

www.jpnim.com Open Access eISSN: 2281-0692 Journal of Pediatric and Neonatal Individualized Medicine 2017;6(2):e060234 doi: 10.7363/060234 Published online: 2017 Oct 24

Abstracts

Selected Abstracts of the 13th International Workshop on Neonatology

THE POWER OF EPIGENETICS TWINS: IDENTICAL BUT DIFFERENT

CAGLIARI (ITALY) · OCTOBER 25TH-28TH, 2017

The Workshop has been organized with the patronage of the Italian Society of Neonatology (SIN), the Italian Society of Pediatrics (SIP), the Italian DOHaD (Developmental Origins of Health and Disease) Society, the Italian Society of Preventive and Social Pediatrics (SIPPS), the Union of European Neonatal and Perinatal Societies (UENPS), the International Federation of Clinical Chemistry and Laboratory Medicine (IFCC), the Union of Middle-Eastern and Mediterranean Pediatric Societies (UMEMPS), the European Association of Perinatal Medicine (EAPM) and lastly the Italian-Romanian Society of Pediatrics.

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HOW TO CITE

[Abstract's authors]. [Abstract's title]. In: Selected Abstracts of the 13th International Workshop on Neonatology; Cagliari (Italy); October 25-28, 2017. J Pediatr Neonat Individual Med. 2017;6(2):e060234. doi: 10.7363/060234.

ABS 1

NEONATAL ACUTE KIDNEY INJURY AND OXI-DATIVE STRESS

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INTRODUCTION

Newborns are especially prone to oxidative stress (OS) [1]. OS can exacerbate several neonatal conditions to the extent that Saugstad coined the phrase "oxygen radical disease of neonatology" such as bronchopulmonary dysplasia, chronic lung disease, respiratory distress syndrome, periventricular leukomalacia, neonatal encephalopathy, retinopathy of prematurity, necrotizing enterocolitis, and acute kidney injury (AKI) [2]. The objective of this research is to study different components of pro-oxidant system and antioxidant defense system in critically ill full-term newborns with and without AKI.

MATERIALS AND METHODS

Fifty-seven critically ill full-term neonates were selected in this study. The first group included 22 neonates with AKI, the second group included 35 newborns without AKI. AKI was detected according to literature [3]. The level of oxidative modification of proteins (OMP) and concentration of malondialdehyde (MDA) in erythrocytes as pro-oxidant markers were established. The concentration of ceruloplasmin (CR), activities of catalase (Ct) and glutamyl transpeptidase (GTP) in plasma, activities glucose-6-phosphate dehydrogenase (G6PD) and glutathione reductase (GR) in erythrocytes as anti-oxidant markers were established.

RESULTS

In the first group the level of OMP was 1.16 ± 0.02 optical density per milliliter of erythrocytes, in the second group was 1.04 ± 0.02 optical density per milliliter of erythrocytes, p < 0.05. The concentrations of MDA in erythrocytes were $25.1 \pm 0.48 \mu$ mol/l and 24.2 ± 0.39 respectively, p > 0.05.

In the first group the level of CP was 195.3 ± 3.91 mg/l, in the second group was 224.1 ± 5.9 mg/l, p < 0.05; activities of Ct were 9.6 ± 0.37 µmol/min•1 and 6.9 ± 0.31 µmol/min•1 respectively, p < 0.05; activities of GTP were 68.0 ± 3.83 UI/l and 107.1 ± 1.16 UI/l respectively, p < 0.05. The activity of G6PD in the first group constituted 1.9 ± 0.06 µmol/min•HB, in the second group – 1.6 ± 0.04 µmol/min•HB, p < 0.05; GR activities were 2.1 ± 0.07 µmol/min•HB and 2.3 ± 0.06 µmol/min•HB respectively, p > 0.05. CONCLUSION

The imbalance of the presented components of prooxidant system and antioxidant defense system in critically ill full-term newborns under conditions of pathological OS is one of the triggering mechanisms of a number of pathological reactions causing the formation of renal injuries in early neonatal period. REFERENCES

 Mutinati M, Pantaleo M, Roncetti M, Piccinno M, Rizzo A, Sciorsci RL. Oxidative stress in neonatology. A Review. Reprod Dom Anim. 2014;49:7-16.
 Saugstad OD. Oxygen and oxidative stress in bronchopulmonary dysplasia. J Perinatal Med. 2010;38(6):571-7.

[3] Selewski DT, Charlton JR, Jetton JG, Guillet R, Mhanna MJ, Askenazi DJ, Kent AL. Neonatal Acute Kidney Injury. Pediatrics. 2015;136(3):E463-73.

ABS 2

LIFE SUPPORT MANAGEMENT IN NEONATES WITH SEVERE OR DISMAL PROGNOSIS: THE OPINIONS OF DOCTORS AND NURSES IN A TERTIARY-LEVEL NEONATAL INTENSIVE CARE UNIT

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INTRODUCTION

Advances in fetal diagnosis and postnatal medical care are increasingly improving survival and offering more hope of recovery, both in extremely premature babies and more mature infants born with severe clinical conditions. Thus, health professionals and parents are frequently challenged to make complex and emotionally demanding decisions, which may ultimately affect whether a baby shall live or die [1, 2]. MATERIALS AND METHODS

An anonymous online, 48-item questionnaire was distributed to doctors and nurses of the NICU in Verona, to find out about the opinions of health professionals regarding the adoption of end-of-life decisions in critically ill infants.

RESULTS

Among the 69 recruited health professionals, we found much uncertainty about who should make end-of-life decisions. For example, while about half of the health professionals considered the Director as the final decision-maker in the NICU setting, though after multidisciplinary meetings (56.5%), in the delivery room there was a greater degree of indecision, identifying either the physician on duty (29%), the medical and nursing team (23.2%), or the Director (33.3%). Conflicts of opinion as a result of end-of-life decisions were reported by 46% of the health professionals.

In evaluating 3 clinical scenarios (any patient, extreme prematurity or severe encephalopathy), we observed significantly different opinions between males and females. In particular, males either strongly disagreed or agreed in providing supportive care in each case, while females were in disagreement or uncertain (**Fig. 1**). Of note, as the age and clinical experience of health professionals increased, uncertainty about life support decisions raised.

CONCLUSIONS

Managing life support in neonates with poor or dismal prognosis is very controversial. Lack of shared scientific criteria complicates the role of the health professionals. Several uncertainties and difficulties in making end-of-life decisions by NICU health professionals clearly emerged from our questionnaire. Tight collaboration between the medical and nursing team, as well as adequate parental involvement are essential [3]. Multidisciplinary meetings and specific training courses are needed to improve knowledge and quality of care in this delicate field.

REFERENCES

 Tripp J, McGregor D. Withholding and withdrawing of life sustaining treatment in the newborn. Arch Dis Child Fetal Neonatal Ed. 2006;91:F67-71.
 Dupont-Thibodeau A, Barrington KJ, Farlow B, Janvier A. End-of-life decisions for extremely low-gestational-age infants: why simple rules for complicated decisions should be avoided. Semin Perinatol. 2014;38(1):31-7.
 Cuttini M, Nadai M, Kaminski M, Hansen G, de Leeuw R, Lenoir S, Persson J, Rebagliato M, Reid M, de Vonderweid U, Lenard HG, Orzalesi M, Saracci R. End-of-life decisions in neonatal intensive care: physicians' self-reported practices in seven European countries. EURONIC Study Group. Lancet. 2000;355(9221):2112-8.

ABS 3

THE METABOLOMICS REVOLUTION IN BRON-CHOPULMONARY DYSPLASIA: PERSONAL EX-PERIENCE

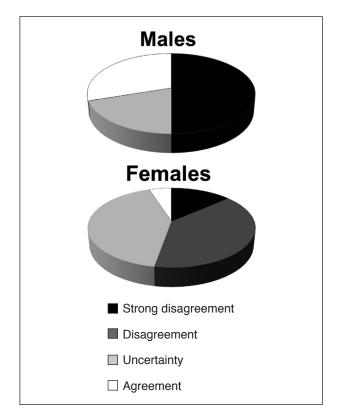


Figure 1 (ABS 2). Example of orientation of males and females in life support decisions in extremely premature babies.

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INTRODUCTION

Bronchopulmonary dysplasia (BPD) is the most common pulmonary complication induced by prematurity and it is one of the major causes of mortality in high-preterm infants. It has a multifactorial etiopathogenesis, and still it is extremely difficult to treat resolutely and effectively infants with this disease. Therefore, to be able to identify precociously patient with greater risk of BPD would improve the outcomes of these little patients and identify effective personalized prevention strategies. Metabolomics is the science that studies the metabolome, the ultimate product of biochemical reactions. In order to analyze the complex metabolic system of an individual, a large number of metabolites from different biofluids, such as blood, urine, sweat, saliva, are examined by spectroscopic techniques. Multivariate

statistical data analysis allows understanding which metabolic information is relevant to the biological characterization of a given physiological or pathological condition. The aim of our study was to characterize the urinary metabolism of newborn babies with BPD, to increase the basic knowledge of the metabolism of these patients and compare the data obtained with those in the literature.

STUDY POPULATION AND METHODS

For the study, 36 preterm infants were recruited altogether below the gestational age of 29 weeks and weighting less than 1,500 g. Patients were divided into two groups: cases and healthy controls. Urine samples were collected by non-invasive methods at the end of the seventh day of life, using a sterile cotton wad inserted in a disposable nappy. 1 ml of urine was then taken, and then prepared for proton nuclear Magnetic Resonance Assay (H-NMR). H-NMR spectra were acquired with a Spectrometer Varian Unity Inova 500. The results of urine spectra were analyzed using a supervised model (OPLS-DA R2Y = 0.81 and Q2 = 0.66) to two classes that highlighted a valid model that discriminates children's urine samples with BPD from those of healthy controls.

RESULTS

Metabolites that significantly contributed to the separation of healthy infants from BPD cases included alanine, betaine (increased in BPD group), glycine, lactate and trimethylamine N-oxide (TMAO) (decreased in BPD group). Alanine synthesis is directly proportional to intracellular pyruvate concentration. In anaerobiosis, pyruvate is partially converted to alanine and partly to lactic acid and in patients with BPD this would correlate with the increase in alanine in the urine. The glucose-alanine cycle is also stimulated by elevated plasma glucocorticoid levels that are increasing during stress, such as the development of BPD. Cori and Krebs cycles alterations would correlate with lactate depletion because lactate is transported to the liver and used as gluconeogenetic precursor. Betaine is one of the major donors of methyl groups and participates in methylation of DNA. This could therefore correlate with the epigenetic bases of the disease. TMAO is one of the products of the intestinal microbial activity. In high-grade preterms, the reduction of TMAO may be due to the immaturity of the microbiota. Glycine is one of the constituents of glutathione, one of the main protective factors of oxidative stress. In high-grade preterms the need for high levels of glutathione could correlate with the reduction of urinary glycine.

DISCUSSION

The metabolomics analysis of the urine therefore has allowed a discrimination between the two groups, suggesting that newborn infants with BPD have a modified metabolic profile, as a result of an altered cellular metabolic activity. The results of our study have determined the possible trend of metabolites in patients who develop BPD with respect to healthy controls and thus to determine a metabolic "fingerprint" that could be used as a predictive and diagnosis tool of this pathology, in the future. Therefore it seems it could be possible to predict accurately which patients will develop BPD so that they can receive individualized treatments, making it possible to manage pediatric disorders such as BPD in a tailored way.

REFERENCES

• Fanos V, Van den Anker J, Noto A, Mussap M, Atzori L. Metabolomics in neonatology: fact or fiction? Semin Fetal Neonatal Med. 2013;18(1):3-12.

• Fanos V, Pintus MC, Lussu M, Atzori L, Noto A, Stronati M, Guimaraes H, Marcialis MA, Rocha G, Moretti C, Papoff P, Lacerenza S, Puddu S, Giuffrè M, Serraino F, Mussap M, Corsello G. Urinary metabolomics of bronchopulmonary dysplasia (BPD): preliminary data at birth suggest it is a congenital disease. J Matern Fetal Neonatal Med. 2014;27(Suppl 2):39-45.

• Baraldi E, Giordano G, Stocchero M, Moschino L, Zaramella P, Tran MR, Carraro S, Romero R, Gervasi MT. Untargeted Metabolomic Analysis of Amniotic Fluid in the Prediction of Preterm Delivery and Bronchopulmonary Dysplasia. PLoS One. 2016;11(10):e0164211.

ABS 4

WHARTON'S JELLY: A SIGNIFICANT DETAIL

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INTRODUCTION

Wharton's jelly is an important substance, which supports and protects umbilical vessels. Its absence or deficiency is usually associated with fetal death or poor neonatal outcome, because umbilical cord is more prone to compression. This is a very rare, but critical condition and in literature are described only few cases.

CASE REPORT

A 37-year-old woman, first pregnancy after assisted medical reproduction (ICSI), was admitted to hospital at 40 week of gestation for induction of labour because of impaired fetal growth. During the expulsive stage of delivery unexpected and serious bradycardia occurred, so that the fetus was immediately pulled out with vacuum extractor. The newborn had two cord's rounds around the neck and the umbilical cord was very thin (8 mm) with lacking Wharton's jelly. Birth weight was 2,390 g (< 3th percentile). The baby was atonic, apneic, with absence of detectable heart rate: she was resuscitated with intubation, ventilation, chest compression, administration of intravenous epinephrine and volume expander. Good heart rate occurred at 17th minute of birth; she was transferred to NICU where began hypothermia. At 20 minutes of life gas analysis revealed important acidosis: pH 6.46, pCO₂ 120, HCO₃ 2.8 mmol/l, basic excess -27, lactate 23. She showed severe hypoxic ischemic encephalopathy with absent reflexes, coma, hypotonia. Electroencephalography revealed a severe alteration of cerebral electrogenesis. In addiction she developed disseminated intravascular coagulation, not responsive to plasma and requiring vasoactive amines to support cardiac output. The clinical condition become progressively severe and the baby died at twenty hours of life. Autopsy showed thin cord with diffuse deficiency of Wharton's jelly, no other anatomical or placental abnormalities were detected.

DISCUSSION

The fetus receives all of its nutrients through umbilical cord, which represents a vital organ for fetus. Umbilical cord abnormalities can lead to fetal death or serious problems during pregnancy and delivery. Thin cord syndrome (TCS) with deficiency of Wharton's jelly is very rare and it's associated with significant fetal or neonatal pathology. The reduction of Wharton's jelly can lead to placental insufficiency without pathological Doppler parameters but with fetal intrauterine growth retardation, as in our case report. The association with oligohydramnios increases the risk of reduced fetal perfusion. In the presence of SGA or IUGR of uncertain cause it is important not only to assess fetal Doppler parameters but also cord morphology. In fact, in rare cases, this can play an important role, thus enabling detection of women who might benefit from a more intensive antenatal monitoring.

REFERENCES

• Ghezzi F, Raio L, Gunter Duwe D, Cromi A, Karousou E, Durig P. Sonographic umbilical vessel morphometry and perinatal outcome of fetus with a lean umbilical cord. J Clin Ultrasound. 2005;33(1):18-23.

• Kulcarni ML, Matadh PS, Ashok C, Pradeep N, Avinash T, Kulkarni AM. Absence of Wharton's jelly around the umbilical arteries. Ind J Pediatrics. 2007;74:787-9.

• Proctor LK, Fitzgerald B, Whittle WI, Mokhtari N, Lee E, Machin G, Kingdom JCP. Umbilical cord diameter percentile curves and their correlation to birth weight and placental pathology. Placenta. 2013;34:62-6.

ABS 5

FROM STORK WINGS TO AIRPLANE'S: OUR EXPERIENCE ON 100 NEONATAL TRANSPORTS

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INTRODUCTION

The neonatal transport service (STEN) is the main transport network for the critical and pathological infant, present throughout Europe, extending over 90% of the Italian peninsula but with of Valle d'Aosta and Sardinia exception. In the latter case STEN could be configured as a regional transport system, but Sardinia's geographical feature imposes the need for other interregional transport systems for the peninsula.

OBJECTIVE

To evaluate the epidemiology of critical infant air transport in Sardinia region, enhancing the Italian Military Aeronautics contribution.

MATERIALS AND METHODS

Interregional transfers carried out over a fiveyear observational period from 2012 to 2017 have been examined. Data were extracted from the Department's paper registers, the SISAR Regional Health Register and the clinical records of the patients transferred.

RESULTS

The total number of neonatal interregional transfers carried out in 2012-2017 is one hundred, including fifty-five from the Neonatal Intensive Care Unit (NICU) and forty-five from the Neonatal Pathology. In the period 2015-2017 the total transfers were forty-nine, twenty-four from the NICU and twenty-five from the Neonatal Pathology. The main conditions that require transport for surgical treatment were abdominal, heart and neurological diseases. The destinations were: Pediatric Hospital Bambin Gesù (34), Pediatric Meyer Hospital (4), Pediatric Paschinucci Hospital in Massa Carrara (2), Hospital IRCCS Policlinico San Donato Milanese (7), Pediatric Gaslini Hospital in Genoa (2).

DISCUSSION

Insularity, high population density in regions near coast, the absence of highly specialized Departments make clear the high percentage of air transfers under critical conditions and lifethreatening patients. All this is dealt with the collaboration of the Italian Military Aeronautics and the medical and nursing staff of the two Departments mentioned above, regardless of service hours. This imposes on the Sardinian institutions considerable costs that remain counterbalanced by benefits, in terms of health, both for patient and family.

CONCLUSIONS

If the image of a stork represents the happiness of birth, instead the wings of an airplane of the Italian Military Aeronautics are hopeful and represent the chance of a normal life for doctors, nurses, parents and their sick children.

REFERENCES

Selected Abstracts of the 3rd Edition of Transport of High Risk Infants;
 Oxford (UK); August 31st-September 2nd, 2017. J Pediatr Neonat Individ Med.
 2017;6(2):e060214.

ABS 6

ANOMALIES OF INTRA-HEPATIC BILE DUCT CILIA IN BILIARY ATRESIA

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INTRODUCTION

Abnormalities in primary cilia of biliary epithelial cells (BEC) have been described in both intra- and extra-hepatic biliary ducts from livers of patients with biliary atresia (BA) and chronic severe liver disease. Pathogenetic significance of these abnormalities has not been yet determined. Aim of the study was to investigate intra-hepatic BEC cilia (IHBC) in liver biopsies of infants with biliary atresia at the Kasai portoenterostomy surgery.

MATERIALS AND METHODS

Surgical biopsy liver tissue sections from 31 cholestatic infants (22 BA, 11 males, ages 39 to 116 days; 9 non-BA, 4 males, ages 0.45 to 16.87

years) were evaluated for IHBC by indirect immunofluorescence (IF) and conventional scanning electron microscopy (CSEM) by a pathologist blinded to clinical information. Controls included infants with choledochal malformations, portal cavernoma and total parental nutrition related cholestasis.

RESULTS

In BA group, hepatic histology showed pictures with variable degrees of fibrosis, ductular proliferation and inflammation. At IF, IHBC were absent or reduced in 19 (86%) BA but only in 3 (30%) non-BA liver sections (p < 0.05) CSEM analysis showed several anomalies in BA samples with IHBC compared to non-BA.

CONCLUSIONS

Our study confirms that significant IHBC anomalies characterize BA livers, which appear to correlate to the progression of liver damage until total BEC cilia loss. Whether BEC cilia might have a role in BA etiopathogenesis and/or disease progression warrants further investigation.

ABS 7

HUMAN HERPES VIRUS 6 INFECTION IN CHILDREN WITH LYMPHOBLASTIC ACUTE LEUKEMIA COMMON: CAUSING AGENT OR CASUAL ASSOCIATION?

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INTRODUCTION

Infections (Hepatitis viruses, Ebstein Barr Virus, Parvovirus, and Human Herpes Virus 6 [HHV6]) are possible causes of acquired Aplastic Anemia (AA). HHV6 potential oncogenic role for Lymphoblastic Acute Leukemia (LAL) has been hypothesized also. We describe a child with AA, possibly caused by HHV6, who developed LAL. CASE REPORT

An 8-year-old male arrives to the emergency room with vomit, diarrhea, and fever since 48 h. At physical examination: pallor, fatigue, and laterocervical lymphadenopathy. Laboratory tests: WBC 1,200/mmc (neutrophils 230/mmc), Hb 7.7 g/dL, reticulocytopenia, PLT 87,000/mmc, CRP 18.2 mg/dL; electrolytes, liver and kidney function tests, LDH normal; serum HHV6 DNA positive (< 1,000 copies), HHV6 IgG positive, IgM borderline. Peripheral blood smear: no quantitative anomalies. Chest X-rays and abdominal ultrasound: within normal limits. After 14 days: WBC 5,050/mmc (N 1,400/mmc), Hb 11.6 g/dL, reticulocytosis, PLT 671,000/mmc. After 30 days: clinical deterioration with pallor, petechiae, laterocervical and axillary lymphadenopathy; WBC 29,360/mmc (N 3.8% and L 84%), Hb 8.4 g/dL, PLT 67,000/mmc. A bone marrow aspirate was performed and LAL common was confirmed.

CONCLUSIONS

The role of HHV6 in the pathogenesis of LAL is not clear. Even though recent studies appear to deny an oncogenic role of the virus, in our case, the hemopoietic transient suppression, likely induced by HHV6 infection, followed by rapid neoplastic transformation, suggests re-evaluating its role.

ABS 8

WHOOPING COUGH IN NEWBORN: A CLINICAL CASE DIFFICULT TO SOLVE

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INTRODUCTION

Whooping cough is an acute and contagious infectious disease of the respiratory system and is caused by a specific organism: B. Pertussis or Bordet and Gengou bacillus. It is a Gram negative bacterium, asporogenous, aerobic. Such disease prefers childhood but also affects adults and even many elderly people, especially following the increase of childhood vaccination practice. Pathogenesis seems primarily attributable to toxins that, by involving the upper airway mucosa, cause edema phenomena followed by superficial necrosis with irritation of the respiratory endings of the vague. The initial lesion is hyperplasia of peribronchial lymphatic tissue and tracheobronchial lymph nodes. Subsequently pharynx, larynx, trachea and bronchi are involved. The typical anatomic-pathological lesion is the

necrosis of the middle and basal layer of the epithelium. The bacterium is found among the lashes of the epithelial cells of the trachea and bronchi. Incubation period is about 5-20 days. In typical forms the course of the disease is six to eight weeks and can be divided into three periods: catarrhal, convulsive or spasmodic, period of decline or convalescence. In the neonatal age and in the first months of life, the symptoms are different, insidious, and much more serious when patient is too small, as well as it is often very difficult to diagnose early. In the first few weeks of life or in the early months, the disease begins with the typical clinical picture. Especially in the first month of life and also later, the cough is little present or absent and is replaced by episodes of apnea with cyanosis or by a "sneezing" crisis with dense mucus emission from the mouth. The mucus can be diagnosed by an expert observer. In fact, in a very small child, sputum is unusual and occurs only in two conditions: pertussis and bronchiectasis. Symptoms often contribute to a diagnostic error, because it is often interpreted as a gastroesophageal reflux (GER). In the first year of life and in the following months the disease may be very serious and dangerous for the patient's life. Death can occur by asphyxia during access and "syncopal episodes", preceded by intense cyanosis. The cause of death is attributed to a severe hypersensitivity of the brain due to repeated access. Although relatively infrequent, pertussis encephalopathy is one of the most well-known and serious complications of the disease and is the consequence of tiny blemishes or diffuse thrombosis or generalized cerebral edema or, more often, anoxic brain injury. Such encephalopathy occurs clinically with seizures and coma, is burdened with considerable lethality in the smallest patients (1/3 of the cases). In survivors it can leave neurological after-effects of various kinds. The cause of pertussis encephalitis is still obscure: it is assumed to be linked to the responsibility of ischemic-hemorrhagic mechanisms and the co-responsibility of immuneallergic mechanisms.

CLINICAL CASE

A 25 days newborn baby was taken to us following a severe GER diagnosis at 20 days old. The newborn presented the following symptomatology: apneas, cyanosis, drowsiness, loss of consciousness, incessant output of dense mucus from the mouth. Despite the therapy for GER, symptoms progressively worsened, requiring

further attention. The small baby had a normal clinical examination; anamnestic result, however, was very alarming. In light of our examinations and above all on the basis of the clinical and instrumental data, we thought of neurological disorder, so we directed the little to a highly specialized structure to carry out a neurological investigation. Notwithstanding negative EEG, cerebral echo, and cerebral MRI, the neurologistpediatrician suspected a "convulsive condition" and undertook anticonvulsant therapy to which the small baby reacted for the first few days. Subsequently, the symptoms resumed more intense than before with the appearance of new symptoms: cough, mucus dense by mouth, syncopal episodes. Such evidences have allowed making diagnosis of whooping cough. Laboratory tests revealed significant leukocytosis (58,000) with marked lymphocytosis (90%) and the diagnosis has been made with isolation of B. Pertussis. Appropriate therapy has allowed the patient a rapid recovery and discharge.

CONCLUSIONS

We believe it is important and appropriate to draw the attention of the pediatricians to this disease that, despite the strong increase in vaccination, still affects our children. It is of utmost gravity to experience diagnostic delays and to practice, in the first days of life, inappropriate and harmful therapies for our newborns.

ABS 9

FROZEN SECTIONS EXAMINATION OF PLA-CENTA IN SEPTIC NEWBORNS. THE GENOA EXPERIENCE

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BACKGROUND

Inflammatory placenta lesions often correlate with a neonate septic state and require prompt antibiotic therapy. However, antibiotic therapy alters the formation of the neonatal microbioma with possible dysfunctions in the later life. Routine hematochemical indexes of inflammation are often considered unreliable.

AIMS

To investigate the reliability of frozen sections examination of placenta in the therapeutic choice of newborn babies with possible or suspected sepsis.

MATERIALS AND METHODS

All placentas submitted for frozen section examination at the Operational Unit of Pathology of the San Martino Polyclinic Hospital in 2011-2015 were considered. For each case a sample of umbilical chord and a sample of amniotic membranes were processed with frozen sections. The presence and intensity of inflammation in the umbilical cord and membranes has been evaluated. The results of the examination have been communicated to the neonatologist in less than 45'. The result of frozen sections examination was then compared with results of permanent slides. RESULTS

A total of 407 placentas were considered with mean maternal age of 32.83 ± 5.69 years and with mean gestational age of 37.94 ± 2.84 wks. The indications to frozen section examination were divided into: absolute (PROM > 12 h = 202; positive vaginal swabs = 59; maternal fever = 48) and relative (tocographic alterations = 81; urgent cesarean section = 72; meconium discharge = 62; others = 75). Frozen sections examination showed funisitis in 38 cases (9.34%) and chorioamnionitis in 182 cases (44.9%; mild = 132; moderate = 42;severe = 8), consequently, according to the flowchart in Fig. 1, antibiotic therapy was given in 39 cases (9.58%) while 27 cases (6.63%) required further clinical exams. Comparing these results with those of the permanent sections, a high correlation was observed for both funisitis (κ = 0.7; sens. = 71.05%; spec. = 97.56%; PPV = 75%; NPV = 97.04%) and chorioamnionitis ($\kappa = 0.65$; sens. = 78%; spec. = 92.72%; PPV = 60%; NPV = 96.78%) resulting in overlapping therapeutic choices.

CONCLUSIONS

In the possible or suspected cases of neonatal sepsis, placental frozen sections examination represent a quick, useful and reliable tool for therapeutic choice, in particular if result negative for inflammation. It allows a drastic reduction in the administration of antibiotics and a significant reduction in unnecessary or unreliable clinical examinations.

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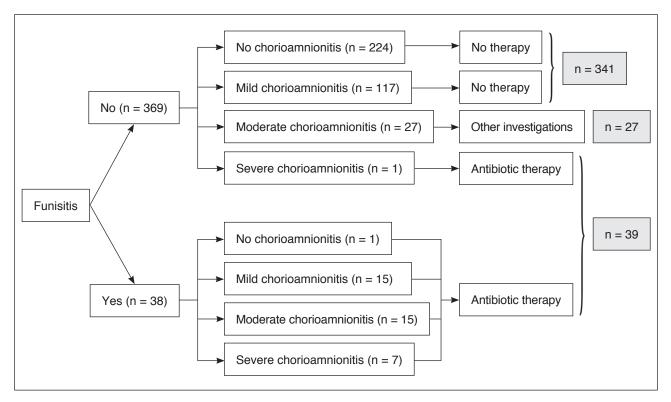


Figure 1 (ABS 9). Frozen sections flow-chart.

ABS 10

THE ACADEMIC PEDIATRICS OF SIENA AND THE GREAT WAR; A STUDY CREATED FOR THE CENTENARY OF THE FIRST WORLD WAR

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The history of the Academic Paediatrics of Siena is useful to understand both the birth and development of the discipline inside the University, and the Italian military healthcare during the Great War; as well as the influence that this has had in society and in the Italian medical field. For this reason, this study was conducted on the occasion of the centenary of the First World War.

WHEN THE WAR WAS FAR AWAY

Until 1917 the Academic Paediatrics of Siena developed in a fast and excellent way, it was one of the first faculties to be set up, dating back to the eighth century in Italy (**Tab. 1**), and it was entrusted to a local professor, Enrico Gagnoni. After his death his position was immediately filled by another professor, an unusual situation in many Universities even prestigious. This path was brutally stopped

a hundred years ago, because of the war. Enrico Gagnoni, born in Siena on January 1st 1870, was the first Pediatric Professor: he was Director of the Pediatric Clinic from 1904 to 1915. When Gagnoni died in Siena on the 13th May 1915, Gino Menabuoni, professor of Florence's pediatric clinic, replaced him. He was a student of Carlo Comba's (1870-1951) school in Florence that trained a number of students who won numerous professorships. Carlo Comba's school did not reach the success of the Sicilian-Parthenopean school by Rocco Jemma (1866-1949), that went on to spread throughout Italy, but it is ranked number two in the pediatric history of Italy over the last 100 years. Menabuoni was very active in the organization of the pediatric clinic: he increased nurses and clinics, and opened the laboratory and "Aiuto Materno" ("Maternal Help") ward to help

Table 1 (ABS 10). Italian universities, in ascending order,when the teaching of Pediatrics was established.

Year of establishment	University			
1882	Florence, Padua			
1886	Naples			
1894	Rome			
1900	Bologna, Parma			
1903	Palermo			
1904	Siena			

pregnant women and children with social issues. On November 26th 1916 Menabuoni inaugurated the new pediatric clinic in the presence of Angelo Roth (1855-1919), undersecretary of Public Education. Menabuoni particularly studied syphilis, tuberculosis and tetanus.

THE WAR

When the war begun, Gino Menabuoni had been dismissed from the military due to health issues, but he was recalled to the battlefield in 1917. This is a really interesting fact as it testifies to the lack of doctors in the Regal Army that forced many doctors, like Menabuoni, born between 1874 and 1884 to join the army. The lack of doctors was influenced by the high number of soldiers mobilized, the Army alone was formed of 5,728,277 soldiers deployed on a wide front hundreds of kilometers wide. When the war started the Italian Army only had 770 doctors serving permanently, therefore they had to call doctors born from 1870, so 45 years old at the time. Despite this, the number of doctors was still insufficient. The difficulty for healthcare in the military was the large number of men needing assistance for different illnesses, more cruel and severe, as it was an extremely cruel war. The innovative elements introduced during the First World War that took unexpectedly Italian military physicians were the increase in artillery; the new weapons such as aerial bombings from airplanes, and chemical weapons with the destructive actions of asphyxiation, blistering, irritation and high toxicity. These elements determined a change also in the type of illness that needed medical attention. If in the past the infectious diseases occupied doctors' time more than the actual injuries, now, the introduction of these new weapons determined a change in the type of illness, and injuries became more relevant especially the ones caused by the artillery that, with howitzers up to 420, created horrible injuries and caused 66% of the total injuries, while only 23.5% were caused by light weapons. It was a cruel war, with 680,071 lives lost, and including the disabled arrived to 1,142,883 people. To understand the value of the military medical care, we can think that it treated 950,000 injured people, which is 16.57% of the total Army size. Menabuoni couldn't cope with the hard military life. In February 1919 he died from an illness while he was hospitalised in a military hospital in Verona. AFTER THE WAR

In 1919, after Menabuoni's death, Angiola Borrino (**Fig. 1**), born in Cossato, on March 27, 1880, once in Novara's county and now Biella, had the direction of the pediatric ward. One of Battista Allaria's (1872-1955) pupils at the Pediatric Wards at the University



Figure 1 (ABS 10). Angiola Borrino.

of Turin, she stayed in Siena during the Academic year of 1923/1924. Angiola Borrino was the first woman Chief of a University Pediatric Ward. Borrino had a excellent career path, later she was appointed professor and director at Sassari and Perugia where she was director of the wars until the Academic year 1949/1950 when she had to leave due to age limits. While Borrino was in Siena, she did fantastic research especially regarding artificial breastfeeding and the addition of acid, but the focus of her studies were children's health issues given by the social issues created by the war. Borrino particularly studied tuberculosis, rachitis, nutritional deficiencies, social and psychological issues of orphans. In January 1921 established in Siena the Committee to help orphans. We can see the strong relationship she had with the city of Siena by looking at the numerous research proposed and published at the Academy of the Fisiocritici of Siena. In 1923 Borrino published, always in Siena, at the Typographical Society of Siena, the second improved edition of her book Manuale di terapia infantile (Guide to the infant's therapy).

CONCLUSIONS

The Pediatric Clinic of Siena was heavily hit by the Great War and the only one to lose its director. This study also wants to show the will of the Academic institutions to keep pediatric studies alive. This is especially shown with the rapid substitution of Gagnoni, after his death, with Borrino, an innovative and forward looking person. Generally there was indeed a big loss of pediatricians, especially from University, because many until 1920 were still enrolled in the army. Therefore they named a woman. Menabuoni's sacrifice offers an interesting addition to understand the context in which the Italian military healthcare worked, while Borrino's work help us to understand the social and welfare issues of childhood after the war.

REFERENCES

• La scuola medica da campo a San Giorgio di Nogaro. Bologna: Cappelli, 1918.

• Allaria GB. La Pediatria in Italia. Milano: Ufficio Stampa Medica Italiana, 1937, pp. 298-303, 351-3.

• Bracci R. Siena. Riv It Pediatr. 1993;19(Suppl 1):18-21.

• Farnetani I. Storia della pediatria italiana – Le origini: 1802-1920. Afragola: Etica Grafica, 2008, pp. 81-2, 109-20.

• Farnetani I, Farnetani F, Masetti A. Contributo dei Medici Militari Pediatri alla Prima Guerra Mondiale. G Med Mil. 2010;160:37-52.

Farnetani I. L'Università castrense nel contesto della sanità militare italiana.

Mem Sci Giuridiche Lett – Accad Naz Sci Lett Arti Modena. 2016;8(19):563-8.
Ferrajoli F. Il servizio sanitario militare nella guerra 1915-1918 (nel cinquantenario della vittoria). G Med Mil. 1968;118:501-16.

ABS 11

BRONCHOPULMONARY DYSPLASIA IN A CO-HORT OF EXTREMELY LOW GESTATIONAL AGE NEWBORNS (ELGANs): 5 YEARS OF EXPERIENCE

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INTRODUCTION

Despite the advances in neonatal care, bronchopulmonary dysplasia (BPD) remains one of the most frequent and serious complications of premature birth and has a significant impact in the quality of life of affected infants. Incidence is variable depending on the characteristics of the population studied and the definition used to make the diagnosis; Stoll in 2010 reported an incidence of 68% in infants born \leq 28 weeks of gestation according to NICDH criteria. The pathogenesis of disease is multifactorial with a wide variety of pre- and postnatal risk factors (**Fig. 1**). Currently there is no effective treatment: our only weapon of choice is prevention.

MATERIALS AND METHODS

The aim of this study is to analyze the management and the incidence of BPD in ELGANs (infants born ≤ 28 weeks of gestation), hospitalized between 2012-2016 in the Neonatal Intensive Care Unit of Cagliari and survived at 36 weeks PMA. We examined prenatal and postnatal risk factors, delivery room stabilization, transfer information, complications, treatments and outcomes at 18-24 month corrected age. Patients were studied retrospectively and divided into two groups: with and without BPD. The diagnosis of BPD was made according to NICDH criteria. RESULTS

We enrolled 86 patients ELGANs, 38 males (44%) and 48 females (56%); mean gestational age (GA) 26.9 (\pm 1.5 SD) w, mean birth weight 879 (\pm 217 SD) g. The incidence of BPD in this population was 52% (45 total cases): 17% mild, 4% moderate and 31% severe. Patients with BPD had significantly lower GA: 26.3 ± 1.6 vs 27.7 ± 0.8 weeks and a lower birth weight: 767 ± 174 vs $1,003 \pm 190$ g. All patients under 26 w GA developed BPD (65% in a severe form). In the group of BPD were present 24 males (53%) and 14 (34%) in controls. Patients with BPD showed a higher incidence of IUGR 16% vs 10%, pPROM 29% vs 22% and perinatal asphyxia; furthermore they required a longer duration of mechanical ventilation 12.7 \pm 12 vs 1.9 \pm 2.7 days, non invasive ventilation 71 ± 27 vs 34 ± 11 days, parenteral and enteral nutritional support. An extrauterine growth restriction has been reported in 60.5% of total patients with greater severity in the first month of life. Patients with BPD presented more frequent and severe complications: hemodynamically significant patent ductus arteriosus (44% vs 15%), retinopathy of prematurity (53% vs 10%), culture-positive sepsis (16% vs 12%), pulmonary hypertension (16% vs 0), severe brain injury (11%) vs 5%), pneumothorax (11% vs 5%), liver abscess (13% vs 0). ELGANs with BPD had a longer duration of hospitalization than patients without BPD (122 \pm 40 vs 70 \pm 11). Total mortality was 1.2% (only 1 patient > 36 w PMA died at 6 months postnatal age because of a severe form of BPD, pulmonary hypertension and CMV infection). Finally we examined 79% of the original sample at 18-24 months: a severe disability was found in 12% of them. This rate increases if we evaluate separately the subgroup of patients with a GA < 26 w PMA (29%) and infants with BPD (23%).

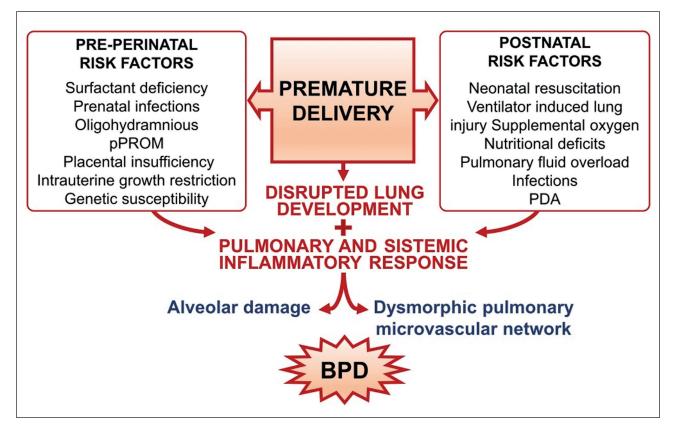


Figure 1 (ABS 11). Pathogenesis of bronchopulmonary dysplasia (BPD).

CONCLUSIONS

In our Department, BPD shows an incidence of 52%in infants born ≤ 28 weeks of gestation and it still represents an important cause of neonatal mortality and morbidity, prolonged hospital stay and severe long-term disability.

REFERENCES

• Collins JJP, Tibboel D, de Kleer IM, Reiss IKM, Rottier RJ. The Future of Bronchopulmonary Dysplasia: Emerging Pathophysiological Concepts and Potential New Avenues of Treatment. Front Med (Lausanne). 2017;4:61.

• Jobe AH, Bancalari E. Bronchopulmonary dysplasia. Am J Respir Crit Care Med. 2001;163:1723-9.

