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FROM WOMB TO ADULTHOOD

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ABS 1

LESS INVASIVE SURFACTANT ADMINISTRATION (LISA). RESULTS IN REA MATERNITY HOSPITAL NEONATAL INTENSIVE CARE UNIT OF ATHENS

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BACKGROUND

Less invasive surfactant administration (LISA) is widely and increasingly used in Europe for the treatment of respiratory distress syndrome in preterm infants. There is a growing body of evidence that LISA-treated infants are at a decreased risk for bronchopulmonary dysplasia compared to intubation and mechanical ventilation and other non-invasive strategies of respiratory support. LISA reduces duration of ventilation and risk of bronchopulmonary dysplasia, both well-defined risk factors of adverse development.

Data on long-term outcome after LISA compared to intubation are scarce.

The results of LISA in the Neonatal Intensive Care Unit (NICU) of Rea Maternity Hospital (Athens, Greece) over the last 2 years are presented below.

METHODS

We compared the standard procedure of surfactant administration, that was applied via endotracheal intubation, and ventilation with a gentler approach called LISA: with LISA technique, surfactant was applied less invasively via a thin catheter, while the infant was breathing spontaneously on CPAP.

We have administrated surfactant at 537 infants over the period April 2019-December 2021.

The infants were further classified in 3 categories according to their gestational age (GA):

- Category 1: mature infants, ≥ 37 GA;
- Category 2: late preterm infants, 34^{+0}-36^{+6} GA;
- Category 3: preterm infants, < 34 GA.

We used Curosurf® at a dose of 200 mg/kg. No premedication was used.

RESULTS

- 237 (44%) of the infants received surfactant with LISA, while 300 (56%) via endotracheal intubation;
- 27 (5%) of the infants that received surfactant with LISA needed secondary intubation due to lack of respiratory improvement;
- 130 children of Category 1 (mature infants ≥ 37 GA) received surfactant, 126 (97%) via intubation, while 4 (3%) via LISA;
- 210 children of Category 2 (late preterm infants) received surfactant, 135 (64%) via intubation, while 75 (36%) via LISA;
- 197 children of Category 3 (preterm infants) received surfactant, 39 (20%) via intubation, while 158 (80%) via LISA.

COMMENTS

- Complications: pneumothorax in one infant after LISA;
- the majority of infants in Category 1 has received surfactant via intubation because nCPAP is not preferred as a method of RDS therapy in mature infants at our NICU and that is why LISA was performed less in this group;
- among the 237 infants that received surfactant with LISA, intubation could be avoided for 210 of them, while 27 needed a secondary intubation.

CONCLUSION

Without doubt, LISA patients showed an immediate improvement of the oxygenation, a shorter duration of any other form of ventilatory support as a lower percentage of intubation and mechanical ventilation in the first 72 hours.

Data on long-term outcome after LISA compared to intubation are rare, so that further follow-up is mandatory.

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ABS 2

ENDOCRINE DISRUPTING CHEMICALS EXPOSURE IN BREAST MILK AND INFANT NEUROBEHAVIORAL AND GROWTH OUTCOMES: THE LIFE-MILCH PROJECT

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The Life MILCH Project [1] is a longitudinal and prospective pilot study that aims to determine the correlation between levels of maternal milk contamination/exposure to endocrine disrupting chemicals (EDCs) and infants' neurobehavioral health in the first year of life in two geographic areas of Italy: Emilia Romagna and Sardinia.
500 pregnant women in the third trimester of pregnancy with no diagnosis of fetal anomalies or gestational pathologies are enrolled in the Parma, Reggio Emilia and Cagliari Hospitals. Mother-infant dyads are examined during the first year of life assessing pre- and post-natal EDCs exposure and infant health outcomes. At recruitment, EDCs exposure pre- and during pregnancy are assessed by 2 questionnaires about maternal clinical history, lifestyle, and nutritional habits referring to the last 6 months. Samples of maternal urine and plasma are collected. At birth, cord blood, placenta and clinical data on newborn's condition at birth are sampled. After delivery, the mother-infant dyads are recalled at 1, 3, 6, and 12 months of age to assess post-natal EDCs exposure through biological samples (maternal urine, infant urine, and breast milk or formula milk) and 2 questionnaires about mother and infants' lifestyle and nutritional habits. At each follow-up visit, infants' growth and neurodevelopment are assessed by neonatologist and psychologist. Infants growth parameters include weight, length, head circumference, size of the bregmatic fontanelle, puberal stages, and anogenital distance. Infant neurodevelopment assessment focus on perceptive, socioemotional, cognitive and behavioral development that are independently and respectively assessed at 1, 3, 6, and 12 months of age. The Visual Preference Paradigm (1 month of age) assesses the infants' visual preference for social or non-social stimuli (upright face vs upside-down face, respectively). The Face-to-Face-Still-Face test (3 months of age) evaluates the infants' socioemotional stress response to maternal unresponsiveness. Fagan test (6 months of age) measures the infants' ability to discriminate between novel and familiar stimuli as an index of cognitive maturation. The Barrier task (12 months of age) evaluates infant behavioral response to frustration during a play situation. At 6 and 12 months of age, Bayley III test is administered to assess the infant global neurodevelopment.
This is an ongoing study and results are not completed yet. A preliminary analysis has assessed possible differences of breastfeeding vs formula milk nutrition on early infant development. Since growth, perceptive, socioemotional, cognitive, behavioral and global developmental areas are susceptible to EDCs exposure [2], this ongoing study may provide early biomarkers of effects in infant neurobehavioral outcomes depending on EDC exposures and allow to evaluate possible differences in exposure between urban and rural areas in Italy.

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ABS 3

CHARACTERIZATION OF HUMAN MILK MESENCHYMAL STROMAL CELLS. POSSIBLE CLINICAL IMPLICATIONS

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Breast milk, by virtue of its composition in biologically active nutritional and non-nutritional factors, is the food of first choice for infant nutrition and the best solution for supporting the health and development of the baby.

Recently, the presence of milk-derived mesenchymal stromal cells (MDMSCs) has been demonstrated in breast milk, but to date the precise characterization of these cells is lacking and their role is still unclear.
In the present study, we characterized MDMSCs and investigated their potential contribution in the mechanisms of immunomodulation, neurodevelopment, and regulation of glucose homeostasis.
Twenty-one milk samples were collected, including 18 of colostrum and 3 of transitional milk. MDMSCs were isolated and cultured in Dulbecco's modified minimum essential medium (DMEM), fortified with 10% fetal bovine serum, in flasks previously coated with fibronectin.
Characterization of MDMSCs was performed by cytofluorimetry evaluating the expression of the main surface markers of MSCs: CD105, CD90 and CD73, and the expression of human leukocyte antigen DR (HLA-DR), which is responsible for the presentation of not-self antigens to the immune system. In order to verify the stem profile of MDMSCs and evaluate their possible fetal origin, gene expression levels of OCT4 and NANOG (pluripotency markers) and SRY were analyzed.
By real-time PCR, the expression levels of TNF-alpha, IDO1 and OPN as immunomodulation markers; BDNF, NT-3 and NT-4 as neurotrophic markers; and GLUT-1 and GLUT-4 as glucose transport markers were determined and compared between MDMSCs isolated from colostrum and transitional milk, and with placenta-derived mesenchymal stromal cells (PDMSCs).
All the cell lines evaluated were positive for the surface antigens CD105, CD90 and CD73, and for mRNA expression of both OCT4 and NANOG; while they were negative for HLA-DR.
Gene expression levels of TNF alpha, IDO1 and OPN were higher in these cells than in PDMSCs, with higher expression levels in colostrum than in transitional milk.
Only BDNF and NT-4, and not NT-3, were positive in MDMSCs, to a lesser extent than in PDMSCs and with higher concentrations in colostrum.
Gene expression of GLUT-1 and GLUT-4 was found to be reduced in MDMSCs compared with PDMSCs, and GLUT-1 also has higher expression levels in transitional milk while GLUT-4 is more highly expressed in colostrum. Both are found to be reduced in cells isolated from milk of pregnancies complicated by GDM compared with physiological pregnancies.
In conclusion, we have shown that human milk is a source of mesenchymal stromal cells, a unique cell population involved in the processes of immune maturation, neurodevelopment, and neonatal glucose homeostasis. Furthermore, the absence of HLA-DR expression makes these cells optimal candidates for the development of allogeneic cell therapies, without the risk of rejection, and for enrichment of pasteurized donated human milk intended for premature infants in intensive care.

ABS 4

REACTIVE ARTHRITIS AFTER *CLOSTRIDIUM DIFFICILE* GASTROENTERITIS: A CASE REPORT

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BACKGROUND

Reactive arthritis (RA) affects one or more joints and occurs several days up to 3 weeks after an infection (e.g. *Y. enterocolitica*, *Y. pseudotuberculosis*, *C. jejuni/coli*, *Shigella spp.*, *Salmonella spp.*, *K. pneumoniae*, *C. difficile* [CD] and *Cryptosporidium spp.*).
A broad spectrum of special tests is required to detect all etiologic agents. It is therefore pivotal for the laboratory to receive a thorough information on the diagnostic hypothesis. We report a case of RA associated with CD infection.

CASE REPORT

A 4-year-old girl was treated with cefixime for 7 days due to cough. After antibiotic therapy she presented abdominal pain and loose stools with mucus. Afterwards she was admitted to hospital for pain in the lower left limb with refusal to stand, functional impotence of left elbow and pain of the four limbs. After a short improvement with ibuprofen therapy, the patient again presented lower limb pain associated with a rash of large and itchy elements. Vital signs were as follows: skin temperature 36.3°C, HR 108/min, BP 107/70 mmHg, SpO_2 99% in room air. Physical examination: good general condition, rare knobs

in the limbs, slightly edematous and painful to palpation right hand, normal hydrated skin, lymph nodes within normal limits in the latero-cervical, supraclavicular and axillary areas, cardiovascular and chest examination negative. Abdomen was soft and tender, but sore at deep palpation in the right quadrants, bowel movements normal. She presented pain at the passive mobilization of the right wrist and hips, unstable "short" gait due to pain with flexed limbs. Normal skin at the joints. Eye examination was negative for anterior uveitis. Blood tests, autoimmunity panel and antibodies for Toxoplasma, CMV, *Borrelia* and EBV were performed, showing a slight increase of C-reactive protein. A stool film array was tested positive for CD. Due to a potential CD post-infectious RA, vancomycin was started, ibuprofen and PPI gastroprotection were given as well. On the third day of treatment, swelling, gastrointestinal and articular symptoms disappeared.

DISCUSSION AND CONCLUSIONS

RA is an immune disease secondary to infection in a susceptible individual. Gastrointestinal, urogenital and respiratory infections are those which can cause this problem. Its pathophysiology is not yet fully clarified. It has been shown that CD toxin A increases the permeability of the intestinal epithelium and then spreads into the bloodstream, causing a localized immune response at the cell level. CD is a spore-forming anaerobic Gram-positive bacillus and it is found in soil and water, as well as in the human intestine and in many other animals. It can be found in the neonatal intestine as well, who might be an asymptomatic transporter. CD RA is rare. However, the current increase in the CD infection incidence makes post-infectious arthritis more likely. For this reason, more studies are needed and we suggest to look for CD infection in RA in a more systematic way.

ABS 5

THE IMPORTANCE OF PRENATAL DIAGNOSIS IN SMALL CHORANGIOMAS: A SERIES OF CASES

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INTRODUCTION

Chorangioma (CM) is a placental benign non-trophoblastic vascular tumor, whose clinical significance depends on the size. Small CMs (< 3 cm) are often asymptomatic. Intermediate ones (3-5 cm) may be associated with polyhydramnios, fetal hydrops, preterm delivery, fetal growth retardation, perinatal death. Giant CMs (> 5 cm) are associated with various pregnancy complications and an unfavourable perinatal course (recurrent spontaneous abortion, fetal coagulopathies). Multiple foci of CM are particularly worrying. Term or late placentas are the greatest risk factors. Appearing as a well-demarcated placental mass usually on the fetal surface of the placenta, CM is an anastomosed network of fetal capillaries surrounded by pericytes and a single trophoblast layer. Increased cellularity, cytologic atypia, and mitotic figures may be observed, though they should not be related to malignancy. Tumor cells immunostain for CD31, CD34, factor VIII, GLUT1, SMA and CK 18, while Ki-67 should be low. Prenatal diagnosis can be made using colour Doppler ultrasonography and magnetic resonance imaging; the therapy includes alcoholic ablation, endoscopic surgical devascularization, interstitial laser coagulation [1-3].

CASES REPORT

Case A

Suggesting child growth arrest, prenatal examinations also evidenced CM, oligohydramnios, and breech fetus in a pregnant woman at 34 weeks + 5 D with Sjogren's syndrome and Ab anti-Ro positive. At birth the female infant weighed 2,780 g and Apgar was 10/10. On histopathological examination, CM (26 mm of diameter), funiculus' congested umbilical vein and early thrombosis were detected in the placenta.

Case B

Manual secondment was administered in pregnant II at 37 weeks + 5 D with spontaneous labor and unexpected MAC rupture. At birth the male infant weighed 3,010 g and Apgar was 10/10. Placental histopathological examination evidenced a 15 mm of diameter CM (immunophenotype: CD34+; CD31+; SMA+; ERG+/-; WT1 +; Ki67 < 5%).

Case C

IUGR, gestational hypertension, and flowmetric changes were diagnosed in pregnant I at 38 weeks + 2 D. Weighing 2,060 g and showing Apgar 9/10, pH 7.249, and BE 0.4, at birth the female infant was transferred to the Neonatal Pathology Unit. In about 10% of the cut surface the placental

chorionic plate showed multiple whitish areas, which at histopathology were multiple CMs, each not exceeding 10 mm. Stage I grade II chorioamnionitis, infarct areas of various ages, umbilical vessel hemorrhaged were also seen.

CONCLUSIONS

Prenatal diagnosis of CM is essential since this entity may cause some concerns even when the tumor is small as it was showed in our cases, all of which harbored severe complications while the size was less than the usual worrisome cutoff of 3 cm.

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ABS 6

APPLICABILITY OF POLYSOMNOGRAPHY IN PEDIATRIC PRACTICE OF SLEEP-RELATED BREATHING DISORDERS IN CHILDREN

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OBJECTIVES

Evaluation of the incidence and characteristics of sleep-related breathing disorders (SRBD) in pediatric patients referred to the Somnology Department within the Pediatric Clinic of the Children's Clinical Hospital "Dr. Victor Gomoiu" during the period March 1, 2019 - March 1, 2021.

EVALUATION METHODS

A number of 90 patients were monitored through a prospective study over a period of 2 years. They were evaluated clinically, paraclinically and the pediatric sleep questionnaire was applied. A polysomnographic sleep study was performed using the Alice6 LDxS device, the SleepwareG3 interpretation program, and the AASM VIII4.B scoring criteria.

RESULTS

Out of a total of 90 patients aged between 1-18 years evaluated for SRBD, 30% presented rhinopathies. Varying degrees of SRBD were detected in 63% of patients, the pathology being more common among males (72%) than females (47%). Moderate-severe forms of RDS are present in 31% of the evaluated children.

All patients showed positive correlation between the clinical symptomatology and the results provided by the analysis of the sleep questionnaire.

A decrease in children's quality of life, quantified by applying the pediatric sleep questionnaire and validated by a polysomnographic study, was reported by 58% of the relatives.

Patients presented with otorhinolaryngology pathology associated SRBD in 75% of cases.

Among patients with neuromuscular diseases, 72.2% presented varying degrees of apnea during the course of the disease. SRBD are present in 83% of children with Prader-Willi syndrome.

Obesity as a risk factor was found in 23% of patients and more than half of them presented apnea in mild, medium or severe form.

CONCLUSIONS

Polysomnography is the gold-standard investigation for diagnosing SRBD in children. These conditions are important due to the consequences they have on the neuropsychic development, physiology and proper functioning of the body with a major impact on the quality of life.

ABS 7

PREGNANCY RISK FACTORS AND PERINATAL OUTCOMES IN IMMIGRANTS, REFUGEES AND GREEK (NATIVE) POPULATION, BEFORE AND DURING THE SARS-CoV-2 PANDEMIC. A SINGLE-CENTRE EXPERIENCE

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OBJECTIVE
The SARS-CoV-2 pandemic seems be an independent risk factor in health care to every population, native or not, in many countries. The aim of our study was to investigate the risk factors during pregnancy and the perinatal outcomes of the immigrant (I), refugee (R) and native (Greek, G) populations before and during the SARS-CoV-2 pandemic.
MATERIAL AND METHODS
An observational retrospective epidemiological study was conducted regarding all neonates born in the Papageorgiou General Hospital between 2019 and 2020. Demographic data and data regarding pregnancy and the perinatal period were recorded from secondary databases of our Hospital.
RESULTS
Our study sample consisted of 2,356 women and their 2,417 newborns. The SARS-CoV-2 pandemic led to higher rates of pregnant women without insurance (R: 95% vs 13.2%, $p < 0.001$; G: 13.1% vs 8.2%, $p = 0.014$), higher rates of maternal hypertension (I: 9.2% vs 3%, $p = 0.029$; G: 6.5% vs 4, $p = 0.049$), lower rates of follow-up during pregnancy (R: 67.3% vs 53.2, $p = 0.001$; G: 5.3% vs 2.7%, $p = 0.008$), higher rates of incomplete follow-up during pregnancy (R: 28.8% vs 17.1, $p = 0.001$; G: 7% vs 4.9%, $p = 0.008$) and caesarean section (I: 48.2% vs 35.2%, $p = 0.038$; G: 55.3% vs 51.7%, $p = 0.004$), as well as to newborns with lower birth weight (G: 3,130 [2,650-3,405] grams vs 3,165 [2,830-3,485] grams, $p = 0.004$) in comparison to the era before the SARS-CoV-2 pandemic.
CONCLUSIONS
This study indicates that the COVID-19 pandemic had a significant impact on perinatal outcomes among pregnant women and their newborns.
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ABS 8

A MONOGENIC CAUSE OF CHRONIC KIDNEY DISEASE WITH MULTIORGAN INVOLVEMENT: CAKUTHED SYNDROME

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INTRODUCTION
Congenital anomalies of the kidney and urinary tract (CAKUT) are among the most frequent congenital malformations in newborns, involving approximately 0.5-1% of live births. CAKUT can be isolated or part of complex syndromes. Despite the high incidence, CAKUTs represent a diagnostic challenge, given that genetic etiological diagnosis is often inconclusive. Only about 20% of CAKUTs have a monogenic basis. Multifactorial genetic diagnosis with no evidence of disease-causing genes are more common [1].
The spectrum of phenotypes is very broad, from asymptomatic forms to the letal ones. Mild forms of CAKUT may be diagnosed by occasional ultrasound, whereas the severe ones may begin with recurrent urinary tract infections or perinatal renal failure, until chronic kidney disease [2].
Despite the different clinical features, most CAKUTs share an embryonic mal-development of nephro-uro-genic tissues and the etiopathogenetic genes are genes for signaling molecules (*ROBO2*), components of extracellular matrix (*FRAS1*, *FREM2*) or transcription factors such as *PAX2* and *PBX1*, whose haploinsufficiency is responsible for renal and collecting system malformations and also extrarenal manifestations [3].
CASE DESCRIPTION
A newborn was admitted to the Neonatal Pathology Unit for respiratory distress in post-term birth with oligohydramnios and prenatal diagnosis of bilateral renal hypoplasia. In the first days of life there was a marked contraction of diuresis, with creatinine levels of 2 mg/dL at 2 days of life and 1.2 mg/dL at 15 days, with progressive normalization of the urine output. Renal ultrasound at 10 days of life confirmed the diagnosis of bilateral renal hypodisplasia.
At 4 months of life he performed the MAG3 sequential renal scintigraphy. total tubular extraction rate (12 mL/min) was below the expected range for age and BSA, with hypofunctional left kidney.
Since the discharge from hospital, the patient performed a close follow-up, which has shown a progressive deterioration of renal function.

Actually, at the age of 14 he presents a peculiar phaces, a mild learning disorder, and a GFR of 35 mL/min/1.73 m^2 creatinine-cystatin C-based CKiD equation.

