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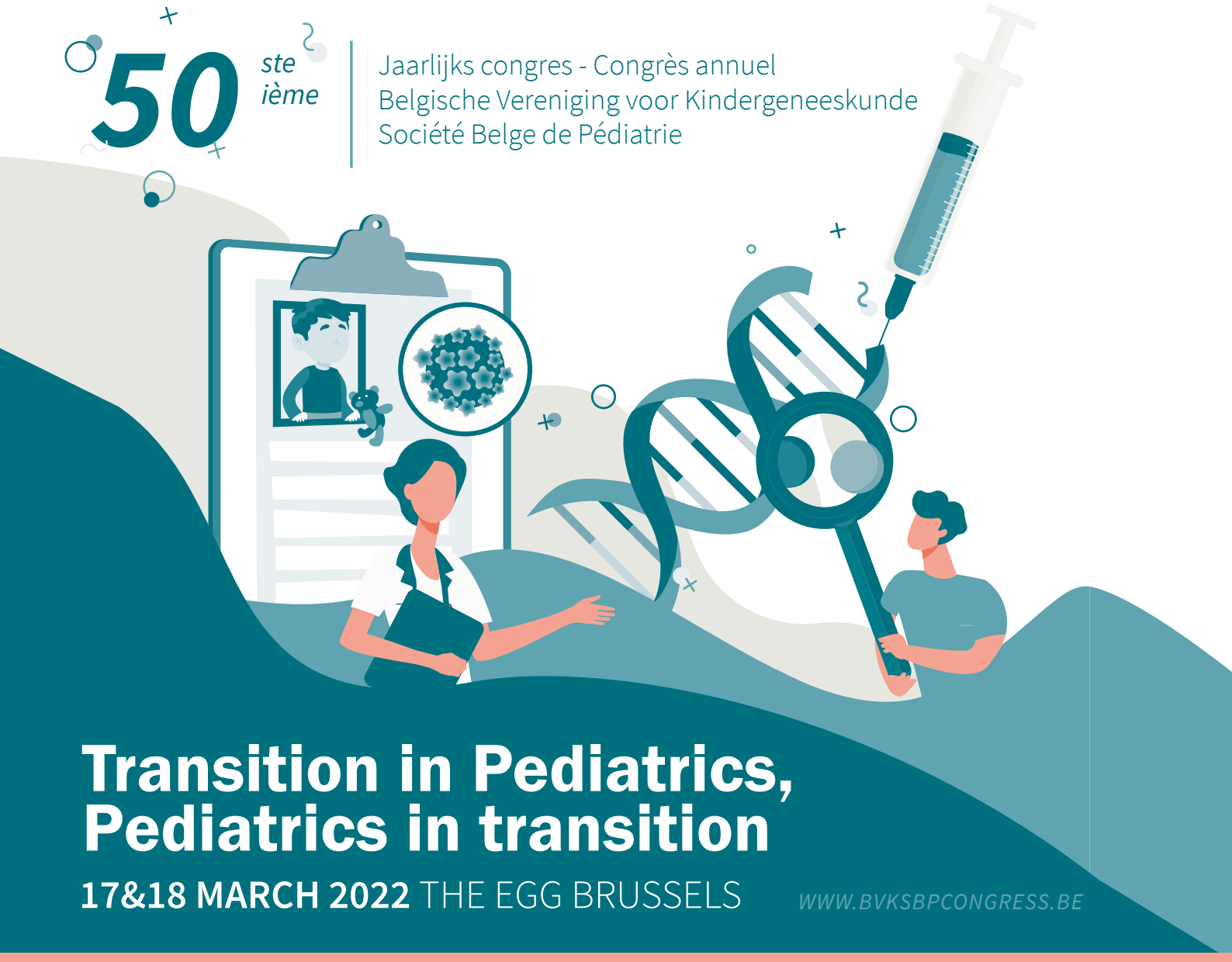

Transition in Pediatrics, Pediatrics in transition

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SO 33.

A snapshot on practices and trends on vitamin K prophylaxis in term neonates in FlandersA snapshot on practices and trends on vitamin K prophylaxis in term neonates in Flanders

K. Keiren, M. van Winckel, K. Allegaert

KU Leuven, UZ Gent, U Gent, Erasmus MC UMC

Introduction

Vitamin K prophylaxis in neonates is relevant to reduce vitamin K deficient bleeding, but practices vary on route of administration, dosing, and repated dosing during breastfeeding. We intended to document current practices in Flanders as an update of the VVK guideline is planned.

Methods

Following ethics approval (MP016741, KU Leuven), and supported by the VVK secretary, an online questionnaire circulated (Q1/2021) to all heads of the relevant departments (paediatrics, neonatology), with an additional search on midwifery units. The questionnaire focused on the current practices on vitamin K prophylaxis in term cases and on recent changes in practices, and collected information on experiences with parental refusal.

Results

Responses from 56/59 Flemish maternities and 17/30 midwifery units that do home deliveries unveiled relevant variability. About 50% of maternities use the intramuscular (1-2 mg), about 50% the oral (1-2 mg) route, with 5 different maintenance doses in breastfed infants. Nine/13 recently (<5 years) changed regimens were a switch from oral to intramuscular. Midwifery units only use oral vitamin K, with 6 different regimens. Both paediatricians and midwives reported personal experience with parental refusal of intramuscular (20/54 and 13/15) or any prophylaxis (11/54 and 16/16) respectively.

Conclusions

This snapshot on vitamin K prophylaxis in term neonates born in Flanders provides a contemporary and reliable overview on the diversity in practices to support the VVK guideline update.

SO 34.

Controlling the flood of study feasibilitiesControlling the flood of study feasibilities

E. Degraeuwe, L. Persijn, L. Nuytinck, E. Levtchenko, N. Deconinck, E. Sokal, S. Vande Velde, O. Gilliaux, K. Logghe, A. Raes, M. Turner, J. Vande Walle

U Gent, UZ Gent, UZ Leuven, HUDERF, UCL, CHU Charleroi, University of Liverpool, AZ Delta Roeselare

Background/Aims

Due to an increase in multicentric paediatric clinical trials after 2007, following the EU Paediatric Regulation there has been a substantial increase in the number of feasibility questionnaires (FQ). A FQ contains questions on site requirements, principal investigator (PI) capabilities and recruitment estimates. A FQ is usually completed by the PI and who ideally is supported by a clinical research coordinator (CRC). It is difficult to complete FQs in a timely manner if the site does not have historical experience and/or research infrastructure. Paediatric clinical trial conduct has made many and unique advances through the conect4children (c4c) pan-European network, funded by the Innovative Medicines Initiative (IMI2). The Belgian National network within c4c is coordinated through center Ghent University Hospital and supports FQs in Belgium. We examined the role of a national, centralized coordination center to pre-fill feasibility questionnaires and quality control responses from sites.

Method

This report describes the 4-year learnings of prefilling and performing quality control for FQ by the Belgian Network representative. Additional information has been included from a broad survey sent to the 15 sites in the Belgian Network, of which 32 responses from 13 unique sites have been collected.

Results

PIs are confronted by between 10 to 50 FQ requests per year, each taking at average 60 minutes. The number of redundant questions asked by sponsors is on average 43% of the FQ. Of the 112 completed feasibilities, approximately 82% required quality control adjustments by our national coordinating center. Inconsistencies were primarily found in PIs' report of experience, number of paediatric studies conducted, recruitment estimates and site qualifications. With a centralized and repetitive collection of FQ, a prefill of 65% of the requested information and potential corrections can be performed. A time reduction of 10 to 46% is estimated when a FQ is facilitated through the national representative.

Conclusion

The increase in paediatric clinical trials has substantially burdened sites within Belgium. Quality control and adjustments by a national central organization could be beneficial to increase feasibility quality and efficiency.

Short Oral Presentation

SO 35.

Pulse oximetry to screen for critical congenital cardiopathy in neonates: current practices in Flanders.Pulse oximetry to screen for critical congenital cardiopathy in neonates: current practices in Flanders.

R. Conelissen, K. Allegaert

KU Leuven

Background

Congenital cardiopathies are the most common group of congenital malformations (prevalence 9.4/1000). About 15-25% of these cases are critical congenital heart diseases (CCHD), any ductus dependent malformation, in need of a surgical or invasive intervention in the first 28 days of life). Pulse oximetry (PO) aims to identify CCHD cases as 'pre-collapse' detection is associated with improved outcome.

Methods

Following ethics approval (MP017253, KU Leuven), and supported by the VVK secretary, an online questionnaire circulated (Q1/2021) to all heads of the relevant Flemish departments (pediatrics, neonatology). The questionnaire focused on current practices, implementation pathway and perceived burdens on PO screening.

Results

The response rate (54/59, 91%) was high. As 48/54 of the maternity wards have a systematic PO screening program, this means that at least 48/59 (81%) of the maternity wards implemented systematic PO screening. Before the VVK guideline (≤2015), there were 2 maternity wards that already conducted PO, with a steady annual increase (+4, +8, +13, +8, +11, +1, one unit has not reported on the year of implementation), until 2021. Other units only screen in the event of abnormal clinical findings (n=4), and 2 of the responders do not have a PO strategy yet, while the majority of these maternities intend to implement systematic PO screening in the next year(s). Commonly reported barriers are limited resources (time, staff, equipment) in the absence of funding, the need for training initiatives, the presence of false-positives, the absence of echocardiographic expertise in the event of a positive screening, or interference with earlier discharge.

Conclusions

Implementation of cardiac screening has been quite successful. Similar data in the French speaking regions are likely of add on benefit, while formal registration as valid screening technique within the existing framework of preventive medicine and screening practices (regional), and subsequent reimbursement are needed for sustainability.

SO 36.

Medical Decisions in Pediatrics in the Context of Social versus Biological parenthood.

J. Toelen, I. Boone, K. Matthijs, S. Lierman, P. Dewinter, D. De Coninck

KU Leuven, Spaarne Gasthuis, Kulak Kortrijk Campus

Background/Aim
Alternative family configurations are becoming more prevalent, yet our current legislative statutory does not support stepparents to play a relevant role in the medical decisions of their stepchildren. In this study, we investigate the opinion of Flemish and Dutch adults regarding the inclusion of stepparents in medical decision making in minors and the factors influencing that decision.

Methods
We used a case-based survey, using four hypothetical but realistic cases from the parental perspective. Two thousand participants completed the questionnaire online in October 2020. Participants were selected by gender (50/50) and age (35-55yr), no other inclusion criteria were appended. We performed a binary logistic regression to conduct a multivariate analysis, using SPSS statistics.

Results
We make two main observations. Firstly, participants allowed stepparents to be involved in cases when medical information had to be shared (76%) or informed consent signed (61%). However, when the stepparent objects against previously approved medical interventions by the biological parent or the treating physician, respondents are considerably less likely to support the stepparent's decision, as shown in the vaccine (38.9%) and blood sample case (26.7%). Secondly, we found that region, gender and having (step)children were the three most significant factors influencing the results. Dutch participants and women were less likely to include stepparents in information sharing than Flemish participants and men, but they were more likely to support the stepparents' decision to disagree with the biological parent. Participants with stepchildren gave more autonomy to stepparents in the information and consent cases than those without stepchildren.

Conclusion
Based on these data, Belgian and Dutch adults are in favor of a relevant role for stepparents in a medical context, despite the fact that the legal framework does not formally allow or support this. However, when there is disagreement between the biological parent and stepparent, the participants leave authority with the former. Future research will have to show to what extent stepparents could be included in medical information and decision making regarding stepchildren.

P 52.

Haematological Alterations in an Adolescent Female Population Cohort with Anorexia Nervosa of the Restrictive Type

M. Docx, M. van den Akker

ZNA Koningin Paola Kinderziekenhuis, UZA, ZNA Middelheim Algemeen Ziekenhuis

Aim
To investigate haematological abnormalities, especially leukopenia (< 4.0 x 10 9 E/L) and compare it with clinical , biochemical and endocrinological risk factors.

Methods
This retrospective cohort study was conducted on 311 anorexia nervosa adolescent females of the restrictive type. These patients with a mean age of 14.6 ± 1.8 years and mean BMI of 15.4 ± 1.8 kg/m² were studied on admission in the acute, restrictive phase of the disease and nine months later after refeeding with the same structural protocol.

Results
Of a population of 311 restrictive anorexic adolescent females, 42 (13.5%) were diagnosed with leukopenia (< 4.0 x 10 9 E/L) and 4 (1.3%) with thrombocytopenia (< 130 .10 E 9/L)

The two major risk factors to develop leukopenia (< 4.0 x 10 9 E/L) with mild leukopenia (1.51 ± 0.46 x 10 9 E/L) were a low BMI mean (14.4 ± 1.4 kg/m²) and serious weight loss mean (25.2 ± 8.2 %). Three minor risk factors are a lower reticulocyte count (32. ± 15.5 %), a low IGF-1 (118.7 ± 67.1 ng/ml) and an increased serum ferritin (136.8 ± 88.1 ng/ml).

Conclusion:
The Leukopenia as well as all haematological alternation ameliorate disappear finally after refeeding.

P 53.

Scrotal Borrelial lymphocytomaScrotal Borrelial lymphocytoma

S. Verbeeck, K. Casteels, L. Sevenants, M. Depypere

UZ Leuven

Interesting case for a poster presentation because of the typical clinical pictures.

A 5-year-old boy was referred to the pediatric clinic with progressive swelling and red discoloration of the left scrotum since two months. There were no associated symptoms and no fever, local pain or pruritus. Past history was negative with no recent infections, no insect bites or other dermatological problems.

On inspection we see a sharply demarcated nodule with a diameter of 2cm.

Laboratory test results show low infectious parameters and positive Borrelia IgG using immunoblot analysis. Diagnosis of Borellial lymphocytoma was made and therapy with oral amoxicillin (50mg/kg) was initiated for 4 weeks. After 4 weeks, the swelling and redness had improved and after 6 weeks it had disappeared completely.

Borrelial lymphcytoma is typically located on the outer ear but it is also important to consider this diagnosis in other locations.

P 54.

Congenital diaphragmatic hernia in an infant: Don't miss the diagnosis!

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Background

Late-presenting Congenital Diaphragmatic Hernia (CDH) is a rare entity. Diagnosis and management remain a challenge. We hereby report a case of a two-month-old infant in whom CDH was diagnosed.

Method and results

A 2 months old infant was admitted to the hospital for failure to thrive. She complained of occasional vomiting and feeding difficulties. Pregnancy was complicated by short long bones (3rd percentile), hydramnios and large muscular ventricular septum defect. Maternal medical history was marked by neurodevelopmental retardation and small size of unknown etiology. Birth was uneventful. However, the baby was admitted to the neonatal intensive care unit for social reasons. Physical exam was normal except of dysmorphic features. Thus, a micro-array CGH was performed and came back normal. The newborn was discharged with organized follow-up.
With 2 months the baby developed respiratory distress additionally to the gastro-intestinal discomfort. Auscultation revealed decreased air entry on the left side of the chest. Chest Xray showed a left posterior-lateral diaphragmatic hernia. Surgical repair was performed. Post-operative outcome was favorable after with improvement of gastro-intestinal symptoms and satisfactory weight gain.

In the classic form of CDH the diagnosis is often made in prenatally or at birth. The typical presentation consists of a classic symptomatic triad (respiratory distress, deviation of heart sounds and flat abdomen). The estimated overall prevalence is 1 case per 2500 to 3500 live births among whom 5-30 % are late-presenting CDH. Physiopathology and mechanism of late-presenting CDH remain unknown. Commonly, diagnosis is suspected in case of acute respiratory distress or gastrointestinal symptoms (GI) such as volvulus. But sometimes the manifestation is non specific, such as important gastro-intestinal reflux, vomiting or recurrent pneumonia. Surgical repair with or without patch closure of the diaphragm is indicated.

Conclusion

This case illustrates a late-presenting CDH with gastrointestinal symptoms and failure to thrive. Late-presenting CDH is a rare and often misdiagnosed condition. Physicians should suspect it when GI and/or respiratory symptoms do not respond to usual management.

P 55.

Cytolysis associated to haematological abnormalities in a neonate with a moderate form of CovId-19 infection: a case report

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HUDERF

Clinical and biological features of neonatal Sars-cov2 disease seem also different from children and adults. We report a neonatal case with biological disturbances detected at the beginning of his hospitalization. A 4-week-old neonate presented with vomitis and decreased feeding and loss of 8% of her weight without fever since 1 week. She was the first child of non-consanguineous parents and was born by caesarean section at 41 weeks of gestation. Her mother reported that she was recently detected positive for covid-19. At admission, she did not show any clinical signs of dehydration or respiratory abnormalities or neurological symptoms. Her body temperature was 36.5°C and O2 saturation was 99%.

The neonate was tested by RT-PCR on pharyngeal swab specimen and found positive.

Laboratory abnormalities evidenced a neutropenia (neutrophil count : 400/µL), a thrombocytopenia (139x 103/µL), very high levels of aspartate amino transferase and alanine amino transferase enzymes (1478 ui/L,733 ui/L) and mild cholestasis (gamma GT 205 ui/L) with negative hepatitis serologies .Coagulations parameters were normal which allow us to exclude hepatocellular insufficiency. She was isolated in our infant unit. A supportive and therapeutic care began. A biological follow up was performed at day 1,day 5, day 15 respectively. An hyperferritinemia (7751 µg/l) were detected at day 1 but the CRP was normal. The absence of fever, splenomegaly on ultrasound as well as normal fibrinogen and normal triglycerides levels were not in favor of macrophage activation syndrome. Digestive symptoms resolved at day 3 and the oral feeding were then started. At day 5, neutrophil and platelets counts were normal and a significant drop of liver enzymes and ferritin was noticed. Given the very good clinical condition of the patient, the weight gain and the decrease of biological disturbances, she returns home. Two weeks later, she was asymptomatic and all abnormal blood tests had returned to a normal range. In conclusion this neonatal case highlights some interesting features. It’s the first case with in the same time detection of a moderate neutropenia and very high levels of liver enzymes; a thrombocytopenia appeared shortly in the course of the ill; a spontaneous normalization of liver enzymes appeared in 15 days .It remains unclear why these biological disturbances appear only in some neonates and not in all. Once more the prognosis of these biological abnormalities stay again unknown.

P 56.

The knowledge of child maltreatment among general practitioners and pediatricians in training: a survey

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Background

Child maltreatment is a common, underreported, worldwide problem. Although the variation of clinical signs and mimickers of child maltreatment are well described in the medical literature, the knowledge about it in clinical practice is less investigated. Given that health care providers play an important role in addressing child maltreatment, this study examined the knowledge on clinical signs of child maltreatment of Belgian general practitioners and pediatricians in training.

Methods

Participants filled in an anonymous, online survey with demographic characteristics and 15 fictional, but realistic cases with either suspicious or non-suspicious signs, presenting in primary care or emergency setting. The overall score, the number of correct answers per case and the median score on the suspicious and the not suspicious cases for child maltreatment were calculated. In addition, the influence of the participant's demographic characteristics on their score was examined.

Results

A total of 70 participants completed the survey. The overall median score was 73.3% (interquartile range, IQR: 20.0%). The suspicious cases were better solved than the non-suspicious cases (median: 85.7% [IQR: 28.6%] versus 62.5% [IQR: 25.0%], p-value <0.001). Type and level of education did not show any significant correlation with the performance on the survey.

Conclusion

The knowledge of the clinical signs of child maltreatment among pediatricians and general practitioners in training is good, but there still is room for improvement. Particular attention should be paid to the mimickers of child maltreatment, as these were less adequately recognized and may lead to false accusations.

P 57.

A broad survey to determine paediatric clinical trial requirements within BelgiumA broad survey to determine paediatric clinical trial requirements within Belgium

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U Gent, UZ Gent, UZ Leuven, HUDERF, UCLouvain, CHU Charleroi, University of Liverpool, AZ Roeselare

Background/Aims
Due to the Paediatric Regulation in 2007, the number of paediatric clinical trials within Europe has substantially increased. Consequently, potential sites for paediatric clinical trials were overrun by trial opportunities, infrastructure pressure and their limited experience. The number of unsuccessful clinical trials in children remained high. Research networks to support sites and consider national adaptations has been proven to increase efficiency and durability. A core example is the pan-European network conect4children (c4c) funded by the Innovative Medicines Initiative (IMI2). The c4c Belgian Clinical trial network is located at the Ghent University (Hospital). The National Network includes both large and smaller sites with limited research experience, requiring intensive facilitations, training and recruitment facility support. To date, little is known about the effect of networks on sites and specific innovation needs a research site has.

Method
A questionnaire was conducted within the c4c Belgian Clinical trial network over a period of 3 months. An inquiry was made into the role of a national hub and other topics for support. Due to multiple responses per site, answers were grouped per site.

Results
Of the 15 connected sites, we received 32 responses coming from 13 unique sites. Surveyor's background included sub-specialised paediatricians and clinical research coordinators. Within the Belgian network, around 4 (30%) of sites do not have a paediatric clinical trial unit to support the trial. All sites agreed or strongly agreed that a national hub is useful for the site to conduct clinical trials. The majority namely 11 (84%) of sites identified human resources as a core improvement need for sites, specifically finding dedicated clinical research nurses and finding time for principal investigators (PI) to perform study tasks aside from clinical work. A strong need for financing of infrastructure from 10 (76%) of sites is acknowledged for consultation areas, imaging, and a biobank structure. On the contrary, 12 (92%) of sites did not regard study-specific courses as a priority.

Conclusion
In order to foster sustainable development of new medicines in paediatric diseases, site need's must to be taken into account and prioritized. The primary need is in human resources and financing of infrastructure. Further investigation into how a centralized organization can support is needed.

P 58.

Ethical theories as Practical Tools for Clinical Practice

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HUDERF

Background

Philosophy and ethical theories in particular offer words to name the daily experience of care. Thus, the main work of a clinical ethicist is to design a consistent ethical gallery through which care givers can walk and analyse each situation under different ethical perspectives without freezing the dynamism of their reflections and their practice or erasing the specificity of each patient.

Yet, perinatal palliative care imposes a frame where our core concepts and values are confronted to their limits. The care and the person at its centre urge us to deploy an ethical dialogue preserving us from their oversight. From this frame, for five years now, on the basis of an active participation in the reality of the field of care, we have been building the architecture of an ethical gallery that we want to be able to respond to the challenges of care, from the bedside to these institutional and systemic scales.

Methods

Three theories caught our attention. They all have the same singular specificity. The field of care has appropriated them at different scales precisely to name and express its daily challenges, realities and experiences.

The first one is narrativity presenting a dialogue between Paul Ricoeur's philosophical narrativity and Rita Charon's narrative medicine. The second one is responsibility presenting a dialogue between Emmanuel Levinas and its appropriation by a nurse's perspective through the reading of Per Nortvedt. And the third one, asked directly by the field of care in which we were evolving, is the ethics of care from Carol Gilligan and Joan Tronto.

Results

Those three theories are involving an internal dialogue with the practice of care and questioning each other to weave an applied ethical framework from clinical ethics consultation to the development of an institutional ethical framework.

From this practice we designed the integration of other concepts: Sense and Share, to link the whole together, give it new depth and new perspectives. This ethical framework ensures and maintains a close connection and the preservation of both the care and the person at its centre.

Conclusion

From professionals to the relatives and the patients themselves, there is a need, a purpose and a duty to ensure and maintain humanity at the core of care. Ethical perspectives through their ongoing and dynamic dialogues draw bridges from theory to practice, frfrom theory to practise, from the bedside to society.

P 59.

Development of a questionnaire to assess the knowledge of midwives and pediatric nurses on maternal use of analgesics during lactation.Development of a questionnaire to assess the knowledge of midwives and pediatric nurses on maternal use of analgesics during lactation.

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KU Leuven, Erasmus MC UMC Rotterdam

Background
There is a need to assess the knowledge of healthcare providers on the use of maternal analgesics during lactation, while valid instruments are not yet available. This study aimed to develop and test a valid questionnaire on the knowledge of analgesics (acetaminophen, ibuprofen, aspirin, tramadol, codeine, oxycodone) during lactation, using a structured, stepwise approach.

Methods
Stepwise, structured approach, using literature screening, Delphi rounds, and subsequent pilot testing.

Results
Literature was screened to generate a preliminary version consisting of a pool of item subgroups. This preliminary version was subsequently reviewed during two focus groups [midwives (n=4), pediatric nurses (n=6)], followed by a two-round online Delphi with experts (n=7) to confirm item and scale content validity. This resulted in an instrument consisting of 33 questions, and 5 specific clinical case descriptions for both disciplines. Based on the assumption of an a priori difference in knowledge between midwives and pediatric nurses related to their curricula (known-groups validity), high construct validity was demonstrated in a pilot survey (midwives: n=86, pediatric nurses: n=73). These questionnaire are available upon request in English and Dutch for teaching purposes (karel.allegaert@uzleuven.be).

Conclusions
A valid instrument to assess knowledge on lactation-related exposure to analgesics was generated, which could be further validated and used for research and educational purposes. As these pilot findings suggest suboptimal knowledge for both professions on this topic, adaptations to their curricula and postgraduate training might be warranted.

Long Oral Presentation

LO 1.

Fecal amine metabolites analysis before onset of severe necrotizing enterocolitis in preterm infants: a prospective case-control study

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Rationale
Necrotizing enterocolitis (NEC) is characterized by a change in host and microbiota metabolism. The potential of specific metabolomics, i.e. amino acids and amino alcohols (AAA), as early diagnostic biomarkers for NEC is largely unexplored.

Methods
In this multicenter prospective case-control study, longitudinal fecal samples from preterm infants (born before 30 weeks of gestation) developing severe NEC (Bell's stage IIIA/IIIB) 1-3 days before diagnosis were analyzed by targeted high-performance liquid chromatography (HPLC) and compared to samples from gestational and postnatal age-matched controls.

Results
Thirty-one NEC cases (15 NEC IIIA;16 NEC IIIB) with 1:1 matched controls were included. Preclinical samples of infants with NEC were characterized by five increased essential amino acids – isoleucine, leucine, methionine, phenylalanine and valine. Lysine and ethanolamine ratios were lower prior to NEC, compared to control samples. A multivariate model was rendered based on isoleucine, lysine, ethanolamine, tryptophan and ornithine, modestly discriminating cases from controls (AUC 0.67; $p < 0.001$).

Conclusion
Targeted HPLC pointed to several specific AAA alterations in samples collected 1-3 days before NEC onset, compared to controls. Whether this reflects metabolic alterations and has a role in early biomarker development for NEC, has yet to be elucidated.

SO 25.

Analgosedation before less invasive surfactant administration: a systematic review

S. Tribolet, N. Hennuy, D. Snyers, V. Rigo

CHU de Liège

Background

Surfactant therapy is the cornerstone of the management of respiratory distress syndrome. Alternatives to endotracheal intubation for surfactant administration currently include “less invasive surfactant administration”. Its effectiveness was demonstrated by meta-analyses and guidelines now recommend it as the optimal method of surfactant administration in spontaneously breathing babies. While it still requires a direct laryngoscopy, the issue of sedation and analgesia during the procedure remains controversial as 52% of European neonatologists do not use any.

Methods

Medline via Ovid, Embase, Scopus and Cochrane Library of Trials were searched for studies of LISA after sedation without any filters or limits independently by two reviewers. Risk of bias (RoB) and quality assessment were evaluated using the RoB2 for RCT or the Newcastle Ottawa Scales (NOS) for cohort studies.

Results

We included eight studies: one RCT, two prospective, three retrospective and two RCT comparing INSURE and LISA after sedation (LISA arms assessed as prospective cohorts), for a total of 908 newborns. Failure, defined as need for intubation or for a second dose of surfactant was no different between sedated and unsedated groups. Infant pain was significantly reduced, with more infants evaluated as comfortable. LISA with sedations led to higher occurrences of intraprocedural desaturation and need for positive pressure ventilation, but need for mechanical ventilation within 24 or 72 h of life was not significantly different. Clinical tolerance and complications (hypotension, mortality, air leaks, BPD...) were similar. Procedural conditions were evaluated as good or excellent in 83% after sedation.

Discussion and conclusion

This systematic review highlighted that analgesia or sedative drugs increase infant comfort and allow good procedural conditions, with a limited impact on the clinical evolution. Many questions remain about best choise of drug and dosage, with the constraint to maintain spontaneous breathing and have a rapid offset. Further good quality studies are needed to provide additional evidence to supplement those limited existing data, given how deleterious awake laryngoscopy can be.

Prospero registration: CRD42020205365.

Short Oral Presentation

SO 26.

QTc intervals are prolonged in (near)term neonates during therapeutic hypothermia

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Introduction
Therapeutic hypothermia (TH, 33-34°C) is standard treatment for neonates (≥ 36 weeks) with perinatal asphyxia (PA) and moderate-to-severe hypoxic-ischemic encephalopathy, but the outcome still needs improvement (add-on interventions). To facilitate drug development, neonatal hypothermia physiology-based PK (PBPK) framework would enable precision dosing. Related to this, reported (patho)physiological observations should be integrated.

Methods
A structured search and literature assessment (PubMed, Embase, Google Scholar) with 'Newborn/Infant, QT and hypothermia' was conducted (October 2021), and retrieved individual values were converted to QTc (Bazett) over postnatal age (day 1-7).

Results
We retrieved 94 QTc observations during HT (n=50, until day 3) and subsequent normothermia (n=44, day 4-7) in 33 neonates form 6 publications. The median (range) of the QTc during TH was 508 (430-678) and 410 (317-540) msec afterwards (difference 98 msec, or +28 msec/°C decrease). Four additional cohorts (but without individual QTc observations) confirmed the magnitude of the effect of body temperature on the QTc time interval.

Conclusions
The retrieved information consistently reports a proportional (temperature mediated) significant prolongation of QTc during HT in newborns. This is of specific relevance for drug development, as QTc prolongation is a commonly explored safety marker. QTc patterns and trends needs specific reflection in the context of any neonatal drug development, even more in the HT setting.

Research supported by the iPREDICT (FWO GOD0520N) grant.

SO 27.

Ventilation and Respiratory Outcome in Extreme Preterm Infants: Trends in the new Millennium

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Objective
To determine changes in respiratory care and disease frequency in extremely premature infants, at a tertiary neonatal intensive care unit in Belgium

Design
A retrospective study in extremely preterm babies born between 24+0 and 27+6 weeks of gestational age (GA) in 2000-2001 (Epoch 1), 2009-2010 (Epoch 2) and 2018-2019 (Epoch 3).

Outcome measures
Incidence of BPD (diagnosed according to three different criteria) or death. Secondary outcomes included the usage of different ventilation modes, changes in pharmacotherapy and the incidence of significant extra-pulmonary morbidities.

