonary resuscitation (CPR) was initiated less than eight hours after the patient's entry to our paediatric clinic. CPR was conducted until the paediatric emergency care team from the university clinic in town arrived. After two unsuccessful thoracenteses, the patient was transported under ongoing CPR to the university clinic and connected to an extracorporeal membrane oxygenation (ECMO). Beta-haemolytic group A streptococcus were later identified in a sample of the pleural effusion.

Conclusion: This case shows that a viral infection can be superinfected with beta-haemolytic group A streptococcus causing TSS. In a hospital serving a predominantly adult population, diagnosing TSS can be impeded by failing to identify red flags or by having delayed access to immediate imaging technology or paediatric intensive care, leading to a fulminant progression of the disease with a nearly fatal outcome.

Disclosure of Interest: None declared

PF16

DOSE-DEPENDENCY OF ECHINACEA IN THE TREATMENT OF ACUTE COMMON COLDS IN CHILDREN 4–12 YEARS

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Introduction: Alternative therapies have a high acceptance in parents for treating minor ailments in their children due to positive safety profiles. Clinical efficacy studies in this vulnerable population are however scarce for natural remedies as for synthetic drugs [1]. In view of frequency and severity of cold symptoms and the associated 30% complication risk, parents as well as physicians urgently seek for clinical efficacy.

Methods: An alcoholic extract of *Echinacea purpurea* was developed into a children-friendly formulation (Echinaforce® Junior tablets, EFJ). 1200 and 2000 mg of *Echinacea* extract were randomly applied in three or five tablets per day to children (4–12 years) with acute cold symptoms. Up to three cold episodes were treated until symptom resolution but no longer than 10 days. Parents recorded respiratory symptoms using a validated scoring method (e-diaries).

Results: A total of 130 cold episodes were treated with EFJ in 79 children during the 5.3 months of observation. The dose increase from 1200 to 2000 mg *Echinacea* extract effectuated a reduction of episode duration from 8.1 ± 3.52 to 6.9 ± 3.48 days (ITT collective, p < 0.05 in Wilcoxon-test, fig. 1). Children allocated to 1200 mg *Echinacea* but with >160% compliance were moved to the 2000 mg group, which further enhanced the treatment effect to 1.7 days (p = 0.020). After 10 days with 2000 mg *Echinacea* 8.7% of episodes remained unresolved in comparison with 23.5% with 1200 mg (p = 0.005). 2000 mg decreased the risk for recurrent infections from 71.9% to 58.1% and the overall cold incidence in susceptible children from 3.6 episodes (as per history) to 1.8 episodes (p < 0.001). Tolerability of EFJ was by 98.5% of physicians and 99.2% of parents rated as "good" or "very good". Thirteen (13) children (19.1%) reported adverse events but none was causally related to EFJ nor serious. Finally, over 80% of parents stated that they would want to take the medication again.
Conclusion: Echinaforce® Junior tablets are a newly developed, safe and efficacious option for the treatment of the common cold in children. Five tablets deliver 2000 mg *Echinacea* extract daily and reduce the duration of cold episodes by 1.2 to 1.7 days. Symptoms are soothed from the first day of treatment and after 10 days 9 of 10 episodes are fully resolved.

[1] Taylor JA, et al. JAMA. 2003;290:2824–2830.

I have read and understand application rules: No

Disclosure of Interest: A. Bächler: None declared, S. Feldhaus: None declared, G. Lang: None declared, P. Klein: None declared, A. Suter Employee of: A Vogel Switzerland, R. Schoop Employee of: A Vogel Switzerland

PF17

LEUKEMOID REACTION IN SUPPURATIVE ADENITIS, A CASE REPORT

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Introduction: Severe infections or reactive conditions can induce leukemoid reaction, which needs to be distinguished from neoplastic processes. Mononuclear cell surface markers together with tissue morphology are needed to rule out the later condition (1). We present a case of leukemoid reaction in a child with submandibular suppurative lymphadenitis.

Methods: 7-year-old girl referred with fever and a right submandibular lymphadenitis refractory to treatment with oral co-amoxicillin 50 mg/kg/d t.i.d. started 4 days before. The child has no particular medical history other than receiving prophylactic tuberculosis treatment 3 years earlier and no animal contact other than a cat. Physical examination shows a huge painful right submandibular adenitis with skin inflammation. Laboratory tests, imaging and histology are performed.

Results: Laboratory tests indicate moderate inflammation (leukocytes 14.6 G/l, total neutrophils 74%, lymphocytes 13%, CRP 19 mg/l, ESR 50 mm/h) without evidence of tumor lysis syndrome. Neck ultrasound confirms the suppurative adenitis with a small collection of pus (<1 ml). Abdominal ultrasound and chest radiograph are normal. Furthermore, serologies (EBV, CMV, Toxoplasma gondii, Bartonella henselae, Yersinia enterocolitica, Francisella tularensis) are negative, as well as MRSA on nasal swab. Intravenous treatment of amoxicillin and clavulanic acid 150 mg/kg/d t.i.d. is installed for 18 days. Slight increase in pus collection allows surgical drainage on day 8 of intravenous therapy with positive culture of methicillin-susceptible Staphylococcus aureus, Acid-fast stain is negative. Histology shows peculiar accumulation of myeloid precursors expressing myeloperoxidase (MPO) without expression of CD34 or terminal deoxynucleotidyl transferase (TdT). CD68 was positive in histiocytes, CD20 and CD3 highlighted normal lymphocytes. The differential diagnosis was either an acute myeloid leukemia or a strong inflammatory reaction. Further analysis of precursor surface markers suggests leukemoid reaction since CD34, CD56 and CD117 were negative on leukemic suspected cells. Course of disease is favorable and further blood samples are normal.

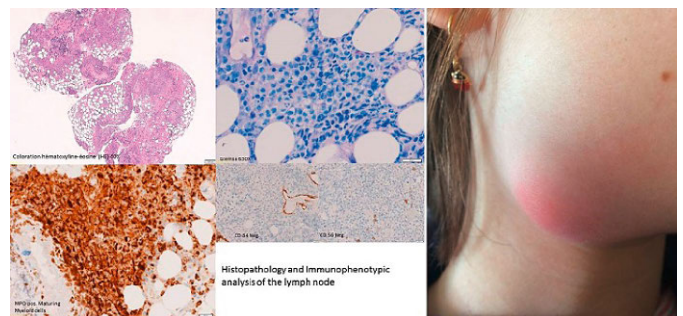


Figure 1

Conclusion: Besides malignancies, the major causes of leukemoid reactions are severe infections, intoxications, severe hemorrhage, or acute hemolysis (2). The diagnostic work-up consists of the exclusion of leukemia/myeloid sarcoma. This situation is illustrated by our case report.

NaChwuchs Prize: I wish to apply

Plaschke Prize: I wish to apply

I have read and understand application rules: Yes

Disclosure of Interest: None declared

PF18

EPIDEMIOLOGY AND RISK FACTORS FOR SERIOUS BACTERIAL INFECTIONS IN CHILDREN AGED 0 TO 36 MONTHS PRESENTING WITH FEVER WITHOUT SOURCE

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Introduction: Children younger than 36 months are at particular risk of serious bacterial infections (SBI), when they present with fever without source. However, the risk of occult bacteremia has been greatly reduced since the introduction of conjugated vaccination against *Haemophilus influenzae* and *Streptococcus pneumoniae*. The aim of this study is to describe the epidemiology and risk factors of SBI in children with fever without source in our setting and to evaluate the performance of our management algorithm.

Methods: We designed a prospective single-center cohort study. Parents of children aged 0 to 36 months presenting with fever without source in our emergency unit were approached for participation in the study. Demographic and clinical characteristics, investigations and management procedures were recorded on a common case-report. Information on clinical evolution, final diagnosis, and immunization history were obtained by calling parents and/or paediatrician 10 days after the inclusion. Potential predictors of SBI were compared between patients with and SBI.

Results: Between October 2015 and September 2017, 173 children were recruited, with a median age of 4.4 months (range 0.2–35.4). One hundred and sixty-six patients (96%) were up to date with their vaccinations and 121 children (70%) had received at least one dose of pneumococcal conjugate vaccine. There were 45 children (26%) with a final diagnosis of SBI which were all urinary tract infections. Presence of chills (OR 3.5, 95% CI 1–12.6), fever for more than 2 days (OR 8, 95% CI 2.3–28.1) and being less than 12 months old (OR 9, 95% CI 2.1–38.8), and recapillary time of >2 seconds (OR 2.2, 95% CI 1–5) were statistically significant predictors of SBI on multivariate logistic regression. Our management algorithm was able to identify all cases of SBI. We measured a sensitivity of 100% (95% CI 92–100%) but a specificity of only 21% (95% CI 15–29%).

Conclusion: We identify a high proportion of urinary tract infections and no single case of occult bacteremia nor other SBI. All cases of SBI were successfully diagnosed with our management algorithm. The identification of independent predictors of SBI could help to adapt our management algorithm in a future attempt to increase its specificity.

Disclosure of Interest: None declared

POSTER FLASH 2

PF19

SEVERE HAEMOCHROMATOSIS IN THREE REFUGEE ADOLESCENTS WITH B-THALASSAEMIA

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Introduction: Beta-Thalassaemia is caused by a point mutation that leads to a decreased production of beta-globin, resulting in defective red blood cells and ineffective erythropoiesis. Complications are microcytic hypochromic anaemia, bone marrow expansion and, due to compensation mechanisms, extramedullary haematopoiesis and increased intestinal iron absorption. Today, complications of Beta-Thalassaemia in developed countries are rare due to early initiation of regular blood transfusions and chelation therapy followed by curative allogeneic stem cell transplantation from a HLA-matched sibling donor.

Methods: We present three clinical cases of refugee adolescents from the middle east with Beta-Thalassaemia who show severe complications due to inadequate therapy during escape from their countries. Details on the management of therapy initiation will be reported as well as first follow-up.

Results: Three refugees, two females and one male at the age of 14, 16 and 17-years presented with severe complications of Beta-Thalassaemia. All had been treated by blood transfusions, two of them with intermittently chelation therapy in their home countries. Clinical examination showed hepatomegaly, growth retardation and absence of puberty and the MRI of the head showed severe hemosiderosis especially in the hypophysis in all of them. One of them has severe ascites and multiple fractures. Due to several fractures of the long bones, one patient has not been able to walk since 1.5 years. Imaging and further diagnostics showed severe iron overload of the liver and heart with cirrhosis of the liver, subsequent liver dysfunction and cardiomyopathy with cardiac dysfunction. Endocrinological sequelae such as primary hypothyroidism was found in one patient and diabetes was found in two patients. We started therapy with regular blood transfusions depending on severity of anaemia. The goal of our treatment is suppression of the ineffective erythropoiesis controlled by low reticulocytes count. Combination chelation therapy was initiated with either deferasirox and deferoxamine in one patient and deferasirox and deferiprone in the other two patients adapted to ferritin values. Patients have a follow-up of two, two and 25 months and show first ameliorations.