The genetic etiology of this malformative and multiorgan phenotype was investigated. The SNP-Array showed in heterozygosity a microdeletion of 622kb in the 1q.23 region, including gene *PBX1*, which is mandatory for regular nephrogenesis and brain development; the haploinsufficiency has been associated with CAKUTHED syndrome.

DISCUSSION AND CONCLUSIONS

The clinical phenotype of CAKUTHED includes neurodevelopmental diseases and hearing diseases; therefore, he underwent an auditory function survey, which revealed a bilateral sensorineural hearing loss on acute and very acute frequencies.

In conclusion, genetic diagnosis of CAKUTHED leads to explain and complete a complex clinical case. CAKUT should be suspected and investigated in cases of kidney malformations, especially if associated with other clinical problems.

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ABS 9

METABOLOMICS PROFILE OF A PEDIATRIC POPULATION SUFFERING FROM PFAPA AND UNDER TREATMENT WITH PIDOTIMOD: A PRELIMINARY, MULTICENTER, OPEN-CLINICAL TRIAL

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BACKGROUND

The periodic fever, aphthous stomatitis, pharyngitis, adenopathy (PFAPA) syndrome is an autoinflammatory periodic disease featured by recurrent febrile episodes and at least one of three following symptoms, including pharyngitis, aphthous stomatitis, and cervical adenitis. To date, there is not a specific treatment modifying the clinical course and outcomes; and only the systemic steroids appear able to abrupt the PFAPA episodes [1]. Recently, in a pilot, prospective, controlled, open, cross-over study trial, we reported that the administration of pidotimod, a synthetic dipeptide molecule, resulted in a significant decrease in the frequency of PFAPA episodes by reducing febrile episodes, episodes of pharyngitis and aphthous stomatitis [2, 3]. Moreover, a significant decrease in rescue medication use, betamethasone, was also recorded. Interestingly, the administration of pidotimod was not associated with severe adverse events in the enrolled patients. However, no data were reported on how pidotimod can induce the above mentioned significant clinical changes [2]. Aiming to fill this gap, we started a preliminary, multicenter, open-clinical trial to investigate the pidotimod-mediated changes in the metabolomics profile of a pediatric population suffering from PFAPA [4, 5].

MATERIALS AND METHODS

Pediatric patients of both genders, aged 3-8 years, with a diagnosis of PFAPA were enrolled in the clinical trial. Each patient was receiving pidotimod for up to 1 month. All patients underwent to anamnestic data collection and clinical examination. A urine sample (10 mL) of each patient has been collected at the baseline, at 15, and 30 days from the start of the treatment. Before analysis, samples were centrifuged for 10 min at 4°C at 12,000 × g to remove solid particles. Then, 630 µL of the supernatant were mixed with 70 µL of potassium phosphate buffer in D2O (1.5 M, pH 7.4) containing sodium 3-trimethylsilyl-propionate-2,2,3,3,-d4 (TSP) as an internal standard (98 atom% D, Sigma-Aldrich, Milan, Italy). Finally, 650 µL were transferred to 5 mm NMR glass tubes for ^{1}H-NMR analysis. ^{1}H-NMR analysis was carried out using a Varian UNITY INOVA 500 spectrometer operating at 499.839 MHz for proton and equipped with a 5 mm double resonance probe (Agilent Technologies, Santa Clara, CA, USA). One-dimensional proton

NMR spectra were obtained by using a 1D Nuclear Overhauser Enhancement Spectroscopy (NOESY) standard pulse sequence to suppress water signals with a relaxation delay of 3 s. For each sample, 256 free induction decays (FIDs) were collected into 64K data points with a spectral width of 6,000 Hz spectral with a 90° pulse, an acquisition time of 2 s, and a mixing time of 100 ms. The FIDs were weighted by an exponential function with a 0.5 Hz line-broadening factor prior to Fourier transformation. MR spectra were phased and baseline corrected using an Advanced Chemistry Development (ACD) Lab (Toronto, ON, Canada) Processor Academic Edition (Advanced Chemistry Development, 1 December 2010) and chemical shifts referenced internally to trisodium phosphate (TSP) at $\delta = 0.0$ ppm. The spectral region comprising the signal of residual water and urea (4.7-6.5 ppm) was removed. The final spectral regions were between 0.5-4.7 ppm and 6.5-9.5 ppm. The ACD Labs intelligent bucketing method was used for spectral alignments.

RESULTS AND CONCLUSIONS

Based on the alignment and a visual comparison of the pathological spectra, it is possible to highlight a "trend of metabolic modification" starting from T0 up to T28 (time points).

To conclude, our next step, for this very preliminary analysis, will be to identify the metabolites that characterize each time point, which may help to uncover specific urinary metabolites associated either with PFAPA or with the pharmacological treatment.

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ABS 10

CELIAC DISEASE IN CHILDREN: DIAGNOSTIC DELAY AND ITS CAUSES. A RETROSPECTIVE ANALYSIS

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Celiac disease (CD) is a chronic immune-mediated enteropathy of the small gut. Exposure to gluten triggers and sustains the pathogenetic pathways in genetically predisposed subjects, and a gluten-free diet allows for the regression of the intestinal damage. With a prevalence of roughly 1% worldwide, CD represents one of the most common autoimmune disorders, and is now considered a "social disease".

In order to evaluate the diagnostic delay of this condition as well as the effect of the COVID-19 pandemic on this issue, we performed a retrospective analysis on 225 patients, divided into two groups depending on whether the diagnosis was made before (2010-2019) or after (2020-2022) the beginning of the pandemic. The diagnostic delay has been analyzed through three different forms: "patient-dependent" (the time from symptoms onset to the first medical consultation), "doctor-dependent" (the time from the first consultation to the definitive diagnosis), and "global" (from symptoms onset to the definitive diagnosis). "Extreme delay" has been defined as a diagnostic delay above the 75th percentile (p75). To describe the continuous data, median and interquartile ranges (IQR) have been adopted. The correlation between parameters and delay has been explored through uni- and multi-variate analysis.

Gastrointestinal (GI) symptoms represented the most common clinical manifestation, followed by iron deficiency anemia, cephalea, and other neuropsychiatric symptoms including specific learning disorders. Regarding the genetical, serological and histological features of the cohort, DQ2 heterodimer, anti-transglutaminase immunoglobulins A (IgA) levels 10-fold higher than the baseline (allowing for CD diagnosis without additional invasive procedures) and villous atrophy have been found in 86.6%, 55% and 80.9% of the patient cohort, respectively. The

average age at diagnosis resulted to be 7 years. The median "global", "patient-dependent" and "doctor-dependent" delay resulted to be of 3 (IQR 1-7), 2 (IQR 0-6), and 1 (IQR 0-2) months, respectively. No significant differences between pre- and post-pandemic time have been found. Among the parameters that influence negatively the "global" diagnostic delay we identified GI symptoms at presentation and growth retardation, while familiarity for CD and autoimmune comorbidity represented protective factors, diminishing all the types of delay examined and "patient-dependent" delay, respectively. Moreover, dermatitis herpetiformis and recent GI infection have been found to increase the "doctor-dependent" delay. Finally, regarding the "extreme" delay, growth retardation resulted to have a negative impact on both "global" and "patient-dependent" delays.

Overall, the study showed that in our cohort COVID-19 did not impact on CD diagnostic delay. Moreover, GI symptoms and growth retardation should always rise the suspect of CD and CD screening test should be performed in children presenting with these clinical manifestations.

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ABS 11

ASSOCIATION BETWEEN UMBILICAL CORD ANGIOMA AND MATERNAL VASCULAR MALPERFUSION (MVM): A CASE REPORT

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INTRODUCTION

Umbilical cord angioma is a very rare benign tumor of endothelial cells often associated with fetal malformations, intrauterine fetal death, vascular compression, and fetal hemorrhage by vascular rupture. Prenatal imaging is not always able to reveal its presence, therefore most morphological lesions are described by the pathologist only after fetal death.

DESCRIPTION OF THE CASE

Not feeling fetal movements for about 24 hours, a pregnant 35-year-old woman at 29 w + 1 d went to the hospital. Ultrasonography (US), performed urgently, revealed fetal death. While at 21 w + 5 d the US had showed an anechoic cyst with a diameter of 15 mm, the vital parameters remained normal until the 28 w + 5 d. The post-mortem examination highlighted normal-shape male fetus weighing 1,030 g, length V-C 30 cm, C-C 30 cm, and features consistent with gestational age. The 19-cm-long umbilical cord measured 7 mm in caliber on the fetal side, 4 cm in the central portion, and 1.5 cm in the distal portion. Devoid of spiralization, the dilated area covered a length of 16 cm. The cut surface showed a cystic dilation of 2 cm associated with a large hemorrhagic area. The viscera appeared regular and *in situ* except for the presence of dolichol-mega-sigma. The ducts of Arantius and Botallo were pervious. In the placenta, thickened and opaque membranes with focal whitish areas were highlighted. The maternal side was greyish in color and showed and prominent cotyledons, though a brownish marginal area was detected. The histological examination showed the proliferation of capillary-like small vessels lined by a single layer of flat endothelial cells inside Wharton's jelly. In addition, we found the presence of neoplastic vascular channels arranged around the largest vessels. Dilation and congestion of the umbilical vessels and focal thrombosis were present. Immunohistochemistry confirmed the vascular origin of the neoformation with immunostaning positivity to SMA, CD31, and CD34. Consequently, the diagnosis was umbilical cord angioma and maternal vascular malperfusion (MVM).

DISCUSSION AND CONCLUSIONS

The rare association between cord angioma and MVM here reported raises some concern about the causality between the two phenomena. In other words, was it possible that the umbilical cord angioma has caused the MVM? The contribution of the morphological diagnosis in cases of rare vascular tumors, which are difficult to diagnose in the prenatal age, is essential for understanding the pathogenesis and the progression. Further reports will be valuable in assessing whether the two concomitant alterations may eventually increase fetal mortality. Thus, the accurate pathological

examination of the fetal annexes, in particular the placenta, is mandatory to deeper understand the US findings.

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ABS 12

NEPHROTIC SYNDROME IN ASSOCIATION WITH VARICELLA IN A 9-YEAR-OLD BOY: A RARE CASE REPORT

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INTRODUCTION

Varicella may present with severe clinical manifestations also in immunocompetent children. Among its complications, nephrotic syndrome is extremely rare, with only 3 pediatric cases reported in the literature.

CASE REPORT

We describe the case of a previously healthy 9-year-old boy who presented with facial and peripheral edema and increased body weight, 5 days after the beginning of varicella exanthema. Laboratory work-up for nephrotic syndrome showed massive proteinuria (22.9 g/24 hours), microhematuria, hypoalbuminemia (serum albumin, 1.50 g/dL) and dyslipidemia (serum cholesterol, 385 mg/dL; triglycerides, 280 mg/dL), with normal renal function (serum creatinine, 0.67 mg/dL). These findings supported the diagnosis of varicella-related nephrotic syndrome. Serological testing confirmed a recent varicella-zoster infection. Corticosteroids are the first-line treatment of nephrotic syndrome, but this therapy was not performed in this case because the infection was in the acute phase. Instead, therapy with intravenous acyclovir was carried out for 5 days to inhibit viral replication. The response to therapy was rapid, leading to clinical improvement, progressive reduction of body weight with disappearance of edema, and normalization of urinalysis. On day 12, the patient was discharged home in good clinical conditions, with a strict follow-up schedule.

DISCUSSION AND CONCLUSIONS

Renal complications of varicella are very rarely reported in the literature, generally in association with disseminated forms of infection. Varicella-related nephrotic syndrome is exceptionally rare, especially in childhood, and, to our knowledge, only 3 pediatric cases have been reported. Similarly, to the patient described by Lin et al., the nephrotic syndrome described by us characteristically occurred during the acute phase of varicella in a previously healthy patient. Moreover, proteinuria of such a high magnitude (22.9 g/24 hours) has not been reported previously to the best of our knowledge.

Considering the favourable course and outcome of our patient, the diagnosis was exclusively based on clinical and laboratory examinations, while renal biopsy was not deemed necessary.

Both the early onset of nephrotic syndrome during varicella infection and its resolution concurrent with the disappearance of exanthema, as well as the response to antiviral treatment, suggest that, in our patient, glomerular injury was a direct result of viral invasion of the renal parenchyma.

In conclusion, the present case highlights that varicella, although generally considered a self-limiting disease, may be rarely associated with renal complications also in immunocompetent patients.

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ABS 13

NEONATE WITH CORNELIA DE LANGE SYNDROME. A CASE REPORT

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INTRODUCTION

Cornelia de Lange syndrome is a congenital anomaly syndrome characterized by distinctive facial dysmorphism, primordial short stature, hirsutism, and upper limb reduction defects that range from subtle phalangeal abnormalities to oligodactyly. It is an autosomal dominant disorder caused by specific gene mutations and occurrence is one in 30,000 to 50,000 children. Various gene mutations, such as *NIPBL* gene, *SMC1A* gene and *SMC3* gene, have been described until now. However, spontaneous mutations are responsible in the majority of the patients.

The case of a late preterm infant with typical dysmorphic characteristics is reported below.

CASE REPORT

A male late preterm neonate with gestational age 35^{+5} weeks has been delivered with cesarean section due to intrauterine growth restriction, with a birth weight of 1,600 g. A hypoplasia of the upper limbs could be detected at the prenatal visit of the second trimester. The neonate showed a normal cardiovascular transition at birth.

Physical examination findings:

1. upper-limb reduction defects;
2. syndactylism between the third and fourth finger on the left foot;
3. tetra-dactylism on the right foot;
4. undescended testicles;
5. short nose;
6. domed hard palate;
7. ear helix abnormalities;

Examinations:

- normal biochemical and haematological blood examinations as well as control for congenital infections (TORCH);
- normal echocardiography;
- normal cranial, abdominal, and lumbosacral spine ultrasound;
- testicles ultrasound revealed both testicles in the inguinal canal;
- neurological-orthopedic examination;
- normal cranial MRI;
- normal opthalmological examination and otoacoustic emissions;
- normal control for hereditary metabolic diseases.

Clinical exome sequencing is presented in **Tab. 1**. This mutation has not yet been reported in the scientific data base and has not yet been described in any international reference. However, this mutation causes such a gene change, that the structure and function of the produced protein may be affected. Mutations of this type have been observed in patients with Cornelia de Lange syndrome.

A genetical examination of both parents revealed a *de novo* mutation of the infant.

DISCUSSION

Cornelia de Lange syndrome is a multisystem disorder with physical, cognitive and behavioural characteristics. A genetical examination in order to search for gene mutations is necessary. Panel sequencing is the most effective way of detecting causative variants in any of the genes known to cause Cornelia de Lange syndrome, and first-line molecular testing should use a panel that contains at least the 7 known genes. It is a complex disorder and it is important that a lead clinician is identified for each patient to ensure coordination of the numerous aspects of care in both childhood and adulthood.

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Table 1 (ABS 13). Clinical exome sequencing.

Gene	Nucleotide change	Amine oxide change	Result	Zygote	Clinical significance
NIPBL	c.3117_3120delTAAA	p.Asn1039LysfsTer4	Frameshift deletion	Heterozygote	Pathogenic

ABS 14

THE EVALUATION OF DIGITAL HEALTH: PEDIATRIC ASSESSMENT FOR EXPOSITION TO DIGITAL MEDIA

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In recent decades, digital technologies have become an integral part of our daily life, indispensable tools for work, school, communication and maintaining

relationships. Conscious use has offered important opportunities for growth, allowing us to have free access to information, stay updated on what is happening in the world, increase knowledge and stay in contact with others, regardless of where they are.

However, the transition from healthy and balanced use to risky behavior is short. Always more often children, preteens and adolescents have been prematurely exposed to a world of digital content not always appropriate for their age, to risks and dangers such as solicitation, cyberbullying and revenge porn.

Recent studies have shown the possible negative effects of early and prolonged exposure to digital content with particular reference to screen time. In fact, excessive use of Digital Media (DM) can have important repercussions on psycho-physical development and health of the little ones, in particular on sleep, sight, hearing, posture, metabolic functions and on parent-child relationships. In addition, Internet-connected devices represent the key to accessing digital subculture, the content of which can have a negative impact on children's psychological and physical development. Given the impact of DM and in particular of "digital subculture" on children, pre-adolescents and adolescents, we propose to broaden the concept of health by including not only the biological, psychological and social dimensions but also the digital dimension and thus the relationship with digital devices.

In this work, we introduce the concept of Digital Health (DH), which refers to the well-being of all individuals, in particular of subjects of developmental age exposed to digital devices, and we provide pediatricians with a new health procedure, called Evaluation of Digital Health (EDH). The EDH, aimed at assessing the digital habits, screen time and digital content viewed by the child, is carried out during periodic check-ups. In fact, we believe that pediatricians have a fundamental role in the protection of all-round health, including DH. The EDH would enable pediatricians, who follow growth from birth to adolescence, to prevent and/or promptly pick up on the signs of any risky digital behavior displayed by parents and their children. From this perspective, the pediatrician may lead to the manifestation of appropriate digital behaviors, thus representing the first DH promotion service and fostering the development of digital awareness in the family.

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ABS 15

PULMONARY CAPILLARY HEMANGIOMATOSIS: CASE REPORT IN NEWBORN

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INTRODUCTION

Pulmonary capillary hemangiomatosis (PCH) is a rare disease, about which little is known so far. Described for the first time by Wagenvoort et al. in 1978, PCH is an atypical proliferation of capillaries, harboring cytologically benign endothelial cells, that can infiltrate even the bronchi, the lobular septa, and the walls of pulmonary vessels. Rarely, capillaries proliferate into the pericardium, the pleura, and the mediastinal lymph nodes.

The average age of presentation is between 20 and 40 years with no gender prevalence. Pulmonary hypertension, hemoptysis, and dyspnea are the main clinical presentation. The outcome is often adverse since patient survival is estimated between 1 and 5 years. Although the congenital presentation of PCH has been even more rarely described in the scientific literature, here we present the case of an infant, who, having died a few hours after birth, showed the characteristic histological picture of PCH.

CASE REPORT

A male infant born through emergency caesarean section at 37^{+4} weeks from a mother who carried

a poorly controlled insulin-dependent gestational diabetes. The newborn's Apgar index was poor (1-0-1), so CPR was performed for 24 minutes. Since the newborn presented 130 bpm HR, he was intubated and admitted at the Neonatal Intensive Care Unit (NICU), where chest-abdomen X-ray revealed an enlargement of the cardio mediastinal area. Over time, the clinical condition progressively deteriorated, and death occurred about 3 hours after NICU admission.

The post-mortem examination showed normo-shaped visceral organs according to gender and gestational age. Being hypo-inflated and dark reddish in color, the lungs appeared shrunken in the chest cavity because of the enlargement of the mediastinal-pericardial area. Globular in shape, especially in the left side, the heart had an increased size, while the myometrium was firm, and the atrioventricular valves were dilated, particularly in the tricuspid site.

The histologic examination revealed congestion, dilatation of pericardial vessels, and foci of intramyocardial capillaries. Still in saccular phase, the lungs showed diffuse congestion and dilatation of septal and lobular vessels. Furthermore, a huge amount of septal nodular capillary vessels protruded into the saccular spaces, matching the features of PCH.

DISCUSSION

Being a rare cause of progressive lung disease, PCH is detected as a nodular proliferation of small capillary vessels. Since the diagnosis can be difficult, the histologic examination is generally required. Indeed, given the differences in treatment, PCH should be distinguished from primary pulmonary hypertension. Nevertheless, PCH should also be included in the differential diagnosis of perinatal autopsies, particularly those characterized by cardiomegaly, right ventricular hypertrophy, and/or endocardial fibrosis, and in newborn or infants with neonatal pulmonary pathology, pulmonary hypertension, cardiomegaly, and/or biventricular hypertrophy.

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ABS 16

UNPLANNED OUT-OF-HOSPITAL BIRTHS: AN UNUSUAL CLUSTER IN ROVIGO HOSPITAL – ITALY

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BACKGROUND

Unplanned out-of-hospital birth (UOHB) counts for 0.19% to 0.60% of all deliveries, with increased mortality and morbidity rate. We present 4 cases of UOHB, during a 2-month period (April-May 2022) at Rovigo Hospital, which counts about 650 deliveries yearly.