Results
184 neonates were included of whom 151 survived until 36 weeks of corrected GA (cGA). Oxygen- or positive pressure dependence increased over time (26.1%, 41.7% and 56.1% respectively), with higher adjusted odds in Epoch 3 for the composite outcome 'BPD or death' (aOR 2.55 [95%CI 1.19 – 5.61]). Severity-based definitions show increasing trends in survivors only. Time spent on invasive mechanical ventilation was similar throughout the years but the use of non-invasive ventilation significantly increased in Epoch 3 (32.0 [95%CI 25.0 – 37.0] vs 27.0 [95%CI 26.0 – 32.0] vs 53.0 [95%CI 46.0 – 58.0] days). Moreover, mortality-adjusted rates of severe IVH, NEC or intestinal perforation and multiple sepsis tended to decrease.

Conclusions
In spite of significant clinical advancements and adherence to novel treatment guidelines in our neonatal intensive care unit, the incidence of BPD is increasing over time. Lung function assessment in survivors of BPD at school age needs to be performed to assess long-term consequences.

P 47.

Invasive candidiasis and candidemia in neonates: about a case.

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Background
Invasive candidiasis and candidemia (IC/C) is the most frequent neonatal invasive fungal infection but data in this population remain limited. This affection has a high morbidity and mortality rate. Here are the characteristics and recent guidelines available about neonatal IC/C.

Case report
A 6-day-old infant, born spontaneously at 26 weeks of gestational age, suddenly presented with fever, increased apnea and bradycardia, hyperglycemia and clinical instability. Candida albicans was found in the blood culture and in the urine by PCR. The risk factors identified were prematurity, previous antibiotic exposure for a suspected early onset sepsis, ventilatory support, central venous catheter (CVC) and parenteral nutrition. CVC replacement and antifungal therapy with fluconazole for 14 days allowed for recovery.

Discussion
Clinical signs of IC/C such as lethargy, persistent fever, bleeding manifestations, feed intolerance, pneumonia or apnea are not specific and fake a bacterial sepsis.

Risk factors (prematurity, premature rupture of membranes with vaginal candidiasis, neutropenia, antibiotic or systemic steroids exposure, central venous catheter, parenteral nutrition, ventilatory support, previous surgical procedures, prolonged hospital stay) enhance suspicion of IC/C.

Laboratory results can also help showing mostly leukocytosis and thrombocytopenia. Urine analysis, lumbar puncture and retinal examination should be performed. In case of persistent positive blood cultures, head, heart and kidneys ultrasonography are strongly recommended.

Recommendations for the treatment of neonates with IC/C promote the use of liposomal amphotericin B (AmB) and deoxycholate AmB, with no significant signs of nephrotoxicity. Fluconazole is useful in case of no previous exposure to azoles to avoid resistance. Echinocandins are gradually more used in the neonatal population, with good tolerance.

CVC should be removed or replaced as soon as possible to avoid dissemination.

Although experts agree that the minimum treatment duration to achieve blood culture sterilization is at least 14 days, it is also important to consider the clinical evaluation of deep tissue infection and resolution of candidemia symptoms.

Conclusion
Studies underline the importance of early and structured approach in the management of IC/C in addition to careful monitoring to mitigate the morbidity and mortality of this condition in the neonatal population.

P 48.

Non-concordant congenital Cytomegalovirus infection in diamniotic dichorionic extremely premature twins: a case report.

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ULB, HUDERF

Cytomegalovirus (CMV) is the most common cause of congenital infection, affecting multiple gestations with a higher incidence than singletons. Although its hereditability has been estimated at 94%, non-concordant congenital infection in twins is not a rare phenomenon, most frequently detected in dizygotic gestations. Furthermore, chorionicity seems to play an important role in CMV transmission. Notwithstanding, cases of late prenatal horizontal transmission from the affected co-twin have been reported and associated with twin placental fusion.

We report the case of a Diamniotic-Dichorionic (DC/DA) gestation of a 20-year-old primiparous woman. Maternal CMV serologies at 8 weeks of gestation showed negative anti-CMV-IgM and positive anti-CMV-IgG. Ultrasound evaluations showed an intrauterine growth restriction of twin A, and no morphologic abnormalities in both male fetuses. An emergent Caesarean section was performed in the 29th week of gestation due to premature rupture of membranes and arm prolapse of twin A. The birth weight of the twin A was 810 g (7th percentile) and Apgar score was 1,1 and 5 at one, five and ten minutes of life respectively. Routine urine culture at day 14 was positive for CMV, and congenital infection was confirmed by retrospective analyses of the Newborn Metabolic Screening sample collected at day 0. A CMV viremia of 35014 UI/mL was detected at day 16 and Valganciclovir was administered from day 17. He presented no signs of classic CMV inclusion disease at birth, but a primary neurophenotype was concluded (microcephaly and symmetric foetal growth restriction). Fundus oculis showed no signs of retinitis, cranial ultrasonography showed a periventricular haemorrhage of first grade, and a cranial magnetic resonance confirmed abnormalities of the periventricular white matter. Auditory brainstem response tests revealed severe central alterations. The birth weight of the twin B was 1200 g (46th percentile) and Apgar score was 3,5 and 7 at one, five and ten minutes of life respectively. Urine cultures were negative for CMV and the infant was completely asymptomatic.

We describe a non-concordant congenital CMV infection in a DC/DA gestation due to a recurrent asymptomatic CMV infection, leading to two very different outcomes in the postnatal period. Due to the extreme preterm resolution of the gestation, prenatal horizontal acquisition of a CMV by the twin B might have occurred if the gestation term would have happened later.

Posters

P 49.

Perinatal hydrometrocolpos, unique diagnosis but different antenatal and postnatal clinical pictures

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CHC Montlégia, UCL

We report 2 cases of prenatal diagnosis of hydrometrocolpos and fetal ascites.

Both our patients had similar ultrasound scans and fetal MRI, which highly suspected a dilated uterus, bladder, and ascites secondary to urogenital malformations. Both amniocenteses came back negative for genetic anomalies. Cases like these are rare since prenatal diagnostic tools have largely progressed in the last decades, and termination of pregnancy can be offered. Both families decided to go through with the pregnancy and it has enabled us an extensive review of the care necessary for these complex cases.

Patient 1 developed severe bilateral pulmonary hypoplasia secondary to the voluminous ascites. A peritoneo-amniotic shunt was placed at 30 weeks gestational age (GA) and was temporarily effective in diminishing the volume of ascites. Unfortunately, it was not sufficient to promote significant lung growth; the child was born at 33 weeks GA by elective caesarean section. At birth, a supra-pubic vesical catheter was inserted and the patient was intubated. Adequate ventilation was only possible by high frequency oscillatory ventilation (HFOV) with elevated pressures. Postnatal clinical examination and imaging showed persistent urogenital sinus. Lung hypoplasia was prominent and led to the infant's death on day 8.

Patient 2 had less severe intrauterine ascites, good lung volumes but abdominal calcifications with a high suspicion of meconium peritonitis. She was delivered by emergency caesarean section due to maternal distress with amniotic fluid embolism at 34 weeks GA. Oesophageal atresia was diagnosed at birth and was surgically cured on her first day of life. She demonstrated bilateral uretero-hydronephrosis and a persistent urogenital sinus with an imperforated hymeneal membrane. High suspicions of intestinal atresias and multiple adherences have prevented any enteral nutrition.

The spectrum of uro rectal septum malformations is vast and isn't very well known in many centers. They can be isolated or be included in a multitude of defects in one same patient. They currently do not have a known genetic origin but are instead thought to be due to embryological defects. The separation between the 3 different tracts occurs between the 6th and the 12th weeks of gestational age and a series of factors have been identified to increase the susceptibility of organogenesis defects. Cases like these are difficult on many levels but remain filled with educational value.

P 50.

A Neonatal Case with a Prenatal Diagnosis of Capillary Malformation-arteriovenous Malformation Syndrome (CM-AVM)

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ULB

Introduction

Capillary malformation arteriovenous malformation syndrome (CV-AVM) is marked by the appearance of vascular lesions on the skin, frequently found on the face and limbs. The main genetic defect in this syndrome is a mutation in the RASA-1 gene transmitted in an autosomal dominant manner. Besides, this syndrome is characterized by a wide intra- and interfamilial variability in clinical manifestations. In fact, little is known about prenatally diagnosed cases of CV-AVM.

Case presentation

Here, we report the case of a 25 year-old woman admitted during her third pregnancy for management of unexplained polyhydramnios. Prenatal genetic analysis (Mendeliom sequencing from amniotic fluid), performed due to two previous unexplained fetal losses, showed a mutation in a heterozygous state in RASA-1 gene c.2367 C>T (p.Arg789*). Hence, an antenatal diagnosis of CV AVM was then made. An emergency cesarean section was performed at 31 5/7 weeks due to complications of the second amnioreduction. At birth, the neonate presented five erythematous lesions with diameters of 1-3 cm, similarly to those found on the father. No signs of arteriovenous malformations was detected on MRI of the brain and spine. No other anomaly has been found.

Conclusion

Despite its autosomal mode of transmission, CV-AVM is still underrecognized due to its variable clinical presentation. Our report underlines that CM-AVM should be considered at the first clinical examination of a neonate revealing atypical capillary malformations especially in the context of unexplained polyhydramnios, previous fetal losses, or family history of clinical presentation of CM. When genetic diagnose is made a follow-up should be organized during the period of early childhood.

P 51.

Bilateral congenital chylothorax with generalized lymphoedema in a newborn with a PIEZO 1 mutation: a case report

S. Denes, D. Snyers, N. Cajgfinger, S. Bulk, A. Lumaka Zola, V. Rigo, S. Tribolet

U Liège, CHU

Introduction

Congenital chylothorax (CCT) is the most common form of pleural effusion in newborn with an incidence of 1 per 10 000 to 24 000 lives births. Neonatal chylothorax can be congenital or acquired, most commonly as a complication of thoracic surgery. Congenital chylothorax can occur isolated, in association with certain genetic syndromes or other lymphatic developmental anomalies.

Case report

A 35-week-old-boy was admitted in our NICU after a pregnancy marked by severe polyhydramnios requiring amniocentesis, prefrontal oedema and bilateral pleural effusion. At birth, he breathed spontaneously but required a non-invasive respiratory support for respiratory distress. Ultrasonography confirmed the presence of bilateral pleural effusion. Lymphocytic predominance and high triglyceride level in the puncture fluid led to the diagnosis of congenital chylothorax. Despite fasting and exclusive parenteral nutrition, we noted respiratory deterioration, requiring continuous chest drainage from D10 of life. Octreotide injections were administrated for 10 days, without improvement. In parallel, a generalized lymphoedema appeared. Lymphography-MRI didn't identify lymphangiomatous malformations, nevertheless there was no passage of contrast product from the subdiaphragmatic to the supradiaphragmatic level. As the classical genetic analysis (CGH-array) was non-contributory, a whole genome sequencing was conducted and highlighted 3 mutations (loss of function) in the PIEZO 1 gene.

Octreotide and diuretics were resumed at the age of 3 months for a period of 3 months. This was associated with a very slow improvement allowing the removal of chest tube at 5 months of life. Gradual refeeding was initiated with low-fat milk rich in medium-chain triglycerides (LIPISTART). A non-invasive respiratory support at sleep is still required at 9 months, due to persistent bilateral effusion.

Discussion and conclusion

Congenital chylothorax is a rare condition with high mortality and morbidity rates. If associated with generalised lymphoedema, hereditary lymphatic dysplasia should be evoked and a whole genome sequencing should be conducted to allow for genetic diagnosis and prognosis counselling. Our case highlighted a slow favourable evolution following long term octreotide, diuretics and dietetic therapy.

LO 2.

Ciliary dyskinesia is present in patients with cystic fibrosis, and would be present from childhood

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Background

Mucociliary clearance (MCC) results from an effective interaction between the mucus layer and the normal coordinated ciliary beating. In cystic fibrosis (CF), MCC is impaired due to dehydrated mucus, but ciliary function is poorly studied. Recent data suggested that abnormal ciliary beating may contribute to poor MCC in CF patients. However, results are controversial: ciliary beat frequency (CBF) might be either decreased or increased, compared with healthy subjects. We aimed to evaluate if ciliary function may have a role in impaired mucociliary clearance in CF patients and if this ciliary dyskinesia was present in childhood.

Methods

Ciliated epithelial samples were obtained by nasal brushing from 22 paediatric (0-17 years) and 15 adult CF patients (18-78 years). Beating cilia were recorded using digital high-speed videomicroscopy at 37°C. Ciliary functional analysis (CFA) was assessed by CBF and the percentage of abnormal ciliary beat pattern (CBP) and was compared with 14 healthy subjects (22-54 years).

Results

There was a significant difference in CBF between adult CF patients and healthy subjects (12.6±2.0Hz vs 14.8±2.2Hz; p = 0.008), but not between paediatric CF patients and healthy subjects (13.9±2.7Hz vs 14.8±2.2Hz; p = 0.300). However, the percentage of abnormal CBP was significantly higher in adult and paediatric CF patients (33±2% (p = 0.010) and 26.2±15.1% (p = 0.049), respectively) compared with healthy subjects (17.3±7.6%).

Conclusion

Our pilot study suggested that an abnormal ciliary function, present in childhood, might contribute to impaired MCC in CF patients, as shown by the higher percentage of abnormal CBP compared with healthy subjects. To evaluate if ciliary dyskinesia is primary or secondary to chronic inflammation, CFA should be repeated after air-liquid interface cell culture.

Long Oral Presentation

LO 3.

Assessing the use of antibiotics and the burden of varicella in Belgium using a retrospective GP database analysis

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MSD Belgium, Real World evidence, IQVIA, Center for observational and real-world evidence, Merck & Co., Inc, MSD Spain

Background/Aims
Varicella is a highly contagious infection that typically occurs in childhood. While most cases have a generally benign outcome, infection results in a considerable healthcare burden and serious complications may occur.

The objective of this study was to characterize the burden of varicella in a real-world primary care setting in Belgium, including the rate of varicella-related complications, medication management and general practitioner (GP) visits.

Methods
The study was a retrospective observational study using data from a longitudinal patient database in a primary care setting in Belgium. Patients with a GP visit and a varicella diagnosis between January 2016 and June 2019 were eligible and data one month prior and three months after the diagnosis were included. Outcomes included varicella-related complications, antibiotic use, antiviral use, and GP follow-up visits. Antibiotic use could be specified by class of antibiotic and linked to a diagnosis. Complications were identified based on concomitant diagnosis with varicella during the study period.

Results
3,847 patients with diagnosis of varicella were included, with a mean age of 8.4 years and a comparable distribution of gender. 12.6% of patients with varicella had a concomitant diagnosis of a varicella-related complication. During the follow-up period, 27.3% of patients with varicella were prescribed antibiotics, either systemic (19.8%) and/or topical (10.3%). The highest rate of antibiotic prescriptions was observed in patients with complications (63.5%) and in patients younger than one year (41.8%). Nevertheless, 5.3% of the patients were prescribed antibiotics without a concomitant diagnosis of another infection. The most commonly prescribed systemic antibiotics were amoxicillin alone or combined with beta-lactamase inhibitor, and thiamphenicol. Fusidic acid and tobramycin were the most prescribed topical antibiotics. Antivirals were prescribed for 2.7% of the study population. 4.7% of the patients needed a follow-up visit with their GP.

Conclusions
This study reports a substantial burden of varicella in a primary care setting in Belgium, with high rates of complications and antibiotic use.

SO 28.

COVID IN THE RESIDENTIAL REHABILITATION CENTER ZEEPREVENTORIUM

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Zeepreventorium

Introduction

Zeepreventorium is a residential center for children aged 0-18 years with various chronic diseases. Up to 160 patients reside in groups of 10, mainly according to age. Throughout the COVID epidemic, the centre remained active. Besides hand hygiene, social distancing, wearing masks, and temperature control, additional measures were taken such as PCR-testing on admission, no contact between groups, restricted return home during the epidemic peak, screening for high risk contacts and symptoms before and after weekends, PCR and later antigen test upon suggestive symptoms plus isolation until results. Vaccination of 12-18 year olds started July 20th 2021.

Aim

to report the COVID PCR-test results and patients' symptoms in our cohort during the COVID19 pandemic.

Methods

retrospective collection of PCR test results and symptoms between March 2020 and September 2021.

Results

during the studied period, 450 children resided at the centre. 936 PCR-tests were done of which 36 (3.8%) were positive in 28 patients (14 boys). Of these, 4 patients were positive on admission.

Mean age ± SD of positive patients was 15.5 ± 2.2 years (y). We never documented infection in the 0-3 and 4-6 age groups, the youngest positive patient being 9.7y. All but 2 infections occurred in patients with severe obesity. We never had suspicion of cross-infection from patient to personnel or vice versa, nor between subjects residing in different groups. Within group patient to patient transmission occurred on 5 instances in 3 different groups: in the eldest age group an 18y old boy most likely infected another boy, later a 17y old girl infected 1 peer; a 15y old boy infected 2 others and 4 months later a 15 y old girl infected another girl; a 13y-boy infected 5 peers. Besides the 4 patients positive on admission, 9/28 positive patients remained isolated cases.

A 17 year old boy had severe dyspnea lasting one day; all other youngsters were either asymptomatic or had mild flu like symptoms.

Conclusion

in the residential rehabilitation center Zeepreventorium, only 3.8% of 936 PCR-tests done returned positive. Although patient to patient transmission occurred on 5 occasions, half of the positive tests concerned isolated cases. Only 1 of 28 infected patients developed short lived respiratory distress. Hygienic and segregation measures plus careful monitoring were helpful to safely continue our mission of rehabilitation of children with chronic disease during the COVID pandemic.

Short Oral Presentation

SO 29.

Application of the KAPAUSA protocol : severe respiratory distress at the pediatric emergency department

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CHU Saint Pierre

Background/Aims

Studies have shown the efficacy of non invasive respiratory support (NIRS) (High Flow Nasal Cannula and Continuous Positive Airway Pressure) especially in bronchiolitis. NIRS seems to reduce the need for invasive respiratory support, thus lowering costs, with clinical advantages and fewer adverse effects. But it isn't clear when and at which conditions NIRS should be started and stopped. We established the KAPAUSA score based on clinical observation (respiratory rate, work of breathing and general condition, in function of age), applicable by medical and paramedical crew. Based on this score and the eventual presence of hypercapnia, the child was treated following KAPAUSA protocol. Primary outcome: identify those children who don't need NIRS, those who could benefit from NIRS and those who should be transferred to an intensive care unit (ICU). Secondary outcomes included the impact of standardization of care on the length of stay (LOS), duration of NIRS, rates of transfer to an ICU and adverse events. We also questioned the feasibility

Methods

A prospective observational study was conducted 01/10/2017 until 15/05/2018, including children under 6 years of age in respiratory distress prone to needing NIRS. We collected demographic data, cardiorespiratory and general parameters, duration of NIRS, LOS and clinical evolution. Due to low number of inclusion, a retrospective collection of data for patients who had benefit from NIRS was added, creating a prospective group where the Kapausa score was realized before starting NIRS, and a restrospective group, where the score was calculated after starting NIRS, based on the informations in their medical file. An anonymous evaluation was filled in afterwards by the (para)medical team

Results

55 patients aged from 1 week to 23 months benefited from NIRS. 6 patients were excluded for insufficient data. No adverse events were observed. Children in the prospective group (n:30) received NIRS earlier and for a shorter period, and stayed 1 day less hospitalized compared to children in the restrospective group (n:19). In the latter, the conditions applied were higher than prescribed by protocol. 88% of the (para)medical group found this protocol helpful in the approach and treatment of the child with respiratory distress

Conclusion

The instauration of a protocol is always beneficial. We saw a trend towards shorter NIRS and LOS and lesser transfers to ICU. A study with simplified protocol will be conducted

SO 30.

Ciliary videomicroscopy at room temperature lacks sensitivity for PCD diagnosis

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Background
Primary ciliary dyskinesia (PCD) is an heterogenic inherited ciliopathy in which respiratory cilia are stationary or dyskinetic. Digital high-speed videomicroscopy (DHSV) is highly sensitive and specific for PCD diagnosis but lacks of standardization. Various laboratories perform DHSV using different temperature, which may influence ciliary functional analysis (CFA). Recent data suggest that ciliary beat frequency (CBF) increases with temperature, but the relationship between ciliary beat pattern (CBP) and temperature has not been extensively studied. However, CBP evaluation is more sensitive and specific for PCD diagnosis, and the European Respiratory Society recommendation stated that CBF should not be used without assessment of CBP in PCD diagnosis.

Aims
To study the effect of temperature during DHSV on CFA.

Methods
Ciliated epithelial samples were obtained by nasal brushing from 5 confirmed PCD patients. Beating cilia were recorded using DHSV at 37°C and at room temperature (25°C). CFA was assessed by CBF and the percentage of dyskinesia (%DK).

Results
4 patients had an abnormal ciliary function at 25°C and at 37°C, as measured by CBF and the %DK, confirming a PCD diagnosis in the 2 conditions of temperature.

However, the 5th confirmed PCD patient presented at 25°C a normal CBF (17.4 Hz ± 2.4; normal value = 14.8 Hz ± 2.2) and a normal %DK (20%; normal value = 17% ± 8). In contrast, this patient presented an abnormal ciliary function when measured at 37°C (CBF = 9.4Hz ± 13.3 and %DK = 92%).

Conclusion
In our cohort of 5 confirmed PCD patients, CFA performed at 25°C would have missed a PCD diagnosis in one patient. Our pilot study confirms, as suggested previously, that some PCD variants present a temperature sensitive ciliary function, and that the diagnosis may be missed if DHSV is performed under 37°C. Due to the heterogeneity of PCD, this result has to be confirmed in a higher number of patients.

Short Oral Presentation

SO 31.

Impact of COVID-19 on viral respiratory infection epidemiology in young children: a single-center analysis

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Background

The COVID-19 pandemic impacts different health aspects. Concomitant with the adoption of nonpharmaceutical interventions (NPIs) to reduce the spread of SARS-CoV-2, global surveillance studies reported a reduction in occurrence of respiratory pathogens like influenza viruses (IAV & IBV) and respiratory syncytial virus (RSV). We hypothesized to observe this collateral benefit on viral respiratory infection epidemiology in young children.

Methods

Respiratory samples of children aged below 6 years, presenting at the outpatient clinic, emergency department, or pediatric infectious diseases department of the University Hospitals Leuven, between April 2017 and April 2021 were retrospectively analyzed. The occurrence (positivity rate), and seasonal patterns of viral respiratory infections were described. Chi-squared or Fisher's exact test were used to explore differences in occurrence between 2020-2021 and previous 12-month periods.

Results

We included 3020 samples (453 respiratory panels, 2567 single SARS-CoV-2 PCR tests). IAV and IBV were not detected from March and January 2020, respectively. For IAV, positivity rate in 2020-2021 (0%, n=0) was significantly lower than 2018-2019 (12.4%, n=17) ($p<0.001$) and 2019-2020 (15.4%, n=19) ($p<0.001$). IBV positivity rate in 2020-2021 (0%, n=0) was not significantly lower than previous periods. RSV occurrence was significantly lower in 2020-2021 (3.2%, n=3), compared to 2017-2018 (15.0%, n=15) ($p=0.006$), 2018-2019 (16.1%, n=22) ($p=0.002$) and 2019-2020 (22.8%, n=28) ($p<0.001$). The RSV (winter) peak was absent and presented later (March-April 2021). Positivity rate of parainfluenza virus 3 (PIV-3) was significantly higher in 2020-2021 (11.8%, n=11) than 2017-2018 (1%, n=1). PIV-3 was absent from April 2020 to January 2021, whereas no clear seasonal pattern was distinguished the other years. Other parainfluenza viruses, adenovirus, bocavirus, human rhinovirus/ enterovirus, human metapneumovirus and common coronaviruses showed no significant differences in occurrence. From March 2020 onwards, 20 cases (0.7%) of SARS-CoV-2 were identified.

Conclusion

These findings reinforce the hypothesis of NPIs impacting the epidemiology of influenza viruses and RSV in young children. Compared to previous periods, no IAV and IBV cases were observed in the 2020-2021 study period, and the RSV peak occurred later. Since the pandemic is still ongoing, continuation of epidemiological surveillance, even on a larger scale, is indicated.

SO 32.

Imported Pediatric Malaria in Brussel. A study on 160 malaria affected children

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Background
Malaria is a major global public health problem. Imported childhood malaria is rising in the malaria non-endemic countries. European data on malaria affected children are scarce.

Methods
Retrospective case review of all laboratory confirmed malaria for children admitted between 2009-2019 in two teaching hospitals in Brussels. Cases were included when any Plasmodium parasite was isolated from the blood sample. All the isolates were submitted to the National Institute of Tropical Medicine for species confirmation. We describe the main epidemiological, clinical and laboratory characteristics of children 0-15 years old with malaria in the city of Brussels.

Results
A total of 160 children with a median age of 6,8 years (range 5-191 months) were identified. There were 111 (69%) Belgian based children that acquired malaria during their visit to malaria-endemic country to visit friends and relatives, and 49 children were visitors to Belgium(15 or newly installed immigrants. Among the travellers to the malaria-endemic areas 99% were of African ethnicity. The incidence of malaria rates increased during the study period. The peak seasonal incidence was during the months of August-September. Plasmodium falciparum was responsible for 89% of the malaria cases. Almost 80% of the Belgian based children visited the travel clinic for advice, but only one third of them reported to have taken the prophylaxis scheme according to the recommendations. One fifth of the patients had seen one or more physicians prior to malaria diagnosis. Based on the WHO criteria approximately one fifth of the total number of children 31(19.3%) developed severe malaria including coma(n=2); alteration of neurologic status (n=6), convulsions (n=1); severe acidosis(n=1) severe anaemia (n= 3); jaundice (n=1); parasitaemia (n=17).

Conclusions
Malaria is a significant cause of morbidity in returning travellers and newly arrived immigrants to Belgium. The majority of the children had an uncomplicated course of disease and all children fully recovered . Physicians should educate families travelling to malaria-endemic areas on the correct malaria preventive measures and prophylaxis.

Keywords
Imported malaria, severe malaria, pediatric

P 19.

When encephalitis complicates a winter viral condition

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Our patient is a 3-year-old girl who is having fever for 4 days associated with behavior disorders. The parents describe agitation followed by apathy. The first neurological examination shows significant agitation. Contact is difficult. The global motricity is normal and she does not present any sign of lateralization.

Biological evaluation shows a mild inflammatory syndrome and a slight hepatic cytolysis. Influenza A is found in the nasopharyngeal aspiration and high level of IgM against EBV in the blood serology. Lumbar puncture shows no pleocytosis and a normal protein content in the cerebral fluid. Microbiological culture and PCR are performed.

Empiric treatment with Acyclovir is started due to strong clinical suspicion of encephalitis. The diffuse abnormally slow rhythms shown by the EEG supports the diagnosis.

Initial evolution is variable with improvements followed by regressions. All multiplex and Herpes Simplex PCR come back negatives. Another lumbar puncture 4 days later shows mild lymphocytic pleocytosis. The cerebral MRI at day 10 come back normal. Neurological evolution is favorable and EEG gradually improves.

Encephalitis is a rare condition that can result in severe and definitive sequelae and even death. It is an inflammation of the brain parenchyma secondary to a neuroinvasive infection in association with clinical evidence of neurologic dysfunction.

The early diagnosis of encephalitis is crucial to eliminate non-infectious causes, identify pathogens and deliver the most appropriate treatment.

Usually, the diagnostic evaluation is guided by epidemiological and clinical clues, brain imaging, EEG findings and laboratory data. MRI of the brain should be performed in all patients. CSF analysis is also essential in all patients with encephalitis.

The most frequently found causes are viruses. However, despite extensive testing, the etiology remains unknow in most patients.

It is important to distinguish between infectious encephalitis and postinfectious or postimmunization encephalitis (e.g. ADEM).

Acyclovir should be started in all patients with suspected encephalitis pending results of diagnostic studies.

If the Herpes PCR result is negative, consideration should be given to repeating the test 3-7 days later.

Encephalitis can be a severe complication of viral conditions like the flu. Therefore, it is important to always stay vigilant in viral conditions.

P 20.

A clinical case of Chlamydia Pneumoniae epiglottitis

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Background

Epiglottitis is most commonly caused by bacterial infection leading to inflammation and edema of the epiglottis and the adjacent supraglottic structure. If untreated, epiglottitis can progress to life-threatening airway obstruction. In the last 30 years, Haemophilus influenzae type B vaccine has significantly reduced the incidence of the infection in children aged between 2 and 6 years.

Clinical case

We report the case of a 12-year-old child with respiratory failure leading to the diagnosis of epiglottitis. The child was presented to the emergency room with an acute onset of a sore throat, hypersalivation, hoarseness of the voice associated with a feverish state. On admission, clinical examination revealed an anxious patient with the audible sound of stridor and inspiratory dyspnea, rapidly evolving to respiratory distress. With epinephrine nebulisation and a bolus of methyprednisolone the situation improved partially. A chest x-ray showed a narrowing of the trachea. Intubation had to be performed due respiratory decline combined with low blood oxygen levels. Laboratory examination revealed mild inflammatory syndrome and a positive serology for Chlamydia Pneumoniae.

Conclusion

Since the introduction of Haemophilus Influenza vaccine, the morbidity of epiglottitis has significantly decreased in the pediatric population. This case highlights the presentation and clinical manifestations of the disease in an adolescent despite a complete course of vaccination. Nowadays, there is an emergence of new pathogens responsible for certain bouts of epiglottitis. It is important to have the right gestures and medical knowledge in this kind of situation. Thanks to the serological analysis, it was possible to highlight an uncommon pathogen in this type of infection.

P 21.

A case of Pediatric Headscarf Pin Aspiration

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Background

Aspiration of foreign bodies is common, especially in children younger than 3 years of age. Peanuts, nuts and other seeds are the most prevalent objects. However, older children and adults, placing small objects in their mouth, form a distinct and increasing group of patients at risk for accidental aspiration. Most foreign bodies are located in the central airways and can be removed with flexible or rigid bronchoscopy.

Case

A 13-year old girl presented to the emergency department with a history of foreign body aspiration: while fixating her headscarf, she placed a safety pin in her mouth and accidentally inhaled it. Except for persistent coughing, the child was asymptomatic. Physical examination was normal. Chest X-ray revealed the pin in a segment of the right lower lobe, just above the diaphragm. A CT-thorax showed its detailed location, in the posterior basal segmental bronchus of the right lower lobe (B8) and suggested its sharp end pointing downwards and lying just outside of the bronchus. No pneumo- or hemothorax, nor a pneumomediastinum was seen on imaging. Removal was attempted 12 hours after inhalation with a 4.2 mm Olympus flexible bronchoscope and the use of a biopsy forceps. In contrast to CT findings, the sharp end was facing the scope. Subsequently, we were able to grasp the end of the pin and remove it, in the axis of the airway. Screening of the airway post-removal did not reveal any post-traumatic lesions, probably due to its favorable position.