Conclusion: Beta-Thalassaemia patients who come as refugees can show extraordinary severe clinical findings that can rarely be observed today. Initiation of treatment needs an interdisciplinary approach with family education as an important point for patient's care.

Disclosure of Interest: None declared

PF20

CASPR2-ANTIBODIES RELATED DISEASE IN A 3-YEAR OLD CHILD

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Introduction: Contactin-associated protein-like 2 (CASPR2) is an essential protein for localization of voltage-gated potassium channels (VGKC). It is expressed in the CNS and peripheral nervous system. Antibodies against CASPR2 are reported in adult patients [1, 2] with neuromyotonia, Morvan syndrome, limbic encephalitis. The clinical spectrum is diverse and yet not well defined. An association with autonomic dysfunction and Hashimoto thyroiditis is also been described. To our knowledge there is still no report about the disease in a young children.

Methods: We describe a 3.5 year old girl that presented with a continuous worsening of general condition, mood changes, apathy and insomnia. She showed a significant arterial hypertension and tachycardia. A primary hyperthyroidism with normal thyroid stimulating hormone (TSH) and a primary Cushing syndrome were found. The etiology remained unclear, probably due to a peripheral hormone resistance. A paraneoplastic cause was ruled out. The hypertension led to the introduction of 3 antihypertensive drugs with almost no effect. The hyperthyroidism was treated with neomercazole. During the hospitalisation progressive dysaesthesia, areflexia and abasia were seen. A demyelinating polyneuropathy was electrophysiologically confirmed. Due to the progressive disease with neurological symptoms and unclear autonomic and endocrinological changes, several autoimmune antibodies were determined. Antibodies against VGKC and CASPR2 were found high positive in blood and cerebrospinal fluid (CSF). MRI scan of the brain showed a progressive supratentorial brain atrophy without changes in the limbic region. We assumed a CASPR2-antibodies related disease and started a high dose corticosteroid therapy and a course of immunoglobulins. This led to a fast and significant improvement of the neurological abnormalities and of the arterial hypertension. Whether the hyperthyroidism got better under neomercazole or steroids is still unclear.

Conclusion: To our knowledge, our patient is the first published case of a CASPR2-antibody related disease in a young child. Symptoms involving the CNS and peripheral nervous system and additional clinical features like insomnia and autonomic dysfunction should raise suspicion of CASPR2 antibodies also in children. Because, as shown in our case, CASPR2-antibodies related disease can be treated, early recognition is important.

Disclosure of Interest: None declared

PF21

CEREBRAL PALSY: ASSESSMENT AND TREATMENT OF THE UPPER EXTREMITY

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Introduction: Cerebral Palsy (CP) describes a heterogeneous group of permanent movement disorders that occur in infancy or childhood. Only currently, more attention is paid to the function of upper extremity. Some patients may have less than optimal function due to delayed or inadequate treatment.

Methods: Patients who were assessed at the upper extremity CP clinic (UEPC) between 2007 and 2017 were reviewed retrospectively.

Results: 95 patients were assessed in the UEPC with only few who had just one visit because no improvement of the therapeutical setting could be offered. The others were formally assessed with the AHA test (Assisting Hand Assessment) by hand therapists and therapeutic goals were defined. Conservative treatment with hand therapy, splinting, constraint movement therapy and aids was adapted accordingly. Botox injection, followed by an adapted hand therapy and a second AHA test, were performed in those patients who potentially qualified for surgery. Only 17 hands in 16 patients (16.8%) were operated at a mean age of 15.2 (7.6–19.6) years. Their Manual Ability Classification Scale (MACS) was II in 5, III in 5, IV in 2 and V in 2 patients. Most had multiple procedures including 3 arthrodesis, 4 sesamoidesis, 20 tendon transfers, 49 tendon releases, 6 muscle slides and 21 surgical muscular lengthening procedures. No reoperations were performed. An improvement of function and appearance was found in all patients with a postoperative assessment (8/16).

Conclusion: Due to a very restrictive patient selection, less than 20% of the 95 patients who were assessed were operated upon. All patients who were assessed postoperatively had an improved appearance and function. Botox injections helped to unmask antagonistic muscles and to optimize exercise. However, the effect is not long lasting and it's most important function is to help adapting surgical strategies. Confirmation and sometimes slight modifications of ongoing therapies as well as reassurance of the families and their therapists are a further essential task of the UEPC team. The complexity of movement disorders of the upper extremity in children with CP demands an interdisciplinary approach including pediatric hand therapists, neurologists, rehabilitation specialists and hand surgeons. Longitudinal observations are necessary because severity and characteristics of CP change over time. The objective of the assessment is to define relevant goals that may improve activity and participation.

Disclosure of Interest: None declared

PF22

DANCING WITH MYSELF

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Introduction: A 9 y.o. healthy Syrian refugee girl came to the ER for abnormal movements of the left arm and leg. Symptoms appeared spontaneously and worsened until she couldn't get dressed and scarcely spoke. There was no history of trauma, febrile illness or other neurological issue except for mood swings for a month. She had sore throat a month ago and had several not treated anginas in Syria in the past. There was no familial history of inherited neurological or autoimmune disease nor consanguinity. Examination showed chorea of the left arm and leg (a video was made), with cerebellar tests hindered by chorea and stepping while walking. There was no heart murmur and no sign of arthritis. Laboratory test showed increased ESR (43 mm/h), ASLO (2820 kU/l) and Streptodornase B (313 kU/l). ANA, thyroid tests and throat culture were negative. Wilson's disease was excluded. Cerebral MRI was normal. Clinical and biological findings were compatible with Sydenham Chorea (SC). Echocardiography showed moderate mitral insufficiency and acute rheumatic fever (ARF) was diagnosed. Treatment of penicillin V was prescribed for ten days, followed by benzathine penicilline once per month until 18 yo. Symptoms were treated by carbamazepine with dramatic improvement in a week and near complete recovery at 6 months.

Results: Discussion: Differential diagnosis for chorea ranges from metabolic disorders to genetic, post-infectious, autoimmune or tumoral origins. SC is the main cause of chorea in children and is more often found in female. Symptoms are generalized but hemi-chorea occurs in 25% of patient. Physiopathology is explained by antibody mimicry after non-treated infection by some strains of Streptococcus, attacking the caudate nucleus. It's one of the major Jones' Criteria, making it mandatory to search for other manifestations of ARF, such as carditis and arthralgia. It is the latest appearing and a rare sign of ARF. The disease is self-limited, resolving in 1–6 months. Treatment is based on penicillin (acute and chronic treatment) to reduce risk of recurrence and cardiac complications. Neurological symptoms are treated by anticonvulsants (valproate, carbamazepine) or neuroleptics (haloperidol). Steroids or IVIG can be used as second-line treatment. **Conclusion:** SC should always be considered when faced with chorea. ARF is going to be more frequent in our region due to migratory flows. Rapid diagnosis and adequate treatment help prevent recurrences and multiple valve surgeries.

Disclosure of Interest: None declared

PF23

ATRIOVENTRICULAR BLOCK DURING MIGRAINE ATTACK – A CASE REPORT

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Introduction: Migraine can be accompanied by dysregulation of the autonomic nervous system (ANS). Symptoms such as nausea, vomiting, diarrhea, cutaneous vasoconstriction or diaphoresis can be seen as well as cardiac arrhythmias.

Methods: In our emergency department a 13 5/12 years old boy sought medical advice because of a headache of the right hemisphere and nausea. This pain preceded a 30 minutes episode of visual change (curtain like effect over the right eye and shimmering). Episodes like this had occurred 3–4 times before. He also described a chest pain that aggravated by inspiration. Initially it was located on the right side and afterwards on the left. The family history was positive for migraine. The clinical examination and the cardiorespiratory parameters were completely normal (heart rate 93/min, respiratory frequency 20/min, blood pressure 116/53 mm Hg, oxygen saturation 100%). The electrocardiogram (ECG) showed a first degree atrioventricular (AV) block with a PQ time of 270 ms. In the control examination the following day the boy was pain free and the AV block was not measurable in the ECG anymore (PQ 170 ms).

Results: –



Figure 1

Conclusion: This case report shows the involvement of the ANS during migraine attacks. Cardiac arrhythmias are rarely documented. In literature, however, electrocardiographic changes like prolongation of QTc and PR interval, T inversion or ST segment abnormalities have been described.

NaChwuchs Prize: I wish to apply
Disclosure of Interest: None declared

PF24

BLISTERING ERUPTIONS IN CHILDHOOD HENOCCH-SCHÖNLEIN SYNDROME: SYSTEMATIC REVIEW OF THE LITERATURE

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Introduction: The occurrence of blistering eruptions in childhood Henoch-Schönlein syndrome has hitherto been addressed exclusively in individual case reports. We aimed at describing epidemiology, clinical presentation, and therapeutic options in Henoch-Schönlein patients ≤18 years of age with blistering eruptions.

Methods: We performed a systematic review of the literature in the National Library of Medicine and Excerpta Medica databases. The review was performed according to the "Preferred reporting items for systematic reviews and meta-analyses" statement.

Results: Ten (1.5%) children with blisters were detected in 7 case series presenting a total of 666 pediatric Henoch-Schönlein cases. We also identified 41 individually documented cases of Henoch-Schönlein syndrome with blistering eruptions. Blistering eruptions and purpura were similarly distributed and blisters developed concomitantly with palpable purpura or with a latency of ≤ 14 days. Most (80%) of the cases remitted within 4 weeks. The clinical course was similar in children managed expectantly and in those managed with steroids. Colchicine positively influenced the disease course in two severe and long-lasting cases.

Conclusion: Blistering eruptions are rare in Henoch-Schönlein syndrome. They can be a source of diagnostic dilemma but do not have any prognostic value since they almost always spontaneously subside within 4 weeks.

Publication

Ramelli V, et al. Blistering eruptions in childhood Henoch-Schönlein syndrome: systematic review of the literature. *Eur J Pediatr.* 2017;176:487–492.

Disclosure of Interest: None declared

PF25

ATRIAL FLUTTER IN A HEALTHY FULL-TERM NEWBORN

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Introduction: Atrial flutter (AF) is a supraventricular tachycardia (SVT) caused by an intra-atrial reentrant circuit without intervention of the atrioventricular (AV) node. Although it is the second most common cause of SVT in healthy foetuses and newborns, AF is an uncommon primary arrhythmia in children and the prevalence is low.

Methods: The patient was a male newborn, born at 40+5/7 weeks of gestation with vacuum due to an abnormal CTG and non-recordable heart rate interpreted as either extreme bradycardia or tachycardia. Adaptation to extrauterine life was good. At 6 hours of life a routine physical examination showed good general condition, postductal oxygen saturation levels above 96%, blood pressure 74/42 mm Hg, respiratory rate 60/min. The heart rate was 210 bpm while the patient was sleeping. The cardiopulmonary, neurological and abdominal status were normal. The pregnancy was uneventful with normal routine CTG registrations and ultrasounds without need for any treatments. Early onset sepsis and electrolyte imbalance were excluded. The ECG showed a narrow QRS-complex tachycardia of 206 bpm with ... sawtooth waves, without isoelectric line and with an AV conduction of 2:1. The echocardiography was normal. A vasovagal manoeuvre confirmed an AF with a 2:1-3:1 AV block. A unique electrical cardioversion with 1J/kg under anaesthesia was successfully performed and sinus rhythm was restored. A prophylactic therapy with beta blockers was introduced for a month. A follow up was normal.