CASE REPORTS

Patient no. 1

Patient no. 1, female, born at the ER, unknown gestational age (GA). Vital signs (VS) at birth: SpO_2 80%, HR > 150 bpm, RR 50/min. Body weight (BW) 2,840 g, skin temperature (T) 36.6°C. Apgar score 8 (1'), 9 (5'). Pediatrician and midwives were called at ER. Soon after birth she was admitted to Neonatal Care Unit (NCU), and blood gas analysis (BGA) was performed: pH 7.27; pCO_2 48 mmHg; pO_2 46 mmHg; HCO_3 22 mmol/L; glucose 46 mg/dL. O_2 (0.4-1 LPM by nasal cannula) was given for 3 hours. At 4 hours of life she was transferred to rooming-in, and discharged 3 days later.

Patient no. 2

Patient no. 2, male, born in the ambulance, GA 39 weeks. BW 2,900 g, Apgar score 9 (1'), 10 (5'). Initial hypothermia in the ambulance. He was admitted to our NCU with skin T 36.7°C. O_2 was given by nasal cannula (0.2-0.5 LPM). BGA: pH 7.38; pCO_2 44 mmHg; pO_2 49 mmHg; HCO_3 26 mmol/L; glucose 65 mg/dL. After 3 days he was discharged.

Patient no. 3

Patient no. 3, female, born in a car at GA 33 weeks. 118 (i.e., the medical emergency number in Italy) was called and mother and baby transported to our NCU. VS were within normal limits, but she presented severe hypothermia (T 33.2°C). Apgar score 9 (1'), 9 (5'), BW 1,480 g. BGA: pH 7.40;

pCO_2 27 mmHg; pO_2 101 mmHg; HCO_3 16.7 mmol/L; glucose 60 mg/dL. Due to hypothermia and low BW she was put in a preterm infant incubator with an initial set T of 37.5°C. Due to persistent hypothermia, hot water bags were put inside. No O_2 was needed. ECG, CXR, abdomen XR and lab tests were normal. She was discharged at 30 days of life, in good clinical conditions.

Patient no. 4

Patient no. 4, male, born at home, GA 38 weeks. 118 was called. Mother and newborn were transported to the NCU. Body T and Apgar score data not available. O_2 has been given due to SpO_2 > 94% for 72 hours. BW 2,500 g. Skin T 36.6°C, RR 48/min. BGA: pH 7.38; pCO_2 42 mmHg; pO_2 47 mmHg; HCO_3 24 mmol/L; glucose 68 mg/dL. After 5 days he was discharged at home.

DISCUSSION AND CONCLUSIONS

Management of UOHB mainly requires prompt temperature and infection control, and effective call for help. 118 needs to be called whenever birth takes place in an ambulance, at home, or in other places outside the hospital. Anaesthesiologist, pediatrician/neonatologist (if NCU available), midwife and gynecologist need to be called whenever birth takes place in a hospital. Whenever birth takes place outside the delivery room, hypothermia is very frequent. Therefore, drying the newborn, covering him with warm sheets, closing windows and using the incubator as soon as possible is very important. A glove filled with warm water and the bare chest of the mother are good initial transient solutions to keep the newborn warm. Importantly is to declare birth time and place in case of UOHB.

ABS 17

INFANT WITH RETROPHARYNGEAL HEMANGIOMA. SUCCESSFUL TREATMENT WITH PROPRANOLOL. A CASE REPORT

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INTRODUCTION

Infantile hemangioma is the most common tumor of infancy. In contrast to vascular malformation, infantile hemangioma is usually absent or inconspicuous at birth. Differential diagnosis with lymphatic malformations should be performed. These are congenital vascular anomalies that result from abnormal development of lymphatic vessels, which present in the head and neck in 48% to 75% of cases. Involvement of the upper airway is rare.

We report one case of symptomatic retropharyngeal tumor that was successfully managed with propranolol.

CASE REPORT

A female infant with gestational age 38^{+6} weeks was delivered with C-section due to disproportion. An orotracheal intubation was necessary 10 minutes after delivery due to acute respiratory distress. The laryngoscopy revealed a retropharyngeal mass with obstruction of the upper airways.

Further diagnosis was based on radiological exam, specifically a contrast-enhanced MRI, which revealed a retropharyngeal tumor, similar to a retropharyngeal hemangioma. The sonography of the pharynx and larynx revealed also a retropharyngeal tumor with increased perfusion. Our patient was assessed by a senior otolaryngologist under orotracheal intubation.

Based on clinical history, clinical examination and radiological exam, a retropharyngeal hemangioma was suspected. On the second day, an oral treatment with propranolol in a dose of 1 mg/kg/day was started and could be escalated until 2 mg/kg/day. No severe complications were noted. A second contrast-enhanced MRI was performed 15 days later and showed a massive decrease of the tumor. Our patient could be successfully extubated on the same day.

No other hemangiomas could be detected neither in the abdominal ultrasonography, nor in the cranial one. Our patient could be discharged at a good clinical condition on the 25th day.

DISCUSSION

Infantile hemangiomas are usually small lesions and can involute spontaneously. Differential diagnoses mostly consist of other vascular tumors and vascular malformations, although infantile hemangiomas may sometimes mimic nonvascular tumors or developmental anomalies.

The majority of infantile hemangiomas can be managed conservatively, but for those requiring active treatment, management has been revolutionized in the last decade by the discovery of propranolol. There are many other several well-established treatments for infantile hemangiomas, such as corticosteroids (either intralesional or

systemic corticosteroids), interferon-α, laser therapy, cryotherapy, and surgical excision.
Our patient could be successfully treated with propranolol in a dose of 2 mg/kg/day as a great remission of the hemangioma has occured. Propranolol is a safe and effective first-line therapy for problematic infantile hemangiomas. Therapy should show significant response by 2 weeks. If no response is observed by 3 weeks, then other treatment should be sought.

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ABS 18

MATERNAL BMI AND FETAL PROGRAMMING: PLACENTAL BIOMARKERS AND SHORT-TERM NEONATAL OUTCOMES

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BACKGROUND

The high prevalence of overweight and obesity in pregnant women is a matter of concern for the health of current and future generations: in fact, the state of obesity not only affects the health of the woman herself, but also increases the risk of perinatal and postnatal morbidity in the offspring, to which is added the risk of developing long-term consequences, thus creating a cross-generational vicious circle. The study of placental biomarkers is extremely interesting, as some of them are associated with several pathological conditions in pregnancy, including maternal obesity, and may contribute to maternal and neonatal risk.

AIMS

To evaluate placental tissue in both normal-weight and overweight/obese patients, assessing endothelial and inflammatory differences; to compare short-term perinatal and neonatal outcomes between normal-weight and overweight/obese groups; to assess for correlations between these possibly epigenetic modifications and clinical outcomes.

METHODS

Normal-weight was defined as a BMI 18.5-24.6, while overweight and obese was defined as a BMI ≥ 25. 35 normal-weight patients, 27 overweight/obese patients and their neonates were recruited. None of the recruited patients had additional comorbidity. For each patient, random biopsies were taken from the placental basal plate. Immunoregulatory modulators (INF-γ, OPN, IDO1) and angiogenesis modulators (VEGF, PIGF, Sflt-1) were evaluated using real-time PCR. Obstetric and neonatal outcomes were also studied, and lastly compared with the expression of placental biomarkers.

RESULTS

Obstetric outcomes showed no significant differences between the 2 groups. Regarding the neonatal data, no significant differences were found in terms of gestational age, neonatal weight, length and head circumference, LGA frequency. Likewise, there were no significant differences in short-term neonatal outcomes, except for mild breastfeeding difficulties. On the other hand, a dysregulation of placental markers was demonstrated in the overweight/obese pregnant group compared to normal weight controls. These alterations, probably of epigenetic origin, could affect the baby's health in the long term despite a "physiological" neonatal condition.

CONCLUSIONS

Taken together, our data relating to the course of pregnancy, labor, delivery, and neonatal course are justifiable by the fact that we are dealing with a group of patients selected from the beginning, with no initial comorbidity. Furthermore, the outcomes reflect optimal obstetric and neonatal management. Therefore, these markers should also be analyzed in the future in the child, thus monitoring the possible association with any health problems during development. Furthermore, the anomalies of epigenetic origin have by their nature a characteristic of potential reversibility and are susceptible to lifestyles changes and diet modifications, cornerstones on which to focus health interventions in children.

ABS 19

SAFFRON IN ADOLESCENT MOOD DISORDERS

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Saffron is an herbal medicine extracted from whole plant that has efficacy for the treatment of mild to moderate depression. Among its active principia, safranal inhibits serotonin reuptake and crocin has neuroprotective abilities, increasing intracellular levels of neurotrophic factors such as brain-derived neurotrophic factor (BDNF) in *in-vitro* and animal models. There are numerous studies confirming its effect both alone and also in combination with mood-modulating drugs [1].
Even with the limitations in subjective detection of symptoms, the use of a 28 mg/day dose of saffron as an alternative to pharmacological treatments (with a significant difference between 28 mg/day and placebo, $p < 0.001$), has been shown to significantly improve mood and symptoms related to stress and anxiety, and improved sleep conditions assessed by the Pittsburgh Sleep Quality Index (PSQI) [2].
In this regard, because phytotherapeutics are often proposed to treat sleep disturbances both during the early falling asleep phase and during the night, regular use of saffron for periods of at least 28 days was seen to be associated with improvements in sleep quality in adults with self-reported sleep disorders [3].
Disorders of psychiatric origin such as anxiety and depression affect not only adulthood, but increasingly the adolescent period as well. According to data provided by the World Health Organization, these disorders currently are among the leading causes of disability in young people. The SARS-CoV-2 pandemic, with lockdown, unchecked use of the Internet and social media, COVID-19 disease-related symptoms, and fear of the disease for self and family members, has worsened the state of mood distress in adolescents. The effect of a saffron extract was evaluated for 8 weeks in a randomized, double-blind, placebo-controlled trial in young people aged 12 to 16 years with mild to moderate anxiety or depressive symptoms [4]. Based on the youth's self-assessment questionnaires, Revised Child Anxiety and Depression Scale (RCADS), the group taking saffron at a dose of 14 mg twice daily had significant improvements in symptoms of separation anxiety ($p = 0.003$), social phobia ($p = 0.023$) and depression ($p = 0.016$). The herbal drug was well tolerated and improved anxiety and depressive symptoms in adolescents with mild to moderate symptoms, according to the adolescent's assessment, and less consistently according to the parents' assessments as well.

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ABS 20

CAESAREAN SECTION: HISTORICAL AND ETYMOLOGICAL ASPECTS

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INTRODUCTION

The term "caesarean section" dates back to Plinius the Elder (23-79 A.D.) who, in the book VII of *Naturalis Historia*, affirmes that breech presentation birth is a bad omen and writes: "*Primusque Caesarum a caeso matris utero dictus, qua de causa et caesones appellati*". However, it is to be excluded that Julius Caesar was born by hysterotomy, given the fact that his mother was still alive at the time of the Gallic war and that at that time it was practically impossible to perform a caesarean section on an alive woman. On the other hand, it could be performed for religious reasons, according to the *Lex Regia*, on a dead woman.

METHODS

An extensive and accurate bibliographic research has been carried out on the topic.

DISCUSSION

Plinius' opinion achieved consensus and over time it was widely assumed that the name Caesar derived from *caesus* in reference to the hysterotomy of his birth. For example, we refer to J. Melber (1482): "*Caesar kaiser, sic dictus, quod ex ventre matris caesus*" (*Vocabularius praedicantium* D 6a), and the first obstetrics printed book by Eucharius Rosslin (*Der swangern frawen und hebammen roszgarten*, Strasbourg 1513): "Thus we read in Roman History that the first emperor called Giulius was extracted with a cut from the mother womb. For this reason, he was called Caesar, which means cut out of the maternal womb". S. Mercuri (1596), T. Raynaud (1637), C. Volter (1680), A. Levret (1770), and C. Stark (1801) all confirmed this history. But it is Francois Rousset (1538-1603) the one to whom we owe the name commonly used today to indicate hysterotomy, even if it seems that he never performed this type of intervention. In the *Traittè nouveau de l'Hysterotomotokie ou Enfantement Caesarien* (Paris 1581), the first monograph on caesarean section, he was the first to talk about the "*section caesarienne*". Rousset believed that the name Caesar derived etymologically from the concept of delivery through incision, too. As did Caspar Bauhin, who in 1586 translated Rousset's writings into Latin: as a result, "*section caesarienne*" became "*sectio caesarea*".

CONCLUSIONS

Starting with Plinius the Elder, the name of Caesar has been considered related to childbirth by incision of the maternal womb. This concept has been supported by the popular belief that histerotomy was the more risky and difficult surgical intervention. The term "caesarean section" has now entered common and scientific usage of every civil language: tautology could be avoided by replacing it with "caesarean delivery".

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ABS 21

THE IMPORTANCE OF A SYSTEMATIC APPROACH TO THE MACROSCOPIC EXAMINATION OF THE PLACENTA: THE EXCESSIVE LENGTH OF THE UMBELICAL CORD CAN INDUCE PERINATAL ASPHYXIA

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The umbilical cord performs an important function throughout fetal development as it is responsible for the transport of oxygenated blood from the placenta to the fetus, through the umbilical vein, and the transport of deoxygenated blood from the fetus to the placenta, through the two umbilical arteries. Umbilical cord development begins at the third week of gestation, and it is fully formed at the seventh week. Any alteration that arises in the period of its development or subsequently can lead in turn to serious alterations in fetal development, sometimes with fetal morbidity and mortality.

The length of the umbilical cord is approximately 50-60 cm at a full-term newborn [1]. Umbilical cords above 70 cm are considered long whereas umbilical cord below 40 cm are short. The length of the cord and the correlation of this finding with adverse neonatal outcome has often been the subject of discussion. The length of the umbilical cord can depend on fetal movements and hyperactivity but also on genetic factors. The short cord, on the other hand, is found more frequently in infants with trisomy 21 or 18 or in infants with neurological deficits.

Excessively long umbilical cords are associated with increased morbidity and mortality for cord accidents, true knots, body or nuchal cord; long umbilical cord can cause blood flow alteration with increased risk of stasis for slow flow and low pressure resulting in fetal hypoperfusion and hypoxia. Growth restriction or fetal demise are significantly increased with long cords [2]. In our experience, most of the long umbilical cords also appear hypospiralized with an increase in the alteration of the blood flow due to the lack of spiral support. For these reasons excessive long umbilical cords are associated with birth asphyxia, low Apgar score, neurological compromission. Perinatal asphyxia is an important cause of morbidity and mortality in the perinatal period, and it is correlated to numerous histologic placental features, for example: thrombosis, maternal vascular malperfusion or inflammation.

The histologic examination of the placenta is essential for the purposes of a correct classification of the neonatal pathology, but it is sometimes a paradoxical histological reading as it is not consistent with the state of health of the newborn. The pathology of the excessively long umbilical cord can be cause of perinatal asphyxia and can only be diagnosed if the histological examination of the placenta is preceded by a detailed, careful and critical macroscopic examination.

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ABS 22

ORBITAL ALVEOLAR RHABDOMYOSARCOMA, ACUTE PRESENTATION IN A 10-YEAR-OLD GIRL: A CASE REPORT

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INTRODUCTION

Rhabdomyosarcoma is an aggressive tumor that can develop in almost every body area, arising from mesenchymal cells which are starting differentiation as muscles. It is the most common type of soft tissue sarcoma among children and young adults, with an annual incidence of 5 cases in 1 million below the age of 20. There are two peaks in the presentation age prevalence; between 2 and 6 and between 10 and 18 years of age, with a slight increase among males and the Caucasian ethnicity [1]. From a histologic point of view the most important types are: embryonal (80%) with a better prognosis, and alveolar with a worse one [2].

MATERIALS AND METHODS

A 10-year-old girl arrived at the ER with pain during left eye movement associated with exophthalmos and slight palpebral edema which appeared 2 days earlier. There were no itching or ocular secretions, only small lacrimation and sporadic blood traces coming from the homolateral nostril. There was pain during palpation of the frontal and maxillary sinuses, while there were no symptoms such as headache, vomit or fever. During physical examination there were exophthalmos of the left eye associated with slight eye movement limitation in the upward and lateral direction, without cutaneous hyperemia, secretions, and negative neurological examination. In order to confirm or deny the hypothesis of orbital cellulitis, blood workup was done and showed negative inflammation indices; the ophthalmologist suggested to start antibiotic therapy anyway. A head CT scan was performed, where a retro-orbital expansive lesion was observed with likely malignant features, given the timing from the onset of symptoms and the important infiltration. Before transferring the patient to the Onco-Hematology Department of the Padua Hospital, more investigations were carried out, such as total body CT scan, blood exams (uric acid, LDH, alkaline phosphatase, fibrinogen: all normal) and ENT evaluation with rhinoscopy.

RESULTS

In Padua, to complete the diagnostic workup, the histologic exam was performed, finding a parameningeal alveolar rhabdomyosarcoma. Chemotherapy protocol was then readily started.

CONCLUSION

Children with orbital rhabdomyosarcoma (almost 10% of all cases) may have as only signs exophthalmos or proptosis and these are often mistaken for signs of sinus or soft tissue infection [3]. However, in the first case patients with this localization show no symptom of inflammation, such as fever and redness. For this reason, imaging is pivotal in the differential diagnosis of unilateral exophthalmos, and should be performed without delay.

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ABS 23

THE EMERGENCE OF CONSCIOUSNESS-STATE DEPENDENT COMPLEXITY: SEPs AND PERTURBATION COMPLEXITY INDEX IN NEWBORNS AND YOUNG INFANTS

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During sleep the conscious perception of sensations disappears, so sleep and wakefulness are examplary models for studying unconscious and conscious sensory processing. During sleep when consciousness fades, EEG responses are stronger as the disruption of effective cortical connectivity occurs: event-related potentials (ERPs) are found to have a greater amplitude during sleep and a less extensive but more diffuse response during wakefulness. This process of segregation of brain responses is an evolutionarily important mechanism that preserves sleep from awakening induced by sensory stimulation. To assess the complexity of brain responses related to consciousness, the perturbative complexity index (PCIst) that is associated with different states of consciousness can be used, and in particular in adults, higher PCIst values have been found in wakefulness than in sleep. In parallel, previous studies have shown that somatosensory evoked potentials (SEPs) influence sleep by determining higher values in sleep than in wakefulness, already at birth. In this study, we researched the emergence of the complexity of brain responses by exploiting a new approach based on PCIst computation on responses elicited by peripheral stimulation. We collected EEG responses to median nerve electrical stimulation during wakefulness and sleep in newborns (N = 9; age = 12-36 hours) and infants (N = 9; age = 3-4 months). Both subjects were recruited from Sant'Anna Hospital in Turin and had physiological conditions. As in previous studies, in newborns' SEPs, the mean amplitude of the middle- and long-latency components was greater in sleep than in wakefulness, in the centroparietal (P1: $p = 0.03$; $t = 2.56$) and frontocentral (P2: $p = 0.05$; $t = 0.05$) clusters. Consistently, infants' results showed the same pattern in early- and late-latency SEPs components, in centroparietal (N1: $p = 0.01$, $t = 3.11$) and parietal (P2: $p = 0.05$, $t = 2.32$) clusters. In contrast, comparing PCIst values, an opposite trend was found: newborns showed significantly higher PCIst values in sleep than in wakefulness ($p = 0.004$; $t = 3.95$); infants, as well as adults, showed higher PCIst values in wakefulness than in sleep ($p = 0.005$; $t = 3.82$). From the results obtained, we can state that while ERP values show greater segregation of brain responses during sleep already at birth, PCIst values suggest that the working structure related to brain complexity is fully developed in the postnatal period: already in the first 3 months infants present the same pattern of adults, with more brain complexity during wakefulness than during sleep.