Conclusion

During the temporary placement of a safety pin between the lips, accidental aspiration may occur and has been reported amongst young Muslim girls wearing headscarves. Inhalation of sharp foreign bodies has the risk of life threatening complications, including pneumothorax, bronchial rupture and pericardial tamponade. 8% of these patients require multiple bronchoscopies or thoracic surgical intervention for the treatment of complications and/or removal of the foreign body. Fortunately, the placement of the pin in the mouth with the spherical head, often leads to a favorable position for safe extraction, with the sharp end upwards. If extraction via bronchoscopy is not successful, alternative techniques including the use of intraoperative fluoroscopy, rigid and flexible endoscopy, with or without the use of magnets, tracheotomy and balloon dilatation of the airway, to facilitate foreign body extraction, are proposed. Sometimes video assisted thoracoscopy or thoracotomy is needed.

P 22.

A rare form of tuberculosis: a case of phalangial osteomyelitis in a 12-year-old child

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A Syrian 12-year-old boy was admitted for a progressive swelling and purulent lesion of the left thumb, which appeared after a traumatism. He complained of left thoracic pain, not irradiating, not linked to breathing nor to physical efforts. There were no cough nor symptoms of systemic disease.

He was in Belgium since 2018 and his vaccination status was unknown.

Physical examination showed only painful local swelling and erythema, surrounding a purulent wound in the left thumb.

The X-ray (XR) identified a spina ventosa image. Chest XR and CT described a chronic lateral pleural compartmentalized effusion in the left pulmonary base.

ESR was slightly augmented, without other signs of inflammation.

The tuberculin intradermal reaction was positive, while the swob wound was negative for Mycobacterium tuberculosis (MT).

The biopsy demonstrated giant cell granulomas with Langhans cells and caseous necrosis. GeneXpert MTB/RIF detected a rifampicin sensible MT.

Anti-tubercular therapy was planned for a total of 9 months.

No side effects were observed, the swelling resolved, with a favorably evolution.

Tuberculosis (TB) is still considered a major threat to children worldwide. Physicians in low-incidence regions must be aware of TB, especially in high-risk populations (migrants from endemic areas, malnutrition and/or immunodeficiency status) or facing unexplained clinical entity.

Osteoarticular involvement is a remarkable manifestation of TB albeit hand tuberculosis is exceptional.

Typically, patients claim a painful or painfulness swelling of the finger involved, associated or not to functional limitation, evolving in several months. A fluctuant abscess, or a discharging sinus can be seen at the time of consultation. Systemic symptoms are infrequent.

Laboratory tests are mostly inconclusive, often showing just a mild increase in white cell counts and ERS.

Four types of XR images are generally associated with TB osteomyelitis: cystic lesions, infiltrative lesions, focal erosions and spina ventosa (an expansive lytic lesion due to progressive absorption of the cortex and subperiosteal hyperplasia).

A histological confirmation of giant cell granulomas with epithelioid cells and caseous necrosis is required.

Posters

Since the lesions are classically paucibacillary, the smear microscopy and the nucleic acid amplification test are often negative.

MT cultures are the gold standard test, but they are limited by the long time required.

Treatment corresponds to that of pulmonary TB

P 23.

An unusual cause of pneumomediastinum

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Introduction

Foreign body aspiration (FBA) can be deadly if left untreated. The occurrence is most common in children under the age of three and is considered to be one of the most common causes of accidental death in infants. The severity varies according to the degree of obstruction and the latency in which the patient is taken care of. In severe cases, patients can be presented with subcutaneous emphysema and pneumomediastinum.

Clinical case

Patient aged 4, with no prior medical history, was presented to the emergency room for right sub clavicular swelling and persistent bouts of cough. The mother observed a progressive increase in size of the sub clavicular mass over a 48hr time lapse. Patient had not recently travelled or been in contact with anyone visiting from abroad. Upon arrival, the vital stats showed moderate polypnea with a breathing frequency of 50, increased blood pressure and good oxygen saturation level. Clinical examination showed signs of chest in-drawing, unequal breath sounds, hyper-resonance with percussion over the chest wall, and decreased wall movement on the right side. Chest x-ray revealed a hydropneumothorax of the right lung and moderate inflammation was expressed in blood work. Pleural drainage was performed and was coupled with a CT scan of the thorax. Emphysema and an important pneumothorax of the right lung was concluded as well as the obstruction of the right segmental bronchus, by a heterogenous hypo dense body. Bronchoscopy was performed and a chunk of cotton wool was extracted from the right segmental bronchus. It was revealed that the patient had ingested the cotton wool from her teddy bear a week prior.

Conclusion

In such situations, the pneumomediastinum occurs when air leaks from any part of the lungs or airways into the mediastinum and emphysema is the consequence of this continuous air flow into subcutaneous tissues. Extraction of the foreign body constitutes the best pathway for a quick relief and clinical stability in patients. Occult FBA should be part of the differential diagnosis of any form of chronic or persistent respiratory symptom that is poorly explained, even in the absence of a previous history of aspiration. A proper clinical examination in combination with the adequate imaging is of the essence for a quick and correct diagnosis of FBA. Bronchoscopy remains the best course of action to alleviate symptomatic patients.

P 24.

Primary ciliary dyskinesia: importance of deep phenotyping and genotype/phenotype correlation to confirm the diagnosis

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Introduction

Primary ciliary dyskinesia (PCD) is an inherited heterogeneous motile ciliopathy in which respiratory cilia are stationary, or beat in a slow or dyskinetic manner, leading to impaired mucociliary clearance and significant sinopulmonary disease. The clinical presentation of PCD is highly non-specific, and PCD diagnosis relies on several concordant tests, since none are sensitive or specific enough alone.

Clinical case

A 15-year-old girl presented with a story of respiratory distress in a late preterm infant, and a chronic wet cough and chronic rhinitis since school age. She suffered from recurrent respiratory infections and had required a long-term antibiotic treatment in childhood. She had recurrent suppurative otitis media and hearing deficiency, resolved in late childhood. Pulmonary function tests were normal. A chest CT showed bronchiectasis with situs solitus, and a sinus CT showed a bilateral pansinusitis. She was referred to the Liège PCD diagnostic center. The PICADAR score, a clinical diagnostic prediction score for PCD, was calculated at 6. Nasal nitric oxide measurement was low on 2 occasions. Ciliary videomicroscopy, using nasal brushing samples, was repeated on 3 occasions, and after air-liquid interface cell culture, and revealed ciliary dyskinesia (CBF = 0 Hz and 100% abnormal CBP, with mainly immotile cilia). Transmission electron microscopy revealed the absence of outer dynein arm, and immunofluorescence staining revealed the absence of DNAH5 and DNAH11. A PCD gene panel identified one pathogenic variant and one variant of unknown significance in the gene DNAH5, in trans configuration.

A multidisciplinary team of clinical and laboratory staff determined that the patient had a positive PCD diagnosis, given the compatible clinical history and the correlation between the results of the different diagnostic tests, sustaining the involvement of the gene DNAH5: DNAH5 encodes outer dynein arm component, and biallelic DNAH5 mutations are associated with static cilia with some residual movement.

Conclusion

PCD diagnosis is challenging, and requires a combination of different tests, as there is no single gold standard diagnostic test. According to the ERS recommendation, biallelic mutations in known PCD causing genes and hallmark ultrastructural defects assessed by TEM can confirm a diagnosis, but the correlation between the clinical picture and the different diagnostic tests results is often needed to ascertain a PCD diagnosis.

P 25.

Covid-19 associated thromboembolic events in the pediatric population

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Case report

We describe the occurrence of pulmonary embolism in a 15-year-old female adolescent infected with SARS-CoV-2 and presenting two important risks factors: being overweight and taking estrogens. Her main complaint was left basi-thoracic pain increased by speaking and breathing. The rest of the medical history and clinical examination were reassuring. The injected CT-scan identified pulmonary embolism. The blood biology matched the diagnosis, showing an increase in C-reactive protein and an increase in D-dimer. This complication was treated with low molecular weight heparin and supplemented with direct oral anticoagulants for 3 months.

Discussion

The SARS-CoV-2 infection also affects children and teenagers who usually develop mild disease. A severe complication is the pulmonary embolism, which occurs preferentially in young female adolescents presenting with thrombotic risk factors such as obesity, use of the estrogen pill, neoplasia, central venous catheter or surgery. The risk of developing a pulmonary embolism is increased by the hyperinflammatory and hypercoagulable actions of SARS-CoV-2. The clinical picture presenting with chest pain of varying severity and/or dyspnea in non-massive forms is complex. Moreover, although D-dimer values are not specific to the complication, they may be combined with the injected CT-scan and contribute to the diagnosis. Due to the lack of guidelines on the effective dose for prophylaxis and on the management of pulmonary embolism, the administration of low molecular weight heparin is made on an individual basis with constant adjustment in order to be effective and to avoid secondary bleeding.

Finally, post-complication treatment can be longer in children if they present persistent high D-dimer levels or other thrombotic risk factors.

Conclusion

The SARS-CoV-2 infection is rarely complicated by pulmonary embolism in children and adolescents. Therefore, attention is needed to a misleading symptomatology and risk factors must be investigated. All diagnostic hypotheses must be explored to avoid dramatic outcomes. There are no guidelines for the management of pulmonary embolism in the pediatric population, so the treatment is based on adult guidelines. It must be continuously adapted to the child's age, weight and comorbidities.

P 26.

When the thymus mimics pneumonia

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Introduction

The thymus is a lymphoid organ located in the anterior mediastinum. It gradually involutes with age. On a standard chest x-ray the thymus can sometimes appear atypical in an infant and can complicate the radiographic image interpretation.

Case report

A 9-month-old child was presented at the consultation with persistent fever associated with symptoms of an ongoing bronchitis, despite oral antibiotic therapy prescribed for 5 days. The blood work showed no sign of inflammation but the chest-X-ray revealed an ongoing pneumopathy of the right upper lobe of the lung. The child was then hospitalised, and treatment was administered through intravenous antibiotics for 3 days , followed by an oral antibiotic treatement to complete a course of 10 days. Six weeks later, control Imaging by X ray showed no major difference. A Bronchoscopy was performed and moderate tracheomalacia and some purulent secretions in the right upper lobe of the lung was revealed. Additional imaging through chest echography and a CT-scan confirmed the presence of a non-pathological right thymic hypertrophy. Control imaging showed the transformation of the 'mass' to a thymus with the indication of a « sail sign ». The 3 year follow up demonstrated residual normal thymic tissue through X Ray. The clinical evolution was excellent.

Discussion

Variability in the size of the thymus in children, could render the interpretation of chest x ray complicated. The Thymus can have a lot of forms, with sometimes showing the presence of a « sail sign » due to right lobe hypertrophia. This could thereby mimic a pneumonia, a mediastinal tumor or a cardiomegaly.

Ultrasound could complete the clinical process and reveal the normal size and echogenicity of the thymus. In some situations and especially when there is an extension to the posterior mediastinum a doubt with a tumor may persist and in this case the workup will generally be completed by a CT scan or a thoracic magnetic resonance imaging (RMN) .

Conclusion

In children the thymus may appear in an array of of forms trough standard chest radiography and an enlarged mediastinum may appear pathological when it is not. To avoid additional invasive and unnecessary examinations, it is important to know how to diferentiate between benign thymus hypertrophy from other pathological masses. The ultrasound approach will be helpful to refine the diagnosis, and will be supplemented if necessary by a CT-scan or a RMN.

P 27.

Biphasic stridor and subglottic hemangioma.

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CHC Liège

Introduction

Upper respiratory distress increasing during the first months of life should not be trivialized and may be indicative of a tracheal obstruction.

Clinical case

Our patient presents a first laryngitis accompanied by bronchiolitis at the age of 3 weeks. During his hospitalization, he develops a biphasic stridor, although being previously asymptomatic. His laryngitis soon recurs at the age of 1.5 months – ENT fibroscopy confirming the presence of laryngomalacia. Polysomnography reveals the presence of obstructive apneas. Despite anti-reflux treatment with omeprazole and inhaled corticosteroids, the child keeps a biphasic stridor with signs of indrawing, eating difficulties, and a break in the height-weight curve. A bronchial fibroscopy, performed at the age of 6 months, confirms the presence of a non-pulsatile subglottic mass with almost complete closure of the tracheal lumen on expiration. Injected CT scan confirms the diagnosis of subglottic angioma. Treatment with beta blocker begins and the child improves rapidly with almost complete amendment of stridor. The beta-blocker treatment is weaned at around 13 months of age without relapse thereafter.

Discussion

Subglottic hemangioma is a rare malformation, representing 1.5% of laryngeal malformations in children. Although these vascular malformations are often asymptomatic at birth, they have a high proliferative potential. There is increased growth of the lesions from the first months of life until the age of 12-18 months, after which they reach a stable size and then regress spontaneously. Symptoms are manifested by upper respiratory distress. Although ultrasound is an excellent screening test, it is the injected chest CT scan and bronchial fibroscopy that confirm the diagnosis. The first-line treatment is Propanolol. It allows rapid resolution of symptoms, with a low rate of complications. When treated early, the prognosis is excellent.

Conclusion

Persistent upper respiratory distress in infants should not be trivialized and subglottic haemangioma should be part of the differential diagnosis. Propanol is the treatment of choice and allows rapid improvement of symptoms.

P 28.

Aseptic peritonitis associated with multisystem inflammatory syndrome in children (MIS-C)

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A 6-year-old child is brought to the emergency department with a history of 3-day fever (maximum 40°C) associated with abdominal pain. Clinical examination revealed right iliac fossa guarding and peritoneal symptoms. Biology showed an inflammatory syndrome (CRP 87 mg/L) with normal leukocyte count. Given the hypothesis of acute appendicitis complicated by peritonitis, a decision was made to undertake emergency surgery. On laparoscopy, there was abundant purulent peritoneal fluid but healthy digestive organs, including the appendix. Following persistent fever at 2 days post-surgery and deterioration in general condition, transfer to the paediatric unit was made. On history, a familial SARS-CoV-2 infection was noted 5 weeks earlier. Clinical examination revealed a diffuse macular rash and cervical adenopathy. Further investigations were then performed. Biology revealed normocytic anaemia (Hb 9.4 g/dL), normal leukocyte count, elevation of all inflammatory markers (CRP 191 mg/L, VS 120 mm/h, fibrinogen 8.36 g/L, procalcitonin 2, D-dimers 2.53 ug/ml, ferritin 366 ng/ml, LDH 312 U/l, IL-6 133 pg/ml), elevated cardiac markers (troponin 0.022 ng/ml, NT pro-BNP 16.599 pg/ml) and hypoalbuminaemia (27 g/L). Abdominal ultrasound showed diffuse inflammation of the intestines. Cardiac ultrasound showed dilatation of the anterior interventricular artery with mitral insufficiency. Haemodynamic monitoring revealed several episodes of arterial hypotension. In the light of all these elements, a diagnosis of MIS-C was made and treatment with immunoglobulins, methylprednisolone and acetylsalicylic acid was started (day 5 of fever). The clinical, biological and cardiac evolution was rapidly favourable with complete normalisation of cardiac ultrasound at day 14.

Discussion

Gastrointestinal symptoms are present in over 80-90% of MIS-C cases, making it the most frequent organ system involved. These symptoms can mimic acute abdomen with pseudo-appendicular presentations, aseptic peritonitis or mesenteric adenitis; leading to unnecessary laparoscopy. According to some recent articles, it would even be advisable to evaluate cardiac function prior to surgical exploration in case of acute abdomen during this pandemic.

Conclusion

Our case of aseptic peritonitis associated with MIS-C highlights the fact that a diagnosis of MIS-C should be considered in the differential diagnosis in children with acute abdomen, especially in case of atypical course and recent exposure to SARS-CoV-2.

P 29.

Don't forget the forgotten disease: Lemierre syndrome, a case report

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UZ Brussel

Background

Lemierre syndrome (LS) is a rare condition, recognized by its typical triad of oropharyngeal infection with bacteremia, septic thrombus of the internal jugular vein (IJV) and possible septic metastases. Most often it affects adolescents and young adults with a blank medical history. In this population the most common pathogen is Fusobacterium Necrophorum.

Case presentation

A 3-year-old boy presented to the emergency department with fever since 3 days, swelling on the left side of the neck, vomiting and decreased intake. Physical examination showed an erythematous pharynx and a hard, 4 x 4cm mass on the left side of the neck. Laboratory testing on admission revealed an increased WBC (21,5 x103/mm3) and increased CRP (188,9 mg/L). A doppler ultrasound of the neck revealed a thrombosis of the IJV and a unilaterally enlarged lymphadenopathy with possible abscess. Treatment was started with IV amoxicillin clavulanic acid combined with metronidazole and clindamycin for suspected LS. Blood and throat cultures were positive for Group A streptococcus (GAS). A CT scan of the neck confirmed the thrombus and abscess. The abscess was drained, punctate fluid also revealed GAS. Metronidazole and clindamycin were stopped after 2 days. Fever resolved after 2 days and progressively the swelling of the neck decreased. After 10 days of treatment patient was discharged home on oral clindamycin, which was continued for another 2 weeks. Follow up ultrasounds of the thrombosis showed a spontaneous decrease in size without the use of anticoagulation.

Conclusion

Clinical suspicion for the diagnosis of LS should be aroused when a febrile patient with an ENT infection develops acute swelling of the neck, sepsis or organ failure due to septic emboli. Doppler ultrasound should be performed to screen for possible thrombophlebitis. One should be aware of the disease to assure a rapid diagnosis and subsequent interventions to avoid lethal consequences. To properly treat LS, we advise a high dose IV antibiotic therapy primarily directed towards gram positive and anaerobic bacteria to protect against the most common micro-organisms. Due to the lack of RCT in the pediatric population to validate the use of anticoagulation therapy, it is currently only recommended in cases with septic disease or thrombus progression despite the use of antibiotics.

P 30.

Takayasu arteritis in a 13-year old girl: a case report

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Background

Takayasu arteritis (TA) is a rare form of large vessel vasculitis. The age of onset is usually in the range of 10-40 years with a strong female predisposition of 8:1. The diagnosis of TA is often delayed due to difficulty in recognizing patients as symptoms are systemic but nonspecific.

Case presentation

A 13-year old girl presented to the emergency department with fatigue, anorexia, vomiting, weight loss and myalgia. Physical examination revealed a weak girl with abdominal pressure pain. Cardiorespiratory parameters at admission were normal, except for a left-right difference in blood pressure. Blood results showed a strong inflammatory blood count: ESR >120mm/h, CRP 180 mg/L, ferritin 454 µg/L and impaired coagulation. Covid-19 PCR, Covid-19 and other viral serology was negative. There were no signs of bacterial/fungal infection. The mycoplasma pneumoniae titer was 1/2480, a macrolide was started. Rheumatological markers RF, ANA, ENA, ANCA and complement were negative, but IgG was markedly increased. Tuberculosis screening was negative. Chest X-ray showed no abnormalities. Abdominal ultrasound and CT abdomen showed a mesenteric adenitis. Echocardiography revealed a mild aortic insufficiency. Due to ongoing vomiting despite anti-emetics, MR brain was performed and was normal. Despite the intravenous administration of broadspectrum antibiotics started at admission, fever and increased inflammatory blood markers persisted during hospitalization. A PET CT was performed which showed an aneurysm (36 mm) and inflammation of the ascending aorta, aortic arch and common carotid artery, which led us to consider TA. Treatment with oral high dose glucocorticoids was initiated with a favorable clinical response. MR angiography showed the known aorta aneurysm and an amelioration of the inflammation. To minimize glucocorticoid-related toxicity methotrexate was initiated and follow-up in pediatric rheumatology was provided.

Conclusion

There needs to be a high index of suspicion by the physician in female adolescents presenting with nonspecific systemic symptoms who have raised inflammatory markers without obvious cause. From a cardiology perspective, further suspicion should be raised for annular dilatation secondary to aortopathy if aortic regurgitation is present with a structurally normal aortic valve. Performing a PET CT to confirm this clinical suspicion and exclude other possible causes of persistent fever can provide an early diagnosis of TA.

P 31.

Impact of refining the U Na/Creat cut-off according to age on the diagnosis of salt depletion in patients with cystic fibrosis

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Objective

Salt depletion without hyponatremia is thougt to be common in patients with cystic fibrosis and could be associated with poor growth. Assessment of sodium status can be done using the least invasive U Na/Creat ratio (mmol/mmol). In case of normal kidney function, a U Na/Creat cut-off of 17 mmol/mmol is widely used in infants. Recent data convincingly suggest that this treshold actually varies with age as muscle mass increases (Declercq et al. J Cyst Fibros 2021). We aimed to evaluate the impact of this approach on salt depletion diagnosis.

Methods

We carried out a retrospective study of urine samples collected in outpatients between january 2020 and december 2021, at clinician's discretion. Salt depletion prevalence was calculated using both a fixed U Na/Creat cutt-off (17) or an age-adapted cutt-off (<6 y : 17.6, 6-<12 y : 14.8, 12-<18y : 11.7, >18 y : 10.3).

Results

193 samples were collected. U Na/creat was < 17 in roughly half the patients. Among 92 patients ⍰ 6 y, salt depletion was documented in 60 (65%) using a cutt-off of 17. Corresponding values were 49 (53.3%) using age-dependent cutt-offs, which avoided overdiagnosis in 11 (12%) patients.

	All	<6 y	6 - <12 y	12 - <18 y	>18 y
Samples (%)	193	101 (52.3%)	29 (15%)	21 (10.9%)	42 (21.8%)
UNa/Creat <17 (%)	101 (52.3%)	41 (40.6%)	17 (58.6%)	11 (52.4%)	32 (76.2%)
UNa/Creat <N age adapted (%)	90 (46.7%)	41 (40.6%)	16 (55.2%)	8 (38.1%)	25 (59.5%)

Conclusions

Sodium depletion is common in cystic fibrosis patients. Interpreting U Na/Creat according to age significantly decreases the prevalence of sodium depletion diagnosis in patients aged 6 years or more.

LO 4.

Multispecies probiotic for the prevention of antibiotic-associated diarrhoea in children: a randomised clinical trial

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Background/Aims
The efficacy of multispecies probiotic formulations in the prevention of antibiotic-associated diarrhoea (AAD) remains unclear. We aimed to assess the effect of a multispecies probiotic on the risk of AAD in children.

Methods
We conducted a randomised, double-blind, controlled trial. Children aged 3 months to 18 years undergoing systemic antibiotic therapy were assigned to receive a placebo or a multispecies probiotic (Ecologic AAD 612) consisting of eight bacterial species for the duration of antibiotic treatment and for the following 7 days . The primary outcome was AAD, defined as three or more loose or watery stools per day in a 24-hour period, caused either by Clostridioides difficile or of otherwise unexplained aetiology, after testing for common diarrhoeal pathogens. Among the secondary outcomes, we assessed diarrhoea regardless of the aetiology, diarrhoea duration, and predefined diarrhoea complications.

Results
A total of 350 children were randomised and 313 were included in the intention-to-treat analysis. Compared with placebo (n=155), the probiotic (n=158) had no significant effect on risk of AAD (relative risk [RR] 0.81; 95% confidence interval [CI] 0.49 to 1.33). However, children in the probiotic group had a lower risk of diarrhoea regardless of the aetiology (RR=0.65; 95% CI 0.44 to 0.94). No differences were observed between the groups for most of the secondary outcomes, including adverse events.

Conclusion
The multispecies probiotic used in this trial did not reduce the risk of AAD when analysed according to the most stringent definition. However, we found a beneficial effect of the formulation on the overall risk of diarrhoea during and 7 days after antibiotic therapy (number needed to treat: 9). The latter outcome corresponds well with the standard approach to AAD in clinical practice. Therefore, the use of the studied probiotic may be considered for diarrhoea prevention during antibiotic treatment in children. Our study also shows that the AAD outcome definition has a significant impact on clinical trial results and their interpretation.

SO 12.

Therapeutic interventions in pediatric eosinophilic esophagitis: a retrospective study.

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UZ Leuven

Background

Eosinophilic esophagitis (EoE) is a chronic, food antigen driven disease with increasing prevalence. As a first-line treatment, a proton pump inhibitor (PPI) is used. In PPI non-responsive EoE, treatment options include topical corticosteroids (tCS) or dietary interventions such as the elemental diet (ELED), the allergy test-directed elimination diet or the empiric elimination diet. In this last diet, one or more subsets of the 6 most commonly associated food triggers (cow's milk, soy, hen's egg, wheat, peanut/tree nuts and fish/shellfish) can be eliminated. A step-up from a 2-food elimination diet (2FED) to a 6-food elimination diet (6FED) approach or a step-down approach is used. The aim of this study is to investigate which treatment is used in our pediatric population and the effect of it on the histological remission.

Methods

A retrospective study was performed with the following inclusion criteria: (1) children up to 18 years, (2) a diagnosis of EoE in UZ Leuven between 1/1/2014 and 31/12/2021 and (3) histological confirmation of >15 eosinophils per high-power field in esophageal biopsy. Nineteen children were included. We performed a medical chart review and relevant data was collected. GraphPad Prism was used for statistical analysis.

Results

Ten out of 19 children had a history of symptoms caused by one or more associated IgE-mediated food allergy'(s): hen's egg (n=4), cow's milk (n=5), wheat (n=3), peanut/tree nuts (n=7), soy (n=2) and fish/shellfish (n=4). Six of them were diagnosed with a pollen allergy. At last follow-up, 16 children obtained histological remission and in 3 APO remained positive despite initial treatment. Monotherapy was successful in 5 patients: PPI (n=2), tCS (n=1) and dietary approach (2FED, n=2). The other 11 children had a combination therapy of which 8 combined two therapeutic strategies (PPI and diet, n=6), diet and tCS (n=2)) and 3 were treated with triple therapy: PPI, tCS and a diet. The 3 children with a remaining positive APO were treated with PPI and tCS (n=2) or diet (6FED) and tCS (n=1). None of the children were treated with ELED.

Conclusion

While EoE is known to be primarily a non IgE-mediated allergic disease, associated IgE-mediated food allergy is common in our population. Moreover, pollen allergy is not infrequent and biopsies in pollen season should be avoided. Furthermore, histological remission is rarely obtained with monotherapy and a combination of different strategies is often needed.

P 60.

Recurrent Mycoplasma pneumoniae infection causing hepatitis and cholestasis in a young girl

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Background/aim
We present a rare case of a five year old girl with recurrent episodes of hepatitis and cholestasis associated with mycoplasma pneumoniae infection.

Methods
The case information was retrieved from hospital reports.

Results
A healthy 5 years old girl, presented a first time with cholestasis, polydipsia and polyuria. Blood results showed: normal glycemia, alanine aminotransferase (ALAT) 90 U/L (0-33 U/L) / aspartate aminotransferase (ASAT) 123 U/L (0-41 U/L), gamma-glutamyl transferase (ɣGT) 45 U/L (4-12 U/L), total bilirubin 2.87/ mg/dl (0-1.2 mg/dl), direct bilirubin 2.4 mg/dl (0-0.3 mg/dl), C-reactive protein (CRP) 1.2 mg/L (< 5mg/L), white blood cells 23500 mm3 (3500-12000 mm3), neutrophils 9188 (1800-6800 mm3) lymphocytes 10928 mm3 (1500-5500 mm3). Hepatitis B (protected), C and A, Epstein-Barr (EBV) virus and cytomegalovirus (CMV) serology were negative. Mycoplasma pneumoniae titer was positive (320 (positive if >160). She was treated with clarithromycin 20 mg/kg/day for five days and symptoms rapidly disappeared allowing to stop the drugs that were started for icterus/pruritus (Desloratadine 5 mg/day and 150 mg ursodeoxycholic acid 3x/day). Exactly one year later, she presented again with exactly the same symptoms. Blood results showed similar abnormalities as the first episode. However, CMV titer IgG was 0 and IgM 1.77 (positive if ≥1,00:) and Mycoplasma pneumoniae titer was also positive, with a much higher titer as during the first episode (2560). As mycoplasma serology can stay elevated for a longer period, we considered this second episode possibly linked to CMV. Abdominal ultrasound and genetic screening for (recurrent) cholestasis panel was normal. We treated her for the cholestasis and itchiness like first episode together with a topical cooling cream and Rifampicin due to persisted itching. One month after treatment, the symptoms persisted as did the cholestasis and elevated transaminase. So clarithromycin 20mg/kg/day was started for five days because, CMV viral load were negative. Like the first episode, within one week she was completely asymptomatic. Within a month her laboratory testing normalised, except for Mycoplasma serology that increased up to 1:5120, two months after the onset of symptoms.

Conclusion
Recurrent Mycoplasma induced hepatitis, to our knowledge, has never been reported. Mycoplasma is a rare cause of cholestasis in children and should be recognized early to enable adequate treatment.

P 61.

An unusual rectal protrusion in a two year old girl caused by a rectal duplication cyst: Case report and literature review.

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Background/aims
We present a rare case of unusual rectal protrusion in a two year old girl, caused by a rectal duplication cyst. How to approach these patients in the clinic in not always easy due to the slight different presentation of symptoms and young age. Therefor we have evaluated the literature about diagnosis and management.

Methods
The case information was retrieved from hospital reports. We used heading and key words of interest to extract articles in English language from PubMed and Google Scholar.

Results
A two year old girl had an unusual rectal protrusion, occurring four months after starting toilet training. The rectal protrusion could be manually repositioned. Digital rectal exam was normal, as the rest of the physical examination. The initial diagnosis was prolapse related to constipation. An ultrasound of the abdomen was normal. Due to parental observation and pictures of deformation of the stools, a polyp was suspected. However, colonoscopy was normal with however an impression of a slight bulging of the mucosa in the rectum. A contrast radiography with gastrographin enema did not show any abnormality. A magnetic resonance imaging of the abdomen was performed and showed a suspicion of a lipoma (10 x 19 x26 mm) pre-sacral against the rectum wall. Multidisciplinary approach was performed with additional advice of different paediatric specialists. Additional blood/urine investigation was normal. A surgical intervention was performed: through a posterior sagittal incision the prerectal mass could be completely removed out of the posterior rectal wall. Pathology result showed a duplication cyst, completely excised. After surgery, the rectal protrusion did not recur during a three month follow-up. We found three other case reports mentioning rectal duplications cysts presenting with a rectal prolaps. However, they had additional clue-symptoms compared to our case like a palpable mass during digital rectal examination and/or rectal bleeding. Literature review shows different diagnostic approaches. For the management of a rectal duplication cyst, a minimal invasive surgery with total excision by a posterior sagittal approach is suggested.

Conclusion
A duplication cyst can be a rare cause of rectal protrusion in young children. Parental documentation and a multidisciplinary approach were key in finding the correct diagnosis, after a normal digital rectal exam led us astray. Diagnostic strategies vary in the literature.

P 62.

Perception of transition in Inflammatory Bowel Disease patients: Point of view of the pediatric and adult care giver.

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Background/Aims

Up to the 15% of patients affected by Inflammatory Bowel Disease (IBD) receive their diagnosis before the age of 20 years. The management of children with IBD will include the transition from the pediatric to the adult IBD care system. At CHC Montlégia Clinic (Liège), the transition is considered as an essential step in the care of a young patient.

The aim of this survey is to collect the perception that the actors of pediatric and adult care have of the transition. The collection of the caregivers' point of view aims to establish a consensus of practices.