Stoll BJ, Hansen NI, Bell EF, Shankaran S, Laptook AR, Walsh MC, Hale EC, Newman NS, Schibler K, Carlo WA, Kennedy KA, Poindexter BB, Finer NN, Ehrenkranz RA, Duara S, Sánchez PJ, O'Shea TM, Goldberg RN, Van Meurs KP, Faix RG, Phelps DL, Frantz ID 3rd, Watterberg KL, Saha S, Das A, Higgins RD; Eunice Kennedy Shriver National Institute of Child Health and Human Development Neonatal Research Network. Neonatal outcomes of extremely preterm infants from the NICHD Neonatal Research Network. Pediatrics. 2010;126:443-56.

ABS 12

EXTREME THROMBOCYTOSIS IN A PRE-TERM TWIN: A CASE REPORT

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INTRODUCTION

Thrombocytosis is defined as platelet count more than 500×10^{9} /L and it is commonly classified as primary or secondary (reactive) thrombocytosis. Reactive thrombocytosis is a more common, and generally transient and benign condition in the paediatric age. The main causes identified are viral or bacterial infections (usually lower respiratory tract infection) or inflammatory condition. Surgical intervention and iron deficiency anemia are other possible condition associated [1]. Few literature focused on neonatal age is available, therefore neonatal platelet references ranges are not universally defined yet. Wiedmeier et al. recently pointed out that published reference ranges for neonatal platelet count may be imprecise, as physiological values can be higher as in neonate pre and post-natal age need to be considered [2]. Wang et al. identified a positively correlation of platelet count and white blood cell but an inversely correlation with red blood cells, most likely due to cytokines involved during inflammation [3]. Clinical

complication as bleeding and thromboembolism were never reported [1, 3].

CASE REPORT

We describe the case of a newborn female born at 29⁺³ weeks of gestational age by an emergency caesarean section for preterm labor and IUGR of this fetus in a dichorionic diamniotic (DCDA) twin pregnancy. The vagino-rectal tampon was not performed due to the gestational age, no other infectious risk was signaled during the pregnancy. At birth the neonate needed minimal resuscitation with continuous positive airway pressure (CPAP), maintained with a nasal mask for only one day during the hospitalization. The blood count at birth showed a thrombocytosis ($816 \times 10^{9}/L$) in absence of other blood alteration. At the following controls platelets value increased progressively till a maximum of 1,650 x 10⁹/L at 12 days of life. White blood cells had the same trend. For the persistence of thrombocytosis infectious investigations were performed (blood research of Epstein Barr Virus, Parvovirus B19, Toxoplasma DNA; urine research of Cytomegalovirus DNA; Adenovirus nasal sample): all resulted negative. The peripheral blood smear was obtained and the analysis showed big platelet but absence of atypical cells. Abdomen and cardiac echography were normal. The echoencephalography was frequently repeated, but no pathologic evidence was found. Histological analysis of placenta was also performed: the exam indicated focal membrane inflammation with no signs of chorioamnionitis.

After 20 days of life the platelet count decreased gradually reaching the value of 365×10^9 /L (in the same time white blood cells decreases) but after few days thrombocytosis recurred. Simultaneously with this evidence, the baby presented liquid and stinking stools. The Rotavirus research was found positive. The newborn maintained good clinical conditions, tolerated enteral nutrition and grew up. By now platelet count is 1,154 x 10⁹/L and the baby has been discharged from hospital with hematological follow-up.

CONCLUSIONS

The case we report is an unusual presentation of extreme thrombocytosis at birth. In consideration of the negativity of all the infective and hematological researches but considering also the presence of inflammation of amniotic membrane at the placenta examination, we can hypothesize that the early thrombocytosis was associated with the inflammation stress of delivery. The second episode, instead, can be realistically related with Rotavirus infection. The last but not the least fundamental consideration is that the newborn didn't report any haemorrhagic or thromboembolic complication, according to the literature available [1, 3] and no treatment was indicated. The case described and the literature suggests that high platelets count in newborns are frequently reactive and transient and that might not lead to major complications. Generally, platelets go down spontaneously in two weeks later. Nonetheless, more studies focused on the development of correct neonatal platelet reference ranges and on a more clear definition of the related condition with thrombocytosis should develop.

REFERENCES

[1] Wiedmeier SE, Henry E, Burnett J, Anderson T, Christensen RD. Thrombocytosis in neonates and young infants: a report of 25 patients with platelet counts of $\geq 1000000 \ \mu$ l⁻¹. J Perinatol. 2010;30:222-6.

[2] Wiedmeier SE, Henry E, Sola-Visner MC, Christensen RD. Platelet reference ranges for neonates, defined using data from over 47 000 patients in a multihospital healthcare system. J Perinatol. 2009;29:130-6.

[3] Wang IL, Huang LT, Wu KH, Lin HW, Ho MY, Liu HE. Associations of reactive thrombocytosis with clinical characteristics in pediatric diseases. Pediatr Neonatol. 2011;52:261-6.

ABS 13

USE OF PARACETAMOL IN PATENT DUCTUS ARTERIOSUS CLOSURE: THE EXPERIENCE OF THE NEONATAL INTENSIVE CARE UNIT OF CAGLIARI

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INTRODUCTION

Patent ductus arteriosus (PDA) often complicates the clinical course of preterm infants, increasing the incidence's risk of other diseases as chronic lung disease (CLD), necrotizing enterocolitis (NEC), and intraventricular hemorrhage (IVH) [1]. It's now known that the main drugs use to induce the closure of PDA, ibuprofen and indomethacin, have important side effects due to inhibition of prostaglandin synthesis, include transient renal failure [2], gastrointestinal bleeding and perforation and reduced brain flow [1]. The contraindications for ibuprofen or indomethacin administration include thrombocytopenia, intracranial hemorrhage, renal failure, necrotizing enterocolitis, co-administration of corticosteroid (risk of intestinal perforation) or hyperbilirubinemia [3]. Ibuprofen, a more recent is as effective as indomethacin has fewer renal sides effects [1]. Recent studies show the efficacy of paracetamol in PDA closure in preterm infants with poor side effects, but currently this drug is not the first choice: its use is limited to patients with contraindications to therapy with NSAIDs [4]. Despite the mechanism of action is not clear it seems that paracetamol acts on peroxidase segment of the PG synthetase by inhibition of prostaglandin synthesis [5, 6].

OBJECTIVE

Our NICU started using paracetamol in the treatment of PDA since 2014, for babies with contraindications to use of ibuprofen. The purpose of this study is to evaluate the efficacy of this drug in PDA closure. MATERIAL AND METHODS

Seventeen preterm infants were included (13 females and 4 males). Three of them were treated with oral administration and fourteen by intravenous route; the dosage was 15 mg/kg every 6 h in both of them. The mean gestational age (GA) was 25.8 \pm 2.1 w; the main weight was 777 g, the main age at the beginning of treatment was 7.6 days and the average time of treatment was 6.4 days. Two infants were African and the others were Caucasian. Paracetamol was effective on closure of PDA in 11 newborns (9 females and 2 males). In two of them the drug was administered by oral route. The mean GA was 23 ± 1.7 w; the main weight was 819 g, the main age at the beginning of treatment was 7 days and the average time of treatment was 7.5 days. In no responder newborns (5 babies, 3 females and 2 males) we gave ibuprofen. In one of them, paracetamol was administered by oral route. The mean GA was 26 ± 2.6 w; the main weight was 808 g, the main age at the beginning of treatment was 7.1 days and the average time of treatment was 8.6 days. Both African babies didn't respond to the only use of paracetamol. In one newborn (female) neither paracetamol or ibuprofen have been effective, so we administered indomethacin.

RESULTS

Paracetamol was effective alone in 11 newborns (64.7%), 5 patients (29.4%) also needed ibuprofen therapy and in 1 patient (5.8%) we had to use both ibuprofen and indomethacin. In our study, there were no adverse effects for the use of paracetamol (liver function abnormality).

CONCLUSIONS

The results of our study are promising for the safe and efficacy of this drug, but further studies are needed to understand if the clinical efficacy of paracetamol on PDA closure may depend on gestational age, age

at the beginning of treatment, duration of treatment, route of administration and ethnic group. REFERENCES

[1] Ohlsson A, Walia R, Shah S. Ibuprofen for the treatment of patent ductus arteriosus in preterm and/or low birth weight infants. Cochrane Database Syst Rev. 2015;(2):CD003481.

[2] Lin YJ, Tsai YJ, Chen JS, Lin JS, Wu JM, Lin CH, Yeh TF. Renal effects and urinary excretion of prostaglandin following indomethacin therapy in premature infants with patent ductus arteriosus. Zhonghua Min Guo Xiao Er Ke Yi Xue Hui Za Zhi. 1995;36(2):104-7.

[3] Ohlsson A, Shah PS. Paracetamol (acetaminophen) for patent ductus arteriosus in preterm or low-birth-weight infants (Review). Cochrane Database Syst Rev. 2015;(3):CD010061.

[4] Dang D, Wang D, Zhang C, Zhou W, Zhou Q, Wu H. Comparison of Oral Paracetamol versus Ibuprofen in Premature Infants with Patent Ductus Arteriosus: A Randomized Controlled Trial. PLoS One. 2013;8(11):e77888.

[5] Allegaert K, Anderson B, Simons S, van Overmeire B. Paracetamol to induce ductus arteriosus closure: is it valid? Arch Dis Child. 2013;98(6):462-6.
[6] Bardanzellu F, Neroni P, Dessì A, Fanos V. Paracetamol in Patent Ductus Arteriosus Treatment: Efficacious and Safe? Biomed Res Int. 2017;2017:1438038.

ABS 14

A CASE OF HEMOPHAGOCYTIC LYMPHO-HISTIOCYTOSIS TRIGGERED BY A CYTO-MEGALOVIRUS INFECTION

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INTRODUCTION

Hemophagocytic lymphohistiocytosis (HLH) is a rare condition characterized by a hyper inflammatory response and aberrant activation of lymphocytes and macrophages resulting hypercytokinemia and loss of immune normal function. HLH can be genetically determinate or secondary to other conditions like infections, the most common but not only of which is an Epstein-Barr virus infection. Diagnosis and treatment have been standardized in the HLH-2004 protocol.

CASE REPORT

A 3-year-old male patient came to our Pediatric ER showing fever since the last 8 days, with remittent pattern in spite of the antibiotic therapy (amoxi-cillin/clavulanic acid). Born to consanguineous

	Hb (g/L)	WBC (#/mm³)	PLT (#/mm³)	LDH (U/L)	CRP (mg/dL)	Ferritin (ng/mL)	Fibrinogen (mg/dL)	AST/ALT	γ-GT
Admission	10.8	1,750	113,000	838	75.6			156/87	
+3 days	11.3	2,270	92,000		77.8				
+6 days	9.7	2,210	107,000	976	88.5	6,566	225	151/83	152

Table 1 (ABS 14). In-hospital clinical chemistry at the beginnning.

parents he had several congenital abnormalities (perimembranous ventricular septal defect, left agenesis and right hypoplasia of the 1st metacarpus, bilateral agenesis of the radium, triangular face). At the admission he appeared suffering, with a diffuse rash, and $T = 39.8^{\circ}C$. Several alterations of the clinical chemistry emerged after blood collection (Tab. 1). Conversely, blood cultures and chest X-ray were negative. Cytomegalovirus (CMV) infection was evidenced by PCR (DNA 267,026 copies/ml). The antibiotic therapy was switched to a 2nd line association due to the hypothesis of a bacterial superinfection but neither the lab data nor the clinical pattern improved. Eventually, the extension of exams showed both fibrinogen consumption, and high levels of ferritin among others. Abdominal US showed splenomegaly. Thus HLH secondary to CMV infection was suspected despite the lack of all criteria for diagnosis. The child was transferred to a tertiary Center for referral where methylprednisolone therapy was started after bone marrow aspiration (hypoplasia of the erythroid series and hyperplasia of granuloblastic series, without hemophagocytosis). The same time genetic tests excluded a familial type of HLH. Some days later the remnant criteria (hypertriglyceridemia, and cytopenia of 3 lineages in the peripheral blood) for appropriate diagnosis emerged some days afterward and the complete protocol (HLH-2004) was started. In round 7 days both the clinical pattern and lab data improved. The child was discharged after 45 days on therapy with steroids and etoposide. He was sent to a shared follow-up between the two Centers involved. At present the conditions are stable and the cycles of therapy are continuing with good response. CONCLUSIONS

Our case shows how HLH diagnosis becomes a diagnostic possibility when the clinical pattern seems to refer to a systemic infection not-responding to antibiotic therapy, severe viral infection, persistent fever during autoimmune/ autoinflammatory disease or cytopenia without blasts in bone marrow. Ferritin levels can help to diagnose HLH earlier and address towards

more extended hematologic analysis. According to literature early diagnosis and appropriate management in secondary HLH are pivotal and can be helpful in leading a complete recovery despite the severity of the condition.

ABS 15

GIOVANNI BUCCI (1928-2016) AND ANTONIO MARINI (1931-2006), LEADING SCHOLARS AND FOUNDING FATHERS OF NEONATOLOGY IN ITALY

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Working group on the History of Pediatrics of the Italian Society of Pediatrics

INTRODUCTION

The Working group on the History of Pediatrics of the Italian Society of Pediatrics wishes to commemorate Giovanni Bucci and Antonio Marini, who may rightfully be considered pioneers of modern Neonatology and Neonatal Intensive Care in Italy. This short contribution aims to stimulate data collection to start an appropriate and objective biographical testimony of the over 50 years of scientific and cultural activity of Giovanni Bucci and Antonio Marini, who were both never-to-beforgotten models for research, clinical care and the academia.

METHODS

This contribution was developed by the Authors through the following steps:

- 1. search in public and private documentary sources;
- 2. interviews with family, friends and colleagues of both Giovanni Bucci and Antonio Marini;
- 3. professional acquaintance of one of the Authors (LC), who worked as treasurer of the board of the Italian Society of Pediatrics during the presidency of Giovanni Bucci (1994-1997) and Antonio Marini (1997-2000). Furthermore, the friendship of the Author (LC) with both Professors allowed him to witness their last four decades of activity.

RESULTS

Documents and testimonies confirmed the contribution that both Giovanni Bucci and Antonio Marini gave to research, education and progress of clinical care in Neonatology and Perinatal medicine. Giovanni Bucci completed his specialty training with Maggioni in 1953 and moved to Boston from 1959. Following his return to Rome in 1961 he became the Director of the Department for preterm newborns of the Pediatric Clinic directed by Arrigo Colarizi (1960-1973). He was firmly resolute in tackling the issue of the treatment of respiratory failure and the challenge of mechanical ventilation in preterm newborns. He cared for five preterm newborns in very severe conditions; four weighted less than 1,500 g, one weighted 2,400 g and was the only one to survive. Autoptic examination showed evidence of severe pneumonia in two of the four newborns who died despite mechanical ventilation. As Professor Bucci explained to the Authors, these were the first attempts at mechanical ventilation in patients in extremely severe conditions: suffice to say that these attempts used to be carried out in the underground floors of the San Matteo Hospital in Pavia, with an Angstrom machine for adults.

Antonio Marini showed to be a motivated researcher in Perinatal medicine when he was still very young: he was only 27 years old when he won the Pfizer award for research (25 September 1958) for his important research on animal newborns. He completed his specialty training in Pediatrics in Bologna (1957) and was later researcher in Pediatric cardiology at the Karolinska Institute in Stockholm (1957-1960). He then worked at the Children's Hospital in Pittsburg. Both the Swedish and the American experiences were fundamental to lay the foundation to the international relationships which he cultivated and put to use in later years. Afterwards he worked with Schwartz-Thiene in Ferrara, then he moved to Pavia in 1961. He finally moved to Milan. He became "aiuto pediatra incaricato" at the end of 1963, then "professore incaricato" of Neonatal Pathology (1972-1980) and finally full professor of Neonatology (1980-2003). His scientific contribution was always continued and effective. His efforts in research, clinical care and education were captivating. His exuberant personality and his ever up-to-date and vast knowledge in fetal and neonatal physiopathology used to allow him to contribute with competence in all areas of Perinatal medicine, Neonatology, as well as other disciplines. Antonio Marini was a charismatic figure and he organized countless national and international workshops and

conferences in his career. He was able to involve both Italian and international neonatologists, as he did in occasion of a well-known workshop which was held in Egypt in 1988. Some of his apprentices merited the direction of important Neonatology Departments in Milan, and one of his apprentices followed his steps in taking over his Professorship.

CONCLUSION

Giovanni Bucci and Antonio Marini well deserve the deep gratitude of those who appreciated their enthusiasm and their contribution to the development of Italian Neonatology. Their work as researchers, clinicians and educators will always remain an example to be followed by Italian neonatologists and medical doctors.

REFERENCES

• Bucci G. Mechanical respiration with intermittent positive-negative pressure in therapy of respiratory insufficiency in the premature infant. Minerva Pediatr. 1960;12:97-9.

• Bucci G. Personal communication, 2012.

 Bucci G. Racconti Romani di Terapia Intensiva Neonatale (1961-2001). In: Cataldi L, Gregorio MG (Eds.). Atti del Convegno del GSSP; Rome, 1 March 2013.

• Cataldi L. Giovanni Bucci e la IVPP nella terapia dell'insufficienza respiratoria del prematuro. Poster at the 18th National Conference SIN; Rome, 3 October 2012.

 Cataldi L. Antonio Marini, una vita per il progresso della ricerca medica per la salute del bambino. Poster at the 63th National Conference SIP; Pisa, 18-20 October 2007.

- Colarizi P. Commemoration of Giovanni Bucci; Rome, 6 December 2016.
- Marini L. Personal communication, 2007.
- Rinaldi G. Vita di un reparto per la vita. Foggia: Ed. TIN Foggia, 2010.
- Sereni F. Commemoration of Antonio Marini; Milan, 18 October 2006.

ABS 16

A FISH... OUT OF WATER

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INTRODUCTION

The word ichthyosis comes from the Greek root ichthys, meaning fish, (referring to the cutaneous scaling that is characteristic of these disorders) which is said to resemble the scales of a fish. Scaling can be localized or generalized and can be associated with a variety of additional cutaneous and/or systemic manifestations. The ichthyoses represent a large group of cutaneous disorders linked by the common finding of abnormal epidermal differentiation. In patients with ichthyosis, the barrier function of the skin is compromised. Consequences can be particularly dangerous, at times life-threatening, with increased susceptibility to infections, secondary to impaired skin integrity and dramatically increased metabolic demands due to increased epidermal turnover and evaporative heat and water loss [1].

CASE REPORT

B., a female infant, was born at term by spontaneous delivery after uneventful pregnancy. At birth, she was small for gestational age with Apgar score 9-10. On admission, her skin was erythrosic and shiny, taut membrane covered her entire body; she presented swollen hands and feet, stiff fingers and crumpled ears. She had good reactivity and vitals signs were stable. The results of the laboratory tests, including complete blood count, liver and kidney function tests, along with the measurement of serum electrolytes were all within normal limits, except hemogasanalysis that showed acidosis and hypocalcemia. X-ray showed a hypoplasia of feet's distal phalanges with membranous syndactilia and arthrogryposis of the right hand's fingers. In the first day of hospitalization dermatologists confirmed collodion baby diagnosis. The patient started supportive and antibiotic treatment. She was placed in a humidified incubator to prevent hypothermia and dehydration. The fluid and electrolyte balance was maintained. Skin hydration was ensured by daily bathing with only water or mild cleanser and emollients application. The second dermatological consult underlined right wrist, ankle and interdigital spaces fissurings, maceration in axillary and popliteal regions but with a generalized improvement of edema. Skin swabs were taken with positivity for S. haemolyticus and K. pneumoniae. On the 19th day of life, she was discharged with unchanged therapy. The baby was then referred to a specialized hospital for further management (genetic tests and skin biopsies that confirmed lamellar ichthyosis).

CONCLUSIONS

Lamellar ichthyosis is an autosomal recessive disorder with variable expressivity. It is the most rare and severe form of ichthyosis, which is invariably present at birth and has an incidence rate of approximately 1: 600,000 live births [2]. Although these disorders are fairly rare, it is important for the neonatologist to have some familiarity with them, as the perinatal period and early infancy represent particularly critical stages for patients with ichthyosis.

REFERENCES

 Craiglow BG. Ichthyosis in the newborn. Sem Perinatol. 2013;37(1): 26-31. [2] Bangal VB, Gangapurwala S, Gavhane SP, Gupta K. Rare Case Report – Neonatal Lamellar Ichthyosis in Newborn – Collodion Baby. Int J Biomed Adv Res. 2014;5(2):120-2.

ABS 17

ULTRA LATE ONSET DISEASE PRESENTING WITH GROUP B STREPTOCOCCUS MENINGITIS IN A PRETERM TWIN: A CASE REPORT

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INTRODUCTION

Group B Streptococcus (GBS) infection in infants older than 90 days is classified as ultra-late onset disease (ULOD). ULOD is extremely rare and may be nosocomial or community-acquired. Literature data suggest that highly virulent strains, underlying host disease and immunological factors increase the risk of this infection. Prematurity seems to be significantly associated with ULOD.

CASE REPORT

A 120-day-old boy was admitted to our Clinic with a history of fever for 10 hours. He was a diamniotic, dichorionic twin born at 35 weeks of gestation by emergency cesarean section (birth weight, 2,505 g; length, 44 cm; head circumference, 33 cm). At birth, he was admitted to the Neonatal Unit for his prematurity. Maternal GBS cultures from the vagina and rectum were negative at delivery. On admission to hospital, the infant presented with fever (38.4°C), irritability, normal anterior fontanel, skin hypoperfusion, normal oxygen saturation, tachypnea, and good muscle tone. Initial laboratory tests detected mild neutrophilic leukocytosis and moderate increase in inflammatory markers (WBC, 21.96 x 10⁹/L; Neutrophils, 67%; C-Reactive Protein, 1.33 mg/dL). Chest X-ray was negative. About 10 hours after admission, clinical conditions worsened with bulging anterior fontanel, nuchal and limb stiffness, therefore the lumbar puncture was performed. The cerebrospinal fluid (CSF) was turbid and showed pleocytosis and increased protein level (54 mg/dL). Laboratory examinations showed a considerable inflammatory markers increase (WBC, 35.4 x 10⁹/L; Neutrophils, 56%; C-Reactive Protein, 18.66 mg/dL).

Clinical and laboratory features were suggestive of the diagnosis of meningitis, and therefore, an empirical treatment with ceftriaxone, vancomycin and netilmicin, combined with dexamethasone, was started. On the third day of hospitalization, fever decreased, and clinical and laboratory features started to improve. The CSF culture was found to be negative, while blood culture was positive for Str. agalactiae. This bacterial agent was sensitive to ceftriaxone, which, accordingly, was continued as the sole antibiotic until day 18. On day 16, laboratory parameters were normalized, and the infant was discharged 3 days later. Subsequent follow-up examinations have not revealed any neurological sequelae. During the patient's hospital stay and in the weeks after discharge, the co-twin was in good health, despite the exposure to infectious sources had been common to both twins.

CONCLUSIONS

ULOD is an extremely rare form of GBS infection, whose pathogenesis is not well understood. This case of ULOD presented with meningitis, which was probably community-acquired, and had a favorable evolution. The present case together with the medical history of the co-twin, who did not develop any infection, support the hypothesis that genetic factors influence the individual's susceptibility to the ULOD.

ABS 18

NEONATAL HYPOTONIA: A CASE OF SEVERE CONGENITAL MYOPATHY

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INTRODUCTION

Neuromuscular diseases frequently present in newborns or infants with hypotonia and weakness. However, these signs also occur in many common disorders such as sepsis, organ failure, and metabolic dysfunction. If these latter conditions can be excluded, hypotonia probably is caused by a primary neuromuscular disorder.

CASE REPORT

A one-day-old baby girl, born by a no consanguineous Sardinian marriage, was admitted with symptoms of severe congenital hypotonia and respiratory distress. She was born at full term with Apgar Score 7 (1') and 7 (5') by an emergency caesarean section because of polyhydramnios and reduced fetal motility. Birth weight was 2.670 kg (9th percentile), birth length was 53 cm (99th percentile). There was no family history of hypotonia or genetic syndromes, except for the mother who likely suffers from Ehlers-Danlos syndrome. On examination, child was found to be macrocephalic with head circumference 35.5 cm (92nd percentile) and hoarse cry, with dysmorphic notes (micrognathia, unique palm groove, long fingertips). Blood tests, karyotype, urinalysis, ABR, view exam, echocardiography and abdomen ultrasound were normal. Genetic tests for SMA syndrome and Prader-Willi/Angelman syndrome were negative. Cranium ultrasound detected asymmetry of brain ventricles, so she performed also a brain MRI that showed ventriculomegaly and reduced myelination for age. EEG showed unorganized pattern for age, without clear abnormalities. Neurologic exam described severe hypotonia and hyporeflexia with discrete reactivity to sensory stimuli, suspected for congenital myopathy. EMG detected an absence of spontaneous activity. Muscle biopsy was compatible with unspecified congenital neuromuscular disease. Currently genetic investigations have been carried out on the DNA of the patient and parents, still ongoing.

CONCLUSIONS

Neonatal hypotonia diagnosis is complex and requires multidisciplinary team and examinations. Actually the child is 10 months old and shows moderate hypotonia, treated with daily physiotherapy. She presents also a psychomotor retardation, and she is fed by percutaneous endoscopic gastrostomy. 2 months ago she performed a Nissen stomach fundoplication because of severe stomach reflux.

REFERENCES

 Bodamer OA, Miller G. Approach to the infant with hypotonia and weakness. Available at: http://www.uptodate.com/contents/approach-to-theinfant-with-hypotonia-and-weakness, date of publication: October 2015, last access: September 2017.

http://www.adhb.govt.nz/newborn/Guidelines/Neurology/Hypotonia.
 htm, last access: September 2017.

ABS 19

PERSISTENT FEVER: DO NOT THINK TO EXOTIC DISEASES ONLY, PUT A PROBE ON THE HEART!

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INTRODUCTION

Ebstein Barr Virus (EBV) infection may affect several organs, among them the heart and mainly pericardium and myocardium. We present a child with EBV infection and pericarditis.

CASE REPORT

Male, 21 months; he arrives to the emergency room due to persistent fever (39.9°C) since 10 days. At physical examination: vital parameters with mild tachycardia and blood pressure at the upper normal limits, edema on bilateral eyelids, rhinitis, chest and cardiac physical examination within normal limits. Splenomegaly (+1 cm), mild enlargement of axillary, lateral cervical and supraclavicular lymph nodes, sovraclavear; hyperemic tympanic membranes and oropharynx. Laboratory tests: WBC 10,450/mmc (Neutrophils 29.1%, Lymphocites 66.3%, Monocytes 3.1%), Hb 9.2 g/dL, MCV 77 fL, Platelet 110,000/mmc, LDH 2,394 u/L, CRP 1.7 mg/dL, ESR 31 mm; kidney function tests, C3, C4 and ASLOT within normal limits; pathologic proteinuria below nephritic range, AST 336 U/L, ALT 160 U/L, GGT 206 U/L; coagulation tests and serum protein normal. Toxoplasma and CMV Ab negative; EBV Ab without seroconversion. Chest X ray, abdominal and tissues ultrasound: reactive lymph nodes. At 10 days of fever: suspecting Kawasaki, IGIV were given; at cardiac ultrasound: left ventricle thickening and pericardium effusion; ECG, troponin normal; serum Coxsackie, Echovirus and Influenza A and B Ab negative; Serum EBV-DNA positive (21,070 viral copies/ml). Therefore Aspirin therapy (80 mg/kg/die) was started. At later controls, progressive clinical and ultrasound resolution of pericardium effusion and ventricle thickening were observed. After six weeks of treatment, Aspirin therapy was discontinued. CONCLUSIONS

In the differential diagnosis of persistent fever, ultrasound is useful not only to rule out complications of Kawasaki disease, but also to diagnose a possible viral myo-pericardium involvement, in order to promptly start aspirin to reduce the risk of longterm complications.

ABS 20

A STRIDOR THAT YOU DON'T EXPECT!

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INTRODUCTION

The vascular rings represent less than 1% of congenital heart disease. There are two types, complete and incomplete, depending on if the vascular anomalies completely encircle the trachea and esophagus or not. The 95% of rings are formed by double aortic arch, sometimes with other cardiac abnormalities, and by right aortic arch with a left ligamentum arteriosum. In 50% of cases there are other comorbidities, like trisomy 21, 22q11 deletion syndrome, anal atresia, tracheal stenosis, congenital lobar emphysema and congenital conotruncal cardiopathies. The symptoms are caused by extrinsic compression of vascular ring on airways and digestive tract. The respiratory symptoms are more frequent (70-95%), mainly wheezing, stridor, respiratory distress, cough and recurrent infections of upper airways; gastrointestinal symptoms are reported in 5-50% of cases, generally dysphagia, vomit and difficult weight gain.

CASE REPORT

C. is a 24-hours-old baby, born by eutocic delivery after normal pregnancy, Apgar score 10-10. She arrives to our Department because of suddenly onset of tirage and cornage, without cyanosis or desaturations; the cornage is better heard when the baby is calm, while it decreases during crying. Blood tests are normal, like chest radiography. For the suspect of laryngomalacia, we request laryngoscopy, in the norm. So, we suspect a tracheal compression by abnormal vessels and we ask a cardiological examination. The echocardiography shows a muscular interventricular septal defect and abnormal shunt with right aortic arch in suprasternal projection (Fig. 1). The next step is magnetic resonance. It confirms the presence of complete vascular ring (double aortic arch) encircling trachea and esophagus; the right arch is dominant and above the left one. The epiaortic vessels originate separately in each side (four vessel sign). The baby is sent to a cardiac surgery center, where receives corrective intervention: section of posterior arch that has also a coartaction at the connection with the anterior one. She is extubated at the first postoperative day, but at the second day she has pleural effusion; the analysis reveals that it's chylothorax. She is fed with nolipids milk and progressively improves; she takes furosemide 3 mg for 3 times/day and she will stop it after 2 months.

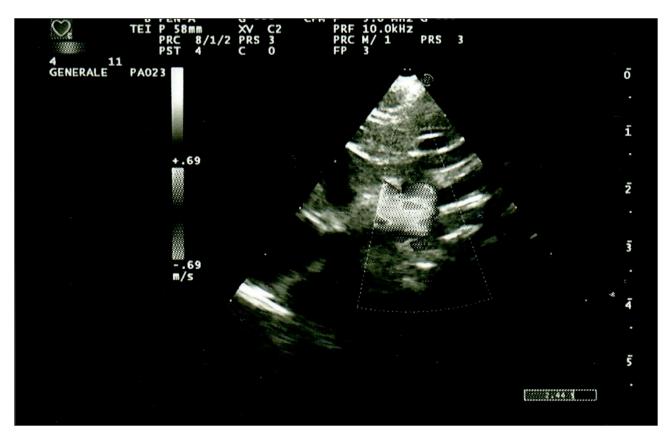


Figure 1 (ABS 20). In suprasternal projection, the echocardiography shows abnormal shunt with right aortic arch.

CONCLUSIONS

The vascular rings are extremely rare disease, often burden by diagnostic delay especially in newborns with congenital stridor and respiratory distress. They can be asymptomatic lifelong, but sometimes the early diagnosis and surgical intervention (with excellent results) can improve the symptoms and reduce the subsequent complications, also the iatrogenic ones.

ABS 21

METABOLOMICS OF HUMAN BREAST MILK: PRELIMINARY RESULTS ON THE IMPOR-TANCE OF SECRETOR PHENOTYPE

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INTRODUCTION

Human breast milk is a complex fluid that has developed to satisfy the nutritional requirements of infants. It contains proteins, lipids, carbohydrates, vitamins, minerals, and biologically active components [1], such as growth factors, antimicrobial factors and stem cells [2]. Human milk changes in composition during the lactation, and varies within feeds, diurnally and between mothers. The most variable components are the human milk oligosaccharides (HMO) the third largest component of human milk. They have several functions, including prebiotic and antimicrobial effects, and are involved in immune modulation [3]. These HMOs are complex and unconjugated sugars containing a lactose core bound to ≥ 1 glucose, galactose, N-acetylglucosamine, fucose, and sialic acid residues. More than 200 different oligosaccharide structures have been found and it is known that every woman synthesizes different subsets of oligosaccharides. The HMO composition mirrors blood group characteristics, which depend on the expression of certain glycosyltransferases. Four milk groups can be assigned based on the Secretor (Se) and Lewis blood group gene (Le). The Se gene encodes for the α 1-2-fucoslyltransferase (FUT2), the Le encodes for the α 1-3/4-fucosyltransferase (FUT3). Individuals

with an active Se locus are classified as Secretors, and their milk is abundant in 2-fucosyllactose (2 FL), lacto-N-fucopentaose I (LNFP I) and other α 1-2fucosylated HMOs. In contrast, non-Secretors lack a functional FUT2 enzyme and their milk does not contain α 1-2-fucosylated HMO [4]. Metabolomics in Neonatology may be useful to investigate the effects of the breast milk in several aspects of neonatal metabolism [5]. It is based on the systematic study of the complete set of metabolites in biological samples [6]. The metabolome of a sample can be viewed as a metabolic "fingerprint", representative of the state of the organism at that time. This technology consists of two steps: an experimental technique, based on mass spectrometry or nuclear magnetic resonance (NMR) spectroscopy, designed to profile low molecular weight compounds, and multivariate data analysis. **METHODS**

The aims of the present study were to study the metabolic profile of samples of human milk from 60 lactating women and compare the results with previous works concerning metabolomics in human milk. 1-HNMR spectroscopy was used to analyse the samples, then principal component analysis was performed in order to identify differences between the samples. Milk samples were obtained from 60 women enrolled in the Alexandra General Hospital of Athens, who gave birth to 48 appropriate for gestational age, 2 large for gestational age and 10 intrauterine growth restricted infants.

RESULTS

3D-Principal component analysis (PCA) identified two clear groups of patients distinguishable on the basis of milk oligosaccharide concentration, who were classified as secretors or non-secretors of fucosyltransferase 2 gene products, according to the concentration of 2-fucosyllactose, lactodifucotetraose and lacto-N-fucopentaose I. Secretor genotype was consistent with expected frequencies, implying protection from neonatal diarrhea due to *E. coli*.

CONCLUSIONS

Together with genomics and proteomics, metabolomics appears to be a promising tool in Neonatology for the identification of biomarkers as early predictors of outcome, the diagnosis and monitoring of various diseases and the "tailored" management of neonatal disorders.

REFERENCES

[1] Jenness R. The composition of human milk. Semin Perinatol. 1979;3(3):225-39.

[2] Hassiotou F, Hartmann PE. At the Dawn of a New Discovery: The Potential of Breast Milk Stem Cells. Adv Nutrition. 2014;5(6):770-8. [3] Thurl S, Munzert M, Henker J, Boehm G, Müller-Werner B, Jelinek J, Stahl B. Variation of human milk oligosaccharides in relation to milk groups and lactational periods. Br J Nutr. 2010;104(9):1261-71.

[4] Bode L. Human milk oligosaccharides: Every baby needs a sugar mama. Glycobiology. 2012;22(9):1147-62.

[5] Marincola FC, Dessì A, Corbu S, Reali A, Fanos V. Clinical impact of human breast milk metabolomics. Clin Chim Acta. 2015;451(Pt A):103-6.

[6] Nicholson JK, Connelly J, Lindon JC, Holmes E. Metabonomics: a platform for studying drug toxicity and gene function. Nat Rev Drug Discov. 2002;1(2):153-61.

ABS 22

LONGITUDINAL WEIGHT GAIN FROM BIRTH TO 18-24 MONTHS CORRECTED AGE IN 66 EXTREMELY LOW GESTATIONAL AGE NEW-BORNS (ELGANS) WITH AND WITHOUT BRON-CHOPULMONARY DYSPLASIA

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INTRODUCTION

To achieve an optimal postnatal growth is a very important and challenging goal in the management of extremely preterm infants. This is especially complex for infant who develop bronchopulmonary dysplasia (BPD) because of increased energy expenditure due to the work of breathing, stress, inflammation and medications: they present a postnatal growth failure in 30-67% of cases. In addition suboptimal nutrition and inadequate weight gain are linked to adverse clinical and neurodevelopmental outcomes.

MATERIALS AND METHODS

Growth was retrospectively assessed for 66 infants born with a gestational age (GA) \leq 28 weeks of gestation hospitalized between 2012-2016 in the Neonatal Intensive Care Unit (NICU) of Cagliari and survived at 36 weeks postmenstrual age (PMA). Anthropometric measures (body weight, length, head circumference) were performed from birth until 18-24 months corrected age. Growth data were extracted from medical records and converted to age-adjusted percentiles using the INeS growth Charts (< 42 w PMA) and Centers for Disease Control and Prevention 2000 growth chart (0-3 years). We also compared growth between infants with and without BPD.

RESULTS

The mean gestational age was 27 ± 1.3 weeks and the mean birthweight was 902 ± 207 g; 45.5% of them developed BPD. At birth, 9% of infants were small for gestational age (SGA); during hospitalization rate of PGF increased from 42.4% in the first month of life to 53% at 36 weeks' PMA, and slightly decreased at 42 w (43.9%). Comparisons between groups of infants shows that preterm infants who developed BPD had more frequent and severe PGF than infants without BPD; moreover, they acquired later nutritional autonomy: mean duration on tube feedings was 91 vs 53 days. Fig. 1 shows the trend of percentiles during the time; in the first month occurred an important loss of the mean weight percentile (more evident for patients with BPD) that continued until 36 weeks. After discharge we observed a progressive gain of percentile and at 18-24 months of corrected age mean values are between the 22nd and 25th percentiles for BDP patients and controls. A similar trend was observed for length and head circumference measured at birth, at 3-6 months and at 18-24 months corrected age. CONCLUSIONS

Achieving adequate growth remains a complex objective to reach, especially in BPD patients. We intend to develop specific protocols to optimize nutrition, improving care and nutritional strategies, in order to reduce the severe growth failure that we observed in our patients especially the first month of life and in more sick infants. These measures should confer some benefits for patients in order to improve short- and long-term complications and a to achieve a better neurodevelopmental outcomes. REFERENCES

• Ehrenkranz RA, Dusick AM, Vohr BR, Wright LL, Wrage LA, Poole WK. Growth in the neonatal intensive care unit influences neurodevelopmental

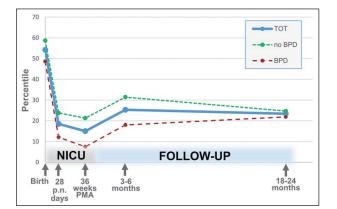


Fig 1 (ABS 22). Longitudinal weight gain from birth to 18-24 months corrected age.

BPD: bronchopulmonary dysplasia; PMA: postmenstrual age.

and growth outcomes of extremely low birth weight infants. Pediatrics. 2006;117(4):1253-61.

 Guimarães H, Rocha G, Guedes MB, Guerra P, Silva AI, Pissarra S. Nutrition of preterm infants with bronchopulmonary dysplasia after hospital discharge. J Pediatr Neonat Individ Med. 2014;3(1):e030116.

 Martin CR, Brown YF, Ehrenkranz RA, O'Shea TM, Allred EN, Belfort MB, McCormick MC, Leviton A; Extremely Low Gestational Age Newborns Study Investigators. Nutritional practices and growth velocity in the first month of life in extremely premature infants. Pediatrics. 2009;124(2):649-57.