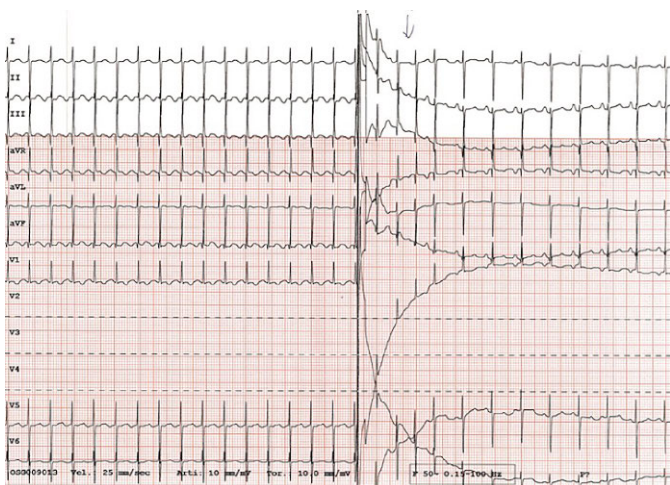


Figure 1

Results: Discussion: Differential diagnoses in a patient with tachycardia are very important to choose the correct treatment. The AF can be mistaken for SVT, which is more common than reentrant AV tachycardia. Different methods can be used to reveal the underlying FA. The simple vasovagal manoeuvre can stop the reentrant AV tachycardia or can reveal the AF, like it did in our case. AF has a low

incidence in paediatrics and most of the cases occur in premature newborns or after invasive procedures like the positioning of central and PIC-lines. Nevertheless it is important suspect it in healthy, full-term newborns. In our patient the cause was primary.

Conclusion: AF is a rare cause of SVT but is more prevalent in fetal and neonatal life. It is the only SVT with electrical cardioversion as first indication. Generally, it has an excellent long-term prognosis.

Disclosure of Interest: None declared

PF26

CYSTIC FIBROSIS IN A PATIENT WITH NEGATIVE NEWBORN-SCREENING

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Introduction: We report the case of a 6-month old female patient, born at term with a birth weight on the 50th percentile. Pregnancy, birth and the neonatal period were uneventful. She was born in Switzerland and the routine newborn-screening (NBS) showed no pathological findings. At the age of 5 months the patient started presenting abdominal discomfort with vomiting. Later on, she refused to eat and drink. A severe weight loss was determined and at 6 months of age she was admitted to our hospital for further assessment. During the hospitalization, the interaction between the parents and the patient was described as problematic, leading to initially interpret the disturbed interaction as the cause of her refusal to eat. After release, the patient lost more weight and developed a cough. A few weeks after discharge she was admitted again because of sudden paleness and hyporeactivity. In the emergency department, severe dyselektrolytemia and metabolic alkalosis were found. The patient had to be resuscitated and intubated. A broad spectrum of investigations was conducted and as the results came all back negative, a sweat test was performed. The pathological sweat test and a diminished pancreatic elastase in the stool confirmed the diagnosis of cystic fibrosis (CF). The salt loss was classified as Pseudo Bartter's Syndrome. A therapy with substitution of pancreatic enzymes, salt and fat-soluble vitamins was started, and she was put on a secretolytic inhalation and physiotherapy. The girl recovered within a few days of treatment and returned to her 50th weight percentile. 6 months later she presents clinically insuspicious without any respiratory or intestinal symptoms, and normal growth.

Methods: –

Results: CF is part of the NBS in Switzerland since 2011 and consists of an IRT-IRT-DNA strategy. In case of a positive result, a confirmatory sweat test is performed. In our patient, the initial IRT-value (37 ng/ml) was far below the cut off value (50 ng/ml) leading to a false negative NBS and misguiding further diagnostic approach. Current data from the NBS data base show a rate of false negative results for classical CF in 4% of all cases.

Conclusion: A negative NBS can delay diagnosis of CF, leading to severe complications. Thus, it is important to remain alert when symptoms compatible with cystic fibrosis are presented in a child of any age, including failure to thrive, intestinal and/or respiratory symptoms, or a salt loss of unclear reason.

Disclosure of Interest: None declared

PF27

RESPIRATORY RATE IN INFANTS WITH CYSTIC FIBROSIS THROUGHOUT THE FIRST YEAR OF LIFE AND ASSOCIATION WITH LUNG CLEARANCE INDEX MEASURED SHORTLY AFTER BIRTH

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Introduction: Lung impairment in cystic fibrosis (CF) starts in infancy. However, tools to monitor lung disease are limited in early life. Respiratory rate (RR) as a key vital sign is easy to assess during sleep and is elevated during acute respiratory disease. Thus, elevated RR could indicate early lung impairment and potentially serve as a diagnostic tool in disease monitoring. The aim of the study was to assess the natural course of respiratory rate during the first year of life in infants with CF compared to healthy controls.

Methods: In a prospective cohort of infants with CF and healthy controls respiratory rate was measured and respiratory symptoms reported weekly throughout the first year of life by parents. Infants performed a lung function measurement within the first three months of life. We used multilevel linear regression to compare respiratory rate between infants with CF and healthy infants, adjusting for respiratory symptoms and other potential determinants.

Results: The analyses included 5656 measurements from 153 infants (43 with CF). RR declined from 43.2 (40.5)/min at 6 weeks of age to 28.3 (24.6)/min at 50 weeks in infants with CF (healthy controls). Infants with CF had consistently higher RR than controls (difference: 4.15/min; 95% CI (2.86, 5.44); $p < 0.001$). In both study groups, RR was increased in infants with higher Lung Clearance Indices (LCI) and during episodes of respiratory infections throughout the study period.

Conclusion: Infants with CF have a higher RR compared to healthy controls during the first year of life. Persistent association with early LCI measurements may indicate tracking of poorer lung function throughout the first year of life by RR. It might thus be an early subtle sign of functional respiratory deficit and could be a sensitive and promising marker to monitor early CF lung disease.

Disclosure of Interest: I. Korten: None declared, E. Kieninger: None declared, S. Yammine: None declared, G. Cangiano: None declared, S. Nylas: None declared, P. Anagnostopoulou: None declared, F. Singer: None declared, C. Kuehni: None declared, N. Regamey: None declared, U. Frey Research grant from: the Swiss National Science foundation, C. Casaulta: None declared, B. Spycher Research grant from: the Swiss National Science Foundation, P. Latzin Conflict with: personal fees from Vertex, Novartis, Roche, Polyphor, Vifor, Gilead, all outside the submitted work.

PF28

PREVALENCE AND CHARACTERISTICS OF NONBLANCHING, PALPABLE SKIN LESIONS WITH A LINEAR PATTERN IN CHILDREN WITH HENOC-SCHÖNLEIN SYNDROME

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Introduction: Linear nonblanching skin lesions are thought to occur very rarely in patients with Henoch-Schönlein syndrome. The aim of this study was to investigate the prevalence and characteristics of linear nonblanching skin lesions in children with Henoch-Schönlein syndrome.

Methods: A prospective case series was conducted at the outpatient clinic of a hospitalist between January 1, 2010, and December 31, 2015, among 31 consecutive children with Henoch-Schönlein syndrome. Children with Henoch-Schönlein syndrome underwent a careful, structured skin examination, with emphasis on the presence of palpable lesions with a linear pattern.

Results: Thirty-one consecutive children affected with Henoch-Schönlein syndrome were included. Among the 31 children in the study (12 girls and 19 boys; median age 6.2 years [range, 3.0–12.0 years]), 8 (26%) had linear lesions on the legs, groin, waistline, wrists, or forearms. Patients with or without linear lesions did not differ significantly with respect to sex, age, and cutaneous, abdominal, articular, or renal involvement.

Conclusion: This study illustrates the prevalence and characteristics of linear skin lesions in patients with Henoch-Schönlein syndrome. Patients with symptoms suggestive of this vasculitis should be evaluated for the presence of nonblanching, palpable lesions with a linear pattern.

Publication

Milani GP, Lava SA, Ramelli V, Bianchetti MG. Prevalence and characteristics of nonblanching, palpable skin lesions with a linear pattern in children with Henoch-Schönlein Syndrome. *JAMA Dermatol.* 2017; 153: 1170-1173.

Disclosure of Interest: None declared

PF29

IDIOPATHIC SYSTEMIC CAPILLARY LEAK SYNDROME IN CHILDHOOD: SYSTEMATIC REVIEW OF THE LITERATURE

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Introduction: Idiopathic systemic capillary leak syndrome (or Clarkson disease) is characterized by episodes of acute increase in vascular permeability resulting in loss of fluid into the interstitium. The typical patient is a previously healthy subject presenting with acute onset of edema associated with the triad of hypotension, hemoconcentration and hypoalbuminemia. The disease usually occurs in subjects with a monoclonal gammopathy.

Methods: Between May and December 2017, we performed a search with no date limits of the Mesh terms "idiopathic capillary leak" OR "Clarkson disease" OR "Clarkson syndrome" OR "primary capillary leak" OR "hyperpermeability capillary syndrome" in the US National Library of Medicine and Excerpta Medica databases.

Results: We identified 20 reports providing data on 28 otherwise healthy children, who experienced a total of at least 65 episodes of acute capillary leak, 59 of which were individually described. Forty-one (69%) attacks were preceded by virus-like symptoms and signs. The condition initially presented in newborn age ($n = 2$, 7%), infancy ($n = 5$, 18%), toddler age ($n = 4$, 14%), preschool age ($n = 6$, 22%) or school age ($n = 11$, 39%) and more frequently affected girls (71%) than boys. They suffered from a mean of 2 (range 1–6) attacks. Hemodynamic shock ($n = 35$, 59%), rhabdomyolysis ($n = 14$, 24%) with ($n = 7$, 7%) or without ($n = 7$, 7%) compartment syndrome, acute kidney injury ($n = 10$, 17%), pulmonary edema ($n = 9$, 15%) and either pleural ($n = 4$) or pericardial ($n = 3$) effusion ($n = 7$, 12%) were, in decreasing order of frequency, the most common complications. A monoclonal protein in blood was never detected. The acute drug management involved intravenous hydration ($n = 32$), catecholamines and vasopressors ($n = 18$), antimicrobials ($n = 14$), albumin ($n = 12$), corticosteroids ($n = 11$), immunoglobulins ($n = 9$), antivirals ($n = 4$), diuretics ($n = 4$), aminophylline ($n = 3$), bicarbonate ($n = 2$) and red blood cells or platelet transfusions ($n = 2$).