Methods

In order to obtain a multidisciplinary view of the concept of transition, 9 medical and paramedical staff members were interviewed: one pediatric dietician, 2 nurses, 4 pediatric gastroenterologists, one adult gastroenterologist and a pediatrician specializing in adolescent medicine. Each of them answered four questions on the "when" and "how" to introduce this transition.

Results

The interviewed professionals agreed that the age of Belgian majority was the “ideal“ time to initiate the transition process. However, other factors should also be taken into account before the transition to adult medicine is undertaken: the young person's autonomy, his or her capacity for discernment, the remission of the disease, adherence to the treatment and also the parents' willingness to “let it go”. The opinion of the carergivers also converges with regard to the number of shared consultations to be carried out with the adult and pediatric gastroenterologist before the transition is made. Two to four joint consultations seem necessary in order to make the transition, depending on the patient's rhythm. The first joint consultation, led by the pediatrician, would in particular make it possible to establish an initial contact with the adult gastroenterologist. Some believe that this first joint consultation should be planned 2 to 3 years before the majority while others would organise it 3 to 6 months before the transfer. Finally, all the care givers considered psychological support for the young person and his family to be essential in this transitional context.

Conclusion

The transition in the care of adolescents/young adults with IBD is a dynamic process that must be anticipated and prepared, taking into account the evolving capacities of the patient. Coordination between the pediatric and adult care teams therefore seems essential to aim for a better quality of care.

LO 11.

Dental development disorders in children who had chemotherapy treatment before the age of 10 for a malignant disease

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U Gent, UZ Gent

Objective
Childhood cancer survivors are at risk to develop long term sequelae caused by the treatment. The aim of this study was to investigate the prevalence of dental abnormalities in survivors of childhood cancer treated with multichemotherapy. In addition, a possible effect of type of malignancy and treatment and the occurrence of more and/or more severe dental abnormalities was examined.

Methods
Eighty-one patients between the age of 6 and 20 years who had been treated with chemotherapy for a malignant disease before the age of 10 and who were off therapy for at least 2 years were examined clinically and radiographically (Planmeca ProMax® 2D). Finally, 69 patients were included in the study. For each individual, the permanent teeth with an abnormal root-to-crown ratio, as well as the number of agenetic and microdontic teeth were counted. With this information, the Individual Defect Index (IDeI) was calculated, which quantifies the severity of the dental developmental disorders. The results of the study were compared to reference values from literature. To examine the possible effect of type of malignancy/chemotherapy, patients were divided into four groups: ALL and T-cell lymphoma (40 pt); Burkitt's lymphoma and A.L.C.L (6 pt); neuroblastoma (4 pt) and other (19 pt).

Results
In the whole study population, at least one tooth development disorder was seen in 66 of 69 patients (95%). Two or more different abnormalities were noted in 83.3% . Agenesis was diagnosed in 7.2% of the study population and microdontia in 30.4%. This is significantly higher than in the normal population where the prevalence of microdontia is 2.5%. A significantly higher mean IDeI score was seen in the study population (score: 13.01 , range 0-43) compared to the normal reference (score: 1.8 , range 0-15).

Differences in the root-to-crown ratio were found between the four groups. Microdontia and/or severe abnormalities in the root-to-crown ratio are most commonly seen in the neuroblastoma group and agenesis in the Burkitt lymphoma pts. Finally, the average IDeI score was highest in the ALL category. However, the differences are not significant in the different groups.

Conclusion
Treatment with multichemotherapy at young age has an explicit negative impact on dental development. We recommend to refer children who undergo chemotherapy treatment at a young age to a specialized pediatric dental team in order to identify developmental disorders in time.

Short Oral Presentation

SO 7.

Comparison of microscopic bone marrow examination versus MIBG scintigraphy in detecting bone marrow involvement in stage 4/4S neuroblastoma patients

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UZ Leuven

Background/aims

In patients with stage 4 or stage 4S neuroblastoma, it is advised to perform a microscopic bone marrow (BM) examination (on aspirates and biopsies) and a [123I]MIBG scintigraphy to detect BM metastases. The aims of this retrospective study was to assess the concordance of [123I]MIBG and microscopic BM examination in detecting BM involvement both at the time of diagnosis and during treatment, namely before autologous stem cell collection (ASCC). In addition, results of BM biopsies and aspirates in detecting BM involvement were compared.

Methods

In this study, the results of 55 pediatric patients with stage 4 or stage 4S disease were reviewed, including the results of 37 patients who received an autologous hematopoietic stem cell transplantation (AHSCT). The concordance/discordance rates between the diagnostic tests (BM aspirate, BM biopsy, [123I]MIBG scintigraphy) were measured and a McNemar test was used to analyze paired binary data to look for a systematic difference.

Results

Acceptable concordance rates for the microscopic BM examination versus [123I]MIBG were found at time of diagnosis and before ASCC (85.3% and 77.1% respectively). Discordant results between both examinations were found in both directions and at both time points. Comparing results of the biopsy versus aspirate at diagnosis, the concordance rate was 80.6%. A much higher concordance rate (94.1%) between the microscopic evaluation of the BM biopsy and the BM aspirate was however found before ASCC. While two biopsies still showed tumor invasion before ASCC, none of the BM aspirates showed residual neuroblastoma cells.

Conclusion

For pediatric patients with stage 4/4S neuroblastoma, our retrospective data show that a microscopic examination of a BM aspirate and biopsy, as well as a [123I]MIBG scintigraphy should be performed as complementary tools in the evaluation of BM involvement because of their diagnostic heterogeneity. These three examinations should be done not only at diagnosis and but also during treatment (before ASCC).

SO 8.

Deciphering the non-coding RNA landscape of pediatric acute myeloid leukemia

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UZ Gent

Pediatric acute myeloid leukemia (pedAML) is a heterogeneous blood cancer of childhood age. Although survival rates have significantly improved over the past decades, still 20-30% of children will succumb due to treatment-related toxicity and relapse. Extensive molecular characterization of the leukemic stem cell, shown to be responsible for relapse, is needed to improve treatment options and survival. Recently, it became clear that also non-coding RNAs, including long non-coding RNAs (lncRNAs) and microRNAs (miRNAs), play a role in the development of human diseases, among which pediatric cancer. Nevertheless, non-coding RNA expression data in pedAML are scarce. Here, we explored lncRNA (n=30168) and miRNA (n=627) expression in pedAML subpopulations (leukemic stem cells (LSC) and leukemic blasts (L-blast)) and their normal counterparts (hematopoietic stem cells and control myeloblasts, respectively). Potential regulatory activity of differentially expressed lncRNAs in LSC (unique or shared with the L-blast comparison) on miRNAs was assessed. Moreover, pre-ranked gene set enrichment analyses of (anti-)correlated protein-coding genes were performed to predict the functional relevance of the differentially upregulated lncRNAs in LSC (unique or shared with the L-blast comparison). In conclusion, the study provides a catalog of non-coding RNAs with a potential role in the pathogenesis of pedAML, paving the way for further translational research studies.

Posters

P 1.

Self Limiting Sternal Tumors of Childhood: about a case

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CHU de Liège

Anterior chest wall tumors in children have various differential diagnoses, including malignant tumors, osteoarthritis, inflammatory disease or benign tumors. Among these, self limiting sternal tumors of childhood (SelSTOC) are entities of sternal, benign, rapidly growing tumors, with no previous history of trauma or infection. They are characterised by spontaneous regression. Imaging can be helpful. Their management is challenging, to avoid aggressive diagnostic investigations and unappropriated treatment without missing any severe differential diagnosis.

We report the case of a 14-months-old girl who was briefly explored in our centre for a rapidly growing sternal tumor clinically and radiologically compatible with SelSTOC. Despite low-grade fever and mild inflammatory syndrome, we opted for a watchful waiting and anti-inflammatory treatment. The patient recovered spontaneously after a few weeks.

In conclusion, facing rapidly growing sternal swelling, paediatricians must consider SelSTOC as differential diagnosis to avoid unnecessary aggressive investigations or treatment.

P 2.

Torticollis : not always a trivial symptom

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Torticollis is a common symptom in children. It is defined as an abnormal neck position. It can be due to a wide variety of pathologies, from benign to life-threatening ones. The association with pathologies of the posterior fossa or the cervical spinal cord is often underestimated leading to diagnosis delays.

A 8-year-old girl presented with frontal headaches evolving for 48 hours accompanied by vomiting. She didn't reported any other acute complaints but cervical pain treated by regular physiotherapy leading to intermittent clinical improvement.

Her clinical examination was reassuring apart from the evidence of a slight contracture of the muscles of the nape of the neck. The MRI showed a brain tumor of the posterior fossa and cervical spinal cord. In this context, surgery was performed. Anatomopathology revealed an ependymoma. The resection was unfortunately incomplete. Radiotherapy has been initiated and is currently in progress.

Acquired torticollis can be the first or the only sign of a wide range of pathological conditions such as muskulo-skeletal disorders or syringomyelia. It can also be a warning sign of a posterior fossa tumor or cervical spine cord tumor and may preceed other neurological symptoms.

Unfortunately, this diagnosis is not part of the initial differential diagnosis and therefore, the exclusion of serious underlined central nervous system lesions including cerebral tumors is commonly delayed.

Cerebral tumors are the most common solid tumor in children, the posterior cranial fossa being the most common localization. Symptoms vary and can include headaches, vomiting and ataxia. The torticollis is frequently overlooked but the occurrence can reach up to 30% of the patients with posterior cranial fossa tumors, and is especially seen in astrocytomas and ependymomas.

Ependymomas represent 6 to 12% of pediatric brain tumors. The management includes surgery (the resection must be as complete as possible) and radiotherapy. The place of adjuvant chemotherapy is still being studied.

The prognosis depends on age, localization and degree of resection of the tumor.

In the presence of a torticollis, the clinical examination must be conducted in a meticulous and systematized way. Indeed, even if the etiology of torticollis in children is mostly benign, one should not forget the possibility of a serious cause that can be life-threatenin

P 3.

Case report : Juvenile trabecular ossifying fibroma in an 12 year old patient

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Background

Juvenile ossifying fibroma (JOF) is one of the rarest entities within the very heterogenous and vast group of benign fibro-osseous tumors. It is often diagnosed in the first two decades of life and is usually asymptomatic, affecting males and females indifferently. JOF commonly arises in the mandibular or maxillary region and exhibits a particularly aggressive behaviour characterized by rapid growth and high risk of recurrence (estimated at 20-30%). Surgical enucleation, followed by curettage and/or osteotomy remains the main treatment strategy.

Case Report

A 12 year old boy presented for investigation of a painful mandibular mass, that was noticed following a minor mandibular trauma. The patient was apyretic and there was no sign of infection. Medical history was uneventful. Physical examination confirmed an inferior-lateral right mandibular mass that was firm and not mobile, without any skin lesions. Complete mouth opening was prevented by the lesion, thus compromising solid feeding.

Blood test showed no sign of inflammation. Serological screening was negative. A mandibular CT scan revealed an encapsulated formation of about 5 cm in width, located within the right ascending branch of the lower mandibular, facing the dental root 48. MRI showed no sign of bone destruction, although a FDG PET-CT imaged a hyperavidity of the lesion with a suspicious adenopathy in IIa, homolaterally.

A biopsy was performed and revealed a tumoral structure with osteogenic differentiation and lamellar osteoid matrice, amid disrupted osteoblastic and connective structure. Histology was consistent with juvenile trabecular ossifying fibroma.

Conservative surgical endobuccal resection is planned, followed by 6 weeks maxillomandibular block to optimise bone consolidation.

Conclusion

Although JOF has no potential of malignant transformation, its rarity and its silent presentation can make diagnosis challenging. Differential diagnosis with odontogenic tumors, sarcoma, hematopoietic neoplasms, langerhans cell histiocytosis or secondary metastasis is important. Surgical resection is the mainstay of treatment, although harbouring its own challenges, such as aggressive surgery sometimes requiring bone grafts, as well as high recurrence rate. Those features contribute to increase JOF's potential morbidity.

P 4.

Case report: Thrombocytopaenia and Down Syndrome – what about the usual suspects?

C. Schmit, C. Geurten

ULG

Down syndrome is a chromosomal disorder renowned for its predisposition to haematological conditions and auto-immunity. Haematological abnormalities in those children should prompt investigations to exclude malignancy, but differential diagnosis should always include other pathologies.

We present the case of a 15 months-old infant with trisomy 21 that presented with isolated moderate thrombocytopaenia (platelet count 66.000/mm3, Hb 11.3 g/dL, WCC 6900/mm3) during an episode of upper respiratory tract infection, with a clinical examination unremarkable except for mild hepatomegaly and diffuse petechial rash. History included a recent episode of infectious mononucleosis and a mild interatrial communication.

Due to worsening of thrombocytopenia and onset of rectal bleeding requiring initiation of treatment, a bone marrow aspiration was performed, showing normocellularity with normal megakaryocytes. Intravenous immunoglobulins were administered without any significant benefit over the course of the following 7 days. The patient did however develop haemolytic anaemia (Hb 7.8 g/dl, reticulocytes 217.0000/mm3, haptoglobin <0.01, schizocytes 54/mm3) and steroids were initiated without significant response. Platelet count increment following transfusion was over 300.000/mm3 and sustained for over 24h. A repeat marrow with trephine confirmed the absence of malignant infiltration. Caryotype was 47XY, +21c, GATA-1, CEBPA, FLT3 and NPM1 were not mutated. Differential diagnosis at this stage included malignancy, Evans' syndrome, and thrombotic microangiopathy. ADAMST13 (a disintegrin and metalloproteinase with a thrombospondin type 1 motif member 13) activity was undetectable (<0.2%), and plasmatic inhibitors were detected (4.25 U Bethesda), confirming the hypothesis of immune thrombotic thrombocytopenic purpura, triggered by an infection, or unravelled by passive transfer of antibodies through the immunoglobulins administered earlier in the course of the disease. Daily plasmapheresis were started with immediate but short-lived benefit, and TTP persistently recurred every 48h after interruption of exchanges, warranting initiation of treatment with Rituximab 375 mg/m2 x4 after 2 weeks.

Acquired or immune TTP is an extremely rare entity in children below 9 (<1/1.000.000), and can be life-threatening through terminal circulation microthrombi leading to organ damage and dysfunction. Prompt recognition of this entity is important to avoid long-term damages.

P 5.

Factor IX and XII combined deficiency: a rarely seen coagulation disease.

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Background

An eight-months-old boy suffering of recurrent petechiae presented a deficiency of both clotting factors IX and XII. Deficiency of factor IX is an inherited-X-linked or acquired disease, well-known as hemophilia type B that mostly affects males and leads to bleedings and petechiaes. Factor XII deficiency is a rare genetic blood-disease not associated with hemostasis disorders but can sometimes lead to thrombosis. However, factor XII deficiency decreases vascular permeabilty (by inhibiting the conversion of kininogen to bradykinine/signaling pathways) and so reduces red blood cells extravasation. The combined deficiency of clotting factors IX and XII is unusual and poorly known.

Methods

We analysed clinical and laboratory data of the eight-months old child admitted in our center at Epicura hospital. We compared the data from our case report with the available literature about factor IX and XII combined deficiency.

Results

We report a case of an eight-months-old boy admitted in our emergency unit with a rash not erased by vitropression ; it appeared on his body less than 24 hours earlier. His clinical examination was normal, except an one centimeter diameter isolated purpuric spot on the chest. The mother described recurrent petechiae but no abnormal bleeding in her child. An uncle would have clotting disturbances not yet investigated. The initial laboratory analysis performed revealed an absence of inflammatory or infectious syndrome, a normal hemoglobin level, a normal platelet count and no liver cytolysis but a clotting disorder. Prothrombin time and INR levels were in normal range. However, the Activated Clotting Time and the Kaolin Clotting Time were increased ; a factor IX and XII deficiency was diagnosed. A familial genetic check-up is in progress.

Conclusion

We reported an uncommon case of recurrent petechiae in a child whose diagnosis is a factor IX and XII combined deficiency. The literature concerning the factor IX and XII combined deficiency is poor. According to our present knowledge, these petechiae can be explained by an isolated factor IX deficiency but not by an isolated factor XII deficiency that usually decreases vascular permeability. The both factor IX and XII combined deficiency can probably explain the intermediate symptomatology (isolated petechiae without bleeding or thrombosis) presented in our child.

P 6.

Pediatric Refractory Acute Immune Thrombocytopenic Purpura Secondary to Asymptomatic SARS-COV2.

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Acute immune thrombocytopenic purpura (ITP) has been revealed as an uncommon complication of COVID-19 in children. Severe bleeding can occur but is rarely life threatening. Management is based on the severity of bleeding symptoms and degree of thrombocytopenia. Initial treatment usually consists in close monitoring in an ambulatory setting. Severe cases may require systemic corticosteroids, intravenous (IV) immune globulins, or both. The use of Rituximab, thrombopoietin receptor agonists, immunosuppressive therapy, splenectomy and other new therapeutic interventions will be discussed in case of refractory ITP.

We report the case of a 7-year-old girl with severe acute ITP secondary to a COVID-19 infection without any respiratory symptoms. We notify a traumatic skull fracture in the medical history. The initial clinical examination showed a large bulging mediodorsal hematoma, purpuric lesions, posterior pharyngeal hemorrhage, and secondary macroscopic hematuria. Laboratory results report severe thrombocytopenia with platelets <1000/mm^3 and detect the presence of antiplatelet antibodies. A recent COVID 19 infection was diagnosed based on positive PCR test.

The patient was hospitalized in a pediatric intensive care unit. Initial medical management consisted of two courses of 1g/kg IV immune globulins and 4mg/kg/day of Solumedrol for 72 hours. Despite the treatment, severe bleeding and thrombocytopenia progressed. The patient also developed neurological symptoms (headhaches, apathy) with normal cerebral imaging and hypopituitarism secondary to corticotherapy. She received 8 units of platelet transfusions in 4 days and 30 mg/kg/day Solumedrol for 72 hours to ensure sufficient hemostasis. Her clinical state improved and we were able to taper the oral corticotherapy to 4 mg/kg/day for 1 day and 1 mg/kg/day for the next 5 days before stopping. Three weeks after the end of the corticosteroids and six weeks after the IV immune globulins she has not experienced relapse.

This case presents a rare and severe acute pediatric ITP secondary to asymptomatic SARS-COV2 which was refractory to initial management. It underscores the importance of COVID19-testing in new ITP diagnoses even in cases without any of the usual symptoms (cough, fever, fatigue). The future scientific challenge will consist in the determination of predictive factors of therapeutic response to optimize medical management and therapeutic options to manage severe bleeding in refractory ITP.

P 7.

Neonatal cholestasis as initial manifestation of primary hemophagocytic lymphohistiocytosis

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Background
Hemophagocytic lymphohistiocytosis (HLH) is severe hyperinflammatory condition characterized by high fever, cytopenia, and hepatosplenomegaly. Primary (genetic) and secondary HLH are clinically indistinguishable, but knowledge on the etiology may be important for treatment choices (e.g. early HSCT). Timely recognition of HLH is crucial as it can result in significant morbidity and mortality. Diagnosis of HLH, however, remains challenging and current diagnostic criteria insufficiently cover the diverse clinical presentations.

Methods
We report an infant in whom an initial presentation of neonatal cholestasis confounded the diagnostic process and delayed identification of un underlying primary HLH.

Results
A female infant born to non-consanguineous Caucasian parents at 35 weeks, was referred at day 4 because of cholestasis, thrombocytopenia, and hepatosplenomegaly. Elaborate work-up remained inconclusive, but a metabolic disorder was suspected. Cholestasis and thrombocytopenia spontaneously resolved in the first weeks, hepatosplenomegaly remained. At the age of 2 months, she had an episode of HLH that resolved rapidly without corticosteroids. Based on the latter, secondary HLH was assumed. A tentative diagnosis of Wolman disease was made, a lysosomal storage disease associated with secondary HLH. However, from the age of 4 months, she presented with increasingly severe episodes of HLH requiring intensive immunosuppressive treatment (HLH-2004 protocol). Additional work-up revealed familial HLH type 3. Unfortunately, by the time the diagnosis was confirmed, she suffered from severe therapy-refractory CNS inflammation to which she succumbed at the age of 8 months.

Conclusion
With our report, we underline that neonatal cholestasis can be an early symptom of primary HLH. Previous reports demonstrated that cholestasis is a relatively common feature of HLH in neonates, suggesting it may not be that atypical as currently assumed. Pediatricians should keep a high index of suspicion especially when associated with cytopenia and splenomegaly. Note that cholestasis is not included in the HLH diagnostic criteria set forth by the Histiocyte Society, and evaluation for HLH is missing in the NASPGHAN-ESPGHAN consensus recommendations on the work-up of neonatal cholestasis. This limitation in the leading diagnostic guidelines should be addressed to better support pediatricians in these complex diagnostic processes.

P 8.

Myeloid Lineage Switching as escape Mechanism to Chimeric Antigen Receptor T-Cell Therapy in Precursor B-ALL with ZNF384-TCF3 fusion: a Case Report

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Outcomes in paediatric precursor B-cell acute lymphoblastic leukaemia (BCP-ALL) are steadily improving in the last decades, with event free survival rates reaching 90% in contemporary clinical trials. Despite the global favourable outcome for the majority of patients, some patients suffer from relapse or refractoriness to conventional intensive chemotherapy regimens. A novel treatment option in relapse/refractory (R/R) patients is immunotherapy targeting the CD19 (cluster of differentiation 19) cell surface protein. Besides Blinatumomab, a bispecific antibody that redirects a patient's T cells to kill CD19-positive cells, also chimeric antigen receptor T-cell (CART) therapy targeting this antigen has recently been approved by the competent authorities. CART cells are engineered to express a CAR, linking an antigen recognition domain with T-cell signalling domains. Upon antigen stimulation, these CART cells produce a cytotoxic T-cell response, killing the antigen-expressing cells. In clinical trials, CART cell therapy resulted in excellent remission rates in R/R B-ALL. However part of the patients treated with CART cells still relapse and different relapse mechanisms have been described. Lineage plasticity and immunophenotypic switching, from a lymphoid to a myeloid immunophenotype, is more rare, although its prevalence is suspected to be higher than reported. Among the subtypes of leukaemia that have a higher risk of lineage switching, Zinc-finger protein 384 – Transcription factor 3 (ZNF384-TCF3) fusions and KMT2A (MLL)-rearrangements are most frequently reported.

We present a case of a seven-year-old girl demonstrating myeloid lineage switch after CD19 CAR T-cell therapy for R/R B-ALL with a ZNF384-TCF3 fusion.

Acute leukaemia with ZNF384-TCF3 fusion is considered high risk in contemporary frontline treatment protocols and will be treated by Hematopoietic Stem Cell Transplantation (HSCT) or Chimeric Antigen Receptor T-cell (CART) treatment in first complete remission.

Although current cytogenetic and molecular work-up of newly diagnosed paediatric acute lymphoblastic leukaemia (ALL) includes the identification of the TCF3-ZNF384 fusion, the importance of including this information in the choice of bridging therapy and the timing of CART treatment, as the high cytokine levels during high grade Cytokine Release Syndrome (CRS) might drive ALL cells to lineage switching, is highlighted by this case.

Short Oral Presentation

SO 9.

Shaken Baby Syndrome: review of a series of 24 cases

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Background/Aims
We propose a review of 24 cases of children with a diagnosis of shaken baby syndrome admitted to the university pediatric service over a period of 12 years.

Methods
We defined the inclusion criteria on the basis of: diagnostic criteria used in the literature, namely multifocal subdural hematomas associated or not with retinal haemorrhages or unifocal subdural hematoma associated with retinal haemorrhages. All clinical situations were taken care of in a multidisciplinary medical and psychosocial way.

Results
Out of 24 children, the sex ratio is 1 and the average age is 3.5 months.

Neonatal factors: 25% (n=6) of children had a twin and two sets of twins had shaken baby syndrome. 37% had stayed in neonatology and 16% were premature < 36 weeks gestational age

Symptoms that led to the diagnosis: 30% had convulsions or even status epilepticus, 35% had severe apnea, 15% had vomiting, 15% a rapid increase in head circumference, 25% associated apnea and convulsions, 5 % were diagnosed following an investigation following the diagnosis made on the twin.

Clinical lesions
60% of the children had plurifocal subdural hematomas associated with retinal haemorrhages, 34% had plurifocal subdural hematomas but no retinal haemorrhages, 6% had unifocal subdural hematomas associated with retinal haemorrhages. 62% of the children were admitted to the intensive care unit and of these 46% benefited from endo-tracheal intubation and invasive ventilation. A neuro-surgical intervention was necessary in 30% of cases. In 33% of cases, bone fractures were found on complete skeletal radiography.

Sequels
41% of children were on antiepileptic treatment three months after discharge from hospital. 12.5% progressed to cerebral palsy, 37.5% developed a developmental delay (attention and learning disorders, language disorders, dysgraphia, dyscalculia, etc.), 21% retained visual sequelae.

Psycho-social factors
Our sample comes from all socio-cultural backgrounds. In 29% of the cases the author admitted the facts during the hospitalization (among these we find 57% of the fathers and 43% of the mothers). A report to the youth assistance services was made in 100% of cases. At the end of the psychosocial assessment, 37.5% of the children had to be placed outside the family environment.

Conclusion

Shaken Baby Syndrome is a complex pathology that is related to child abuse. Its clinical and psychological consequences in the short and long term are considerable.

P 9.

Clinical case: Discovery of Shaken Baby Syndrome in the course of Streptococcus Agalactiae Septicemia

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Background/Aims

Shaken Baby Syndrome (SBS) is an abusive head trauma entity and represents a severe form of child abuse. We describe the clinical case of a 2-month-old child who, in the course of hospitalization for Streptococcus Agalactiae sepsis, presented an unfavorable clinical course leading to the accidental discovery of SBS and detail the clinical and diagnostic elements allowing it to be detected and managed in a multidisciplinary manner.

Methods

This is a male child, the couple's first child, the mother already having 3 other children from previous unions. His antecedents are marked by prematurity at 30 weeks 3 / 7th, late neonatal sepsis at day 6 of birth due to E. Aerogenes with associated meningitis. A brain MRI performed before the child was discharged from the neonatal ward was found to be normal. He is admitted at the age of 2 1/2 months of actual age and 3 weeks of corrected age with a clinical picture of severe malaise in the infant and septic shock. Additional examinations show an Hb at 6.6g / dL, a hematocrit at 27.3%, white blood cells at 2270 / m3, platelets at 137,000 / mm3, a CRP at 75 mg / l. The blood culture will come back positive for Streptococcus Agalactiae, the lumbar puncture will be negative. The child was initially be treated by broad-spectrum antibiotic therapy and then on amoxicillin for 14 days. He evolved well infectiously with rapid defervescence of both temperature and inflammatory syndrome, reflecting a correct response to antibiotic therapy. However, axial and peripheral hypotonia persisedt as well as fluctuating eye tracking. These clinical signs were explored by transfontanellar ultrasound which will highlight images of echogenic subdural collection 3 mm thick on the left and anechoic fluid collection 2 mm thick on the right.

Results

A brain MRI confirmed subdural plurifocal hematomas and a fundus of bilateral retinal haemorrhages diagnosing shaken baby syndrome.

During the assessment, one of the parents will admit having shaken the child violently on several occasions. Clinically and in longitudinal follow-up, the child will develop an epileptic syndrome treated with Valproic Acid as well as a psychomotor delay associating delay in walking and speech, as well as fine motor disorders and will benefit from multidisciplinary follow-up.

Conclusion

Shaken Baby Syndrome is a form of serious child abuse that needs to be recognized and diagnosed in order to initiate prompt medical care and protection for th

P 10.

Acute motor axonal neuropathy with bulbar symptoms- Case report

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Background

Acute motor axonal neuropathy (AMAN) belongs to the axonal variants of Guillain-Barré syndrome (GBS). GBS is a leading cause of acute flaccid paralysis. An autoantibody-mediated immune response underlies axonal degeneration. AMAN is associated with a more severe course than acute inflammatory demyelinating polyradiculoneuropathy (AIDP) but responds more favourably to IVIg.

Methods

Herein we present an AMAN case with bulbar symptoms.

Results

A 14 year old boy was brought to the emergency room with reduced strength, mainly in lower limbs. Initially mainly noticeable at the left side. Furthermore, he choked during the feeding. On physical examination, he couldn't walk on his toes and the Gowers's sign was positive. There was no loss of sensibility, reflexes were intact, good sphincter control and cardiorespiratory stable. Cerebrospinal fluid protein level was normal and no cells were detected. Stool culture was negative for Campylobacter jejuni. Viral serology was negative. Anti-ganglioside antibody determination was not possible. Toxicology screening was negative. Magnetic resonance of the brain and myelum showed no abnormalities. Electromyography was consistent with a motor axonal polyneuropathy of the four limbs. Initial treatment regime was 400 mg/kg/day intravenous immunoglobulin (Privigen) for five days. Follow-up two weeks later showed regression of all symptoms.

Conclusion

Axonal GBS is an important variant of classical GBS. AMAN is a pure motor variant without involvement of the sensory or autonomic nerves. Often characterized with a more severe course. IVIg should be considered because of the underlying hyperreactive humoral response.

Key words

Acute severe motor axonal neuropathy- Guillain-Barré syndrome- Paralysis- Bulbar- Paediatric- Case report

P 11.

A peculiar gait for a 5-year-old child

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Introduction

Abnormal gait is a frequent complaint among children and can occur in many conditions, which are usually benign but can sometimes be more serious.

Peculiar gait in children should always be assessed, especially when persisting or worsening.

Clinical case

5-year-old boy presenting at the emergency room for right knee pain with slight limp for 2 days after falling on his knees.

His medical background includes chronic diarrhea and an upper respiratory tract infection within the last month.

Clinical examination shows little pain when mobilising the right knee without local sign of inflammation. The remaining physical examination is normal.

A radiography of the right leg comes back normal.

Given the story and the clinical exam, the patient is discharged with nonsteroidal anti-inflammatory drugs and instructions of physical rest. A clinical reassessment is scheduled 48h later.

He comes back 2 days later with an abnormal gait and random bouts of pain in both legs and lower back not responding to level I analgesics. There is no fever or other symptoms.

Clinical exam now reveals a clear ataxia, weakened muscle stretch reflexes in the lower limbs without strength loss, adiadochokinesia nor dysmetria. The cranial nerves testing is normal.

Suggested differential diagnoses are: posterior cord syndrome (due to spinal cord compression), viral myositis, Guillain-Barré syndrome (GBS).

An initial assessment includes a normal blood test with normal creatine phosphokinase, and a negative nasopharyngeal swab for viruses such as SARS-CoV2, influenza A/B.

A cerebral CT scan is performed before the lumbar puncture, showing carotid-jugular and spinal lymphadenopathies. The cerebrospinal fluid analysis reveals hyperproteinorachia at 1320 mg/dL with albuminocytological dissociation, evocative of a GBS. The electroneurography shows a demyelinating sensorimotor polyneuropathy, confirming the diagnosis.