ABS 23

A CASE OF SUCCESSFUL PROPRANOLOL TREATMENT OF NEONATAL HEPATIC HEMAN-GIOMATOSIS

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INTRODUCTION

Hemangiomas are the most frequent benign vascular tumors of infancy and can be asymptomatic or need treatment for functional reasons and/or local complications/disfigurement; they usually proliferate during the first year of life and undergo involution after this period. Hemangiomatosis is cutaneous and/or visceral and can be characterized by focal, multifocal or diffuse lesions. Some complications are rarely associated, such as heart failure, liver disease, compartmental abdominal syndrome or consumptive hypothyroidism due to of 3-desiodase ectopic secretion. The first line treatment is pharmacological and the β -blocker propranolol is the drug of choice (which is effective, relatively safe and determines excellent response, especially under eight months of age); in case of failure, topical timolol or pingyangmycin or intravenous vincristine can be efficacious. Not pharmacological treatments include laser, surgery or cryosurgery and radiotherapy [1, 2]. CASE REPORT

S. is the 1st born of a twin pregnancy. An urgent caesarean section was made at 35 weeks of gestational age (weight 2,110 g, Apgar 9'-10'). During an abdominal ultrasound, performed for prematurity, it was occasionally discovered a circular angiomatic formation (5 x 5 mm) in the V hepatic segment (S5). At 7 weeks of age, she repeated abdominal

ultrasonography and multiple vascularized hepatic formations have been described. Abdominal MRI confirmed the hepatic polyangiomatosis and evidenced 10 circular formations (10-20 mm of diameter), the major localized in S8 (20 x 12 mm), S2-S3 (20 x 15 mm) and S6 (14 x 14 mm). The lesions resulted hypointense in T1, with centripetal enhancement after gadolinium. In the family of S., paternal grandmother and aunt are also affected by hepatic polyangiomatosis. By mutual agreement of the neonatologist, dermatologist and cardiologist, S. started a therapy with propranolol at the dose of 1 mg/kg/day in two daily administrations for the first week, to increase to 2 mg/kg/day in the second week and 3 mg/kg/day from the third. ECG, blood pressure and glycemia have to be evaluated during the therapy. After three wks from the beginning of the treatment, the lesions showed a slight dimensional reduction: the ultrasound examination after three months of propranolol showed a great therapeutic success, evidencing both a numerical and a dimensional reduction of hemangiomas to two residual lesions, the first located in S8 (6 mm) and the second in S6 (3.6 mm). S. is still assuming propranolol at the full dose; her blood pressure and glycemia always resulted in range and ECG monitoring has never showed alterations.

CONCLUSIONS

In the treatment of infantile hemangiomas, the early approach is fundamental for therapeutic success. The therapeutic effect of propranolol in reducing hemangiomas volume was serendipitously discovered in 2008. It is now used as a first line therapy in the treatment of cutaneous and visceral forms and it is efficacious also on hepatic lesions.

REFERENCES

[1] Zhang L, Yuan W, Zheng JW. Pharmacological therapies for infantile hemangiomas: A clinical study in 853 consecutive patients using a standard treatment algorithm. Sci Rep. 2016;6:21670.

[2] Lee K, Bercovitch L. Update on infantile hemangiomas. Sem Perinatol. 2013;37:49-58.

ABS 24

NEONATAL ANTHROPOMETRIC CHARTS FOR TWINS BORN IN ITALY: UPDATE AND COM-PARISON

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INTRODUCTION

The target population is the population on which the chart is built and to which the chart will apply. In absence of exclusion criteria regarding risk factors for fetal growth, a chart based on such population is a reference. Most neonatal charts in use, including the Italian ones, are essentially descriptive references. Our aim is to define, update and compare neonatal anthropometric charts for twins, based on data reflecting current medical management.

MATERIAL AND METHODS

A cross-sectional study, using electronic anonymized data covering neonatal unit admissions from 2011 to 2015, was carried out. Weight, length and head circumference (HC) where measured within 12 h from delivery by trained personnel. Implausible values for gestational age due to errors in measurement or in recording were identified and excluded. Birth growth centiles were constructed using the LMS method [1] and implemented using GAMLESS in the software programming language R. Data from the Italian Neonatal Study [2] and from Bertino et al. [3] were used for comparison.

RESULTS

Data were received from 18 neonatal unit in Piedmont, comprising 5,486 valid birth weight, 5,253 valid birth length and 5,337 valid birth HC of 5,554 twins born from 28 to 40 week's gestation. 66 twins with gestational age lower than 28 week were excluded. Heterogeneity in Italian geographical area and nationality observed in our study population, as well as temporal trend, do not seem to affect weight, length an HC at birth. Difference with the past standard [3] could be related to sample size. As expected, difference in growth is observed between singleton and twin data, starting from 31 weeks of gestational age. LMS figures by sex and gestational age were estimated for weight, length and HD. A sample of our results is summarized in **Fig. 1**.

CONCLUSIONS

Our neonatal anthropometric charts are update national references charts constructed according to an accurate methodology and could be used in current clinical care. LMS figure can be transformed into a Z-score for a more precise growth assessment of twins at birth.

REFERENCES

[1] Cole TJ, Green PJ. Smoothing reference centile curves: the LMS method and penalized likelihood. Stat Med. 1992;11:1305-19.

[2] Bertino E, Spada E, Occhi L, Coscia A, Giuliani F, Gagliardi L, Gilli G, Bona G, Fabris C, De Curtis M, Milani S. Neonatal anthropometric charts: the Italian neonatal study compared with other European studies. J Pediatr Gastroenterol Nutr. 2010;51(3):353-61.

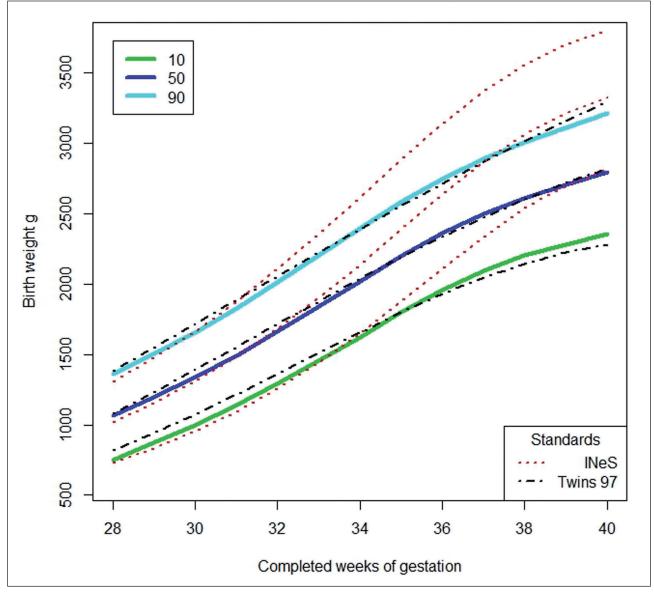


Figure 1 (ABS 24). Female twin birth weight and comparison with standards.

[3] Bertino E, Bagna R, Licata D, Luzzi MB, Murru, P, Prandi G, Coscia A, Leone A, Nicocia M, Martano C. Standard antropometrici del neonato da parto bigemino. Ital J Pediatr. 1997;23:98-105.

ABS 25

BLADDER EXSTROPHY: RECOGNITION AND MANAGEMENT OF A RARE DISEASE

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INTRODUCTION

Classic bladder exstrophy (CBE) is a complex congenital anomaly involving the genitourinary,

reproductive and gastrointestinal tracts, musculoskeletal system and bony pelvis. It is one of three disorders within the exstrophy-epispadias complex, a spectrum of malformation ranging in severity from epispadias to exstrophy of the cloaca. The reported incidence of CBE ranges from 3 to 5 per 100,000 live births and occurs more often in males than females (1.5-2.5:1). The incidence increases in offspring of affected individuals to a rate of 1 in 70 live births and this supports a genetic predisposition. The pathogenesis of CBE appears to be due to an embryological defect in migration of the mesenchymal tissue towards the midline. During the fourth gestational week, the subsequent rupture of the cloacal membrane results in herniation of the lower abdominal components to the surface of the abdominal wall. Bladder exstrophy can be diagnosed prenatally with fetal transabdominal ultrasound between the 15th-32nd weeks of pregnancy. Absence of bladder filling, a low-set umbilicus, widened pubic rami, small genitalia and a lower abdominal mass are all characteristics of CBE on antenatal imaging. Other investigations could be performed to detect associated anomalies: renal ultrasound to examine congenital anomaly of the kidney or urinary tract; spinal ultrasound scan or MRI to assess neurospinal defects; karyotyping to discern newborn's gender.

CASE REPORT

In this report we describe the case of S., born in Sardinia in 2016 at 39 weeks by vaginal delivery. No sign of distress at birth, Apgar 9-10. Neonatal weight was 3.340 kg, height was 50 cm, head circumference was 33 cm. Family history reveals an older age of parents (mother was 40 years old, father was 37 years old). The pregnancy's course was physiological without signs of infection. At birth, the sub-umbilical region exhibited a cleft in abdominal wall. Exposed bladder mucosa was red-colored and urethral meatus were visible from the outside. Distally, bladder neck and urethra were also exposed, labia majora were laterally displaced and anus was anteriorly. Thighs were abducted and external rotated. Advanced maternal age was the only significant potential maternal risk factor.

DISCUSSION

Children with CBE should be treated at tertiary care centers with availability of experienced pediatric urology and pediatric orthopedic surgery teams. A range of surgical techniques (complete vs staged repairs) has been used to achieve successful primary bladder closure with or without pelvic osteotomies. CBE is a rare multisystem birth defect, in the past it was considered fatal for the newborn. Using current techniques, the majority of patients can obtain full continence, adequate genitourinary function and improvement in quality of life. Recognition and management of infants who are diagnosed at birth is difficult, for this reason it is important to provide accurate information.

REFERENCES

• Arlen AM, Smith EA. Disorders of the bladder and cloacal anomaly. Clin Perinatol. 2014;41(3):695-707.

• Goldman S, Szejnfeld PO, Rondon A, Francisco VV, Bacelar H, Leslie B, Barroso U, Ortiz V, Macedo A Jr. Prenatal diagnosis of bladder exstrophy by fetal MRI. J Pediatr Urol. 2013;9:3-6.

• Reinfeldt EG, Mantel A, Fossum M, Nordenskjold A. Maternal and fetal risk factors for bladder exstrophy: A nationwide Swedish case-control study. J Pediatr Urol. 2016;12(5):304.e1-e7.

ABS 26

AMBIGUOUS GENITALIA IN NEWBORN: A CASE REPORT

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INTRODUCTION

The "disorders of sex development" are congenital conditions characterized by an altered development of chromosomal, gonadal or anatomic sex. Clinical evaluation is based on family and prenatal history, a detailed physical examination with a special attention to any dysmorphic features. First-line testing includes karyotype, abdominopelvic ultrasound, measurement of sexual hormones, serum electrolytes and urinalysis [1].

CASE REPORT

We report the case of an infant born preterm at 35 weeks from caesarean delivery for asymmetric intrauterine growth retardation (IUGR). He was a second child of 36-year-old non consanguineous parents. Family history reported MTHFR mutation and hypertension in the mother, treated with methyldopa, acetylsalicylic acid, deltacortene and nadroparin. APGAR score was 7 at the 1st minute and 9 at the 5th minute, respectively. Prenatal ultrasounds revealed a female foetus. At birth, weight was 1,460 g, length was 42 cm and occipitofrontal circumference was 29 cm (all the parameters < 3rd percentile). Resuscitation included ventilation with neopuff with 30% of oxygen needs. The infant was admitted to our Neonatal Intensive Care Unit for dysmorphic features in a premature baby small for gestational age. The newborn examination showed triangular face, large bregmatic fontanelle, facial asymmetry with large nose root and tip, microretrognathia, low-set ears, ambiguous genitalia with hypospadia and "uninhabited" scrotum, anterior anus, sacral dimple and partial syndactilia of the 2^{nd} and 3^{rd} finger of the left foot. On the 1st day of life, cerebral ultrasound was normal; abdominal ultrasound detected the presence of testicles in inguinal canal, while cardiac ultrasound revealed a ostium secundum atrial septal defect and a patent ductus arteriosus, which closed on the 5th day of life. Laboratory findings were normal except a slight increase of aspartate aminotransferase (61

UI/L) and of gamma-glutamyltransferase (94 UI/L) on day 8, with normal bilirubin levels. Chromosome analysis demonstrated a normal male karyotype (46 XY). During his hospitalisation, the infant did not need breathing assistance and showed a good food tolerance. It is currently in good general conditions with a growth below the 3rd percentile. Genetic investigations are ongoing for the delineation of the clinical picture.

CONCLUSIONS

The management of a newborn with ambiguous genitalia requires immediacy and a great sensitivity. It should be evaluated, when possible, by a multidisciplinary team composed by neonatologists, paediatric urologists, endocrinologists, geneticists and child psychiatrist/psychologists; this team should work closely with the family. Surgery is useful to make external genitalia compatible with the karyotype, but its role in the neonatal age is very limited [2].

REFERENCES

 Lee PA, Houk CP, Ahme, SF, Hughes IA. Consensus statement on management of intersex disorders. Pediatrics. 2006;118(2):e488-500.

[2] Lambert SM, Vilain EJ, Kolon TF. A practical approach to ambiguous genitalia in the newborn period. Urol Clin North Am. 2010;37(2):195-205.

ABS 27

PEDIATRIC HEADACHE: SIMPLE OR SIMPLY DIFFICULT TO MANAGE?

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INTRODUCTION

Pseudotumor cerebri (PTC), also known as idiopathic intracranial hypertension, is characterized by elevated intracranial pressure (ICP) without any evidence of masses, lesions or cerebrospinal fluid (CSF) obstructions on neuroimaging. Normal CSF composition is also the rule [1].

PTC prevalence is 1/100,000 individuals and few studies are reported on pediatric series.

In adults and post-pubertal children PTC more frequently than others affects overweight women.

In the pre-pubertal ages PTC relates less frequently to obesity and male to female prevalence is similar. Criteria for the diagnosis are: papilledema, normal neurologic examination except for cranial nerve abnormalities, normal MRI, normal CSF chemistry but ICP > 28 cmH₂O. Empty sella and swelling of the optic nerves confirm the pattern [2, 3]. We report the case of a girl with headache and confounding factors.

CASE REPORT

A 12-year-old pubertal girl (161 cm, 75th-90th percentile; 60 kg, 90th-95th percentile) came to our observation because of frontal and retro-orbital headache (Wong-Baker 6/10) persisting since 2 months. Her history was silent except for a mild head-trauma one year before. The family history reported strong disagreement between the parents. The ENT and neurologic consultations were normal. EEG (basal and in sleep) showed no alterations. Nevertheless a deflection of mood was detected. Clinical chemistry excluded other conditions. The fundus oculi showed papilledema (Frisen scale: stage 3 right, 2 left) while the electroretinogram was normal. Conversely, the visual-evoked potentials showed an altered conduction of the optical nerve. At MRI a minimal increase of CSF in the perioptical spaces and a partially empty sella, without other alterations (Fig. 1). The pattern was identified as a PTC and a 3rd level referral was arranged. After the exclusion of a secondary PTC from autoimmune or hormonal conditions (Ab anti-phospholipid, vitamin A, circulating hormones), the girl was started on acetazolamide 900 mg/day after the final diagnosis with lumbar puncture (CSF: clear and colorless, normal chemistry, $ICP = 29 \text{ cmH}_2O$). Follow-up. The empty sella and the optic nerves swelling improved rapidly and the fundus oculi as well. Two weeks later metabolic acidosis emerged thus acetazolamide was reduced (500 mg/day) and oral sodium bicarbonate was introduced. One more episode of headache is reported to-date resolved by painkiller and her follow-up continues as stated. Simultaneously, psychological support was started to improve the girl's mood. CONCLUSIONS

The diagnosis of PTC in a pediatric patient is becoming more common so it has to be included in the differential diagnosis of any child with newonset or chronic headaches. Neuroimaging and CSF examination must be performed to make a diagnosis. A proper early management is necessary to avoid major complications as permanent visual loss and chronic pain.

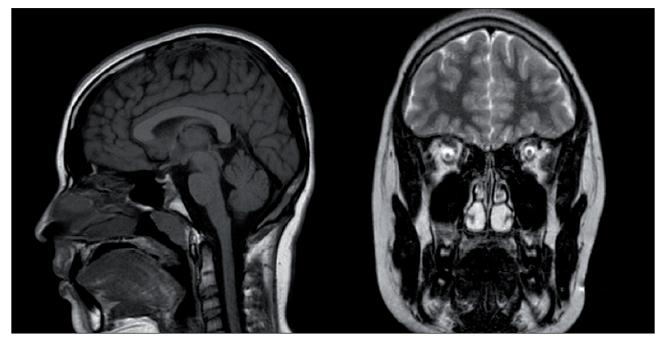


Figure 1 (ABS 27). Partially empty sella and swelling of perioptical spaces.

REFERENCES

 Rook BS, Philips PH. Pediatric pseudotumor cerebri. Curr Opin Ophthalmol. 2016;27(5):416-9.

[2] Rogers DL. A review of Pediatric Idiopathic Intracranial Hypertension. Pediatr Clin N Am. 2014;61:579-90.

[3] Aylward SC, Reem RE. Pediatric Intracranial Hypertension. Pediatr Neurol. 2017;66:32-43.

ABS 28

SUBCUTANEUS FAT NECROSIS AND HYPER-CALCEMIA: A CLINICAL CASE

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INTRODUCTION

Subcutaneous fat necrosis (SCFN) of the newborn is a rare inflammatory disorder of the fat tissue, presenting in newborn, consisting in a form of panniculitis associated to painful, hard and erythematous-violaceous nodules. Usually it has a favorable prognosis, with complete auto resolution of subcutaneous lesions within several weeks or months, but it can be complicated by metabolic alteration.

CLINICAL CASE

A full-term infant girl (39⁺⁴ gestational weeks) was born by cesarean section for fetal cardiac anomalies. Birth weight was 3,550 g. Gravidic history was unremarkable. The patient needed cardiopulmonary resuscitation at birth for neonatal asphyxia, Apgar score was 4 at 1 min, 7 at 5 min. Cord blood gas analysis showed metabolic acidosis (pH 7.03, PCO₂ 65, HCO, 17.2 mmol/l, BE -13.6). At 3 hours of life for the appearance of upper limb clones and convulsions at CFM monitoring, the hypothermic treatment was started for a total duration of 72 hours with a rectal temperature of 33.5°C. During the procedure she performed sedation with fentanyl and phenobarbital. On second day of life, the patient presented painful subcutaneous hard nodules covered by erythematous skin. These lesions were localized in the gluteal region, elbows and shoulders and were consistent with nodules of subcutaneous fat necrosis. Serum calcium was increased (12.5 mg/dl), parathormone was 5.5 pg/ml, calcitonin 52 pg/ml. The hypercalcemia was treated with hyper hydration, furosemide and prednisolone. Basing on clinical history, physical examination, and biochemical parameters, the erythematous lesion were diagnosed as SCFN confirmed by cutaneous biopsy. The neurological examination was normal at discharge. DISCUSSION

SCFN of the newborn is a benign condition occurring in the neonatal period, characterized by inflammation and necrosis of subcutaneous fat tissue. Lesions may be isolated or clustered and are typically localized on shoulders, back, buttocks and face; the evolution is subcutaneous calcifications. Etiology of this disorder is unknown, but it is linked to multiple risk factors (perinatal asphyxia, umbilical cord prolapse, meconium aspiration, therapeutic hypothermia, neonatal sepsis, maternal diabetes, maternal medi-

cations, preeclampsia). Sometimes it may be associated with thrombocytopenia, hypoglycemia, hypercalcemia: these metabolic derangements may represent a possible risk of serious complications. Hypercalcemia is found in 25% of cases. The first 6 weeks of life represent the time frame at highest risk for clinically significant hypercalcemia in SCFN. Usually, serum calcium starts to rise as SCFN lesions begin to regress; sometimes, hypercalcemia is already detectable before the subcutaneous lesions. Newborn with hypercalcemia present lethargy, hypotonia, irritability, vomiting, polyuria, polydipsia, constipation, dehydration. Hypercalcemia is caused by increased prostaglandin activity, release of calcium from necrotic fat tissue and increased secretion of 1,25-dihydroxyvitamin D3 from subcutaneous lesions, leading to an increased intestinal uptake of calcium. If left untreated, hypercalcemia may lead to nephrocalcinosis, nephrolithiasis, renal failure, calcification of skin, myocardium and gastric mucosa. Treatment of hypercalcemia consists in hyperhydration and diet with low levels of calcium and vitamin D; sometimes drugs like furosemide and prednisolone are needed to reduce serum calcium; bisphosphonates may be employed to treat moderate to severe hypercalcemia when other measures have been ineffective.

CONCLUSIONS

SCFN of the newborn is a rare inflammatory disorder of the fat tissue, presenting in newborn, consisting in a form of panniculitis associated to painful, hard and erythematous-violaceous nodules. It is usually a transient and self limited condition. However, it may be complicated by a number of metabolic alterations like thrombocytopenia, hypoglycemia, hypercalcemia, hypertriglyceridemia. It is important to monitor newborns with SCFN to avoid the risk of serious complications, with particular reference to hypercalcemia. Regular monitoring of serum calcium is recommended until the age of 6 months.

ABS 29

PHENOTYPIC DISCORDANCE IN DIGEORGE MONOZYGOTIC TWINS

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INTRODUCTION

DiGeorge Syndrome (DGS) is the most common microdeletion syndrome, with a frequency of 1:4,000 live births. Even monozygotic (MZ) twins can exhibit a level of discordance, suggesting that the phenotypic variability could not be explained on a genetic basis alone.

CASE REPORT

L. and T. are a set of MZ twins from a monochorionic and diamniotic pregnancy. Their family history was unremarkable, without any cases of cardiac disease. The US scan at the 16th gestational week showed a right sided aortic arch in T. Twin. T. also showed less fetal movement than L. The mother suffered from gestational diabetes, treated only with diet modification. T. and L. weighted respectively 2.210 kg and 2.270 kg and they were 46 cm long. Heart US scan at 1st day of life confirmed that T. had right sided aortic arch, with normal cardiac parameters. At the age of 1 year and a half the parents noticed in both infants a developmental delay that was confirmed by the neurological examination. A genetic cause was suspected. The genetic analysis showed a 22q microdeletion (FISH), and the MLPA detected a microdeletion of 1.5 MB, with no difference between them. Further phenotypic features were concordant.

CONCLUSION

We describe a twin pair discordant for heart anomaly but genetically identical. According to the other cases in literature, the twin with the heart anomaly has often a lower weight at birth. In our case was shown a phenotypic difference, even though less significant. Moving from this observation, it can be presumed that this discordance could be due to differences in the utero environment, such as growth disadvantage, disturbance of laterality and possible placental anastomoses with consequent twin-to-twin transfusion. Goodship et al. advocate that haploinsufficiency for the region 22q11.2 may be sufficient to cause the dismorphic facies, but the occurrence of the cardiovascular defect may require a second hit on the controlateral allele. Moreover, the most likely mechanism for the second hit could involve epigenetic changes. The highly variable expressivity in DGS patients can be clarified by slightly altered levels of TBX1 expression among different individuals with the same deletions. This heterogeneity of presentations may be caused by random environmental events or sequence variations of genetic modifiers, like PAX3, CRKL, VEGF and RALDH2. Other genes such as NKx2.5, transcription factors or chromatin modifiers like

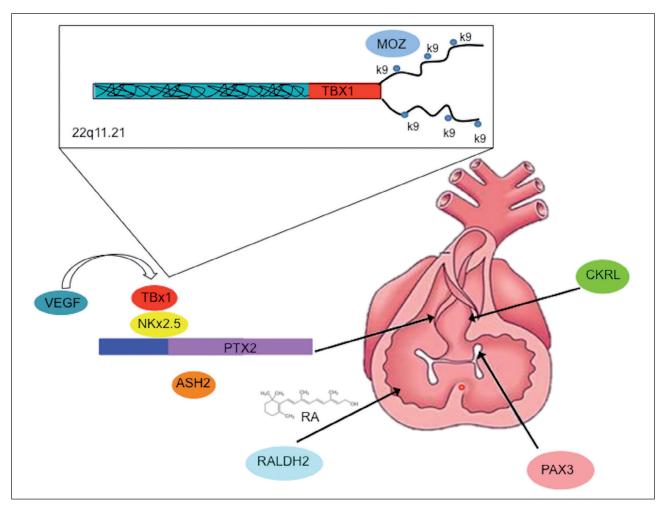


Figure 1 (ABS 29). Schematic representation of major genes and regulators of transcription during heart development, that can be involved in DGS.

MOZ or ASH2 can be further studied for the discordance of the syndrome presentation, because they can change the expression of Tbx1 (**Fig. 1**). In fact a loss of function, caused by epigenetic mutations or stochastic events, could modify the expression of this gene and could also explain the different presentation in MZ twins.

REFERENCES

• Goodship J, Cross I, Scambler P, Burn J. Monozygotic twins with chromosome 22q11 deletion and discordant phenotype. J Med Genet. 1995;32:746-8.

• Liao J, Kochilas L, Nowotschin S, Arnold JS, Aggarwal VS, Epstein JA, Brown MC, Adams J, Morrow BE. Full spectrum of malformations in velocardio-facial syndrome/DiGeorge syndrome mouse models by altering Tbx1 dosage. Hum Mol Genet. 2004;13:1577-85.

• Voss AK, Vanyai HK, Collin C, Dixon MP, McLennan TJ, Sheikh BN, Scambler P, Thomas T. MOZ Regulates the Tbx1 Locus, and Moz Mutation Partially Phenocopies DiGeorge Syndrome. Dev Cell. 2012;23(3):652-63.

ABS 30

FETAL MAGNETIC RESONANCE PARAMETERS IN INTRAUTERIN GROWTH RESTRICTION

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INTRODUCTION

The early identification of the intrauterine growth restriction (IUGR) is one of the main goals of prenatal care. Although there are limited indications about the management of fetal growth restriction *in utero*, the early recognition of fetus at risk can lead to improved perinatal outcome. Nowadays, ultrasonography is the gold standard for fetal screening. A second-line method is fetal Magnetic Resonance Imaging (f-MRI) [1], which provides more information about the development of individual organs, such as the lung [2]. The aim of our study is to evaluate if the f-MRI is useful to identify IUGR fetuses and to predict their perinatal respiratory outcome.

METHODS

All f-MRI performed between 2014-2016 at "S. Maria alle Scotte" Hospital, in Siena, were analysed. Eight pregnancies with IUGR were recruited together with a control population (n = 10) coupled for gestational age at the time of f-MRI. The ultrasound evaluation of the fetal weight (EFW) and babies' information was collected. The MRI protocol consists of T2 weighted images. 6 ROI (Region of Interest) were placed as follow: 2 on the lung, 2 on the liver and 2 on the amniotic fluid (**Fig. 1**). The signal intensities (SI) of each ROI were measured. A ratio comparing the SI of the lung with structures at a comparable depth used as a reference was introduced [3]. Thus, SI lung

to liver ratio (SI lung/liver) and SI lung to amniotic fluid (SI lung/amniotic fluid) ratio were obtained for each fetus. SI lung/liver and SI lung/amniotic fluid were related to fetal anthropometric measures and compared between IUGR and control group. In order to compare the fetuses that recover their growth from those who born small for gestational age (SGA), the IUGR population was also divided in AGA and SGA. All the results were also studied in relation to the subsequent perinatal respiratory outcome. RESULTS

ESULIS Llung/liver uv

SI lung/liver was linearly related to gestational age at the time of f-MRI (Rho = 0.858; p < 0.001) and to EFW (Rho = 0.794; p < 0.001). SI lung/amniotic



Figure 1 (ABS 30). Coronal SSFSE T2-weighted image where 6 ROI were placed.

fluid was significantly higher in the IUGR group than in the control group (respectively: 0.80 ± 0.06 and 0.65 ± 0.03 ; p = 0.043). Looking at the respiratory outcome, the IUGR population required a higher amount of days of oxygen needs (IUGR: 12.25 ± 8.13 vs Control: 1.9 ± 1.01 ; p = 0.005). In contrast, among the IUGR fetuses, lower values of SI lung/amniotic fluid were found in the SGA population than in the AGA group (respectively: 0.63 ± 0.07 and 0.9 ± 0.06 ; p = 0.036). The days of oxygen supply were higher in the SGA newborns than in the AGA group (30.66 ± 18.52 and 1.2 ± 0.96 ; p = 0.028).

CONCLUSIONS

SI lung/liver increases with fetal lung maturation and it appears to be useful to estimate intrauterine fetal growth. SI lung/amniotic fluid seems to be a reliable predictive index to identify the IUGR newborns that can recover their growth from those that will born SGA. The f-MRI represents a promising frontier for the IUGR fetus outcome prediction, contributing to ameliorate the perinatal management of IUGR pregnancies.

REFERENCES

[1] Perrone A, Savelli S, Maggi C, Di Pietro L, Di Maurizio M, Tesei J, Ballesio L, De Felice C, Giancotti A, Di Iorio R, Manganaro L. Magnetic resonance imaging versus ultrasonography in fetal pathology. Radiol Med. 2008;113:225-41.

[2] Kasprian G, Balassy C, Brugger PC, Prayer D. MRI of normal and pathological fetal lung development. Eur J Radiol. 2006;57:261-70.

[3] Brewerton LJ, Clari RS, Liang Y, Bhargava R. Fetal lung-to-liver signal intensity ratio at MR imaging: development of a normal scale and possible role in predicting pulmonary hypoplasia in utero. Radiology. 2005;235(3):1005-10.

ABS 31

DRY LUNG SYNDROME AND SEVERE BRON-CHOPULMONARY DYSPLASIA WITH FATAL OUTCOME IN TWO PRETERMS OF 29 WEEK'S GESTATION

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INTRODUCTION

Dry lung syndrome is a lung functional hypoplasia consequent to oligohydramnios [1]. The lung, for a correct growth, needs an adequate amniotic fluid volume. According to some authors [2] oligohydramnios causes a rise of chest compression and a reduction of foetal breathing movements, which bring an outflow of lung fluid. Furthermore, the contraction of lung muscle cells and correlated stretch and shear forces regulate gene expression for proliferation and differentiation of lung cells. The lack of these mechanisms and the resultant hypoplasia, at birth, lead to respiratory problems that go from distress to pulmonary insufficiency. Oligohydramnios can be the result of a prolonged premature rupture of membranes (PROM), that happens about 0.4-0.7% of pregnancies, in particular from 23 to 27 week's gestation. From rupture to delivery might be a long time, also 14 days. During these days often occurs infections and this represents another important element able to reduce foetal breathing movements and to determine an alteration in pulmonary vascular development [1, 3]. PROM and infections, in turn, are important risk factors for bronchopulmonary dysplasia (BPD), a chronic inflammatory lung disease of preterm newborn marked by different degrees of respiratory distress. BPD is usually correlated with mechanical ventilation and oxygen damage but it's known that the introduction of antenatal steroids, intratracheal surfactant and less aggressive ventilation has led to a change in the pathology and in clinical presentation [4]. The lung injury decreased in larger and more mature babies and increase in newborns of 24-27 week's gestation. In these weeks the lung is in canalicular stage and an arrest in the development, consequent to the injury, cause an alveolar simplification (fewer and larger) and an impaired vascular growth (who results in pulmonary hypertension) [5]. Gestational age is inversely related to incidence, morbidity and mortality of BPD; in a baby of 22 weeks the rate of survival is only 6% versus the 92% of 28 weeks [6]. These case reports show the story of two preterm infants of 29 week's gestation. Both of them develop a severe BPD with negative outcome despite the "good" gestational age. They had a very similar prenatal story, characterized by dry lung syndrome and chorioamnionitis.

CASE 1

The first baby was a male, born at 29 week's gestation with weight of 1,000 g. During pregnancy, at 25 week, the mother had PROM and chorioamnionitis. Born by emergency caesarean section, had an Apgar scores of 1 at first minute, 2 at fifth minute and 4 at tenth minute; amniotic fluid was clear but there was a sever oligohydramnios. The baby was immediately intubated and reanimated with external cardiac massage. At the admission in our NICU he started HFVO (high frequency oscillation ventilation) and made surfactant. After few hours we tried to extubate the baby and to start BiPAP (biphasic positive airway pressure) but the baby needed to be reintubated and to make another administration of surfactant. During the hospitalization we tried several time to extubated him but the baby, for the most of time, was ventilated in HFOV with need of oxygen around 60%. Echocardiography at 2 months of life showed pulmonary hypertension (PH) so he started sildenafil and inhaled nitric oxide (iNO). For BPD he made six cycle of dexamethasone, caffeine, hydrochlorothiazide and spironolactone. Despite therapy and gestational age the respiratory state has only got worse until the death of the baby. CASE 2

The other baby was a female of 29 week's gestation with birth weight of 960 g and a history of PROM and chorioamnionitis at 23 week's gestation. Due to chorioamnionitis, the Obstetricians decided to perform a caesarean section. She had an amniotic fluid cloudy and anhydramnios. Her Apgar score was of 3 at first minute, and 8 at fifth minute. As in the previous case she was immediately intubated and during the hospitalization we tried several time to extubate her, but overall the most of time the baby was ventilated in HFOV with need of oxygen around 60%. From the first days of life echocardiography showed PH with fast progression and it was immediately started therapy with dobutamine and after iNO and sildenafil. The respiratory state was very similar to the other baby and she made the same therapy with six cycle of dexamethasone, caffeine, hydrochlorothiazide and spironolactone. In the same way, despite the therapy and the gestational age the respiratory state has only got worse until the death of the baby at six months of life.

DISCUSSION

BPD is the most common and one of the prognostically most severe sequelae of preterm birth [7]. But in accordance to current definition of the "new BPD" [4, 8] the most complicated frameworks are in newborn with gestational age between 24-27 weeks. [5]. So, babies with a gestational age of 29 weeks should have a BPD with a good outcome. However, if in the antenatal story of the baby other conditions (such as a dry lung syndrome and chorioamnionitis) are present, the outcome of BPD change and get worse. The presence of different pathological condition can led to a respiratory state more serious and more difficult to manage. In the cases we showed, the clinical evolution was not determined by BPD but rather by a more complicated state caused by dry lung syndrome and chorioamnionitis.

REFERENCES

 McIntosh N. Dry lung syndrome after oligohydramnios. Arch Dis Child. 1988;63(2):190-3.

[2] Williams O, Hutchings G, Hubinont C, Debauche C, Greenough A. Pulmonary effects of prolonged oligohydramnios following mid-trimester rupture of the membranes – antenatal and postnatal management. Neonatology. 2012;101:83-90.

[3] Sehgal A, Francis JV, Ang H, Tan K. Dry Lung Syndrome: A Distinct Clinical Entity. Indian J Pediatr. 2010;77:1029-31.

[4] Jobe AH, Bancalari E. Bronchopulmonary dysplasia. Am J Respir Crit Care Med. 2001;163(7):1723-9.

[5] Abman SH. Bronchopulmonary dysplasia: "A vascular hypotesis". Am J Respir Crit Care Med. 2001;164:1755-6.

[6] Stoll BJ, Hansen NI, Bell EF, Shankaran S, Laptook AR, Walsh MC, Hale EC, Newman NS, Schibler K, Carlo WA, Kennedy KA, Poindexter BB, Finer NN, Ehrenkranz RA, Duara S, Sánchez PJ, O'Shea TM, Goldberg RN, Van Meurs KP, Faix RG, Phelps DL, Frantz ID 3rd, Watterberg KL, Saha S, Das A, Higgins RD. Eunice Kennedy Shriver National Institute of Child Health and Human Development Neonatal Research Network. Neonatal outcomes of extremely preterm infants from the NICHD Neonatal Research Network. Pediatrics. 2010;126:443-56.

[7] Jensen EA, Schmidt B. Epidemiology of broncolpulmonary displasia.Birth Defects Res A Clin Mol Teratol. 2014;100(3):145-57.

[8] Collins JJP, Tibboel D, de Kleer IM, Reiss IKM, Rottier RJ. The Future of Bronchopulmonary Dysplasia: Emerging Pathophysiological Concepts and Potential New Avenues of Treatment. Front Med (Lausanne). 2017;4:6.

ABS 32

FETAL MAGNETIC RESONANCE IMAGING AP-PLICATIONS IN GASTROINTESTINAL CON-GENITAL ABNORMALITIES

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INTRODUCTION

Magnetic Resonance Imaging (MRI) is the study of choice in the characterization of gastrointestinal congenital abnormalities highlighted by fetal sonography (USf).

MATERIALS AND METHODS

USf is the routine screening study of all fetal anomalies; most fetal anomalies are diagnosed by ultrasound and not require a complementary imaging study. US remains the predominant modality of evaluating disorders related to pregnancy but it has technical limitations; such limitations may require MRI as alternative and complementary imaging method. MRI has superior soft tissue contrast resolution and multiplanar imaging; however, fetal study may give limited diagnostic information in early gestational age due to the small size of the fetus and fetus movements.

RESULTS

MRI of fetal gastrointestinal tract is based on contrast of meconium and ingested amniotic fluid.

MRI confirms or completes USf inconclusive findings, verifying the presence, severity and extent of gastrointestinal congenital abnormalities with high sensibility and specificity. The indications for MRI in gastrointestinal fetal pathology assessment are identifying site of bowel obstruction, bowel atresia, bowel stenosis, Hirschsprung disease, meconium ileus, evaluation of intestinal ischemia (in case of bowel volvulus), and malrotation.

CONCLUSIONS

The data in the literature shows that MRI has an additional value to fetal US in delineation and characterization of abdominal abnormalities thanks to the high contrast resolution and to the excellent anatomical detail.

REFERENCES

• Gupta P, Sharma R, Kumar S, Gadodia A, Roy KK, Malhotra N, Sharma JB. Role of MRI in fetal abdominal cystic masses detected on prenatal sonography. Arch Gynecol Obstet. 2010;281(3):519-26.

• Rodriguez RM. Fetal MRI: thoracic, abdominal and pelvic pathology. RAR. 2012;76(1):1-20.

• Saleem SN. Fetal MRI: an approach to practice: a Review. J Adv Research. 2014;5:507-23.

ABS 33

NOT ALL CAUSES OF BAD BREATHING ARE RELATED TO LUNGS

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INTRODUCTION

Total anomalous pulmonary venous connection (TAPVC) is a rare congenital heart defect in which pulmonary venous blood directly flows into the right side of the heart or into the systemic veins [1]. There are four types of TAPVC: supracardiac (connected to left superior vena cava [LSVC] or right superior

vena cava); cardiac (coronaric sinus or right atrium); infracardiac (portal veins or inferior vena cava [IVC]); mixed pattern (at least 2 different locations). The most common type is the supracardiac. The presentation and outcomes are influenced by the presence or not of obstruction, which is a surgical emergency. Generally, in these cases newborns show cyanosis, respiratory failure and shock; it may be difficult to distinguish TAPVC from respiratory distress syndrome (RDS); timing of onset can help differentiate between these disorders: RDS immediately after birth, TAPVC delayed onset after 12 hours of life. Diagnosis is echocardiographic: no pulmonary venous connections to the left atrium; collecting vein with a connection to systemic veins; direct connection with right atrium or coronary sinus; right atrium and ventricle dilated; right-to-left interatrial shunting. Chest radiography can show the "snowman sign" in supracardiac TAPVC. ECG is non-specific. Management is focused on stabilization of the patient and surgical correction [2].

CASE REPORT

N. was born at term (40 w) with a birth weight of 4.100 kg, APGAR 9-10. His mother had gestational diabetes and positivity of vaginal-rectal swabs for Str. beta hemolytic. After 20 hours he presented polypnea and peripheral oxygen saturation 90%. For that reason he was transferred to our Neonatal Intensive Care Unit (NICU) and CPAP support was established with variable oxygen requirements. Emogasanalysis was normal. Initial chest X-ray showed microgranular appearance. The first 2D-echocardiogram (performed by an adult cardiologist) was initially suggestive for acute heart failure and persistent ductus arteriosus with bidirectional shunt. In our NICU, pulmonary venous connections to a dilated right atrium were observed; right ventricle was dilated as well; there was an interatrial defect (7 mm) with right to left shunt. A common pulmonary venous chamber was identified behind the left atrium. A vertical vein drained directly to the superior vena cava (dilated). No obstruction was recognized with pulsed Doppler, and the diagnosis of non-obstructed supra-cardiac TAPVC was established. There were no other associated cardiac defects. The newborn was then transferred to a cardiac surgery unit to perform elective surgical correction.

