

MEETING ABSTRACTS

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## MEETING ABSTRACTS

### A1

#### Use of 13-valent pneumococcal conjugate vaccine in children older than 5 years of age

Nicola Principi<sup>\*</sup>, Susanna Esposito  
Pediatric Department of Pathophysiology and Transplantation, Università degli Studi di Milano, Fondazione IRCCS Ca' Granda Ospedale Maggiore Policlinico, Milan, Italy

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The first pneumococcal conjugate vaccine (PCV7) included 7 serotypes and was licensed for the use in infants and in children < 5 years. Recently, preparations containing a greater number of pneumococcal serotypes have been developed and one of them, that including the greatest number of serotypes (PCV13), has been licensed for the use in older children and adolescents. Compared to young children and the elderly, children and adolescents aged 6-17 are at lower risk of pneumococcal disease. Disease incidence falls markedly after the second year and reaches its lowest point in adolescence. Moreover, also carriage of *Streptococcus pneumoniae* (Sp) also declines as children reaches adolescence. Consequently, it is thought that vaccination of older children and adolescents could only marginally reduce the number of Sp disease in these subjects and poorly contribute toward herd immunity for unvaccinated individuals. However, some recent data regarding carrier state of healthy older children and adolescents vaccinated with PCV7 during infancy and living in areas with vaccination coverage lower than 70% seem to suggest that, at least in some particular situations, Sp carriage can be higher than expected and regard most of the serotypes included in the vaccine. This could indicate that mucosal protection offered by PCV7 wanes in time and that after some years, vaccinated children when exposed to serotypes included in the vaccine can be re-colonized with these serotypes and become potential source of infection for unvaccinated subjects. Vaccination with PCV13 could be useful to reduce this risk. However, the most striking evidence of the potential beneficial effect of vaccination with PCV13 of older children and adolescents regards the so called subjects at risk. There is a not marginal number of subjects 6-17 years old with chronic heart, kidney, liver and respiratory disease, diabetes, cochlear implants or cerebrospinal fluid leaks, functional or anatomic asplenia or immunosuppression for whom an increased risk of severe pneumococcal disease is demonstrated or rationally supposed. In these subjects, administration of PCV13 is today strongly recommended, with different schedules according to the previous use of the 23-valent polysaccharide vaccine (PP23). On the other hand, PP23, since long time suggested for protection of subjects at risk >2 years of age, has several problems including an absolute poorer immune response to common serotypes in comparison to PCV13, an hyporesponsiveness to repeated doses, and an uncertain efficacy in risk groups.

### A2

#### A new meningococcal B vaccine

Rocco Russo<sup>1,2</sup>

<sup>1</sup>Maternity and Pediatrics Services, Local Health Units Benevento, Italy;

<sup>2</sup>Maternity and Pediatrics Services – Local Health Units Naples<sup>1</sup>, Italy

E-mail: roccorusso@tin.it

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Neisseria meningitidis is an important cause of invasive bacterial infection in children worldwide, and is a rare example of a bacterium that has evolved to become an obligate human commensal which commonly colonizes the oropharyngeal mucosa. Carriage is age-dependent and appears to be very common in young adults. The relationships between carriage and invasive disease are not completely understood [1].

Each year approximately 1.2 million cases of invasive meningococcal disease (sepsis or meningitis) with 135,000 deaths are estimated to occur worldwide [2].