During the hospitalisation, the patient develops a symmetric ascending weakness with abolished reflexes and nocturnal neuropathic pain.

The treatment consists of intravenous immune globulin at 2g/kg for 5 days, level I-II analgesics in association with clonazepam and carbamazepine.

The management also includes physical therapy, regular follow-ups and social services involvement.

Conclusion
GBS is a rare acute immune-mediated polyneuropathy but needs to be part of the differential diagnosis of rapid-onset abnormal gait.

Keywords : neuropediatrics, Guillain-Barré syndrome

P 12.

Febrile focal status epilepticus as a presentation of Sturge-Weber syndrome type 3

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Background

A 18 month old infant without previous medical history, presented with a status epilepticus. Seizures started at home with right-sided clonic convulsions with secondary generalization. The seizure could be aborted with a single dose of intravenous (IV) lorazepam. No fever was noted.

At the emergency department, she developed again clonic convulsions but now on the left side, and had a temperature of 39.3ºC. Intravenous lorazepam, a loading dose of 40 mg/kg IV levetiracetam and 20 mg/kg IV phenytoin were needed to stop these recurrent convulsions. The status epilepticus lasted 40 minutes. Airway, breathing and circulation were never compromised during the status epilepticus.

Evolution

A CT of the brain showed (sub)cortical calcifications in the left occipital region. Broad spectrum antibiotics and acyclovir were started after the lumbar puncture. Levetiracetam 20mg/kg/day was continued as maintenance therapy. She was admitted to the Paediatric Intensive Care Unit for close monitoring.

During hospitalization, no new seizures were observed. Lumbar puncture remained negative and antimicrobial therapy could be discontinued. She had no new episodes of fever during her admission. A brain MRI was performed which confirmed the cortical calcifications and showed leptomeningeal angiomatosis in the left occipital zone suggestive of Sturge-Weber syndrome (SWS) type III. There were no signs of SWS on ocular examination.

Currently, she has a normal psychomotor development.

Conclusion

- SWS is a congenital neurocutaneous disorder and is characterized by a port-wine stain, leptomeningeal angiomatosis and ophthalmic abnormalities. SWS can be classified according to the presence/absence of facial and leptomeningeal angiomas, and ophthalmic anomalies.
- Radiographic features (CT and MRI) of SWS are superficial cerebral calcifications, hypertrophy of choroid plexus, cerebral atrophy and leptomeningeal enhancement.
- SWS type III is a rare disease and is characterized with isolated leptomeningeal angioma.

P 13.

Unusual presentation of idiopathic intracranial hypertension in an 11 year-old girl.

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Jessa ziekenhuis Hasselt

Background and aims

Idiopathic intracranial hypertension (IIH) is a rare diagnosis in children with an estimated annual incidence around 1 per 100.000 children. IIH is a disorder defined by clinical criteria that include symptoms and signs isolated to those produced by increased intracranial pressure (ICP) (eg, headache, papilledema, vision loss), elevated ICP with normal cerebrospinal fluid (CSF) composition, and no other cause of intracranial hypertension evident on neuroimaging or other evaluations. It can present with a variation of symptoms. Cranial nerve function can be impaired in IIH and patients with IIH may report intermittent or continuous horizontal diplopia. This is typically due to a unilateral or bilateral sixth cranial nerve palsy. Other nerves are usually unaffected.

Methods

case-report

Results

We report on an 11 year-old female presenting with diplopia and right sided shoulder and neck pain since two weeks. She also complained of muscle weakness and numbness in the right hand. There was no history of head trauma and no medication use. Physical examination revealed muscle weakness of the right hand and binocular diplopia. Ophtalmology was consulted and she was found to have bilateral papilledema and sixth nerve palsy right > left. MRI showed bilateral papilledema and signs of elevated ICP without evidence of hydrocephalus or an intracranial space occupying lesion. A lumbar puncture was done which revealed an opening pressure of 350 mm of water. CSF analysis showed a cell count of < 3/µl, CSF protein of 190 mg/dL, and CSF glucose of 50 mg/dL. Lab analyses showed low infection parameters and negative infectious serology. All cultures remained sterile. Screening for auto-immune disease came back negative. During lumbar puncture a volume of 15 ml cerebrospinal fluid was evacuated. A course of Acetazolamide was started. After the lumbar puncture and initiation of pharmacological treatment, neurological complaints fully recovered.

Conclusion

The patiënt described in this case-report didn't present with the classical symptom of headache, which is present in 90% of cases. The muscle weakness and numbness are not usually seen in IIH. According to some definitions of IIH (Modified Dandy criteria) there should be no localising symptoms, with the exception of a sixth nerve palsy. Shoulder pain is also not a classical symptom. Since all symptoms recovered fully after starting treatment, we considered IIH still to be the diagnosis in this case.

P 14.

Malignant hyperthermia in child with Williams Beuren syndrome

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Background

Malignant hyperthermia (MH) is an autosomal dominant pharmacogenetic disorder characterized by hypermetabolic crisis occurring when a predisposed patient (mainly patients with skeletal muscle disorder) is exposed to triggering agent: depolarizing muscle relaxant or a volatile inhalational agent.

Williams Beuren syndrome (WBS) is caused by the deletion of genes on chromosome 7q11.23. It is characterized by supravalvular aortic stenosis, distinctive facial features, hypercalcemia and neurodevelopmental deficit.

Case report

We report the case of an 11-year-old child with WBS, hospitalized to receive dental care under general anesthesia. During the anesthesia by halogen derivatives and curare, hypercapnia and involuntary contraction of the masseters occurred. Temperature was not measured at the time. In the immediate post-operative period, urine were red. Urinary sediment showed an absence of red blood cells. Completion of biology showed mild renal failure and moderate hepatic cytolysis in a context of acute rhabdomyolysis (Myoglobinemia > 120,000 mcg/L, CK at 122,286 UI/L). Medical management consists of intravenous hyperhydration, administration of Dantrolene and urine's alkalization. Evolution was favorable. A search for myopathies and a genetic panel are in process.

Discussion

The particularity is that the child had 3 times previous anesthetic exposures, all of which were uneventful. Recent review of literature suggests that MH occurs as a spectrum varying from mild asymptomatic to fulminant MH with sinus tachycardia, hypercarbia, rapid temperature increase, muscle rigidity, dark colored urine, tachypnea and hyperkalemia. As described in the literature for patient of his age category, our patient symptomatology was incomplete which led to a delay in diagnosis.

To our knowledge, this case is the fourth report of MH in patient with WBS. Several factors can potentially support this association: a) genetic mutations are shown to be closely associated with some form of MH susceptibility (17q11.2-q24, 3q13.1, 5p, and notably 7q21-q22); b) disturbance of calcium homeostasis are frequent in WBS patients; c) abnormalities of the musculoskeletal system have been reported in WBS patients.

Conclusion

This report underlines the need of vigilance for this anesthetic complication in WBS patients and for the potential incomplete presentation in pediatric population in general.

P 15.

Central Apneas in Rett Syndrome

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Introduction

Rett syndrome is a progressive neurodevelopmental disorder associated with sleep disorders that needs to be detected.

Clinical case

A 7-year-old female patient with Rett syndrome presented to the sleep consultation for sleep disorders, with numerous apneas, fragmented sleep during the night and daytime sleepiness. A polysomnography performed after the consultation shows the presence of numerous central apneas (960 central apneas, 2 obstructive apneas and 0 obstructive hypopneas) accompanied at each one by a desaturation of >3%, which corresponds to an apnea-hypopnea index of 102.9/h. The sleep structure was also disturbed with less REM sleep. Mean saturation was 94.8%. The transcutaneous PCO2 measurement was normal. After discussion, it was decided to start chronic oxygen therapy at 1L/minute. The child's general condition improved with a clear improvement in daytime awakening and interaction. The polysomnography control under oxygen therapy shows no more central apneas and only one obstructive apnea with an apnea-hypopnea index of 0.3/h.

Discussion

Our patient presents a typical Rett syndrome polysomnography with a relative increase in N3 sleep, a decrease in N2 and REM sleep time, a decrease in the sleep efficiency index and central sleep apneas. These apneas seem to be linked to a dysfunction of the central chemoreceptors in the brainstem resulting in an instability of central respiratory drive. While the use of non-invasive ventilation by BIPAP is usually recommended for patients with central apneas, it has been shown that the use of oxygen therapy alone can reduce the apneas in Rett patients. That’s what we observed in our patient who saw her apnea-hypopnea index decreased drastically under oxygen therapy. Positive repercussions were also observed on her sleep efficiency index. However, no impact was observed on the distribution of the sleep phases.

Conclusion

Patients with Rett syndrome have a characteristic sleep stage architecture and may present with apneas of central origin of varying severity. Oxygen therapy is an alternative to non-invasive ventilation as first-line treatment for these apneas.

P 16.

Severe hypotonia and developmental delay due to an EBF3 pathogenic variant : clinical implications of a molecular defect.

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ULiège

Hypotonia Ataxia and Delayed Development Syndrome (HADDS) is a neuro-developmental syndrome due to missense pathogenic variants of the EBF3 gene located on chromosome 10q26.3. In most cases, these variants appear de novo and the transmission is autosomal dominant. HADDS would concern around 200 people around the world.

Case

A Caucasian girl was born at 36 weeks with a slight respiratory distress. Her evolution was marked by major hypotonia, slow weight gain and significant failure to thrive. She also presented a grade II left vesicoureteral reflux, a pathological gastroesophageal reflux associated with severe eating difficulties, a chronic constipation and a right convergent strabismus. Some dysmorphia with a broad forehead and low implanted ears were reported. Despite a multidisciplinary management, her evolution remains marked by a severe hypotonia and a significant psychomotor delay leading to perform a whole exome sequencing. A de novo missense EBF3 pathogenic variant c.626G>A (p.Arg209Gln) was identified.

Discussion

HADDS is characterized by a quite clinical variability in the different symptoms : major hypotonia, failure to thrive, psychomotor delay, digestive and eating disorders, vesicoureteral abnormalities, strabismus, and moderate facial dysmorphia. Although our knowledge is still limited, the significance of these symptoms seems to rely upon the EBF3 expression in different tissues during embryogenesis. Based on animal studies, EBF3 appears to play a critical role in neurogenesis and neuronal migration. It interacts with CDKN1A, NEUROD and ARX regulation pathways. Regarding to diaphragmatic and vesicoureteral dysfunction as well as hypotonia, EBF3 seems to be involved in the myocytes calcium metabolism. Further studies with patient's muscular biopsies would be useful to improve the physio-pathological understanding. Furthermore, EBF3 has been recently reported as a novel tumor suppressor gene in some cancers. Until now, no previous reported HADDS cases have been described with an oncological disease.

Conclusion

HADDS is an extremely rare genetic syndrome caused by EBF3 pathogenic variants. It is primarily characterized by a severe hypotonia and developmental delay. Other clinical signs have been reported with a large inter-individual variability. Further research on the EBF3 gene and the associated pathological pathways are still needed to improve our understanding of HADDS and to offer appropriate care in such rare diseases.

P 17.

CD59 deficiency: a very rare, potentially curable form of infection induced neuromotor regression

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UZ Gent

Whole exome (WES) based gene panels caused a revolution in the diagnosis of rare genetic diseases. However, for the correct evaluation of found variants multidisciplinary interaction between geneticists, biochemists and clinicians remains of uttermost importance.

A 2,5-year-old girl presented to the outpatient clinic because of progressive walking problems. She was born as the first child of healthy consanguineous parents. The pregnancy was uneventful, and she had a birth weight of 2600 grams. Her early motor development was normal until the age of 15 months when she developed a viral upper airway infection with fever. When she recovered from the infection the parents noted that she lost the ability to stand without support. On clinical examination in a peripheral hospital ataxia was noted and the diagnosis of post viral cerebellar ataxia was made. She was treated with corticoids and intensive physiotherapy. However, this only brought minor improvements.

On first presentation at the pediatric neurology, we saw an active and reactive toddler with a wobbly gait and slow movements of the arms. She could only walk a few steps without support. Rossolimo's and Babinsky's signs were present but Achilles and patellar reflexes were rather weak. Krabbe and metachromatic leukodystrophy were excluded enzymatically. MRI of the brain and the spine were normal as well as a routine metabolic screening. Genetic testing with an inhouse Ataxia Spasticity gene panel returned normal. Yet, on reanalysis of the WES data a homozygous frameshift variant was found in CD59. CD59 can prevent C9 from polymerizing and forming the complement membrane attack complex. CD59 deficiency is known to cause the very rare hemolytic anemia with immune-mediated polyneuropathy, with neuromotor regression typically occurring during infections. The patient never suffered from hemolytic crises, but increased reticulocyte count and decreased haptoglobin were present on routine blood examination. Additionally, cytoflow demonstrated a complete absence of CD59 on the surface of red blood cells and thus conforming the pathogenetic nature of the mutations. Eculizumab is a humanized antibody that inhibits the terminal pathway of complement by blocking the activation of C5. Several case reports suggested that eculizumab could ameliorate neuromotor function in patients with CD59 deficiency. After consent of the parents, eculizumab was started and her neuromotor function is followed up closely.

P 18.

Juvenile ALS with crystalline retinopathy caused by a de novo mutation in SPTLC2 causing a shift in substrate specificity of Serine Palmitoyl Transfer

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An eight-year-old girl presented to the clinic with slowly progressive spasticity of the lower limbs. On clinical examination she had normal intelligence and no dysmorphism. She was small for her age. She had a spastic diparesis, a low amplitude tremor in the hands and fasciculations of the tongue. MRI of the brain and the spine were normal but on fundoscopy a crystalline retinopathy was observed. Optical Coherence Tomography (OCT) showed thinning of the retina with loss of Müller cells and presence of small cysts.

By trio-WES we found a de novo mutation in SPTLC2 coding for a subunit of Serine Palmitoyl Transferase (SPT). SPT catalyzes the first and rate limiting step of ceramide -and thus of sphingolipid-synthesis by joining palmitoyl-CoA and serine. SPT consists of 4 subunits coded by SPTLC1 and SPTLC2/3, SPTSSA/B and ORMDL3. Mutations in SPTLC2 were previously described as a cause for hereditary sensory and autonomic neuropathy (HSAN) IC, where a shift in substrate specificity from serine to alanine leads to accumulation of the neurotoxic deoxysphinganine (DoxSA). Recently mutations in SPTLC1 were described in juvenile ALS without crystalline retinopathy and linked to sphingolipid overproduction. Similar to our case a combination of retinopathy and neurological regression is seen in the Stellar mouse. In this mouse model a missense mutation in Sptssb causes excess synthesis of sphingolipids with a 20-carbon long chain bases (LCB). Unlike patients with HSAN IC, our patient did not have an increase of DoxSA in plasma. Nonetheless, an increase in desoxysphinganine (DosSA) and desoxysphingosine (DosSO) (downstream products of glycine+palmitoyl) were seen. Moreover, similar to the Stellar mouse, the 20-carbon LCB forms of GlcCer, LacCer and Gb3 were increased.

We hypothesize that the SPTLC2 mutation in our patient causes a shift in substrate specificity of SPT towards longer chain acyl-CoA's, explaining the difference in clinical symptoms with HSAN and juvenile ALS.

LO 12.

Short term follow-up after cardiac dysfunction in pediatric inflammatory multisystem syndrome associated with COVID-19 infections

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Background/Aims

Pediatric inflammatory multisystem syndrome (PIMS) is a new disease first described in April 2020, which occurs weeks after COVID 19 infections. The syndrome is caused by an abnormal immune system response secondary to the infection and manifests as impairment of several organ system including the heart. We report our single center experience of the short term follow-up of patients who had cardiogenic- and /or distributive shock in the context of PIMS.

Methods

From September 2020 to January 2022, we enrolled prospectively and followed-up 15 patients diagnosed with a PIMS and who had hemodynamic shocks at their initial presentation. During the acute phase, we collected clinical, biological and cardiological data. Patients were assessed regularly: 5 had a whole year follow up, 5 were followed up for 3 months, 1 was lost of follow up before the 3 months evaluation, and 4 were recently diagnosed with no follow up yet. The 3 months evaluation included a cardiac MRI, a complete functional echocardiography and a physiological evaluation with ergospirometry.

Results

All out of the 15 patients studied had high CRP levels, 80% had elevated troponin levels and 92.9% elevated Pro-BNP levels. Initially, all except one had echocardiographic signs of systolic and diastolic ventricular dysfunction, 46.7% had ECG anomaly and 73.3% other cardiological findings (coronary dilation, pericardial effusion). At 3 months follow up, no patient had residual symptoms, 9% had remaining ECGs anomalies, 18.1 % had slightly abnormal echocardiograms with marginally altered GLS, and cardiac MRIs were normal. 62.5% of the patients had VO2max values that were superior to P90 of the expected value.

Conclusion

Myocardial damage is one the main characteristics of PIMS, the majority of patients being admitted in PICU with hemodynamic instability and left ventricular dysfunction. Our report seems to confirm recent data that suggest that the majority of patients have a complete recovery in a few weeks after PIMS. This completely new disease needs to be further investigated in long term follow-up protocols in order to give accurate advice to patients and pediatricians on how to manage exercise limitation in this population.

Short Oral Presentation

SO 18.

QTc intervals are not prolonged in former ELBW infants at pre-adolescent age.

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Background
Whether preterm birth is associated with cardiac conduction or repolarization abnormalities in later life is still poorly explored, with conflicting data on QTc prolongation in former extreme low birth weight (ELBW, <1000 g) infants.

Methods
Twelve lead electrocardiograms (ECG) at rest, collected in the PREMATurity as predictor of children's Cardiovascular-renal Health (PREMATCH) study in former ELBW cases and term controls during pre-adolescence (8-14 years) were analysed on corrected QT time (QTc, Bazett) and QT dispersion (QTd). ECG findings were compared between groups (Mann-Whitney), and associations with clinical and biochemical findings were explored (Spearman). In ELBW cases, associations between QTc and perinatal characteristics (at birth, neonatal stay) were explored (Mann-Whitney, Spearman).

Results
QTc and QTd were similar between 93 ELBW cases and 87 controls [409 (range 360-465) versus 409 (337-460); 40 (0-100) versus 39 (0-110)] ms. Age, height, weight or body mass index were not associated with the QTc interval, while female sex (median QTc was 415 to 401 median difference 11.4 ms, p=0.001) and lower potassium (r=-0.26,95% -0.41 to -0.09, p=0.003) were associated with longer QTc interval. We could not observe any significant association between QTc interval and perinatal characteristics.

Conclusions
There were no differences in QTc or QTd between ELBW and term controls in ECGs at rest in pre-adolescents.

SO 19.

Cardiac isomerism: a 20 year retrospective cohort at a single institution

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Background and Aim

Cardiac isomerism or 'isomerism of the atrial appendage' is found in about 0,4-2% of congenital heart defects and is associated with a complex spectrum of anomalies. We reviewed isomerism cases from the last 20 years at a single institution.

Methods

Retrospective review of 48 patient records (Jan 2000-Dec 2020). Case selection was based on echocardiographic findings suggesting cardiac isomerism. Definitive diagnosis was made post-mortem or peri-operatively by identifying symmetrical morphological left or right atrial appendages.

Results

Left atrial isomerism (LAI) was diagnosed in 24 (10 confirmed), right atrial isomerism (RAI) in 24 cases (18 confirmed). Prenatal diagnosis was made in 33 cases (17 LAI; 16 RAI). Unconfirmed cases consist of 5 terminations of pregnancy were no autopsy was performed and 15 live-born patients who did not undergo cardiac surgery or autopsy.

In LAI, 21 cases (87,5%) have anomalous systemic venous return, being the only cardiovascular abnormality present in 10 patients. Conduction or rhythm abnormalities were present in 11 cases (45,8%). In RAI, only complex univentricular heart defects were found with a large proportion of AVSD, transposition of the great arteries and pulmonary stenosis/atresia. Splenic function is disturbed in 86% of all live-born patients. While other extra-cardiac defects are more incidental, malrotation is more prevalent in this cohort (>10%) than in the general population.

After exclusion of termination of pregnancy (n=11) and patients lost to follow-up (n=2), overall survival for isomerism patients is 60%, with a median time to follow-up of 9 years. Compared to RAI, survival rates in patients with LAI are much higher: 84% (median follow-up 6yrs) compared to 31% (median follow-up 12yrs). However, for those LAI patients that need univentricular palliation (n=3), survival rates are low (33%) and similar to RAI. Main causes of death are primary cardiac failure, abstinence of life-prolonging care and severe sepsis.

Conclusion

Survival rates for patients with cardiac isomerism are low for those who need univentricular palliation. These results can be used when counseling parents. Additional pathology like (functional) asplenia and malrotation add to the burden of disease.

Short Oral Presentation

SO 20.

Right atrial myocardial remodeling in children with congenital cardiac defect depends on the type of hemodynamic load

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Introduction

Myocardial remodeling in infants with congenital cardiac defect involves expression of inflammatory cytokines, growth factors and regulators of apoptosis and fibrosis, respectively.

We aimed at investigating the role of different forms of hemodynamic overload on this process.

Methods

Concentrations of gene mRNA of the early stress response (HSP-70), inflammation (IL-1β, IL-6, IL-10), growth (IGF-β, CT-1, VEGF), apoptosis (Bcl-xL, Bak), and fibrosis (TGF-β, collagen) were measured in the right atrial myocardium (RA) of 13 children. Eight had ventricular septum defect (VSD) with congestive heart failure but without volume overload of the RA and 5 had atrial septum defect (ASD) with volume overload of RA (n=5).

Results

Patients with VSD had significantly higher RA-expression of mRNA coding for IL-1β ($p<0,0001$) and HSP-70 ($p<0,01$) but lower expression of BcL-XL ($p<0,05$) than patients with ASD. Expression of CT-1-mRNA tended to be lower in patients with VSD than in those with ASD ($p<0,1$).

Conclusion

Our results suggest that in patients with VSD and heart failure, RA-myocardial remodeling involves inflammatory- and early stress gene response due to the sustained systemic inflammatory reaction related to congestive heart failure whereas in patients with ASD genes involved in the inhibition of apotosis and growth predominate, probably as a consequence of RA distention.

This observation confirms differential myocardial remodeling in children with congenital cardiac defect depending on the type of hemodynamical load suggesting the role of local stretch stimuli and of circulating mediators.

SO 21.

Recovery kinetics of gas exchange parameters and heart rate after maximal exercise in children with repaired Coarctatio Aortae compared to controls

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Background and aim

Children after succesful repair of Coarctatio Aortae (rCoA) still demonstrate a decreased exercise capacity. Although the physiological processes during exercise are thoroughly investigated, little research exists on the recovery after exercise. In this study we evaluate the recovery kinetics of VO2, VCO2 and HR after maximal exercise in children with rCoA compared to healthy controls.

Methods

65 children after rCoA and 65 matched controls performed a maximal cardiopulmonary exercise test. In the subsequent 6 minute recovery period, kinetics of VO2, VCO2 and HR were analysed. The half-life time (T1/2) of the exponential decay and the patterns of percentages drop per minute were compared between groups.

Results

Exercise performance was reduced in the rCoA group with lower VO2peak (40.0 ± 7.7 vs. 41.1 ± 8.8 ml/min/kg; p=0.012) and lower percentages of predicted value of VO2peak (89.0 ± 18.4 vs. 100.3 ± 13.7 %; p=0.002) and maximal load (79.8 ± 19.9 vs. 93.0 ± 18.0 %; p=0.004). Maximal HR (183 ± 15 vs 190 ± 12 bpm; p=0.003) was also lower in the rCoA patients.

The recovery kinetics of VO2 and VCO2 was faster in rCoA patients with lower T1/2 values compared to controls (T1/2VO2: 38.1 ± 11.8 vs. 44.9 ± 9.4 s; p<0.001 and T1/2VCO2: 55.5 ± 15.2 vs. 61.8 ± 10.9 s; p<0.001). Examining the patterns of percentages drop revealed a significant interaction (p<0.001) between group and time, indicating that the presence of rCoA altered the recovery course.

T1/2HR was lower in the rCoA group (54.1 ± 21.4 vs. 68.3 ± 20.7 s; p<0.001), demonstrating faster HR recovery. The values of percentages drop were also higher in the rCoA group. Looking at the recovery pattern, a significant effect of group as well as interaction of group and time (p<0.001) was found, confirming the faster recovery in the rCoA patients.

Conclusion

Despite a lower exercise tolerance, rCoA patients exhibit faster recovery kinetics of VO2, VCO2 and HR after maximal exercise. An altered oxygen supply-demand balance might induce a higher local muscle metabolism leading to faster recovery. Further research will need to clarify the underpinning mechanisms leading to faster recovery in rCoA patients.

Short Oral Presentation

SO 22.

Evaluation of motor competence and physical activity of children and adolescents with a univentricular heart

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Background and aim
Physical activity (PA) and fitness are important predictors of health in later life. To achieve a satisfactory level of activity and fitness, one is dependent on adequate motor competence (MC), i.e., the degree of proficiency in performing a wide array of motor skills as well as the underlying processes such as coordination, control, and quality of movement. Development of MC requires dedicated practice and active exploration, preferably in organized sports (or PE) settings. Given their medical history and possible risk of overprotection by the environment, individuals with a univentricular heart (UVH) may be at risk of reduced MC. The current study set out to examine MC, PA and level of intrinsic motivation towards sports in a sample of children and adolescents with a UVH.

Methods
Twenty participants with UVH (mean age: 10.6, range: 4-17), monitored by the Pediatric Cardiology unit of the Ghent University Hospital, were assessed with the Movement Assessment Battery for Children 2 (MABC2) and surveyed with the Flemish Physical Activity Questionnaire (FPAQ) and an age-appropriate version of the Behavioral Regulation in Exercise Questionnaire (BREQ). The results of the MABC2 were compared against the published reference values, those of the FPAQ and BREQ with an age-matched control sample of 104 children from our own database.

Results
The average MABC2 score of the participants with UVH was at the 23rd percentile (range: 0.1-75). In 13 of them (60%), motor competence was “at risk of a motor problem”. Furthermore, the degree of PA (32 minutes/day, range: 5-70) was lower than the control group (63 minutes/day, range: 11-195; $t(120)=3.604$, $p<0.001$). Only 2 participants with UVH (10%) met the WHO guideline of 60 minutes PA per day compared with 50% of the control group. Finally, intrinsic motivation towards sport was relatively high in the UVH group (3.75 on a 5-point-scale, range: 2-5), however lower than in the control group (4.20, range: 0-5, $t(119)=2.074$, $p=0.04$).

Conclusions
In view of the role of motor competence in ensuring a healthy lifestyle, actions are recommended to promote motor development and physical activity in children and adolescents with UVH.

SO 23.

Diagnosis of severe congenital heart defects in 2 consecutive periods: increased prenatal detection rate but similar mortality rate

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Aim
to determine the actual prenatal detection and mortality rate in patients with severe congenital heart diseases (CHD) and to compare these data with an earlier study period (2006-2014).

Methods
single center retrospective study in patients with severe CHD diagnosed prenatally or postnatally between 2015 and 2020.

Results
a total of 379 patients were included. Univentricular heart (20%), coarctation of the aorta (18%), transposition of the great arteries (17%) and tetralogy of Fallot (15%) were the most prevalent. 194 diagnoses (52%) were made prenatally, with large differences among CHD types. Prenatal detection rate was the highest for univentriculair heart defects (85%) and the lowest for coarctation (15%).

Overall mortality rate was 30% mostly due to prenatal mortality including termination of pregnancy (TOP; 48%) and postnatal compassionate care (17%). In the group who underwent surgery, mortality rate was 8% (<30d postop: 5%). Postnatal mortality was higher in patients born after prenatal diagnosis than in those with a postnatal diagnosis (32% vs 6%; p<0.001).

Compared to the earlier study period, the prenatal detection ratio increased from 30% to 52% (p<0.001) with a marked improvement in the prenatal detection of tetralogy of Fallot (23% vs 40%) and transposition of the great arteries (24% vs 45%). Mortality rate remained similar both for overall and postnatal mortality. In the prenatal diagnosis group, TOP rate dropped from 41 to 29% (p=0.001).

A molecular diagnosis was found in 18% of the patients that were genetically tested (N=296). In patients born alive, additional abnormalities were found in the gastro-intestinal tract (4%), the airway tract (2%), the renal system (6%) or the nervous system with or without neurodevelopmental problems (11%).

Conclusion
prenatal diagnosis of CHD has improved significantly in the last years. Despite this improvement, overall mortality remained similar with major contributors still being TOP and compassionate care. Genetic abnormalities and extra-cardiac pathology were present in an important amount of patients indicating that prenatal and postnatal counseling should include the possibility of having an underlying genetic disorder or associated extra-cardiac defects which could impact the overall outcome.

Short Oral Presentation

SO 24.

Differential secretion of PF4 and platelet derived TGF-β by in children with congenital cardiac defects

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Introduction

Platelet-derived TGF-β, a cytokine regulating fibrosis, has been implicated in cardiac remodeling and cardiac dysfunction secondary to ventricular pressure overload. The role of platelet-derived TGF-β in myocardial remodeling in children with congenital cardiac defect has not been investigated yet. Therefore, we aimed to investigate platelet secretion of TGF-β in children with congenital cardiac disease and pressure overload of the right ventricle.

Methods

Ex vivo activation of thrombocytes was performed in 11 infants with large ventricular septum defect (VSD) (n= 5) or tetralogy of Fallot (TOF) (n= 6). Ten age-matched healthy children in whom blood was withdrawn before scheduled INT surgery served as controls. Concentrations of TGF-β and PF4, a marker of platelet activation, were measured in the culture supernatant. TGF-β-thrombocyte production was assessed by TGF-β/PF4-ratio.

Results

Control subjects showed the highest TGF-β/PF4 (3.9 ± 0,7 (mean ± SEM)), in comparison to patients with VSD (2,6 ± 1,17) and patients with Tetralogy of Fallot (2,52 ± 0,69). There was no significant difference between both patient groups but between patients and controls (p= 0.0014).

Conclusions

Our results show differential secretion of TGF-β in children with congenital cardiac defects in comparison to healthy children. Lower TGF-β/PF4 ratio in patients than in healthy controls suggests exhaustion of TGF-β secretion that might be secondary to chronic platelet stimulation by intra-cardiac turbulent flow and might impact the mechanisms of myocardial remodeling in this context of congenital cardiac disease.

P 70.

A late diagnosis of vascular ring : Right Sided Aortic Arch with Kommerell Diverticulum and Aberrant Left Subclavian Artery in a 17 Year Old Girl

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Background

Right-sided aortic arch with Kommerell diverticulum and aberrant left subclavian artery is a rare congenital malformation. It may lead to a symptomatic compression of the esophagus and/or the trachea whereas it is often associated with atypical clinical presentation that delays diagnosis.