CONCLUSIONS

Not all children badly breathing have respiratory problems. Always think about a congenital heart disease, especially if symptoms start after 12 hours from birth. In the evaluation of congenital heart diseases the role of pediatric cardiologist is crucial [3].

REFERENCES

[1] Zhang Z, Zhang L, Xie F, Wang B, Sun Z, Kong S, Wang X, Dong N, Wang G, Lv Q, Li Y, Li L, Xie M. Echocardiographic diagnosis of anomalous pulmonary venous connections: Experience of 84 cases from 1 medical center. Medicine (Baltimore). 2016;95(44):e5389.

[2] Files MD, Morray B. Total anomalous pulmonary venous connection: preoperative anatomy, physiology, imaging, and interventional management of postoperative pulmonary venous obstruction. Semin Cardiothorac Vasc Anesthes. 2017;21(2):123-31.

[3] Singh Y, Gupta S, Groves AM, Gandhi A, Thomson J, Qureshi S, Simpson JM. Expert consensus statement 'Neonatologist-performed Echocardiography (NoPE)'-training and accreditation in UK. Eur J Pediatr. 2016;175: 281-7.

ABS 34

NEONATAL FEMUR FRACTURE AFTER CAE-SAREAN SECTION AND BREECH PRESENTA-TION

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INTRODUCTION

Breech vaginal delivery is a major risk factor for many birth injuries. However, also caesarean section because of breech presentation may rarely cause traumatic birth events. Long bone fractures occur in 0.1% of caesarean deliveries and in 0.5% of vaginal ones.

CASE REPORT

Male newborn, 37^{4/7} weeks gestation, born by urgent caesarean section because of breech presentation and premature rupture of membrane. Birth weight 2,800 g, Apgar scores 1': 6; 5': 8. At birth he was ventilated with Neopuff because of ineffective breathing pattern for a few seconds. At first physical examination at well-baby nursery, the neonatologist during Ortolani maneuver noticed an unusual click and instability of left femur. On the second day of life, soft tissue above left femur appeared swollen with hyperemic upper skin. Therefore a left femur X-ray was performed and a composed diaphysis fracture between third medium and third proximal of left femur was confirmed. No peripheral nerves deficiency occurred. Immediately, the newborn was referred to orthopedic surgeon for management and Bryant's traction was applied for three weeks. During hospitalization, we ruled out vitamin D and calcium deficits. A second left femur X-ray was performed after 3 weeks of traction and showed a generous fracture callus.

DISCUSSION AND CONCLUSIONS

Predisposing factors for foetal bone fractures during delivery are: twin pregnancy, fetal macrosomia, inadequate uterine incision, uterine myomas, breech presentation and prematurity. It is important to notice any unusual click sound at birth during Ortolani maneuver, since a long bone fracture may be shown. Caesarean section reduces the incidence of foetal injuries in comparison to vaginal delivery, chiefly in the case of breech presentation, although a long bone fracture is always possible. Several mechanisms have been proposed.

REFERENCES

• Basha A, Amarin Z, Abu-Hassan F. Birth-associated long-bone fractures. Int J Gynaecol Obstet. 2013;123(2):127-30.

• Capobianco G, Virdis G, Lisai P, Cherchi C, Biasetti O, Dessole F, Meloni GB. Cesarean Section and Right Femur Fracture: A Rare but Possible Complication for Breech Presentation. Case Rep Obstet Gynecol. 2013;2013:613709.

• Thakkar UG, Mishra VV, Vanikar AV, Agrawal AS, Kadam PG. Fracture of shaft of left femur in a newborn delivered by caesarean section. Sri Lanka J Child Health. 2015;44(1):53-5.

ABS 35

DASH DIET SCORE AND DISTRIBUTION OF BLOOD PRESSURE IN A LARGE COHORT OF SOUTHERN EUROPEAN CHILDREN

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INTRODUCTION

High blood pressure (BP) is a health and lifethreatening process, which often begins early in childhood [1]. The Dietary Approaches to Stop Hypertension (DASH) Trial shares clinical evidence about the impact of a specific diet pattern (i.e. low salt/fat/processed foods and high fruit/vegetables intakes) on BP [2]. The effects of compliance on BP with a DASH-type diet in a paediatric population sample has not yet been studied, so that this study aimed at evaluating this relationship.

METHODS

We used cross-sectional data from a study of boys and girls aged 10-15 years (n = 839; females 52.6%). Participants completed a physical examination, including 5 standardized clinical BP recordings over a three years period, to screen only those with persistently high BP [3]. Diet quality was assessed by using a DASH dietary score, constructed from a standard Food Frequency Questionnaire. Lower scores indicated less healthy diets. Hypertension was defined as clinic BP \ge 95th percentiles according with specific pediatric nomograms.

RESULTS

The prevalence of hypertension (9.4%) was higher than previously usually reported in paediatric age (1-5%). Furthermore, the most widely diffused subtype of hypertension in our young population was the isolated systolic. Inverse associations were detected between DASH score and systolic BP. Specifically, there was a difference in systolic BP of 7.3 mmHg and 5.0 mmHg in boys and girls, between the highest and lowest DASH quintiles. At multivariate regression analysis, DASH score was inversely associated with SBP (β = -1.29; 95% CI = -2.48 to -0.11; p = 0.03). Other differences in systolic BP were evident across DASH quintiles (**Fig. 1**).

CONCLUSIONS

Our findings suggest that adherence to DASH diet can decrease BP also in paediatric age. Public policies promoting a DASH-style diet may have a positive effect on population health because reducing systolic BP and so its harmful consequences.

REFERENCES

[1] Bassareo PP, Mercuro G. Pediatric hypertension: An update on a burning problem. World J Cardiol. 2014;6:253-9.

[2] Harrington JM, Fitzgerald AP, Kearney PM, McCarthy VJ, Madden J, Browne G, Dolan E, Perry IJ. DASH diet score and distribution of blood pressure in middle-aged men and women. Am J Hypertens. 2013;26:1311-20.

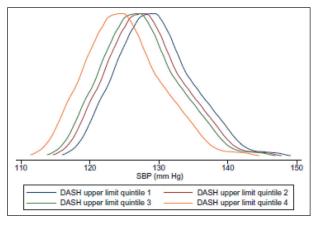


Figure 1 (ABS 35). Distribution of systolic blood pressure by DASH score in boys and girls.

[3] Marras AR, Bassareo PP, Ruscazio M. The prevalence of paediatric hypertension, emphasising the need to use specific population references: the Sardinian Hypertensive Adolescents Research Program study. Cardiol Young. 2009;19:233-8.

ABS 36

THE CORRELATIONS BETWEEN THE WIDTH OF THE NEPHROGENIC ZONE AND MATERNAL AND NEONATAL CLINICAL DATA

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INTRODUCTION

The dramatic increase in the incidence of endstage renal disease focused our attention on the conditions predisposing to the insurgence of renal failure. Prematurity probably represents the most important factor predisposing adults to develop chronic kidney disease and hypertension. The oligonephronia, in particular, represents a possible link between preterm birth and susceptibility to develop kidney disease in adult life. The renal stem cell represents the most important structure of the developing kidney. In particular, in the sub capsular zone of fetal organ, was identified as the most important stem cell niche, called "Blue Strip". The multipotent mesenchymal cells, located in this zone, represent the nephron progenitor population of the newborn kidney, capable to originate all segments of the nephron, except the collecting ducts. The aim of this study was to correlate the width of the Blue strip with maternal and neonatal clinical data, in order to identify possible factors that may cause oligonephronia.

METHODS

In this work, we examined 50 kidney samples (62% men, 38% females) belonging to 2 fetuses, 36 preterm births and 12 births at term and 50 respective maternal medical records.

RESULTS

We found an important inter-individual variability between the width of the blue strip in the kidney samples and their gestational age, with a greater width in low gestational age. The low weight at birth resulted correlated with a greater width of nephrogenic zone. In particular, by comparing

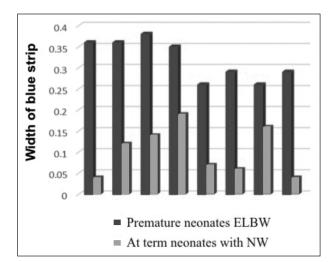


Figure 1 (ABS 36). Comparison of the width of blue strip between 8 at term neonates with normal weight (NW) and 8 preterm neonates with extremely low birth weight (ELBW).

the width of blue strip between 8 neonates at term with normal weight and 8 preterm neonates with extremely low birth weight, it resulted wider in the first group (p = 0.05) (Fig. 1). No correlation was found between the blue strip width and maternal ages or nulliparity and multiparity.

CONCLUSIONS

In conclusion, we confirmed an extreme complexity in the nephrogenesis. Several factors seem to affect kidney development. In our study, we confirmed the important linkage between prematurity and low weight and oligonephronia. Further studies are needed to verify the correlation between the width of nephrogenic zone and, for example, maternal and paternal disorders or drugs and food taken during pregnancy. In future, the identification of possible risk factors during perinatal period might help us to extend nephrogenesis and to increase the number of nephron at birth.

REFERENCES

· Crobe A, Desogus M, Sanna A, Fraschini M, Gerosa C, Fanni D, Fanos V, Van Eyken P, Faa G. Decreasing podocyte number during human kidney intrauterine development. Am J Physiol Renal Physiol. 2014;307(9): F1033-40

· Faa G, Gerosa C, Fanni D, Nemolato S, Locci A, Cabras T, Marinelli V, Puddu M, Zaffanello M, Monga G, Fanos V. Marked interindividual variability in renal maturation of preterm infants: lessons from autopsy. J Matern Fetal Neonatal Med. 2010;23(Suppl 3):129-33.

• Faa G, Gerosa C, Fanni D, Monga G, Zaffanello M, Van Eyken P, Fanos V. Morphogenesis and molecular mechanisms involved in human kidney development. J Cell Physiol. 2012;227(3):1257-68.

• Faa G, Gerosa C, Fanni D, Nemolato S, Di Felice E, Van Eyken P, Monga G, Iacovidou N, Fanos V. The role of immunohistochemistry in the study of the newborn kidney. J Matern Fetal Neonatal Med. 2012;25(Suppl 4):135-8.

• Fanos V, Loddo C, Puddu M, Gerosa C, Fanni D, Ottonello G, Faa G. From ureteric bud to the first glomeruli: genes, mediators, kidney alterations. Int Urol Nephrol. 2015;47(1):109-16.

ABS 37

EPSTEIN-BARR VIRUS ENCEPHALITIS: A CASE REPORT

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INTRODUCTION

Epstein-Barr virus (EBV) usually causes mild, asymptomatic, and self-limited infections in children and adults. However, severe complications such as neurological diseases, malignant diseases, hepatic failure, and myocarditis have been rarely described. The incidence neurologic complications of following symptomatic EBV infection (infectious mononucleosis) is less than 0.5%. The pathogenesis of EBV-induced neurologic injury remains incompletely understood, although most evidence suggests that immunological mechanisms rather than active viral replication are primarily involved [1].

CASE REPORT

We describe a case of EBV encephalitis of a polish 4-year-old girl, infected by EBV in Poland, who was admitted to our hospital with 7-day history of fever, twisting and repetitive oral movements, and decreased activity. On admission, she was drowsy and presented with normal vital signs, sore throat, tonsil hypertrophy and cervical lymphadenopathy. In the first hours following admission, she exhibited seven episodes of fixed and open eyes, lockjaw, flushing, oral rumination movements, legs stiffness and unresponsiveness to verbal and painful stimuli, spontaneously resolving after few minutes. Cerebrospinal fluid (CSF) analysis revealed prominent lymphocytic pleocytosis (55 cells/mm³), hyperproteinorrachia (80 mg/dL), and a glucose level of 61 mg/dL. Serologic testing was positive for Viral Capsid Antigen (VCA) IgM and negative for VCA IgG. EBV-DNA was detected in CFS by PCR. Blood, urine, and CSF cultures were negative. Empirical intravenous antibiotic (ceftriaxone) and acyclovir (20 mg/kg per day for a total of 16 days) were administered under the suspicion of EBV encephalitis. An electroencephalogram (EEG) showed generalized slow waves, especially in waking. Brain CT scan demonstrated nonspecific evidence of brain edema. A brain MRI revealed signal abnormalities in thalami, mesencephalon (sparing pyramidal pathways and red nuclei), superior cerebellar peduncles, posterior portion of pons and medulla oblongata and hippocampus. In agreement with the pediatric neurology specialist, corticosteroids (methylprednisolone) and anticonvulsants (clobazam and levetiracetam) were introduced. The patient gradually recovered, and no further convulsions occurred. On day 21, EEG was normalized, and brain MRI was improved. At the same time, the neurological examination only revealed mild ideomotor dyspraxia, abnormal left fine movements with distal tremors, and altered left leg osteotendinous reflexes. On day 25, she was discharged home.

CONCLUSIONS

Encephalitis is a rare complication of EBV infection, whose pathogenesis and treatment are so far controversial. Previous reports suggest that acyclovir and corticosteroid therapies may be considered reasonable [2], and the good response to them, in our patient, seems to confirm it. However, prognosis is uncertain and long-term follow-up is needed to identify possible neurodevelopmental sequelae. REFERENCES

 Hoover SE, Ross JP, Cohen J. Epstein-Barr virus. In: Sheld WM, Whitley RJ, Durack DT (Eds.). Infections of the Central Nervous System, 3rd ed. Philadelphia: Lippincott-Raven Publishers, 2004, pp. 175-83.

[2] Doja A, Bitnun A, Jones EL, Richardson S, Tellier R, Petric M, Heurter H, Macgregor D. Pediatric Epstein-Barr Virus-Associated Encephalitis: 10-Year Review. J Child Neurol. 2006;21(5):385-91.

ABS 38

METABOLOMICS PATHWAYS IN SONS OF MOTHERS WHO ASSUMED NEUROPSYCHO-TROPIC DRUGS DURING PREGNANCY: A PI-LOT STUDY

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INTRODUCTION

The neuropsychotropic drugs are substances able to pass the hematoencephalic barrier and to have effects on the nervous central system. It is a heterogeneous category of drugs, including antidepressants, antipsychotics, mood stabilizers and drugs of abuse (such as cocaine, methamphetamine and opioids). If taken by a mother during pregnancy, they can exert different effects on the fetus and the underlying pathophysiological mechanism of many of them is still unclear. The application of metabolomics in this field can enlighten the metabolic alterations induced by these drugs in the fetus. The aim of our study was to investigate the possible urinary metabolome alterations of sons of mothers that took neuropsychotropic substances during pregnancy compared with a group of controls.

MATERIALS AND METHODS

In total, we collected 21 samples of urine around of the 5th day of life. The patients were divided in two groups: one included 11 samples from sons of mothers who assumed neuropsychotropic drugs during pregnancy and the other included 10 samples from a group of controls, matched for gestational age, sex and type of delivery. The first group was composed of 3 newborns exposed to valproate, 3 to methadone, 2 to antidepressants, 1 to methadone, valproate and antipsychotic, 1 to benzodiazepines and 1 to cigarette smoke. In this group, nine were born at term (6 AGA and 3 SGA), and two were preterm (both AGA). Concerning the type of delivery, five were born with spontaneous vaginal delivery and six with caesarean section. The samples were collected with a non-invasive technique, by a cotton ball in the diaper. After that, 2-3 mL of urine were withdrawn with a sterile syringe, transferred in a sterile test tube and then stored at -80°C. Then the samples were prepared for the metabolomics analysis and 1H NMR was performed. The data collected were statistically analyzed by principal component analysis (PCA). RESULTS

As a result, we did not find a significant difference between the two groups. The only exception was one case that was exposed to methadone during pregnancy. This neonate was the only one with meconium-stained amniotic fluid. His metabolomics profile was characterized by increased levels of lactose and aromatic compounds, and lower levels of formate. According to literature, increased levels of lactose were found in urine samples of rats addicted to methamphetamine [1]. As regards the aromatic compounds, the most likely is the tryptophan, precursor of the serotonin and melatonin, which is found increased in schizophrenic patients [2], and in rats addicted to heroin [3]. The increased levels of tryptophan may indicate a disruption in the pathway of the serotonin and oxidative damage. Even the purines can be implied in the increase of the aromatic compounds. Purines have a role in the energy metabolism and can be a signal of inflammation

process. Purines, such as guanine and xanthine, are found increased in the plasma metabolome of patients in therapy with methadone [4]. The formic acid is implied in the tetrahydrofolate pathway and in the metabolism of the gut microbiota. The formate is found related to the pulmonary function in patients affected by COPD [5]; it is also included in a panel of six metabolites that can distinguish between patients with major depression from patients with bipolar disorder [6].

CONCLUSIONS

In conclusion, the effect of the neuropsychotropic drugs on the fetus are still unclear, thus further studies are needed and metabolomics could be able to reveal physiopathological pathways still unknown since it is a noninvasive technique that can be easily applied in the field of neonatology and pediatrics.

REFERENCES

[1] Zaitsu K, Miyawaki I, Bando K, Horie H, Shima N, Katagi M, Tatsuno M, Bamba T, Sato T, Ishii A, Tsuchihashi H, Suzuki K, Fukusaki E. Metabolic profiling of urine and blood plasma in rat models of drug addiction on the basis of morphine, methamphetamine, and cocaine-induced conditioned place preference. Anal Bioanal Chem. 2014;406:1339-54.

[2] Fukushima T, Iizuka H, Yokota A, Suzuki T, Ohno C, Kono Y, Nishikiori M, Seki A, Ichiba H, Watanabe Y, Hongo S, Utsunomiya M, Nakatani M, Sadamoto K, Yoshio T. Quantitative analyses of schizophrenia-associated metabolites in serum: serum D-lactate levels are negatively correlated with gamma-glutamylcysteine in medicated schizophrenia patients. PLoS One. 2014;9:e101652.

[3] Zheng T, Liu L, Aa J, Wang G, Cao B, Li M, Shi J, Wang X, Zhao C, Gu R, Zhou J, Xiao W, Yu X, Sun R, Zhou Y, Zuo Y, Zhu X. Metabolic phenotype of rats exposed to heroin and potential markers of heroin abuse. Drug Alcohol Depend. 2013;127:177-86.

[4] Mannelli P, Patkar A, Rozen S, Matson W, Krishnan R, Kaddurah-Daouk R. Opioid use affects antioxidant activity and purine metabolism: preliminary results. Hum Psychopharmacol. 2009;24(8):666-75.

[5] McClay JL, Vunck SA, Batman AM, Crowley JJ, Vann RE, Beardsley PM, van den Oord EJ. Neurochemical metabolomics reveals disruption to sphingolipid metabolism following chronic haloperidol administration. J Neuroimmune Pharmacol. 2015:10:425-34.

[6] Chen J, Liu Z, Fan S, Yang D, Zheng P, Shao W, Qi Z, Xu X, Li Q, Mu J, Yang Y, Xie P. Combined Application of NMR- and GC-MS-Based Metabonomics Yields a Superior Urinary Biomarker Panel for Bipolar Disorder. Sci Rep. 2014;4:5855.

ABS 39

PEDIATRIC DIFFERENTIATED THYROID CAN-CER: EXPERIENCE OF A SINGLE CENTRE IN NAPLES

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INTRODUCTION

Thyroid nodules are uncommon in children and adolescents, however when diagnosed, carry a greater risk of malignancy compared to those in adults. Pediatric differentiated thyroid cancer (PDTC) is rare but aggressive carcinoma. Therefore, it is important to diagnose it at initial stage to prevent recurrence and/or metastasis. The objective of this study was to analyse the clinical features, the treatment and the outcome of patients with PDTC referred to our center.

METHODS AND PATIENTS

A retrospective analysis was conducted of all PDTC diagnosed in patients aged less than 18 years at the time of diagnosis. We identified 23 patients (female = 21, 91%; male = 2, 9%) with PDTC with a mean follow-up of 7.9 ± 5.3 years. During the follow-up, recurrent disease was defined on the basis of positive biochemical (thyroglobulin under TSH-suppression or after rhTSH-stimulation) and/or imaging (neck ultrasound, whole body scan and, when necessary, CT and/or PET/CT) findings.

RESULTS

8 patients (35%) had history of autoimmune disease (5 patients with Hashimoto's thyroiditis, 3 patients with type 1 diabetes) and 3 patients (13%) of radiation to treat hematological disease. 7 patients underwent total thyroidectomy, 16 patients total thyroidectomy and central and/or lateral cervical lymph node dissection. 8 patients had a micro carcinoma (≤ 10 mm), 15 had a carcinoma. In all cases the histological diagnosis was papillary thyroid cancer; 70% had classical variant and 30% follicular variant. 4 patients were free of disease after surgery and iodine-131 ablation; 12 patients had no evidence of recurrence after surgery and 2 (6 patients), 3 (5 patients) and 4 (1 patient) therapeutic doses of iodine-131; 7 patients underwent another surgery. At the end of follow-up only 4 patients (87%) had evidence of disease: 1 patient pulmonary metastasis and 3 patients persistence of Tg or AbTg (antibodies anti-Tg).

CONCLUSION

In our series papillary carcinoma is the most common PDTC. It needs surgical treatment more extensive and cumulative iodine-131 activities are higher compared to adults. However, most of patients are free from the disease and have a good prognosis. Pediatric patients had more frequently than adults history of autoimmune diseases or radiation, therefore in these cases it is recommended to perform neck ultrasound in children to identify thyroid cancer at initial stage.

ABS 40

GASTRIC PERFORATION IN AN INFANT WITH PATAU SYNDROME: A CASE REPORT

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INTRODUCTION

Trisomy 13 (Patau syndrome) is a congenital disorder with an incidence of 1:16,000. It is associated with severe neurological impairment and with diffuse physical abnormalities, including orofacial clefts, cardiac malformations, gastrointestinal and urogenital system defects [1]. Approximately 50% of infants with trisomy 13 die during the first weeks of life; only 5-10% infants survive during the first year of life [2].

CASE REPORT

Male, first child of non-consanguineous parents. Fetal MRI and ultrasound at 29 weeks revealed right microphthalmia, cerebral ventriculomegaly, prevalence of right cardiac chambers, a suspected left kidney agenesis, cryptorchidism and hypospadia. The patient was born at 35 weeks of gestation by spontaneous vaginal delivery characterized by prolonged membrane rupture, oligohydramnios and placental infarction. At birth APGAR score was 1 at the 1st minute and 6 at the 5th minute, respectively. Birth weight was 2,300 g, length was 46 cm and occipitofrontal circumference 42.2 cm (50th percentile for all the parameters). Resuscitation included intubation at 1st minute and extubation after 7 minutes. He was admitted to our neonatal intensive care unit due to respiratory distress and physical abnormalities. The newborn examination showed narrow bitemporal diameter, right microphthalmia, anteverted nostrils, post-axial polydactyly of hands and right foot with syndactyly of the 5th and the 6th finger, low-set thumb, micropenis, cryptorchidism and a sacral dimple. Laboratory findings showed: leucocytes 16,000/mm³, C-reactive protein (CRP) 0.9 mg/dl, procalcitonin (PCT) 6.53 ng/ml; an intravenous antibiotic therapy was started. Cerebral, cardiac and abdominal ultrasound on the 1st day

of life revealed a mild asymmetry of the cerebral ventricles, a patent ductus arteriosus and a small ptotic left kidney with the presence of both testes in the inguinal canal. The infant received a noninvasive ventilation and nasogastric enteral nutrition was started on day 2. A clinical worsening happened on the 4th day of life, when the infant had a significant abdominal distension with respiratory distress; the abdominal radiograph showed abdominal effusion with pneumoperitoneum. CRP increased to 52 mg/ dl. An evacuative paracentesis was performed, draining about 60 milliliters of milk. The baby was immediately transferred to paediatric surgery where he underwent a surgical intervention for gastric perforation with the placement of a peritoneal drainage for 5 days. After his return from surgery, his clinical conditions worsened progressively; he was intubated with high oxygen needs; laboratory test showed further increases of serum CRP and PCT and creatinine; he required platelets and blood transfusions; for the occurrence of tonic movements of upper limbs, phenobarbital therapy was introduced. The infant died on the 21st day of life. Chromosome analysis demonstrated trisomy 13 (47, XY +13).

CONCLUSIONS

Infants with Patau syndrome might have gastrointestinal malformations. In our case, gastric perforation, a rare event in the neonatal population [3], could be a consequence of the chromosomal abnormality.

REFERENCES

 [1] Jones KL, Jones MC, Del Campo M. Smith's Recognizable Patterns of Human Malformation. 7th ed. Philadelphia, PA: Elsevier Saunders, 2013, pp. 14-16, 20-21.

[2] Rasmussen SA, Wong L-YC, Yang Q, May KM, Friedman JM. Population-based analyses of mortality in trisomy 13 and trisomy 18. Pediatrics. 2003;111(4 pt 1):777-84.

[3] St-Vil D, LeBouthillier G, Luks FI, Bensoussan AL, Blanchard H, YoussefS. Neonatal gastrointestinal perforations. J Pediatr Surg. 1992;27:1340-2.

ABS 41

PLEOMORPHIC ADENOMA OF THE SUB-MANDIBULAR GLAND: A CASE REPORT IN A CHILD

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INTRODUCTION

Salivary gland tumors are rare in children. Pleomorphic adenoma is the most common pediatric salivary gland tumor and the most common benign tumor in childhood. Mainly it occurs in the parotid gland, whereas the palate represents the most common site of minor salivary gland involvement. An associated chronic sialadenitis has been also reported in children.

CASE REPORT

We report a case of pleomorphic adenoma occurring in the submandibular gland in a 12-yearold girl with an underlying chronic sialadenitis. The tumor, presented as an oval swelling in the left submandibular region, gradually increasing in size, during the last 9 months. Submandibulectomy was performed. On gross examination the tumor appeared tan-white in color, surrounded by a fibrous capsule. The microscopic appearance showed a fibrous capsule, with variable thickness, surrounding the tumor that was formed by an admixture of epithelial and mesenchymal cells. Epithelial elements were organized in nests or glandular like structures. Tumor cells showed large polymorphous atypical nuclei, with irregular nuclear membrane. The mesenchymal components were characterized by a spindle nucleus, with dense compact chromatin, and were embedded in a myxoid matrix, with focal areas, of chondroid differentiation. The tumor showed pushing margins, focally extending into the fibrous capsule. Some scattered foci of capsular infiltration were occasionally detected (Fig. 1). The proliferative index of tumor cells, detected by Ki67 immunostaining, reached levels around 10% in some tumor areas characterized by the highest frequency of atypical tumor cells. Mitoses were rare. No hemorrhages, nor intratumoral necrosis have been observed. The residual submandibular gland showed a lymphomonocytic interstitial infiltrate, sometimes organized in lymphoid nodules. A diagnosis of atypical pleomorphic adenoma was performed. After 1 year of follow-up, the patient is in good health, in the absence of any recurrence.

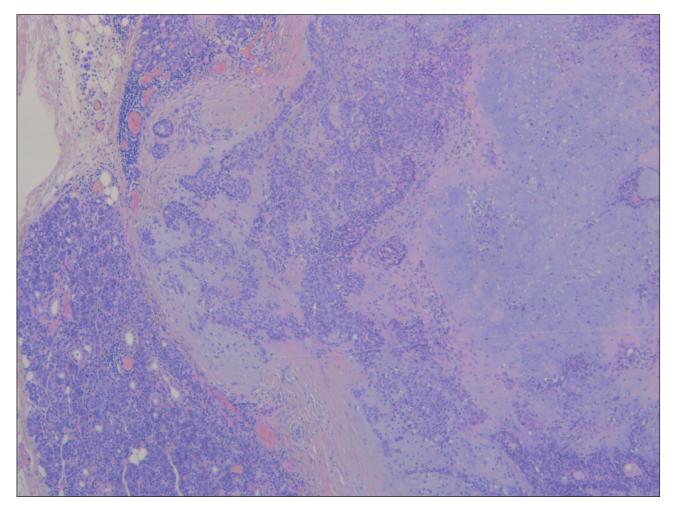


Fig. 1 (ABS 41). The tumor pushing margins were characterized by cones of pressure inside the capsule.

DISCUSSION

The case here reported confirms the peculiarities of clinical and pathological features of the pleomorphic adenoma arising in childhood. This tumor, when it occurs in pediatric age, has an increased risk of an aggressive behavior, and an higher frequency of atypical histological and immunohistochemical features. A careful examination of the eventually capsular infiltration, an evaluation of the proliferative index of tumor cells, with Ki67 immunostaining, appear mandatory, in order to identify the tumors with a possible aggressive potential.

ABS 42

PRIMARY CONGENITAL HYPOTHYROIDISM IN DISCORDANT TWINS: THE ROLE OF EPI-GENETICS. A CASE REPORT

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INTRODUCTION

Twins are widely reported to have more morbidity than singletons, mainly because of a higher preterm birth rate, that can affects thyroid function. Congenital primary hypothyroidism (CH) is estimated to occur in about 1:2,500 newborns and surprisingly, monozygotic (MZ) twins are usually discordant for CH due to thyroid dysgenesis. A possible explanation for this phenotypic discordance is the existence of epigenetic differences. To address this issue, we report a case of CH in one of two MZ twins.

CASE REPORT

J. and D. are MZ twins born at 32 w. CH screening test by capillary blood sampling was performed for both of them 48 hours after birth and judged to be suspicious for discordant CH. J's TSH level resulted pathologically increased and CH hypothyroidism was then confirmed by venous blood sampling (TSH = 75 μ IU/mL, fT4 < 0.3 ng/dL). Replacement therapy was started with levothyroxine at an initial daily dose of 12 μ g/kg b.w. D. has been re checked, according Congenital hypothyroidism guidelines, at 2 w of life, because of fetal blood mixing that can occurs between twins. Laboratory re-evaluation was performed on their 12th day of age and D. showed a normal thyroid function. Thyroid ultrasound evaluation on J. showed thyroid agenesis. D. presented a left-sided renal ectopy. Both J. and D. have normal neurological development, according to their age.

CONCLUSIONS

The INRICH (Italian National Registry of Infant with Congenital Hypothyroidism) data show a more than 3-fold higher frequency of twins in the CH population than in the general one, especially in low birth weight and low gestational age twins, reporting a discordant rate of 95.7% [1]. MZ twinning is associated with an increased rate of epigenetic alterations. Literature shows no precise risk factors with a maternal origin. The occurrence of the disease could be related to no inheritable post zygotic events including epigenetic modifications, such as specific methylation/ demethylation patterns in thyroglobulin gene and TSHR gene. Moreover it is possible that the post zygotic events create two different cell populations in the inner cell mass, stimulating the twinning process. If there is a unequal distribution of these different cell populations between the two embryos, the development of specific organs, such as the thyroid, could differ between MZ twins. The stochastic nature of these postzygotic events is highlighted by our case index twins, they are also discordant for left-sided renal ectopy [2]. Olivieri et al. hypothesize a possible competitive conditions between the twins about the metabolic factors in utero, the blood supply, and the nutrients (especially iodine) that could differentiate the development of the gland [2]. Future studies should address the specific mechanisms responsible for the observed epigenetic drift of MZ twins. REFERENCES

[1] Cassio A, Corbetta C, Antonozzi I, Calaciura F, Caruso U, Cesaretti G, Gastaldi R, Medda E, Mosca F, Pasquini E, Salerno MC, Stoppioni V, Tonacchera M, Weber G, Olivieri A; Italian Society for Pediatric Endocrinology and Diabetology; Italian Society for the Study of Metabolic Diseases and Neonatal Screening; Italian National Institute of Health; Italian National Coordinating Group for Congenital Hypothyroidism; Italian Thyroid Association; Italian Society of Pediatrics; Italian Society of Neonatology; Italian Society of Endocrinology; Associazione Medici Endocrinologi. The Italian screening program for primary congenital hypothyroidism: actions to improve screening, diagnosis, follow-up, and surveillance. J Endocrinol Invest. 2013;36(3):195-203.

[2] Olivieri A, Medda E, De Angelis S, Valensise H, De Felice M, Fazzini C, Cascino I, Cordeddu V, Sorcini M, Stazi MA; Study Group for Congenital Hypothyroidism. High risk of congenital hypothyroidism in multiple pregnancies. J Clin Endocrinol Metab. 2007;92(8):3141-7.

ABS 43

THE EFFICACY OF VENTED BASE FEEDING BOTTLE IN NEWBORNS: A RANDOMIZED

CLINICAL TRIAL. THE ROLE OF A RESEARCH PEDIATRIC NURSE

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INTRODUCTION

Vented-base feeding bottles allow a homogeneous milk flow more adequate to newborns requests thus producing a sucking pattern more similar to that of breastfeeding [1]. In our Neonatal Intensive Care Unit (NICU) a pilot study conducted on 10 newborns showed that standard feeding bottles are associated to shorter feeding time (p = 0.04) with less organized swallow-respiration patterns while vented-base feeding bottle are associated with a lower number of desaturation and reflux events (p = 0.04) [2, 3]. The subsequent clinical trial (ClinicalTrials.gov ID: NCT03031288) currently going on in our NICU is designed for the active participation of a pediatric nurse. Therefore, a scholarship funded by the University of Turin was assigned to a research pediatric nurse from April 2017. Aim of the present study is to describe the role of a research pediatric nurse and to evaluate her impact on our NICU.

MATERIALS AND METHODS

In this descriptive study a modified Italian Clinical Trials Nursing Questionnaire (CTNQ) [3] was proposed to 5 members of a medical-nursing team selected for their involvement in research projects. Each item of the CTNQ was scored by the interviewed subjects to describe the type of activity, the amount of work and the contribute of the research nurse to daily nursing activities. RESULTS

The CTNQ was completed by all the 5 members: a clinician, a junior medical doctor, the head nurse and two nurses. The research pediatric nurse contributes to coordinate the clinical trial identifying the eligible newborns and participating in the consensus form collection. The nurse follows the enrolled newborns along their participation into the study managing cardiorespiratory monitoring, administering the feeds with the different devices and informs and communicates with the parents. She is responsible for the collection of data and the management of the database and contributes to the analysis of data and the creation of reports.

CONCLUSIONS

The research pediatric nurse represents a connection between daily clinical and nursing practice and research activities. Her involvement guarantees a uniform conduction of the trial and provides the information flux among different people involved into the trial. She allows research activities to be carried out without overburdening nursing staff involved in daily care activities.

REFERENCES

[1] Geddes DT, Sakalidis VS, Hepworth AR, McClellan HL, Kent JC, Lai CT, Hartmann PE. Tongue movement and intra-oral vacuum of term infants during breastfeeding and feeding from an experimental teat that released milk under vacuum only. Early Hum Dev. 2012;88(6):443-9.

[2] Cocchi E, Zavattero C, Maggiora E, Peila C, Coscia A, Bertino E, Cresi F. Vented base feeding bottle in preterm infants with GERD symptoms: effects on respiration-deglutition patterns and reflux. In: Selected Abstracts of the 12th International Workshop on Neonatology; Cagliari (Italy); October 19-22, 2016. J Pediatr Neonat Individual Med. 2016;5(2):e050247.

[3] Catania G, Poirè I, Dozin B, Bernardi M, Boni L. Validating a measure to delineate the clinical trials nursing role in Italy. Cancer Nurs. 2008;31(5):E11-15.

ABS 44

A CASE OF NORMOCYTIC ANEMIA IN AN IN-FANT WITH VITAMIN B12 DEFICIENCY

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INTRODUCTION

Vitamin B12-deficiency can occur as a consequence of prolonged exclusive breastfeeding in infants whose mothers are vegetarian, and often becomes clinically evident between 4 and 12 months. This vitamin deficiency presents with hematological (megaloblastic anemia and, in severe cases, neutropenia and thrombocytopenia) and neurological features (hypotonia, lethargy or irritability, neurocognitive retardation and, rarely, brain atrophy). We report a case of an 11-month-old girl with severe vitamin B12 deficiency, admitted to our Clinic with severe anemia, leucopenia, hypotonia and lethargy. CASE REPORT

The patient was born at term after an uncomplicated pregnancy and delivery (birth weight: 3,630 g; length: 52 cm; head circumference: 35.5 cm). She was exclusively breastfed until hospital admission, as weaning was never started. Her mother was a strict vegetarian, who was not under regular treatment with vitamin B12, except for a course of supplementation that had been carried out during the previous pregnancy due to the finding of macrocytic anemia. Neurological development and growth of the patient were reported to be normal until the age of 7 months. The infant was evaluated by the family pediatrician, due to the occurrence of increasing pallor, hypotonia and weakness. Moreover, severe normocytic anemia (Hb, 6.7 g/dL; MCV, 80 fL), and leucopenia (WBC, 3,300/µL; Neutrophils, 8.8%) were found. On admission, the severe anemia required a transfusion of packed red blood cells resulting in a prompt increase of Hb concentration (Hb: 10.6 g/ dL). Under the suspicion of leukemia, bone marrow needle aspiration was carried out. The bone marrow examination showed a polymorphic picture, with no evidence of leukemic blasts but rather particularly rich in megaloblasts. The finding of low serum vitamin B12 levels (< 83 pg/mL), together with the characteristic bone marrow features, supported the diagnosis of severe vitamin B12 deficiency. Two intramuscular doses of vitamin B12 (1 mg each) were given 5 days apart, leading to the resolution of leucopenia (WBC 9,480/ μ L, neutrophils 2,500/ μ L) and the normalization of vitamin B12 serum level (215 pg/mL), on day 10 of hospitalization. The neurological picture showed a progressive improvement in reactivity and muscle tone, even if these findings were not completely normal for age, at discharge. During hospitalization, weaning was started.

CONCLUSIONS

The mother's diet and her hematological status during pregnancy and breast-feeding should be always carefully evaluated. It is also important to assess the nutritional status of infants with prolonged exclusive breastfeeding for the early prevention and treatment of nutritional deficiencies. Our report shows that normocytosis can be a misleading feature, hiding the typical hematological features of vitamin B12 deficiency. Therefore, the serum level of this vitamin should be measured when anemia and neurological manifestations are associated, also in absence of macrocytosis.

ABS 45

THE CHANGING PATTERN OF HEAD INJURY: REPORT OF TWO CASES

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INTRODUCTION

Head injury (HI) is a common presentation in pediatric emergency rooms (PER) and when associated to brain injury (BI) it's a leading cause of death or disability. In mild HI (90-95% of overall cases) BI emerges in 5% of cases [1]. CT scan is the gold standard for BI assessment while MRI is a second line exam based upon CT abnormalities and/or clinical conditions [2]. The latter can change quickly after traumas. We report two cases of HI occurred to two 14 years old girls in whom clinical observation was pivotal in the decision-making process and final diagnosis. CASE 1

The patient came to our PER after a car crash in which she reported an orbital laceration wound. She was well and conscious despite amnesia about the accident. Brain CT showed a left parietal fracture, apparently uncomplicated. Initial neurological examination was normal even if incomplete due to bandages. The girl was sutured, then admitted to the pediatric unit for clinical observation and monitoring. On day 2 mild asymmetry of smile and tiny hypomobility of the left side of the face emerged. On day 3 some bandages were removed thus the pattern became evident with asymmetry of smile and incomplete closure of the left eye. MRI was performed therefore. It showed a small left fronto-parietal subdural hematoma and a smooth enhancement of the intramastoid tract of the facial nerve, consistent with post-traumatic edema (Fig. 1A). Notably, the parietal fracture did not affect the facial nerve course. The facial palsy was favorably treated with oral steroid therapy and pyridoxine. CASE 2

The girl came to our PER because of an occipital trauma due to an episode of postural syncope. At the PER neurological examination was normal but she complained relevant headache, vomiting and an occipital wound. A brain CT was performed at admission showing an occipital fracture without BI. Despite painkillers the headache persisted so a CT scan was repeated. A 5mm cortical junctional contusion at the right frontal lobe was detected. Therefore a brain MRI was performed. Despite confirming the CT scan, MRI images showed a complex pattern characterized by diffuse contusion (**Fig. 1B** and **Fig. 1C**) foci at the frontal and the parietal right lobes, small bitemporal subarachnoid hemorrhages, and a few leaks of frontal subdural cerebrospinal fluid. The EEG showed

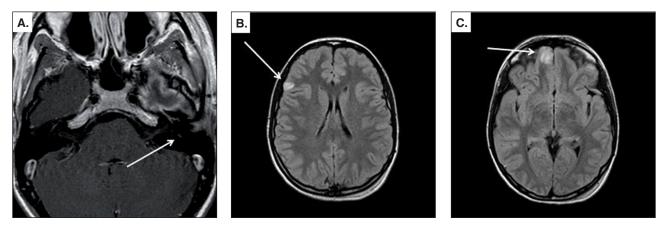


Fig. 1 (ABS 45). Two cases of head injury. A. Case 1: post traumatic edema of the left facial nerve. B, C. Case 2: brain contusions in the right frontal lobe.

a slightly asymmetric activity, without epileptic features. The girl was treated by continuing the pain killer therapy, obtaining headache improvement and EEG normalization. She was discharged on day 5. An MRI will be scheduled for follow-up.