ABS 24

BARTONELLA HENSELAE RELATED ENCEPHALOPATHY: DIAGNOSIS AND CLINICAL COURSE

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INTRODUCTION

Bartonella henselae (BH), is the etiologic agent of cat-scratch disease (CSD). The illness is typically self-limited, characterized by regional lymphadenitis following cutaneous inoculation, and variably accompanied by low-grade fever, malaise, and headache [1].

CASE DESCRIPTION

An 8-year-old female child with lateral-cervical lymphadenitis was admitted to hospital for convulsive episode with clones in the upper limbs, flexion and torsion movements of the head, muscle hypertonus, and progressive reduction of consciousness leading to transfer to the Intensive Care Unit. Rachicentesis showed: clear cerebral-spinal fluid (CSF) at normal pressure, hyperglycorrachia, 3 cells, negative film array for viruses and bacteria, and negative autoimmunity and oligoclonal bands. Brain MRI was negative, and EEG showed diffuse slow abnormalities. For the anamnestic data of contact with cats and the presence of lymph-nodes enlargement, serum BH antibodies IgG and IgM were dosed, resulting: positive at high titer and leading to the diagnosis of BH related encephalopathy. Specific antibodies in CSF were yet negative. The therapy was azithromycin 10

mg/kg/day and rifampicin 10 mg/kg BID. After 24 hours patient showed progressive and rapid improvement of the general conditions and state of consciousness, with resolution of the convulsive manifestations. EEG revealed disappearance of the previously reported alterations in 2 days. To date the patient does not present any sequelae.

DISCUSSION AND CONCLUSIONS

Although it is generally considered a self-limiting disease, CSD cases can be accompanied by serious complications. The infection can spread to several organs and present systemic symptoms, such as febrile spikes, hepatosplenomegaly and abdominal pain [2]. Neurologic involvement in CSD is rare, occurring in 2.0% of cases; with onset occurring from a few days to 2 months after infection. CSF is usually normal or with mild mononuclear pleocytosis. CT or MRI can be negative or occasionally reveal focal or transient abnormalities, EEG is typically nonspecific, demonstrating diffuse slowing that returns to normal in nearly all patients. Awareness of the relationship between CSD and encephalopathy remains limited.

The pathophysiologic mechanisms responsible are unknown. Possibilities include a deficient host immune response, elaboration of a neurotoxin, or direct CNS invasion. In consideration of normal or minimally abnormal CSF, and an uneventful, rapid neurologic recovery in most cases, the possibility of direct CNS bacterial invasion appears less likely. Different antibiotic regimens have suggested, and convulsant therapy can be indicated. Prognosis for recovery is usually excellent [1, 2]. This case underlines the absence of signs of CNS invasion together with encephalopathic manifestations reversible with treatment, and suggests the importance of considering CSD as differential diagnosis in any case of acute neurological manifestations with compatible anamnestic history [3].

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ABS 25

A MALE INFANT WITH NATAL TEETH. A CASE REPORT

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INTRODUCTION

Teeth present at the time of birth are called natal teeth while the teeth which erupt within 30 days of birth are neonatal teeth. The incidence ranges from 1: 2,000 to 1: 3,500.

Exact etiology for the premature eruption or for appearance of natal and neonatal teeth is not known. In the past, neonatal teeth were merely considered cysts of the dental lamina of the newborn. It was also suggested that they occur due to inheritance as dominant autosomal trait. Endocrine disturbance resulting from pituitary, thyroid, and gonads also may be one of the key factors.

Early detection and treatment of natal teeth are recommended because they may induce deformity or mutilation of tongue, dehydration, inadequate nutrients intake by the infant, and growth retardation.

A case report of a male infant with natal teeth is reported below.

CASE REPORT

A male infant with gestational age 37^{+2} weeks was delivered by C-section due to cephalopelvic disproportion. Natal teeth were present at the clinical examination. All the teeth were white to yellowish white in color, mobile, and were devoid of roots.

A maternal medical history revealed the presence of hypothyroidism treated with thyroxine as well as a treatment with prednisolone due to rheumatoid arthritis. A paternal relative with the presence of natal teeth in both father and grandfather was reported.

After delivery, the infant was transported to the Neonatal Intensive Care Unit in order to control the infant's milk intake.

The routine laboratory tests were normal.

Further examinations: normal cranial ultrasound; normal X-ray of the bones.

Due to the paternal medical history, a genetical examination (next generation sequencing) was performed.

RESULT

Genetical examination is presented in **Tab. 1.** A genetical examination of both parents was suggested. The mother was negative for the mutation c.812A>C, while the father was heterozygote for

Table 1 (ABS 25). Genetical examination.

Gene	Nucleotide change	Amine oxide change	Result	Zygote	Clinical relevance
KDF1	c.812A>C	K271T(p.Lys271Thr)	Missense	Heterozygote	Unknown

the same *KDF1* mutation. Furthermore, a genetical examination of the paternal grandfather has also revealed the same nucleotide change at the *KDF1* gene in heterozygous genotype.

DISCUSSION

Keratinocyte differentiation factor 1 (*KDF1*) is a recently identified gene related to tooth development, but it has been little studied. *KDF1* mutations are related to tooth development, including ectodermal dysplasia cases accompanied by tooth loss as well as non-syndromic cases with tooth agenesis. The nucleotide change c.812A>C has not yet been reported in the international scientific resources. This mutation is supposed to cause changes in the structure and function of the produced protein and can be assorted in the category of changes of unknown clinical relevance. A further research is necessary, in order to find out how far this mutation is correlated with the appearance of natal teeth and how far the development of teeth, hair, nails and other ectodermal tissues can be affected.

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ABS 26

HUMAN HERPES VIRUS 6 HEPATITIS IN A CHILD WITH ANEMIA AND SHIGA TOXIN *E. COLI* (STEC) GASTROENTERITIS

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INTRODUCTION

Human herpesvirus (HHV-6) is a Betaherpesvirus of the Herpesviridae family. Seroprevalence is over 90% in many parts of the world [1]. It typically manifests itself between 6 months and 3 years as exanthema subitum associated with or without febrile seizures [2]. More rarely, during primary infection, there are other more serious manifestations such as hepatitis, even fulminant, gastroenteritis, myocarditis or neurological complications [3].

CASE REPORT

A 14-month-old patient with beta thalassemia trait was transferred from a first-level hospital to our pediatric ward because of recurrence of microcytic anemia, mild hypertransaminasemia, multiple vomits, diarrhea and fever (38.5°C) for 2 days. Diagnosis at first glance was a possible metabolic disease. At admission the liver function was examined both through complete lab tests with evidence of hepatocellular necrosis and signs of cholestasis (ALT 241 U/I, AST 418 U/L, GGT 226 U/L, total bilirubin 2.2 mg/dL, direct bilirubin 1.9 mg/dL) and through abdominal ultrasound showing increased echogenicity of the liver associated with gallbladder hydrops. Microcytic anemia (Hb 7.5 g/dL, MCV 50.6 fL) was associated with iron deficiency. As a result of a close monitoring, there was a progressive clinical improvement, with resolution of vomiting and diarrheal disease, and almost normalization of laboratory tests, with the last tests performed on the 4th day of hospitalization showing ALT 120 U/L, AST 87 U/L, GGT 161 U/L, total bilirubin 0.65 mg/dL, direct bilirubin 0.48 mg/dL. On the 1st day of hospitalization there was also a stable normal temperature followed by a non-itchy papular macular rash spread over the trunk and limbs, which persisted for about 48 hours.

The microbiological investigations carried out during the hospital stay showed positivity in the film array for *E. coli* producing toxin (STEC) on faeces, and positivity on blood PCR for HHV6 with high viral load.

CONCLUSIONS

The initial "metabolic" diagnosis was actually composed of three different diseases, which unusually occurred at the same time in the same

patient: acute gastroenteritis caused by STEC, liver inflammation caused by HHV6 and microcytic anemia in beta thalassemia trait associated with iron deficiency. Although the above conclusions are the most plausible, it cannot be excluded that acute gastroenteritis picture could be caused by HHV6 as well and that *E. coli* could be considered as microbial colonization.

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ABS 27

NANOG IS HIGHLY EXPRESSED IN THE DEVELOPING HUMAN HEART

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INTRODUCTION

Nanog is a stem cell transcription factor expressed by pluripotent stem cells in mice. It is named after the mythological celtic land "Tir Na Nog" (the land of the ever young). Nanog expression has been linked to the regulation of self-renewal in embryonic stem cells. During development, Nanog is crucial for the pre-implantation phase and for regulation of development, being involved in cell proliferation and apoptosis [1]. After birth, Nanog is silenced in most tissues through life-span. Limited data are available of Nanog expression in early human development and in fetal development, Nanog expression being restricted to testes and ovaries [2]. Recently, Nanog was reported to be expressed in mesenchymal stem cells isolated from human fetal hearts [3].

MATERIALS AND METHODS

In this study, we analyzed, by immuno-histochemistry, the expression of Nanog in the heart of 10 human fetuses whose gestational age ranged from 13 up to 26 weeks of gestation. Paraffin tissue sections were immunostained for Nanog utilizing a commercial antibody (clone AB80892). As a negative control, a sample of an adult heart was used. Results. Nanog expression was detected in all the fetal myocardial samples analyzed. Nanog protein was mainly expressed along the cell membrane of myocardiocyte precursors. Regarding the intensity of the immunoreaction, the highest levels of Nanog expression were detected in the youngest embryos, at 13 weeks of gestation. No reactivity for Nanog was observed in the adult heart.

CONCLUSIONS

Our study evidence that Nanog is highly expressed in the developing human heart, particularly in the early phases of human development. These preliminary findings indicate that *Nanog* gene plays a major role in the differentiation of myocardial precursor cells during human development. Further studies will better characterize Nanog expression during all the phases of pregnancy, in order to describe cardiac Nanog expression in all stages of intrauterine development.

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ABS 28

TALKING ABOUT TO SUPPORT SCREENING IN POSTPARTUM DEPRESSION

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Postpartum depression (PPD) is a mood disorder that affects up to 1/5 of new mothers. It manifests itself with the typical symptoms of major depression and typically occurs within 6 months of delivery. The consequences of this pathology affect both the health of the mother and that of the child, which could undergo alteration of the affective, behavioral and social development, but

also on the relationship with the partner (including the paternal postpartum depression). An early diagnosis that allows for the timely establishment of an adequate therapy is therefore essential.

Talking About (TA) is a Speech Emotion Recognition project (SER) made by G.P.I. (Trento, Italy) which consists of a series of artificial intelligence algorithms that have the purpose of reliably recognizing emotions and/or stress through audio streams of the human voice (e.g. telephone calls, telemedicine). Voice acquisition takes place by recording an audio while the mother describes an image chosen from a series.

The study aims to compare the results obtained through TA and the score obtained with the Edinburgh Postnatal Depression Scale (EPDS) which is the most widely used tool for screening for PPD, in order to evaluate the possible correspondence of the results. Both tests (EPDS and TA) are administered 1-2 months after delivery through the Haumea platform, created by G.P.I. specifically for this purpose, which allows the administration of the tests both in presence, through the use of a tablet, and remotely via video call.

In total, 64 women were recruited: 53 in the puerperium, close to childbirth, and 11 by telephone. Consequently, postnatal recruitment proved to be slightly more effective. To date, 30 of these 64 patients have performed the test (46.8%). Only 4 of these 30 patients (13.3%) were able to carry out the test in presence, the remaining 26 (86.7%) chose, for convenience, to carry out the remote test via video call.

The test was on average 52 days after delivery.

The administration of the test through Haumea is divided into 3 phases:

1. collection of statistical data (age of the mother, gestational age, weight of the newborn at birth, etc.) and more specific data relating to any risk factors for PPD (family history of depression, complications in the peripartum, health problems of the newborn, etc.) in order to stratify the cases examined;
2. EPDS administration;
3. administration of the TA test.

A preliminary analysis of the data obtained so far shows how TA is able to provide a specific profile for each new mother. These profiles are presented as a specific combination of 5 different categories that describe the emotional state of each patient: "I feel good", "It is a good day", "I do not agree", "I am tired", "I am pessimistic". Specifically, each of these categories is expressed as a percentage, thus highlighting different aspects of the same mother.

Examples of profiles obtained are presented hereafter.

Example 1: "I feel good" 24.10%; "It is a good day" 32.3%; "I do not agree" 40.8%; "I am tired" 0.1%; "I am pessimistic" 2.7%.

Example 2 (patient who did not sleep the whole night): "I feel good" 9%; "It is a good day" 0%; "I do not agree" 0.7%; "I am tired" 90.3%; "I am pessimistic" 0%.

Thus, it is interesting to note how with TA it is possible to highlight different emotional facets of a subject through a single test, unlike the single score obtained with the EPDS which inevitably, being a screening tool, is based on specific cut-offs useful for stratifying the risk of PPD. In fact, from the administered tests, it emerges that women who obtained the same score on the EPDS presented more or less different profiles at the TA, precisely because this test investigates different aspects compared to the EPDS. Therefore, it remains to be investigated whether, once the results obtained with TA have been standardized, there is a good overlap of the different profiles belonging to patients who have scored the same score on the EPDS and a correlation between the 2 tests can therefore be identified. This could be of interest in order to further improve the screening process and hypothetically allow for an ever earlier diagnosis of PPD.

ABS 29

SEVERE ACUTE HEPATITIS OF UNKNOWN ORIGIN ASSOCIATED WITH BONE MARROW HYPOPLASIA IN A 4-YEAR-OLD BOY: A RARE CASE REPORT

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INTRODUCTION

On March 31, 2022, Public Health Scotland was alerted to 5 previously healthy children hospitalized with severe acute hepatitis of unknown origin (SAHUO). As of July 8, 2022, 1,010 probable cases of this disease have been reported to the World Health Organization (WHO).

The WHO has defined a probable case as a person ≤ 16 years old presenting with an acute hepatitis with aspartate aminotransferase (AST) or alanine aminotransferase (ALT) levels > 500 IU/L, excluding cases due to hepatitis A-E or other explanations.

Clinical presentation may vary from mild symptoms to acute liver failure. Most affected children recover fully with supportive care, but liver transplantation may be required for cases with severe liver failure.

CASE REPORT

We describe a recent case of a previously healthy 4-year-old boy presenting with jaundice and scleral icterus. His parents reported an infection with SARS-CoV-2 approximately 15 days prior to hospital admission. Laboratory work-up for hepatitis showed an important increase in markers of liver cells injury (AST, 1,606 IU/L; ALT, 2,305 IU/L; γ-glutamyl-transferase, 209 IU/L) and in serum levels of total bilirubin (7.11 mg/dL) and direct bilirubin (4.72 mg/dL).

The search for HAV, HBV, HCV, HEV, EBV, CMV, HSV, and parvovirus yielded negative results. SARS-CoV-2 RT-PCR testing performed on nasopharyngeal swab sample was repeatedly negative; moreover, the patient tested positive for anti-SARS-CoV-2 IgG, while tested negative for anti-SARS-CoV-2 IgM.

An autoimmune profile showed weak positivity for serum anti-nuclear antibodies (1:80) and anti-smooth muscle antibodies (1:40). Blood levels of alpha-1 antitrypsin, ceruloplasmin, cupremia, and biochemical test for lysosomal diseases were all within the normal range. The coagulation profile revealed an increase in INR (1.79) and deficiency of vitamin K-dependent factors.

Abdominal ultrasound scan and MRI cholangiography documented hepatomegaly in the absence of biliary tract dilatation. Complete eye examination and cardiological evaluation were unremarkable.

The patient was successfully treated with rehydration, ursodeoxycholic acid, and intravenous vitamin K, with gradual resolution of clinical manifestations and normalization of markers of liver cells injury, bilirubin and coagulation. On day 25, the patient was discharged home.

During the follow-up period, progressive leucopenia and thrombocytopenia were observed, with nadir values (WBC count, 1.99×10^3/μL; PLT count, 28.0×10^3/μL) at about 30 days after discharge. Therefore, a bone marrow aspirate was done and revealed bone marrow hypoplasia. Accordingly, the patient was referred to a tertiary academic pediatric center to be evaluated for a possible bone marrow transplant.

DISCUSSION AND CONCLUSIONS

The association between seronegative autoimmune hepatitis and aplastic anemia has already been reported in the literature. In some cases, affected patients become reliant on blood transfusions and/or require bone marrow transplant.

Our rare pediatric case of acute hepatitis of undetermined origin associated with bone marrow hypoplasia is still to be precisely classified, as it has features in common with both the recently described SAHUO and the seronegative autoimmune hepatitis. Moreover, it highlights the importance of a prolonged follow-up, considering that severe hematological complications may occur even after the resolution of liver disease.

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ABS 30

FEEDING THE LATE PRETERM INFANT: IMPACT OF USING A VALVED FEEDING BOTTLE WITH AN ERGONOMIC TEAT ON THE COORDINATION OF THE SUCKING, SWALLOWING AND BREATHING PROCESS

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INTRODUCTION

The coordination of sucking-swallowing-breathing is the main physiological element that guarantees safe oral feeding, without the risk of adverse cardiorespiratory events.

In the late preterm infant this coordination may not be mature, consequently this population has greater risk of complications associated with oral feeding.

The introduction of rear-ventilated valve bottles with ergonomic teat, which guarantee the outflow

of milk only if activated by sucking, could result in a more physiological sucking-swallowing-breathing pattern.
The purpose of this study is to evaluate the impact of using these bottles on the coordination of sucking-swallowing-breathing in the late preterm infant.
MATERIALS AND METHODS
A randomized controlled study was conducted at the Neonatal Intensive Care Unit (NICU) of the Sant'Anna Hospital in Turin from 25 November 2020 to 15 April 2022 on late preterm infants of gestational age 34^{+0}-36^{+6} fed, partially or exclusively, with the bottle. The infants were assigned to two groups: valved bottle ergonomic teat (B-EXP) and standard bottle (B-STD).
The sucking-swallowing-breathing process was assessed by non-invasive synchronous polygraphic monitoring at the time of a bottle-fed meal in the first 24-48 hours of life (T1) and repeated at 72-96 hours of life (T2).
The primary outcome of the study was the assessment of the suck-swallow-respiration ratio.
RESULTS
20 infants were enrolled, with mean gestational age 35.05 ± 0.82 weeks, 10 (50%) in the B-EXP group and 10 (50%) in the B-STD group. No differences were observed between the two groups in the main outcome analysis. The secondary outcome analysis found greater meal volume and duration (16 ± 5.67 vs 21 ± 6.58 mL and 183 ± 124.78 vs 308.8 ± 204.2 s, respectively) and lower milk flow in the B-EXP group (12.24 ± 11.3 vs 8.566 ± 4.09 mL/min) ($p > 0.05$). The frequency of apneas was higher in the B-STD group (0.946 ± 0.82 vs 0.696 ± 0.64 apneas/min) while the respiratory rate was higher in the B-EXP group (33.04 ± 12.7 vs 37.51 ± 13.24 acts/min) ($p > 0.05$). The percentage of dangerous swallows on inspiration was higher in the B-STD group (0.219 ± 0.13 vs 0.169 ± 0.12%) ($p > 0.05$). The composite analysis of secondary outcomes was statistically significant at T1 ($p < 0.05$), not statistically significant at T2 ($p > 0.05$). The prevalence of breastfeeding at discharge was higher in the B-EXP group, 70% vs 20% ($p < 0.05$).
CONCLUSIONS
This study has shown that the use of these rear-ventilated valve bottles with ergonomic teat, during the first days of the infant's life, results in better coordination and lower risk of cardiorespiratory events.
We have also shown how these infants, getting used to the mechanism of sucking with suction, are then able to approach breastfeeding more easily than infants who have used a standard bottle.