Methods and results

A 17 year-old girl treated for repeated ear-nose-throat infections and gastro-esophageal reflux since early infancy complained about exertional dyspnea, chronic cough with expectoration and dysphagia. Apart of a severe obesity the clinical examination was normal. A chest computed tomography angiography was performed to exclude bronchiectasis that incidentally showed right-sided aortic arch with a Kommerell diverticulum and aberrant retro-esophageal left subclavian artery. Barium swallow confirmed a posterior esophageal obstruction of about 30%. Tracheo-bronchoscopy was normal.

The patient underwent resection of the Kommerell diverticulum and of the ligamentum arteriosum and anastomosis between the left subclavian artery and the ascending aorta by interposition of a 8 mm GORETEX® prosthesis. The patient mentioned immediate relief of dysphagia.

Right-sided aortic arch with Kommerell diverticulum and aberrant left subclavian artery is a rare congenital malformation with a prevalence of up to 0,5%. It results from the persistence of the right fourth branchial arch and incomplete regression of the fourth pharyngeal arch artery. The Kommerell diverticulum is the aneuvrysmatic origin of the aberrant left subclavian artery. Kommerell diverticulum, left aberrant retro-esophageal subclavian artery and ligamentum arteriosum create a complete vascular ring that may compress esophagus and trachea.

Conclusion

This case illustrate well that in presence of a right aortic arch easily diagnosed at echocardiography and respiratory or digestive symptoms, the possibility of a vascular ring must prompt to identify aortic arch vessels by computed tomography angiography.

P 71.

Pulmonary embolization of a floating pedunculated tricuspid valve thrombus in a 2 ½ months old infant

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Background

Right intracardiac thrombi are uncommon but known complications of central venous catheterization in children. Whereas they usually respond to anticoagulation, indication for thrombectomy may be discussed depending on thrombus size and -mobility.

Clinical Case

A 2 ½ months old infant with RSV Bronchiolitis required invasive ventilation. A subclavian catheter was inserted that was complicated by a subclavian vein thrombus. Echocardiography performed at day 5 post-admission showed incidentally a pedunculated thrombus (6x5 mm) on the anterior leaflet of the tricuspid valve that swung across the valve with the heart beats but without any valve dysfunction. Wide spectrum AB-therapy was given while several blood cultures returned negative. Anti-coagulation therapy (iv heparin then sc enoxaparin) was immediately started and sub-clavian thrombus resolved. Regular echocardiographic controls showed growth of the hypermobile tricuspid thrombus (14,5X6 mm) despite effective anticoagulation. Thrombectomy was therefore planned. However, at day 25 post-admission, thrombus embolized in the lungs with a fragment (6x3mm) identified in the left pulmonary artery by echocardiography.

The patient had stayed asymptomatic without any sign of pulmonary hypertension. Lung CT performed at day 26 post-admission was normal. Anti-coagulation was switched from enoxaparin to rivaroxaban (Xarelto®) for 1 month.

Discussion

Right intracardiac thrombus is a rare and potential severe complication of central venous catheterization in infants. The indication of anti-coagulation is always given as first line treatment in accordance to the current guidelines. There is no evidence-based recommendation for thrombectomy in the absence of response to anticoagulation, the decision being guided on individual basis and depending on thrombus size and -mobility.

Conclusion

Our case illustrates a delayed positive outcome in an infant with iatrogenic large floating pedunculated and hypermobile tricuspid valve thrombus secondary to a central venous line that initially did not respond to anti-coagulation.

P 72.

Atrial fibrillation in the pediatric ward: rarely a benign condition

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Atrial fibrillation (AF) is uncommon in children in absence of congenital heart disease. Epidemiological data are lacking. The presence of AF in a young patient requires careful etiological work-up.

A 15-year-old girl was hospitalized for a very recent diagnosis of diffuse large B cell lymphoma. She presented suddenly with thoracic oppression plaints. Monitoring showed a heart rate varying between 200 and 220bpm with normal blood pressure. On ECG, AF was diagnosed. Previously, she underwent TTE that showed a small atrial septum defect, normal biventricular dimensions and function. Previous ECG was normal, particularly no pre-excitation or early repolarisation. Blood gas excluded ionic perturbations. Given the hemodynamic tolerance, she was charged with Amiodarone 800mg/m² orally. After 6h, heart rate dropped to 180bpm but there was still no sinus rhythm. She developed pallor, hepatomegaly, blood pressure dropped to 79/43mmHg. TTE showed normal function, completed with a transesophageal echocardiography in order to rule out intracardiac thrombus. We proceeded to electrical conversion, which was obtained after one shock of 1J/kg. Cardiac MRI showed tumoral invasion in the lateral wall of the left atrium.

In contrast to adults, with left atrium (LA) dilation and myocardial fibrosis causing LA dysfunction and electromechanical conduction delay, the substrate for AF in children is often different. AF can be an early manifestation of a cardiomyopathy or unrecognized channelopathy, or be associated with any of the genetic and extracardiac causes of AF, as in our case. In absence of other underlying extra-cardiac trigger, such as hypertension, hyperthyroidism, pulmonary embolism, viral infection, sepsis or drug overdose, we considered neoplastic invasion of the LA wall as etiological mechanism of AF.

Although conversion to sinus rhythm by antiarrhythmic drugs has been observed, we experienced that synchronized electrical cardioversion was the most straightforward procedure to rapidly establish sinus rhythm. The risk of recurrence of AF is related to the underlying pathology. There was no recurrence in our case, the oncological condition being treated.

In patients without cardiac condition before onset of AF, its etiology should always be investigated. Oncological conditions are part of the differential diagnosis as we showed in this case report. Cardiac MRI can be a helpful tool. Electrical cardioversion is a rapid and very effective treatment.

P 73.

Catheter-based treatment of pulmonary embolism after pneumectomy in a child with univentricular physiology.

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University Hospital Saint-Luc

Pulmonary artery stump thrombosis is a well-known complication after pneumectomy, as well as thromboembolic events in patients with superior cavopulmonary connection (SCPC). Management protocols are lacking, especially for antithrombotic therapy in children.

A 7-year-old boy with univentricular heart in right isomerism syndrome with complete non-balanced AVSD was referred for evaluation before total cavopulmonary connection (TCPC). At the age of 15 months, he underwent surgical correction of a partial abnormal pulmonary return of both right pulmonary veins, as well as a SCPC with interruption of the native right ventricular outflow tract. Cardiac catheterization showed correct drainage of the superior caval vein to the right pulmonary artery (RPA). Left pulmonary artery (LPA) had retrograde perfusion from multiple aorto-pulmonary collaterals. Left pulmonary veins were totally occluded. In the meantime, several collaterals were embolized. After left pneumectomy, he was treated with Sildenafil and low molecular weight heparin LMWH which was switched to Aspirin 10 days later. On post-operative day 12, he developed acute respiratory distress with severe hypoxia. Angiography showed multiple thrombi in the right superior and inferior lobe and an important thrombus in the left stump of the ligatured LPA. LPA stump was totally occluded proximally by a 16mm vascular plug, to exclude the thrombus and the thrombi on the right side were aspirated. At the end of the procedure, pulmonary pressure dropped from 20 to 13mmHg. LMWH was relayed by Xarelto 2 weeks after percutaneous thrombectomy. 4 weeks later, pulmonary pressure remained low and angiography showed homogenous perfusion and drainage of the right lung, permitting TCPC.

Antithrombotic therapy after pneumonectomy in patients with CHD is not standardized. Pulmonary embolisms (PE), typically originating from thrombi in the contralateral stump, are a life-threatening complication in this population. The risk of thromboembolisms is even higher in univentricular physiology where decreased flow and absence of pulsatility characterizes the pulmonary vasculature. Percutaneous treatment of massive PE by endovascular embolectomy is feasible. Moreover, we showed that occluding the contralateral pulmonary artery stump could avoid recurrence.

Patients after SCPC and pneumectomy are at very high risk of PE. Adequate anticoagulation therapy is mandatory. We showed that endovascular embolectomy can be life-saving.

SO 10.

Therapeutic window in monosymptomatic and non-monosymptomatic enuresis

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UZ Gent, U Gent

Background
Nocturnal enuresis is caused by nocturnal urine production and functional bladder capacity mismatch. ICCS suggests patient classification into MNE and NMNE, where MNE is likely to respond to desmopressin and/or alarm. This led to the misconception that NMNE could not benefit from desmopressin. With the recent ICCS standardization, most patients are now labeled NMNE. Desmopressin's anti-diuretic effect and renal concentrating response have no direct correlation with bladder dysfunction and should be evaluated independently. Many patients have LUTS and nocturnal polyuria and could benefit from desmopressin in combination therapy.

Aim
identify patients in a tertiary center who might benefit from desmopressin (defined as urinary osmolality (Uosmol)<850 mOsm/l) and study the timing overnight.

Methods
retrospective analysis of 398 enuretic children who performed a 24h-urine concentration profile at home (4 daytime (D1-D4), 4 nighttime urine collections (N1-N4)).

Results
212 children (>50%) had Uosmol<850 mOsm/l at the 1st-night collection (N1), and would benefit from a short-term desmopressin activity; however, in a significant percentage, Uosmol is low later in the night (181 N2, 169 N3, 167 N4), needing a longer action duration. 50 patients didn't reach Uosmol>850mOsm/l over 24h, suggesting lower maximal renal concentration capacity of the normal spectrum or high 24h fluid intake.

Conclusion
Classification into MNE and NMNE is mainly bladder/LUTS driven and is widely accepted to predict the anti-enuretic effect of therapy, thus an indication for desmopressin. However, many patients have a combination of LUTS and abnormal circadian diuresis pattern. Desmopressin's anti-diuretic effect may be expected in most patients with high diuresis and low Uosmol overnight. >50% of patients have a low Uosmol early the night, hence a therapeutic window for desmopressin. In 1/3 patients, Uosmol remains longer low, needing longer-acting V2-stimulation, without risk of too prolonged action. It is evident that desmopressin's PK/PD characteristics do not fulfill these promises.

Short Oral Presentation

SO 11.

More Illness-related Parental Stress and Lower Quality of Life in Transplanted Children with Chronic Kidney Disease: A Multi-centric Study

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Objectives
Monitoring the psychological well-being of children with chronic kidney disease (CKD) is seen as standard care in pediatric nephrology, as many studies have shown that CKD has a great psychological impact. This multi-centric cross-sectional study investigated quality of life (QoL) and illness-related parental stress in this population by 1/ comparing mean levels of these two variables between several CKD categories, and 2/ exploring their correlation.

Methods
We recruited children with CKD and their parents, followed at the 6 Belgian revalidation reference centers for child nephrology. Children's QoL was assessed by the PedsQL 4.0 Generic Core Scales, parental stress was measured by the Pediatric Inventory for Parents (PIP). All patients were divided in categories based on their CKD diagnosis: 1/congenital diseases 2/ tubulopathies and metabolic diseases, 3/ nephrotic syndromes, 4/ acquired diseases with proteinuria and hypertension, and 5/ kidney transplantations.

Results
In total we included 295 children (176 boys; M age= 11.8, SD = 3.7) and 285 parents. Fifty-seven children (19%) had transplant in the past. There were no significant differences in QoL between CKD categories as reported by the children ($p>.05$). In contrast, there were significant differences between CKD categories in QoL ($F(4, 220) = 3.46$, $p<.01$)) and stress ($F(4,269) = 2.92$, $p<.05$), reported by parents, with transplant patients having lower QoL ($t(220) = - 3.31$; $p = .001$) and higher parental stress ($t(269) = 2.30$; $p = .02$). Finally, there were significant negative correlations ($p < .001$) between QoL and parental stress.

Conclusions
This multi-centric study showed lower levels of QoL and higher levels of parental stress in transplanted children, compared to children without transplant, when based on parent reports. More parental stress is associated with worse QoL in the child. These results highlight the importance of a multidisciplinary team with special attention for the parents.

P 37.

Atypical presentation of nephrotic syndrome in a young boy

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UZ Gent, AZ West

Background/aims
Nephrotic syndrome is defined by the clinical triad of heavy proteinuria, hypoalbuminemia and generalized edema. Children with atypical presenting features should be referred for specialist pediatric nephrology assessment, including renal biopsy and genetic analysis.

Methods
Case description.

Results
A Caucasian boy of 1,5 years old with a blank medical history was referred to our hospital because of nephrotic range proteinuria in combination with haematuria. There was a quite atypical presentation with only minimal periorbital edema in the morning since one month but no other symptoms. Urine sediment showed significant proteinuria (13,6 g/g creat) with mildly elevated alfa-1-microglobuline (16,16 mg/L) and significant haematuria (5261/µL). Lab results showed a normal kidney function (creat 0.35 mg/dL, urea 29 mg/dL) and low albumin (21 g/L). Clinical examination showed mild periorbital edema but no generalized edema and furthermore a stable clinical condition. He did have significant hypertension for which he was started on Amlodipine.

Investigations were performed to identify the underlying cause. Duplex ultrasound of the kidneys showed no vascular abnormalities, but bilateral enlarged kidneys with increased echogenicity and swollen aspect of the cortex. Ophthalmic screening showed no signs of uveitis or other abnormalities. Cardiac screening was negative. A kidney biopsy was performed, and while waiting for the results, he was started on corticosteroids orally (60mg/m2/d).

Kidney biopsy showed an image of mesangial hypercellularity and diffuse mesangial sclerosis with negative immunofluorescence, suggestive for an underlying hereditary cause. Genetic analysis is performed, results are following.

Conclusion
We present a case of a young boy of 1,5 years old presenting with nephrotic-range proteinuria and macroscopic haematuria. Clinically only mild periorbital edema and hypertension. Lab results showed a normal kidney function with significant proteinuria and hematuria in urine. Kidney biopsy revealed a rare image of mesangial hypercellularity and diffuse mesangial sclerosis, genetic analysis is following.

Our clinical case shows that it is essential that small children who present with atypical features not fitting the classic clinical triad of nephrotic syndrome need specific pediatric nephrological assessment.

Posters

P 38.

Case Report Juvenile Nephronophtisis

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A 15 year old girl presents at the office of her general practitioner with generalized joint pain, muscle pain and pronounced fatigue. From her medical history we note an episode at the age of 7 years old of polyuria and polydipsia. Renal ultrasound showed normal kidneys, serum creatinine was the upper limit of the normal value and a fluid deprivation test was inconclusive. A diagnosis of a habitual drinker was made.

Diagnostic investigations demonstrated an iron deficiency anemia, a severe chronic kidney disease (GFR 15 ml/min/1.73m2) and an important hypocalcemia and hyperparathyroidism. Renal ultrasound revealed hyperreflective kidneys with multiple small cysts and a decrease in volume; suggestive for kidney dysplasia.

Except of a bilateral amblyopia, ophthalmologic screening was normal. Genetical testing showed an autosomal recessive homozygous deletion at 2q13 in the NPHP1 gene by which the diagnosis of juvenile nephronophthisis was made.

The NPHP gene encodes structural components of cilia. When there is a mutation present in this gene, the body is not capable of concentrating the urine and reabsorbing natrium allowing the development of a chronical tubulointerstitial nephropathy which leads to end stage renal failure. The juvenile variant shows a typical progression of polyuria, polydipsia and secondary enuresis at the of 4 to 6 years old, leading to end stage renal disease at a median age of 13 years old. Most likely this girls' episode of polyuria and polydipsia in 2013 was already part of the disease progression. In 20 percent of the patients extrarenal manifestations are present caused by ciliary dysfunction, for example retinitis pigmentosa. In this case there was only renal pathology.

The treatment is symptomatic.

As key message of this case, we would like to point out it is important to ensure additional investigations and a proper follow-up when complaints of polydipsia and polyuria are expressed, especially in combination with a slightly raised serum creatinine.

P 39.

From a harmless IV catheter to limb necrosis: a thromboembolic complication in an infant with nephrotic syndrome.

E. Hermans, J. Vande Walle, A. Prytuła, J. Dehoorne, E. Dhont, E. Snauwaert, L. Dossche, A. Raes

UZ Gent

Background

Thromboembolism (TE) is a well-known complication in nephrotic syndrome (NS) patients. Risk factors include the disease-related hypercoagulability, pro-thrombotic state and risks related to treatment (intravenous catheters and/or diuretics). The reported incidence of TE in nephrotic children is lower than in adults, ranging from 1.8 to 4.4%. This report discusses a thromboembolic complication in an infant with congenital NS.

Methods

We present the case of a boy of two months old who had been recently diagnosed with congenital NS of the Finnish type. During the initial in-hospital management of the severe proteinuria and hypoalbuminemia (including low-molecular-weight heparin), a transient episode of sudden paleness and coldness of the left forearm occurred. Two days before a peripheral IV catheter had been placed in the left forearm. The day after this episode, the same signs reappeared with additional marbling of the skin. During the removal of the IV catheter, a pulsatile flow was noted, confirming that the catheter had accidentally been placed in the brachial artery. A Doppler ultrasound showed a thickened arterial wall at the site of the IV catheter. Despite administration of a continuous heparin infusion, he developed compartment syndrome of the forearm for which multiple fasciotomies were performed. Necrosis of the skin of the forearm and the tip of the index finger occurred. Extensive wound care, silicone and pressure garment therapy were continued until 1 ½ year after the incident. The boy was treated with acetylsalicylic acid until the age of 22 months.

Results

This case describes an arterial thromboembolic complication in the limb of an infant with nephrotic syndrome. Thromboembolic complications are generally venous, whereas the occurrence of (peripheral) arterial thromboembolism in NS is rare. However, multiple cases have been described in the literature, each associated with significant morbidity and/or mortality. The precedent of an arterial puncture appears to be an important risk factor.

Conclusion

Thromboembolisms are a serious complication in children with nephrotic syndrome and can cause significant morbidity and mortality in all age categories. A high index of suspicion is required as the clinical features may be subtle. In general, physicians should recognize the risk of (accidental) arterial punctures, especially in patients with additional risk factors.

Posters

P 40.

A complication of nephrotic syndrome that should not be missed.

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U Gent, UZ Gent

Background/aims
Nephrotic syndrome is characterised by the clinical triad of proteinuria, hypoalbuminaemia and oedema. Children suffering from nephrotic syndrome are at risk of venous thromboembolisms, including pulmonary embolism. Risk factors for venous thromboembolism are a hypovolaemic state and a hypercoagulable state. This last can be explained by urinary loss of anticoagulant proteins such as antithrombin III, protein C and S, and increased synthesis of prothrombotic factors such as factors V and VIII.

Methods
A case report.

Results
A 10-year-old boy with a medical history of steroid dependent nephrotic syndrome since 2013 and minimal change disease on kidney biopsy, received a first dose of rituximab due to insufficient control of nephrotic syndrome under tacrolimus and steroid therapy. Shortly after rituximab infusion, he developed a relapse with hypoalbuminaemia and proteinuria. In the following days he developed progressive tachypnea and decreased tolerance to physical activity. At presentation in the clinic, an ill-appearing boy was seen, with severe tachypnea (35/min), tachycardia (135/min), oxygen saturation of 93%, severe peripheral oedema and poor peripheral circulation. RX thorax showed pleural fluid on both sides. Venous blood gas pointed to respiratory alkalosis and D-Dimers were elevated to 2470 ng/ml. Consequently CT imaging was performed, confirming severe bilateral pulmonary embolism and pleural effusion. He was urgently transferred to the paediatric intensive care unit for acute thrombolysis and anticoagulation therapy. Post hoc, we identified a raise in haematocrit with a raise in haemoglobin from 13,3 to 17 g/dl, together with a mineralocorticoid excess UK/(UNA+K)-ratio >90%, documenting hypovolaemia.

Conclusion
This case report describes the case of a 10-year-old boy who presents himself with tachypnea, dyspnea and tachycardia, due to pulmonary embolism, a complication related to hypercoagulability of nephrotic syndrome. The observed hypovolaemia was certainly a co-factor. This case underlines that there should be awareness for possible life-threatening complications of nephrotic syndrome, such as pulmonary embolism.

P 41.

Luminous Fecalomas in an Adolescent Girl with Peritoneal Dialysis.

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An 18-year-old severe retardated girl was admitted tot he transition clinic of nephrology with severe abdominal pain in the right fossa iliaca. She was born after an uncomplicated pregnancy to consanguinous Moroccan parents. Mental developmental index <50 on the Bayley scales of infant and toddler development II (BSID-II), corresponding to a development of three months at a calendar age of eight months. Brain NMR confirmed the diagnosis of Joubert syndrome with in the transverse images of the midbrain the characteristic view of the “molar tooth sign”.At the age of 7 years she was hospitalized for a cholecystitis. Genetic testing demonstrated a missense mutation c.1888T>C in the gene TMEM 67 on chromosome 8q22.1. The patient developed a chronic thrombocytopenia and leukopenia at the age of eleven-years old. At the age of 14 years old due to end stage renal failure nocturnal continuous peritoneal dialysis was started. Patient had terminal renal failure(eGFR(CKD-EPI):6.1 ml/min/1.73 m²) due to nephronophtisis. She developed a severe hyperphosphataemia and a very high level of PTH. The severe hyperphosphatemia does not respond tot he tradional phosphate binders like calcium carbonate. Since 2015 at the age of 15 years-old lanthanum carbonate. Suddenly at the age of 18 years old she was admitted to the emergency department with biliary vomiting, and severe colicky abdominal pain and subfebrilitas. A diagnosis of a subobstruction was made. X-ray of abdomen showed multiple radio-opaque concretions throughout the stomach, the right hemiabdomen and rectal sigmoid .X-ray of the pelvis showed a huge radio-opaque fecaloma (height 11 cm with 9 cm). Due to the patient’s mental status, the size of the fecaloma and the possibility of external compression of the outflow of the Tenkhoff catheter, a coloscopy under general anesthesia was performed with manual removal of the fecalomas.A few weeks later she developed a peritonitis. She was treated with broad-spectrum antibiotics. A month later she died from hepatic coma .

Lanthanium is a rare earth metal. In Mendeliev's table, lanthanium has an atomic number 57 and atomic mass 139, an atomic number higher than barium. (56) Because lanthanum has an atomic weight almost identical to that of barium, it absorbs X-rays and also has a density four times greater than that of calcium.This radiopacificity of lanthanum carbonate may be misinterpreted , especially in children and young people with mental disabilities.

Posters

P 42.

Harmful or not? Proteinuria in a child with Familial Mediterranean Fever

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KU Leuven

A child who presented with possible Familial Mediterranean Fever and proteinuria at an early age is rare. Here, we present a case report of a three year old boy who presented with mild proteinuria, suspected FMF and a normal kidney function. Secundary amyloidosis was suspected but not confirmed in a subsequent kidney biopsy. Subsequently his sister was also screened for FMF which was negative, but she presented with proteinuria as well. Familial genetic evaluation for proteinuria revealed a rare terminal mutation in the CUBN gene, resulting in a reduced function of the cubilin protein with a phenotype of isolated chronic proteinuria. With this case report, we want to stress the importance of fully understanding an odd presentation and why further investigation such as genetic evaluation is necessary to comprehend an unexplained feature.

P 43.

Nocturia: the link with enuresis during childhood and beyond

B. Borgonjon, J. Vande Walle, L. Dossche

UZ Gent, U Gent

Introduction

Both enuresis in childhood and nocturia in adults are well studied, and the impact on quality of sleep and life makes treatment indispensable. Despite the clear overlap between pathophysiological mechanisms, the link between both conditions remains unclear. Therefore, it urges the question if nocturia is a relevant health problem in children, and if it is linked with enuresis.

Materials and Methods

A literature search was conducted using electronic databases Medline, Embase and Google Scholar, using search terms; nocturia, enuresis, children and sleep disorders. Additional articles were also obtained using citation-based search and the 'snowball' method. A total of 38 different articles were selected.

Results

Only 2 articles reported on the prevalence of nocturia at least once a week in childhood, which was around 5-8% at school age and therefore comparable with enuresis. The majority of studies identify that a history of enuresis during childhood is linked with nocturia in adulthood. Additionally, a history of enuresis is correlated with a more severe course of nocturia in adulthood. Moreover, alarm treatment for enuresis in children may lead to the development of nocturia. The main difference between both conditions is sleep and arousal mechanisms.

Conclusion

Nocturia in children has been studied only to a limited extent in comparison with enuresis, while the restricted evidence indicates that it is at least as prevalent as enuresis. The link between both conditions advocates for an optimized transition from pediatric to adult care.

Posters

P 44.

Congenital nephrotic syndrome: a case of an unusual favorable disease course.

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UZ Gent

Background

Congenital nephrotic syndrome (CNS) is a rare kidney disease characterized by the typical triad of edema, proteinuria and hypoalbuminemia within the first 3 months of life. Most cases are due to monogenic defects of the structural proteins that form the kidney filtration barrier. Defects in the NPHS1 gene are the most prevalent, resulting in CNS of the Finnish type. This report discusses the unusual disease course of a boy with CNS of the Finnish type.

Methods

We present the case of a boy of 2 ½ years old with CNS of the Finnish type. He was diagnosed at the age of 2 months after being admitted to the hospital with general deterioration, vomiting and ascites. Genetic analysis of the NPHS1 gene revealed compound heterozygosity for the mutation C.1868G>T p.(Cys623Phe) and c.1913A>G p.(Tyr638Cys). The boy was treated with intermittent regular albumin infusions and diuretics to control edema and angiotensin-converting-enzym inhibition (ACE-I) for the hypertension. The course of this CNS was favorable with tapering of the albumin infusions and complete withdrawal at the age of 11 months. Diuretics were discontinued 7 months later. To this date, his renal function remains normal, with stable mild proteinuria, stable low-normal serum albumin and normal blood pressure under ACE-I inhibition.

Results

This case of a boy with CNS of the Finnish type describes an unusual course of this disease. The typical CNS of the Finnish type, caused by Finmajor (p.Leu41fs*91) and Finminor (p.Arg1109*) mutations, demonstrates relatively little phenotypic variation with massive proteinuria at birth and rapid progression to end-stage renal disease. This boy showed partial remission of CNS, probably linked to the different NPHS1 mutations. To the best of our knowledge, only eight other isolated cases of CNS of the Finnish type with a favorable outcome are reported in the literature. Additionally, one specific family with multiple cases of CNS and a specific ethnic group have been described to have a milder phenotype.

Conclusion

CNS of the Finnish type can rarely present with a mild disease course with prolonged preservation of renal function.

P 45.

Case presentation of an anti-factor H antibodies induced thrombotic microangiopathy

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Case Presentation

A 7-year-old boy without a relevant medical history presented himself at the general pediatrician with fever, pruritus and petechiae. Blood tests showed hemolytic anemia with presence of schistocytes, thrombocytopenia and acute kidney injury, diagnostic for a thrombotic microangiopathy (TMA). Further examinations showed normal ADAMTS13 and negative screening for verotoxin induced TMA (faeces and serology). During hospital admission a progressive deterioration of his kidney function was seen, for which peritoneal dialysis was started. Given the serious clinical presentation, the absence of arguments suggesting a verotoxin positive TMA and a normal ADAMTS13, further etiologic elaboration was started. One dose of Eculizumab (complement C5 inhibitor) was given after administration of meningococcal vaccination, while waiting for further results. After receiving Eculizumab a complete resolution of the TMA course was seen with recuperation of the kidney injury and possibility to stop peritoneal dialysis. Elaboration showed high titre levels of anti-factor H antibodies (titre: 19 584 u Arb, normal < 150 u Arb), diagnostic for an atypical hemolytic uremic syndrome (HUS). Genetic testing showed a homozygotic deletion of CFHR1 and CFHR3, which is a known predisposing factor for anti-factor H antibodies induced TMA. Having regard to the autoimmune basis of the condition prednisolone and mycophenolate mofetil were associated to the treatment and Eculizumab was continued. This resulted in a favourable decline of anti-factor H antibodies to < 200 u Arb so Eculizumab could be discontinued after three months of treatment.

Conclusion

Early recognition and treatment of thrombotic microangiopathy is essential to optimise the outcome of these patients. Each patient with TMA should get an urgent and extensive elaboration with verotoxin screening and ADAMTS13 diagnostics. In this manner, patients in whom verotoxin positive TMA is excluded can qualify for treatment with Eculizumab, a treatment that drastically improved the prognosis of these children.

Posters

P 46.

Extensive review of pharmacokinetic and pharmacodynamic properties of ACE-inhibitors and angiotensin receptor blockers in hypertensive paediatric patients

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Background/Aims
Globally, hypertension is among the key preventable causes of premature death. Currently, childhood hypertension is estimated around 2 to 4% of the paediatric population and expected to further increase due to the obesity epidemic. The leading cause for childhood hypertension is renal impairment, described in over 90% of the Western cases. ACE-inhibitors (ACE-I) and Angiotensin Receptor Blockers (ARBs) are most frequently prescribed for blood pressure reduction. Both therapeutics are currently being used off-label, despite being on the market for the past three decades. Multiple incentives have been put forward by the European Medicines Agency to stimulate receiving appropriate labelling. However, none were approved for a hypertensive renally impaired population due to a lack of appropriate study designs and results. The aim of this study is to retrospectively summarize and compare findings of pharmacokinetic (PK) and pharmacodynamic (PD) studies investigating all drugs of the ACE-I and ARB classes, including a potential recommendation for improved study design.

Method
This review focused on the clinical trials investigating pharmacokinetic and pharmacodynamic properties of ACE-I and ARBs over the past 30 years in response to regulatory initiatives. A total of 60 studies were selected, including 19 randomized controlled trials. The study population included a total of 3660 hypertensive children. Analysis was conducted with a focus on trial design and endpoints, drug dosing, safety, efficacy and drug indication.

Results
Between ACE-I and ARBs, geographical location, drug intake and formulations were comparable. Study population differed, where studies on ARBs focused on both primary and secondary hypertension, whereas studies on ACE-I focused on secondary hypertension. Sampling regimens differed, where studies investigating the PK of ARBs were more frequently based on single dosing at non-steady state. For both classes, low reporting of estimated glomerular filtration rate (eGFR) (23.3%) and the exclusion of participants with an eGFR under 30 was apparent. Individual antihypertensive effects of ACE-I could be verified in 77 children, where around 90% achieved a blood pressure decrease of ≥ 6 mmHg. ACE-I were generally well tolerated when considering safety parameters and serious adverse events. No studies investigated the long-term effects of ACE-I and ARBs on cardiovascular morbidity and mortality.

Conclusion
Standardization of methodology and reporting of results is imperative for both PK and PD studies, to allow a better comparison of results and to aim towards appropriate labelling. Stratification for and inclusion of different age categories and eGFR ranges is recommended.

LO 5.

INSENODIAB Study: Determinants and characteristics of insulin dose requirements in children and adolescent with new-onset type 1 diabetes

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Cliniques universitaires Saint-Luc

Aims

In children with newly diagnosed type 1 diabetes (T1D), insulin dose regimens vary substantially. According to current ISPAD recommendations, the initial total daily dose should range from 0.7 to 1 UI/kg body weight/day. Adjusting and stabilizing this dose to achieve normal blood glucose concentration can take several days. The goals of this study were (1) to assess how patient characteristics influence insulin dose requirements and (2) to establish predictive models of those insulin requirements in newly diagnosed children with T1D.