CONCLUSIONS

Both cases show that, even without consciousness, clinical signs and symptoms indicate the emerging of a secondary or late involvement of central nervous system after an HI. Thus even according to the literature [3] it seems very difficult, if possible, to schedule checks of imaging after an HI with a standard timetable as clinical observation is the main indicator for emerging complications.

REFERENCES

[1] https://www.nice.org.uk/guidance/cg176, last access: June 2017.

[2] Da Dalt L,Bressan S, Parri N, Nocerino A, Amigoni A, Selmin S, Manara R, Peretta P, Vardeu M P. Approccio in Pronto Soccorso al bambino con Trauma Cranico Linea Guida basata sull'evidenza a cura della Società Italiana di Medicina di Emergenza-Urgenza Pediatrica e della Società Italiana di Pediatria. Riv Urg Emerg Ped. 2015;2;16-104.

[3] Aziz H, Rhee P, Pandit V, Ibrahim-Zada I, Kulvatunyou N, Wynne J, Zangbar B, O'Keeffe T, Tang A, Friese RS, Joseph B. Mild and moderate pediatric traumatic brain injury: replace routine repeat head computed tomography with neurologic examination. J Trauma Acute Care Surg. 2013;75(4):550-4.

ABS 46

PEMPHIGUS VULGARIS IN CHILDHOOD: A CASE REPORT

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INTRODUCTION

Pemphigus vulgaris (PV) is an uncommon autoimmune bullous dermatosis affecting the skin and mucous membranes. The disease is caused by autoantibodies against desmoglein 1 and 3, which are keratinocyte adhesion proteins located in the epidermis and external mucosae. PV usually affects adults; the pediatric variant is very rare, and has been classified into childhood PV (0-12 yrs) and adolescent PV (13-18 yrs).

CASE REPORT

A previously healthy 9-year-old boy was evaluated for the occurrence of multiple blisters in mucous membranes and skin during the last 2 months. Initially, blisters were located in the scalp, navel and genital area, and then spread throughout the body and oral mucosa. On examination, skin lesions in different evolutionary phases included flaccid vesicles and blisters, erosions, and multiple crusted lesions predominantly in the scrotum and neck root. The Nikolsky's sign was positive. Painful erosions on the oral mucosa were found. In suspected autoimmune bullous dermatosis, the following laboratory tests were carried out: Complete Blood Count (CBC), inflammatory markers, C3, C4, protein electrophoresis, ferritin, vitamin B12, LDH, TORCH, immunodiffusion, autoimmunity markers such as c-ANCA, p-ANCA, ENA screen, thyroid profile, anti-Thyroid Peroxidase antibody, celiac profile, and tuberculin skin test. To exclude paraneoplastic syndromes, chest X-rays, abdominal US scan and blood smear were performed. All laboratory and instrumental examinations were negative except for antidesmoglein 1 (196.57 RU/mL) and 3 (349.00 RU/mL) antibodies, and ANA. Histological examination of skin biopsy specimens showed intraepidermal suprabasal acantholysis, and lymphoplasmacellular infiltrate in the superficial dermis. Direct immunofluorescence revealed the positivity of keratinocytes for IgG and C3. Clinical, histological and laboratory features confirmed the diagnosis of PV. Therefore, prednisone therapy was administered at the daily dose of 2 mg/kg for 2 weeks, with partial response. Thus, dapsone was associated with an initial dose of 0.8 mg/kg per day, followed by a gradual increase to 1.5 mg/kg per day. During therapy, blood pressure, body weight, blood glucose, MetHb and CBC were monitored. Furthermore, oral clarithromycin and IV teicoplanin and gentamicin were given due to the culture positivity (S. epidermidis and K. oxytoca) of skin swabs. Combined therapy (prednisone and dapsone) induced a gradual but incomplete regression of skin and mucous lesions. After discharge, the patient has been enrolled in a follow-up program.

CONCLUSIONS

Pediatric PV is a very rare condition that usually has a favorable prognosis, with the majority of patients achieving remission within weeks to months. The treatment consists of corticosteroids alone or in combination with dapsone; in our patient, the response to combined therapy has been only partial. In severe cases, additional therapies (IVIG, cyclosporine, azathioprine, and rituximab) have been used.

ABS 47

WHAT ROLE DOES ULTRASONOGRAPHY PLAY IN THE STUDY OF PEDIATRIC CHEST?

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INTRODUCTION

In some cases the ultrasonography (US) may be useful applied as a complement to chest X-ray, as follow up study and for therapeutic management. MATERIALS AND METHODS

The main applications of MRI in the study of chest pathology are pleural effusion (anechoic, echogenic without disruption, with septa, internal solid components), thoracic empyema, complicated pneumonia (necrosis areas, abscess); in these cases US is also useful in therapeutic management. Other indication for MRI is follow up of pneumothorax and peripheral pulmonary solid lesions. More generally, the US is applied with high sensitivity and specificity in the study of mediastinal masses, thyme evaluation, and thoracic wall lesions. A recent application is the follow-up of inflammatory pulmonary lesions based on a first radiographic examination.

RESULTS

The US has a limited role in the study of lung disease due to the acoustic barrier caused by the bone structure of the chest cavity and lung air content, which does not allow ultrasound transmission and does not allow detailed study of lung, airways and pulmonary interstitial.

CONCLUSIONS

The advantages of lung US study are of interest especially in terms of reducing exposure to ionizing radiation, as well as repeatability, ease of execution, but there are still many perplexities. There is currently no evidence of high sensitivity and specificity of the US study in the diagnosis of emergency lung disease, for example in intensive care.

REFERENCES

• Ball L, Vercesi V, Costantino F, Chandrapatham K, Pelosi P. Lung imaging: how to get better look inside the lung. Ann Transl Med. 2017;5(14):294.

• Tomà P. Lung ultrasound may not be reliable alternative to chest radiography in childhood pneumonia. Chest. 2017;151(1):244.

• Tomà P. Sonographic diagnosis of pneumonia: it is time to change. Br J Radiol. 2017;90(1069):20160799.

ABS 48

CLINIC-PATHOLOGICAL FEATURES OF PE-DIATRIC MYCOSIS FUNGOIDES: TWO CASES DESCRIPTION

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INTRODUCTION

Mycosis fungoides (MF) is the most common cutaneous T-cell lymphoma, its incidence is 1 per 1,000,000 per year in adults. The occurrence in children is rare and it's estimated from 0.5% to 7% with only the 5% of all cases observed before

the age of two [1]. There are several distinct clinical forms of MF, which have clinical and histological resemblances with some common inflammatory skin disorders, such as eczema, psoriasis, tinea, pityriasis versicolor. As a consequence, the diagnosis of pediatric MF is commonly delayed.

CASE REPORTS

Two female children presented to our Department, the first with a history of atopic dermatitis lasting since the age of 6 months, characterized by pruritic poikilodermic finely scaly roundish 5-10 cm plaques over her trunk, groins and proximal extremities. A biopsy taken revealed a CD8+ CD4- mycosis fungoides (Fig. 1A and Fig. 1B). The second patient, a 2-year-old girl, came to our attention complaining of an eczematous sharply circumscribed verrucous plaque on the dorsal aspect of her hand. The mother first notice the lesion at the age of 1 and subsequently the size and elevation of the plaque gradually increased. A biopsy taken showed an epidermotropic infiltrate of CD8+ atypical lymphocytes with hyperchromatic and pleomorphic nuclei. Infiltrating as individual cells or clusters in the epidermis

featuring a rare variant of MF, the localized pagetoid reticulosis, also known as Woringer-Kolopp disease (**Fig. 1C** and **Fig. 1D**).

CONCLUSIONS

Delayed diagnosis of pediatric MF is mostly attributable to the rarity of the disease but also to the different clinicopathological features that can be challenging for both the dermatologist and the pathologist. Clinical presentation can be classical with erythematous patches and plaques but also atypical variants are described, often even combined [2]. Histologically, it is characterized by the proliferation of atypical T lymphocytes confined to the basal layer of the epidermis, with also epidermotropism and Pautrier microabscesses. Unlike adults, pediatric MF most frequent shows the cytotoxic CD8+ CD4phenotype especially in the hypopigmented variant. The pagetoid reticulosis differently express prominent pagetoid epidermotropism of atypical lymphocytes with hyperchromatic and hyperconvoluted nuclei. Furthermore, pediatric MF rarely demonstrates the clonal T-cell receptor gene rearrangement. Pediatric MF is an uncommon and challenging disease,

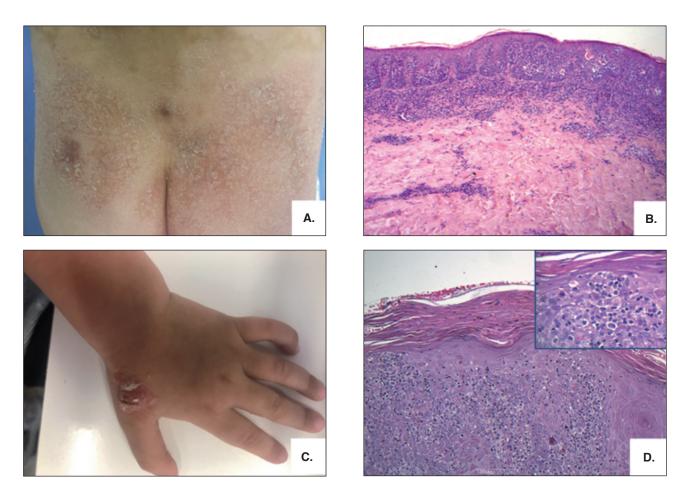


Fig 1. (ABS 48). A, B. First girl with a history of atopic dermatitis: a biopsy taken revealed a CD8+ CD4- mycosis fungoides. **C, D.** Second girl with an eczematous sharply circumscribed verrucous plaque on the dorsal aspect of her hand. A biopsy taken showed an epidermotropic infiltrate of CD8+ atypical lymphocytes with hyperchromatic and pleomorphic nuclei.

which resembles frequent skin disorders; therefore suspicious skin lesions should be biopsied to avoid diagnostic delays, especially in children. REFERENCES

 Gameiro A, Gouveia M, Tellechea Ó, Moreno A. Childhood hypopigmented mycosis fungoides: a commonly delayed diagnosis. BMJ Case Rep. 2014;2014:bcr2014208306.

[2] Matsuzaki Y, Kimura K, Nakano H, Hanada K, Sawamura D. Localized pagetoid reticulosis (Woringer-Kolopp disease) in early childhood. J Am Acad Dermatol. 2009;61:120-3.

ABS 49

EXPLORING LIM1 AMONG OLD AND NEW FUNCTIONS

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INTRODUCTION

In mammals, genital tract embryogenesis is a complex multistep process and alterations in each of these stages can cause different levels of damage to the organ development. Although numerous genetic and molecular determinants taking part in these processes have been already identified, further studies are needed to fully clarify all the responsible factors. Lim1 is a LIM-Homeodomain transcription factor whose expression is involved in the development of many organs, including several structures of the female genital tract. This work focuses on the importance of Lim1 during the endometrial development.

UTERINE EMBRYOGENESIS

Oviducts, uterus, cervix and the higher vaginal portion develop from Müllerian Ducts (MD), which originate from mesonephric epithelial-like cells resident in the area between pro- and mesonephros [1, 2]. MD formation occurs through three phases: initiation, invagination and elongation; many transcription factors and signalling pathways contribute to these processes [2, 3]. During initiation, expression of Lhx1 starts in the cranial mesonephric progenitor cells that will develop into MD epithelium [3]. The second phase is the invagination of Müllerian cells expressing Lhx1, up to reach WD and the structures of urogenital sinus. Elongation phase starts from the contact of invaginated MD and WD; for this reason, the genes involved in WD development, such as Lim1, result crucial also for MD formation [1-3]. Endometrium initially consists in a packed columnar epithelium, while the formation of the glands occurs successively. Myometrium and Fallopian tubes develop from the mesenchimal tissue around MD [2].

LIM1 (A.K.A. LHX1) FAMILY

Lhx1 is a LIM-Homeodomain transcription factor crucial in WD and MD development; its absence leads to the failure of urogenital structures formation. Lhx1 also occurs in the differentiation and development of head, neural and lymphoid tissues, kidney and retina formation, cerebellar cell support [1, 2]. Lhx1 knockout (cKO) female mice are sterile, show a short oviduct and the absence of uterus, cervix and the upper vagina; moreover, cellular death and low proliferation rate have been detected in MD epithelium, contributing to the elongation defects. The evidence attests the essential role of Lhx1 for female reproductive tract formation and also suggests the necessity to fully understand epithelial and mesenchimal tissues interactions during the uterine development[1].

HUMAN PATHOLOGIES LHX1-RELATED

Five types of Lhx1 mutation are related to Mayer-Rokitansky-Küster-Hauser (MRKH) syndrome, characterized by the absence of uterus and upper vagina [3]. Moreover, the heterozygosis missense Lhx1 mutation has been detected in Müllerian aplasia (uterine and vaginal absence) [1].

CONCLUSIONS

Specific markers expressed only by adult stem cells (SC) have not been demonstrated. Lim1 is fundamental during endometrial development, so that we supposed it could also be identified as a possible marker of endometrial SC.

REFERENCES

[1] Huang CC, Orvis DG, Kwan KM, Behringer RR. Lhx1 is required in Müllerian duct epithelium for uterine development. Dev Biol. 2014;389(2): 124-36.

[2] Rey R, Josso N. Chapter 7. Sexual Differentiation. Available at: http:// www.endotext.org/pediatrics/pediatrics7/pediatrics7.html, date of publication: 2011, last access: September 2017.

[3] Mullen RD, Behringer RR. Molecular genetics of Müllerian duct formation, regression and differentiation. Sex Dev. 2014;8(5):281-96.

ABS 50

NEONATAL ATRIAL FLUTTER AND SYN-CHRONIZED ELECTRICAL CARDIOVERSION: YOU CAN DO IT IN NEONATAL INTENSIVE CARE UNIT!

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INTRODUCTION

Fetal dysrhythmia is reported in approximately 1-2% of all pregnancy. The most common fetal arrhythmia is fetal extrasystoles accounting for 85-90%, these are often paroxysmal and usually they resolve spontaneously [1]. The most common tachyarrhythmia is supraventricular tachycardia (SVT); atrial flutter (AF) is a rare arrhythmia that usually occurs later in pregnancy compared to SVT. AF is sustained by a circular macroreentrant pathway within the atrial wall, whereas the atrioventricular (AV) node is not part of the reentry circuit. Atrial rate ranges between 300-500 beats per minute (bpm), which is commonly associated with 2:1 AV conduction and ventricular rates between 150-250 bpm. Usually echocardiography shows absence of structural heart disease. Electrocardiogram (ECG) diagnosis is straightforward with saw-tooth flutter waves [2]. AF is usually tolerated and fetal hydrops and death are uncommon. The treatment aim during pregnancy is to suppress the arrhythmia or, if this is not achieved, to slow the ventricular rate. If the fetus is non-hydropic usually medical transplacental therapy, with either oral digoxin or flecainide, is effective [3]. When AF persists after birth, since medical therapy is controversial, sinus rhythm can be restored by synchronized electrical cardioversion. CASE REPORT

N. is a male newborn, first child of nonconsanguineous parents. The mother had a history of surgery for interatrial defect and ablation for SVT. At 33 week of gestational age the fetal echocardiogram showed tachyarrhythmia with mean heart rate (HR) of 280 bpm, neither structural heart disease nor hydrops. The mother started therapy with oral digoxin and flecainide with just a reduction of fetal HR. The pregnancy was monitored and a cesarean section was performed at 35⁺² weeks. Apgar 8-9, weight 2,629 g. He was admitted to Neonatal Intensive Care Unit (NICU). ECG showed tachyarrhythmia with 280 bpm, echocardiogram confirmed no congenital heart disease but a poor contractility and an important atrial dilatation. The diagnosis of AF was performed after an adenosine bolus that increased AV block and showed more clearly the typical atrial waves with mean atrial rate of 320 bpm. The patient remained hemodynamically stable. First amiodarone was administered (bolus and continuous infusion) without suppression of AF. After 24 hours conversion to sinus rhythm was obtained by synchronized cardioversion (1 Joule/kg), prior anesthesia (Fig. 1). No cardiac therapy was administered after cardioversion. The baby ECG and Holter-ECG confirmed stability of sinus rhythm, no recurrence of AF. On follow-up the infant had good clinical condition and was discharged to home on day 20.

CONCLUSIONS

This case report suggests that synchronized electrical cardioversion is a safe procedure that can be executed in a NICU in the presence of neonatologist and cardiologist.

REFERENCES

[1] Rasiah SV, Ewer AK, Miller P. Prenatal Diagnosis, Management and Outcome of Fetal Dysrhythmia: a tertiary Fetal Medicine Center Experience over an Eight-Year Period. Fetal Diagn Ther. 2011;30:122-7.

[2] Jaeggi E, Öhman A. Fetal and Neonatal Arrhythmias. Clin Perinatol. 2016;43(1):99-112.

[3] Crochelet AS, Jacquemart C, Massin M. [Repeated electrical cardioversions and amiodarone for recurrent neonatal atrial flutter]. [Article in French]. Arch Pediatr. 2015;22(10):1032-4.

ABS 51

PULSE OXIMETRY SCREENING FOR CON-GENITAL HEART DEFECTS IN THE FIRST 24 HOURS OF LIFE

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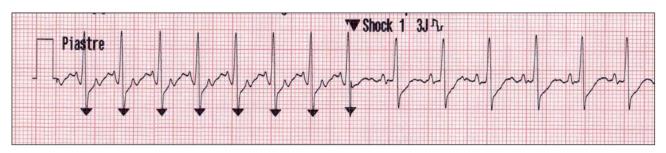


Figure 1 (ABS 50). ECG showing the shock: restored sinus rhythm.

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INTRODUCTION

Congenital heart defects (CHD) are the most common group of congenital malformations. As symptoms and signs could be unclear and aspecific, 20-30% of CHD are still missed by postnatal physical examination. Pulse oximetry measuring of arterial oxygen saturation (SatO₂), between 24 and 48 hours of life, is an established screening tool for early detection of CHD. Since several cases of CHD may decompensate during the first day of life, earlier testing may help to detect these patients before clinical deterioration. Aim: To assess the usefulness of pulse oximetry screening within 24 hours of life for early detection of CHD.

MATERIALS AND METHODS

A prospective study was conducted in all healthy newborns consecutively born in the Neonatal Unit of Santa Maria alle Scotte General Hospital (Siena) between February 16th, 2016 and April 8th, 2017. SatO₂ measuring was performed preductally and postductally on each newborn prior to physical examination between the second and the twentyfourth hour after birth. Established cut off for positive values of the screening were: a) preductal or post-ductal SatO₂ < 90%; b) pre-ductal or post-ductal SatO₂ < 95% in three consecutive registrations; c) a difference between pre-ductal or post-ductal SatO₂ > 3% in three consecutive registrations. Newborns with positive results were referred for an echocardiogram and for further investigation, including hemogasanalysis, hemochrome, C reactive protein and blood culture. RESULTS

During the reference period, 859 newborns were screened; among them, 14 (1.6%) had positive POS results. Among the newborns with positive POS, 8 (57.1%) were affected by CHD (1 aortic coartaction, 1 anomalous partial venous return, 1 interventricular defect [VSD], 2 patent ductus arteriosus, 2 interatrial defects and 1 aneurysm of the interatrial septum). The analysis of the risk ratio showed that pulse oximetry screening has a statistically significant probability to detect CHD (p < 0.001). The Relative Operating Characteristic (ROC) curve analysis showed a positive predictive value with a considerable trend toward significance (p = 0.065). All the newborns with positive POS but not affected by CHD, were affected by other conditions requiring further intervention, such as

perinatal infection or lung diseases: the ROC curve analysis for detection of non cardiac diseases by POS showed a highly significant positive predictive (p < 0.001).

CONCLUSIONS

Pulse oximetry screening within 24 hours of life resulted an accurate, non-invasive test for early detection of CHD. Although earlier testing has an higher rate of false positive results in comparison to later screening, our study demonstrated the usefulness of POS in the detection of diseases, such as respiratory problems and infections, which are potentially serious illnesses, representing an additional advantage to earlier screening.

ABS 52

TAKE CARE OF CHILDREN. THE MEDITERRA-NEAN DIET IN THE FIRST 1,000 DAYS OF LIFE. A HEALTHY LIFE. WITH THE CONTRIBUTION OF "SARDEGNA RICERCHE" UNDER THE PROJECT "SCIENTIFIC SCHOOL 2017"

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The importance of nutrition as a way to live a long, healthy and happy life is well known since the Ancient Greeks. In fact, about 2,500 years ago Hippocrates stated: "let food by thy medicine and medicine by thy food". Today, it is acknowledged that nutrition is fundamental since the beginning of life of an individual, starting from pregnancy or even before. According to the theory of "fetal programming" and Developmental Origins of Health and Disease (DOHaD), the health outcome of a human being is determined during pregnancy and the maternal nutritional state is one of the crucial variables involved. In fact, in a very recent study, it has been found that high adherence to Mediterranean Diet prior to pregnancy seems to guarantee a gestational weight gain inside the Institute Of Medicine recommendations [1]. Furthermore it seems that following this nutritional regimen during pregnancy may low the risk for the newborn to develop atopy, wheezing, asthma and

gastroschisis and reduced waist circumference, which is a marker for obesity. More generally the Mediterranean Diet is considered to be the golden standard in preventive medicine. Indeed, it may be useful to prevent cardiovascular diseases, metabolic disorders, certain cancers and to preserve cognition even later in life (reduction of symptoms of Alzheimer disease from animal studies) [2]. It had also been found that poor nutritional choices in preschoolers (2 to 4 years old children) with food high in fat, salt and sugar are associated with reduced score in verbal and cognitive ability [3]. One of the main actors of Mediterranean Diet is olive oil, which has been used since 2000 BC, according to documents found in Syria that state that its cost was five times higher than the wine. It was thought to be so beneficial that both in the Bible and in the Quran is defined as a gift from God [4]. Nowadays, there are good evidences that its constituents (polyphenols in particular) possess antioxidant capacity according to in vitro studies. Additionally, olive oil consumption reduces the risk of developing metabolic syndrome by reducing hyperglycemia, hypertension and lipid peroxidation [5]. Furthermore, in a study in which a diet enriched with extra virgin olive oil was administered to obese women, their fat loss was 80% higher than in obese women that did not assume the oil and they presented reduced diastolic pressure. Maybe, in the future it could be proposed as a treatment for obesity. Based on these premises, the Scientific School titled "Take care of children. The Mediterranean diet in the first 1,000 days of life. A healthy life" was organized by some of the authors (V.F., E.B., G.T., A.N., P.B.). It took place from 20 to 23 September 2017. It is a 4 days workshop in which the leading Italian experts in nutrition and food lectured about all aspects of these topics to a selected audience. The importance of a proper nutrition and the health benefits of the Mediterranean Diet from pregnancy and breastfeeding to adult life were discussed.

The most recent discoveries in nutritional field were illustrated, especially those coming from the applications of the "omics" sciences, nutrimetabolomics, microbiomics and sportomics in particular [6]. Indeed, the interaction between food intake and gut microbiota has an important role as well for the health outcomes of a patient. This was an excellent opportunity to share knowledge and to grow as a scientific community in order to develop better and better strategies to take care of the patients using nutrition both as a preventive and therapeutic tool in all phases of life.

REFERENCES

[1] Koutelidakis AE, Alexatou O, Kousaiti S, Gkretsi E, Vasios G, Sampani A, Tolia M, Kiortsis DN, Giaginis C. Higher adherence to Mediterranean diet prior to pregnancy is associated with decreased risk for deviation from the maternal recommended gestational weight gain. Int J Food Sci Nutr. 2017;14:1-9.

[2] Martinez-Gonzalez MA, Martin-Calvo N. Mediterranean diet and life expectancy; beyond olive oil, fruits, and vegetables. Curr Opin Clin Nutr Metab Care. 2016;19(6):401-7.

[3] Leventakou V, Roumeliotaki T, Sarri K, Koutra K, Kampouri M, Kyriklaki A, Vassilaki M, Kogevinaz M, Chatzi L. Dietary patterns in early childhood and child cognitive and psychomotor development: The Rhea mother-child cohort study in Crete. Br J Nutr. 2016;115(8),1431-7.

[4] Uylaşer V, Yildiz G. The historical development and nutritional importance of olive and olive oil constituted an important part of the Mediterranean diet. Crit Rev Food Sci Nutr. 2014;54(8):1092-101.

[5] Trapani G, Vagliano L, Giribaldi M, Cavallarin L, Coscia A. Olive oil: maternal and pediatric health. J Pediatr Neonat Individual Med. 2017;6(1):e060133.

[6] Dessì A, Cesare Marincola F, Masili A, Gazzolo D, Fanos V. Clinical metabolomics and nutrition: the new frontier in neonatology and pediatrics. Biomed Res Int. 2014;2014:981219.

ABS 53

THE EFFICACY OF LOW LEVEL LASER THERAPY FOR THE PREVENTION AND TREATMENT OF CHEMOTHERAPY-CAUSED MUCOSITIS IN PEDIATRIC PATIENTS

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INTRODUCTION

Oral mucositis is one of the most common and debilitating condition in pediatric patients undergoing antineoplastic and radiotherapy treatments. Its frequency is highly variable: it occurs in 30-70% of patients who underwent chemotherapy and up to 90% of patients who had an hematopoietic stem cells transplantation (HSCT). The typical clinical inflammatory outcomes of mucositis exhibits pain, multiple painful ulcers that limit the patient's mastication and swallowing, and determines the increase of opioid prescriptions and use of parenteral nutrition due to poor nutritional status. The aim of this study is to evaluate the preventive and therapeutic efficacy of low-level laser therapy (LLLT).

MATERIALS AND METHODS

The patients included in the study were aged between 2 and 17 years old and they showed a grade 2 or greater mucositis during their antineoplastic treatment. Chemotherapy had been performed three weeks before. Treatment was administered using K-lase cube 3, whit combined wave lengths of 660-970 nm, average power of 3.2 W (6.4 W pulsed at 50%), time duration of irradiation 3'51", frequency from 1 to 20,000 Hz, spot size 1 cm 2. During phase 1 the therapeutic effect of laser was tested on 10 patients. A daily session was performed for four consecutive days; a standardized physical examination of oral cavity (subdivided in 9 sectors) had been performed at day 0 (T0), day +3 (T1), day +7 (T2). During phase 2 the preventive efficacy was tested on 9 patients by performing a session before, during and after the chemotherapy, a week apart from each other. For each site we recorded a score from 0 to 4 according to the extension of ulcers measured in centimeters, and from 0 to 3 according to the severity of erythema. Mucositis grade was assessed with the visual analogue scale (VAS).

RESULTS

Phase 1 results: the average grade of mucositis showed a regression from 3.2 on T0, to 1.95 on T1, to 0.5 on T2 with a reduction of respectively 40.62% and 84.37%. The average grade 3.21 of ulceration showed a regression of 66.76% at T1 (mean score 1.16) and of 84.45% at T2 (mean score 0.8). The average grade of erythema 2.5 showed a regression of 19.2% (2.05) and of 68.47% at T2. The average pain grade according to VAS scale was 6.5 at day 0, 2 at T1 and 1 at T2 with a reduction respectively of 69.23% and 84.62%. Phase 2 results: the incidence of mucositis was 22% (counter 30-90% found in scientific literature) a mean grade of 1.65 and an average duration of 3 days found in 3 patients. CONCLUSIONS

This study shows the efficacy of LLLT for the prevention and treatment of chemotherapy-caused mucositis. We found a significant reduction of mucositis grade, ulcers grade and pain (over 80%) and a decrease of approximately 70% in the erythema grade.

ABS 54

ENDOMETRIAL STEM CELLS: ANGELS OR DEVILS?

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INTRODUCTION

Pregnancy and woman reproductive physiological functions are allowed and finely regulated by the immense and surprising endometrial ability of regeneration and repair. This tissue is able to grow and renew itself, thanks to the great potential of Endometrial Stem Cells (ESCs) [1].

Although these great benefits, recent evidence suggest that a deregulated ESCs activity contributes to the pathogenesis of endometriosis and could play a role in the development of human leiomyomas and malignant tumors [2].

WHAT IS NEW IN ENDOMETRIAL REGENERA-TION

Functional endometrial layer, under the stimulation of progesterone and estradiol, is monthly regenerated and shed during woman's reproductive life. ESC can be classified in: Epithelial SC, Mesenchymal SC and Endothelial SC and have been found in both basal and functional layers. Those who reside in basal layer migrate into functional layer to replace glands, stroma or vessels. This pool is constituted by endometrial residing multipotent SC, in addition to other SC derived from different tissues [1]. The deregulation of SC interaction with their niches seems to have an important role in the determination of several pathologic conditions [2]. For example, ESC can generate endometriotic implants and could be involved in all the mechanism proposed to explain endometriosis pathogenesis, such as retrograde menstruation or coelomic metaplasia. Moreover, both monoclonal cells and polyclonal cells have been detected in endometriotic lesions [3]. Recently, it has been advanced the hypothesis that uterine leiomyomas, the most frequent benign pelvic tumor in fertile women, could originate from uterine SCs. Leyomioma's progenitors may be monoclonal transformed myometrial smooth muscle cells deriving from mutations occurred in leiomyoma-derived side population (LMSP), although several studies are still needed to confirm this theory. At least, the relation between cancer stem cells (CSCs) and normal SCs has been widely studied; CSCs could represent SCs that lose the dependence from their niches and regulation factors or represent differentiated cells that, through several mutations and transformations, gained new functions, "stemness" characteristics and probably immortalization [3]. Cancer Stem Cells (CSCs) can renew themselves and give origin to a differentiated aberrant progeny, not responding to homeostatic control and probably able to induce metastatization in murine models. CONCLUSIONS

ESCs, with their high proliferation rate and low immunogenic power, show an enormous therapeutic potential and result very promising for clinical experiments and applications. For this reason, further studies are needed to fully understand ESC's features [3]. Unfortunately, the enormous regenerative potential of endometrium is not exempt from an ugly side, coexisting with the good one as in a two-faced Janus and wasting the regenerative mechanism that allows reproduction and the miracle of life itself.

REFERENCES

[1] Gargett CE, Schwab KE, Deane JA. Endometrial stem/progenitor cells: the first 10 years. Hum Reprod. 2016;2:137-63.

[2] Kessler M, Zietlow R, Meyer TF. Adult stem cells niches-Stem cells in the female reproductive system. In: Wislet-Gendebien S (Ed). Adult Stem Cell Niches. Available at https://www.intechopen.com/books/adult-stem-cell-niches/adult-stem-cells-in-the-female-reproductive-system, date of publication: 2014, last access: September 2017.

[3] Xu Y, Zhu H, Zhao D, Tan J. Endometrial stem cells: clinical application and pathological roles. J Clin Exp Med. 2015;8(12):22039-44.

ABS 55

IMBALANCE OF PRO- AND ANTI-INFLAMMA-TORY CYTOKINES IN COMMUNITY ACQUIRED PNEUMONIA OF CHILDREN WITH DIFFERENT LEVELS OF PHYSICAL DEVELOPMENT

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INTRODUCTION

Physical development (PD) of children and adolescents reflects the pattern of body growth processes, its morphological and functional state for each age period. Despite the large number of studies in this field, the problem remains unclear concerning the peculiarities of the immune reaction to acute inflammatory process of children with different levels of PD. Taking into account the high incidence rate for community-acquired pneumonia (CAP) among children of different age groups and the lack of clear data on the peculiarities of its course among patients with different PD levels, the relevance of the study is beyond doubt.

MATERIALS AND METHODS

Having examined 171 children (of 3-14 years old) CAP diagnosis was made due to the Order of the Ministry of Health of Ukraine "On Approval of Protocols for Provision of Medical Care to Children in the field of "Children's Pulmonology". The assessment of PD level of children was made under body mass index (BMI). Taking into account PD level patients were divided into groups: 1st group (n = 50), with overweight (OWT); 2^{nd} group (n = 50), with body weight deficiency (BWD); 3^{rd} group (n = 51), with average physical development indices (APDI). The control group consisted of 20 apparently healthy children of correspondent age. The cytokine profile (IL-1 β , IL-4 and TNF- α) was evaluated using a solidphase immunoenzymometric assay in serum on 3-4 day and 7-8 day from onset of the disease. The research was carried out according to international bioethical standards.

RESULTS

Analysis of results of cytokine studies among patients with CAP showed that the level of proinflammatory interleukins (IL-1 β and TNF- α) was significantly higher among children with OWT and BWD than among children of control group. The analysis of IL-1 β levels among children with different PD levels, found that level of IL-1 β in the group with OWT and BWT was significantly higher in 1.4 times $(50.07 \pm 6.04 \text{ pg/ml} \text{ and } 48.47 \text{ })$ \pm 5.73 pg/ml) (p < 0.05) compared to patients with APhDI (34.9 \pm 4.23 pg/ml). There were no significant differences between patients with OWT and BWT (p > 0.05). Rate of increase of the content of proinflammatory cytokines among girls with OWT ($[34.8 \pm 4.21]\%$; p < 0.05) significantly exceeds (in 3.3 times) than among boys ([11.1 \pm 1.2]%; p < 0.05). It makes it possible to establish the differences in metabolism of IL-1 β in response to the inflammatory process in the lungs and the dependence on PD level. The level of TNF- α was found to be $56.1 \pm 4.7\%$ among patients with OWT, $47.5 \pm 3.2\%$ among patients with BWT and $39.1 \pm 2.7\%$ among children with APDI (p > 0.05). The maximum increase of IL-4 level among children with this elevated level, was found among children with BWD (47.5 \pm 3.2%), while patients with OWT had significantly less (in 1.7 times; p < 0.05, among $26.8 \pm 2.2\%$ of patients), however, the elevated level of cytokine in the group of patients with APDI (21.7 \pm 1.9%), in 2.2 times less (p < (0.01) than among patients with BWD and in (1.2)times less than in the group with OWT.

CONCLUSIONS

Assessment of characteristics of the immune response to acute inflammation among pediatric patients with CAP shows a clear imbalance between pro- and anti-inflammatory cytokines and has a clear dependence on the level of PD of the child. Thus, there is an increase in productivity of infectious cytokines (IL-1 β , TNF- α) in the groups of children with OWT and BWD, as well as a significant increase of anti-inflammatory cytokine rates (IL-4), which is appropriate to patients with BWD.

ABS 56

THINK FIRST ABOUT WHAT'S COMMON

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INTRODUCTION

Cardiac myxoma is a benign neoplasm unusual in neonates. It's an endocardial mass that occupies the cardiac chamber. Most of myxomas are found in the left atrium representing a risk factor for thromboembolism. Right atrial thrombosis (RAT) is a rare event in neonates. There's a strong association between central venous catheters and venous thromboembolic events. Incidence is still not clear (1.8-8.3%) [1]. Risk factors for RAT include stasis, vascular injury and hypercoagulable state (congenital heart disease, parenteral nutrition, prematurity, protein losing state and malignancies). There are no guidelines on the management of RAT in the neonatal and pediatric population. The clinic varies from asymptomatic cases to respiratory distress and arrhythmia. Some authors classify patients in high, moderate or low risk, considering some features of thrombi (dimension > 2 cm, pedunculated, mobile or snake-shaped). Neonates and children with RAT who are asymptomatic, hemodynamically stable and at low risk could be followed with radiological monitoring with or without anticoagulation. Some authors prefer to use unfractionated heparin, which is more reliable and predictable in children. For symptomatic or high-risk patients, management decisions should be considered on a case-by-case basis. Surgical thrombectomy or thrombolytic therapy have significant risk of surgical mortality and bleeding.

CASE REPORT

P. was referred to the Department of Neonatology at birth because preterm (34 w), male infant of diabetic mother treated with insulin. No family history of cardiac and hematologic disease.

Physical examination: heart rate 140/bpm, respiratory rate 45 breaths/min, peripheral oxygen saturation 98%. No dyspnea. Laboratory tests: leukocytosis, moderated elevation of C-reactive protein and procalcitonin. He started ceftazidime e.v. Electrocardiography was normal. Because of persistent hypoglycemia umbilical vein catheter (UVC) had been placed. Chest X-ray shows UVC in the vein port, so has been removed by 2.5 cm. On the third day of life he developed jaundice requiring phototherapy. Echocardiography examination showed an echo contrast, floating, oval shaped mass, sized 5 x 6 mm in the right atrium. The differential diagnosis was between thrombosis and atrial myxoma. For that reason we moved the baby in a third level center where he was evaluated for a potential cardiac surgery. After 6 days of followup they resigned the baby and decided to continue cardiological monitoring outpatient. The next cardiological control echocardiography reveal spontaneous resolution of the atrial mass.

CONCLUSIONS

Considering the spontaneous resolution and the anamnestic data of UVC, prematurity and parental nutrition, the most likely diagnosis was RAT. UVC are associated with complications such as infections, thrombosis and trauma; although intracardiac thrombosis is rare in children, the most common cause of atrial mass is thrombosis associated with UVC. Take home message: remember to place special attention in the placement of UVC! REFERENCES

 Yang JY, Williams S, Brandão LR, Chan AK. Neonatal and childhood right atrial thrombosis: recognition and a risk-stratified treatment approach. Blood Coagul Fibrinolysis. 2010;21(4):301-7.

ABS 57

A COMPLEX CASE OF ANOMALOUS LEFT CORONARY ARTERY ORIGIN FROM THE PULMONARY ARTERY (ALCAPA)

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INTRODUCTION

Anomalous left coronary artery origin from the pulmonary artery (ALCAPA) is a not frequent congenital heart disease (CHD); the age of onset and the clinical presentation vary depending on the degree of myocardial perfusion ensured by collateral circles. It can be surgically treated, with the anastomosis of the ectopic artery to the aorta [1]. CASE REPORT

G. a male neonate, was born at 35 wks of GA, weight 2,270 g, Apgar 9'-10'. His father and paternal aunt are affected by Alagille Syndrome (ALGS). Hospitalized for prematurity, in the following days he developed jaundice. At 5 days, G. presented high values of transaminases (AST 124 U/L) and cholestasis indices (γ GT 1,183 U/L). Cardiac ultrasound evidenced an aneurysm of the interatrial septum, without other apparent anomalies. The ECG showed non-specific repolarization abnormalities. At 11 days, high γ GT persisted (987 U/L) and the parents decided for the discharge in spite of the sanitary advice, without concluding the diagnostic iter for the suspected ALGS.