ABS 31

FOUR CHARACTERS IN SEARCH OF AN AUTHOR. PEDIATRIC "FICTITIOUS" SYNDROMES DURING COVID-19 PANDEMIC

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INTRODUCTION
Psychophysic stress caused by COVID-19 pandemic has increased the prevalence of psychiatric and psychological pathologies (and, among others, of simulation disorders) in the pediatric population [1]. Scientific literature has demonstrated how this vulnerable age group is more susceptible to the psychosocial impact of the pandemic rather than to the actual disease.
CASE REPORTS
Case no. 1
A 12-year-old boy arrives at the ER reporting constant headache and hypertension. Investigations carried out included blood pressure Holter, CT scan, MRI, renal vessels ecography, EEG, blood workup for pheochromocytoma, all with negative results. During a second hospitalization for the same clinical presentation, bruising on the arm at the level of where the blood pressure was measured was witnessed. The patient during every blood pressure measurement voluntarily contracted the arm muscles, in order to increase the measured value.
Case no. 2
A 10-year-old boy arrives at the ER for weakness, sudden inability to walk, apparent inability to locate tactile and pain stimuli. For the suspicion of spinal lesion an MRI was performed, with poor quality of the image due to movement artifacts. While communicating to the parents the necessity to repeat the exam, the patient suddenly recovered from all the referred symptoms.
Case no. 3
A 14-year-old girl arrives at the ER for long standing fever (20 days) and headache with

consequent great impact on school attendance. During hospitalization, the patient was always apyretic and presented only one episode of headache resolved by infusion of saline.

Case no. 4

A 7-year-old boy arrives at the ER for diplopia lasting for 20 days, treated with prednisone, without any reported improvement. During eye examination, no pathological signs were found, and correction of the symptom was achieved by applying neutral lenses.

DISCUSSION AND CONCLUSIONS

This small group of cases shows some examples of simulation disorders observed in the Emergency Department. The simulatory nature of the diseases was supported by a negative physical examination and symptoms resolution after placebo administration.

Many studies have demonstrated how in pediatric age the COVID-19 might be the trigger of monotony, anxiety, depression and neuropsychiatric disorder in general [2]. The results are long standing illnesses, numerous accesses to the ER, improper hospitalizations and negative impact on school attendance and daily activities [3]. Future research should explore all the aspects of the psychological burden of COVID-19 pandemic in order to put in place adequate mental health strategies for infants and adolescents, consisting of direct or digital collaborative networks of psychiatrists, psychologists and pediatricians.

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ABS 32

IMPACT OF CONGENITAL HEART DISEASES IN MALFORMATIVE SYNDROMES IN CENTRAL-SOUTHERN SARDINIA: EXPERIENCE OF A 3-YEAR PERIOD

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Congenital heart diseases have a complex and heterogeneous etiology: 70% of congenital heart diseases occur as isolated malformation, while 30% correlate with extracardiac malformations and chromosomal syndromes. In Europe, in 2017, the prevalence of newborns with congenital heart diseases was 0.69%, while the prevalence of newborns with chromosomal syndrome was 0.168% (EUROCAT data).

This study considered the medical data of newborns admitted in the 3-year period 2016-2018 at the University Hospital of Monserrato (AOU Cagliari), the main birth center of central-southern Sardinia, equipped with a Neonatal Intensive Care Unit.

In a cluster of 7,880 live births in the 3-year period 2016-2018 in the Metropolitan City of Cagliari, 2.2% newborns presented birth malformations. Of 171 babies with congenital malformation, 38 had isolated congenital heart diseases (0.48% of the population studied) and 31 had a chromosomal syndrome (0.39% of the population examined). Of 31 babies with chromosomal syndrome, 30 had at least one congenital heart abnormality, which means 96.77% of the cases. The most frequently identified syndromes were Down syndrome (13 out of 31 cases), Cayler syndrome (3 out of 31 cases) and Noonan syndrome (2 out of 31 cases). Three cases were labelled as "suspected syndrome" as the diagnosis of chromosomal syndrome by genetic investigation had not yet been confirmed at the patient's discharge. They all presented the association with at least one congenital heart disease, except the case of Wolf-Hirschhorn syndrome.

The most important fact is surely that 52% of the newborns with chromosomal syndrome presented a clinical situation characterized by at least two concurrent congenital cardiological abnormalities. In most cases, the cardiological situation was the first sign of alarm and criticality of the newborn, and countless cases required even multiple interventional and surgical procedures for the resolution of critical cardiological frameworks, compromising the early stages of development of the newborn and infant.

According to this study, central-southern Sardinia recorded an incidence of 0.39% of live birth babies suffering from chromosomal syndrome, a data that dramatically clashes with the 0.168% of incidence in Europe recorded by EUROCAT. In Sardinia, there is no register of congenital malformations: it would be desirable that a unique

and accurate data collection could be formulated as soon as possible to investigate the etiology and the clinical and anamnestic data of subjects suffering from congenital malformations. This would allow more realistic values to be reported in European statistics, which could also consider data coming from realities such as Sardinia.

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ABS 33

LANGUAGE DISORDERS AS COMORBIDITY IN ENURETIC CHILDREN: PREVALENCE AND IMPACT ON TREATMENT

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PURPOSE

Several studies have recorded an association between nocturnal enuresis (NE) and developmental delays. Language disorders are one of the main comorbidities in enuretic children, affecting therapy outcome and prognosis.

METHODS

The study was carried out within our Pediatric Outpatient Clinic in Campus Bio-Medico University Hospital Foundation of Rome, enrolling a total of 226 enuretic children, of which 21 with a language disorder. Data concerning NE were collected through questionnaire and interview with parents and a bladder diary. The language disorders were secondary investigated. The chi-squared test was used to evaluate the correlation between therapy outcome and language disorders.

RESULTS

Our sample was composed of 226 enuretic children, with a mean age of 7.67 years. 217 children were subjected to pharmacological treatments, while 9 children, with mild clinical symptoms, did not take any drugs. It was observed that 49% of enuretic children did not respond to therapy or relapsed, while 51% responded in the first cycle. 21/226 (9%) of enuretic children had a language disorder, of which 15 subjected to pharmacological therapy: 12/15 (80%) did not respond to therapy and 3/15 (20%) responded ($p = 0.024$).

CONCLUSION

Language disorders could impact NE therapy response, underlying the influence of comorbidities and the importance of a global assessment in enuretic children.

ABS 34

IF ANEMIA RELAPSES... LOOK AT THE BOWEL!

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An 8-year-old patient with no past medical history was admitted for hypochromic, microcytic anemia (Hb 5.6 g/L, MCV 51 fL, ferritin 3 ug/L, blood iron 10 ng/dL). Physical examination was unremarkable except for pallor. She was diagnosed with iron deficiency anemia, received one packed RBC transfusion and was discharged with oral iron replacement therapy. Despite an adequate response to iron supplementation (Hb 12 g/L), there was a recurrence of anemia with rapid drop in hemoglobin levels by the end of the treatment. She was screened for hemoglobinopathies, hemolysis and possible causes of gastrointestinal bleeding such as coeliac disease, *Helicobacter pylori*, or inflammatory bowel disease: these investigations revealed negative results. She performed repeated fecal occult blood test (FOBT) that turned out positive, therefore her anemia was attributed to gastrointestinal blood loss. She underwent esophagogastroduodenoscopy (EGD) and colonoscopy, with negative results. Meckel scintigraphy and Tc-99m labeled RBC scintigraphy were performed and a gastrointestinal bleeding site in the ileal mucosa was detected. The patient then underwent video-capsule endoscopy (VCE), that showed a lesion of the jejunal tract, with histological aspect of well-differentiated gastric mucosa. VCE

was then repeated with balloon-assisted endoscopy (BAE): 2 polypoid tumors with ulcerated surface on the jejunal mucosa were detected. Resection was performed under laparotomy. Histopathological evaluation revealed 6 fundic cystic polypoid tumors with heterotopic gastric mucosa with physiological beta-catenin membrane positivity. The patient is now asymptomatic and presented with adequate Hb levels at follow-up.

Iron deficiency anemia is the most frequent form of anemia in children. Iron-deficiency anemia due to gastrointestinal bleeding needs to be suspected in case of refractory, relapsing anemia or positive FOBT. Source of bleeding is located in the small intestine in 75% of cases in pediatric age and it usually underlies polyps, vascular malformation, Meckel diverticulum or inflammatory bowel diseases [1]. Diagnostic investigations include Tc-99m labeled RBC scintigraphy, EGD and coloscopy. In case of negative results, VCE and BAE are recommended since they provide a higher diagnostic yield especially when combined together. VCE and BAE were fundamental to achieve a diagnosis of polypoid gastric heterotopia in our patient, even if surgery was still needed. To our best knowledge, there is limited literature concerning polypoid gastric heterotopia in children. Polyps are described as isolated lesions whose clinical manifestation depends on the size and their anatomical location [2]. Considering the multiple polypoid lesions found in our patient, genetic investigation has been performed to rule out familial adenomatous polyposis (FAP). Indeed, a new FAP clinical variant associated with *APC* mutation and characterized by gastric polyposis has recently been described [3].

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ABS 35

MADELUNG DEFORMITY: A STRANGE SWELLING

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A 10-year-old girl is accompanied by her parents to the Pediatric Office because she fell and suffered an injury to her right forearm and wrist. The child is in good general condition. The parents report that she now has pain in her arm from the fall that occurred while playing, but that she had been complaining of mild pain in her right wrist for several months. This symptom was not disabling, more of a discomfort that temporarily prevented her from performing common daily activities.

The fall the day before had accentuated the pain by increasing and making the functional impotence persistent. On objective examination, nothing special was observed except for a turgor at the level of the right wrist, but it was not the swelling typical of juvenile bone fractures referred to as "green wood", it looked more like a deviation of the axis of the forearm in relation to the hand, but not attributable to the trauma suffered.

Anthropometric data from the child: kg 46 (90^{th}-95^{th} centile), H 141 cm (50^{th}-75^{th} centile), BMI (kg/m^2) 23.1, Haycock method body surface area 1.35.

The child had been fostered by these parents for only a few months, so no family history data could be traced. The child recounted that she occasionally "could not use her right hand well according to the movements she was making or if she had to make any effort", i.e., she had some difficulty in supination with decreased strength. Full flexion extension was maintained while prone supination was maintained but with some difficulty, but it was unclear whether this was attributable to the trauma.

Sent to the Hospital, she was first X-rayed on the right forearm and wrist. No fracture line was detected, but an incorrect alignment between the hand and wrist was observed. Consequently, a CT scan was performed, which yielded an axial deviation of the hand with prominence of the caput ulnae. The diagnosis was therefore Madelung's deformity.

This disease was first described by Otto Madelung in 1878. The malformation is transmitted by a dominant gene with incomplete penetrance (gender distribution: 4 females/1 male). It can be unilateral or bilateral, but more often it is unilateral. Radiologically, a greater inclination of the distal part of the radius may be observed. The

ulna may be longer than the radius. The proximal semilunar has a triangular, deformed, compressed face between the radius and ulna. The carpal is subluxated on the volar side. It can be isolated or associated with multiple exostoses (Ollier's disease, presence of multiple enchondromas), or posttraumatic or postinfectious. Treatment can only be surgical, particularly with early diagnosis the prognosis is always good; if neglected in the young, it can give much pain and functional impotence [1, 2].

In early onset, girls should be treated to avoid irreducible deformity of the distal forearm associated with reduced range of motion that may appear after puberty. So this little girl underwent surgery with wedge osteotomy opening of the distal third diaphyseal of the radius after section of Vickers ligament and synthesis with APTUS® Medartis plate, finally application for 30 days of plaster shower. Restitutio ad integrum was complete. In adult patients, an ulnar shortening osteotomy is operated to relieve wrist pain on the ulnar side in adult patients with Madelung deformity [3].

Madelung deformity should therefore not be considered a benign disease but should be carefully observed in the pre- and pubertal stages to intervene before it can cause irreversible damage in young girls.

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ABS 36

INBORN ERRORS OF IMMUNITY DUE TO DAMAGING MUTATIONS IN *NFKB2*

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Inborn errors of immunity (IEI) are a heterogeneous group of disorders mainly caused by monogenic mutations affecting the development and function of the immune system, thus leading to severe and recurrent infections, autoimmunity, atopy, and malignancies. One of the most common forms of symptomatic IEI characterized by hypogammaglobulinemia is common variable immunodeficiency (CVID). CVID diagnostic criteria are based on clinical manifestations and laboratory findings (including low serum immunoglobulins and poor response to vaccines) and require the exclusion of all other possible causes of hypogammaglobulinemia (e.g., infectious diseases, drug-induced, systemic disorders with protein loss or hyper-catabolism). Of note, the underlying genetic defects causing CVID, in most cases, remain undefined.

The case study describes a 28-year-old patient diagnosed with CVID in childhood. The patient presented with antibody deficiency, recurrent and atypical infections, including cerebral toxoplasmosis, and autoimmune manifestations, specifically vitiligo and psoriasis. Whole exome sequencing (WES) revealed a heterozygous nonsense mutation in the *NFKB2* gene (c.2557C>T, p.R853*) which has been confirmed with Sanger sequencing. The NF-kB signal transduction pathway is widely known for regulating inflammatory and immune responses. This gene is a key player in the non-canonical NF-kB signaling pathway crucial for peripheral lymphoid development, B cell differentiation, and antibody production.

We compared our findings with the data reported in the literature. Recently, Klemann et al. reviewed 50 reported cases with *NFKB2* mutations, out of which 46 have been diagnosed with IEI, 2 are asymptomatic, 1 individual has isolated ACTH deficiency, and 1 had rheumatoid arthritis onset at age 48. Of note, our patient does not have central adrenal deficiency, although Klemann et al. showed the concomitant presence of ACTH deficiency in 44% of the *NFKB2* patients' cohort. Overall, these findings expand the clinical spectrum of germline mutations in the non-canonical NF-kB signaling pathway as a genetic cause of CVID.

Further studies are required to increase our understanding of the genetic and molecular mechanisms underlying CVID pathogenesis for the development of targeted therapeutic interventions (the "precision medicine" approach)

based on the use of small molecules and biologics to target specific cell functions.

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ABS 37

ONCE UPON A TIME ORAL MICROBIOTA: A CINDERELLA OR A PROTAGONIST IN AUTISM SPECTRUM DISORDERS?

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INTRODUCTION

Autism spectrum disorder (ASD) is an early-onset heterogeneous, multifactorial neurodevelopmental disorder evolving over the lifetime of patients. ASD core features are deficits in social communication and interaction, associated with restricted, repetitive, and sensory-motor behaviors and interests requiring lifelong support. This work aims to discuss different aspects of salivary and dental microbiota between ASD and neurotypical children (NT), establish the oral microbial taxa associated with ASD and evaluate a possible correlation between oral microbiota and the biologic and etiopathogenetic features of ASD. Most studies of the ASD microbiome have focused on the gastrointestinal tract and the oropharynx is less considered.

ORAL-GUT-BRAIN AXIS

After the entry into the bloodstream, oral bacteria may reach the brain where they alter the neuroimmune activity and promote inflammation. Routine dental interventions could cause transient bacteremia, and some pathogens may cross the blood-brain barrier (BBB) colonizing the central nervous system (CNS). The bacterial migration from the mouth to the brain is facilitated by the loss of integrity of the BBB in ASD individuals due to microglia deterioration. Oral dysbiosis in ASD could be reflected by changes in the metabolic profile with the overexpression of metabolites closely related to the pathogenesis of ASD [1].

RESULTS

In animal models, it was demonstrated a close relationship between dysbiosis and abnormalities in social behavior; the reactivation of the gut ecosystem balance may significantly contribute to mitigating ASD symptoms. Transplantation of gut microbiota from human donors with ASD into mice may promote autistic behaviors, modifications in brain function, and host metabolism in recipient mice. Mice receiving oral microbiota from the ASD donor showed significantly different microbiota structures in their oral cavity and intestinal tract, compared with those receiving from controls and those not receiving any bacterium [2].

CONCLUSION

Oral dysbiosis in ASD could be reflected by changes in the metabolic profile with the overexpression of metabolites closely related to the pathogenesis of ASD. There is growing evidence that the analysis of the oral microbiota and metabolome is crucial for the early diagnosis and management of ASD. This approach allows the identification of subclinical or clinical subgroups of ASD patients with gastrointestinal disease, autoimmune diseases, or inflammation [3].

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ABS 38

NOT ONLY ENURESIS: DO NOT DISREGARD ORGANIC DISORDERS

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Nocturnal enuresis (NE) is a common condition in the pediatric age. NE is defined as an intermittent

bedwetting with any frequency while sleeping in children. NE is classified into primary form (patient never had achieved nocturnal urinary control) or secondary form (children with a period of 6 consecutive months of night-time urinary control before incontinence, which is generally associated with organic or psychological causes). Moreover, NE could be monosymptomatic (MNE) or non-monosymptomatic (NMNE), depending on the presence of daytime incontinence or any other lower urinary tract symptoms (LUTS).

We report a 7-year-old female with a history of recent onset of sphincter troubles and recurrent low urinary tract infections. She presented urinary urgency associated to daytime incontinence, bedwetting almost every night in the previous 3 months and sometimes encopresis. The physical and neurological examination was silent, no psychological or social problem intercurred. As first approach, she was treated with deamino-delta-D-arginine vasopressin (dDAVP) 120 mcg associated with oxybutynin 5 mg and educational therapy, for 3 months without benefits. So, she underwent a magnetic resonance imaging of the spinal cord, that highlighted the presence of hydrosyringomyelia from D6 to D10, lipoma of the terminal filum and the presence of synovial cyst between L5-S1.

This case remarks that in secondary NMNE, any possible organic cause must be investigated.

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ABS 39

REPORT OF A CHILD WITH SYNDROMIC INTELLECTUAL DISABILITY AND A NOVEL NONSENSE MUTATION IN *KAT6A*

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Heterozygous variants in the *KAT6A* gene have recently been described to be associated with a newly identified syndrome (MIM#616268) characterized mainly by global developmental delay, intellectual disability of variable severity, poor or absent speech, microcephaly, cardiac defects and gastrointestinal complications [1]. Since 2015, 82 patients have been described in the published literature. We report a patient with a novel autosomal dominant likely pathogenic variant in *KAT6A* gene (c.3553C>T; p.Gln1185Ter) and a clinical phenotype consistent with Arboleda-Tham syndrome, including delayed psychomotor development with independent walking at 6 years, intellectual disability, absent speech, microcephaly and growth delay, abnormal muscle tone with axial hypotonia associated with hypertonicity in the legs, early feeding problems (reflux) and constipation underlining a dysfunctional bowel motility problem and heart malformation. The cardiac defect was characterized by mitral valve prolapse with severe insufficiency (never reported before) and atrial septal defects that have required surgery intervention (mitral valvuloplasty and ASD closure), recalling the need for early cardiology assessment in these patients [2]. This report aims to contribute to further understanding phenotype/genotype correlations, which are fundamental for the interpretation of data made available by NGS panel/exome sequencing for the diagnosis of neurodevelopmental disease. Certainly, further functional study assessing the pathogenic role of this variant are required.

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ABS 40

CHILDREN IN IRAQ AFTER 40 YEARS OF WAR AND SANCTIONS

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During the late 1970's and early 1980's, Iraq was one of the most developed countries in the Middle East. The healthcare system of 172 hospitals and over 1,200 primary health centers in addition to well-regarded medical education and physician training provided excellent care to Iraq's adults and children [1]. Before the 1990s, Iraq also had the best education system in the Middle East; metrics demonstrated superiority in educational access, literacy and gender equality. Iraq provided free education to students from primary to university levels.

In September of 1980, the Iran-Iraq War began decades of destabilization for both child health and education systems. Funding was diverted from both systems to support the war efforts. The Gulf War and the debilitating economic sanctions that followed led to further decline of Iraq's healthcare and education systems. The loss of oil revenues which supported public schools and universities caused a massive shortage of learning resources. Teachers' salaries dropped to US $6 per month. Many hospitals, healthcare and school facilities were destroyed by bombings targeting civilian infrastructure.

The 2003 US-led invasion and ensuing years of war further worsened the situation. The education system was crippled by insecurity, inadequate facilities and a shortage of qualified teachers. Approximately 18,000 Iraqi doctors fled the country. With fewer doctors and educators, the prospect of children growing up healthy and educated is threatened.

Currently in Iraq, 45% of the population are children under 15 years of age; 17% are under 5 years of age. Yet, the Millennium Development Goals, as they pertain to child health, appear to have passed by Iraq. Under 5 mortality has stagnated since the 1990's; Iraq's rate is twice that of other Middle Eastern countries.