Methods

INSENODIAB is a monocentric, retrospective observational study over a 7-year period from January 2013 to February 2020. Chart review was conducted for children (6 months-18 years) admitted in Cliniques universitaires Saint-Luc for a new diagnosis of T1D during the observational period. Demographics, clinical and laboratory data, including insulin dosage were collected for all patients. Univariate and multivariable linear regression models were used to examine the impact of patient variables on insulin total daily dose, using a nominal Type I error of 5% as threshold.

Results

Complete clinical records were available for 103 patients with median body weight of 27 kg (Q1-Q3: 18.2-39.1, Mean: 30.0 kg). Median Insulin total daily dose was 26.8 units (Q1-Q3: 18.8-48.3 units, n=103) during hospitalization and 24.5 units (Q1-Q3: 17.0-45.3 units, n=94) on the day of discharge. Median duration of hospitalization was five days (range 1-10 days). In multivariable analysis, the main variables found to impact optimal insulin total daily dose were age, veinous bicarbonates levels at admission, body mass index and percentage of weight loss at diagnosis. The same factors remained after adjusting the model to insulin dose per day per kilogram body weight.

Conclusions

In newly diagnosed children with T1D, percentage of weight loss and veinous bicarbonates levels at admission, in addition to age and body mass index, influence the Insulin total daily dose necessary to reach glycemic control. Those results helped us developing a dosing algorithm which could potentially reduce the number of days currently needed to stabilize glycemic control in children and adolescents with new-onset T1D.

Long Oral Presentation

LO 6.

Placental SRC-2 methylation and expression profile is affected by prenatal exposure to BPA and BPS in female rats

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U Liège

Epigenetic mechanisms are influenced by the early life environment and play an important role in the fetal set up of homeostatic processes. Changes in placental physiology can trigger an adaptive response, supporting the involvement of placenta in programming. We hypothesized that placental epigenome could provide early markers of exposure to endocrine disrupting chemicals.

We have shown that prenatal exposure to a high dose of Bisphenol A (BPA) (10mg/kg/day) is associated with DNA methylation changes in CpG islands at different loci in a sexually dimorphic manner. By combining placental RNAseq and DNA methylation arrays, we identified the nuclear receptor coactivator 2 (Src2) as a potential biomarker of exposure to BPA.

We aimed at studying DNA methylation and mRNA expression of Src2 in the female placenta after exposure to two low environmentally relevant doses of BPA. Because of the increasing presence of Bisphenol S (BPS) as a BPA substitute, we further studied the epigenetic and transcriptional Src2 placental changes after exposure to the same doses of BPS.

Pregnant rats were orally exposed to two doses of BPA and BPS (25 ng/kg/day, a very low dose that affect puberty timing and 4 µg/kg/day, the defined tolerable daily intake dose) one week before mating and during gestation. Placenta were harvested at GD 19. Male and female placentas were identified using classical PCR for SRY expression. Following DNA bisulfite treatment, we did a targeted sequencing of the Src2 gene promoter regulatory region in female placenta to study DNA methylation. Moreover, to further study placental transcriptional changes, we studied the expression of Src2 and DNA methylation enzymes (Dnmt1, Dntm3a, Dntm3b, Tet1 and Tet2) by qPCR.

Our results show that Src2 was significantly downregulated by both doses of BPA and BPS. Furthermore, the lower dose of BPS was found to significantly alter 6 out of 23 CpG sites of the Src2 promoter as compared to controls. Finally, exposure to both doses of BPS and the highest dose of BPA significantly downregulated Tet2 mRNA expression in the placenta. Neither BPA nor BPS affected Dnmt1, Dnmt3a, Dnmt3b or Tet1 expression.

In conclusion, our transcriptional and epigenetic analysis allowed the identification of the Src2 gene as a potential placental biomarker of BPA and BPS exposure. Src2 is a key mediator of placental response to estrogens. Overall,our data shows that placental physiology is vulnerable to environmental exposure to BPA and BPS.

SO 13.

New-onset type 1 diabetes in children and adolescents before and during Covid-19 pandemic in Belgium

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HUDERF, ULB

Background
Viral infections have been implicated in the development of type 1 diabetes. There have been reports of Covid-19 induced new-onset diabetes. In this context, this study aimed to describe and compare the characteristics at diagnosis of children who develop type 1 diabetes before and during the Covid-19 pandemic in Belgium.

Methods
This observational study compares two groups of children and adolescents who develop type 1 diabetes: the first includes new-onset diabetes before the pandemic (1 March 2018 and 31 December 2019) and the second during the pandemic (1 March 2020 and 31 December 2021) in HUDERF, Brussels, Belgium.

Results
In our center, the number of new-onset type 1 diabetes in the pre-pandemic period was 87 and in the pandemic period was 147 (p=0.010). During the Covid-19 pandemic, patients with new-onset diabetes were more often male (62 % versus 44 %; p=0.010) and had a slightly higher pH at admission [7.35 (7.22 – 7.41) versus 7.34 (7.18 – 7.38); p=0.043] than before the pandemic. There was no difference in age, BMI SDS, HbA1c or severity of ketoacidosis at type 1 diabetes diagnosis (Table 1). During the pandemic, 2 children (2 %) had a positive SARS-CoV2 PCR test on admission but were asymptomatic.

Conclusions
The number of new cases of type 1 diabetes in our center during the Covid-19 pandemic was significantly higher than before the pandemic. These new patients were mainly male. Longer-term and national follow-up is needed to assess the role of Covid-19 in the development of type 1 diabetes.

Short Oral Presentation

SO 14.

Absence of association between hyperuricemia and aberrant metabolic parameters in Belgian children and adolescents with overweight or obesity

E. Nauwynck, J. Vanbesien, W. Staels, S. Ryckx, S. Verheyden, B. Rutteman, J. De Schepper, I. Gies

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Background/ Aims

The level of serum uric acid (SUA) is directly related to the risk of type 2 diabetes and cardiovascular disease in adults. Hyperuricemia is present in 12.4-40% of obese/overweight children and adolescents. SUA is associated to different components of the metabolic syndrome (MS) in pediatric and adult obesity, especially in females. The aims of this retrospective study were 1) to compare the prevalence of hyperuricemia in obese versus overweight children and adolescents, and 2) to evaluate correlations between SUA and HOMA-IR, a measure for insulin resistance, and the different components of pediatric MS.

Methods

Fasting SUA, plasma glucose and insulin, total cholesterol, HDL cholesterol, and triglycerides were measured in 389 overweight (BMI 1.3 – 2 SDS) and obese (BMI SDS > 2) children and adolescents. Median age of the 283 obese children was 10.7 years, while median age of the 106 overweight children was 10.2 years (range 6-16). 62/106 (58.5%) in the overweight group and 158/283 (55.8%) in the obese group were females. Routine anthropometry and blood pressure measurements were performed. SUA was measured by enzymatic colorimetry. Hyperuricemia was defined by age and gender reference ranges. Metabolic syndrome was defined by the IDF criteria.

Results

Median (range) SUA was 4.9mg/dL (2.5-8.6) in the overweight group vs 4.7 mg/dL(0.5-9.5)(P 0.324) in the obese group . SUA values were comparable between females and males. MS was present in 8.5% of the overweight patients and in 12% of the obese patients (P 0.351). Hyperuricemia was as prevalent in the obese (19.4%) as in the overweight group (22.6%, P 0.5). Gender distribution, median age, BMI SDS, waist SDS and birth weight SDS were not significantly different between patients with or without hyperuricemia. Median blood pressure SDS, HDL cholesterol, triglycerides, glucose, and HOMA IR did not differ significantly between subjects with normal or elevated SUA concentrations.

Conclusion

Twenty percent of overweight and obese children and adolescents have hyperuricemia, irrespective of gender and age. No significant correlation between serum uric acid and the different components of MS was found. We hypothesize that beside ethnic origin, lifestyle (sedentarism) and dietary factors (fructose/ carbohydrate intake) as well as obesity-related conditions, such as chronic inflammation or duration of overweight, may favor the association between hyperuricemia and MS found in other studies.

SO 15.

Transition of adolescents with type 1 diabetes from pediatric to adult health care systems in Belgium

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UZ Gent

Background and Objective

Young adults with type 1 diabetes (T1D) who transition from pediatric to adult care face a higher risk of adverse outcomes. The main objective of this study is to inform on current transition practices in Belgium and describe the experiences and perceived barriers to transitional care reported by pediatric and adult endocrinologists.

Research design and methods

We conducted an electronic survey to pediatric and adult endocrinologists identified through the Belgian Society for Pediatric Endocrinology and Diabetology (BESPEED) and the Belgian Endocrine Society (BES).

Results

Sixty-four physicians responded (20%), 21 of whom were pediatric endocrinologists (63%) and 43 adult endocrinologists (15%). The majority of the respondents were female (60.9%), > 45 years old (59.5%) and worked in a non-academic hospital (62.5%). According to most respondents, the ideal age for transition is between 16 and 18 years (48.2%). In 72 % of the cases, the current age of transfer is significantly higher than the considered ideal age (P=.003). A written, structured transition program is used by only 11.1% of the respondents, and 17.2% of the respondents have a transition coordinator. Main perceived barriers to successful transition were lack of time and resources for organizing transitional care. Patients' knowledge and self-management skills, and complete medical information transfer were the most important factors for successful transition. The majority of adult endocrinologist stated that they almost always receive a written medical summary upon transition (60%). Still, a psychosocial summary is rarely or never received, although 81.4 % of the adult respondents want one. Respondents who received training were more likely to use a structured transition program (P=.014), and were more likely to have a transition coordinator in their team (P=.025).

Conclusions

Our study reveals a wide variation and gaps in transitional care for adolescents and young adults with T1D and a need for a more structured approach to transition care in Belgium. Main barriers are lack of time and resources and lack of transition protocols. The development of a diabetes transition care program tailored to the Belgian situation can help to overcome the perceived barriers. Education on transition has a positive impact on the implementation of a structured transition program and should be promoted.

Short Oral Presentation

SO 16.

Did Sars-CoV-2 pandemic change incidence or patient and admission characteristics of children with de novo type 1 diabetes? Preliminary results

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Background and Aims

In 2020 Sars-CoV-2 caused a worldwide pandemic. Some centers reported changes in incidence and severity of illness at presentation of de novo type 1 diabetes (T1D) in children, whilst others did not. The aim of this study is to provide data on incidence and severity of illness at presentation of de novo T1D in children in one large center.

Methods

We conducted an observational, single center study including children of 16 years or younger with de novo T1D, diagnosed between March 2020 and February 2021 (n= 51) and controls admitted between January 2015 and February 2020 (n= 203). Patient characteristics and biochemical values were evaluated and compared data using non-parametric tests.

Results

No change in incidence of de novo T1D was observed in the first year of the Sars-CoV-2 pandemic (n= 51) compared to 5-years before (n=33 to 52). We observed no significant difference in age, glycaemia, HbA1c, C-peptide, ketone bodies, bicarbonate and base-excess at diagnosis during the first year of the pandemic compared to five years before. Duration of signs and symptoms before presentation was, however, longer (29 days vs 20 days, p=0.011) and pH at diagnosis was lower (7.25 vs 7.30, p=0.016) during pandemic compared to five years before, but without increased need for intensive care admission (11.3% vs 19.6%, p=0.123). Finally, the total daily insulin dose at discharge did not differ compared to five years prior to the pandemic.

Conclusions

We conclude that Sars-CoV-2 pandemic did not change the incidence nor age distribution of de novo T1D in children in our center. Signs and symptoms of T1D were present longer and pH was lower during the pandemic than before, possibly reflecting delay in presentation, but not resulting in increased need for intensive care. Efforts should be made to obtain nationwide data.

SO 17.

Self-reported tolerance and adherence to statin treatment in adolescents with familial hypercholesterolemia (STAFF study).

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Background and aims

Current Belgian guidelines recommend statin treatment in adolescents with familial hypercholesterolemia (FH) from the age of 10 years. In adults statin nonadherence is considered a major challenge to the prevention of cardiovascular disease. Data on real-world adherence and tolerance of statins in adolescents are scarce. The aim of this study was to measure real-world statin tolerance and adherence in adolescents with FH in two Belgian university hospitals.

Patients and methods

38 adolescents with FH had regular follow-up in the UZ Gent or UZ Brussel. 18 (12 males) /38 subjects, who had received treatment with statins for at least 12 months, completed questionnaires on demographic characteristics, adverse events (AE) at initiation, and adherence. Drug tolerance was assessed based on the occurrence of targeted AEs, such as muscle pain, headache, or gastrointestinal symptoms, in the first months of treatment. Adherence was measured with the Medication Adherence Report Scale (MARS-5), with higher scores indicating higher reported adherence.

Results

The mean (range) age of the respondents was 14.6 (10.2-17.7) years. Median (range) duration of statin treatment was 3 (1-7) years. In 4 /18 subjects, statins were started under 10 years of age. 15/18 adolescents received rosuvastatin (7 patients at an initial dose of 10 mg, 8 patients at 20 mg) and 3/18 received atorvastatin (1 patient at 10 mg, 2 patients at 20 mg). 7/18 patients reported AE in the first months of treatment. Transient muscle pain was the most common AE reported by 2/15 patients on atorvastatin and 2/3 on rosuvastatin. No patient discontinued treatment. Three patients used smartphone reminders and 1 a pillbox for improved adherence. The median adherence score by MARS-5 (range = 5-25) was very high at 24. The lowest reported MARS-5 score was 18, observed in only one patient. MARS-5 scores did not correlate with age, gender, duration of treatment, baseline or 1-year LDL cholesterol levels.

Conclusion

Adolescents with FH report good tolerance and very high adherence to statins in the first years of treatment and the within-group variance is low.

Posters

P 63.

Syndromic tall stature associated with a novel heterozygous variant in TGFβ3

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Background

Tall stature including disproportion and dysmorphism might be a difficult diagnosis. Monoallelic mutations in the TGFβ3 gene (transforming growth factor beta-3) are associated with a rare heritable disorder of connective tissue mainly affecting the skeletal system and cardiovascular system. The phenotype is variable, including aortic abnormalities (aneurysm, dissection), a bifid uvula or cleft palate and skeletal abnormalities (pectus deformity, scoliosis, cyphosis, arachnodactyly, tall stature).

Case report

A 14,5-year-old girl born to Romanian, non-consanguineous parents (father: 180 cm; mother: 171.9 cm; target height: 169.2 cm,+ 0.9 SD), with normal measurements at birth (weight: 3kg150; length: 51 cm) was referred to our department for tall stature.

She had an attention deficit disorder, learning difficulty and a special needs education.

Height was 181.5 cm (+ 3,2 SD), weight was 79,8 kg, BMI: 24.2 kg/m2 (+1.4 SD), head circumference: 56,7 cm (+1.3 SD). She had a marfanoid habitus including long legs (upper/lower segment: 0.84), long and flat feet and long hands with long digits.

She had crowded teeth, a high-arched palate but no bifid uvula.

Physical examination also revealed moderate joint laxity, spaced toes, easy striae, thorax deformity without scoliosis. Breast was at tanner staging 4 since more than 2 yrs, but the patient had primary amenorrhea.

Hormonal evaluation was normal including normal IGF-1 at 517 µg/L.

Bone age was of 15 years.

Cardiovascular ultrasound showed aortic valve dysplasia.

Results

The exome sequencing identified a novel heterozygous c.926G>A; p.(Arg309His) variant in the TGFβ3 gene, inherited from her mother.

It also showed a heterozygous c.5531T>A; p.(Leu1844His) variant in the TNXB gene.

Conclusion

Our patient shares some clinical features with the reported patients with TGFβ3 mutations: tall stature, aortic valve dysplasia, moderate hyperlaxity of joints, long feet and long hands, pectus deformity. The mother has tall stature and a bifid uvula but no marfanoid habitus, her morphotype might illustrate incomplete penetrance or variable clinical expressivity.

Not only biallelic but also monoallelic TNXB pathogenic variants could lead - according to some reports - to joint hypermobility, vascular fragility and abnormal skin texture. The monoallelic TNXB variant, not inherited from her mother, found in our patient could contribute to hypermobility.

P 64.

Diagnosis and long-term outcome of GH therapy in a Belgian child with a GHGHR gene defect

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Background

Growth hormone releasing hormone receptor (GHRHR) gene mutations are emerging as a common cause of familial isolated growth hormone deficiency (GHD). Most GHRHR deficient patients are of consanguineous parents or from certain ethnic backgrounds. We describe the skeletal and body composition status after a long-term growth hormone (GH) therapy, which was started at a very young age in a Belgian girl with a pathogenic GHRHR mutation.

Methods

Persistence of GHD was evaluated in a 14 year old girl. GH therapy had been started at the age of 16 months for an isolated GH deficiency (documented persistent growth failure since the age of 3 months, a retarded bone maturation, very low basal serum IGF1 and GH levels and a hypoplastic adenohypophysis at brain imaging). However, there was no history of neonatal hypoglycemia and no confirmation by a GH stimulation test. A dramatic response to GH initiation (17cm height gain in the first year of treatment and rapid loss of subcutaneous fat) was observed. No other pituitary hormone deficiencies developed during follow up. Parental history showed a normal height in the father (186cm) and mother (162cm), who were both from West Flanders origin and not known to be related.

Results

Standing height was 155.8cm. DXA of showed an aerial BMD z scores 0.7 at the lumbar spine and hip and a whole body a fat percentage of 32%. Serum IGF1 was 46µg/L (z score -6.07). PRL, FT4, cortisol, LH and FSH were within normal limits, while TSH was slightly increased (7.5mIU/L). Basal GH was 0.05µg/L and did not increase significantly after insulin induced hypoglycemia (peak value 0.18µg/L). The extremely low IGF1 and post stimulation GH levels and the slightly elevated TSH in combination with a hypoplastic anterior pituitary at MRI lead to the suspicion of a GHRHR gene defect. A homozygous previously reported pathogenic frame-shift mutation (c.674_677delinsGCTGTTGGCAGAAG p.(Val225Gly*fs165) in exon 7 of the GHRHR gene was found by exome sequencing.

Conclusion

In conclusion, GH therapy normalized adult height, body proportions , bone mineral content and body fat content in a Belgian girl with a documented GHRHR gene defect. A GHRHR gene mutation analysis should be performed in children with isolated GHD due to a hypoplastic adenohypophysis without neonatal hypoglycemia, frontal bossing or midfacial hypoplasia, when showing very low basal and stimulated GH levels at retesting in children.

P 65.

Recurrent convulsions due to insulinoma-related hypoglycemia

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UZ Brussel

Background
Severe hypoglycemia is a known cause of seizures, especially in diabetics on insulin therapy. However, hyperinsulinemic hypoglycemia can also be caused by insulinomas. These rare pancreatic endocrine tumors can present isolated or in the context of multiple endocrine neoplasia type 1 (MEN1) syndrome. We present a boy with recurrent seizures who was initially diagnosed with epilepsy but found to have an insulinoma as part of MEN1 syndrome that caused his neurological symptoms.

Case
An 11-year-old boy with recently diagnosed epilepsy presented to our outpatient clinic for second opinion. His neurological complaints had started eight months ago and recurred despite antiepileptic drugs. The episodes included focal and tonic-clonic seizures, unconsciousness, dilated pupils, sweating, regular headaches, and behavioral problems. Aside from this he had considerable weight gain. Previous investigations showed normal serum tests (normoglycemia, electrolytes, low inflammatory markers, negative hemocultures and serology tests), negative metabolic serum and urine tests. CSF analysis showed no pleiocytosis, but glucose levels were low (35.7 mg/dL). Liquor cultures were negative. CT-scan and MRI were normal, while EEG showed signs of mild encephalopathy.

In our waiting room, the boy suddenly had generalized seizures and rapid screening exams revealed low blood glucose levels at 38 mg/dl.

Results
Laboratory tests at the time of hypoglycemia showed high insulin and C-peptide levels with normal growth hormone, ACTH and cortisol. No ketonuria and toxicology screening was negative. Previous genetic tests including mendeliome sequencing in the hospital where he was initially followed, had identified a heterozygous de novo variant in MEN1 which had not been linked to his neurological complaints. Indeed, by abdominal MRI we found a solitary pancreatic nodule that was enucleated laparoscopically and confirmed to be an insulinoma. Serum glucose levels stabilized, his behavior and weight normalized, and his neurological complaints resolved.

Conclusion
Acute hypoglycemia can cause convulsions. Variable timing of hypoglycemia and absence of ketonuria is suspicious for hyperinsulinism. Insulinoma can be the first presentation of MEN1 syndrome in children and mendeliome sequencing can be a useful diagnostic tool to approach difficult clinical presentations.

P 66.

Hungry Bone Syndrome occurring after vitamin D and calcium supplementation in children with seizures on hypocalcemia with hypovitaminosis D

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Background

Initiation of vitamin D and calcium (Ca) therapy can lead to Hungry Bone Syndrome (HBS), defined as hypocalcemia, as a result of Ca flow from bloodstream to bones, lasting more than 4 days despite Ca supplementation. HBS is mainly described in post-operative complications of parathyroidectomy for hyperparathyroidism.

With these cases, we demonstrate the pitfalls of managing hypocalcemia in order to avoid a HBS.

Case 1

A 6 month old boy presented with short seizures.

Biology showed hypocalcemia (1,4mmol/L), elevated parathyroid hormone (PTH) and alkaline phosphatase (PAL), vitamin D deficiency. He had rickets and cardiomegaly with delated left ventricle.

He was treated with intravenous (IV) calcium gluconate and oral colecalciferol.

During the first few days, the hypocalcemia worsened, and the child presented with new seizures. Amount of IV Ca and vitamin D administered were increased and the child required an IV bolus of calcium chloride to improve his blood Ca and reach normal blood Ca levels on day 5.

Case 2

A 19-day-old boy presented with short seizures. Biology showed hypocalcemia (1.58 mmol/L), hypovitaminosis D, low PTH (39ng/L) and high PAL. The child received a bolus of calcium gluconate but hypocalcaemia worsened. Intraveinous Ca was increased and the child received alfacalcidiol and colecalciferol. PTH was increased (48,5ng/L). The child presented with new seizure and a cardiogenic shock requiring calcium chloride boluses, respiratory and inotropic support. At day 6, normocalcemia was achieved.

Discussion

HBS is an uncommon cause of hypocalcemia in children. In these cases, the persistence of hypocalcemia during treatment with Ca and vitamin D suggested a possible HBS. The hypothesis is that IV Ca initiates a flow of Ca from blood to bones, reversing the process of bone resorption to bone reabsorption. The first patient had several risk factors described in adults studies : elevated ALP and PTH, bone damage due to rickets.

In practice, HBS should be considered when initiating treatment for symptomatic hypocalcaemia. The patient's risk factors of HBS should be assessed, to establish the patient's treatment and monitoring strategy. Treatment consists in Ca and vitamin D administration. Ca should be started as a bolus and continuous IV solution. Serum Ca levels should be monitored several times a day to adjust the treatment dose. If blood Ca levels drop after Ca administration, it suggests a HBS and treatment must be intensifed.

P 67.

Everything in excess is opposed to nature, even vitamin D: a case report

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Background/aims
Vitamin D intoxication in children is rare but its incidence is increasing as vitamin D is supplemented more often and in higher doses. Children with cystic fibrosis (CF) are at risk for vitamin D intoxication due to incorrect compounded preparations of liposoluble vitamins. It should be suspected in every child receiving vitamin D supplements when complaining of vague abdominal symptoms, fatigue, and polydipsia - the classical presenting symptoms of hypercalcemia.

Results
A 4-year-old girl with CF and exocrine pancreatic insufficiency, presented with suprapubic abdominal pain, decreased appetite, constipation, fatigue, weight loss, polyuria, and polydipsia for 10 days. Clinical examination was unremarkable. The laboratory results showed an elevated serum calcium and 25(OH)D, which were suggestive for a vitamin D intoxication. The prescription of ADEK supplements clearly read 800 IU vitamin D per capsule, but re-examination of the preparation schedule by the pharmacist revealed a manufactured dose of 8,000,000 IU vitamin D, i.e., a 10,000-fold of the prescribed dose due to a dilution error. Intravenous hyperhydration was started immediately and loop diuretics were administered. After 31 hours, calcium had decreased only slightly (-0.22mmol/L, to 4.57mmol/L) and a single dose of pamidronate disodium (1mg/kg) was infused. The symptoms resolved gradually and after 4 days serum calcium was normal. Serum 25(OH)D progressively decreased but remained elevated after 4 months. Kidney and heart ultrasounds were normal over the disease course.

Conclusion
Vitamin D intoxication should be ruled out when patients with CF present with acute polyuria, constipation, and weight loss. In the case of a vitamin D intoxication, prompt treatment is necessary to avert life-threatening complications. To prevent this iatrogenic disorder, we advise regularly measuring serum calcium and 25-hydroxyvitamin D (25[OH]D) concentrations in children with CF receiving ADEK supplements during their follow up.

P 68.

Congenital hyperinsulinemic hypoglycemia (HH) requiring treatment as the presenting feature of Kabuki syndrome

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Cliniques universitaires Saint-Luc

Congenital HH is a neonatal disorder mostly caused by monogenic diseases with mutations in genes involved in the regulation of insulin secretion. However, HH is also a feature of various genetic disorders as Beckwith-Wiedemann, Sotos and Kabuki syndromes.

Kabuki syndrome is rare with 1-9/100 000 prevalence. This condition is characterized by dysmorphic facial features, cardiac malformations, skeletal abnormalities, postnatal short stature, and neurodevelopmental delay. Feeding difficulties are often present. Genetic mutations (KMT2D, KDM6A genes) are found in 70% of cases. HH is described in 0,3 to 1% of Kabuki patients. KDM6A variant is more likely associated to HH than KM2TD variant. Glycemic control in patients with HH is mostly achieved with diazoxide. However, some patient still present hypoglycemia under diazoxide treatment. Long-acting Lanreotide is an efficient treatment improving glycemic control.

We describe the case of a girl born at 37 weeks 6/7 of gestational age after a non-complicated pregnancy, without any gestational diabetes. Birth weight was 3950 g, birth length 50 cm and Apgar score 6/8/9. On day one, she presented refractory hypoglycemia. Additional tests revealed a HH. IV glucose, nasogastric feeding with enriched milk and diazoxide were required. Further investigations (MRI, EEG, metabolic tests, and cardiac US) did not reveal any anomaly except a persistent oval foramen. Due to persistent feeding difficulties, a gastrostomy was placed. At 6 months old, she presented progressive neurodevelopmental delay. Diazoxide induced feeding difficulties, justifying the prescription of somatostatin analogues. Lanreotide (increased up to 90 mg/month) treatment was introduced at 7 months old, which improved glycemic control. Genetic hyperinsulinism panel was negative but complementary genetic screening revealed a heterozygous truncating variant in the KDM6A gene, known as cause of X-linked dominant Kabuki syndrome.

In this case, the persistence of congenital HH was the presenting feature of Kabuki syndrome. In case of persistent HH, especially if associated with other features (alimentation difficulties, neurodevelopmental delay, dysmorphic features), syndromic HH should be considered and therefore complementary genetic analyses should be realized. Glycemic control will mostly be achieved with diazoxide. In case diazoxide-resistant HH, long acting-lanreotide is an efficient and well-tolerated alternative for normalisation of glucose control.

P 69.

Delayed puberty in a girl with a 46, XX gonadal dysgenesis due to a mutation in the PSMC3IP gene

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We report the case of a patient with a 46 XX gonadal dysgenesis caused by a homozygous frameshift variant in the PSMC3IP gene.

A 13-year-olg girl was referred to the endocrine clinic because of absence of pubertal development. Clinical examination revealed almost complete absence of puberty characterized by Tanner stage M1 P2. She had normal female external genitalia. She had no dimorphic features.

Parents are consanguineous. Age at menarche was 12 years for the mother and a paternal aunt presented late menarche (16 years). There were no reproductive problem.

Hormonal evaluation showed high basal gonadotropin levels: LH 43.22 IU/L and FSH 147.68 IU/L. Estradiol was undetectable and testosterone level was very low (0.14 ng/mL). DHEA-S and delta 4 androstenedione levels were normal, respectively 862 ng/ml and 0.21 µg/ml. 17-OH-Progesteron was also in the normal range (0.15 µg/L). Thyroid function was normal as well as growth function IGF-1 (264 ng/ml).

Karyotype was 46, XX. Abdominal ultrasound and magnetic resonance imaging showed undetectable gonads and severe uterine hypoplasia. Bone age was evaluated at 9.58 years for a chronological age of 12.7 ans. Heart ultrasound was normal as well as renal ultrasound. Genetic testing detected a homozygous frameshift variant in the PSMC3IP gene. This variant was classified as likely pathogenic. Management of this patient with primary ovarian insufficiency consisted in estrogen replacement therapy to induce breast development and regular menstruation at final stage.

46 XX female gonadal dysgenesis (46 XX GD) represents a very small part (around 5%) of 46 XX disorders of sexual differentiation. Clinical presentation is heterogeneous and most cases remain without a clear genetic diagnosis. In our case, the genetic analysis identified a variant in the exon 8 of the PSMC3IP gene, a gene involved in meiosis and DNA repair. Until now, only two mutations in the PSMC3IP gene have been reported in two consanguineous families with 46 XX GD. As mutations in specific genes affect only few women, functional studies and additional reports on affected women will allow to better understand the pathogenesis of 46 XX GD.

Long Oral Presentation

LO 7.

Pediatric Procedural Sedation and Analgesia (PROSA) in the Leuven University Hospitals: An efficacy and safety analysis.

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Background
The hospital can be a stressful and pain-inducing environment for a child. Nociception and stress during procedures can be minimized with, both non-pharmacological (e.g. vacuum mattress, distraction, glucose) and pharmacological (e.g. nitrous oxide, dexmedetomidine, chloral hydrate) techniques. Since there is an increase in demands for pediatric procedural sedation, the PROSA team initiative was started in the University Hospitals in Leuven in 2014. This team consists of trained and skilled nurses and performs sedation and/or analgesia on children. In this study, we assessed the efficacy and safety of this PROSA project in our hospital setting.

Methods
Demographic (age, sex) and clinical (procedure, sedation method) data were prospectively registered by the dedicated PROSA team. Anonymized data (11/2014- 6/2021) were entered in a database for analysis. We hereby defined efficacy as 'successful procedure' and safety as 'the absence of an adverse event' (saturation decrease with oxygen supply or prolonged stay).

Results
After excluding adults and neonates on the neonatal (intensive) care, data on 5090 procedures were available. The median age was 4.8 years (0.1-17.9 years), 53.1% were boys. Based on American Society of Anesthesiology classifications, 68.8% were class I, 24.9% class II, 4.7% class III and 0.1% class IV.

The procedures assisted by the PROSA team were 31.3% gastro-coloscopy, 14.2% DMSA scan/EC scan, 10.0% CT scan, 9.1% MRI scan and 4.8% bronchoscopy. A vacuum mattress was used in 21.6%, distraction in 21.3%, nitrous oxide in 15.5% and dexmedetomidine in 9,9%.