At 6 wks, during a programmed visit, clinical condition was very poor, with signs of heart failure, dyspnea, tachypnea, pallor, feeding intolerance, poor weight gain (3 kg). Echocardiogram evidenced globular left ventricle with severe hypokinesia (FE 34%); considering the marked hyperechogenicity of papillary muscles and ECG evidence of deep Q waves in DI and aVL, ALCAPA has been suspected and confirmed by cardiac catheterism. A normal origin of right coronary artery has been described, giving origin to collateral circles to supply a backward perfused left coronary, originating from the posterior sinus of pulmonary artery trunk. At 8 wks G. presented FE 25% and cardiac surgery has been performed. The postoperative course required Extra Corporeal Membrane Oxygenation (ECMO) for 10 days and therapy with milrinone for severe biventricular hypokinesia; it has also been complicated by a serrated stenosis at the origin of the right pulmonary branch and by the occurrence of chylothorax. At 13 wks G. was discharged, continuing therapy with furosemide and captopril. Echocardiography detected FE 60%, hyperechogenicity of papillary muscles and of medioseptal and posterior areas. Moreover, slight aortic shrinkage (gradient 30 mmHg) and a narrowed origin of the right pulmonary branch (gradient 50 mmHg and diastolic run-off) were detected. At 14 wks, therapy with furosemide was reduced.

At 18 wks, G. presents good weight gain (4.280 kg), polypnea, systolic murmur 3/6 and is treated with ursodeoxycholic acid for the persistence of high γ GT values. Genetic assessments for ALGS is still ongoing.

CONCLUSIONS

ALGS is a complex autosomal dominant disorder, characterized by almost 3 elements among cholestasis, CHD (mainly regarding pulmonary branches), posterior embryotoxon in the eye, facial features and butterfly vertebrae [2]. In our opinion, this could be a case of ALGS associated to ALCAPA. ALCAPA can be surgically resolved, but complications can occur.

REFERENCES

[1] Guzeltas A, Ozturk E, Cansaran Tanidir I, Kasar T, Haydin S. Evaluation of anomalous coronary arteries from the pulmonary artery. Braz J Cardiovasc Surg. 2017;32:29-37.

[2] Saleh M, Kamath BM, Chitayat D. Alagille Syndrome: clinical perspectives. Appl Clin Genet. 2016;9:75-82.

ABS 58

SLEEP-DISORDERED BREATHING IN CHIL-DREN WITH RECURRENT WHEEZING/ASTHMA: A SINGLE CENTRE STUDY

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BACKGROUND

The relationship between asthma and sleep-disordered breathing appears to be bidirectional due to common risk factors that promote airway inflammation. Therefore, obstructive sleep-disordered breathing and recurrent wheezing/asthma are conditions that affect mainly the upper and lower respiratory system, respectively. The aim of the present study was to investigate the respiratory polygraph parameters that characterize recurrent wheezing/asthma condition. METHODS

A retrospective study concerned all children older than 2 years who undergone, between January 2014 and November 2016, an in-laboratory overnight polygraph study. The children were matched between those who have or not recurrent wheezing/asthma disease.

RESULTS

Clinical records of 137 children were examined. Among them, 8 patients were excluded because neurological and genetic conditions. Recurrent wheezing/asthma children (n = 28) were younger (p = 0.002) and leaner (p = 0.013) than non-affected children (n = 98). Moreover, wheezing/asthma children had a comparable Obstructive Apnea-Hypopnea Index (p = 0.733) and Oxygen Desaturation Index (p = 0.535) than unaffected children. Logistic regression analysis in which asthma was a dependent variable and demographic (age, sex, BMI Z-score) and polygraph parameters (Obstructive Apnea-Hypopnea index, Central Apnea Index, SpO₂ and snoring) as covariates showed that the probability to have a high Central Apnea Index (Wald 6.845; p = 0.009) was increased in the asthma group (Exp[B] = 2.212). Fig. 1 shows the plotted distribution with regression lines between the Central Apnea Index and age of the 126 children categorized as recurrent wheezing/asthma (red circle, n = 28) and non-asthma (black circle, n = 98) condition.

CONCLUSION

The main findings of the present study were that children with recurrent wheezing/asthma showed an increased Central Apnea Index than unaffected children. Recurrent wheezing/asthma can affect Central Apnea Index in these children. The selection of a sample of affected and unaffected children, balanced for age and Sleep Disordered Breathing severity, may confirm this observation. This data may suggest a dysfunction of the breathing control in the central nervous system during sleep. Systemic or central inflammation may take a role.

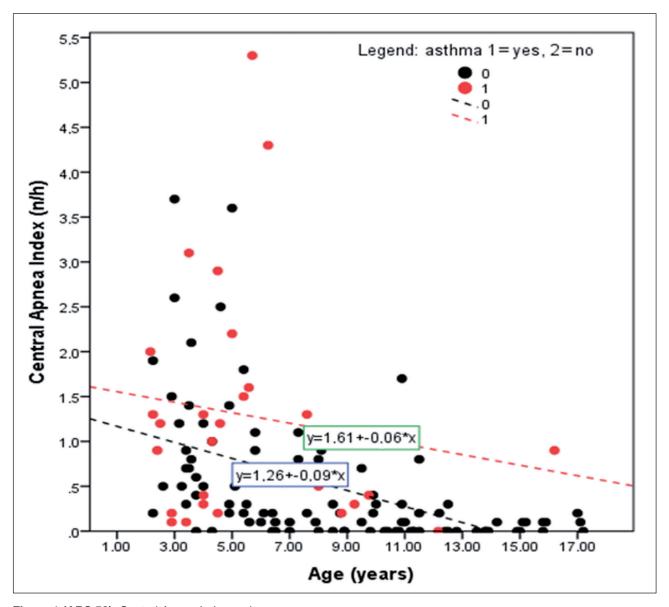


Figure 1 (ABS 58). Central Apnea Index and age.

ABS 59

UNUSUAL GLOMERULAR SHAPE IN TRISOMY 21

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INTRODUCTION

Patients affected by Down syndrome are characterized by a high incidence of renal disease in adulthood. The cause of this predisposition has not been clarified yet. In previous studies from our group, it has been hypothesized that susceptibility to nephropathy in subjects with trisomy 21 might be related to major changes in renal fetal development, resulting in structural glomerular changes. The aim of this study was to describe glomerular morphology in a human fetus with Down syndrome.

PATIENT AND METHODS

A 15 week-old fetus underwent therapeutic abortion following the diagnosis of trisomy 21. At autopsy, edema of the neck, low-set ears, prognathism and brachydactyly were observed. Samples from both kidneys were formalin-fixed and paraffin embedded. Tissue sections were stained with H&E.

RESULTS

At histology, fetal kidneys were characterized by the presence of active nephrogenesis, well evidenced by the persistence of metanephric mesenchymal cells in the subcapsular zone (the so called "blue strip"). Major changes were found regarding the glomerular shape in both kidneys. Glomeruli appeared voluminous, with a narrow capsular space. Podocyte precursors were characterized by large nuclei, with dark clumped chromatin. The glomerular tufts frequently showed multiple adherences to the epithelial capsular cells. Moreover, glomerular vascular tufts were characterized by abnormal shapes, ranging from bi-lobated to multilobated appearance (Fig. 1). We also frequently observed glomeruli adherent to each-other, with the Bowman capsule in common.

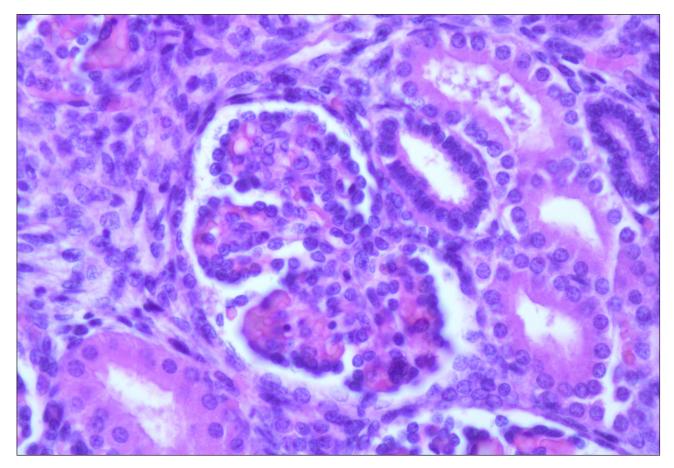


Figure 1 (ABS 59). Glomerular vascular tufts were characterized by abnormal shapes, ranging from bi-lobated to multilobated appearance.

CONCLUSIONS

Our case confirms previous hypothesis regarding the possibility that susceptibility of patients with Down syndrome to develop renal disease later in life, might be related to the presence of kidney structural changes originating during the intrauterine life. These findings, when confirmed by a large series of patients carrying trisomy 21, clearly indicate fetal programming as a major etiological factor in the pathogenesis of renal disease presenting in adult patients carrying trisomy 21.

ABS 60

A STEP TOWARDS A SAFER DISCHARGE: THE EVALUATION OF CARDIORESPIRATORY STABILITY IN PRETERM NEWBORN

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INTRODUCTION

The discharge of a preterm newborn from a Neonatal Intensive Care Unit (NICU) is depending on stable weight gaining, safe oral feeding and cardiorespiratory stability. Despite the complexity of this evaluation, no specific protocol does exist and, in most NICUs, the decision to discharge relies on clinician experience and judgment. Aim of the study is to propose a protocol to assess cardiorespiratory stability at discharge and to evaluate its impact on NICU.

MATERIALS AND METHODS

A 3-step protocol to evaluate cardiorespiratory (CR) stability was made up of clinical evaluation (CE), clinical monitoring (CM) and instrumental monitoring (IM) and was applied in an observational prospective study on preterm newborns (gestational age < 32 weeks) admitted to our NICU from May to December 2016. All newborns near to be discharged (increasing weight > 1,600 g, exclusive oral feeding and spontaneous breathing) were consecutively enrolled and underwent CE. All newborns with positive CE then underwent 24 h CM. Newborns with silent CM but risk factors for CR instability (gestational age ≤ 28 weeks, age at evaluation \leq 34 weeks, mechanical ventilation \geq 24 h) underwent IM with GETEMED Vitaguard 3100®. The results of CE, CM and IM were compared with a control population of newborns discharged during a period of 12 months prior to the study.

RESULTS

Fourty newborns with a median gestational age of 28.1 (26.4-29.9) weeks were enrolled and underwent CE and CM. The CM identified 3 (7.5%) unstable babies. Twenty-eight (70%) performed IM and 556 events were detected, 41 (7.37%) of which were detected by CM as well. The number of events/patient detected by IM (9.0 [3.0-20.6]) was higher (p < 0.05) than CM (0.0 [0-1.3]) and was related to CT90 values. Seven (25.0%) patients with CT90 > 3% were identified. Compared with the control group, enrolled newborns showed a lower time of hospitalization (42.5 [30.5-67.6] vs. 49.0 [32.0-66.0] days) and fewer admissions to emergency department up to 3 months after discharge.

CONCLUSIONS

Applying a protocol to assess CR stability contributes to a safer discharge and allows to individualize the time for discharge without lengthening the time of hospitalization.

ABS 61

NASAL OBSTRUCTION AND SLEEP DISOR-DERED BREATHING DURING CLINICAL PRAC-TICE IN CHILDREN

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BACKGROUND

Nasal congestion typically results in a switch to oral breathing predisposing to sleep disordered breathing (SDB). Testing for nasal occlusion severity takes part of the clinical evaluation of children with SDB. Unfortunately, it is not clearly defined if the nasal obstruction should be considered a risk factor or a mechanism for obstructive sleep apnea syndrome. (OSAS). The aim of the present study was to evaluate the relationship between nasal obstruction and sleep disordered breathing in children during clinical practice.

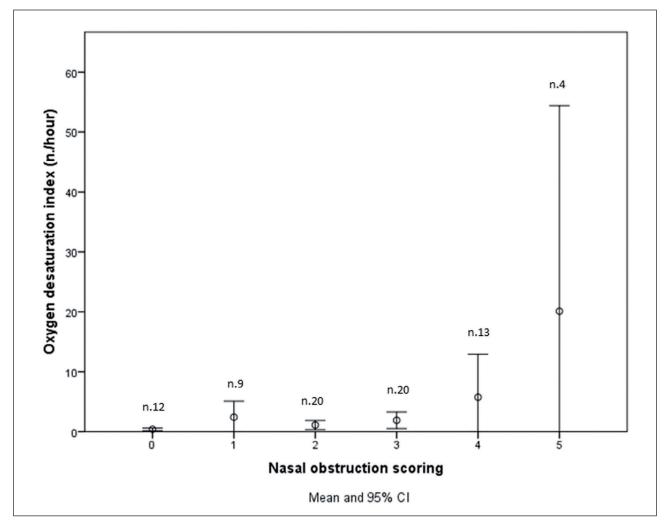


Figure 1 (LECT 61). Oxygen desaturation index and nasal obstruction scoring. Data are shown as mean and 95% Cl.

METHODS

The present study concerned the enrollment of all compliant ambulatory children, 3 years and older, who undergone between January 2016 and May 2017 an in-laboratory overnight polygraph study. Nostrils obstruction test allowed to be scored the severity between 0 (none) to 6 (completely obstructed) [1]. Children with neurological and genetic conditions were excluded.

RESULTS

78 children (58% males; aged 6.0 \pm 2.7 years) were enrolled. Linear regression analysis in which Oxygen Desaturation Index (n./hour) or Apnea-Hypopnea Index (n./hour) were dependent variables and age, BMI Z-score, nasal obstruction score, and allergy as covariate showed that both correlated with nasal obstruction score (p = 0.001). **Fig. 1** shows the relationship between nasal obstruction score results and oxygen desaturation index (ODI). Data are shown as mean and 95% CI.

CONCLUSION

One component of the pathophysiology of OSAS that is among the least understood and studied is the role of the nose. Daytime nasal obstruction was reported as an independent risk factor for OSAS. The patency of the nose can influence the patency of the pharynx. Finally, the nasal obstruction test may be a useful, time sparring ambulatory assessment that can predict SDB severity in children.

REFERENCE

[1] Liistro G, Rombaux P, Belge C, Dury M, Aubert G, Rodenstein DO. High Mallampati score and nasal obstruction are associated risk factors for obstructive sleep apnoea. Eur Respir J. 2003;21(2):248-52.

ABS 62

IS THE BODY POSITION REALLY HELPFUL FOR THE STABILIZATION OF PRETERM BABIES WITH PATENT OF DUCTUS ARTERIOSUS IN NURSING CARE? A LITERATURE REVIEW

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INTRODUCTION

Patent ductus arteriosus (PDA) in preterm infants is one of the most important factors influencing survival. The care of these patients is complex and is a challenge for the pediatric nurses [1]. It is known that the full maturation of the central nervous system is directly related to the organization of awake and mostly sleep state. However, during Neonatal intensive care unit (NICU) hospitalization, preterm infants are routinely exposed to many invasive procedures causing frequently awakening, pain and stress. In infants affected by PDA cardiorespiratory ad behavioural stability help the closure of ductus arteriosus and support to maintain it closed [2]. Can pediatric nurse effectively promote this situation through Newborn Individualized Developmental Care Assessment Programme (NIDCAP) method and postural care?

MATERIALS AND METHODS

Literature research was undertaken on PubMed. This review considered all type of article in English language. No temporal limits were applied. The key words: patent ductus arteriosus, preterm baby, postural care, stress, sleep, NIDCAP, physiological stability. RESULTS

We found no studies evaluating, in PDA, the hemodynamics variables in changing body position, furthermore no findings about the correlation of PDA with postural care and sleeping states so we considered the literature regarding preterm babies and the effects of body position on sleep and stress responses. The research showed a Cochrane and two systematic reviews that support the prone position for improvement of physiological stability and sleep promotion; a literature review stating that midline position and flexion reduce stress and promote sleep state. An RCT affirmed that flexed posture has more positive effects on the daily sleep quantity and decreases wakefulness time. Other studies agree on the effectiveness of prone and flexed position in neonatal stress reduction and in a longer sleep.

CONCLUSION

Our review showed that postural care is considered part of the individualized care for the neonate development and has a central role in increasing the amount of sleep and reducing stress. The

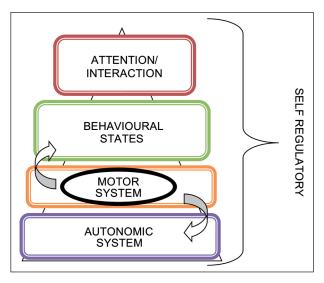


Figure 1 (ABS 62). The synactive theory by H. Als suggests that the development of the newborn proceeds through the constant and continuous interaction of the illustrated subsystems. The motor system represents a powerful stabilizer of the others.

literature point out that postural care and NIDCAP meet and integrate perfectly with every phase of infant care and every clinical situation [3]. Why? Because the preterm baby has special biological and neurobehavioral features so that this concept is essential (Fig. 1). Furthermore, we stress the necessity for nurses of a in-depth education and adequate knowledge in pathophysiology of PDA, in anatomic, physiological and neurobehavioral peculiarities of preterms, in order to assess, plan, and implement patient-centered care based to the gestational age too. However, standing to our review, it is not well known if the body position can influence hemodynamic in babies with PDA. Future researches may determine if a correlation between postures and hemodynamic changes exists in this specific population.

REFERENCES

[1] Samuel BP, Stuive AJ, Eding DM. Nursing Assessment, Education, and Care of Extremely Premature Neonates with Patent Ductus Arteriosus. Curr Pediatr Rev. 2016;12(2):106-9.

[2] Rivas-Fernandez M, Roqué I Figuls M, Diez-Izquierdo A, Escribano J, Balaguer A. Infant position in neonates receiving mechanical ventilation. Cochrane Database Syst Rev. 2016;11:CD003668.

[3] Jarus T, Bart O, Rabinovich G, Sadeh A, Bloch L, Dolfin T, Litmanovitz I. Effects of prone and supine positions on sleep state and stress responses in preterm infants. Infant Behav Dev. 2011;34(2):257-63.

ABS 63

P57 EXPRESSION IN MALE AND FEMALE PLACENTAS

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INTRODUCTION

In previous studies, differences between male and female human placentas have been reported regarding the growth and gene expression and gene expression [1, 2]. The aim of this study was to verify at immunohistochemical level the presence of gender-related differences regarding the development of cytotrophoblast cells. To this end in 4 male placentas and 4 female placentas (from fetuses and newborns) we investigated the reactivity for p57.

MATERIALS AND METHODS

P57 is a protein involved in a cell cycle inhibition, expressed in the cytotrophoblast cells in normal human placenta [3]. We analyzed archival paraffinembedded samples at different gestational age between 12 and 40 weeks by immunostaining with a Dako Autostainer. We used a prediluted p57, anti-p57 Kip2 (Kip10) mouse monoclonal primary antibody (Cell Marque, key code CMC46170001). RESULTS

Our data shows that p57 immunostaining was fully in the syncytiotrophoblasts absent whereas in cytotrophoblasts p57-reactivity involved both peripheral perivillous cytotrophoblasts and central intravillous cytotrophoblasts (**Fig. 1**). In these two areas, we found the greatest differences between the male and female placentas, in particular at the intermediate gestational ages (21 and 30 weeks).



Figure 1 (ABS 63). Immunohistochemistry of p57 in human placenta villi: syncytiotrophoblasts are p57-negative, peripheral perivillous cytotrophoblasts and central intravillous cytotrophoblasts are p57-positive.

CONCLUSIONS

Our immunohistochemical data lay stress on the existence of gender-related differences in the human placenta even at cellular level. Further studies are needed in order to clarify if the cellular genderrelated differences are restricted to cytotrophoblasts or if other cells types are involved.

REFERENCES

[1] Mc Keown T, Record RG. The influence of placental size on foetal growth according to sex and order of birth. J Endocrinol. 1953;(10):73-81.

[2] Cvitic S, Longtine MS, Hackl H, Wagner K, Nelson MD, Desoye G, Hiden U. The human placental sexome differs between trophoblast epithelium and villous vessel endothelium. PLoS One. 2013;8(10):e79233.

[3] Castrillon D, Sun D, Weremowicz S, Fisher RA, Crum CP, Genest DR. Discrimination of complete Hydatidiform mole from its mimics by immunohistochemistry of the partenally imprinted gene product p57 KIP2. Am J Surg Pathol. 2001;25(10):1225-30.

ABS 64

IRON SUPPLEMENTATION IN PRETERM NEWBORNS: SULFATE OR BISGLYCINATE CHELATE IRON?

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INTRODUCTION

More than 65% of preterm newborns develop an iron deficiency (ID); they have poor iron stores at birth, as 80% of the iron is acquired by the foetus in the third trimester of pregnancy. The frequent blood sample drawings associated with an inadequate intake of iron and rapid postnatal growth contribute to developing an ID and subsequent iron deficiency anemia (IDA). Strategies to prevent anemia in preterm newborns include drawing fewer blood samples, the use of recombinant human erythropoietin and iron supplementation. Although sulfate iron is the most commonly used pharmaceutical formulation for iron supplementation, there are few studies comparing different iron salts in newborns. The objective of this study was to compare the efficacy of bisglycinate chelate iron to sulfate iron in preterm newborns receiving erythropoietin.

MATERIALS AND METHODS

Three-hundred newborns of gestational age ≤ 32 weeks were enrolled: 225 were supplemented

with sulfate iron and 75 were supplemented with bisglycinate chelate iron. As in usual local clinical practice, iron supplementation was started when the Reticulocyte Hemoglobin content (CHr) values reached 30-32 pg. Prophylaxis was carried out according to the indications of the pharmaceutical companies manufacturing the two products: sulfate iron 3 gtt/kg/day (equivalent to 3 mg/kg/day of elemental iron), bisglycinate chelate iron 3 gtt/kg/ day (equivalent to 0.75 mg/kg/day of elemental iron). When the CHr values were less than 30 pg, the iron dose was increased by 1 gtt/kg/day. The effect on erythropoiesis was assessed with a general linear model that estimates the response variables (values for Hemoglobin, Hematocrit, absolute values and percentage Reticulocytes, Reticulocyte Hemoglobin content) based on treatment, time, birth weight, and gestational age.

RESULTS

Supplementation with bisglycinate chelate iron at a dose of 0.75 mg/kg/day demonstrated a therapeutic efficacy comparable to sulfate iron at a dose of 3 mg/kg/day in both populations of preterm newborns. The two cohorts have similar erythropoietic response, without significant differences.

CONCLUSIONS

The higher bioavailability of bisglycinate chelate iron resulted in a lower load of elemental iron, a quarter of the dose, and obtained equivalent therapeutic efficacy compared to sulfate iron. Bisglycinate chelate iron appears to be a therapeutic alternative to sulfate iron in the prevention and treatment of preterm newborn anemia.

ABS 65

LIVER BIOPSY INTERPRETATION IN WILSON DISEASE IN CHILDHOOD: A REPORT OF 12 CASES

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INTRODUCTION

The histological picture of liver biopsy in children affected by Wilson Disease (WD) may be extremely variable, showing a wide range of patterns: from no detectable microscopic pathology to acute liver failure and chronic hepatitis. In the majority of children, liver biopsy shows steatosis, associated with a variable degree of portal and/or lobular inflammation, bridging fibrosis and, eventually, cirrhosis. Hepatocyte ballooning, Mallory-Denk bodies, liver cell apoptosis, stellate cell hyperplasia and glycogenated nuclei have been reported as well [1-3]. None of these features is specific for the diagnosis of WD, thus the histological picture should be interpreted in the medical context and the clinical setting. The aim of this work was to analyze the histological pattern of liver disease occurring in Sardinian children.

PATIENTS AND METHODS

12 liver biopsies from pediatric patients, from 3 to 18 year-old, 5 female and 7 male, were analyzed in this study. The following elementary histological lesions were evaluated: steatosis, hepatocyte ballooning (Fig. 1), fibrosis, inflammation, glycogenated nuclei, Mallory-Denk bodies, and nuclear pleomorphism. Steatosis and ballooning were scored in 4 categories (< 5%, 6-33%, 34-66% and > 67%) and zonation; steatosis was subdivided into macrovesicular and microvesicular. Sinusoidal, peri-terminal, portal, bridging fibrosis, regeneration, and cirrhosis were also analyzed. Grading of the inflammatory component was based on the number of foci of inflammatory cells (absent, $\leq 2 = \text{mild}$, 3-4 = moderate, > 4 = severe) and on their location (acinar and/or portal). Glycogenated nuclei, Mallory-Denk bodies, nuclear pleomorphism and additional features were also assessed.

RESULTS

Steatosis and hepatocyte ballooning were the most representative lesions, being detected in all but one liver biopsy (see Fig. 1). In 10 out of 12 cases, higher levels of steatosis were paralleled by lower levels of ballooning, and vice versa. Steatosis was heterogeneous in the majority of liver biopsies, being either macro- and micro-vesicular. Fibrosis was periterminal in 4 biopsies, bridging in 3, periportal in one, whereas cirrhosis was observed in 3 cases. Mixed inflammatory infiltrated was found both in the portal and inside the acinar zone. Glycogenated nuclei were observed in all liver biopsy, Mallory-Denk bodies in 7, nuclear pleomorphism in 4. Additional features including lipogranulomas, apoptosis, eosinophils and ductular reaction, were also occasionally observed. CONCLUSIONS

Our data confirm that the histopathological diagnosis of Wilson disease in childhood is not specific, showing major differences from one case to the next. In the vast majority of pediatric patients

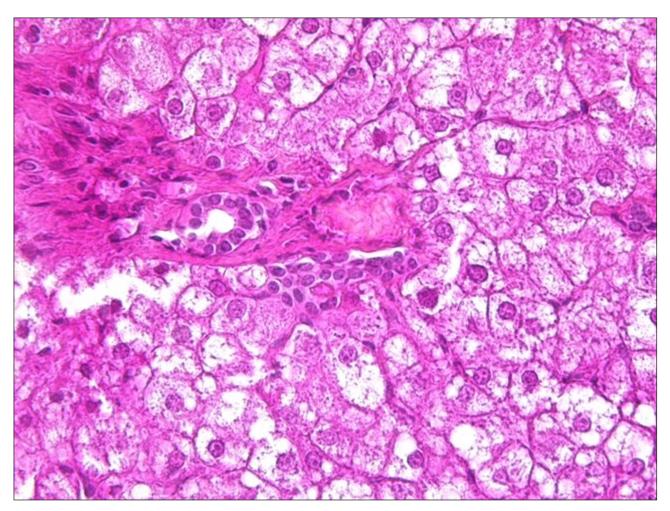


Figure 1 (ABS 65). Hepatocyte ballooning in the zone 1 of the hepatic lobule.

affected by WD, the histological picture appeared as a mimic of NAFLD/NASH. The evolution of liver disease was strikingly different from a child to the next, irrespectively of their age, ranging from simple steatosis to cirrhosis. On the basis of our data, the differential diagnosis of WD and NAFLD/ NASH, when only based on liver biopsy, may be challenging, due to the absence of morphological differential features.

REFERENCES

[1] Stromeyer FW, Ishak KG. Histology of the liver in Wilson's disease: a study of 34 cases. Am J Clin Pathol. 1980;73(1):12-24.

[2] Langner C, Denk H. Wilson disease. Virchows Arch. 2000;445:111-8.

[3] Loudianos G, Gitlin JD. Wilson's disease. Semin Liver Dis. 2000;20(3);353-64.

ABS 66

A NEW APPROACH TO REDUCE CHILDREN'S FEAR

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INTRODUCTION

The fear of medical procedures is broadly widespread among people of all ages. There is a large number of medical procedures which can cause fear, especially in children, such as: pediatric visits, blood draws, vaccines and injections in general, etc. Among the others, the fear of the dental work is one of the most common, given the large number of instruments, which can trigger fear reactions in the pediatric patient. The fear is a big impediment to obtain a good cooperation between the patient and the doctor. Strategies that can reduce the impact of fear at its minimum to improve the cooperation from the pediatric patient are needed. This is often a key ingredient to ensure a successful therapy. In this study we used a mobile application based on augmented reality, able to turn the medical office into a place where playful adventures take place

	Group A		Group B	
Cases	Number of patients	Average age (months)	Number of patients	Average age (months)
Treated at 1 st visit	4	60.0	5	61.2
Treated at 2 nd visit	7	59.1	9	67.6
Treated at 3 rd visit	6	57.3	7	59.0
Treated at 4th visit	5	54.4	6	61.0
Total treated	22	-	27	-
No cooperation	5	57.6	5	57.6

Table 1 (ABS 66). Characteristics of the patients of the groups A and B.

with the pediatric patient as the protagonist. During these adventures, (s)he is involved in mini-activities having as final goal the success of the therapy. The study we are presenting has been focused on the fear of the dental work, and thus has been carried out in a dentist office.

MATERIALS AND METHODS

We tested the "Super Powers" method with 43 pediatric patients (23 females and 20 males) between 3-7 years old. We have included in the study only patients who reported anxiety or fear of the dentist. The number of sessions required to obtain the full cooperation of the patient was adopted as a criterion for evaluating the results obtained. The selected patients were divided according to various criteria:

- presence of painful phenomena occurring at the time of the first visit;
- fear of doctors in general;
- fear originated from previous negative medical and/or dental experiences.

RESULTS

Within the statistical sample, we have identified three patient characteristics:

- A. 21 of 43 patients (48.8%) were presented with pain in the first visit;
- B. 27 of 43 patients (62.8%) were afraid of doctors in general in the first visit;
- C. 32 of 43 patients (74.4%) had been scared by previous experiences.

Results are presented in **Tab. 1**. We have found that, through our study on 30 patients, the success rate was higher than 88% thanks to the "Brave Potions" solution.

CONCLUSION

In conclusion, we have observed numerous advantages: "Super Powers" provides to pedodontist and generic dentist the possibility of approaching the pediatric patient without the need of more complex psychological techniques. The patient, feeling an active part in the treatment process, will form a bond that can deal with any problem that might arise in the future.

ABS 67

SYSTEMIC PROPRANOLOL IN THE TREAT-MENT OF INFANTILE HEMANGIOMAS: EX-PERIENCE OF A MULTIDISCIPLINARY TEAM IN CAGLIARI

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INTRODUCTION

Infantile hemangiomas (IH) is a benign vascular tumor that in some cases can lead to serious complication. Its prevalence is 4-5% reaching 23% in preterm low birth weight infants. Until now oral prednisolone has been the treatment of choice for these problematic lesions and recently beta-blockers have been demonstrated effective and safe, with propranolol becoming first line therapy in complicated IH. Early recognition and treatment of problematic lesions helps to minimize potential complications

MATERIALS AND METHODS

From January 2010 to August 2017 at the dermatology outpatient vascular anomaly clinic, out of 3,249 patient, 269 (8.27%) vascular anomaly have been diagnosed and, between them, 200 (74%) infantile hemangiomas. Forty-nine (24.5%) underwent medical treatment, 21 with systemic propranolol and 28 with topical timolol.

RESULTS

Treatment and outcome of 19 patients with infantile hemangiomas treated with oral propranolol were analysed. Propranolol was given as a first-line treatment for potentially complicated IH. Propranolol was administered since the age of 5 weeks (corrected for preterm babies) at the standard protocol of 2-3 mg/ kg/day divided in two doses. The first administration has been done, after a Doppler sonography and cardiologic screening, in patients hospitalized in a pediatric environment, till the final dose of 3 mg/kg/ day. Subsequently we clinically assessed treatment outcome, by serial photography and monitoring Doppler sonography and adverse events, adjusting the dosage according with the child weight. In all our patients the hemangiomas stopped growing from the beginning of the treatment. 6 patients demonstrated significant clinical reduction in size starting from the first week of treatment and in all the others the reduction was evident at the first month follow-up. Treatment regimen was maintained for 6-12 months (mean 9 months) with lesser duration and minor aesthetic defect when the treatment was started earlier. Complete cure was obtained in 9 children, partial in 5 and 7 are still under treatment No significant adverse events were noted and no patient had to stop therapy. CONCLUSIONS

Identification at-risk IH with close follow-up is crucial in the first weeks of life in order to start treatment as early as possible, to avoid potential complications and to obtain the better response.

REFERENCES

• Goelz R, Poets CF. Incidence and treatment of infantile haemangioma in preterm infants. Arch Dis Child Fetal Neonatal Ed. 2015;100:F85-91.

• Léauté-Labrèze C, Harper JI, Hoeger PH. Infantile haemangioma. Lancet. 2017;39:85-94.

ABS 68

WT1, A MARKER FOR RADIAL GLIAL CELLS IN THE DEVELOPING HUMAN CEREBELLUM AT DIFFERENT GESTATIONAL AGES

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INTRODUCTION

Wilms' tumor 1 (WT1) gene, firstly cloned in 1990 in the childhood kidney cancer Wilms' tumor, is a gene located on chromosome 11p13, which encodes zincfingers protein characterized by multiple alternative isoforms, with important regulatory function in cell growth and development [1]. The WT1 protein regulates the transcription of numerous genes and functions both as activator and as transcriptional coactivator or as repressor of gene expression. WT1 protein was initially shown as a marker of stem cells in the mouse embryo [2]. Evidences showing the expression of WT1 protein during normal kidney development at different stages of nephrogenesis suggest a possible role for WT1 in mesenchymalepithelial transition (MET), as well as in the development and maturation of podocytes [3]. Recent studies have shown that WNT signaling pathway is a major target of WT1. WT1 probably negatively regulates WNT/β-catenin signaling during kidney development. Moreover, WT1 plays a complex role in the regulation of cell proliferation and apoptosis. A strong WT1 cytoplasmic immunostaining in developing skeletal and cardiac muscle cells, in endothelial cells, in the sympathetic system and in the gastro enteric nervous system has been reported in human fetuses. Recently, it has been suggested a potential role of WT1 protein in the development of the human nervous tissues, in particular in the cerebral cortex, during early developmental phases of gestation. The aim of this study was to investigate the immunohistochemical expression of WT1 in the development of human cerebellum in order to better evaluate the human ontogeny.

MATERIALS AND METHODS

To this end, we analyzed the immunoreactivity for WT1 in human fetuses at different gestational ages. Cerebellum samples were obtained from 5 human fetuses, ranging from 15 (**Fig. 1**) to 38 weeks of gestation. The fetuses have been completely sampled and histologically and immunohistochemically studied. Samples were fixed in 10% buffered formalin, paraffin-embedded and immunostained for WT1. RESULTS

Our data show that, at the 18th week, in the surface of the cerebellar cortex, WT1 immunostaining was particularly strong in the cytoplasm of radial glia fibers extending from the ventricular zone towards the pial zone. At the 21st week, WT1 cytoplasmic expression was detected in radial glia fibers extending from the ventricular and subventricular zones, evidencing a decrease in the number of radial glia fibers. At the 34th weeks, a mild WT1 cytoplasmic immunoreactivity was detected in the ventricular and subventricular zones, as well as in the cortical layers. A strong WT1 expression was observed in the epithelium of small blood vessels in the cortical layers. At the 38th week, whereas a mild reactivity for WT1 was detected in cell extensions in the cortical surface, the immunostaining for this antibody was mainly observed in the epithelium of cortical small blood vessels.

CONCLUSIONS

Our data show that, whereas by the 34th week a mild immunoreactivity for WT1 was detected in the

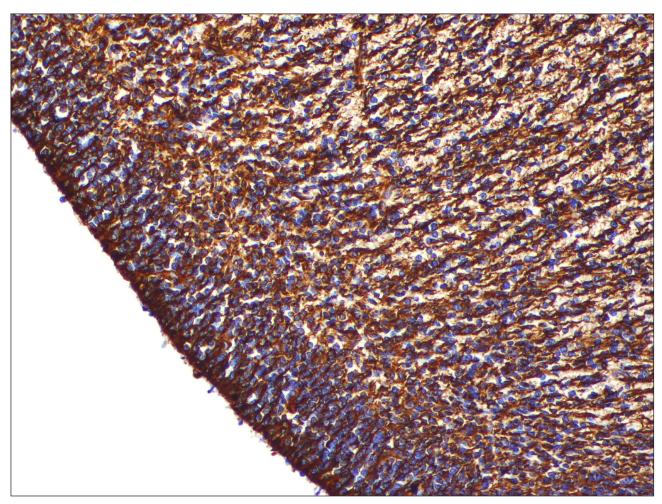


Figure 1 (ABS 68). Immunoreactivity of WT1 in radial glial projections in the ventricular zone (VZ) and subventricular zone (SVZ) in the human fetus at 15 week of gestation.

developing human cerebellum, a strong reactivity for WT1 was observed in radial glial fibers during early stages of cerebellar development. In this study, the strong expression of WT1 in radial glial fibers in the first half of gestation indicate a possible role for this transcription factor in cell migration and differentiation during neurogenesis. These data confirm other findings from previous studies carried out on human fetuses in which WT1 was found in radial glial fibers in the developing human cerebral cortex. On these bases, WT1 may be considered a useful marker of radial glial cells during human brain development.

REFERENCES

 Call KM, Glaser T, Ito CY, Buckler AJ, Pelletier J, Haber DA, Rose EA, Kral A, Yeger H, Lewis WH. Isolation and characterization of a zinc finger polypeptide gene at the human chromosome 11 Wilms'tumor locus. Cell. 1990;60(3):509-20.
 Kreidberg JA. WT1 and kidney progenitor cells. Organogenesis. 2010;6:61-70.

[3] Fanni D, Fanos V, Monga G, Gerosa C, Locci A, Nemolato S, Van Eyken P, Faa G. Expression of WT1 during normal human kidney development. J Matern Fetal Neonatal Med. 2011;24(Suppl.2):44-7.

ABS 69

PEDIATRIC PROCEDURAL SEDATION DURING AMBULATORY TREATMENTS: COMPARISON BETWEEN INHALATION SEDATION WITH NI-TROUS OXIDE AND INTRAVENOUS SEDATION WITH MIDAZOLAM

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INTRODUCTION

Procedural sedation (PSA) is the delivery of sedating or dissociative medications to produce a state of depressed consciousness, with or without opioid analgesics. PSA should allow the patient to maintain continuous and independent ventilation without a loss of protective reflexes. Although adults may be able to tolerate some medical treatments without sedation, the management of pediatric patients needs more attention in order to eliminate pain and fear during the medical session and consequently to avoid the development of future trauma and/ or to make possible some diagnostic/therapeutic procedures in uncooperative children. Children are more prone to anxiety in the acute setting and benefit greatly from sedation and analgesia. Sedation has many uses in pediatrics; PSA is helpful in case of severe pain and anxiety associated with diagnostic tests and it may be necessary in cases of relatively painless procedure, that can be still quite traumatic for the pediatric patient: for this reasons it is often used in pediatric emergency departments. PSA is also used in pediatric dentistry to make possible the management of some minor ambulatory interventions and surgical procedures with uncooperative patients. The choice of sedative agents depends upon the allowable amount of motion during the procedure, comorbidities, age and development level, ability to cooperate, degree of anxiety. Any previous problem with specific medications. The aim of this pilot study is to compare two different type of anesthetic technique used in uncooperative children who need dental treatments: nitrous oxide, which is a colorless gas with a sweet taste widely used as dental and surgical anesthetic for over 150 years and midazolam which is an intermediate-acting benzodiazepine that can be administered by intravenous, intramuscular, oral and intranasal route.

METHODS

The sample consists of 20 children (12 males and 8 females) of average weight between 20 and 35 kg, and between 3 and 13 years old. The sample has been examined in a private pediatric dental clinic. The sample has been divided in two groups, group A and group B each group composed of 10 children: group A clinical condition made the examined patient classified as ASA Group I. The ASA physical status classification system is a system for assessing the clinical condition of patients before surgery. In this case it corresponds to a state of normal health, with no organic, biochemical or psychiatric disorder. It was used an inhaled mixture of fixed equimolar 50% N₂O and 50% O₂ prepared in cylinders (Group A 10 children). For intravenous route it was used midazolam at a dose of 0.25 mg/kg (Group B 10 children).

RESULTS

In group A there was a faster and sedative action and a better awakening. The inhalation procedure was effective both in a sedative and analgesic levels but limited to mildly and moderately painful pediatric dental procedures. Furthermore, the use of nitrogen protoxide appeared safe when administered for very short procedures (about 15 minutes) and only in sufficiently cooperative children. It's not to be neglected that the N₂O appears to have genotoxic potential mechanism that can be reduced with low number and duration of exposures but it remains a problem for the health of the dental team.