Conclusion: Idiopathic systemic capillary leak syndrome exists not only in adulthood but also in childhood. Management is mostly supportive. The supposed pathogenetic role of a monoclonal gammopathy is not confirmed in children.

Disclosure of Interest: None declared

PF30

HYPERAMMONEMIA ASSOCIATED WITH DISTAL RENAL TUBULAR ACIDOSIS OR URINARY TRACT INFECTION: SYSTEMATIC REVIEW

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Introduction: Hyperammonemia usually results from an inborn error of metabolism or from an advanced liver disease. Individual case reports suggest that both a distal renal tubular acidosis and a urinary tract infection may also result in hyperammonemia.

Methods: Stimulated by our experience, we performed a review of the literature on hyperammonemia ($>50 \mu\text{mol/L}$) secondary to the mentioned renal tract diseases in the National Library of Medicine and Excerpta Medica databases. We identified 39 scientific reports published between 1980 and 2017.

Results: Hyperammonemia was detected in 13 children with distal renal tubular acidosis and in an adult patient with distal renal tubular acidosis. In these patients, a negative relationship was observed between circulating ammonia and bicarbonate ($P < 0.02$). Alkali therapy corrected both acidosis and hyperammonemia. In 31 patients (19 children and 12 adults) with an anatomically abnormal urinary tract, an acute urinary tract infection was complicated by hyperammonemia and symptoms and signs of acute neuronal dysfunction such as an altered level of consciousness, convulsions and asterixis, often associated with signs of brain edema such as anorexia and vomiting. Urea-splitting germs were isolated in 28 out of the 31 cases (most frequently *Proteus* species and *Corynebacterium* species).

Conclusion: This study points out that both an altered distal renal tubular acidification and a urinary tract infection may be associated with relevant hyperammonemia both in children and in adults.

Publication

Clericetti et al. Hyperammonemia associated with distal renal tubular acidosis or urinary tract infection: a systematic review. *Pediatr Nephrol* doi 10.1007/s00467-017-3829-7

Disclosure of Interest: None declared

PF31

D-LACTIC ACIDOSIS IN SHORT-BOWEL SYNDROME: SYSTEMATIC LITERATURE REVIEWD. G. Bianchetti¹, M. G. Bianchetti¹, G. D. Simonetti¹, S. A. A. G. Lava²*¹Pediatrics, Pediatric Department of Southern Switzerland, Bellinzona; ²Pediatrics, Universitätsklinik für Kinderheilkunde, Bern, Switzerland

Introduction: Since human cells only contain L-lactate dehydrogenase, they almost exclusively synthesize L-lactic acid. By contrast, some colonic carbohydrate-fermenting bacteria form D-lactic acid (among other organic acids). D-lactic acidosis is an uncommon and challenging form of metabolic acidosis that may develop in short bowel syndrome. It has been documented exclusively in case reports and small case series.

Methods: Stimulated by our experience, we performed a review of the literature in the National Library of Medicine and Excerpta Medica databases and identified 84 original reports published between 1977 and 2017.

Results: D-lactic acidosis was observed in 98 individuals ranging in age from 7 months to 86 years (45 children and 51 adults) with short bowel syndrome. The clinical presentation included Kussmaul breathing, confusion, slurred speech and gait disturbances. Furthermore, among 99 consecutive patients with short bowel syndrome, 21 reported having episodes with symptoms consistent with D-lactic acidosis.

Conclusion: In humans with short bowel syndrome, D-lactic acidosis is likely rather common and under-recognized complication. The identification of D-lactic acidosis is challenging because the usual laboratory test detects L-lactic acid but not D-lactic acid. This diagnosis should be included in the differential diagnosis of unexplained high-gap metabolic acidosis where the anion causing the acidosis is not known.

PublicationBianchetti DGAM et al. D-lactic acidosis in humans: systematic literature review. *Pediatr Nephrol*. doi 10.1007/s00467-017-3844-8**Disclosure of Interest:** None declared

PF32

PRIMARY HYPEROXALURIA (PH): TYPE 1, 2, 3 OR NONE?A. Ménétrey¹*, T. Neuhaus¹, S. Nikolai¹, B. Röthlisberger¹
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Introduction: Children with urolithiasis and/or nephrocalcinosis need careful metabolic investigation. In particular, hyperoxaluria has to be kept in mind.

Methods: *Case Report:* – Case 1: 7 month-old girl with first febrile urinary tract infection. Ultrasound (US) showed medullary nephrocalcinosis. Urinary analysis revealed increased excretion of oxalate and glycerate. Genetic analysis showed a heterozygous mutation in the AGXT gene, consistent with PH type 1. – Case 2a: 3 year-old boy with first episode of nephrolithiasis (stone analysis: 100% Calcium-oxalate-monohydrate). US was normal, but urinary analysis revealed increased excretion of oxalate and glycerate. Genetic analysis showed a homozygous mutation in the GRHPR gene, confirming PH type 2. – Case 2b: His identical twin brother is asymptomatic. – Case 3: 21 month-old boy with bladder stone (analysis: Calcium-oxalate-mono/dihydrate); one year later further episode of nephrolithiasis. US was normal, but urinary analysis revealed persistent hyperoxaluria and minimally elevated glycerate. Genetic analysis showed a homozygous mutation in the HOGA1 gene, confirming PH type 3. – Case 4: 3 year-old boy with acute right flank pain and microhaematuria, suggesting nephrolithiasis. US was normal, but urinary analysis showed persistent hyperoxaluria. Genetic analysis for PH type 1/2/3 did not show any mutation. All patients have normal renal function and avoid dietary oxalate excess. Cases 1 to 3 are on long-term oral citrate

Conclusion: Children with urolithiasis and/or nephrocalcinosis need careful investigation including urinary oxalate. If hyperoxaluria persists, genetic analysis is required to confirm primary hyperoxaluria. There are at least 3 types of PH with great genotypic/phenotypic variability: Whereas PH type 1 often leads to end-stage renal failure with subsequent combined kidney-liver transplantation, PH type 2 and 3 have a more favourable course. Life-long medication with oral citrate to inhibit calcium-oxalate crystallization may both prevent urolithiasis and maintain renal function.

Disclosure of Interest: None declared

PF33

ACUTE HEART FAILURE IN A 10-YEAR-OLD-CHILD: A RARE CASE OF LEFT VENTRICULAR NONCOMPACTIONJ. Lourenco¹*, D. Schaller¹, M. Beghetti², N. Farhat²
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Introduction: Isolated left ventricular noncompaction (LVNC) is a rare disease with a large array of clinical presentation. It can be found incidentally in asymptomatic patients, or with acute heart failure in patients without history or clinical findings suggestive of heart disease. It accounts for 5 to 9% of newly diagnosed cardiomyopathy.

Methods: A healthy, physically active 10-year-old girl presented with persistent epigastric pain and vomiting for the last 9 days, without fever or stool change. Personal history was positive for laparoscopic appendectomy 2 years ago. Initial assessment only showed dehydration and mild tachycardia, with painful abdomen. Patient was admitted for observation with suspicion of abdominal surgical pathology. Adhesive ileus, diabetes, liver disease and pancreatitis were excluded with radiological and biological exams. Within 48h of hospitalisation, she gained 3kg and developed oliguria. Examination showed narrow pulse pressure, pericardial fremitus and diffuse edemas. Pleural effusions and ascites were suspected and heart failure signs were searched. Chest-ray showed cardiomegaly. Echocardiogram revealed severe heart failure with a shortening fraction of 10%, a severely dilated left ventricle and an image compatible with noncompaction of left ventricle. Diagnosis was confirmed by cardiac MRI. Heart failure was treated with diuretics and inotropic agents and patient was stabilized. Genetic testing is under evaluation.

Results: LVNC is a rare form of cardiomyopathy. It is characterized by prominent trabeculae arising in the ventricular cavity and deep intertrabecular recesses within the myocardium inner wall, creating a spongy aspect of the inner myocardium. It seems to be due to an abnormal embryologic development, with an arrest of the heart compaction process. Diagnosis is usually made with typical image on echocardiography and confirmed by MRI. Several genes have been associated with this form of cardiomyopathy. Symptoms and complications vary greatly. It can be found in asymptomatic patients, but initial presentation can be dyspnea, thromboembolism, arrhythmias and heart failure. Life-threatening arrhythmias should be checked.

Conclusion: Acute abdominal pain may be due to low cardiac output syndrome and heart failure should be part of differential diagnosis.

Disclosure of Interest: None declared

PF34

AGED TENASCIN-C KNOCKOUT MICE HAVE INCREASED LUNG VOLUME AND SEPTAL SURFACE BUT UNCHANGED LUNG FUNCTIONS. Gremlich¹*, T. Cremona², J. Schittny², M. Roth-Kleiner¹
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Introduction: Tenascin-C (TNC) is an extracellular matrix protein belonging to matricellular proteins having more regulatory than structural functions. TNC is highly expressed during organogenesis, but its expression is turned off in adulthood. Only after injury, its expression is upregulated and stimulates tissue repair. Previous studies in our laboratory have shown that TNC inactivation induced an increase in static compliance coupled with a decrease in α -smooth muscle actin (SMA) expression around small airways in newborn and adult lungs. The aim of this study was to evaluate the changes occurring in aged lungs.

Methods: Female and male WT (SV129) and TNC KO mice aged 1–1.5 years (Forsberg, PNAS 1996) were intubated (oro-tracheally) and ventilated for one hour with a rodent ventilator (FlexiVent), using a high tidal volume ventilation (25 ml/kg). Lung function parameters were collected. Lung tissue was collected for molecular biology, histology/immunohistochemistry and flow cytometry analysis.

Results: Static compliance, total inspiratory capacity, K-factor and hysteresis of aged TNC KO lungs were the same as WT ones, in basal conditions and during high tidal volume ventilation, whether in males or in females. Morphological aspect of the lungs was assessed for both sexes and both genotypes, but no difference was noticed. Interestingly, morphometrical analyses showed that TNC KO lungs had higher lung volume and increased parenchymal volume and septal surface, compared to WT lungs. Lung senescence, evaluated using beta-galactosidase activity, was visible in bronchial epithelium and in some cells throughout parenchyma. Beta-galactosidase activity was lower in TNC KO compared to WT lungs. Finally, flow cytometry experiments revealed that TNC KO mice showed lower frequencies

of myofibroblasts while higher frequencies of smooth muscle cells compared to WT mice, in the less proliferative SMA+ cell population.

Conclusion: Contrary to newborn and adult mice, aged TNC KO mice had identical lung function parameters with WT mice. However, aged TNC KO lungs showed morphometric and cytological differences indicating that the absence of TNC during aging probably induces an imbalanced ratio between proliferation and apoptosis/necrosis/autophagy in lung parenchyma, potentially resulting from modifications in cell death pathways. Additional experiments will bring more details on the potential beneficial effect of TNC inactivation during the aging process in lung.