The most recent UNICEF figures on primary school enrollment show more boys (93%) are enrolled than girls (87%), with the overall total falling far short of Iraq's 2015 Millennium Development Goal target of 98% [2]. Fewer than half of children who enroll in primary education finish school. With each successive year, fewer children continue their education. Just under half of secondary school age children go to secondary school. This corresponds to an increase in children involved in hazardous child labor and early marriage.

United Nations estimates of the child mortality rate in Iraq and neighboring countries by 5-year periods, 1970-2015 are available [3].

In pediatric practice, it was common to see children with cancer coming from Kurdistan, where the Halabja chemical attack had taken place in 1988. Over the years, more children from Tikrit and areas of southern Iraq were diagnosed with cancer and congenital anomalies. It is noted that the rates of birth defects in Iraq now exceed those seen in Hiroshima.

US military intervention heavily damaged Iraqi infrastructure and ecologies that sustain child health. Military intervention in Iraq was accompanied by unprecedented waste abandonment and waste burning: discarded vehicles, excess weapons, discarded clothing and much more were all left in Iraq's land, water or air. It is no surprise that widespread cancers and congenital anomalies, along with other major health issues are present in the civilian population. The medical resources to manage children with cancers and birth defects are impacted by the enduring effects of war, the targeting of an entire population and their environment, rather than military installations.

Notably, medical education itself has been impacted. University students in the 1980's held a strong command of English; this is no longer the case. Training in many instances is inadequate. Variability in the standards of medical education have led to poorly trained practitioners who often misdiagnose patients and prescribe treatments with no evidence base.

Many Non-Governmental Organizations (NGOs) have come to help Iraq rebuild its health and education infrastructure for children. NGOs such as the Iraqi Children Foundation, War Child Iraq, and Save the Children have supported programs that help improve both health and education for Iraq's most vulnerable children.

Ongoing education for child health practitioners has been a priority. As a non-official scientific group, the IQ-PEACE (Iraqi Pediatricians Across the Continents Education) group started a series of educational online scientific activities for practicing pediatricians. These include topics of child protection and safeguarding aimed at

increasing awareness about child protection and scaling up strategies to address developmental and behavioral health in children.

Future efforts toward rebuilding could include international partnerships between academic centers for teaching and provision of pediatric subspecialty care. The infrastructure of knowledge and competency in these areas would be welcome by Iraqi pediatricians, who would develop to tools to better serve their population of children.

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ABS 41

INTRACARDIAC THROMBOSIS IN A NEWBORN WITH A HOMOZYGOUS METHYLENETETRAHYDROFOLATE REDUCTASE (MTHFR) DEFICIENCY

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INTRODUCTION

Neonatal thrombosis is a rare event, but it is increasingly recognized as a complication of contemporary neonatal care, and it contributes to neonatal morbidity and mortality.

CASE REPORT

A 34-year-old secundigravida was admitted at Emergency Department and a cesarean section was performed at 36 weeks of gestation due to a suspected fetal distress. At birth, the female neonate required ventilation support. Apgar score was 4 at the first minute and 8 at the fifth minute. Due to the persistent respiratory distress and severe skin pallor, the newborn was admitted to the Neonatal Intensive Care Unit (NICU) on oxygen support (FiO_2 0.4). The obstetric history revealed a previous pregnancy ended in a stillborn at 41 weeks of gestational age due to severe intrauterine growth restriction. Therefore, she took low-dose aspirin medication until 34 weeks of gestation and low molecular weight heparin (LMWH) until delivery. In NICU, the newborn required oxygen and respiratory support with continuous positive airway pressure (CPAP). A blood gas analysis showed severe anemia, with a hemoglobin of 5 g/dL and a hematocrit of 15.2%. An umbilical venous catheter was placed, and a transfusion of concentrated irradiated red blood cells was immediately initiated in the first hours of life. An echocardiogram revealed pulmonary hypertension and a patent ductus arteriosus with bidirectional shunt. On the 2nd day of life (DOL), CPAP respiratory support was ended, and echocardiography demonstrated merely a mild pulmonary hypertension. On the 3rd DOL, a complete blood count revealed a thrombocytopenia (82,000/mmc) and normal hemoglobin/hematocrit levels (Hb 14.1 g/dL, HCT 44.4%). On the 7th DOL, cardiac ultrasound showed a hyperechoic mass in the left atrium (diameter 0.37 x 0.65 cm). The patient was transferred to a tertiary referral hospital where a diagnosis of intracardiac thrombosis was confirmed, and anticoagulant therapy was started with LMWH. Homozygous mutation in the *MTHFR* gene was detected in thrombophilia screening. On the 12th DOL, cardiac angio-CT showed no evidence of intracavitary cardiac mass. On the 16th DOL, the infant was discharged home with a prophylactic dosage of LMWH and hematologic follow-up.

DISCUSSION

Neonatal thrombosis is a rare event that can be correlated to several risk factors such as central venous or arterial catheter, infections, prematurity, asphyxia, dehydration, liver dysfunction, maternal problems, polycythemia, major surgery, metabolic disorders, congenital heart disease, congenital nephrotic syndrome, inherited thrombophilia (antithrombin deficiency, protein C or S deficiency, factor V Leiden mutation, prothrombin G20210A, *MTHFR* mutation) [1, 2].

The overall incidence of thromboembolism in hospitalized newborn infants is approximately 2.4 per 1,000 admissions [3]. The causative role of *MTHFR* in neonatal thrombosis is unknown but it may have a contributory role in the pathogenesis of thrombosis in neonates [2].

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ABS 42

CHILD-FRIENDLY HOSPITAL ENVIRONMENT – NEW APPROACH WITH THE USE OF HOLOGRAM AND AUGMENTED REALITY

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INTRODUCTION

Hospital admission can be a scary experience for a child because of needles, intravenous lines, blood procedures and closed spaces, but several studies show that there is evidence that a child-friendly environment has a great impact in patient experience and health outcomes. "There is moreover increasing evidence that the display of visual art can have positive effects on health outcomes, including shorter length of stay in hospital, increased pain tolerance and decreased anxiety" but that works as a temporary distraction strategy: after few days the effect is reduced. In the past years we saw that Augmented Reality (AR) allow children to live, every time, a different experience. But now another technology could be even more effective. Tartu University Hospital in Estonia put together a case study proving that holographic cartoon characters can help to alleviate fear and worry in kids at hospitals. The results showed that thanks to the holographic distraction, children's pain levels were reduced by 95%. Not only did it significantly reduce the children's fear, but also increased the work efficiency of the doctors and nurses. Our goal is to determine the effect of hologram technology and AR on fear and anxiety during pediatric procedures.

MATERIALS AND METHODS

Brave Potions srl is a startup that developed a mobile application called Super Powers, that let children live a magical experience during medical treatment and hospitalization. Brave Potions srl created posters and stickers that interact with the AR app letting children see magic characters and super powers appear in front of them. We elaborated a new experience, integrating the holograms box in the waiting room. The hologram box will show the characters that have been developed by the Brave Potions srl.

General surveys, test such PANAS and others to evaluate pain, using different pain rating scales (faces, numeric, behavioral, behavioral/physiological) will be conduct.

RESULTS

Super Powers has been installed in different hospitals and ambulatories, and we are evaluating how children and families interact with the different materials and AR. In the next phase we will install also the hologram box and evaluate the difference with these two technologies. After this phase we will start to conduct our surveys in order to understand if families can use the technology without the help of the staff. Then we will start the evaluation.

CONCLUSIONS

Hospitals aim to improve the quality of care for their patient population. We expect to see that hologram and AR can provide a positive effect on health outcomes, a reduction of sedation during medical treatments, confirming the fact that improving hospital environment can have great impact on the patient health outcome.

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ABS 43

DON'T FORGET THE MILK! BREAST MILK AND GROUP B STREPTOCOCCAL INFECTION

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INTRODUCTION

GBS is one of the most frequent causative agents of sepsis in newborns. Routinary screening of pregnancies through vaginal and rectal swabs and the use of intrapartum antibiotic prophylaxis have significantly reduced the incidence of early-onset sepsis. The impact in late-onset neonatal sepsis (defined as sepsis occurring after 72 hours of age) does not appear to be affected by those measures. The transmission routes are various: neonatal colonization at birth, horizontal transmission from the mother, from other caregivers, or healthcare workers, or in a few cases through contaminated breast milk [1].

CASE REPORT

We report the case of G., a female neonate delivered at 24^{+5} weeks of gestation through the caesarian section for umbilical cord prolapse and PPROM. Apgar scores were 1 at birth, 1 at 5 minutes, and 7 after 10 minutes, her weight at birth was 570 g. Vaginal-rectal swab culture was negative for *Streptococcus agalactiae*. She was immediately admitted to the Neonatal Intensive Care Unit.

G. showed a good adaptation to extrauterine life. She was fed with pasteurized breast milk until 30 weeks, and frozen breast milk afterward. Less than 48 hours after taking frozen milk, G. showed clinical signs of sepsis, such as desaturation, alteration of skin color and seizures. The blood colture resulted positive for *Streptococcus agalactiae* and enteral nutrition was stopped.

In the same week, G.'s mother showed clinical signs of mastitis and molecular analysis performed on frozen breast milk was positive for *Streptococcus agalactiae*, instead molecular analysis carried out on pasteurized breast milk did not show any pathogens. Previous samples of frozen breast milk during the first week of life were negative.

G. was treated with intravenous antibiotic therapy and when she was better, she resumed feeding with pasteurized breast milk.

DISCUSSION

Streptococcus agalactiae is so called because it's a common causative agent of mastitis in cattle, leading to a decrease in milk production. In women, it is a less common cause of mastitis. Many studies described the presence of *Streptococcus agalactiae* in breast milk (with or without mastitis) and concomitant streptococcal late sepsis in newborns. However, in most cases, it was difficult to determine whether the source of the infection was the milk or the baby itself, with subsequent colonization of the mammary gland during breastfeeding [2].

CONCLUSIONS

Not all authors agree with recommending routine milk screening when mastitis is present. But it seems reasonable to obtain milk culture in the case of high-risk preterm infants or in infants with recurrent GBS sepsis fed with fresh or frozen breast milk. When the presence of the pathogen is documented, some authors recommend to interrupt breastfeeding, others prefer to administer pasteurized breast milk to preserve the many health benefits of breastfeeding [3].

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ABS 44

PULSE OXIMETRY SCREENING FOR CRITICAL CONGENITAL HEART DISEASE IN DUHOK CITY

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BACKGROUND

Delayed diagnosis of critical congenital heart disease in neonates increases morbidity and mortality. Pulse oximetry is a useful, inexpensive, non-invasive method for routine monitoring of the neonate, and it can also be used for raising suspicion of critical congenital heart disease.

OBJECTIVE

The aim of the study was to assess the use of routine pulse oximetry within 24 hours postpartum, and to study its feasibility as a screening test for critical congenital heart disease.

METHODS

The study was carried out in maternity hospital in Duhok city, from 7th of April, 2019 to 28th of August, 2019. Newborns between 4 to 24 hours

of age were included in the study. Pre-ductal (right hand) and post-ductal (either foot) oxygen saturation was taken, babies with low oxygen saturation less than 90%, in whom oxygen saturation was between 90-94% with 3 reading separately with 1-hour apart, and babies with more than 3% difference between either extremity were sent to echocardiography.

RESULTS

Total of 630 newborns were included in the current study, 296 (47%) of them were female and 334 (53%) were male, 389 (62%) were living in an urban area while 241 (38%) were from a rural area. During the screening process 50 cases (7.9%) with low oxygen saturation were found. All the newborn with positive screening result were sent to echo study and 10 (1.6%) of them were diagnosed with critical congenital heart disease and 7 (1.1%) had simple congenital heart disease, while 33 cases had negative echo result (other diagnosis), they were: 7 (21.2%) cases of respiratory distress syndrome, 6 (18.2%) cases of transient tachypnea of newborn, 5 (15.2%) cases of congenital pneumonia, 3 (9%) cases of diaphragmatic hernia, 3 (9%) cases of sepsis, 3 (9%) cases of pneumothorax, 4 (12.1%) cases of meconium aspiration pneumonia, 1 (3%) case of coanal atresia and 1 (3%) case of right side Erb's palsy.

CONCLUSION

Pulse oximetry screening has a significant role in detecting critical and non-critical congenital heart disease and also in detection of some non-cardiac diseases.

ABS 45

INTERACTIONS BETWEEN MICROBIOTA AND GUT-BRAIN AXIS IN CHILDREN WITH OBESITY

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Obesity represents one of the most important public health issues. Its prevalence and its complications are increasingly frequent, particularly in industrialized countries. Nowadays, obesity is widespread even among children. In fact, it affects up to 13.8% of preschool children and 19% of American adolescents [1]. The presence of obesity since childhood is accompanied by several comorbidities and consequences. Therefore, it is fundamental to understand the underlying causes, in order to develop new therapies. The microbiota and its alterations have been studied both as a possible cause and as a possible result of obesity and of its complications during childhood. Furthermore, what happens during the first years of life, including the neonatal period, is essential to define microbiota composition.

The microbiota of obese children shows significative differences compared to that of non-obese peers. More specifically, the microbiota of obese children exhibits an increased *Firmicutes*/*Bacteroidetes* ratio, abundance of *Enterobacteriaceae* and *Prevotella spp.*, and low quantities of *Desulfovibrio spp.*, *Akkermansia muciniphila*-like and *Bacteroides vulgatus*. The diversity index is reduced [2-4]. A prospective study has shown that children who maintained normal weight in adulthood, had a greater number of *Bifidobacteria* during childhood, compared to those who later became over-weight or obese. Differently, this last group showed a high number of *Staphylococcus aureus* [5].

The microbiota actively participates to the development and maintenance of weight gain, by modifying the quantity of calories absorbed by food, the energy metabolism, fat deposition and eating habits. This is caused by several mechanisms, including the production of short-chain fatty acids and other substances with endocrinological or neurotransmitter activity (GABA, serotonin, glutamate, etc.), the modification of gastro-intestinal hormones' release, the increase in gut permeability, the promotion of chronic systemic inflammation and the alteration of vagus nerve activity [6]. Microbiota also plays a key role in the proper functioning and homeostasis of the individual. As a consequence, its alteration can modify the metabolism and determine effects in different organs. Research has long focused on the possible interactions between microbiota and brain, both regarding physiological processes, such as the neurodevelopment itself, and its involvement in pathological conditions. Since the metabolic and immunological alterations of obesity act on developing structures, and this is particularly evident at the level of brain structures, this information is even more valuable talking about children. Anatomically, obese children have a reduction in brain volume, particularly

in the temporal regions and hippocampus [7]. Regarding functionality, the main features are the dysregulation of hypothalamus-pituitary-adrenal axis [8], the altered blood brain barrier permeability, microglia dysregulation and chronic low-grade inflammation [9]. All these aspects together lead to an increased risk of autistic spectrum disorder, stress-related disorders, depression, and decreased cognitive performances in obese children compared to healthy controls [10].

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ABS 46

WHEN DYSPNEA IS NOT JUST A SYMPTOM

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INTRODUCTION

In pediatrics, respiratory pathologies represent the second cause, after traumatic events, of access to the Emergency Room. Futhermore, half of the children hospitalized in Pediatric Intensive Care Unit present a diagnosis of respiratory failure [1]. We talk about respiratory failure when lungs fail to provide appropriate respiratory gas exchanges. The time of onset allows distinguishing between acute, chronic and acute exacerbation of chronic respiratory failure. Respiratory failure is more common among pediatric population than in adulthood, in particular during the first year of life. The right evaluation of the child, combined with the cause assessment, is essential for the therapy, which aims at correcting hypoxemia or supporting child's ventilation to improve respiratory dynamics [2].

CASE REPORT

We present the case of a 7-year-old boy who suffers from frequent bronchospasm (about 10 attacks a month) and who has already recovered from bronchial pneumonia once in the past. He came for the first time to our Emergency Room in June 2022, with a yellow code of priority, presenting dry cough and worsening dyspnea. The initial oxygen saturation was 90% with 10 L/min O_2, heart rate 170 bpm and respiratory rate 60 bpm. He was pale and presented intercostal, subxiphoid and jugular retractions. On auscultation, wheezes were diffused on the whole thoracic wall. We immediately started aerosol therapy with physiological solution and beta-2 agonists. The chest X-ray showed bilateral thickening of bronchial walls, especially in superior hilar horns. After that, we started oxygen therapy at 10 L/min and his oxygen saturation started improving (from 90% to 93%) and respiratory rate passed from 60 to 45 bpm. Then we combined muscarinic antagonists with beta-2 agonists so that oxygen saturation reached 97% with 6 L/min of oxygen and later, when we arranged his admission to Pediatric Intensive Care Unit, with 3 L/min.

His second access to our Emergency Room was clearly worse than the previous one. This time at the triage it was assigned a red code, considering the patient's critical conditions: continuous cough attacks, significant and diffused inspiratory retractions, important skin paleness, irritability, and apparently absence response to beta-2 agonists and muscarinic antagonists. No breath sounds could be heard on chest auscultation. It

was immediately started aerosol therapy with beta-2 agonists at a higher dosage and then with adrenalin, on the suspect of laryngitis. Air penetration improved and on auscultation we could hear coarse crackles and wheezes, especially on the right side of the thoracic wall. Considering the persistence of dyspnea, we administered intravenous corticosteroid and the same dosage of beta-2 agonists for aerosol. He finally started breathing better and the cough stopped. After the anesthetist resuscitators' consulting, we administered an intravenous bolus of beta-2 agonists. He finally underwent aerosol therapy with beta-2 agonists associated with muscarinic antagonists but, even though the air penetration on the left side was sufficient, his admission to Pediatric Intensive Care Unit was arranged.

CONCLUSIONS

In this abstract we showed how important is the prompt evaluation and treatment of respiratory failure. Clinicians have several useful tools to face and manage pediatric acute and acute exacerbation of chronic respiratory failure. One of the main objectives of our future research is the study of more effective and less invasive procedures, the standardization of treatments actually in use and, last but not least, the improvement of the collaboration between family pediatricians and the emergency team.

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ABS 47

THE ENDOSCOPIC AND HISTOLOGICAL FEATURES OF CHILDREN WITH *HELICOBACTER PYLORI* INFECTION

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INTRODUCTION

Helicobacter pylori (*H. pylori*) is acquired in childhood and plays a pivotal role in the development of various gastric and extra-gastric disorders.

AIM AND METHOD

The aim of this study was to analyze the endoscopic and histopathologic changes of the gastric mucosa in *H. pylori* infected children. We performed a prospective study of 132 children (81 girls, age range 6-18 months) with abdominal complaints who underwent esophagogastroduodenoscopy in our clinic between January and December 2019. Clinical symptoms and socioeconomic conditions were analyzed.

H. pylori infection was confirmed by at least 2 standard invasive tests: urease rapid test and histological examination. The endoscopic findings were classified according to the present criteria: normal appearance, hyperemia, edema, erosions, nodular and paving stone aspect. For each patient 2 biopsy specimens were obtained from antrum and corpus. The histological diagnosis of gastritis was based on the updated Sydney system as: inflammation, neutrophil activity, atrophy, *H. pylori* colonization and metaplasia. The endoscopic appearance was evaluated by the same physician and the histologic examination was made by one pathologist.

All statistical analysis was performed with SPSS® version 20.0. A p-value less than 0.05 was considered as statistically significant.

RESULTS

H. pylori infection was identified in 79 of 132 children (59.85%). The rate of *H. pylori* colonization was inversely correlated with the socioeconomic status ($p < 0.005$) and directly correlated with age ($p < 0.002$). All of the children had endoscopic appearance of gastritis, predominantly non-atrophic antral gastritis (76 of 132 children, 57.58%), followed by non-atrophic pangastritis (54 of 132 children, 40.91%) and lastly non-atrophic fundic gastritis (2 of 132 children, 1.52%).

In the *H. pylori* infected children, the histologic findings were distributed as following: netrophil activity in all of 79 cases (100%), antral inflammation in 49 cases (62.03%), intestinal metaplasia in 0 cases and mucosal atrophy in 3 cases (3.80%). Endoscopic nodular gastritis was observed in 57 of 79 infected children (72.15%) compared with 22 of uninfected ones (41.51%).