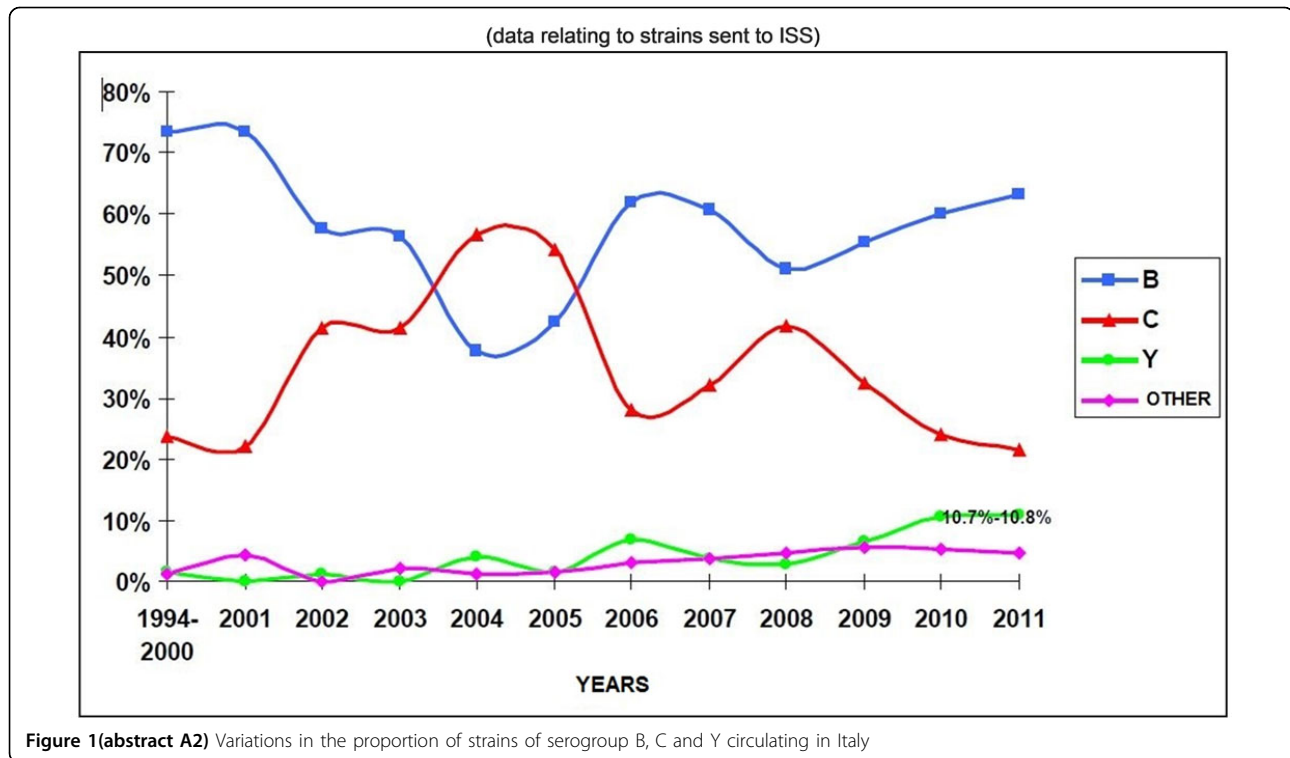
Epidemiology and serogroup distribution differs geographically, as the example in Figure 1 shows, with invasive disease mainly affecting young children, older children and young adults. In addition to age, another individual risk factor includes underlying immune deficiencies; the deficiency of complement components is a known risk factor for invasive infection. Crowding and concurrent upper respiratory tract infections might also contribute to the disease.

In early 2013, a new vaccine developed specifically to prevent disease caused by group B meningococci (MenB) was licensed in Europe (4CMenB, Bexsero<sup>®</sup>, Novartis Vaccines, Italy). This vaccine is protein-based and, therefore, compared to meningococcal conjugate vaccines, has a different mechanism of action, along with different safety, reactogenicity and immunogenicity profiles in the various age groups.

The vaccine, developed by reverse vaccinology, contains three surface-exposed recombinant proteins (fHbp, NadA, and NHBA) and outer membrane vesicles derived from the NZ98/254 strain and has the potential to reduce mortality and morbidity associated with serogroup B meningococci infections, but uncertainty remains about the breadth of protection the vaccine might induce against the diverse serogroup B meningococci strains that cause disease: Meningococcal Antigen Typing System predicted that 78% of all MenB strains would be killed by postvaccination [3].

In Italy (updated April 2014) the Basilicata Region [4] recommend Bexsero for the routine vaccination of infants and will have an active call to parents that includes providing the vaccine free of charge. The Board of Calendario per la Vita, comprising of the country's foremost scientific societies, has recommended Bexsero for all infants with three doses in the first year of life and one dose at 13 months of age [5].

For parents and clinicians, the predicted benefits of 4-component meningococcal group B vaccine (4CMenB) outweigh existing uncertainties



about the potential impact of the MenB vaccine against invasive disease, but future introduction of the vaccine must be followed by rigorous post-implementation surveillance to assess its value to health systems.