Disclosure of Interest: None declared

PF35

FEASIBILITY AND RELIABILITY OF CAROTID INTIMA-MEDIA THICKNESS MEASUREMENTS IN NONSEDATED INFANTS

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Introduction: Carotid intima-media thickness (CIMT) is a surrogate marker for atherosclerosis. It is increased in adolescents and young adults at risk for future cardiovascular disease. However, it remains unclear if it can be considered as a surrogate marker for atherosclerosis in infancy as very few studies have been performed in infants.

Objectives: Our objective was to assess the feasibility and interobserver reproducibility of CIMT measurement in nonsedated infants.

Methods: We measured CIMT in 81 infants less than 1 year of age. Repeated measurements were obtained by a second observer in 24 children. The analysis was performed with semiautomated edge detection software. Measurements with over 95% edge detection over a length of 1 cm were considered as valid. We further compared the measurements using the semiautomated method with measurements using the manual electronic caliper method in a subgroup of 10 infants.

Results: Carotid ultrasound recordings and intima-media thickness measurements were obtained in 79% of infants (n = 64). Mean CIMT of the 64 infants measured by the first observer was 0.44 mm (SD: 0.04). In the 24 participants with measurements by two observers, the mean interobserver difference was 0.001 mm (SD: 0.026). The interobserver coefficient of variation was 5.9%. CIMT measurements obtained with the manual method (mean: 0.35; range: 0.29–0.39) were slightly lower than measurements obtained

with the semiautomated method (mean: 0.38; range: 0.32–0.44). Measurements with both methods were highly correlated (r: 0.87). **Conclusion:** Measurement of CIMT in nonsedated infants less than 1 year of age is feasible in the majority of infants with good interobserver variability.

Disclosure of Interest: None declared

PF36

ANTIBODY-MEDIATED RENAL ALLOGRAFT REJECTION AFTER THE TRANSITION PERIOD FROM PEDIATRIC TO ADULT CARE

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Introduction: Non-adherence to immunosuppressive treatment after kidney transplantation can result in the production of *de novo* donor-specific HLA antibodies (DSA) and allograft loss. Moreover, because non-compliance or poor adherence to therapy is a well-known concern in adolescents and young adults, the gradual transition of care from the pediatric to the adult transplant clinic is critical in this setting.

Methods: We have compared in a retrospective manner, the evolution of two groups of ten adolescents with kidney transplants, who received a cross-match negative renal graft, with conventional immunosuppression and who were transferred from pediatric to adult clinic care during the period 2007 to 2017. In the first group (n = 4), the patients were transferred to the adult transplant clinic between 2007–2011, without any structured transitional period. In the second group (n = 6), patients were transferred to adult care between 2012 and 2017, in the context of a structured multidisciplinary “transition care clinic”.

Results: From 2007 to 2017, a total of 21 children received a kidney transplant. Ten of them (47%) were transferred from pediatric to adult clinic care. The four adolescents in the first group, transferred without “any structured transitional period”, displayed poor adherence to immunosuppressive therapy, and they developed *de novo* DSA. Patient survival after transition in this group was 100%, however all of these patients lost their grafts due to chronic antibody-mediated rejection. The six adolescents transferred in the context of a formal “transition care clinic”, with a structured and progressive transition program, had a good outcome, without transplant loss or *de novo* DSA, with a median follow-up of 22 months. Patients and graft survival were 100% in this second group.

Conclusion: The implementation of a structured multidisciplinary transition kidney transplant clinic improved adherence and appears to be a key factor to improve outcomes in that setting.

Disclosure of Interest: None declared

POSTER FLASH 3

PF37

IMPACT OF DIAGNOSTIC DELAY IN PEDIATRIC CROHN'S DISEASE PATIENTS COMPARED TO ADULT PATIENTS

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Introduction: The length of diagnostic delay in Crohn's disease (CD) is a risk factor for bowel strictures and intestinal surgery. We aimed to assess if diagnostic delay has a similar impact in pediatric CD patients when compared to adult patients.

Methods: Retrospective review of data from the Swiss IBD cohort study (SIBDCS). Pediatric patients were diagnosed with CD <18 years of age whereas adult patients were diagnosed with CD >18 years of age. Frequency of CD-related complications were assessed in the long-term follow-up in both groups. Long diagnostic delay was defined as delay >75th percentile.

Results: From 2006 to 2016, a total of 1,550 patients were included (387 pediatric and 1,163 adult CD patients). Median (IQR) diagnostic

delay was 3 (1–9) months for the pediatric and 6 (1–24) for the adult group, respectively. At diagnosis, children presented less complications than adults [any complications: stenosis, perianal fistula, internal fistula, resection surgery, fistula surgery; 14.8% vs. 26.5% for long diagnostic delay (p <0.01) and 11.3% vs. 14.6% for short diagnostic delay (p >0.05)]. However, no significant difference could be observed after 15 years of disease evolution regarding stenosis, perianal fistulas and internal fistulas. Adults with long diagnostic delay more frequently underwent fistula and abscess surgery during the first 5 years following diagnosis when compared to pediatric patients.

Conclusion: Pediatric CD patients are characterized by shorter diagnostic delay and less complications during the early years after diagnosis when compared to adult CD patients. However, at 15 years of disease evolution the difference with respect to complications does no longer exist when comparing pediatric CD patient to adult CD patients (irrespective of the length of diagnostic delay).

Disclosure of Interest: None declared

PF38

A MULTIDISCIPLINARY APPROACH OF CHRONIC OSTEOARTICULAR PAIN IN CHILDREN AND ADOLESCENTS: THE LAUSANNE EXPERIENCE

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Introduction: Chronic pain disorder is a common and under-recognized problem who is increasing in the pediatric population. Osteoarticular pain is one of the most common symptoms. This is a significant problem leading to a decrease in quality of life, school absenteeism and social withdrawal. A multidisciplinary approach is essential to evaluate and manage those patients who have an unsatisfactory evolution despite primary medical care. Starting in 2014, a group of 4 medical specialists (pediatric rheumatologist, pediatric orthopedist, child psychiatrist and pediatrician specialist in adolescent) and one physiotherapist started a joint outpatient clinic assessing such children. The aim of this study was to describe and outline some characteristics of all patients seen at this platform up to now.

Methods: A retrospective descriptive study was performed based on medical records of patients seen at our center in Lausanne between November 2014 and January 2018. Epidemiological, clinical and therapeutic data was collected and analyzed accordingly.

Results: A total of 35 patients were reviewed. The patients were in most cases (82%) referred by the pediatric rheumatologic or orthopedic surgeon. The sex ratio F/H was 3.3 (27 girls for 8 boys). The average age at the time of consultation was 12.8 years (9 to 18 years). A triggering event was found in 48% of our patients with the notion of trauma in 35% of these cases. The average duration of symptoms was 3.6 years prior to referral to the platform (1–11 years). Chronic pain affected more than 5 joints in 75% of the cases. School absenteeism was noted in 22% of the children with 2 cases of withdrawal. The platform revealed in 52% a primary pain disorder, in 28% an associated orthopedic problem, in 25% a difficult psychosocial situation and in 5% a rheumatologic problem. Therapeutic proposals were mainly personalized physiotherapy or mind body approaches such as hypnoses and depending on the findings, follow-up in orthopedics, rheumatology, child psychiatry or adolescent specialist consultation.

Conclusion: A long delay before children with chronic pain reach the platform was noticed. This could explain the severity of the presentation and the significant impact on school attendance and social life. The integrative clinical approach highlighted the multifactorial aspects of chronic pain and led to the development of an adapted multidimensional treatment to improve the prognosis and reduce the negative impact of chronic pain. We encourage pediatricians to detect and refer such patients early.

NaCHwuchs Prize: I wish to apply

I have read and understand application rules: No

Disclosure of Interest: None declared

PF39

TRANSGENDER YOUTH: IMPLEMENTATION OF A SPECIALIZED MULTIDISCIPLINARY TEAM CARE

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Introduction: Gender dysphoria (GD) refers to discomfort or distress caused by a discrepancy between gender identity and sex assigned at birth. Over the last decades, many clinics have reported an increase in the number of youths seeking medical help for GD. However, few clinicians are trained to take care of these adolescents.

Methods: This is the report of our new program for transgender children and adolescents. The aim is to highlight the needs of this specific population and to provide specialized multidisciplinary care.

Results: Our team consists of pediatric and adolescent medicine specialists, child psychiatrists, pediatric endocrinologists, pediatric gynecologists and specialists of reproductive medicine. The diagnosis of GD is confirmed according to the DSM-V criteria by a child psychiatrist from our clinic. GnRH analogs in order to block puberty are considered at age >12 years and/or pubertal stage Tanner 2. Gender-affirming steroids are considered for patients >16 years of age. We are currently following 7 pediatric patients with confirmed GD. Four out of 7 (57%) present a female-to-male (FTM) GD and 3/7 (43%) a male-to-female (MtF) GD. Close to half (43%) reported having felt they belonged to the opposite gender before the age of 5 years (early onset). Mean age at first endocrinologic evaluation was 14.7 years, with a puberty stage Tanner 4–5. GnRH analog injections were started at a mean age of 15.4 years. Only one patient (MtF) was interested in fertility preservation. A majority of adolescents (86%) presented with severe anxiety, depression, and suicide attempts. Two reported a sexual abuse during childhood. Five reported to be bullied at school (71%) and 3 patients had to be hospitalized in a psychiatric unit despite having started medical care.

Conclusion: We successfully implemented a specialized multidisciplinary team for transgender children and adolescents with coordination of care for assessment, treatments, and follow-up. The vast majority of these patients with GD also suffered from severe mental health issues and school bullying that required child psychiatric and adolescent specialist support. GnRH agonist treatment to block puberty is a cornerstone of today's treatment. However, our patients were referred quite late with an engaged puberty. These results show the importance of raising awareness and the need for early detection and referral of GD patients to a specialized multidisciplinary team.

Disclosure of Interest: None declared

PF40

HEALTH CARE USE OF UNACCOMPANIED MINORS RESETTLED IN THE CANTON OF GENEVA: A ONE-YEAR FOLLOW-UP STUDY

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Introduction: Unaccompanied minors (UM) are considered as the most exposed and vulnerable group among the entire refugee population. The number of UM applying for asylum in Switzerland grew rapidly during the period 2015–2016. All UM between 11 and 18 years who arrived in the canton of Geneva were referred to our university adolescent outpatient clinic for a systematic post-arrival health assessment. Follow-up appointments were offered for both physical and mental health concerns. The aim of this study was to describe health services access and use among UM following their initial 12 month post-arrival period.

Methods: We conducted a retrospective chart review of all UM who were assessed at the Geneva University Adolescent Outpatient Clinic between 01.01.2015 and 30.06.2016. We investigated data concerning visits to the primary care provider, the emergency department and the subspecialty outpatient clinics. We considered a follow-up duration of 12 months for each patient. We also examined hospitalizations during the same follow-up period. We computed descriptive statistics using the IBM SPSS Statistics Program.