Endoscopic nodular gastritis was significantly associated with increased activity and chronic inflammation observed during histologic examination ($p < 0.001$).

CONCLUSION

This endoscopic series reveals that the rate of *H. pylori* infection (59.85%) is still a significative problem in developing countries. Our study confirms the "gold standard" role of the endoscopic examination of children with dyspeptic symptoms suggestive for an organic etiology. Endoscopic nodular gastritis was strongly associated with *H. pylori* infection (72.15%) and with higher grades of gastritis severity ($p < 0.001$).

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ABS 48

NATAL TOOTH IN A 7-DAY-OLD CHILD: MANAGEMENT AND POSSIBLE ASSOCIATION WITH SYNDROMIC PICTURES

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INTRODUCTION

Natal teeth are dental elements present at the time of birth, while if teeth erupt within 30 days of birth they are called neonatal teeth. The first ones erupt more commonly in the jaw, especially in the anterior region (85%). Incidence of this type of condition ranges from 1:1,000 to 1:30,000 newborns. The ratio of natal to neonatal teeth is 3:1. Their presence can lead to some problems such as: insufficient feeding to the infant, injury to the mother while feeding, and development of ulcers on tongue near these teeth. Their management often involves extraction, even during the very first days of life. Syndromic pictures are often associated with the presence of these particular teeth. The aim of this work is to report a case of natal tooth with presence of a not traumatic tongue lesion.

CASE PRESENTATION

A small 7-day-old patient came to our attention due to the presence of a small white hard mass on palpation on the anterior inferior alveolar ridge. His twin brother showed no oral alterations. At an objective examination it was possible to appreciate the presence of a small dental element, which fortunately had not yet created traumatic lesions in the adjacent tissues. The patient came from the Neonatal Intensive Care Unit of the University Hospital of Cagliari. It was also possible to notice the presence of a thick white lesion present on the whole lingual dorsum. During the first visit, it was not possible to understand whether the diffused white area was removable or not. A tooth extraction operation, to be performed in a protected hospital setting, was scheduled after a few days to allow the baby to feed properly. The 1 week and 1 month follow-up showed excellent tissue healing. The white patina at the lingual level had been removed with a small gauze almost completely on the day of the extraction, confirming a diagnosis of tongue localized mycotic infection. The patient had undergone topical antifungal therapy.

DISCUSSION

The presence of natal and neonatal dental elements has been associated with various syndromic pictures, even quite rare, such as Riga-Fede disease, where the dental elements create trauma on soft tissues, especially lingual ones, Van der Woude syndrome and congenital pachyonichia. This last comprises a group of autosomal dominantly inherited conditions showing several signs such as nail thickening, palmoplantar keratoderma, follicular keratoses, and mucosal leukokeratosis, overall in the dorsum of the tongue. For this reason, the diagnostic hypothesis of an initial syndromic picture of pachyonichia in the patient described was taken into consideration. Currently there are no other elements that would suspect a syndromic picture; the patient will be monitored with periodic visits.

CONCLUSION

Recognizing syndromic pictures in extremely early times is one of the goals of the pediatric oral pathologist and the documentation of rare cases can be of great help for other clinicians.

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ABS 49

PLASTIC AND MICROPLASTIC IN PLACENTA

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The influence that the environment has on gestation is without a doubt underestimated. Despite being equipped with great intelligence, mankind possesses some significant cognitive limits that don't allow it to evaluate events correctly and to relate them to the distant future.

It is challenging to measure human intelligence, as it is subject to numerous interdependent variables, some of which are:

- good nutrition;
- regular schooling and school quality;
- fortification of certain food products;
- laws establishing safe levels of pollutants, like lead;
- musical training in childhood;
- socioeconomic status;
- incidence of infectious diseases.

In the last century, the global production of plastic has grown exponentially, reaching over 350 million tons per year produced all over the world. This greatly contributes to environmental pollution.

Recently our group published a study that demonstrates the presence of microplastics (MPs) in intracellular compartments of human placentas in photos for the first time. Moreover, we were able to localize the MPs and to prove that important morphological and structural alterations of the cellular intracytoplasmic organelles were associated with their presence.

Using variable pressure scanning electron microscopy (SEM) and transmission electron microscopy (TEM), we detected MPs within lipid membranes. They can be easily confused with cell organelles such as lysosomes, peroxisomes, lipid droplets and multivesicular bodies. These have never been seen before even though there are many specialists who look at placentas, but between looking and seeing there is an important difference. This amply confirms Goethe's (1749-1832) acknowledgement: "We only see what we know".

In all the observed samples, the stress of the endoplasmic reticulum is evident, seen as being dilated (cribriform aspect of the syncytium trophoblast cells); there are many vesicles discreetly electrodense, with secretory material inside, covered by ribosomes and not (degranulation) communicating with each other.

Intracytoplasmic organelles alterations, together with MPs demonstration in all the samples examined, are a very important item, since endoplasmic reticulum stress and mitochondrial dysfunction could play a decisive role in human non-transmissible disease (NTD) progression. This gives rise to the hypothesis that plastic environmental pollution is partially responsible for the epidemic of NTDs that characterizes the modern world.

Of great concern is the suspicion that exposure to MPs during critical periods when adipocytes are differentiating and pancreas, liver, brain are developing can induce effects that could manifest themselves later in life, often as full-blown disease.

Developing organisms are sensitive to the presence of foreign substances such as plastics in general or MPs. These interferents, like other polluting objects, can lead to abnormal gene expression in tissues, in terms of number of cells, position and imbalance between the cell types, as well as impaired organ structure and incorrect hormone signaling, thus leading to an increased susceptibility to disease/dysfunction throughout adult life.

There is ample evidence to support the fact that many chronic diseases, including obesity, diabetes, and metabolic syndromes, may be linked to epigenetic changes in cells and tissues during intrauterine development.

Currently, we are studying placentas from normal pregnancies to better understand the functional consequences of MPs detection in developing human beings, especially in relation to the current evolution of the global burden of disease, with increasingly high rates of NTDs.

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ABS 50

WHEN THE SPLEEN IS MISSING: A RARE CASE OF NEONATAL DIAGNOSIS OF ISOLATED CONGENITAL ASPLENIA

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INTRODUCTION

Congenital asplenia is a rare malformation that can occur as isolated (isolated congenital asplenia, ICA) or in association with other complex visceral defects, in the setting of the Ivemark syndrome (asplenia/ polysplenia, congenital heart disease, situs viscerum inversus and abnormal lung lobation).

The incidence of ICA is approximately 0.51 per million new births per year. This finding is potentially underestimated as it may go undiagnosed in asymptomatic people [1].

Between 1956 and 2016, 73 cases of congenital asplenia were described: 32 cases were sporadic and 41 were familial forms. In the latter, ICA has been related to autosomal dominant (by haploinsufficiency) mutations of the *RPSA* gene that encodes ribosomal protein SA, a component of the small subunit of the ribosome, that has an essential role in human spleen development [2].

ICA is a life-threatening condition during the first 2 years of life. Later in life, the adaptive immune system seems to provide some protection. In 78% of cases, the first manifestation of ICA is severe bacterial sepsis with a mortality rate of 48% [3].

Imaging studies (ultrasound, CT, MRI) highlight the absence of the spleen. The diagnosis is then confirmed by detecting Howell-Jolly bodies (intra erythrocyte residues usually removed by the spleen) on blood smears. Monocytosis, lymphocytosis and/or thrombocytosis can also be found.

Treatment is based on preventing infections: timely vaccinations (especially pneumococcal and meningococcal vaccines), annual influenza vaccine, antibiotic prophylaxis and education about risk situations and other protective measures. Essential is early medical consultation in case of febrile episodes in order to begin an empiric broad-spectrum antimicrobial therapy before obtaining the result of blood cultures [4].

CASE REPORT

A girl was born at 31 weeks + 6 days of gestational age (birth weight: 1,970 g) by parents of Maghreb origin. She was admitted to the Neonatal Intensive Care Unit for respiratory distress and prematurity.

On the second day of life, she developed clinical signs of sepsis. Blood tests showed an elevated procalcitonin level (49.9 ng/mL, normal values < 2 ng/dL) for this reason she was treated with intravenous antibiotics for 6 days.

On the first day of life, white blood cell count was 16,880/μL, hematocrit was 37.4% and platelet count was 253,000/μL. The platelet count increased over the next weeks to 828,000/μL and, on day 36 of life, was 923,000/μL.

Ultrasonography (US) of the abdomen performed at 23 days after birth revealed a small spleen, with splenic length of 10 mm, width of 5 mm, and depth of 5 mm. Follow-up US at 32 days of life confirmed the absence of a normal-sized spleen.

A prophylactic course of antibiotics with an oral cephalosporin was started on day 31 of life.

On day 36, the peripheral blood smear showed RBCs containing Howell-Jolly bodies, a finding compatible with splenic dysfunction.

CONCLUSIONS

Newborns with ICA, without antibiotic prophylaxis, vaccination, and patient and family education, are prone to developing severe, recurrent infections. Despite being a rare condition, it is important to try to make a diagnosis of ICA as early as possible, thus avoiding an onset characterized by severe sepsis or clinical picture resulting from significant thrombocytosis.

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ABS 51

ADENOTONSILLECTOMY AND OBESITY DEVELOPMENT: A PEDIATRIC CASE CONTROL STUDY

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INTRODUCTION

Childhood obesity is one of the most important problems in our society and it has been recognized as a global pandemic [1]. It has a multifactorial etiology and it is influenced by environment, socio-economic status, diet and lifestyle [2]. Adenoidectomy and tonsillectomy are among the most frequent pediatric surgeries, mainly due to recurrent tonsillitis and sleep apnea syndrome [3]. Primary aim of our retrospective study is to evaluate the possible relationship between tonsillectomy and/or adenoidectomy and the subsequent development of childhood obesity. Secondary aims are to evaluate sex distribution of obesity between case and control groups and the possible effects of control group's therapy (L-thyroxin) in the development of children obesity.

METHODS

Our retrospective case control study involves 258 children aged 3-17 years followed from 2016 to 2022 at the Endocrinology Center of F. Del Ponte Hospital, Varese, Italy.

The case group comprised 158 children (98 male, 60 female) with essential obesity (BMI equal or greater than the 97th percentile, according to WHO criteria of 2006-2007) and no evidence of obesity complications (such as hypertension, hepatic steatosis and insulin resistance) at the first endocrinology visit (T0). The control group includes 100 non obese children (37 male, 63 female) with subclinical hypothyroidism or L-thyroxin treated and controlled hypothyroidism (4 patients): TSH > 4.5 mIU/L, FT4 in range: 8-17 pg/mL. We collect anthropometric parameters at T0 such as height, weight, BMI. We subsequently research the past surgical exposure (46 case subjects and 7 control patients, from 2004 to 2019) with pre- and post-surgical anthropometric measures. We reported anthropometric parameters on Cacciari or WHO growth charts depending on age and ethnicity.

RESULTS

Means of height, weight and BMI are higher in the case group (n = 158) than the control one (n = 100).

Previous otolaryngologist surgery results to be more frequent in the case group than the control one: n = 46 (29%) vs n = 7 (7%), with OR 5.5 and p-value < 0.0001.

The most frequent surgery in our case study is adenotonsillectomy in both groups: n = 27/46 (59%) for the case group vs n = 4/7 (57%) for the control one. The surgery age mean is equal in both groups: 5 years old (SD 1.8 for the case group vs 1.0 for the control group). All 46 case children register a BMI increase after a mean time range of 3.5 years post-surgery (range: 6 months-10 years) while no one of the control children develops subsequent obesity.

Male sex is statistically more affected in the case group than the control one (62% vs 37%, p-value < 0.001). The onset of obesity occurs later for the case group (mean value: 10.9 ± 3 years vs 9.5 ± 4.3 years, p-value = 0.02), but the age of the first endocrinological visit is higher in the case group (aged 6-10 years: 96% vs 73%).

CONCLUSION

Our study has tried to research one of the possible contributors to children obesity development such as adenoidectomy and/or tonsillectomy, which are frequent pediatric surgeries. The surgical exposure seems to be a risk factor for obesity development. Actually, the weight gain

might be related to improvement in lifestyle and in olfactory-taste perception. Based on these results, one future prospective might be to establish a more careful follow-up of the patients undergoing otolaryngologist surgery. According to the literature, male seems to be the sex more affected due to environment exposure, diet, lifestyle and socio-economic status [4]. Finally, appropriate L-thyroxin therapy seems to be a protective factor against obesity by controlling thyroid function.

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ABS 52

HEMORRHAGIC STROKE CAUSED BY ARTERIOVENOUS MALFORMATIONS (AVMS) IN PEDIATRICS

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Brain arteriovenous malformations (bAVMs) are weak tangles of arteries and veins that don't have capillaries connecting them together and can break and bleed, causing hemorrhagic stroke. This event is quite rare in the pediatric population. Despite this, the rate of this vascular rupture seems higher in children than in adults, perhaps because the majority of bAVMs in pediatrics are detected almost only when they lead to a symptomatology, and so after breaking.

M. is a 10-year-old male taken to the Emergency Unit for presenting unstoppable vomit and a syncopal episode that lasted 5 minutes while he was doing a workout. When he arrived at the Hospital, he presented GSC 11 and responded only to painful stimulus. He had shaking chills and was drowsy and hyporeactive. We started intravenous infusion of antiemetic. Then, we began to see heart rate variability from 47 to 90 bpm. We carried out a CT scan that showed a left anterior intraparenchymal hematoma with outlet on ipsilateral anterior frontal cone and causing a tetra-ventricular flooding with a consequent shift to the right of anterior median structures and imprint on the homologous ventricular horn. So, we did immediately a CT angiography, which revealed a 32 x 13 mm bAVM fed by branches of the callosomarginal, anterior cerebral and pericallosal artery of the left side, and some venous and arterial aneurysmal dilations. The patient was then admitted to the Department of Neurosurgery to undergo a craniotomy for the evacuation of the hematoma and to place a ventricular catheter. It resulted in the re-expansion of contiguous tissues. The bAVM detected was treated with endovascular embolization that led to the exclusion of the vascular malformation from the bloodstream and permitted to maintain the patency of the left anterior blood circulation. 19 days later the NMR showed an hypoperfusion of the anterior and basal frontal lobe in the left side.

After 13 months, the CT scan highlighted outcomes of embolization with adjacent malacia and consequent modest ex-vacuo enlargement of the homologous frontal horn. The MR angiography showed, in addiction, thin area of peripheral gliosis and a vein of mesial cortical discharge which from the nidus reached the anterior portion of the superior sagittal sinus.

Hemorrhagic stroke is a possible event in pediatric patients and it's often due to the presence of a bAVM, which becomes evident when it breaks. Early detection of bAVMs is very important to start early treatment and to avoid serious cerebral sequelae.

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ABS 53

WEANING FROM INSULIN TREATMENT TO ORAL SULPHONYLUREAS IN A 4-MONTH-OLD INFANT WITH NEONATAL DIABETES

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INTRODUCTION

Neonatal diabetes mellitus (NDM) onset is within 6 months of life. Mutations involving ATP-dependent K channel (i.e., *KCNJ11*) can be treated with sulphonylureas (SU). This channel is expressed also on brain cells, hence the mutation affects the neurological outcome.

To date, there is no agreement on the NDM insulin regimen nor guidelines on the shifting from insulin to SU [1]. SU receptors are spread in the brain, hence SU can have positive effects on neurodevelopment, while insulin cannot [2].

CASE REPORT

C. had a normal neonatal course and weight deceleration at 3 months.

At 4 months of age, C. started vomiting and had fever (38°C), with increased diuresis and hypotonia and polypnea. Thus, he was referred to the Emergency Department.

Blood gas showed metabolic acidosis (pH 6.968) and hyperglycemia (755 mg/dL), with ketonuria and glycosuria.

After stabilization, he still had marked hypotonia, particularly regarding head control.

An insulin pump (CSII) with continuous glucose monitoring (CGM) sensor was positioned. CSII was set to manual mode with low glucose suspension (LGS) and predictive LGS (PLGS).

Monogenic NDM was suspected.

Basal insulin was set to 0.025 U/h and a 0.025 U bolus was administered when blood glucose was higher or predicted to raise > 250 mg/dL.

Insulin was always administered after feeding. Insulin requirement slowly increased and basal insulin was set to 0.1 U/h.

Glibenclamide treatment was later started at the dose of 0.2 mg/kg, while insulin treatment was administered at 0.025 U/h, but soon increased (0.05 U/h) because of hyperglycemia. SU increase was 0.2 mg/kg daily until reaching the full dose of 1 mg/kg, divided into 4 administrations. When it was 0.8 mg/kg, insulin was decreased to 0.025 U/h and then discontinued at 1 mg/kg.

The daily dose was later split into 6 per day. At this dosage, the optimal time in range (TIR) was obtained and C. was dismissed.

The genetic test showed a heterozygous *KCNJ11* mutation (Val59Met).

DISCUSSION

Our treatment aimed to avoid hypoglycemia, and the initial dose was started according to the minimum viable dose (0.025 U). Then, insulin was gradually increased in order to obtain a better TIR.

When SU was available, basal insulin was set back to the minimum to avoid hypoglycemia. Then, it was soon increased to 0.05 U/h due to frequent hyperglycemia and then decreased accordingly to SU increase. Hypoglycemia often occurred.

Once the full dose was reached and insulin discontinued, the child still experienced hypoglycemia soon after SU administrations, with rebound hyperglycemia due to glucose infusions. Since SU triggers insulin secretion, the dosage was split into more administrations, keeping the regimen dose of 1 mg/kg. This division led to better control. SU also gradually improved the infant tone.

A few cases in the literature described such a young baby affected by NDM shifting from insulin to SU, which is the optimal choice also to obtain a better neurological outcome.

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ABS 54

A DIAGNOSIS THAT ANY PEDIATRICIAN SHOULD NOT MISS: A CASE REPORT OF HEMATOCOLPOS

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CASE REPORT

A 12-year-old female teenager presented to our Pediatric Emergency Medicine Department because of pain and median swelling of the lower quadrants of the abdomen. She reported that the

pain raised 20 days earlier and was on and off, not radiated. No history of any other disease was known. Primary amenorrhea. On physical examination, the hypogastrium was swollen, of a tense-elastic consistency, but not painful. The abdominal ultrasound showed a large ovular, hypoechogenic formation with a maximum diameter of 160 mm, and hypoechoic content. This formation dislocated and limited the distension of the bladder. The uterus was enlarged, and was also displaced antero-superiorly. The endometrial cavity was dilated as well, with evidence of initial hematometra. There was evidence of bilateral dilatation of bilateral calico-pyelic cavities, likely secondary to compression of the distal tract of the ureters. No stones were seen.

Patient was admitted. Hymenotomy and surgical drainage of the hematoma was performed, and she was discharged 2 days after the operation with estradiol gel as home therapy.

DEFINITION

Abdominal and pelvic pain is common in adolescent age group. Differential diagnosis could be gynecological, gastro-intestinal, renal, and psychosomatic causes. We reported a case of hematocolpos and hematometra, defined as the collection of blood in the vagina and uterine cavity, respectively. These conditions can occur at the time of the first menstruation when there is a congenital imperforation of the hymen or later in life secondary to acquired conditions such as stenosis or scarring of the neck and uterine ostium [1].

EPIDEMIOLOGY

Imperforate hymen is a rare condition with an incidence of 1:2,000; however, it represents one of the most common obstructive lesions of the female genital tract [2]. This is a sporadic condition, although both dominant and recessive inheritance have been described in some families [3].

CLINICAL APPROCH

In the case we present, the female teenager reported on and off abdominal and pelvic pain, which occurs in 60% of cases. Imperforated hymen may also be associated with an increase of the uterus volume, in absence of menstrual flow [4, 5]. The marked distension of the vagina can cause pain in the sacral region (38-40%), due to compression of the sacral plexus, constipation, fecal incontinence, or difficulty in urination [6-11]. Physical examination usually shows the palpable mass in the hypogastrium. Exploration of the perineum reveals the swollen and bluish hymenal membrane. Imperforated hymen is not considered an emergency unless the patient is unable to urinate or is febrile with obstructed infected blood [6, 8].