The sedation was successful in 98.0% of patients, be it classified as 'technically difficult' in 3.3%. Of the 2% of failed procedures, 69% were due to inadequate sedation and 31% to logistic reasons unrelated to sedation (such as puncture problem, poor bowel preparation).

The overall adverse event incidence was 2.0% and occurred predominantly during gastro-coloscopy (2.7% of all gastro-coloscopies) or bronchoscopy (15.6% of all bronchoscopies). Pethidine/midazolam IV was used in 81 cases, nitrous oxide in 10 cases. A saturation decrease with oxygen supply was the documented adverse events in 83 cases. There were no deaths or permanent damage reported.

Conclusion
With the current training and implementation of the team, PROSA is effective with a success rate of 98.0% and with a low adverse event rate, clustered in gastro-, colo- or bronchoscopy.

LO 8.

Success of a tertiary care program for children with severe obesity

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Zeepreventorium

Zeepreventorium is a residential centre that helps children with chronic disease and their families to work on specific rehabilitation needs such as adherence to therapy, physical condition, psychosocial balance, social functioning and school attendance. Children reside in groups of 10 according to age. The focus is on adopting a healthy life style. Apart from group activities and coaching, the program includes individual therapeutic sessions with a psychologist, dietitian and physiotherapist.

Aim
to prospectively evaluate the effectiveness of a tertiary care program for children and adolescents with severe obesity using objective parameters.

Methods
predefined, objective parameters were prospectively collected in the weeks after admission and the weeks before discharge. Inclusion criteria were admission and discharge in the year 2020 and residential stay of at least 3 weeks. Parents and youngsters gave informed consent to this study. Domains studied included medical comorbidities, psychological and social functioning, strength and fitness. Data are expressed as mean ± SDS.

Results
Data on 54 youngsters (25 males, 29 females) were collected; mean age: 14.5 ± 2.5 years, mean stay duration: 269 ± 102 days.

Worsening physical functioning and poor adherence were the most frequent rehabilitation needs.

At least 10% loss in BMI was achieved in 76% of the patients. However, those with a stay <2 months did not achieve this target. Mean body fat measured by DEXA-scan decreased from 51 to 39% ($p<.0001$). Mean number of comorbidities decreased from 5 to 2 in children >12 years and from 4 to 1 in those <12 years.

Mean quality of life score assessed by PEDSQL improved from 65 ±13 to 77±12 (paired t test $p<.001$). Youth self-report questionnaire and eating disorder examination questionnaire score improved respectively in 89% and 75% of the patients. Parental education charge questionnaire ameliorated in 93% of patients.

Functional strength measured by the Bruininks Oseretsky test improved in 88 to 98% of patients, highest for V-up and lowest for push-ups. Fitness measured by intermittent shuttle run or Cooper test improved in 96% of the patients (chi square test $p<.001$)

Conclusion
children with severe obesity benefit from a residential stay in a rehabilitation centre by improving their physical and mental health and enhancing their strength and condition.

Long Oral Presentation

LO 9.

Simplified management for suspected allergy to oral beta-lactams in children

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CHC MontLégia, CHR Huy

Abstract

Objective of the study: To assess the efficacy and safety of simplified management of suspected oral beta-lactams allergy in children.

Methods

We carried out a retrospective study based on data collected prospectively from February 2016 to January 2022, referred to the consultation for suspicion of allergy to beta-lactams. In children presenting benign and/or delayed reactions, a 3-day oral challenge test (OCT) was performed directly. Intradermoreaction (IDR) were performed only when the reaction was severe or after a positive OCT. When the OCT was positive, an alternative treatment was implemented.

Results

232 patients younger than 17 years were included in the study. The mean age was 56 months (±44 months). 102 (44%) patients were female. The suspected antibiotic was amoxicilline for 172 (74%) patients, amoxiclav for 49 (21%), cefuroxim for 7 (4%), cefadroxil for 3 (1%) and flucloxacillin for 2 (1%). 33 patients (10%) presented a history of immediate reactions. Two hundred thirty (99%) of whom completed an OCT. IDR were performed in 12 children, but were positive in 4 patients only (2 before OCT, and 2 after OCT). 2 patients with a positive OCT were not subjected to an IDR. OCT was positive during the procedure or within a few hours after the completion of the test in only 7 patients, and after 7 days in 1 patient. 2 patients had subjective symptoms and were thus subjected to a double blind OCT that was negative. Therefore, the final diagnosis of allergy to beta-lactams was confirmed in only 8 patients (3.4%).

Conclusion

Beta-lactam allergy is suspected in many children taking antibiotic treatments. In general, the management of the allergic reaction is adapted from the standard procedure in adult care that includes the realization of IDR. However, this kind of allergy is rarely confirmed after OCT. IDR are time-consuming and painful, and should be performed only in some cases that present a higher risk. OCT must be carried out in a hospital environment by an experienced team and with an intensive care unit nearby. Acute generalized exanthematous pustulosis, Lyell and Stevens-Johnson syndromes remain a contraindication to the realization of IDR and OCT.

LO 10.

Child, adolescent, and parent mental health in general population during a year of COVID-19 pandemic in Belgium: A cross-sectional study

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Université Libre de Bruxelles

Background

More than two years after the discovery of the SARS-Cov-2 in end 2019, the COVID19 pandemic is still the focus of discussions. In many countries, lockdown was the first solution to limit the spread of the virus. The epidemiologic impact of these national decisions brought quick positive results but the psychological and developmental impacts on children and adolescents were not initially considered. Many studies emerged to address these gaps. This study aims to evaluate the mental health status of children, adolescents and their parents during the first year of COVID-19 pandemic in Belgium.

Method

A cross-sectional online survey was conducted between May 2020 and April 2021. Two hundred and eighteen adults and 273 children, aged from 7 to 17, fully completed the survey. Analysis compared results before and during the second national lockdown, started on November 2nd 2020. Multiscore Depression Inventory for Children (MDI-C) was used as a self-report scale to evaluate depressive symptoms in children and adolescents. Parents completed the Depression, Anxiety and Stress Scale (DASS-21) to assess depressive and anxious symptoms. Optionally, parents could complete the Child Behavior Checklist (CBCL) to report externalized or internalized symptoms in their children.

Results

Almost one out of five children (17.9%) presented moderate-to-severe scores of depression. Adolescents presented a higher level of depression than children. The rate of moderate-to-severe depression scores (10.8% to 21%, $p = 0.007$) and internalized symptoms increased during the second lockdown ($p < 0.001$). Parents' depression ($p < 0.001$) and anxiety ($p = 0.027$) levels also increased during the second lockdown. Logistic regression showed that the use of psychotropic medication in parents and parents' depression scores were risk factors for worse children's depression scores.

Conclusion

Although the first lockdown has disrupted the daily life of many families, the second lockdown appears to worsen the effects of the pandemic on children's and parents' mental health. Scores in MDI-C were suggestive of depression in almost half of the children while more than half of the parents present symptoms of depression. There is a need to implement specific interventions targeting both children/adolescents and their parents to support them during lockdown periods and improve mental health outcomes.

Short Oral Presentation

SO 1.

A framework for interprofessional collaboration towards sustainable paediatric drug development

E. Degraeuwe, D. Christiaens, L. Persijn, R.M. Fernandes, T. van der Geest, L. Nuytinck, A. Raes, M. Turner, J. Vande Walle

U Gent, UZ Gent, University of Lisbon, Radboud Institute for Health Science, University of Liverpool

Background/Aims

Close collaboration is of utmost importance to design, coordinate and execute high quality clinical trials in children. This leads to multiple initiatives, such as the conect4children (c4c) network, an Innovative Medicines Initiative 2 (IMI2) funded program facilitating paediatric clinical trials in 20 European countries, through National Hubs and their host National Networks. However, aside from investing in a durable and rooted clinical trial network the same investment must be made into the investigators and clinical coordinators involved, since the experience of site, coordinators and investigators is still unsatisfactory due to the relatively recent increase of paediatric clinical trials.

Method

Within the c4c network, the Belgian National Research network, managed through the coordinating center at Ghent University Hospital, plays a prominent role for innovation and study conduct. The network leads the development of a Young Investigators' Community (YIC) within the c4c network and the development of a National Clinical Research Coordination (CRC) Community in Belgium.

Results

The YIC kicked-off in May 2021 and regular meetings have occurred since then. An agenda and working plan were developed collaboratively based on both formal and informal approaches. Over 30 people have joined the meetings from 18 countries. Learnings on how to educate, motivate and improve clinical study conduct are shared. Process optimalisation of feasibility conduct and national sustainable models are optimized and piloted directly within the coordinating centers per country. Feedback indicates the need for further connection considering the current remote home-working environment and demonstrated increase in efficiency and problem-resolving. The first meeting of the Belgian CRC occurred in March 2021. All 15 sites in Belgium that are connected within the network have a CRC delegate. Topics within the meeting include overview of site needs, proposed and conducted trials, data harmonization and recruitment improvements.

Conclusion

The enthusiasm within both YIC and CRC communities reflects the need to bring together and train young people. The CRC community creates an essential level within the Belgian Clinical Trial network, aiming to harmonize and improve drug development in children. Both interconnecting community platforms pave the way to sustainable and improved drug development in paediatrics.

Short Oral Presentation

SO 2. B

oth medical and context elements influence the decision making processes of pediatricians.

L. Schurmans, D. De Coninck, B. Schoenmakers, P. de Winter, J. Toelen

KU Leuven, UZ Leuven, Spaarne Gasthuis

Aim
We wanted to investigate the relationship of medical and non-medical factors with the clinical decision-making of pediatricians. We hypothesize that the addition of relevant medical information (be it alarming or reassuring) will influence the physician's decision making, but that the addition of non-medical information will also play a role in this process.

Methods
We designed an online questionnaire that contained four demographic questions (gender, age, years of experience, and the number of biological children) and ten clinical case-based scenarios, of which five focused on medical factors and five on non-medical/context factors, each scored on a 5-point Likert scale. The collected data was subjected to a descriptive analysis and linear regression analyses (one per case) to assess what pediatricians' demographic characteristics are linked to pediatricians' decision making in each case.

In total 113 participants completed the online questionnaire. The participants were Belgian pediatricians that are members of the Vlaamse Vereniging voor Kinderartsen or pediatricians in training at KU Leuven.

Results
Overall, the additional medical and non-medical/context factors were considered relevant to change the initial decision in most cases. Additional information of an alarming nature induces the physician to become more worried, whereas reassuring information decreases this worry. In some cases with the medical factors, the gender and the age of the pediatrician does have some effect on the clinical decision making. The number of children does not seem to have an influence.

Conclusions
Medical decision making is affected by multiple intrinsic and extrinsic factors that differ between physicians. Our data indicate that these non-medical factors must be considered when making a medical decision, as it is crucial to be aware that they have a substantial influence on that decision making.

Short Oral Presentation

SO 3.

A tool for home-based cow's milk reintroduction in children with non-IgE-mediated cow's milk allergy: The revised Flemish milk ladder

L. De Vlieger, T. Bosmans, M. Diels, L. Nuyttens, I. Hoffman, M. Raes, J. Leus, S. Verelst, D.M.A. Bullens, K. Coppens

KU Leuven, UZ Leuven, Jessa Ziekenhuis Hasselt, AZ Maria Middelares, Imelda Ziekenhuis

Background
The majority of cow's milk allergic children tolerate extensively heated milk, referred to as "baked milk", and consuming baked milk has shown to accelerate tolerance towards unheated milk. In the UK, clinicians developed a "milk ladder" as a tool to gradually reintroduce milk-containing foods from lowest to highest allergenicity in non-IgE mediated cow's milk allergic children. This milk ladder forms the basis of the NICE-guideline recommendations on cow's milk reintroduction. However, this "British milk ladder" requires country-specific adjustments, as it contains several traditional British dishes. Hence, we designed a Flemish version of this milk ladder containing foods more representative for the Belgian eating habits.

Objective
We investigated the utility and user-friendliness of the home-based gradual reintroduction of cow's milk in children with non-IgE mediated cow's milk allergy (CMA), using the newly designed Flemish milk ladder (s59587).

Methods
The Flemish milk ladder was developed for two age groups, children aged 1-1.5 years and 1.5 years and older. In the youngest children cow's milk was reintroduced through 15 steps, while in older children the milk ladder contained 17 steps. The milk-containing foods at each step were consumed for at least 3 days, in agreement with the physician. The utility and user-friendliness was evaluated through surveys, open interviews and expert panels composed of academics, clinicians and dieticians.

Results
We received 22 surveys from pediatricians and parents, with an average satisfaction score of 75%. Complete cow's milk tolerance was achieved in 17/22 children, whereas 2 children developed merely partial tolerance for fermented milk. Three children were put back on an elimination diet due to diarrhea, cramps or eczema, but also experienced more severe symptoms at initial presentation. The main suggestions of experts in the field and parents were incorporated into a new version of the milk ladder, which included: combining the two ladders for different age categories into one, reducing the number of steps to six, eliminating pasteurized milk as most parents weren't familiar with it, offering "healthier" foods (e.g less sugar) within each step along with recipes and adding the choice to reintroduce growth milk and infant formula.

Conclusion
we present a helpful and safe guide to gradually reintroduce cow's milk at home in children with non-IgE mediated CMA, considering the Belgian eating

Short Oral Presentation

SO 4.

Clinical decision making in adolescents: parental perspectives.

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KU Leuven, UZ Leuven

Background/Aims

Medical decision making in adolescents can lead to family conflicts on autonomy and privacy. As the law in The Netherlands and Flanders allows adolescent involvement in healthcare decisions, it is necessary to have a more detailed insight in the overall parental opinion on confidentiality and consent regarding minors. Although research on this topic has been growing the last decade, few studies have used a case-based questionnaire to identify specific parental preferences in this context. The aim of this study was to get a representative view of the Flemish and Dutch parental opinion on this matter and to identify demographic variables with a significant correlation. Finally, we tried to identify patterns in these reactions and compare them to specific parenting styles.

Methods

We presented 6 cases (3 on confidentiality and 3 on consent) to 1.000 Belgian (Flemish) and 1.000 Dutch participants and asked them to respond as if they were the parent of the described minor. Inclusion criteria were age (35-55 years old) and gender (50/50 gender distribution). Other demographic variables such as education, number of children and family composition were also collected. We performed descriptive statistics, a multivariate logistic regression and a latent class analysis on the data.

Results

In both populations the majority of respondents follow the adolescent's perspective in the context of consent (migraine medication, decision on surgery). However, they diverge from the adolescent's preference with regard to confidentiality (cases regarding STD, depression and ultrasound report) and one consent case (ADHD medication). Surprisingly, higher educated respondents would rather not be informed about an STD or depression. Our latent class analysis shows that the respondents could be classified as authoritative (47%), permissive (30%) or authoritarian (17%) regarding their parenting style.

Conclusion

This study shows that Flemish and Dutch respondents are willing to grant autonomy to an adolescent in medical decisions, but rather regarding consent than confidentiality. They could be classified in putative parenting styles based on their responses. Future research could provide concrete data to help physicians in their interaction with adolescents and their parents during medical decisions.

Short Oral Presentation

SO 5.

Health Literacy Among Caregivers of Children With IgE-mediated Allergy at Risk of Anaphylaxis

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VUB, UZ Brussel, AZ Maria Middelares Gent, UZ Antwerpen, UZ Leuven, KU Leuven

Background

Anaphylaxis is an acute, life-threatening, allergic reaction of which the prevalence is rising. Previous studies in anaphylaxis management at school age have shown that the self-efficacy score can be improved by education. We hypothesized that a training session about anaphylaxis can improve the level of health literacy of caregivers of children at risk of anaphylaxis, by increasing their level of knowledge and self-efficacy.

Methods

In a prospective cohort of caregivers from children with IgE mediated food or insect venom allergy, we investigated the effect of a training session about anaphylaxis on top of standard care provided by the regional hospital, on their knowledge and self-efficacy. This effect was measured by means of a three parted questionnaire: 1) knowledge score, 2) validated Dutch-translated self-efficacy (S.PER.SE-FAAQ) score (both pre and post training) and 3) a subjective score on the additional question about the added value of the training session. We used non-parametric statistics for the data analysis.

Results

The training session was attended by 140 caregivers, 116 consented for participation, 71 completed both questionnaires. Baseline knowledge in the total group (non-responders of the post training questionnaire= partial responders included, n=116) was high (median 66.7%), similar in complete and partial responders, significantly higher in younger caregivers (<55 years, n=49/71) and tended to be higher in first-degree relatives (n=29/71). Baseline self–efficacy was high in the total group (80%) without differences amongst the different subgroups. Median score of knowledge improved significantly after training in the complete responder group (from 66.7% to 83.3%; $p<0.001$) as well as in all subgroups separately. Median score of self-efficacy also increased significantly both in the total group (from 80% to 85%; $p<0.001$) as in all subgroups after the training. This improvement in the complete cohort, was considered clinically relevant. Most of the participants indicated that the training session had a high added value and made them feel more confident.

Conclusions

Education by a training session offered by regional hospital allergologists on top of standard care given by pediatricians significantly improved the total score of knowledge and self-efficacy of all caregivers of children with IgE-mediated allergy at risk of anaphylaxis. This indicates that training sessions should become standard of care for those caregivers.

SO 6.

Psychiatry for Transitional Age Youth (16 to 24 years old): innovative outpatient care organised between paediatrics and adult mental health services

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HUDERF, Erasme Hospital, Brussels, ULB

Background/Aims

Transitional age youth (TAY) are a particularly at-risk population in mental health. They have specific needs, not currently covered between child and adolescent mental health services (CAMHS) and adult mental health services (AMHS), mainly because of existing barriers. This retrospective study was carried out to describe sociodemographic and clinical characteristics of patients who attended a new outpatient program specifically conceived for TAY in Brussels, Belgium.

Methods

The total sample included 243 TAY (16 to 24 y.o.) who presented for care in this outpatient program between October 2019 and May 2021. Outcomes related to trajectories of psychiatric care were retrospectively extracted from each patient's electronic medical records and analysed, such as leading symptom, consultation's referral and requester, and final orientation.

Results

The sample was mainly composed by female (59.3%); the average age was 18.7 (± 2.0) years. General practitioner (18.9%), child and adolescent psychiatrist (18.1%), psychologist (11.5%) and adult psychiatrist (7.4%) referred patients. Leading symptoms were divided into three dimensions: internalizing (67.5%), externalizing (21.8%) and psychotic (10.7%). Leading symptom differed according to sex ($p<0.001$), with internalizing symptoms more frequent in women, externalizing and psychotic symptoms more frequent in men. Patients presenting psychotic symptoms were significantly older than both those with internalizing ($p=0.016$) and externalizing symptoms ($p=0.008$). After first assessment, 81.5% of youth were followed-up in our outpatient program, without any difference according to sex ($p=0.081$) or leading symptom ($p=0.092$). Overall, at the end of data collection, youth were orientated towards ongoing follow-up in this program (37%), AMHS care (21.8%), end of psychiatric care (17.3%) and CAMHS care (4.1%). 19.8% patients discontinued the proposed care.

Conclusion

Classical boundaries, determined by artificial variables such as age or type of psychopathology, do not seem to be efficient criteria to achieve a good quality psychiatric evaluation and continuity of care in TAY. This psychiatric outpatient program for TAY represents an innovative contribution to reinforce CAMHS-AMHS interface in Brussels' French-speaking services. The analysis of trajectories in psychiatric care suggests positive outcomes of this TAY-tailored clinical program to achieve high quality standard of care in youth mental health.

P 32.

Juvenile dermatomyositis with mildly elevated muscle enzymes

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UZ Gent, AZ Maria Middelares

Juvenile dermatomyositis is a rare systemic autoimmune disease characterised by inflammation of small vessels within muscle, skin and major organs. The clinical presentation of juvenile dermatomyositis can be quite diverse. We would like to present a young patient with an atypical presentation of juvenile dermatomyositis.

A Caucasian 4 years old boy was referred because of a painful and abnormal gait. At first, he complained of pain after walking, but more and more he started walking with waddling steps or refusing to walk at all. Parents also noticed depigmentation in his elbow folds and dry dark skin on legs and arms. He had no gastrointestinal complaints but he did lose weight. Full spine and brain MRI showed no abnormalities. Lab examination showed mild elevated inflammatory markers, and mild elevated CK and LDH. Clinical examination showed a rash fitting with ichtyosis, clear desquamation at both elbows and inguinal region of both legs, and discrete erythematous rash at the dorsal aspects of some metacarpophalangeal joints. He had flexion contractures of wrists, elbows, hips, knees and ankles with painful movements of all these joints. He could only walk a couple of steps with a waddling gait. Laboratory examination showed mildly elevated inflammatory markers, normal CK, slightly elevated aldolase and LDH. Auto-immune screening showed ANF 1+, anti-ENA -, and negative myositis-specific antibodies. He was started on Naproxen. MRI of both legs showed diffuse myositis at the pelvic girdle and both legs without any signs of arthritis. Capillaroscopy showed a scleroderma-like pattern. Based on the combination of the clinical picture, abnormal capillaroscopy and MRI, the diagnosis of juvenile dermatomyositis was made.

Establishing the diagnosis of juvenile dermatomyositis can be quite challenging. We report a 4-year-old boy with an atypical presentation. Initially his complaints were most suggestive of a neurological condition but normal muscle enzymes were found so myositis was assumed less likely. Also, the skin manifestations in this patient were not typical for dermatomyositis. The diagnosis was made based on the combination of the clinical picture and an abnormal MRI and capillaroscopy. Our case shows that muscle enzymes not always correlate with the degree of inflammation, especially when time without therapy increases. In the future, it might be useful to include imaging to the diagnostic criteria, as it is not invasive but has a high sensitivity.

P 33.

Pediatric mistreated child care paths: criteria for hospital care

S. Pannizzotto

CHU Liège

Background
Pediatric services are on the front line when there is an emergency, safety and placement provisions are applied to minors. Likewise, when the clinical lesions are so serious that they require often intensive somatic management, the issue of monitoring the child once somatically "healed" is crucial.

Method
In addition to intra-family mistreatment situations which can be followed on an outpatient basis, there are clinical situations relating to child mistreatment which require a medico-psycho-social assessment in the hospital. Because the hospital is not a place for a child to live, this type of care must meet strict criteria. The Cellule Maltraitance pediatric unit of the CHU Liège pediatric service, provides this type of care. An assessment can be initiated according to three different care paths identified as follows:

- The request for a hospital assessment by the SAJ or the Justice Department.
- The arrival of the child via the emergency room.
- Screening for a situation of mistreatment or at risk of being mistreated on one of the sites of the CHU Liège pediatric service.

However, in order to offer an effective therapeutic transition, it is necessary to take into account specific hospitalization criteria:

- Failure of a prior outpatient assessment
- The physical and / or psychological danger to which the child is exposed at home, thus preventing any possible outpatient work.
- The need for the child to be supported in a multidisciplinary manner in a neutral environment in order to free and respect his word without any contamination or possible influence. - The need for intensive care both individually and in terms of family dynamics for which the various hospital specialties are required.
- The need for a strict framework allowing effective parental collaboration.
- The lack of a reception structure currently available from which care could be taken.

Results
Around 50 to 60 assessments are carried out per year within the Cellule Maltraitance service. On average, we observe: - 53% physical abuse - 14% sexual abuse - 11.5% serious neglect - 21.5% psychological abuse, parental conflicts. The required hospital stay is rarely exceeded (less than 5%). On average, the orientation given is 70% return to the family and 30% long-term placement orientation.

Conclusion
The care path for an abused child must be considered according to criteria that will allow adequate care and a beneficial development.

P 34.

Transition process from pediatrics to adult medicine services: support systems in pediatric surgery

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HUDERF

Background
In the surgical department of The Queen Fabiola Children's University Hospital (HUDERF), several professionals have thought about a transition programme for patients operated on oesophageal atresia or diaphragmatic hernia at birth. The main objective of this program is to maintain continuity of care. The transition process coincides with adolescence which is characterized by a period of crisis.

Methods
The reflexions have started with a review of the literature about the transition. The medical and the psychological views have been discussed to create a programme in collaboration between the surgeon and the psychologist of the surgery service. A time is always set aside on each consultation for the parents as their presence can be a valuable support and help. Our program starts with a consultation with the referring doctor of the patient (gastro-paediatrician, pneumo-paediatrician and paediatric surgeon) during which the transition issue is discussed more concretely. There is then a co-consultation with the referring doctor and the psychologist. Objective aspects are discussed such as knowledge of the disease, the concrete organisation of adult care, but also more subjective aspects concerning the representation of the body or the patient's current situation and the perspectives to be considered. Then, there is a first meeting between the patient and the adult health care team in the presence of the paediatric team (referring doctor). A medical file tracing the patient's health history is transmitted to the adult doctors to ensure continuity of information. The paediatric and the psychology services also organise a day dedicated to the transition with the presence of the patient, his family, the paediatric and the adult team and some former paediatric patients who have experienced the transition.

Results
At the hospital, we consider the patient and his family at the heart of our concerns. This program was developed with those preoccupations to ensure the continuity of the care despite the shift change and the psychological and relational changes related to the adolescence. It still needs to be evaluated.

Conclusion
The literature has highlighted a lack of transition program protocols and consensus around the concepts. The proposed program is built on continuity, multidisciplinarity, temporality, narrative activity and rituality. This program could be used as a basis for thinking about other transitions.

P 35.

From conception to birth of a paediatric research community

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Background/Aims
Academic research has traditionally progressed in one direction, where promotor supervision and conceptualizing of research projects have been performed with minimal input from other scientific stakeholders and/or community. Young researchers have expressed the need to gain knowledge and information from peers and colleagues through a community platform. Informal networking with colleagues and peers, including teachings of vast evolving meta-skills such as patient engagement, has been identified as essential in all stages of a scientific career.

Methods
The PhD Community of Internal Medicine and Paediatrics is a unique community by and for PhD Students, residents, and professors. The PhD community is located within the Department of Internal Medicine and Paediatrics at Ghent University. Through a community platform knowledge, expertise, and (multidisciplinary) collaboration opportunities can be induced. By combining formal and informal meetings, the colleague community spirit can be improved between scientific profiles at our institution. This report describes the establishment and progression over the past 18 months.

Results
Since the establishment in August 2020, 24 sessions have been organized with over 350 participants in total. Sessions are both informal and formal style and include a bi-annual open session which is distributed to international and multidisciplinary stakeholders. After an initial start-up with paediatric academics, an expansion to Internal Medicine was made in May of 2021. The number of members has substantially grown to 230 members in our department. Over a short period, the community has grown to be a well-rooted platform for academics to learn new capabilities, present their research, and inspire collaborations. The PhD Community of Internal Medicine and Paediatrics has collaborated within other student networks in medicine such as the Belgian Medicine Students' Association to fill potential educational gaps and stimulate advocacy efforts for a scientific career. A welcome kit for new PhD Students and a mentorship program is expected for March 2022.

Conclusion
A PhD-Community platform in a paediatric setting has been shown to improve collaborations, knowledge, and general well-being of its science faculty. Similar establishments are necessary within other departments and/or universities to aim for a sustainable and substantial impactful scientific community within Belgium.

P 36.

Challenges of creating a patient engagement group in a paediatric setting

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Background/Aims
After the 2007 Paediatric Regulations, a vast increase in the amount of paediatric clinical trials has been initiated. Many trials were considered unsuccessful, partially related to poor study design, but mostly due to insufficient recruitment rates and low trial completion. Off-label use of drugs in a paediatric setting still remains around 60%. However, incorporating patients' perspectives and insights within the whole drug development process has proven to improve clinical trial design, recruitment and retention rates, and overall resulted in better health outcomes and benefits for all the stakeholders involved. Yet patient engagement (PE) is currently not considered standard practice. European countries such as France, Spain and Italy have a well-developed paediatric PE network. A pilot study was designed focused on a PE group within the field of paediatric nephrology at Ghent University Hospital.

Method
This study describes the challenges in creating PE groups within a paediatric setting. As case-study, the start-up of the PE group within paediatric nephrology at Ghent University Hospital will be portrayed.

Results
PE group design and recruitment methodology should be carefully standardized and improved. When involving children and young patients in drug development, the social, cultural, and economic environment of the participant should be considered. For the start-up of our local Belgian PE group, support from the European Young Person's Advisory Group Network (eYPAGnet) was provided to form a common framework. However, when locally applying this framework, the development was confronted with additional challenges. An alternative approach to receive funding had to be taken. PE session materials, such as tailored questionnaires and age-appropriate educational materials on clinical trials were locally developed. Collaborations with the children psychology department has proved to be beneficial. Moreover, ethical committee submission process had to be initialized considering the novelty of the PE group design.

Conclusion
Aside from being widely agreed upon that PE of paediatric patients and their families is important, translation to actual national and local groups has not been up taken. Recently, Ghent University Hospital has successfully designed a PE group in paediatric nephrology, paving the way for other national PE initiatives. Local build-up experience and know-how should be broadly disseminated and standardized

The profile and future expectations of young paediatricians and paediatric trainees in Belgium: a first national surveyThe profile and future expectations of young paediatricians and paediatric trainees in Belgium: a first national survey

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Background/Aims
As paediatrics is a constantly evolving specialty, the profile and expectations of future paediatricians in Belgium are also changing over time. Previous surveys have mapped this profile and expectations among paediatric trainees. However, these surveys were conducted in 2012 and 2015 and were limited to the Flemish universities, making it unclear how these data evolved in time, nor if they are representative on a national level.

Methods
Between October 1st, 2021, and December 6th, 2021, the youth representatives of the Flemish Society of Paediatrics (Jong VVK) conducted a comprehensive survey on (future) professional preferences among paediatric trainees and recently graduated paediatricians in Belgium. The survey comprised 25 questions and 9 statements, and the emphasis was placed on subspecialisation and (future) work perspectives.

Results
The survey was completed by 287 respondents (53.3% Flemish, 46.7% French-speaking), of which 60% were trainees. The average age of all respondents was 29.3 years old and the majority were female (82.5%). This survey was the first to include data from all universities in Belgium with a representative spread over the 7 different universities. In line with previous surveys, 80% of the respondents indicate wanting to subspecialize and only 1% prefer to work exclusively in private practice. The 3 most preferred subspecialties were neonatology (13.2%), pneumology (11.9%), and infectiology (10.8%) with ‘interest in the field’ repeatedly (75.6%) declared as the primary motivation for wanting to subspecialize. Interestingly, large majorities indicate that a central overview of available fellowships (preparing for a subspecialty) would be very useful (93%) and a separate statute for fellows is mandatory (85%). For 2 out of 3 respondents, the presence of subspecialty criteria would affect their ambition to subspecialize.

Conclusion
Our unique dataset provides valuable insights into the desires and needs of the future generation of paediatricians in Belgium. Better structuring of fellowships and subspecialties is warranted to enhance the transition from training to graduation and optimize the career flow of future paediatricians