Results: We identified 163 UM (males 89%, mean age: 16.4 years). Two thirds of them were from Eritrea (37%) or Afghanistan (35%). The median number of visits to the primary care physician was 3 (visit range: 1–13). Almost one third of the UM returned multiple times (≥3) to their primary care clinician after the initial assessment appointment. Ninety percent of them had consultations with a nurse for immunization and growth follow-up (median visit: 4, range: 0–15). One quarter of them were referred to a mental health professional for emotional disorders. Almost two thirds of them were referred to see at least one subspecialist, mostly a dermatologist or a cardiologist. Twenty-two percent of them visited multiple subspecialists (≥3). Nearly half of them visited at least once the emergency department because of trauma (33%), psychological distress (29%), miscellaneous symptoms (24%) and infectious diseases (14%). Nine patients (12%) visited the emergency room multiple times (≥3). One out of ten UM had to be hospitalized, mainly for a surgical intervention or alteration of emotional status.

Conclusion: UM represent a vulnerable population group with high need of health care services. Health care providers should be aware of the particular health concerns of this population in order to appropriately address their needs.

Disclosure of Interest: None declared

PF41

IS VULNERABILITY ASSOCIATED TO POORER HEALTH AMONG YOUNG PEOPLE?

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Introduction: To assess whether vulnerability among young people is associated to poorer health.

Methods: Data were drawn from the baseline (2015) wave of the GenerationFRee study, a cohort study among 5179 students aged 15–24 years in post-mandatory education in the canton of Fribourg. A vulnerability scale (VS) was created based on three variables: having a poor relationship with parents, reporting a below average socio-economic status, or being a below average student. Given the low number of participants reporting either 2 or 3 vulnerability variables, we aggregated these participants to obtain a final three group scale: no vulnerability variable (Control group; N = 3977; 76.8%), 1 vulnerability variable (VV1; N = 1012; 19.6%), two or more vulnerability variables (VV2+; N = 190; 3.7%). Groups were compared on perceived health status, emotional wellbeing, and frequent weight problems, headaches, stomachaches, articular pain, sleeping troubles and fainting in the past 12 months. Significant variables ($p < .05$) at the bivariate level were included in a multinomial logistic regression controlling for age, gender, family structure and academic track. Results are given as relative risk ratios (RRR) and 95%CI.

Results: At the bivariate level all health problems increased as the VS increased. At the multivariate level, compared with the control group, VV1 were significantly more likely to report a poor health status (RRR: 2.31 [1.54/3.47], low emotional wellbeing (1.72 [1.41/2.10]) and sleeping troubles (1.60 [1.24/2.07]). Youths in VV2+ also reported poorer health (5.60 [3.13/10.3]), low emotional wellbeing (2.60 [1.74/3.89]) and sleeping troubles (2.47 [1.57/3.89]).

Conclusion: Vulnerability is associated with poorer perceived health and lower emotional wellbeing. When controlling for confounding variables, sleep troubles is the only symptom that remains significant. The likelihood of all health problems increase with a higher level of the VS. Health professionals dealing with young people with difficult to explain symptoms (and specially sleeping troubles) should assess their vulnerability level.

Disclosure of Interest: None declared

PF42

PRECISION MEDICINE FOR PEDIATRIC DIABETES THROUGH A RAPID GENETIC TEST USING NEXT GENERATION SEQUENCING

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Introduction: Monogenic diabetes (MD) accounts for at least 3% of all pediatric diabetes cases. Because of its wide phenotypic spectrum, MD is often misdiagnosed as type 1 or type 2 diabetes. While clinical and biochemical parameters can suggest MD, a definitive diagnosis requires genetic analysis. We conducted a broad study to diagnose MD cases. Then, with the gained knowledge, we designed a new diagnostic tool to obtain a comprehensive analytical instrument for the diagnosis of MD. A correct diagnosis of MD is crucial to optimize treatment and thereby improve metabolic control.

Methods: Diagnostic tool (Haloplex technology): This custom assay, designed based on liquid phase capture, allows for the trapping of all coding regions of the 42 genes and the respective splicing regions.

Results: Here, we developed a new diagnostic panel of 42 genes, including all the genes causing Maturity onset diabetes of the Young (MODY) and neonatal diabetes. The panel was validated with independent samples of known MD patients. We have now analyzed the first 16 consecutive patients, sent in from all over Switzerland, and identified a monogenic disease in 56% of the subjects. We found three different variants in the *GCK* and *HNF1A* genes, one in the *ABCC8*, *EIF2AK3* and *PAX4* genes. In seven patients the result remained negative. Interestingly, the *ABCC8* variant was reported to be functionally inactivating and therefore compatible with the congenital hyperinsulinism presented by the newborn patient. This is in contrast to activating *ABCC8* mutations known to cause neonatal diabetes.

Conclusion: Our newly developed next generation diagnostic panel shows a pick-up rate of 56% in the first 16 consecutive patients, which is above the published rates of 21% to 37% in the UK and 25% to

30% in France. The panel detects missense variants, insertions, and deletions, as well as activating or inactivating mutations. These results indicate that the three patients with *GCK* diabetes didn't need any treatment, the patients with *HNF1A* mutations could be switched to oral sulfonylurea or glinides. The congenital hyperinsulinism could successfully be treated by diazoxide and the neonatal diabetes due to the *EIF2AK3* mutation needed insulin injections. These cases illustrate how applied precision medicine can tailor treatment to the needs of the individual patient with the aim to reduce long-term complications.

Disclosure of Interest: None declared

PF43

WEIGHT OUTCOME AT FIVE YEARS OF ADOLESCENTS TREATED IN A MULTIDISCIPLINARY GROUP THERAPY FOR OBESITY

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Introduction: Obesity is a serious health problem in Switzerland affecting approximately 5% of the adolescents' population. Since 2008, multidisciplinary intensive group therapy (MGT) for obesity is reimbursed by Swiss health insurances. The aim of our study was to assess the BMI zscore evolution of adolescents who participated to a MGT at 5 years.

Methods: All adolescents (n = 79) aged 12 to 18 years who participated between 2009 and 2012 to a MGT for obesity were contacted. MGT consisted in 6 months of family-based behavioral therapy with weekly psycho education sessions and physical activities. At the end of the intensive program, a regular medical follow up was proposed to all of them. The adolescents were reassessed at 1 year (T1) by a physician. In 2017 (T2), all participants who accepted to meet our research nurse were asked to answer a questionnaire about diet and physical changes they were able to maintain since the MGT, as well as their actual weight follow up. Body weight (kg) and height (cm) were measure. BMI was calculated as weight/height squared (kg/m²), and z-score were derived using the World Health Organization references.

Results: A total of 57 (72%) adolescents (mean age 19.9 years; female 59.6%), accepted the follow up meeting. Mean time since the beginning of the program was 5.7 years. Among those who lost weight at T1 (50.9%), 58.6% have maintained or decrease their IMC z-score at T2. Among those who gained weight at T1 (5.3%), 66.7% have decreased their IMC z-score at T2. Most of them have maintained a regular physical activity at T2 (62.7%), with a positive impact (stability or decrease) on the IMC zscore, particularly when performed on a daily basis ($p < .001$). At T2, 46.2% of the participants have reported to maintain diet changes, with a positive impact on their BMI z-score in 61.1% of the participants. Only 28 adolescents (49%) have monitored their weight with the help of a health professional. 29 (50%) did not weigh themselves.

Conclusion: Multidisciplinary group therapies have a positive impact on the IMC zscore at five year for more than half of the adolescents, particularly for those who maintained a daily physical activity. However, only few of them kept a follow up with a physician. Obesity being a chronic disease with medical co morbidities, further efforts should be made to maintain a regular weight and medical follow up within this population.

Disclosure of Interest: None declared

PF44

CURRENT MEDICAL CARE OF CHILDREN AND ADOLESCENTS WITH DISORDERS/DIFFERENCES OF SEX DEVELOPMENT IN SWITZERLAND

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Introduction: Since 2000 understanding of biology of sex development increased tremendously thanks to genetic research. This lead to new classification for persons with disorders/differences of sex development (DSD) based on genetics, and guidelines from the UK recommend revising medical care for persons with DSD by setting up interdisciplinary DSD teams. In Switzerland, persons with DSD asked for better care, stimulating the Swiss National Ethics Commission in 2012 to recommend improved treatment of DSD persons. We aimed to describe the current situation of medical care of DSD in Switzerland.

Methods: A questionnaire was sent to pediatric endocrinologists of eight hospitals in Switzerland who were either part of the Working Group DSD of the Swiss Society for Pediatric Endocrinology and Diabetology (AG DSD SGPED) or committed to set up a DSD cohort. We asked them to estimate numbers of treated DSD persons, indicate specialists involved in DSD care and report DSD-related research projects.

Results: The eight clinics cover >85% of all newborns in Switzerland. Each year, around 24 newborns and 24 children and adolescents were diagnosed with complex DSD, e.g. ambiguous genitalia. In total, the eight clinics care for about 750 children with a DSD diagnosis according to the Chicago consensus classification. Of those, 90 had complex DSD, 130 congenital adrenal hyperplasia, and 130 Turner syndrome. Most clinics used the UK guidelines and 7/8 had established interdisciplinary DSD-teams including specialized pediatricians from many areas, geneticists, general psychologists or social counselors. Only few clinics had specialized psychologists, social counselors, nurses, or ethicists (fig. 1). Young adults with DSD were transitioned to adult medicine at age 16–25 years, but it was unclear who can provide the optimal care for them, because there were hardly any adult physicians specialized on DSD. All participating clinics indicated to establish a Swiss DSD cohort, and several clinics had their own research projects, including basic science, ethics and epidemiology.

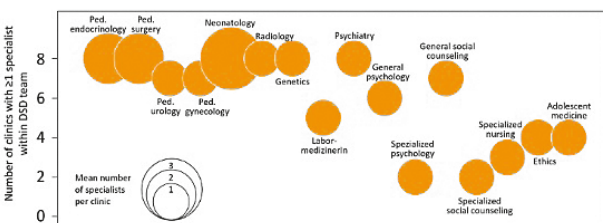


Figure 1: Specialists within DSD team: Number of clinics who have at least one specialist within DSD team or in supply network. The area of the circle reflects the mean number of specialists per clinic.

Figure 1

Conclusion: We identified gaps in psychological care, in the collaboration with adult medicine and with associations of DSD individuals, and in epidemiological monitoring of DSD. Few clinics had already included specialized psychologists in their DSD team. The new Swiss DSD cohort will help to answer open questions on epidemiology of DSD in Switzerland. However, sustained funding is lacking.

NaChwuchs Prize: I wish to apply
Disclosure of Interest: None declared

PF45

“I THINK IT COULD BE GOOD TO CREATE A CAMPAIGN TO PREVENT THE MISUSE OF SEXTING BECAUSE, IN THE END, THAT’S THE PROBLEM”

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Introduction: To gather and compare the opinions of youths, parents and teachers on how sexting can be defined.

Methods: Exploratory qualitative research including 32 youths (17 males) aged 16–21 years, 11 parents of children aged 11–18 years and 18 8th grade or higher teachers who participated in eleven focus groups. Discussions were recorded, transcribed verbatim and a thematic content analysis was performed.