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**A3**

**Chickenpox vaccination**

Giovanni Vitali Rosati  
 Florence, Italy

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Varicella is a common viral disease affecting almost the entire birth cohort. Some cases of varicella can be serious, with 2-6% of complications. The hospitalization rate for varicella in Europe ranges from 1.3 to 4.5 per 100,000 population/year and up to 10.1% of hospitalized patients report permanent or possible permanent sequelae.

In countries where routine childhood vaccination against varicella has been implemented, it has had a positive effect on disease prevention and control. Mathematical models indicate that this intervention strategy may provide economic benefits for the individual and society[1]. Despite this evidence and recommendations for varicella vaccination by official bodies such as the World Health Organization and scientific experts in the field, in Italy the last National Plan for Vaccine Prevention 2012-2014 says that vaccination against chickenpox will be included among those active and free offer for all new born Italian from 2015.

Actually only eight Italian Regions undertook universal varicella vaccination programs, in many cases with widespread use of the vaccine MPRV.

The data of Tuscany[2], where the universal vaccination began in the end of 2008 using the quadrivalent vaccine MPRV co-administered with meningococcal C, show a marked reduction of chickenpox in the 2009-2011 period, compared to the prevaccination era (2005-2007). The avoided cases are estimated approximately 42,000 and the hospitalizations are decreased from 3.5 / 100,000 to 2,6 / 100,000. The varicella avoided hospitalizations can be evaluated as 115 in three years.

The reactogenicity data regarding the quadrivalent vaccine measles-mumps-rubella-varicella (MPRV) are discussed because an increased relative risk of febrile seizures after this vaccination, respect to the administration separate and simultaneous MMR and varicella monovalent, is reported.

The Tuscan pharmacovigilance data on MPRV in the last three years 2009-2011, did not indicate any increase of the risk of adverse events compared to the administration of the separate two preparations.

For these reasons, although a note of the Ministry of Health in 2012 invited to use vaccination separate MMR + V in the first dose, Tuscany decided to continue to use MPRV vaccine also in the first dose.

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#### A4

##### Tuberculosis in children

Susanna Esposito<sup>1</sup>, Nicola Principi  
Pediatric Highly Intensive Care Unit, Department of Pathophysiology and Transplantation, Università degli Studi di Milano, Fondazione IRCCS Ca' Granda Ospedale Maggiore Policlinico, Milan, Italy  
*Italian Journal of Pediatrics* 2014, **40(Suppl 1):A4**

The impact of tuberculosis (TB) worldwide remains a serious concern with an estimated 8.7 million new cases (13% co-infected with HIV) and 1.4 million deaths due to TB (430,000 in HIV-infected individuals) in 2011. Assessing the impact of TB in children is particularly challenging since there is no universal diagnostic algorithm. The identification of TB cases in children usually results from a combination of clinical criteria and a non-specific TB test. In 2011, an estimated 490,000 TB cases occurred among children (about 6% of the all cases). Each year, 64,000 children die from TB, making it one of the top ten causes of childhood death.

The global burden of childhood TB is under-reported due to paucibacillary disease which makes diagnosis by sputum smear microscopy and culture difficult. In 2007, the World Health Organization showed that smear-positive TB in children accounted for 0.6-3.6% of reported cases. These data underestimate the true burden of pediatric TB since incidence is estimated using smear-positive cases. The majority of cases in children less than 12 years of age are smear-negative, and smears are seldom performed in high-burden countries. In low-burden countries, childhood TB constitutes about 5% of TB cases compared to the 20-40% in high-burden countries.

Multidrug-resistant TB (MDR-TB) is another important issue for childhood TB that affects global TB control. The global estimate of pediatric MDR-TB is around 40,000 cases per year. Almost 60% of these cases were in India, China, and Russia. The drugs recommended for these cases are off-label for children. Thus, MDR-TB is also a problem for children in close contact with adults who have MDR-TB.

Despite undeniable advances in identifying markers of TB in recent years, many problems pediatricians face in managing TB remain unsolved. The most important difficulty lies in early diagnosis because treatment can completely cure the majority of cases where TB is suspected early. Achieving a cure is more difficult when treatment is delayed and when MDR pathogens are the cause of the disease. In these cases, prognosis is poor, particularly in children, because what can be done to treat MDR-TB is unclear. New studies of diagnostic tests and optimal treatment for children are urgently needed with the final goal of developing an effective anti-TB vaccine. In the meantime, an aggressive attitude must be adopted for both diagnosing and treating a child with suspected TB because TB can be a devastating disease for children.