In group B, anesthesia was obtained in mild or not cooperating children, with the advantage of being able to perform in tranquility common and complicated dental treatments of long lasting. The procedure intravenously has required more time and can be performed only by experienced anesthesiologists in pediatric range. Moreover another big benefit is that the use of the intravenous route avoids the poisoning of the environment and of all the dental team.

CONCLUSIONS

Recently, epidemiological studies have assessed that the exposure to nitrous oxide can interfere with the action of vitamin B12. Therefore, as a result of chronic exposure to high doses of this gas (a big problem in health care workers), neurological, hematological and immunological disorders, such mieloneuropathies, megaloblastic anemia and leukopenia may occur. A precocious and repeated exposure of N_2O can contribute to negative outcomes on neurological development of children.

ABS 70

A SI-DOHaD-CCM PROJECT: ENVIRONMENT, EPIGENETIC FETAL PROGRAMMING AND PRE-VENTION OF CHRONIC DISEASES

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In many cases, the search of biological biomarkers in tissues or biological fluids has proven to be a precious completion, if not an overcoming, of the process of estimation based on environmental measures and constitutes a fundamental approach in the characterization and management of the health risk. Recent studies concerning the relationship between environmental contamination and biological effects from molecular and epigenetic to tissues and systemic level highlighted how much the main risks of human exposition need to be detected prenatally (environmental fetal programming), since it can result in several developmental abnormalities and affect the health up to the adult life of an individual. Among the pollutant agents, the fetal-maternal exposition and the absorbance of some heavy metals such as arsenic (As), cadmium (Cd), mercury (Hg), chromium (Cr), lead (Pb) - and persistent pollutants such as pesticides, polyaromatic hydrocarbons (PHC) present in each area of the ecosphere and food-chainsresults in significant epigenetic effects overall on in development organisms and, in particular, on central and peripheral nervous system, considering their ability to overcome the placental and the blood brain barrier and to accumulate in embryo-fetal tissues. The aim of this DOHaD project is to deepen the knowledge concerning the fetal-maternal exposition to environmental pollutants and the main molecular and biological mechanism of damage (in particular epigenetic) during the first 1,000 days of life. The innovative potential of this project concerns the adoption of a structured multidisciplinary approach that aims to individuate the presence of early exposition and damage biomarkers starting from fetal level and in particular, placental level usable in neonatology and pediatrics to develop a surveillance system with the aim of primary prevention and early diagnosis of the main disturbances and emerging chronic pathologies. For this purpose, in each partner region (Sardinia, Tuscany, Lombardy and Puglia) 40 pregnant women will be enrolled together with their neonates: 20 will have normal pregnancies and 20 will have pregnancies characterized by alterations of the embryo-fetal development. Normal pregnancies will be characterized by physiological course and normal fetal growth, confirmed by an adequate weight at birth. Pregnancies at risk will be selected on the basis of an accurate anamnesis (exposition to particular

situation of environmental pollution and maternalfetal stress), and signs of actual suffering (problematic pregnancy, prematurity, alterations of fetal growth, low weight related to the gestational age, placenta with sign of phlogosis and functional deficit etc.). In this regard, a questionnaire will be administered that allows obtaining information concerning the presence of previous exposition factors and during the pregnancy. Only singleton pregnancies with elective cesarean section without labor will be included and cases with chromosomopathies and infections will be excluded. Samples of placenta, hair and urine will be collected from mothers. Samples of exfoliated buccal mucosal cells and urines at birth and at 6 months of life will be collected from neonates. On the samples of placenta and exfoliated buccal mucosal cells, the epigenetic investigations will be performed. In the placental samples, the concentration of the main heavy metals (Pb, Cr, As, Hg, Cd...) PAH and PFAS through the ICP-MS technique (Inductively Coupled Plasma-Mass Spectrometry) will be measured. Through 1H-NMR spectroscopy combined with the multivariate statistical analysis, the metabolome of placenta and urine of mothers and urine of neonates collected at birth after 6 and 12 months of life will be characterized. Histological and immunohistochemical analysis will be performed on placentae, in order to evaluate the complex relations between cytotrophoblast and syncytiotrophoblast during development. Ultimately, the state and the regulation of mitochondria will be characterized on placental samples, given their crucial role of energy producers of the cells and their susceptibility to environmental stress events: their content and the methylation of mitochondrial DNA and the activator of biogenesis will be analyzed. The comparison and the crossed analysis of the epigenetic, metabolic, morphological and quantitative compositional profiles for the characterization of the exposition to heavy metals and other epigenotoxic pollutants collected from pairs of at-risk mother-neonate and in controls will allow to: a) detect significant placental and fetal alterations, to correlate: upstream with specific biomarkers (epigenetic, of mitochondrial stress, metabolomics...); downstream with signs and symptoms of prenatal and postnatal suffering; b) demonstrate the existence of biomarkers and early signs that can be used for primary prevention strategies, early diagnosis, individualized counseling and intervention focused ond disturbances and chronic pathologies with possible epigenetic/fetal origin (fetal programming). Thus, the desired result of this project will be the development of an analytical methodology to be used as a model to elaborate a simple ergonomic and economic test/ kit for a serial determination of biomarkers to be used in clinical practice, at patient's bedside and/ or at home. The results of this study will be widely nationally and internationally diffused thanks to an accurate plan of communication and dissemination planned in the project. In particular, the publication of the result of this study and innovation is planned on the Open Access and on scientific journals of European and international relevance in order to favor a further progress in this field.

ACKNOWLEDGMENT

Granted in 2017 by CCM, Ministry of Health, Italy. **REFERENCES**

• Berti C, Agostoni C, Davanzo R, Hyppönen E, Isolauri E, Meltzer HM, Steegers-Theunissen RP, Cetin I. Early-life nutritional exposures and lifelong health: immediate and long-lasting impacts of probiotics, vitamin D, and breastfeeding. Nutr Rev. 2017;75(2):83-97.

• Burgio E. Environment and Fetal Programming: the origins of some current "pandemics". J Pediatr Neonat Individual Med. 2015;4(2):e040237.

• Faa G, Manchia M, Pintus R, Gerosa C, Marcialis MA, Fanos V. Fetal programming of neuropsychiatric disorders. Birth Defects Res C Embryo Today. 2016;108(3):207-23.

• Faa A, Ambu R, Faa G, Fanos V. Perinatal heart programming: long-term consequences. Curr Med Chem. 2014;21(27):3165-72.

• Fanos V, Van den Anker J, Noto A, Mussap M, Atzori L. Metabolomics in neonatology: fact or fiction? Semin Fetal Neonatal Med. 2013;18(1):3-12.

• Luyckx VA, Perico N, Somaschini M, Manfellotto D, Valensise H, Cetin I, Simeoni U, Allegaert K, Vikse BE, Steegers EA, Adu D, Montini G, Remuzzi G, Brenner BM; writing group of the Low Birth Weight and Nephron Number Working Group. A developmental approach to the prevention of hypertension and kidney disease: a report from the Low Birth Weight and Nephron Number Working Group. Lancet. 2017;390(10092):424-8.

• Puddu M, Fanos V, Podda F, Zaffanello M. The kidney from prenatal to adult life: perinatal programming and reduction of number of nephrons during development. Am J Nephrol. 2009;30(2):162-70.

ABS 71

CONGENITAL DIAPHRAGMATIC HERNIA: EX-PERIENCE OF A TERTIARY NICU

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INTRODUCTION

Congenital diaphragmatic hernia (CDH); with an estimated prevalence of 2.3 in 10,000 births, has a very high mortality. The aim of this study is to

evaluate the cases with CDH retrospectively and to determine the prognostic factors in Ondokuz Mayıs University Faculty of Medicine Neonatal Intensive Care Unit.

METHODS

Hospital records of 31 cases diagnosed as CDH between April 2006-August 2016 were retrospectively evaluated. The prognostic value of demographic and clinical data for determining survival was analyzed.

RESULTS

Data of 18 (58.1%) boys and 13 (41.9%) girls were evaluated. 23 (74.1%) were born term and 8 (25.8%) were preterm. 18 (58%) patients had surgical repair. Three (9.6%) patients had genetic abnormalities, 27 (87.1%) patients had additional abnormalities, and 21 (67.7%) had cardiac abnormalities. The CDH was left sided in 26 (83%) and right sided in 2 (6.4%) cases. 13 patients (41.9%) received HFOV treatment, 13 patients (41.9%) were given surfactant and 19 patents (61.3%) received HFO and/or surfactant. 14 patients (45.2%) survived, where 17 (54.8%) died. Cases treated with HFO and/or surfactant treatment had higher mortality when compared the ones who did not require these treatment modalities (73.7% vs 25%, p = 0.008). The major prognostic factor determining mortality was the presence of a cardiac abnormality.

CONCLUSIONS

42% of cases in our series died before corrective surgery. The survival ratio in patients after corrective surgery was 80%. The need for HFO and/ or surfactant treatment and the prescence of a major cardiac anomaly determined survival. This data is important for counselling families both prenatally and postnatally.

REFERENCES

 Snoek KG, Capolupo I, van Rosmalen J, Hout Lde J, Vijfhuize S, Greenough A, Wijnen RM, Tibboel D, Reiss IK; CDH EURO Consortium.
 Conventional Mechanical Ventilation Versus High-frequency Oscillatory Ventilation for Congenital Diaphragmatic Hernia: A Randomized Clinical Trial (The VICI-trial). Ann Surg. 2016;263:867-74.

• Zani A, Zani-Ruttenstock E, Pierro A. Advances in the surgical approach to congenital diaphragmatic hernia. Semin Fetal Neonatal Med. 2014;19: 364-9.

ABS 72

AN INTERACTIVE TOOL SHOWS COMOR-BIDITIES OF CHILD AGGRESSION WITH OTHER CHILDHOOD PSYCHOPATHOLOGIES. UTILITIES AND STATISTICS OF THE WEBSITE WWW.ACTION-EUPROJECT.EU M. Mauri¹, A. Hendriks², V. Fanos³, D.I. Boomsma², M. Bartels²

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The ACTION project (Aggression in Children: Unraveling gene-environment interplay to inform Treatment and InterventiON strategies), funded by the European commission (FP7/2007-2013), under grant agreement no. 602768, brings together a team of scientists from 8 countries and 12 research partners. The main objective of ACTION is to improve the understanding of the causes of individual differences in aggression among children in order to better inform the development of prevention and treatment strategies. A strong focus in this collaboration is on gene-environment interplay and biomarkers at the level of metabolomics. Innovative studies that integrate genetic and environmental risk indicators are possible in the large paediatric twin cohorts, in combination with clinical cohorts comprising patients with positive and negative treatment responses, and population-wide databases that link to outcome variables later in life. Aggression inflicts a huge personal, psychological and financial burden on affected individuals, their relatives, and society at large [1, 2]. Despite large scientific, preventive, and treatment investments, no decrease in aggressive behavior is seen. It is known that childhood aggression often occurs with other childhood psychopathologies [3, 4]; the databases built by the ACTION partners in the years and made available to the consortium, allow for testing comorbidities within different countries across Europe. The ACTION consortium has access to large twin-family cohorts with databases including longitudinal prospective information on aggression and related emotional and behavioural problems and also large-scale genetic, environmental, and biological information. Recently the ACTION consortium released an interactive tool showing the comorbidities of child aggression with other childhood psychopathologies [5]. It is based on the first results of the ACTION consortium that will be published in forthcoming scientific papers. The interactive tool can be browsed by questionnaire (i.e., ATAC, CBCL, DCB, SDQ, MPNI) or by psychopathology (i.e., ADHD, Autism, ODD, OCD, Attention problems, Withdrawn, Anxiety,

Depression, Emotional reactivity, Somatic complaints, Sleep problems, Social problems, Rule breaking, Thought problems, Social isolation, Emotional lability, Dependency, Physical coordination problems, Hyperactivity, Peer problems, Inattention, Impulsivity). This interactive tool collects in an innovative way, the parent- or selfreported questionnaires on children aged between 3 to 16 years old, and allows looking at descriptive statistics (Fig. 1). Since its kick-off in 2014, the ACTION project's results and outcomes are shown to a broad public of researchers and parents in its official website [6]. Day by day the website offers news on events, publications and presentations. The comorbidity tool has been visited more than 1,000 times, it is under constant improvement and it will be integrated during the project's lifetime if new results will be available. This was the last outcome of the project made available through the ACTION website which counts 8 main sections and several webpages dedicated to the research on aggression in children. The website has been visited more than 11,000 times (in the last two years) by people/visitors from 91 countries in the world (Netherlands, Italy, US, UK, India, Germany, Finland, France, Spain, Australia are in the "top ten") and from 583 cities (Amsterdam, Cagliari, Leiden, Rome, Boston, Helsinki, Brisbane, Ashburn, London, Rotterdam are in the "top ten") [7] (Fig. 2). The website offers also an aperiodic free newsletter to the subscribers, which contains the most recent studies carried out within the project [8].

ACKNOWLEDGEMENTS

This work has been supported by the ACTION project, funded by the European Union's Seventh Framework Programme under grant agreement no. 602768. **REFERENCES**

[1] Dretzke J, Frew E, Davenport C, Barlow J, Stewart-Brown S, Sandercock J, Bayliss S, Raftery J, Hyde C, Taylor R. The effectiveness and costeffectiveness of parent training/education programmes for the treatment of conduct disorder, including oppositional defiant disorder, in children. Health Technol Assess. 2005;9(50):iii, ix-x, 1-233.

[2] Scott S, Knapp M, Henderson J, Maughan B. Financial cost of social exclusion: follow up study of antisocial children into adulthood. BMJ. 2001;323(7306):191.

[3] Marmorstein NR. Relationships between anxiety and externalizing disorders in youth: the influences of age and gender. J Anxiety Disord. 2007;21(3):420-32.

[4] Marsee MA, Weems CF, Taylor LK. Exploring the association between aggression and anxiety in youth: A look at aggressive subtypes, gender, and social cognition. J Child Fam Studies. 2007;17:154-68.

[5] http://www.action-euproject.eu/ComorbidityChildAggression, last access: September 2017.

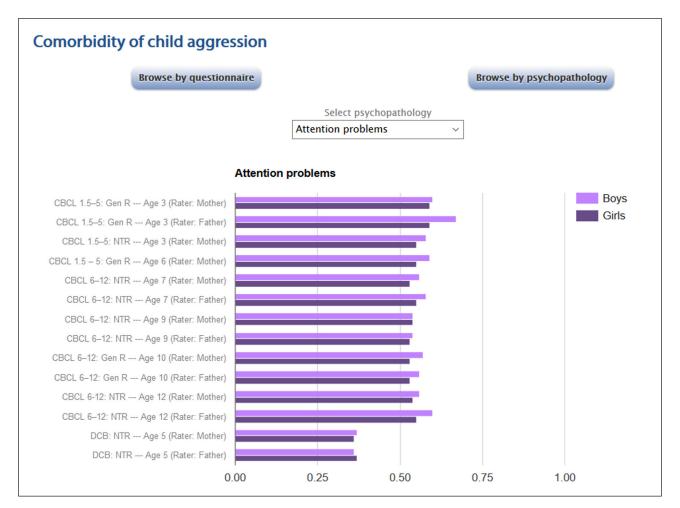


Figure 1 (ABS 72). A snapshot of the ACTION interactive tool that shows comorbidities of child aggression.

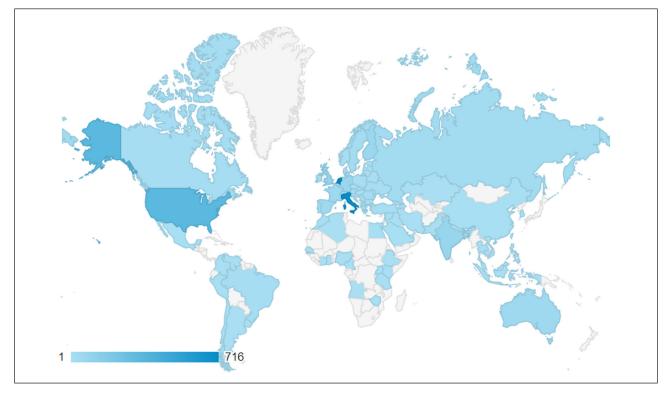


Figure 2 (ABS 72). The geographical distribution of the ACTION website's visitors.

[6] http://www.action-euproject.eu, last access: September 2017.

[7] Google Analytics results. Available at: https://analytics.google.com, last access: September 2017.

[8] ACTION newsletter subscription form. Available at: http://www.actioneuproject.eu/content/action-newsletters, last access: September 2017.

ABS 73

EVALUATION OF MOLECULAR BEACON FLUORESCENT PROBE FOR LABORA-TORY DIAGNOSIS OF CONGENITAL CYTO-MEGALOVIRUS (CMV) INFECTION

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INTRODUCTION

Cytomegalovirus is a species of DNA virus that belongs to Herpesviridae family and represents the leading cause of congenital infection worldwide. It occurs in 0.2% to 2% of live births and 1/5 of these could present long-term health problems. Infected patients spread the virus in cervical mucus, feces, breast milk, saliva even if viral detection of urine has been considered the most reliable method for diagnosis of CMV in newborn. Rapid and accurate laboratory diagnosis of CMV is a crucial point to support the right therapy and proper management of the infection. In this context, the molecular approach by real-time PCR method is a widely available procedure for viral detention/ quantification. This procedure is traditionally based on linear molecular probes (SYBR Green, TaqMan) able to issue fluorescence during the DNA amplification, although different authors reported technical problems in specificity and reproducibility by using these fluorescent probes. In other cases, these molecules require an expensive laboratory equipment, not suitable for example in developing countries. The objective of the present study was to compare traditional PCR TaqMan kit for CMV with a circular probe called Molecular Beacon (MB). The MBs are hairpin-shaped oligonucleotide probes, and have the property of fluorescing upon hybridization with a specific DNA sequence (Fig. 1). Their hairpin shape increases target specificity providing a lower background signal compared to that found in other linear probes previously cited.

METHODS

145 urine samples from 101 patients, 65 new cases and 36 from follow-up, were collected from February 23rd 2016 to November 7th 2017 at Neonatal Intensive Care Unit, AOU Cagliari. After collection, in order to prevent DNA degradation, specimen was centrifuged at 3,000 rpm for 10 minutes at 4°C. DNA extraction was performed by using 0.2 mL of urine pellet with "GenProof Pathogen Free DNA Isolation PCR Kit" (Brno, Czech Republic), following the urine extraction procedure. The oligonucleotide primers and the MB fluorescent probe, used to perform the quantitative real-time RT-PCR, were designed in the CMV glycoprotein B gene viral region, GenBank accession n. X17403, by using oligo 6 (MedProbe, Oslo, Norway) and Mfold programs. A DNA fragments, representative for CMV target sequence, was used as quantitative standard and it was serially 10-fold diluted to contain molecule numbers ranging from $6 * 10^6$ to $6 * 10^{-2}$ copies per µL. PCR reaction has been performed with Premix Ex Taq PCR kit (Takara, USA) following the manufacture instruction. As described before, all samples were then evaluated, for a comparative study, with a commercial kit, CMV DNA QT Kit (Dia-Pro, Italy).

RESULTS

Of 101 patients, 13 were CMV positive (12.8%). The results showed that 28 out of 28 urine positive with MB (on 145) were also positive by CMV commercial Kit, indicating a good agreement between the 2 molecular methods. The detection limit for both procedures was about 0.6 copies μ L. On the contrary, MB procedure showed a better dynamic quantitation range, **Fig. 2**. Positive patients showed a range of CMV copies/ μ L from

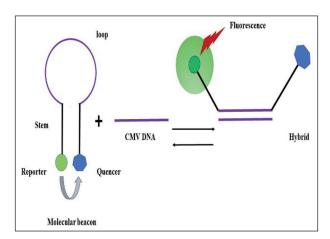


Figure 1 (ABS 73). Schematic representation of a Molecular Beacon (MB) reaction.

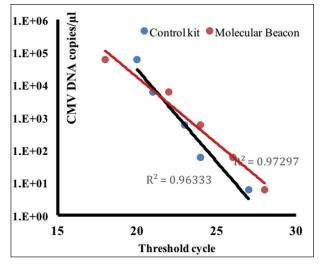


Figure 2 (ABS 73). CMV quantitation curves by using Molecular Beacon (MB) and control kit.

105 to 10, corresponding to $5 * 10^7$ and $3 * 10^3$ CMV copies/mL urine. We have observed negative results using biological samples positives for other Herpes virus (HHV 1, 2, 3, 4).

CONCLUSIONS

In summary, the sensitivity/specificity of the MB procedure developed in this study could be a good diagnostic method for congenital CMV infection. This procedure could be used also as end point analysis by traditional PCR for avoiding the agarose gel procedure and the expensive PCR equipment. REFERENCES

• Orrù G, Ferrando ML, Meloni M, Liciardi M, Savini G, De Santis P. Rapid detection and quantitation of Bluetongue virus (BTV) using a Molecular Beacon fluorescent probe assay. J Virol Methods. 2006;137(1):34-42.

• Tyagi S, Kramer FR. Molecular beacons: probes that fluoresce upon hybridization. Nat Biotechnol. 2006;14(3):303-8.

https://www.cdc.gov/cmv/congenital-infection.html, last access: September 2017.

• http://www.molecular-beacons.com/, last access: September 2017.

ABS 74

THE ETIOLOGY OF PROLONGED JAUNDICE

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INTRODUCTION

Prolonged jaundice (PJ) is a common problem in pediatric practice. Although the most common etiology is breast milk jaundice (BMJ), the need to exclude underlying serious etiologies is dilemma for the pediatricians. NICE and Turkish Neonatal Society (TNS) advice direct and conjugated bilirubin measurements, complete blood count with peripheral smear, blood group determinations of mother and baby, Direct Coombs test of the baby and urine culture (UC) in babies with PJ [1]. TNS also advise screening for Glucose-6-phosphate dehydrogenase deficiency (G6PD). Both groups warn to ensure that screening for congenital hypothyroidism has been performed and inquire about pale stools and dark urine. In this study we aimed to investigate the etiology of PJ in an outpatient clinic and to determine whether basic work-up would be enough.

METHODS

Babies attending to neonatal outpatient clinic of Ondokuz Mayıs University with PJ between 01.10.2013 and 31.10.2015 were included. Prolonged jaundice was defined as jaundice persisting longer than 14 days for term, 21 days for preterm babies. After a detailed history and physical examination, babies were investigated for the etiology of PJ according to the unit and TNS protocol; including serum TSH and fT4, G6PD and UC by catheterization. Babies with hepatosplenomegaly and/or direct hyperbilirubinemia underwent detailed investigation.

RESULTS

Of 100 babies attending the clinic with PJ, 56% were boys and 44% were girls. Mean birth weight was $3,185 \pm 406$ (2,000-4,020) g, average gestational age was 38.6 ± 1.6 (32-41) weeks and mean age at admission was 27 ± 12 (14-68) days. Jaundice was first noticed on 5 ± 4 days (1-21). 89% were exclusively breastfed, 9% were mix fed and 2% were formula fed. Average serum total bilirubin level was 12 ± 4 (2.6-27) mg/dl. Two patients had direct bilirubinemia. The etiology of PJ is shown in **Tab. 1**.

CONCLUSIONS

67% of cases with PJ were due to inadequate breast milk and poor weight gain or due to BMJ [2]. Blood group incompatibilities were also common (18%). We conclude that many babies with PJ can be managed with basic diagnostic work-up including a detailed feeding history and physical examination.

REFERENCES

[1] https://www.nice.org.uk/guidance/cg98/chapter/1-Guidance#care-ofbabies-with-prolonged-jaundice, last access: September 2017.

[2] Rodie ME, Harry C, Taylor R, Rodie ME, Harry C, Taylor R, Barclay AR, Cochran D, Simpson JH. Rationalized assessment of prolonged jaundice is safe and cost-effective. Scott Med J. 2012;57(3):144-7.
 Table 1 (ABS 74). Etiology of prolonged jaundice (PJ).

Etiology	n (%)
Breast milk jaundice	54
Breast feeding jaundice	13
Blood group incompatibility	18
Hemolytic anemia	3
Hereditary spherocytosis	2
Congenital hypothyroidism	2
Biliary atresia	2
Cephalohematoma	1
Congenital CMV infection	1
Breast milk jaundice+ hemolytic anemia	1
UTI + AB0 incompatibility	1
UTI	1
Criggler Najar syndrome	1
Total	100

UTI: urinary tract infection.

ABS 75

CHILD-FRIENDLY HOSPITAL ENVIRONMENT – NEW APPROACH WITH THE USE OF 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 work as a temporary distraction strategy: after few days the effect is reduced. Augmented Reality (AR) allows children to live, every time, a different experience. The aim of this study is to explore the impact of creating a child-friendly hospital environment using AR.

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 created posters and stickers that interact with the AR app letting children see magic characters and super powers appear in front of them and drip bags and other medical devices transform into magic potions. During the study Brave Potions materials will be used to decorate hospital rooms and wards. General surveys, test such PANAS and others to evaluate pain, using different pain rating scales (faces, numeric, behavioural, behavioural/ physiological) will be conducted.

RESULTS

Super Powers has been installed in different hospitals and ambulatories, and we are evaluating how children and families interact with the different materials. 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 AR can provide positive effects 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.

REFERENCES

• Hamdan AB, AlShammary S, Tamani JC, Peethambaran S, Hussein M, AlHarbi M. The Impact Of Creating A Child-Friendly Hospital Environment In Pediatric Cancer Patients And Their Families In Comprehensive Cancer Center At King Fahad Medical City. Available at: http://www.currentpediatrics.com/articles/articles/the-impact-of-creating-a-childfriendly-hospital-environment-in-pediatric-cancer-patients-and-their-families-in-comprehensive-cance. html, date of publication: 2016, last access: September 2017.

 Lankston L, Cusack P, Fremantle C, Isles C. Visual art in hospitals: case studies and review of the evidence. J R Soc Med. 2010;103(12):490-9.

ABS 76

DUCTUS VENOSUS IN EXPECTANT MAN-AGEMENT OF EARLY IUGR FETUSES AND RISK OF ADVERSE NEONATAL OUTCOME; A SURVIVAL ANALYSIS

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INTRODUCTION

Ductus venosus (DV) is usually used for timing the delivery in IUGR fetuses. The aim of this study was to investigate the relationship between abnormal blood flow in DV and intrauterine fetal demise (IUFD), cord blood pH, base excess (BE) at birth

considering time lasted between ultrasound and birth.

MATERIALS AND METHODS

150 consecutive early IUGR fetuses who underwent ultrasound assessment of DV were considered. The DV was related to IUFD and fetal acid base status at birth. An abnormal DV was considered that with absent or reverse A-wave. Time between first ultrasound examination and birth was considered. BESULTS

The mean gestational age at delivery was 29.4 weeks (± 1.75) and the mean gestational age at first ultrasound was 28.8 weeks (± 2.00) in the group with normal DV and $28.43 (\pm 2.09)$ in the group with abnormal DV. Mean fetal weight at birth was 875.69 grams (± 224.62). Abnormal DV was present in 44 cases (29.33%). There were 6 (4%) IUFD, abnormal cord blood pH was present in 70 (48.61%), and BE in 63 (43.75%). Perinatal mortality rate was 21.33% (32/150). Fetuses with abnormal DV had a significant increased risk of IUFD with a cumulative risk of 12% (95% CI: 0-22%) during the first day vs 0% in normal DV (p < 0.05). In normal DV fetuses the cumulative risk of IUFD at 10 days was 5% (95% CI: 0-11%) and was lower respect to abnormal DV (p < 0.05). In the abnormal DV there was an increased risk of abnormal cord blood pH or BE (p < 0.05). In the abnormal DV neonatal death was not associated with abnormal cord blood pH or BE at birth. The only significant correlation was between abnormal cord blood pH at birth and neonatal mortality in the normal DV waveform group (p < 0.05). No significant differences were found in morbidity between the two groups.

CONCLUSIONS

This study shows how a normal or abnormal DV does not reduce IUFD, but is related to a worse acid-base status of the fetuses. This result should be considered as the mortality rate may increase in DV abnormalities while this in not significant for morbidity.

ABS 77

COMPARISON OF pRIFLE AND AKIN CRITERIA FOR DIAGNOSING ACUTE KIDNEY INJURY IN NEWBORNS

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INTRODUCTION

Although newborns are a vulnerable population, the definition and diagnosis of acute kidney injury (AKI) is still not standardized. We aimed to evaluate the prevalence of AKI as defined by paediatric Risk, Injury, Failure, Loss of kidney function and End-stage kidney disease (pRIFLE) and Acute Kidney Injury Network (AKIN) classifications in neonatal intensive care unit (NICU) and to compare these criteria for the diagnosis of AKI.

METHODS

A retrospective analysis of patients admitted to NICU between January 1st, 2012 and March 1st, 2013 was performed. AKI was classified according to AKIN and pRIFLE criteria. Urinary output (ml/kg/h) was also obtained for nRIFLE. RESULTS

815 babies with an average gestational age of 36 \pm 3.3 weeks (48.5% preterm) and birth weight $2,602.5 \pm 833.2$ g were included in the study. AKI was diagnosed in 85 (10.4%) babies according to AKIN and in 115 (14.1%) babies according to pRIFLE criteria. The etiology of AKI was 59.5% prerenal, 39.2% renal according to pRIFLE; whereas 55.5% prerenal, 43.5% renal according to AKIN criteria. The most common time to develop AKI was 2-7 days of life (57% both for AKI and pRIFLE critreia). On logistic regression analysis, risk factors for AKI according to pRIFLE were higher CRIB scores, lower 5th minute Apgar score, sepsis and for AKIN higher CRIB scores and lower 5th minute Apgar score. When we compared AKIN, pRIFLE, AKIN + urinary output and pRIFLE + urinary output, pRIFLE and pRIFLE + urinary output were found superior compared to the other criteria for the diagnosis of AKI (Fig. 1). The mean length of NICU stay was longer, need for mechanical ventilation and mortality rates were higher in patients with AKI when compared to ones without.

CONCLUSIONS

pRIFLE is superior to AKIN and nRIFLE for the diagnosis of AKI newborns. Measurement of urinary output also seems feasible for this at risk population. Neonatal staging systems (preferably including biomarkers) is mandatory since neonatal AKI might have long term consequences.

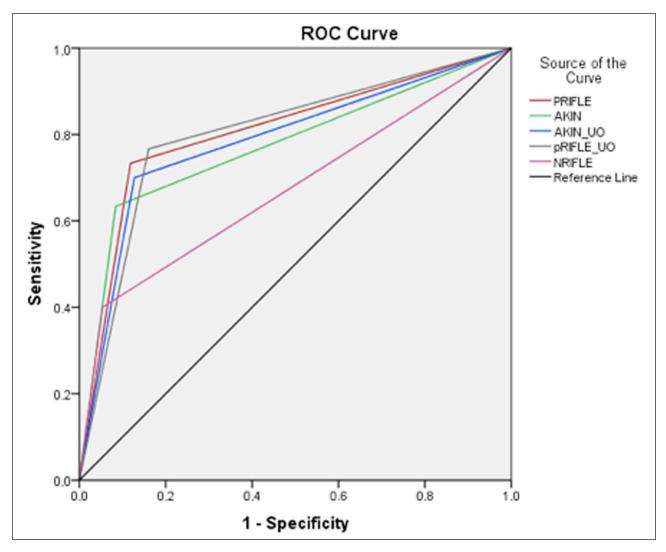


Figure 1 (ABS 77). pRIFLE and pRIFLE + urinary output were found superior compared to the other criteria for the diagnosis of acute kidney injury (AKI).

REFERENCES

• Low Birth Weight and Nephron Number Working Group. The Impact of Kidney Development on the Life Course: A Consensus Document for Action. Nephron. 2017;136:3-49.

• Nada A, Bonachea EM, Askenazi DJ. Acute kidney injury in the fetus and neonate. Semin Fetal Neonatal Med. 2017;22:90-7.

ABS 78

EATING DISORDERS IN TEENAGERS: A CASE REPORT OF ANOREXIA NERVOSA

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INTRODUCTION

Anorexia nervosa is an eating disorder, which is increasing in frequency, especially among adolescent females. At the moment a consensus concerning the causes of the disorder is not available. Social, psychosexual, family emotional climate, biological theories, and a regression hypothesis [1] have been suggested to explain the pathogenesis. The major characteristics of the disease are well described in the literature: 25% loss of body weight, use of various means to lose weight, weight phobia, preoccupation with food, body image disturbances. Again, many medical conditions are associated: bradycardia, hypotension, dehydration, hypothermia, electrolyte abnormalities, amenorrhea, metabolic changes, and abdominal distress.

CASE REPORT

We present the case of a teenage girl aged 15, hospitalized in our clinic for investigations, presenting weight loss (11 kg in 3 months). Clinical examination on admission revealed paleness, fat loss, muscle atrophy, diminished tendon reflexes, bradycardia, epigastric pain and secondary amenorrhea. An initial suspicion of abdominal tumor was invalidated by abdominal CT. Electrolyte disturbances, hypercholesterolemia, low levels of FSH and LH and prolactin in normal ranges, increased urinary and blood amylase in association with normal hypothalamic MRI and neuropsychiatric examination established the diagnosis.

CONCLUSION

A favorable evolution was noticed after treatment with antidepressants and anxiolytics in association with a refueling program and psychotherapy, resulting in weight gain and reappearance of menses.

REFERENCE

 Crisp AH, Burns T, Bhat AV. Primary anorexia nervosa in the male and female: a comparison of clinical features and prognosis. Br J Med Psychol. 1986;59(Pt 2):123-32.

ABS 79

ECHOCARDIOGRAPHIC DIFFERENCES BE-TWEEN SGA AND AGA FETUSES

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INTRODUCTION

The aim of the study was to analyze right and left cardiac function in intrauterine growth restriction (IUGR), Small for gestational Age (SGA) and appropriate for gestational age (AGA) fetuses.

MATERIALS AND METHODS

This was a prospective study performed at the Department of Woman's and Child's Health of the University of Padua in the period between 2015 and 2017. The patients were enrolled during the third ultrasound scan or in suspected growth restricted fetuses. SGA fetuses were defined those with an Estimated Fetal Weight (EFW) between the 3th and the 10th percentile without fetal Doppler alterations; AGA fetuses as EFW between the 10th and the 90th percentile. Conventional echocardiography was performed. at a median gestational age of 34 weeks in the Cardiologic Unit of the University of Padua. All the data collected were those of a complete cardiac exam. Neonatal outcome was registered. RESULTS

Twenty SGA and 62 AGA fetuses were studied. There were no differences about the Global longitudinal systolic peak strain 2D of the left and right ventricle between SGA and AGA fetuses (p 0.33), while left and right indexed cardiac output (ml/mim/kg) were significantly lower in SGA than AGA (p 0.05). Moreover, the Left Ventricle Shortening fraction (%) 2D and Left Ventricle End Diastolic Volume (ml) were lower in SGA compared to AGA (p < 0.05).

CONCLUSIONS

Late preterm SGA fetuses showed signs of cardiac remodeling. These results lead to reconsider the role of low birth weight even in a category of fetuses that not presented intrauterine fetal Doppler abnormalities.

ABS 80

ENDOSCOPIC TREATMENTS IN PEDIATRIC DIGESTIVE PATHOLOGY

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INTRODUCTION

Pediatric endoscopy has developed since its beginnings as fiber endoscopy in 1970s [1]. An important role for the evolution of pediatric endoscopy was the technological improvements in endoscope design and endoscopic devices [2]. The technique requires a well-trained team: pediatric endoscopist, surgeon, anesthesiologist, radiologist, nurses and technicians. This is the most frequent technique presently used in the 5th Pediatric Clinic of Saint Mary Children Emergency Hospital, Iaşi (esophageal stenosis dilatations, polypectomy, etc.). CASE REPORTS

CASE REPORTS

We present 4 cases from our clinic where upper or lower digestive endoscopy played an important role in the diagnosis, treatment and follow-up of the disease: an acute presentation due to gastric pseudoobstruction in a child with gastric Crohn's disease, a case with both esophageal and pyloric stenosis, another case with esophageal stenosis where dilatations with Savary bougies were needed and a family case of an adenomatous polyposis.

CONCLUSIONS

Digestive endoscopy remains a keystone in the structure of modern gastroenterology. Development of therapeutic endoscopy allows efficient, quick and barely aggressive cure of certain diseases, which were only surgically curable before.

REFERENCES

[1] Gilger MA. Gastroenterologic endoscopy in children: past, present, and future. Curr Opin Pediatr. 2001;13:429-34.

[2] Tringali A, Thomson M, Dumonceau JM, Tavares M, Tabbers MM, Furlano R, Spaander M, Hassan C, Tzvinikos C, Ijsselstijn H, Viala J, Dall'Oglio L, Benninga M, Orel R, Vandenplas Y, Keil R, Romano C, Brownstone E, Hlava S, Gerner P, Dolak W, Landi R, Huber WD, Everett S, Vecsei A, Aabakken L, Amil-Dias J, Zambelli A. Pediatric gastrointestinal endoscopy: European Society of Gastrointestinal Endoscopy (ESGE) and European Society for Paediatric Gastroenterology Hepatology and Nutrition (ESPGHAN) Guideline Executive summary. Endoscopy. 2017;49: 83-91.

ABS 81

WHICH DOPPLER PARAMETER DURING THE SECOND TRIMESTER ULTRASOUND SHOULD BE ASSESSED AS PREDICTOR FOR SMALL FOR GESTATIONAL AGE FETUS?

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OBJECTIVE

To assess which Doppler parameter during the II trimester ultrasound scan might predict small for gestational age (SGA) fetuses at term of pregnancy. MATERIALS AND METHODS

We prospectively analyzed pregnancy and neonatal outcomes of 114 singleton pregnancies who underwent a second trimester ultrasound scan at a III level University Hospital from January 2016 to January 2017. All singleton pregnancies were included in this study. During ultrasound examination, all Doppler parameters for each fetus has been assessed: umbilical artery, middle cerebral artery, ductus venosus (DV) and uterine arteries. We excluded all pregnancy related to hypertensive disorders, twin pregnancies and fetal malformations. SGA was defined as a neonatal weight < 10th percentile.

RESULTS

Mean gestational age at ultrasound examination was 21.26 weeks (\pm 1.29) while the mean gestational age at delivery was 38.25 weeks (\pm 2.35). There were no significant differences in gestational age at ultrasound or at delivery between SGA and normal pregnancies. Umbilical and middle cerebral artery did not differ and thus excluded for the prediction of SGA fetuses at term. DV mean PI was 0.58 (\pm 0.16) and was the best predictor for SGA fetuses at term.

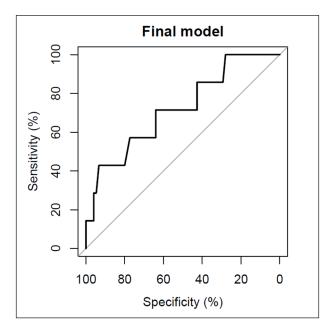


Figure 1 (ABS 81). Final model: sensitivity and specificity

The OR in univariate analysis of DV PI to predict SGA was 0.04 (95% CI 0-0.76; p < 0.05) and in multivariate analysis was 0.04 (95% CI 0.002-0.841; p < 0.05). The area under the curve was 75% (95% CI 51-93%). Results are presented in **Fig. 1**. CONCLUSIONS

Sonographic examination of DV during the II trimester ultrasound might predict SGA fetus at term. This may be important for obstetricians particularly to optimize further ultrasound scans to detect SGA fetuses as well to improve management of these fetuses during labor.

ABS 82

CAPICE: CHILDHOOD AND ADOLESCENCE PSYCHOPATHOLOGY: UNRAVELING THE COM-PLEX ETIOLOGY BY A LARGE INTERDISCI-PLINARY COLLABORATION IN EUROPE

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BACKGROUND

The objectives of the CAPICE project are: 1) identify hereditary variants associated with the event and course of common childhood psychopathology including depression, anxiety and Attention Deficit Hyperactivity Disorder related traits, and set up the hereditary overlap within childhood psychiatric disorders and with adult psychiatric disorders, 2) unwind the mechanisms underlying the associations between early lifestyle factors and childhood psychiatric disorders, 3) recognize new medication targets, and 4) build a risk prediction model that identifies groups of children that are at highest risk to have persistent symptoms. To perform the multisite analyses most efficiently, a facility will be build that allows analyzing all data available over sites without necessarily having access to the raw data. Finally, the results of (epi)genetic and transcriptomic analyses will be summarized and the relevance will be explained to the general audience. This poster provides an overview of the plan to achieve these goals.