Results: Sexting was defined in terms of actions (sending, receiving, spreading), supports (text, audio, photo, video), sexual characteristics and contexts. Two interpretations were given to define sexting in terms of actions. First, mostly in the groups of youths, sexting was defined as a positive activity between two consenting persons with only two actions (sending and receiving) and as a risky behavior but not necessarily leading to negative consequences. “Let’s say there is a risk [...]. But it is not inherent to the thing. For me, when you send a photo, you do not tell yourself: ‘Ah! Maybe it will be shared with others!’[...]” (Male 18 years). For these participants, the term sexting should not be used to talk about problems linked to it. “When it deviates it becomes harassment, does it not? [...]” (Female 18 years). Second, mostly in the groups of parents and teachers, sexting was defined as a violent and deviant behavior per se in a context of threat, blackmail or harassment and the action of spreading a message by sharing it with others was also considered as part of the definition. “Just before, we were talking about sexual harassment, it is a violent term. And precisely, I have the impression that the word Sexting is a word that made harassment common place.” (Teacher).

Conclusion: It is necessary to develop a precise and consensual definition of sexting by separating its above-mentioned elements and by using a specific vocabulary according to the youth’s perceptions. Prevention messages should aim to reduce the risks by combating the problems of consent, pressure, blackmailing, harassment and dissemination rather than prohibiting the sexting per se. Using a suitable vocabulary and a precise definition based on the perspective and the practice of young people themselves would help to better direct the prevention and accurately measure the phenomenon. A clear definition of sexting and its possible abuses could also lead to a better understanding for reference adults.

Disclosure of Interest: None declared

PF46

SOME ECONOMICAL CHARACTERISTICS OF HAVING A CHRONIC CONDITION

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Introduction: To assess economic factors related to having a chronic condition among youths.

Methods: Data were drawn from the 2017 wave of the GenerationFree longitudinal study conducted in 11 post-mandatory schools in the Canton of Fribourg. Participants (N = 2208, mean age 17.7 [16–25], 46.3% females) filled out a web-based self-administered anonymous questionnaire assessing their lifestyle. Participants were asked whether they had a chronic condition that limits their daily activities (CCLim, N = 54, 2.45%), that does not limit their daily activities (CCNonLim, N = 255, 11.55%), or no condition (NoCC, N = 1899, 86.01%). Groups were compared on perception of their family financial situation compared to others in Switzerland (below average, average, above average), of having enough money of their own (not enough, enough, more than enough), whether they had debts (yes/no), how much pocket money they had available each month (mean), and whether they had access to public financial support. Groups were compared at the bivariate level and then at the multivariate level using a multinomial regression analysis to assess the variables associated with having a chronic condition using NoCC as the reference category, controlling for age, gender, and academic track. Data are presented as relative risk ratios (RRR) with 95% CI.

Results: At the bivariate level, compared to NoCC, CCLim and CCNonLim groups were significantly more likely to perceive their family financial situation as below average (19.98% and 11.47% vs. 7.90%), not having enough money themselves (33.75% and 20.95% vs. 16.31%), and to receive public financial support (6.21% and 4.66% vs. 2.02%). At the multivariate level, compared to the NoCC group, CCLim were more likely to think that they did not have enough money (RRR: 1.63 [1.16:2.28] p = 0.004); and the CCNonLim group had the same trend although not significant (1.20 [0.98:1.48]; p = 0.07).

Conclusion: These results show that economic links with having a chronic condition that limits daily life are mostly individual as these youths believe not to have enough money. This might be associated to the feeling of being different from their healthy peers; and having less money might even accentuate this feeling of being different. Further research should explore the economic aspects with more objective indicators. When screening for psychosocial aspects among adolescents with chronic conditions, health professionals ought to assess for individual economic level and whether help is needed in this regard.

Disclosure of Interest: None declared

PF47

HEALTH BEHAVIORS OF WOMEN WITH TURNER SYNDROME

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Introduction: Women with Turner Syndrome (TS) have an increased risk of cardiovascular morbidity and mortality, mainly due to congenital cardiac anomalies and coronary artery disease. Atherosclerosis may already begin to develop in childhood and is strongly related to cardiovascular risk factors. Lifestyle-related risk factors for atherosclerosis include physical inactivity, smoking and excessive alcohol consumption. The lifestyle of young women with TS is currently unknown. We therefore aimed to investigate health behaviors in Swiss women with TS.

Methods: In 2012 we sent a postal questionnaire to 79 women with TS aged ≥18 years and living in Switzerland by the time of the survey. The survey was part of the nation-wide Swiss Growth Study, that included patients treated with recombinant growth hormone since 1985 in 11 Swiss centers of paediatric endocrinology. We compared reported frequencies of smoking, alcohol consumption and physical activity to age-adjusted women of the Swiss Health Survey 2012, a representative general population survey.

Results: Forty-five of the 79 contacted women with TS answered the questionnaire (response rate 57%). These women smoked less compared to controls: 80% of TS vs. 58% of control women had never smoked and 13% of TS vs. 8% of controls were ex-smokers. Only 7% of women with TS vs. 34% of controls were current smokers (all $p < 0.01$). Women with TS drank equally often alcohol than controls ($p > 0.05$) but reported less often than controls to engage in binge drinking (66% vs 29%). About a third (29%) of women with TS reported monthly binge drinking compared to 69% of controls ($p < 0.001$). Women with TS engaged equally often in sports and they were equally active in their daily life as controls, but engagement was less than officially recommended ($p < 0.05$).

Conclusion: Women with TS smoked less and engaged less in binge drinking than controls, and their physical activity level was comparable. These results may indicate that health care professionals motivated women with TS to have a healthy lifestyle. However, women with TS had lower levels of physical activity than recommended by the World Health Organisation (WHO) and the Centers for Disease Control and Prevention (CDC). We thus need further strategies to encourage physical activity, in these patients at high risk for cardiovascular diseases.

NaCHwuchs Prize: I wish to apply

Plaschke Prize: I wish to apply

I have read and understand application rules: Yes

Disclosure of Interest: None declared

PF48

VITAMIN D PRACTICE CHARACTERISTICS OF SWISS PEDIATRICIANS: A NATIONAL INQUIRY

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Introduction: Five years ago, the recommendation that Swiss infants receive a supplementation of vitamin D₃ during the first year of life has been extended. Nowadays, vitamin D₃ is recommended also for the second and third year of life and exclusively during the winter months for older children and adolescents. Furthermore, both "alcoholic" preparations and new "oily" preparations of vitamin D₃ are presently commercialized. The purpose of the study was to identify how pediatricians prescribe vitamin D₃.

Methods: In 2015, a close-ended 20-item not commercially sponsored questionnaire was designed, which was subsequently tested and corrected by a group of Swiss-Italian pediatricians. Native translators specialized in medicine translated the final questionnaire from Italian into French and German using the forward-backward method. Between December 2016 and March 2017, 795 (52%) of 1530 invited pediatricians answered the questionnaire.

Results: Ninety-seven% (N = 769) of respondents almost always prescribe vitamin D in infants 12 months or less of age, whereas the prescription is significantly lower for children 2 to 3 years of life (72%) and especially for children older than 3 years (3.2%; $P < 0.0001$).

The "oily" formulations of vitamin D₃ are largely more popular than the traditional alcoholic (90%, $p < 0.0001$). Finally, the majority of the respondents (66%) prescribe vitamin D in a once-a-day regimen.

Conclusion: This study indicates that most of the pediatricians usually don't prescribe vitamin D supplements in children of three or more years of age. Oily vitamin D₃ preparations have nowadays replaced the "alcoholic" preparations and are usually prescribed in a once-a-day regimen.

NaCHwuchs Prize: I wish to apply

Plaschke Prize: I wish to apply

I have read and understand application rules: Yes

Disclosure of Interest: None declared

PF49

LET'S THINK ABOUT TRANSITION

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Introduction: In the literature, several diabetic transition-programs are described, mostly based on experiences and expectations of health professionals worried by the wellbeing of their patients. The patient perspective has also been studied, mostly by questionnaires. Through a pilot project, we try to develop a different setting to better integrate our adolescent diabetic patients into the process of transition to adult care.

Methods: In view of an imminent transfer to the adult clinic, all type 1 diabetic adolescents aged 16 to 18 years are invited, in small groups of 8 to 11 youngsters, to a half-day meeting. We then propose a structured intervention including a playful introduction, three practical workshops about medical, technical and dietetic aspects of diabetes and a session of brainstorming about their questions and expectations concerning their transfer to an adult clinic. The intervention ends with a lunch in common.

Results: Here, we present the results of our first three interventions with 29 adolescents, nine of which have transitioned to adult care since the intervention. Starting by describing their diabetes using "colours", "feelings" or "animals", the adolescents confronted their visions with their peers; through unexpected questions about their lived experiences with a chronic illness, we then stimulated the debates among them. This first part was most appreciated by our patients; they enjoyed meeting other adolescents having similar life experiences as themselves and adored the opportunity to speak about their disease in a quite unconventional way. The second part of our intervention consisted of three workshops developing aspects on nutrition, calculation of insulin doses, decision making in diabetes care and lipodystrophy; they allowed them to strengthen basic knowledge about diabetes self-management. Finally, when asking them about their concerns and expectations for the transfer to adult care, they pointed out a lot of practical aspects and also focused on the fact that the role of their parents will change.

Conclusion: The proposed interventions helped the adolescent patients and health professionals to initiate the process of transition. The intervention gave them the opportunity to share about their life experiences and to focus on the adolescent's expectations. The participants were grateful to be considered as experts in the field of their own disease.

Disclosure of Interest: None declared

PF50

TRANSITION TO ADULT HEALTH CARE: IS THERE SUFFICIENT COLLABORATION BETWEEN PAEDIATRIC SPECIALISTS AND GENERAL PAEDIATRICIANS IN PRIVATE PRACTICE?

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Introduction: Guidelines for transition of adolescents with chronic disorders from paediatric to adult health care (AHC) include general paediatricians in private practice (GPP) as an important source. However, little is known about collaboration between paediatric specialists and GPP regarding the transition process. The aim of this pilot study was to investigate how consultant paediatric specialists at hospitals (CPS) and GPP collaborate during the transition process of these adolescents to AHC.

Methods: A questionnaire, specifically developed for this survey, was completed online by CPS at the University Children's Hospital Zurich and GPP of the Canton of Zurich.