DIAGNOSIS

Abdominal ultrasound is the gold standard for the diagnosis of hematocolpos and hematometra. It allows to differentiate the different types of obstructive utero-vaginal anomalies [12, 13]. The typical ultrasound appearance, in longitudinal and transverse supra-pubic view, is characterized by a cystic mass posterior to the compressed and displaced uterus [6]. The hypoechoic appearance of the mass is given by its corpuscular nature. The magnetic resonance of the pelvis can help in better delineate the pelvic anatomy, hence it is useful to highlight other congenital malformations of the reproductive system prior to surgery [4, 11].

THERAPY

The management is surgical. It consists of a vertical or cross incision of the hymenal membrane under total or partial anesthesia followed by evacuation of the blood material [5, 9, 10]. Usually the results of the intervention are favorable and relapses are rare [14, 15].

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ABS 55

SLEEP DISORDERED BREATHING AND QUALITY OF LIFE IN PAEDIATRIC PATIENTS UNDERGOING TONSIL AND/OR ADENOID REMOVAL: RELATIONSHIP WITH PRE-EXISTING FACTORS

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INTRODUCTION

Obstructive sleep apnea syndrome (OSAS) is a respiratory disorder that occurs during sleep characterized by episodes of partial or total disruption of the upper airways and accompanied by signs or symptoms. It affects about 2% of the pediatric population in the age of 2-8 years. The main cause of OSAS is adenotonsillary hypertrophy. Other risk factors include obesity, allergic rhinitis, craniofacial abnormalities, genetic syndromes, inflammation and perinatal factors. Children with OSAS may develop metabolic, cardiovascular, and neurobehavioral issues. The gold standard instrumental examination for diagnosis is nocturnal polysomnography. Clinical history, objective examination and screening tools are also important for diagnostic framing. The main treatment is represented by the intervention of adenotonsillectomy or adenoidectomy alone.

AIMS

I. To search possible risk factors of persistence of the disorder evaluated through positivity to the Pediatric Sleep Questionnaire (PSQ) after the surgery;
II. to assess the difference in quality of life in patients with positive PSQ compared to the group with negative PSQ.

MATERIALS AND METHODS

Included in the study were 100 children aged 2-11 who underwent adenoidectomy or adenotonsillectomy surgery between November 2018 and December 2021. The data were collected through the consultation of the medical records kept in the Department of ENT or Pediatrics and through a telephone interview with the parents of the patients in the study. The information obtained from the medical records concerns the pre-intervention characteristics (anthropometric parameters, objective ENT examination, execution of instrumental examinations) and the characteristics of the intervention (diagnostic indication, type of intervention, complications). Through the telephone interview, anamnestic information (comorbidities, anthropometric parameters) and data concerning the perinatal period were collected. In addition, 2 questionnaires were proposed: the PSQ, 22 items that ask about snoring frequency, loud snoring, observed apneas, difficulty breathing during sleep, day-time sleepiness, inattentive or hyperactive behaviour, and other paediatric OSAS features, and a part of the Pediatric Quality of Life questionnaire (PedsQL) to assess physical, emotional, social, and school functioning.

RESULTS

No variable analyzed found p-value < 0.05. Despite this, the age at the call ($p = 0.07$) and the birth weight ($p = 0.06$) obtained values close to the statistical significance. The multivariate analysis showed the BMI z-score at the intervention (OR = 7.94) as a possible predisposing factor. In patients with persistent respiratory disorders, PedsQL results show lower values than the control group (0.81/0.68 and 0.92/0.86, respectively).

CONCLUSIONS

No variable could be determined in this study as a risk factor for persistence of respiratory sleep disorder after surgery. The study confirmed the role of respiratory disorders in reducing quality of life.

ABS 56

LONGITUDINAL IMPACT OF OBSTRUCTIVE SLEEP APNEA SYNDROME ON THE NEURO-BEHAVIORAL PROFILE IN CHILDREN

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SUMMARY

Sleep breathing disorders (SBD) are a spectrum of sleep-related breathing disorders characterized by obstructive sleep apnea, snoring, and noisy breathing. The most severe form is obstructive sleep apnea syndrome (OSAS), characterized by recurrent episodes of upper airway obstruction and intermittent desaturations. The estimated incidence of OSAS is 1-3% in preschool and school age children. It is known that the disease has important cardiovascular repercussions secondary to the production of free radicals and systemic inflammation. In addition, fragmentation, deprivation and reduction of sleep quality also have important repercussions in daytime activities, according to changed behavioral patterns. On long term those also affect cognitive skills, with altered attention and academic performance. In SBD, contrary to what happens in real neuropsychiatric pathologies of the developmental age, with the resolution of the respiratory obstruction these negative repercussions could be reversible, with consequential improvement in academic and learning performance.

AIMS

The goal of this study is to investigate psychosocial functioning and different behavioral patterns in a group of children with SBD.

SUBJECTS AND METHODS

A group of preschool and school age children with varying degrees of OSAS severity were enrolled. The group was evaluated with nocturnal respiratory polygraph study at the time of enrolment. At the first clinical follow-up, 3 questionnaires were administered for each subject: the Pediatric Sleep Questionnaire in its 22-item reduced form (PSQ-SRBD), focused on sleep quality and quantity; the Pediatric Quality of Life questionnaire (PedsQL 4.0), focused on daily activities, wellbeing and cognitive-learning aspects; and the Conners' questionnaire in short form for parents (CPRS-R:S), which is proved by many authors to be a sensible instrument to easily detect the different behavioral patterns.

RESULTS

Of the children who had a previous diagnosis of OSAS, those in which there has not been a regression of the SBD have a worse quality of life, partially due to poorer academical and learning performance and partially due to ADHD-like behavioral patterns. Of the children who were SBD negative on the polygraphic study, those who developed SBD have a worse quality of life and poorer physical performances, without showing altered scoring at CPRS-R:S. Of the patients who had operation of adenoids and/or tonsils, only 52.4% had a regression of the SBD. At follow-up, patients who have had a non-resolving surgery have a worse quality of life, worse academical and physical performance, greater risk of daily-life repercussions due to ADHD-like behaviours.

CONCLUSIONS

SBD cause a worsening in neurobehavioral profiles and quality of children life. This can also cause some neurocognitive alterations. Surgical intervention determines an improvement in many of these areas, but it causes regression of the disease only in half of the patients.

In future studies it would be interesting to analyse neurocognitive functioning in children with SBD.

ABS 57

IDIOPATHIC PULMONARY ARTERIAL HYPER-TENSION IN CHILDREN AND ADOLESCENTS IS STILL A SERIOUS CHALLENGE

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INTRODUCTION

Paediatric pulmonary hypertension (PH) is a miscellaneous disease which causes significant morbidity and mortality if left untreated [1]. PH in paediatric age is defined as in adulthood, i.e. as the combination of mean pulmonary artery pressure (mPAP) at rest ≥ 20 mmHg, pulmonary

arterial wedge pressure (PAWP) < 15 mmHg, and pulmonary vascular resistance indexed (PVRI) > 3 Wood units × m^2 in subjects with biventricular physiology undergoing right heart catheterisation (RHC) [2]. This definition does not take into account the presence or absence of pulmonary vascular disease (pulmonary arterial hypertension, PAH) and does not differentiate between precapillary (PAWP ≤ 15 mmHg) and postcapillary (PAWP > 15 mmHg) PH forms. Systemic blood pressure in paediatric age also varies a lot with age and height in comparison to adults. As such, pulmonary-to-systemic pressure ratio > 0.4 is considered suggestive of PAH in children as well [3].

Idiopathic pulmonary arterial hypertension (IPAH) is a very rare subtype of PAH in paediatric age. It belongs to group 1 PH along with PH caused by congenital heart disease (CHD). IPAH can manifest in the neonatal life or infancy. It is often diagnosed and treated as persistent pulmonary hypertension of the newborn (PPHN). For this reason, when PAH persists beyond the first few weeks of life, early presentation of IPAH should be suspected. In these young patients, lung histopathology is very similar to the changes seen in IPAH at an older age.

Limited data are available on long-term outcomes for IPAH children. The study aims at evaluating the clinical, echocardiographic, RHC features and outcome in a small cohort of IPAH children and adolescents in comparison to a control group of peers with PAH caused by CHD.

METHODS

Twelve IPAH children and adolescents were compared with 12 CHD counterparts affected by PAH caused by CHD concerning clinical, echocardiographic, RHC features and outcome.

RESULTS

IPAH patients were younger than PAH CHD subjects ($p < 0.001$). There was no difference in terms of saturation on room air (p = ns). On echocardiography, right ventricular contractility, expressed in form of tricuspid annular plane systolic excursion, was lower and right ventricular systolic pressure higher in IPAH group ($p < 0.001$ and $p < 0.001$, respectively). On RHC, mean arterial pressure and pulmonary vascular resistance were higher (both $p < 0.001$) in IPAH. Lastly, concerning the outcome, there were 4 deaths in the IPAH group and none in the PAH CHD individuals ($p < 0.0001$). Eight IPAH subjects have undergone double lung or heart-lung transplant and none in the PAH CHD group ($p < 0.00001$).

CONCLUSION

IPAH is an aggressive and rapid disease, leading to cardiac and respiratory impairment as testified on both echocardiography and RHC. Though medications (phosphodiesterase-5 inhibitors, dual endothelin receptor antagonist, and prostanoids) given in combination therapy can slightly slow down the disease progression, IPAH outcome is still transplant or death.

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ABS 58

"WHEN IS IT POSSIBLE TO TRY AGAIN?" HOW TO MANAGE THE REINTRODUCTION OF EGG IN THE DIET OF INFANTS WITH ACUTE FPIES

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BACKGROUND

Acute food protein-induced enterocolitis syndrome (FPIES) is a non-IgE-mediated food allergy, characterized by episodes of prolonged and repeated vomiting, onset is typical of childhood. Symptoms generally occur from 1 to 4 hours after ingestion of the offending food: vomiting is usually associated with pallor and lethargy and diarrhea can follow within 24 hours, more often in 5-10 hours. These clinical manifestations define mild-to-moderate acute FPIES, but a severe subtype can also occur with marked dehydration which leads to hypotension and metabolic acidosis until shock. The diagnosis is based on the International Consensus

Guidelines of 2017, which require that the patient presented both vomiting in the 1- to 4-hour period after ingestion of the offending food with the absence of classic IgE-mediated allergic skin or respiratory symptoms (major criterion) and at least 3 manifestations among: a second (or more) episode of repetitive vomiting after eating the same food, repetitive vomiting episode 1-4 hours after eating a different food, extreme lethargy, marked pallor, need for Emergency Department visit, need for intravenous fluid support, diarrhea within 24 hours (usually 5-10 hours), hypotension or hypothermia (minor criteria). Acute FPIES with mild-to moderate symptoms is usually treated by the administration of ondansetron and oral rehydration, while the severe subtype could require fluid resuscitation in the Emergency Room. The primary management of FPIES is recommending dietary elimination of the offending food, until tolerance is achieved. Timing depends on the subtype of FPIES and age of the patient. The tolerance acquisition can be tested by performing an oral food challenge (OFC) with the administration of the offending food in a healthcare environment.

Egg is one of the most reported solid food causing acute FPIES: in Italy 6% of cases are due to egg intake, in the third place after fish and rice. Although studies performed in different countries obtained discordant results about the age of tolerance acquisition in egg (USA: 42 months; Spain and Australia: 63 months; Italy: 53 months), a recent Italian study hypothesized that the mean age may change depending on whether and how the trigger food is cooked, anticipating the mean age to 30 months with hard-boiled egg followed by the acquisition of tolerance in raw egg at the mean age of 42 months.

CASE PRESENTATION

We describe the case of a girl that was referred to our Allergy Unit when she was 15 months old in 2019. Since she started to introduce egg in her diet at 10 months of age, she had experienced 3 episodes of prolonged and repeated vomiting followed by diarrhea after 2 hours from the ingestion of the offending food (the first 2 times after ingestion of egg yolk and the latter after the intake of a whole egg). On these occasions she never presented lethargy and she never required fluid administration in the Emergency Room. After 1 month from the last episode, she performed testing for food sIgE, which resulted negative for egg yolk and white. At 13 months of age, a first attempt of OFC with hard-boiled egg (i.e., boiled for 10 minutes) was tried, but it failed when the girl presented profuse vomiting after 3 hours from the ingestion. At 14 months of age, she accidentally came in contact with the offending food again after the intake of biscuits containing egg and she experienced a renewed relapse of symptoms. In her past medical history, no other allergic diseases were reported. Skin prick tests were performed during the first evaluation at our clinic, all negative. According to the International Consensus Guidelines 2017, we made diagnosis of acute FPIES based on the finding of the major criterion and 3 minor criteria, and egg-free diet was suggested. At 30 months of age, a new attempt of OFC was suggested considering the proposals of the latest scientific studies, but it was only possible to perform it just at 42 months of age due to pandemic-related delays. The OFC was performed giving 1/6 of a hard-boiled egg 3 times every 20 minutes. The dose administered was 60 g total containing 4 g of protein. After 4 hours of observation the girl was examined again, and she was discharged home given the absence of pathological elements. At home she did not show late reactions and in the following days she experienced hard-boiled egg in her diet introducing 1/6 of it for 2 days and then increasing the dose by 1/6 every 2 days until the dose of half an egg was reached. After 2 weeks, the dose was further increased by 1/6 every 2 days until the dose of a whole egg was reached. Given the age of the girl and the absence of clinical manifestation, the diet was liberalized.

CONCLUSIONS

The case report then agrees with the latest evidence about the cooking influence in tolerance acquisition in egg-induced acute FPIES: the tolerance was reached at 42 months of age and we can affirm that also raw egg tolerance was acquired as the girl showed no more episodes after the liberalization of the diet. OFC can be attempted much earlier than 53 months of age, after a period of egg-free diet, starting with cooked egg until the introduction of the raw one, allowing a better quality of life for the patient.

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ABS 59

PEDIATRIC SCURVY, STILL A MODERN DISEASE? A CASE REPORT

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BACKGROUND

Scurvy is a well-known clinical condition caused by vitamin C deficiency. It is a rare clinical entity in modern age, which presents in children as irritability, pseudoparalysis, failure to thrive and gingival hemorrhage. Scurvy has been recently increasingly reported in children, especially in those with abnormal dietary habits, mental or physical disabilities. Although the entity is well described in literature, the diagnosis is often difficult due to the disease broad clinical presentation.

CASE PRESENTATION

We report a case of a 6-year-old male with Down syndrome who presented to the ER with copious gum bleeding, widespread bruising and petechiae. He showed some difficulties to walk. He was firstly evaluated by the general practitioner, who suspected a bacterial gingivostomatitis and treated the condition with antibiotics, without any result. He underwent extensive diagnostic testing, then a more accurate anamnesis was performed and it was evident that the kid, in the past 2 months, had refused to eat properly, he only fed with biscuits, milk and yogurt. Based on this new information a multivitamin deficiency was suspected. Ultimately ascorbic acid returned undetectable, as well as folic acid, and blood exams also showed iron deficiency anemia. He had immediate and complete improvement upon starting vitamin C, folic acid and iron supplementation. He was also provided with a multivitamin and protein formula. The boy was finally evaluated by a dietician, speech therapist and psychiatrist to start a personalized feeding program and psychological follow-up.

CONCLUSIONS

This case shows that although the incidence of scurvy has been greatly reduced in Western Europe, this pathology can still manifest itself, especially in vulnerable children. Only early diagnosis can avoid unnecessary investigations and potentially fatal complications of the disease. The real prevention in these patients consists in food education and an adequate nutritional intake in global health support, under a multi-specialistic approach.

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ABS 60

FROM TYPICAL TO ATYPICAL

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CASE REPORT

F., 12 years old, had a history of congenital left megaureter and previous typical hemolytic-uremic syndrome (HUS) in 2020. The patient was successively hospitalized, in January 2022, for the appearance of red colored urine, low fever and vomiting. Molecular swab for SARS-CoV-2 is positive. Pale, jaundiced skin, some petechia in the abdomen. In laboratory tests, evidence of acute kidney injury (sCr 1.3 mg/dL, BUN 71 mg/dL, and in the following 12 hours sCr 1.6 mg/dL, BUN 85 mg/dL), thrombocytopenia (13,000/micro-liters) and high values of LDH (3,074

IU/I), AST (112 IU/L) and bilirubin tot 4.4 mg/dL (mainly indirect). Normal PT and aPTT, D-Dimer 17,710 ng/mL (FEU). Anemia (Hb 9.8 g/dL, Hct 29.2%) and consumption of haptoglobin (31.7 mg/dL), C3 and serum immunoglobulins in the norm; negative coproculture for pathogenic *E. coli*. Due to suspicion of atypical HUS in evolution and for SARS-CoV-2 positivity of the patient and patient's parent too, the patient is moved to a Level III Pediatric Center, where a study of ADAMS13 (activity and antibodies) is carried out, which is normal and confirms the presence of schistocytes in the sediment. For the previous episode of HUS it was decided to treat with eculizumab (4 weekly doses at a dosage of 900 mg, then maintenance at a dosage of 1,200 mg every 2 weeks, still ongoing). Subsequent progressive improvement in renal function, in thrombocytopenia and anemia. Blood samples were taken (patient and parents) for genetic testing with whole-exome sequencing method and bioinformatic filtering for a panel of genes associated with atypical HUS.

DISCUSSION

Atypical HUS, now defined as non-STEC HUS, represents today 10% of pediatric HUS and has an estimated prevalence of 7/1 million children in Europe [1]. It is due to genetic mutations of complement factors but it may also be due to the development of antibodies against complement factor H, mutations of the epsilon gene of the diacylglycerol kinase, inborn errors of cobalamin C3 metabolism. Mutations occur by loss of function in a regulatory gene (*CFH*, *CFI*, or *CD46*) or by gain of function in an effector gene (*CFB* or *C3*). Following a triggering event, such as an infection (usually of the upper airways) in a susceptible individual with one or more gene variants or antibodies to complement proteins, uncontrolled activation of the alternative pathway occurs with consequent formation of the attachment complex membrane (MAC), with damage to the endothelium and thrombotic microangiopathy [2].

CONCLUSION

The advent of eculizumab in the treatment of non-STEC HUS has improved significantly patient prognosis. It is a humanized mono-local antibody that binds to the complement protein C5, blocking its cleavage and leading to the reduction of complement activity and has become the treatment of choice in all patients with non-STEC HUS [3].

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ABS 61

EARLY CAPILLARY BLOOD PARAMETERS IN PRETERM AND TERM NEWBORNS

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BACKGROUND

There is growing evidence that transitional phase represents the crucial step from intra- to extrauterine life. In this scenario, the main monitoring parameters available for the frontline physician are the changes in blood pH and gas analytes (BPGA). Up to now, no studies offer reference curves of BPGA in capillary blood corrected for the main perinatal outcomes. The aim of the present study was to investigate whether BPGA parameters, in a cohort of healthy preterm (PT) and term (T) newborns, were gestational age (GA) and weight at birth (BW), delivery mode and gender dependent.

METHODS

We conducted an observational study in 99 PT and 300 T newborns. At birth, newborns fulfilling all the following criteria have been classified as normal: no maternal illness; no signs of fetal distress; pH 7.2 in cord or venous blood; and Apgar scores 7 at 1 and 5 minutes. Clinical and laboratory parameters (capillary blood pH, partial carbon dioxide venous pressure [pCO_2], partial oxygen venous pressure [pO_2], base excess [BE]) were recorded in all infants in the first 12 hours from birth.

RESULTS

BPGA analytes levels differed between PT and T newborns and were GA, BW, delivery mode and gender dependent. Multivariable analysis showed that pH, pO_2, pCO_2, and BE correlated with GA and BW among a series of perinatal parameters.

CONCLUSIONS

The present results, showing that a series of perinatal outcome can affect BPGA in the first hours from birth, open the way to further investigations aimed at providing longitudinal BPGA reference curves in the transitional phase, empowering the diagnostic and therapeutic strategy efficacy.