METHODS

CAPICE is an international training network, funded by an EU Marie Curie grant, in which 12 PhD students will be trained in psychiatric genomics. This network will elaborate on the EArly Genetics and Lifecourse Epidemiology (EAGLE) consortium.



Figure 1 (ABS 82). The Capice website: www.capice-project.eu/index.php.

EAGLE is a well-established collaboration of many birth and adolescent population based (twin and family) cohorts, with unique longitudinal information on lifestyle, family environment, health, and emotional and behavioral problems. Phenotypic and genome-wide data are available for over 60,000 children, in addition to genome-wide data for over 20,000 mothers and epigenome-wide data for over 6,000 children. Analyses will include (but will not be limited to) twin analyses, genome-wide association meta-analyses, polygenic analyses, Mendelian randomization, and biological pathway analyses. We will use statistical learning techniques (classification trees, support vector machines of Naïve Bayes Classifiers) to investigate the association between the exposure (e.g., risk factors early in life) and the outcome (e.g., adolescent mental health symptoms). The translation of the results to guide clinical treatment will be achieved by the development of a prediction models.

RESULTS

We expect that the outcomes will give knowledge into the etiology of mental health symptoms in children and adolescents and shed light on possible targets for prevention and intervention (e.g. by medication target approval or by tailoring treatment based on the risk for persisting symptoms). The dissemination of the project will be performed with the website: www. capice-project.eu/index.php (**Fig. 1**).

DISCUSSION

It is notable that psychiatric symptoms in childhood be the precursor of numerous psychiatric disorders during adulthood. Longitudinal population based cohorts provide a good opportunity to show how hereditary factors influence development over the ages. Since these disorders are the extreme end of the continuum, collaborations with case-control samples may strengthen the results.

ACKNOWLEDGEMENTS

This work has been supported by the CAPICE project, funded by the European Union's Horizon 2020 Research and Innovation Programme under the Marie Sklodowska-Curie grant agreement no. 721567.

ABS 83

URINARY METABOLOMICS IN TERM NEW-BORNS DELIVERED SPONTANEOUSLY OR WITH CESAREAN SECTION: PRELIMINARY DATA

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INTRODUCTION

In the last years the uncritical attitude towards cesarean section has been associated to the fast emergence of "modern" diseases such as early pediatric obesity, asthma, type 2 diabetes mellitus and dermatitis [1]. It is increasingly understood that in newborns delivered by cesarean section, gut microbiota diversity and origins differ from those of infants born by vaginal delivery. In particular, several potentially beneficial bacterial species, such as bifidobacteria species, are in lower abundance, and may eventually link to perturbation of normal development and maturation of a child's immune system. Amongst emerging systems biology approaches, metabolomics is foreseen as the "Rosetta Stone of microbiomics" [2, 3]. The aims of this study was therefore to assess the influence of the mode of delivery in term newborns on urinary metabolic profiles as measured by proton nuclear magnetic resonance spectroscopy, and to evaluate possible perturbations of the host-microbial metabolism during the first few days of life.

MATERIAL AND METHODS

This study was carried out on urine samples from 42 patients admitted to the Neonatal Intensive Unit and Neonatal Pathology of "S. Giovanni Calibita" Hospital Fatebenefratelli (Rome, Italy). The study protocol was approved by the local ethical committee and written informed consent was obtained from the parents before enrolment in the study. According to the type of delivery, term neonates with similar gestational age and birthweight were divided in two groups: (1) born by spontaneous vaginal delivery, n = 21; (2) born by elective cesarean delivery, n =21. Urine samples were matched for sex, gestational age and birth weight, and were collected within 8 h of birth. The clinical data of each patient was recorded in the hospital registers. Each urine sample (volume around 1 mL) was collected by a noninvasive method: a ball of cotton was inserted into the disposable diaper; then urine was aspired with a syringe and transferred to a sterile 2-mL vial. After collection, all the vials were immediately frozen and stored at -80°C until metabolomic analysis.

RESULTS

Newborns delivered spontaneously, showed markedly higher urinary levels of subaric and se-

bacic acids (2 dicarboxylic acids derived from the fatty acid oxidation) and oxaloacetate (involved in several metabolic pathways), metabolites of primary physiological importance related to energy metabolism when compared to C-section born infants. Such increased energy metabolic status seems to be due to the high energy demand of the neonate. In addition, the urine of the children showed lower urinary losses in free choline (which plays a fundamental role in brain fitness meaning that it is involved in myeline synthesis) and myoinositol (which is involved in several physiological pathway, insulin for instance). Several host-bacterial metabolites showed different urinary patterns, including lower hippurate, formate but also higher urinary excretion of methylamines. FUTURE PERSPECTIVES

Metabolomics and microbiomics data integration provides insight into the differences in the pathways of different delivery ways in term newborns [4]. Further studies are required to better understand the level of complexity of how these 2 omics are linked with the exposome, namely the diet, in Perinatology

[5, 6].

REFERENCES

[1] Kulas T, Bursac D, Zegarac Z, Planinic-Rados G, Hrgovic Z. New Views on Cesarean Section, its Possible Complications and Long-Term Consequences for Children's Health. Med Arch. 2013;67(6):460-3.

[2] Fanos V, Atzori L, Makarenko K, Melis GB, Ferrazzi E. Metabolomics application in maternal-fetal medicine. Biomed Res Int. 2013;2013:720514.

[3] Fanos V, Van den Anker J, Noto A, Mussap M, Atzori L. Metabolomics in neonatology: fact or fiction? Semin Fetal Neonatal Med. 2013;18(1): 3-12.

[4] Shaffer M, Armstrong AJS, Phelan VV, Reisdorph N, Lozupone CA. Microbiome and metabolome data integration provides insight into health and disease. Transl Res. 2017 Jul 14. [Epub ahead of print].

[5] Moco S, Collino S, Rezzi S, Martin FP. Metabolomics perspectives in pediatric research. Pediatr Res. 2013;73(4 Pt 2):570-6.

[6] Martin FP, Moco S, Montoliu I, Collino S, Da Silva L, Rezzi S, Prieto R, Kussmann M, Inostroza J, Steenhout P. Impact of breast-feeding and high- and low-protein formula on the metabolism and growth of infants from overweight and obese mothers. Pediatr Res. 2014;75(4):535-43.

ABS 84

ANALYSIS OF TRACE METAL CONCENTRA-TIONS IN HUMAN'S MILK. A PILOT STUDY

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INTRODUCTION

Human milk represents the most important and principal nourishment of the newborn. It is mostly composed by water and sugars (lactose), proteins, fat, and small amounts of vitamins, mineral salts, hormones, antibodies and enzymes. Its composition changes according to the age of the newborn, the mother diet and her nutritional condition and importantly to environmental factors. Some studies have shown that exposure to pollutants during prenatal and postnatal life can determine adverse health effects in children and modify human milk composition. No data are available on the presence of heavy metals in human milk in the Sardinian population. Heavy metals are generally defined as metals with relatively high densities.

OBJECTIVE

To evaluate the feasibility of the determination of the presence of small amount of heavy metals in human milk. To identify correlations between maternal life styles and the presence and concentrations of metals in the milk.

MATERIALS AND METHODS

After mother's informed consent, milk sample of 22 subjects were collected and stored at -20°. Questionnaires on life style, nutrition, work activity and living areas (urban, rural or mining areas). The analysis method set up was performed at the Department of Life and Environmental Science, University of Cagliari. The following heavy metals concentrations were evaluated using a Plasma Mass Spectrometer: antimony, arsenic, aluminum, silver, copper, zinc, nickel, bismuth, boron, barium, cadmium, chromium, mercury, manganese, iron and lead.

RESULTS

15 metals were present in all samples. The most represented metal were zinc, boron, aluminum, copper and iron. Manganese concentrations were inversely correlated to maternal age (p = 0.0045), while zinc concentrations were correlated to maternal age (p = 0.0471). Some metals concentrations were different depending on alimentary or environmental factors.

CONCLUSIONS

Heavy metals are present in human milk and their concentration can be evaluated. Nevertheless the high variability of the metals values between samples and their small number does not consent any definitive conclusion on the role of maternal nutrition and/or occupational and living areas on their concentration. Larger studies are needed to determine these aspects and even to correlate any possible positive or negative effect on newborn growth and development. In particular mother living in mining areas should be monitored as the infants could be at risk of multiple exposure to mining related toxic metals, such as lead, mercury, cadmium and arsenic, through breast milk intake, in addition to *in utero* exposure.

ABS 85

METABOLOMIC ANALYSIS OF PLACENTA TISSUE IN OBESE PREGNANCIES

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INTRODUCTION

The placenta plays a pivotal role as it represents the connection between mother and foetus. It acts as a carrier (nutrients and oxygen, waste products and carbonic anhydride) as well as a barrier against infection. Pregnancy related metabolomics has examined different biofluids in order to reach deeper insights into pregnancy pathologies [1]. Amniotic fluid, urine and plasma were the most studied, while placenta tissue has been the subject of few studies [2]. In this report, a method to analyse the placental metabolome through the GC-MS (Gas Chromatography-Mass Spectrometry) technique is described.

MATERIALS AND METHODS

Placenta tissues were collected after elective caesarean section from obese and normal weight mothers, snap frozen and stored at -80°C until analysis. Each sample was homogenized with a mixture of solvents (chloroform, methanol, water), centrifuged, and the obtained phases were separated and processed following two different protocols. The hydrophilic phase was vacuum dried and treated with two derivatizing reagents, methoxyamine hydrochloride and MSTFA (N-Methyl-N-[trimethylsilyl] trifluoroacetamide), to give a mixture of trimethylsilylated metabolites. The lipophilic phase was dried and treated with boron trifluoride in methanol to obtain a mixture of FAME (Fatty Acid Methyl Esters). The two phases were separately analysed on a GC-MS platform, applying different instrumental conditions. The chromatograms obtained were analysed with the free software AMDIS (Automated Mass Spectral Deconvolution and Identification System) using a lab-made library comprising 200 metabolites.

RESULTS

The analysis of the placenta tissue allowed to identify essential metabolites. In the hydrophilic fraction, 78 metabolites were detected: they mainly belong to the classes of amino acids (18), carboxylic acids (13), sugars and sugar-related compounds (11), phosphorylated derivatives (9). The lipophilic fraction analysis allowed to define the fatty acid profile of the placenta extract: the saturated fatty acids palmitic and stearic, the monounsaturated oleic, and the polyunsaturated (PUFA) linoleic, arachidonic, 8,11,14-eicosatrienoic and 4,7,10,13,16,19-docosahexaenoic acid (DHA) were the most representative. Statistical analysis of the data matrices derived from the hydrophilic and lipophilic fraction analysis were performed in MetaboAnalyst 3.0 [3], to investigate the possibility of identifying characteristic metabolic differences between the different phenotypes. Placentae from obese mothers were compared with those from normal weight mothers to get new insights into the correlation between the metabolite content and metabolic status.

CONCLUSIONS

The reported analysis method allowed to examine different metabolites in placenta samples, divided into two phases, hydrophilic and lipophilic. All metabolites were analysed through the same GC-MS platform to provide interesting informations about the placenta tissue composition.

REFERENCES

 Fanos V, Atzori L, Makarenko K, Melis GB, Ferrazzi E. Metabolomics application in maternal-fetal medicine. Biomed Res Int. 2013;2013: 720514.

[2] Dunn WB, Brown M, Worton SA, Davies K, Jones RL, Kell DB, Heazell AEP. The metabolome of human placental tissue: Investigation of first trimester tissue and changes related to preeclampsia in late pregnancy. Metabolomics. 2012;8:579.

[3] Xia J, Sinelnikov IV, Han B, Wishart DS. MetaboAnalyst 3.0 – making metabolomics more meaningful. Nucleic Acids Res. 2015;43(W1): W251-7.

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PATAU SYNDROME: RADIOLOGICAL CHAR-ACTERISTICS

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INTRODUCTION

Patau syndrome (Trisomy 13) is a rare chromosomal anomaly caused by the presence of an extra chromosome 13 and characterized by multiple somatic and visceral malformations, associated with severe metal impairment. Its incidence is estimated around 1:16,000 newborns and only 5 to 10 percent of children with this condition live past their first year. Postpartum diagnosis of trisomy 13 has been drastically reduced by the prenatal screening tests. CASE REPORT

We're presenting the case of a patient with multiple malformations, who was diagnosed with complete trisomy 13 (karyotype: 47 XY +13). An augmented risk of chromosomopathy was found during first semenster screening tests and confirmed by fetal US scan and MRI. Nevertheless, parents refused to undergo any invasive procedure to determine the disease. At birth the little patient presented scaphocephaly with patent bregmatic fontanelle, microphthalmia and micrognathia. Both hands had a subcentimetric appendage attached to the ulnar side and the right foot showed hexadactyly with syndactyly of V-VI toe. Bilateral cryptorchidism was also noted. Pre and postnatal US scans evidenced hypoplasia of the left kidney and malrotation of the right one, persistence of the Botallo duct and a suspect of interatrial defect with left to right shunt. MRI showed cerebral pachygyria and absence of both crystalline lenses. The clinical conditions were critical from birth, since the patient presented respiratory distress, and worsen at the third day of age, when the patient developed a gastric perforation and underwent an emergency surgery. After several episodes of desaturation, the little patient died at day 21.

CONCLUSIONS

The aim of this case report is to recall the clinical and radiological characteristics of this rare genetic syndrome, since the postnatal diagnosis is becoming unusual in our daily practice.

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A CASE REPORT OF BECKWITH-WIEDEMANN SYNDROME: THE LENS OF THE RADIOLOGIST

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INTRODUCTION

Beckwith-Wiedemann Syndrome (BWS) is a pediatric overgrowth disorder, with an increased risk of childhood tumors. The incidence of BWS is estimated to be 1 out of 13,700 and is caused by various epigenetic and/or genetic alterations. It is characterized by many additional clinical features (major criteria) such as hypoglycemia, macrosomia, macroglossia, hemihyperplasia, cardiomiopathy, omphalocele, visceromegaly, adrenocortical cytomegaly, renal abnormalities, etc. The diagnosis of BWS is established with three major criteria or an epigenetic or genomic alteration leading to abnormal methylation at 11p15.5. Molecular genetic testing for this alteration can be done also on amniocytes in prenatal test (in high risk pregnancies due to positive family history) as well as ultrasound examinations to assess fetal growth and to detect other abnormalities characteristic of BWS (no high risk pregnancies). Most important differential diagnosis with other overgrowth syndromes like: Costello, Sotos, SGB type I. Thanks to the recent improvements in syndrome recognition and treatment the overall mortality rate of BWS is about 10-20% with most deaths occurring early secondary to congenital malformations or prematurity.

CASE REPORT

A preterm (27 w and 1.4 kg) female, born by caesarean section, showed omphalocele, right emihypertrophy and respiratory distress (also seen on first X-ray). Ematochemical analysis showed: hypoglycemia, increase of bilirubin (tot 10 mg/ dl), all cholestasis index (GGT 190 U/I, LDH 1,085 U/I), CPK (682 U/I) and WBC (12,000/µl). Suspecting other organs abnormalities, a study of abdomen with Ultrasound (US) was required. US showed an important hepatomegaly (right lobe 77 mm), bilateral dishomogeneous hypoechoic mass in

adrenal loggias (max 35 mm on the right) referable to hemorrhage, hypertrophy of pancreas and right kidney; it also showed perihepatic effusion and herniation of the abdominal contents into the base of the umbilical cord (omphalocele). No significant finds on echocardiography (hemodynamic overload) and cranial sonography. Having not evacuated meconium, in suspicion of intestinal occlusion, we proceeded to digestive tract X-ray with contrast: no progression of contrast medium persisted on stomach after few hours. On the third day the child died due to numerous complications.

CONCLUSIONS

Although only 10-20% of risk BWS could be mortal. The use of US (in prenatal test too) and other diagnostic methods like X-ray could help in recognition and early treatment of disease as well as on the final diagnosis of BWS, together with genomic studies.

ABS 88

THE TYPE OF DELIVERY STRONGLY IMPACTS ON THE COLOSTRUM MICROBIOTA COM-POSITION

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INTRODUCTION

Human colostrum is a rich nourishing substance that is essential for the initial development of newborns. Colostrum contains numerous microorganisms including *Staphylococcus*, *Streptococcus* and *Bacteroides genera*, that together with probiotic bacteria such as *Alloiococcus spp.*, can reduce the incidence and severity of several gastrointestinal infections by both competitive exclusion mechanisms and production of antimicrobial compounds [1]. The aim of the present study was to assess the effect of delivery mode on the microbiota composition of colostrum samples. Specifically, we evaluated both bacterial abundance and interactions (microbiota network) within colostrum samples associated with either C-section or vaginal delivery by a next generation sequencing (NGS) approach applied to Auto-Contractive Map (AutoCM).

METHODS

Twenty-nine Italian mothers (15 vaginal deliveries vs 14 Cesarean sections) were enrolled in the study. The study was conducted following ethical approval by the hospital committee, in accordance with Italian standards (Ethical Committee of the Azienda Ospedaliera of Verona, Italy, approval No. 1288). Informed consent was obtained from all subjects. Information about mother and newborn pairs were recorded during the study. The microbiota of colostrum samples was analyzed by next generation sequencing (Ion Torrent Personal Genome Machine), as previously described [1]. The colostrum microbiota network associated with Cesarean section and vaginal delivery was evaluated by means of the Auto Contractive Map (AutoCM), a mathematical methodology based on Artificial Neural Network (ANN) architecture. RESULTS

differences Numerous between Cesarean section and vaginal delivery colostrum were observed. Vaginal delivery colostrum had a significant lower abundance of Pseudomonas spp., Staphylococcus spp. and Prevotella spp. when compared to Cesarean section colostrum samples. Furthermore, the mode of delivery had a strong influence on the microbiota network, as Cesarean section colostrum showed a higher number of bacterial hubs if compared to vaginal delivery, sharing only 5 hubs. Interestingly, the colostrum of mothers who had a Cesarean section was richer in environmental bacteria than mothers who underwent vaginal delivery. Finally, both Cesarean section and vaginal delivery colostrum contained a greater number of anaerobic bacteria genera.

CONCLUSIONS

The mode of delivery had a large impact on the microbiota composition of colostrum. Further studies are needed to better define the meaning of the differences we observed between Cesarean section and vaginal delivery colostrum microbiota.

REFERENCE

[1] Drago L, Toscano M, De Grandi R, Grossi E, Padovani EM, Peroni DG. Microbiota network and mathematic microbe mutualism in colostrum and mature milk collected in two different geographic areas: Italy versus Burundi. ISME J. 2017;11(4):875-84.

ABS 89

THIRD BRANCHIAL CLEFT CYST: A POTENTIAL DIFFICULT RADIOLOGIC DIAGNOSIS

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INTRODUCTION

Third branchial cleft cyst is a very rare type of branchial cleft cyst and represent an abnormal persistence of the branchial apparatus. Typically is affected the left side (97%) 1 and they are typically asymptomatic; symptoms occur only in case of enlargement or infection and usually are diagnosed in infancy and childhood. We present a case of neonatal diagnosis of third branchial cleft cyst extended in retropharynx space with a fistula between the cyst and the pharyngeal lumen, symptomatic after ten days of the birth. CASE REPORT

A male at-term neonate came to our observation for a left laterocervical swelling, rapidly growing in recent days, and fever. Prenatal and postnatal examination performed did not show any noteworthy element. The ultrasound examination showed a single ipo-anechoic mass (36 x 23 x 20 mm), septated, with defined limits and thickened and intensely vascularized walls (4 mm); locoregional lymphadenopathies are observed. The mass is localized posterior to the sternocleido muscle, posteromedial to the common carotid and posterior to the left thyroid lobe, anteriorized, and with a retropharyngeal extension. MRI demonstrated in the posterior cervical space a large homogeneous low attenuation mass with well-circumscribed margins and hyper intensity relative to the muscle on T2-weighted images; the thickened walls show intense hyper intensity in DWI sequences. From the medial side of the cyst it seems to be observed a fistula that open into the pharyngeal lumen. After antibiotic treatment fever dropped and the cyst decreased in dimensions.

CONCLUSIONS

The third branchial cleft cyst is a vestige of the branchial apparatus, which appears during the fourth week of gestation like six paired sets of arches, each with an associated internal pouch and external cleft. These anomalies can manifest themselves as any combination of fistula, sinus or, commonly, cyst. Cyst arising from the third or fourth branchial cleft are extremely rare (the majority of branchial anomalies arise from the second cleft) but represent the second most common congenital lesion of the posterior cervical space of the neck after cystic hygroma. The radiological diagnosis may be difficult, but the superficial location of some of these cysts makes them well suited for sonography examination (hypo-anechoic mass but infection and hemorrhage will increase echogenicity). CT scan and MR imaging are the imaging modalities of choice to assess the extent and depth of these lesions. Histopathologic examination of the specimen from our patient confirmed the diagnosis of a third branchial cleft cyst. The surgical excision is necessary to prevent recurrence.

ABS 90

SOLITARY KIDNEY: OUTCOME OF 209 PA-TIENTS IN PEDIATRIC AGE

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INTRODUCTION

Solitary kidney (SK) is a congenital or acquired condition characterized by a functional or anatomical absence of the controlateral kidney [1].

Defects in kidney development are part of the Congenital Anomalies of the Kidney and Urinary Tract (CAKUT) [1]. CAKUT include several clinical entities, from complete renal agenesis to hypodysplasia or renal dysplasia.

The purpose of this study is to evaluate the clinical development and the long-term outcome of SK in children.

METHODS

We retrospectively evaluated 209 children affected by SK followed in our center in the last 20 years (1998-2017), 150 males (71.7%) and 59 females (28.2%). All patients underwent urine exam, glomerular filtration rate (GFR), cystatin C, ultrasound monitoring, sequential renal scintigraphy with Tc-99 Mercaptoacetyl-triglycine (MAG3), and blood pressure control.

Voiding cysto-urethrogram (VCUG) was performed only in children with dilated urinary tract to exclude the presence of other CAKUT (vesicoureteral reflux, duplex kidneys).

RESULTS

Out of a total of 209 children, prenatal diagnosis was performed in 152 patients (72.7%); 126/209 patients (60%) presented dysplasia abnormalities (multicystic dysplastic kidney, MCDK).

Out of a total of 209, 52 patients (24.8%), 39 males and 13 females, had complex single kidney.

Contralateral kidney compensatory hypertrophy was found in 184/209 patients (88%). 6/209 (2.87%) of all the patients had associated syndromes or delay of neurologic development.

Two children (0.9%) had a reduced glomerular filtrate compared to the lower limit expected for their ages. In 4/209 patients (1.9%) proteinuria (100 mg/dl) was found in the urinary sediment. Systolic blood pressure values greater than the 95th percentile for the respective ages were found in 3 patients (1.4%). Only 17/209 children (8.1%) developed recurrent urinary tract infections. We found no malignant degeneration. CONCLUSIONS

Most children with SK are in good health [1]. In cases of uncomplicated SK, natural history shows a favorable prognosis [2] and there is no indication of nephrectomy [1] except in the presence of a dysplastic kidney with a size that can cause significant abdominal encumbrance in the postnatal period. VCUG is no longer a recommended instrumental examination for all patients with SK [3] and should only be performed in selected cases. Follow-up should be performed with renal function monitoring, urinary tract examination, arterial blood pressure control [4] and ultrasound evaluation once a year and more rigorously in patients with complex SK who exhibit a higher risk of developing urinary tract infections and chronic renal failure [5]. REFERENCES

[1] Schreuder MF. Life with one kidney. Pediatr Nephrol. 2017 May 29. [Epub ahead of print].

[2] Cardona-Grau D, Kogan BA. Update on Multicystic Dysplastic Kidney. Curr Urol Rep. 2015;16(10):541.

[3] Calaway AC, Whittam B, Szymanski KM, Misseri R, Kaefer M, Rink RC, Karymazn B, Cain MP. Multicystic dysplastic kidney: is an initial voiding cystourethrogram necessary? Can J Urol. 2014;21(5):7510-4.

[4] Mavinkurve-Groothuis AM, van de Kracht F, Westland R, van Wijk JA, Loonen JJ, Schreuder MF. Long-term follow-up of blood pressure and glomerular filtration rate in patients with a solitary functioning kidney: a comparison between Wilms tumor survivors and nephrectomy for other reasons. Pediatr Nephrol. 2016;31(3):435-41.

[5] Westland R, Schreuder MF, Ket JC, van Wijk JA. Unilateral renal agenesis: a systematic review on associated anomalies and renal injury. Nephrol Dial Transplant. 2013;28(7):1844-55.

ABS 91

THE ROLE OF NEONATOLOGIST IN PRE-NATALLY DETECTED NEWBORNS WITH POS-TERIOR URETHRAL VALVES

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INTRODUCTION

Posterior urethral valves (PUVs) are the most common congenital obstructive abnormalities of the genito-urinary tract with an incidence of 1 to 8,000 males born and represent the most common cause of chronic renal failure (CRF) in pediatric age [1-3]. Etiology is still unknown; it appears to be a multifactor gene-mediated embryopathy during the first weeks of intrauterine life [4]. Complications include vesicoureteral reflux (VUR) [5], recurrent urinary tract infections (UTI) and myogenic bladder dysfunction up to CRF by progressive obstruction [6]. PUVs are the most common cause of chronic kidney disease (CKD) and renal transplantation in pediatric age [1].

DIAGNOSIS

The management of this pathology firstly requires proper prenatal diagnosis in the search for oligoidramnios, indicative of severe obstruction [7]. Prenatal diagnosis of fetus with PUV allows medical staff to arrange delivery in the most appropriate way and place. The goal in the followup of these patients is to empty the bladder the best way possible from birth to adulthood in order to limit CRF that leads to renal replacement therapy in about 40% of cases.

After birth, ultrasound scan normally show enlarged bladder and pelvis and ureters dilation (**Fig. 1** and **Fig. 2**). Urinary volume, specific gravity, osmolality must be considered. When we are able to catheterize the bladder, it is no longer a surgical emergency. A voiding cysto-urethrogram (VCUG) must be subsequently performed (**Fig. 3**). After the first month of life a Tc^{99m} MAG-3



Figure 1 (ABS 91). Pelvis and calyces dilation in a 2 months baby with PUV.

scintigraphy could give a better evaluation of renal function.

THERAPY

In the postnatal period, neonatologists must provide an adequate hydration, antibiotic prophylaxis and accurate renal function evaluation through serum creatinine and cystatin C; the latter is a better indicator of renal function than creatinine. Cystatin C level is not affected by maternal values and is maximum at birth [8, 9]. After delivery, the bladder must be catheterized if possible, with 6 FR feeding tube.

Severe forms require transurethral ablation of flap valves by endoscopy, eventually associated with cystostomy or pielostomy [10].

CONCLUSION

PUVs therefore require multidisciplinary care, with a team including a gynecologist, neonatologist,



Figure 2 (ABS 91). Ureter dilation in a 2-month-old baby with PUV.

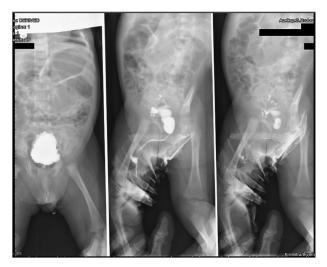


Figure 3 (ABS 91). VCUG: dilated bladder with trabeculation and diverticula; no VUR.

urologist and pediatric nephrologist for prompt diagnosis and therapy to prevent the onset of CKD. REFERENCES

 Diamond DA, Chan IHY, Holland AJA, Kurtz MP, Nelson C, Estrada CR Jr, Bauer S, Tam PKH. Advances in paediatric urology. Lancet. 2017;390(10099):1061-71.

[2] Imaji R, Moon D, Dewan PA. Congenital posterior urethral obstruction.
In: Prem P (Ed.). Newborn Surgery, 2nd ed. London: Arnord, 2003, pp. 856-66.
[3] Krishnan A, de Souza A, Konijeti R, Baskin LS. The anatomy and embryology of posterior urethral valves. J Urol. 2006;175:1214-20.

[4] Elder JS, Shapio E. Posterior urethral valves. In: Ashcraft KW, Holcomb GW, Murphy JP (Eds.). Pediatric Surgery, 4th ed. Philadelphia: Elsevier Saunders, 2005, pp. 781-792.

[5] Parkhouse HF, Barratt TM, Dillon MJ, Duffy PG, Fay J, Ransley PG, Woodhouse CR, Williams DI. Long-term outcome of boys with posterior urethral valves. Br J Urol. 1988;62(1):59-62.

[6] De Gennaro M, Capitanucci ML, Capozza N, Caione P, Mosiello G, Silveri M Detrusor hypocontractility in children with posterior urethral valves arises before puberty. Br J Urol. 1998;81(Suppl 3):81-5.

[7] Reddy A. Nischal Prasad. A clinical study of posterior urethral valve and its impact on renal function. J Evid Based Med Healthc. 2015;2(21):3163-8.

[8] Dharnidharka VR, Kwon C, Stevens G. Serum cystatin C is superior to serum creatinine as a marker of kidney function: a meta-analysis. Am J Kidney Dis. 2002;40:221-6.

[9] Argyri I, Xanthos T, Varsami M, Aroni F, Papalois A, Dontas I, Fanos V, Iacovidou N. The role of novel biomarkers in early diagnosis and prognosis of acute kidney injury in newborns. Am J Perinatol. 2013;30(5):347-52.

 [10] Elder JS. Management of antenatally detected hydronephrosis. In: Prem P (Ed.). Newborn Surgery, 2nd ed. London: Arnord, 2003, pp. 793-808.

ABS 92

BLADDER BOWEL DYSFUNCTION IN CHILDREN: AN UNDERESTIMATED CONDITION?

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The term "bladder bowel dysfunction" (BBD) is a generic term used to describe children and adults with a variety of lower urinary tract symptoms (LUTS) associated with constipation [1], and urinary tract infections (UTIs). BBD is a common problem in the pediatric age. Children with BBD account for about 40% of patients who are visited at pediatric urology clinics and are present in a large proportion of the school population (15-20%). This increasingly common condition is a potential cause of considerable physical and psychosocial weight for children and families. Despite its frequency, this condition is greatly undervalued and therefore

underestimated by families, school staff, and pediatricians.

The association between bladder and intestinal dysfunction can be explained by the common embryonic origin of the bladder and intestine, the position of the pelvis, the same innervation and functional relationship between the pelvic floor and the levator ani muscles.

Normal childhood frequency is between 4 and 7 micturition/day. In preschool age, girls show early sphincter control over males, but are also the most at risk for postponement of empty the bladder. The first healthcare provider who can deal with BBD in children must be the pediatrician, using, according to International Children's Continence Society (ICCS), simple tools like anamnesis, examination (with focus on the lumbar region and the genitals) and first level tools (urine exam, micturition diary). After first evaluation, the pediatrician can decide whether or not the child is to be directed to a pediatric nephro-urology clinic, where the diagnosis will be completed.

Constipation is characterized by alterations of the bowel: reduced weekly frequency of evacuations, episodes of encopresis, painful evacuation, and large feces [3]. Underestimated constipation plays a role in the higher frequency of BBD associated with recurrent UTIs [1].

ICCS recommends applying the Rome III Criteria for the diagnosis of functional evacuation disorders. The diagnosis of functional constipation, with or without encopresis, is fundamentally clinical and is carried out according to Rome III Criteria, different for children under the age of 4 or older.

Bristol Stool Form Scale (BSFS) is also a practical and extremely useful tool for diagnosing and evaluating the response to constipation therapy.

The frequency of evacuations in infants decreases from an average of 4 per day in the first week of life to 1.7 per day at the age of two.

Unfortunately, also constipation in the absence of painful symptoms is underestimated by parents and therefore not reported to pediatricians. Parents and educators are most impressed by the urinary and fecal incontinence episodes. Fecal incontinence is three to six times more common among males than females and has a significant impact on quality of life (QoL), resulting in loss of self-esteem, social isolation and depression [3].

The success of BBD treatment is based on the bladder retraining associated with constipation therapy. In resolved cases, there is also a consequent disappearance of UTIs [4] recurrences.

Urinary and fecal incontinence have a significant impact on the children's QoL. Treatment requires a complete program with bladder re-training, use of macrogol and a proper diet. All of these approaches can be combined to better effect and customize on the individual patient.

Primary prevention of BBD in pediatric age is a key stage in avoiding major diseases in adulthood. REFERENCES

 Masnata G, Manca V, Chia L, Esu F. News on pediatric urology. J Pediatr Neonat Individual Med. 2015;4(2):e040225.

[2] Issenman RM, Filmer RB, Gorski PA. A review of bowel and bladder control development in children: how gastrointestinal and urologic conditions relate to problems in toilet training. Pediatrics. 1999;103(6 Part 2):1346-52.

[3] Abi-Hanna A, Lake AM. Constipation and encopresis in childhood. Pediatr Rev. 1998;19(1):23-31.

[4] Masnata G, Manca V, Chia L, Ledda G, Esu F, Danjou F, Pusceddu P. The role of non-invasive urodynamic assessment in children with LUTS and functional costipation. In: 39th Annual Congress of the Italian Urodynamic Society (Continence, Neuro-Urology, Pelvic Floor), Nicolaus Hotel, Bari, Italy 18-20 June 2015. Neurourol Urodyn. 2015;34(Suppl 2):S1-55.

ABS 93

MILLE GIORNI DI TE E DI ME... MAMMA E FIGLIO (THOUSAND DAYS OF YOU AND ME... MOTHER AND CHILD). THE SCIENTIFIC SCHOOL "TAKE CARE OF CHILDREN: THE MEDITERRANEAN DIET IN THE FIRST 1,000 DAYS OF LIFE. A HEALTHY LIFE" IN THE EVALUATION OF THE PARTICIPANTS

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INTRODUCTION

Good nutrition is essential for good health. Nutritional choices of children and their caregivers are crucial to maintain and enhance it.

How nutritional interventions could be used by Pediatricians and health-care providers to enhance children's health, interacting with families? Do they have updated information?

The University of Cagliari in collaboration with the University of Turin organized a Scientific School named "Take care of children – *Prendersi cura dei bambini*. *Dieta mediterranea nei primi* 1.000 giorni di vita. Una vita in salute" (i.e. "The Mediterranean

diet in the first 1,000 days of life. A healthy life"). For the title of our abstract, we mentioned a song of an Italian singer (Claudio Baglioni). We want to underline the need of 1,000 days in which a team composed by mom, family and other specialized figures "take care of the newborn" during the first phases of his life.

Aim of the meeting was an interactive course that covered every aspect of pediatric nutrition and Mediterranean diet, giving to the participants the ability to apply this knowledge in every-day clinical practice.

SCIENTIFIC SUMMER SCHOOL 2017

As future Pediatricians, we could not miss this opportunity. The course took place in Pula at "Sardegna Ricerche, Parco Scientifico e Tecnologico della Sardegna".

In four days, Pediatricians and Nutritionists from Cagliari and Turin met with different nutritionrelated topics. Through interactive lessons, themes as Mediterranean diet, features of oil and milk and breastfeeding, that we thought to know in a satisfying way, have been redefined making clear their nature. The Scientific School did not stop to nutrition key topics, dealing also with more recent concepts (microbiomics, stem cells and perinatal programming) and new science and their innovative techniques (metabolomics), confirming the importance of nutrition and presenting these methods as potential therapeutic and predictive tools.

The strengths of the Scientific School "Take Care of Children" have been:

- caliber of the involved teachers;
- small participants' number, which created optimal learning conditions;
- a suitable content to establish a scientific debate;
- relevant and updated topics.

For Pediatricians, having the tools for a correct evaluation of food quality, to properly inform and educate the families of our small patients, is crucial. Currently, it is desirable that professional reference figures are formed and updated on nutrition, in order to explain these topics in a clear and exhaustive form, going beyond the wrong widespread beliefs and gaining parents' trust.

The Sardinian geographic area provides excellent and competitive products which represent the basis of Mediterranean diet. During the Scientific School we had the opportunity to visit the milk production plant of Arborea dairy products and taste local extra-virgin olive oil. The idea of promoting these activities came from the need to know the Sardinian land potential to valorize its products and the quality of the foods that we eat and "prescribe". Yes, we prescribe. Nothing new. Hippocrates already exhorted us to consider food as our medicine. Today we know, and daily learn, that a healthy nutrition can contribute to prevent many multifactorial diseases, a topic deeply explained during the Summer School lessons. The future research will help us to fully understand these interactions, to reach the possibility of an individualized, personbased, tailor-made, optimal diet.

CONCLUSIONS

The Scientific School allowed the participants to deepen their knowledge about conscious nutrition and the novelties of scientific research about nutrition in the first 1,000 days of life. The meeting with schoolchildren at the end of the fourday course has been inspirational. So, in the next Scientific School, as future pediatricians, we would like to teach them what we have learned during the course and explain the basis of Mediterranean diet. We hope to repeat this interactive experience and to extend it to other colleagues, in order to further improve the preparation about a fundamental topic of Paediatrics: the nutrition from the maternal womb to early childhood.

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HOMEOPATHY USE IN PEDIATRICS: THE EXPERIENCE OF THE PEDIATRICIANS OF THE ITALIAN FEDERATION OF PAEDIATRIC PHYSICIANS (FIMP)

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INTRODUCTION

The use of Complementary Alternative Medicines (CAMs), and more specifically of homeopathy, is common among the paediatric population in Italy. MATERIAL AND METHODS

In order to verify the attitude of Family Paediatricians towards the use of homeopathy, a survey has been carried out between the approximately 5,500 Family Paediatricians enrolled in the Italian Paediatric Physicians' Federation (FIMP) by means of an online questionnaire created with Google Forms.

The questionnaire was sent in May 2016, with a communication on a mailing list, and remained accessible for a month. The response rate to the questionnaire was 23.19% (1,252 Paediatricians), far above what the literature considers reliable for a study based on data collection across the Web, typically between 6% and 15%. Again, the response has highlighted that 29.4% of the Paediatricians who took part in the survey use homeopathic medicine to cure their patients.

The study has also analyzed several other parameters related to the use of homeopathy, including doctors' age, most commonly treated diseases, pharmaceutical forms and possible adverse effects.

Among the parameters evaluated, the first to reflect is the age of doctors who use CAMs in daily practice: the majority (30%) are between 49 and 67 years (third and fourth decades of practice since graduation), followed by those in the second decade of practice since graduation (25%).

Almost 92.4% of the Family Paediatricians use in an integrated way different therapies with the aim of identifying a therapeutic approach that is appropriate to the diagnosis and at the same time as customized as possible, as the Complementary Medical Study Group (GSMC) of the Italian Association of Paediatrics had underlined in a previous communication on the occasion of the National Congress of 2014.

CONCLUSIONS

The complexity of these data should not be ignored by the Establishment, due to both the number of health professionals who use CAMs and the use of this type of medicines by millions of citizens. REFERENCES

 Lozar Manfreda K, Bosnjak M, Berzelak J, Haas L, Vehovar V. Web Surveys Versus Other Survey Modes: A Meta-Analysis Comparing Response Rates. Int J Market Res. 2008;50(1):79-104.

Trapani G, Zanino L, Careddu D, Gabbanelli G, Gandus S, Annibalini M, Prandi P, Miccichè GM, Calzavara M, Griseri S, Ventura M, Villa I, Sbizzera I, Salvioli GP. Medicine complementari e malattie croniche: risultati preliminari. Comunicazione al 70° Congresso Nazionale SIP – Sessione medicina integrata in Pediatria a cura del Gruppo di Studio sulle Medicine Complementari della SIP (GSMC); Palermo, 13 Giugno 2014.