Results: 59 GPP and 28 CPS completed the survey (participation rate 41% for GPP and 76% for CPS). Both GPP and CPS were very satisfied/mainly satisfied with collaboration (GPP 84.8%, CPS 85.7%). GPP were more satisfied with collaboration with paediatric specialists in private practice than with CPS ($p = 0.012$). The current way of correspondence was highest rated for the combination of email & letter via mail (GPP 37.3%, CPS 28.6%), although both GPP and CPS indicated clear preference for email only correspondence (GPP 33.9%, CPS 28.6%). While GPP indicated they would prefer a complementary role (47.5%) or the main responsibility (16.9%) in the transition planning from paediatric to specialist AHC, 42.9% of CPS assigned no role to GPP in the transition process to specialist AHC. GPP felt rather well (42.9%) or partially well informed (35.7%) about the CPS's transition planning for transfer to specialist AHC. Specialists felt rather insufficiently informed (32.1%) or not informed at all (53.6%) about the GPP's transition planning for transfer to an adult general practitioner. Just 28.5% of GPP and 0% of CPS said that the timing of transfer was always/often discussed with each other in order to prevent concurrent transfer. Duration of transition period was estimated shorter by GPP than CPS: GPP 55.9% 6 months or shorter, CPS 78.6% 6 months or longer ($p < 0.0001$). The majority of both GPP and CPS perceived the ages 18–20 or older as the best age for transfer to AHC. Among the perceived reasons for difficult transition, insufficient communication with patients/parents and between paediatric and adult physicians outweighed insufficient communication between GPP and CPS.

Conclusion: The tasks of GPP and CPS should be defined to complement professional support for adolescents during the transition to AHC.

Disclosure of Interest: None declared

PF51

HOW DO PEDIATRICIANS RULE OUT UNDERLYING ORGANIC ETIOLOGIES AMONG ADOLESCENTS WITH THE FIRST EPISODE OF PSYCHOTIC SYMPTOMS?

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Introduction: Psychotic symptoms may come with a number of medical diseases. A comprehensive differential diagnosis, including underlying organic causes, is the cornerstone of the medical management by pediatric, child psychiatric or adolescent medicine specialist. The aim is avoiding a mistaken attribution of psychotic symptoms to a psychiatric syndrome and not providing adequate treatment for organic condition. There is a gap in guidelines for this initial medical assessment. Our goal is to provide pediatricians with an algorithm to rule out organic causes, in adolescents having the first episode of psychotic symptoms.

Methods: We performed a review of the literature until December 2017, from MEDLINE, CINAHL and databases from the Guidelines International Network and the National Guideline Clearinghouse searching for such guidelines. Inclusion criteria were management and assessment of first psychotic symptoms among children and adolescents. We also gathered current practice and recommendations from local experts in adolescent medicine, hospital pediatrics, psychiatry, neurology, immunology, metabolism, infectious diseases and endocrinology.

Results: We found no evidence-based guidelines on this question, but some narrative papers discussing treatment and side effects in first episodes of psychosis. Only a few provided diagnostic strategies. Majority of authors and experts reported a consensual agreement that no further investigation is needed for "typical" manifestations of psychosis. However, they suggested further investigation for "atypical" manifestations (e.g. abrupt onset, catatonia, visual hallucination, confusion, treatment resistance). In the presence of atypical manifestations, a comprehensive assessment (e.g. blood test, cerebral MRI, EEG) is essential in order to rule out several organic conditions that can be associated with psychotic symptoms, such as late-onset inborn errors of metabolism, substance abuse, autoimmune or infectious diseases, neurological and endocrine disorders.

Conclusion: For atypical manifestation of psychosis in a first episode, we suggest ruling out potential underlying organic causes. An evidence-based guidance for this assessment is needed, since some organic conditions could be difficult to diagnose and clinicians may be unfamiliar with some rare conditions. The sooner a right diagnosis is obtained, the sooner an appropriate treatment may be given.

Disclosure of Interest: None declared

THE ACUTE TOXICITY PROFILE OF A TEETHING GEL CONTAINING SALICYLAMIDE IN TODDLERS: OBSERVATIONAL POISONS CENTRE-BASED STUDY

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Introduction: In Switzerland a teething gel containing 80 mg salicylamide and 1 mg lidocaine per gram (line of 2.5 cm) is available (tube of 25 g). The therapeutic dose is age-dependent: A line of 2 cm in children <11 month, 2.5 cm in children of 11–14 month, 3 cm in older children. The toxicity of lidocaine is well known, and with ingestion of 25 g of the gel a toxic dose of lidocaine is not reached. The product contains also a non-toxic caring ingredient. The aim of the study was to determine the acute toxicity profile of this gel in overdose, since available information of salicylamide is limited.

Methods: A retrospective review of acute overdose of a salicylamide containing gel in toddlers (≤ 3 y), reported to our poisons centre from 1997–2017 with evidence of sufficient exposure and high causality. The severity of observed symptoms was graded according to the Poisoning Severity Score.

Results: 123 patients, 4 unknown gender, 71 (58%) females, 48 (39%) males with a mean age of 1.8y (0.8–3.7y), were included. No effects were reported in 13 cases (11%), minor symptoms in 100 cases (81%), moderate symptoms in 10 cases (8%). There were no severe or fatal cases. In 86 cases the ingested amount of gel was known and ranged from 6–25 g gel (mean 18.8 g; 0.7–2.8 g/kg). No symptoms occurred after ingestion of 1–2.7 g/kg (mean 1.7 g/kg), and mild symptoms were observed after ingestion of 0.5–2.8 g/kg (mean 1.6 g/kg). Moderate symptoms occurred after ingestion of 0.8–2.5 g/kg (mean 1.6 g/kg). Observed signs and symptoms were ataxia ($n = 61$), somnolence (61), vomiting (38), muscular hypotonia (20), drowsiness (17), dizziness (11), paleness (9), short lasting sopor (8), abdominal pain (6), agitation (4), nausea (3), confusion (2), bradycardia (2), tachycardia (2). All symptoms resolved spontaneously. In 42/123 cases the duration was known and was reported to be 5–360 min (mean 120 min), which is in accordance to the half-life of 72 min of salicylamide. In 54/123 cases the latency between the ingestion and the onset of symptoms was known and was reported to be 10–165min (mean 30 min; Tmax of salicylamide: 12–120 min).

Conclusion: This salicylamide containing teething gel seems to have a favorable acute toxicity in overdose up to 25 g (2 g salicylamide), although short-term sopor can be very frightening to care takers. Signs and symptoms were consistent with described adverse effects of salicylamide. A short-time observation at home without gastrointestinal decontamination is reasonable, and a medical check is recommended only in case of sopor.

Disclosure of Interest: None declared

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CONCERTINO: USING TABLETS TO EMPOWER PEDIATRIC PATIENTS AND TO REDUCE LONELINESS

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Introduction: In October 2015, during a Hackathon by the Port association at CERN the idea was born to create a mobile application that could empower patients by keeping them informed about their hospitalization and by increasing communication with the care team. The pediatric application, Concertino, was created to: 1) Improve the quality of life of hospitalized patients, 2) Facilitate contact between patients and their social support structure, 3) Provide pain relief with distraction during treatments and procedures, and 4) To facilitate patient education.

Methods: A multidisciplinary working group conducted a literature review to identify strategies that diminish stress and anxiety both during and after hospitalization. A targeted needs assessment on the use of tablets was then conducted with patients, families, and medical personnel. An agile, user-centered, software development method was used to build the application.

Results: The literature review identified that pediatric patients are vulnerable while hospitalized because they are in an unknown environment, are unfamiliar with hospital processes, and have little control over their plan of care. 17 families and 36 medical personnel participated in the needs assessment. Both groups wanted multi-media tablets to: facilitate patient information and education, as a distraction tool for treatments and procedures, and to increase

communication with friends and family who are not present during the hospitalization. A personal avatar was created to guide the patient through the six modules of the mobile application. 1) A personalized hospital agenda extracted from the hospital information system including the recreational activities in the hospital. 2) A personalized presentation of the care team for the patient to easily identify the various actors. 3) A logbook of questions asked to the healthcare team. 4) A personalized medical atlas that provides interactive medical information based on the patient's diagnosis. 5) Practical information concerning the hospital stay. 6) A multi-media electronic post card delivery system to encourage social support. Beta testing is currently in progress. An evaluation of the application is currently being conducted to measure the impact on patient's situational awareness and shared decision-making.

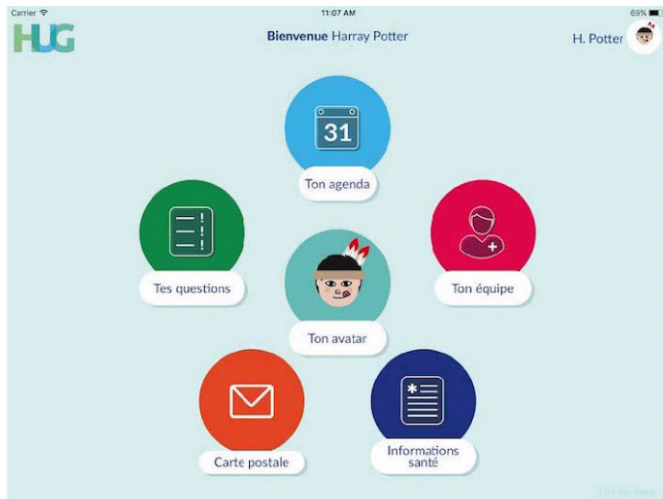


Figure 1

Conclusion: A mobile application targeted for the needs of hospitalized pediatric patients has the potential to empower hospitalized children. The impact of the mobile application is currently being assessed.

Disclosure of Interest: None declared

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EARLY MINDFULNESS-BASED INTERVENTION FOR VULNERABLE ADOLESCENTS: PROJECT FOR A FMRI RANDOMIZED CONTROLLED TRIAL

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Introduction: Early intervention strategies in “at-risk” populations are nowadays central for the prevention of psychiatric illnesses. Among risk factors, stress reactivity is a parameter that could be modulated by early interventions that aim to decrease anxiety levels and to normalize activity in affected brain circuits, such as mindfulness-based interventions. However, findings regarding the cognitive and neural mechanisms underlying their efficacy are inconsistent. Therefore, we designed a study to longitudinally assess, among vulnerable adolescents presenting increased anxiety and stress reactivity, the impact of a mindfulness-based intervention on the activity of brain circuits involved in emotion regulation when submitted to a neuroimaging psychosocial stress task. We will also assess its clinical impact on psychological and functional symptoms.

Methods: We will randomize anxious adolescents recruited from multidisciplinary consultations for adolescents into mindfulness intervention group and standardized care group, and compared with low-risk adolescents from the general population. Pre- and post-treatment stress reactivity will be assessed in a longitudinal design at 3 times points (pre-, post-intervention, after 18 months). Half of each cohort will undergo a 12-weeks mindfulness-based intervention with one session every week and individual daily practice. After re-test of clinical and neuroimaging protocol, the other halves of each cohort will also benefit from 12 weeks mindfulness-based intervention. The protocol will include a thorough clinical characterization, then subjects will perform two MRI sessions, the first with structural, DTI, and spectroscopy data acquisition. After a break, they will perform in the MRI a well-known stress task with a social component and adapted to target stress reactivity. Complementary physiological measures will also be recorded.

Conclusion: We expect that after the mindfulness-based intervention, at-risk adolescents will show a decrease in clinical non-specific symptoms, in particular anxiety and stress levels, and a normalization of activity and connectivity in regions pertaining to emotion regulation circuits. We expect this effect to be specific on the activity of networks elicited by psychosocial stress.

Disclosure of Interest: None declared