#### A5

##### Healthcare associated pathogens in a changing world

Caterina Mammìna<sup>1</sup>, Daniela Maria Geraci, Laura Saporito, Giorgio Graziano, Marco Scognamiglio, Celestino Bonura, Mario Giuffrè  
Department of Sciences for Health Promotion and Mother-Child Care "G. D'Alessandro", University of Palermo, Palermo, Italy  
E-mail: caterina.mammìna@unipa.it  
*Italian Journal of Pediatrics* 2014, **40(Suppl 1):A5**

In developed countries about 10% of the hospitalizations are complicated by a healthcare-associated infection [1]. Up to 75% of these infections are due to multidrug-resistant organisms (MDROs) [1]. Antimicrobial resistant bacterial infections are associated to higher morbidity, mortality and healthcare costs than those caused by susceptible organisms [1]. The findings of the point prevalence survey in European acute care hospitals published in 2013 by the European Centre for Disease Control and Prevention (ECDC) show large variations between countries and between different regions of the same country, with Italy being allocated within the high-endemic areas for both MRSA and MDROs [2].

Despite antimicrobial resistance affects most bacterial species, MDR Gram negatives represent the most serious threat. In a few years *Enterobacteriaceae*, mainly *Escherichia coli* and *Klebsiella pneumoniae*, have evolved from extended spectrum  $\beta$ -lactamase (ESBL) producing to carbapenem-resistant organisms [3]. Simultaneously, *Acinetobacter baumannii* has quickly become extremely or pan-drug resistant [4]. Carbapenem resistant Gram negatives heavily impact on clinical outcomes with mortality rates significantly higher than the susceptible strains of the same species [1]. Of further concern, very few antimicrobial agents are available for an effective treatment of these infections and new agents active against these organisms are not currently in development.

Many intertwining factors are driving these epidemiological changes, involving patients, healthcare delivery systems, infection control practices and, most important, misuse and inappropriate use of antibiotics in all healthcare facilities, in community and in animal husbandry. In particular, the transition of the healthcare delivery systems from a hospital-centered model to a healthcare facility network has gradually blurred the borders between hospital and community and the patients' travel within this network has critically contributed to disseminate MDROs [5]. As a consequence, antimicrobial resistance is now as common, if not more so, in post-acute clinical facilities, such as long term care settings and nursing homes [5]. The "revolving door" is the very efficacious image used as the paradigm of the spreading routes of organisms with hospital and community reservoirs, as *E. coli* or MRSA. The revolving door, indeed, enlightens how the colonized patients entering back and forth several healthcare settings drive the amplification of the antibiotic resistance [6]. Stringent infection control and prevention practices and wise use of antibiotics are unanimously agreed as the key actions to fight MDROs. Of course, we need new antibiotics, but first we have to learn how to protect them from a precipitous erosion of their effectiveness.

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#### A6

##### Body composition measurements

Angelo Pietrobella<sup>1,2</sup>, Paolo Brambilla<sup>3\*</sup>

<sup>1</sup>Pediatric Unit, Verona University Medical School, Verona, Italy; <sup>2</sup>Pennington Biomedical Research Center, Baton Rouge, LA, USA; <sup>3</sup>Azienda Sanitaria Locale (ASL) Milano 2, Melegnano, Milan, Italy  
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Accurate assessment of body composition is important in obesity and in many areas of nutrition-related research. The foundation of the development of body composition analysis has been the two-compartment model, dividing body mass into fat mass (FM) and fat-free mass (FFM). A range of techniques are available. Some of them are producing very accurate data, so called "gold standard" or reference methods, have disadvantages of cost, limited availability. Simpler techniques are well tolerated, portable and therefore can be employed in the clinic, patient's bedside or in the community, though may be less accurate. Densitometry determines body density by measuring weight after whole-body immersion in water. It has been crucial as a reference technique in development of body composition studies. Computed Tomography (CT) and Magnetic

Resonance Imaging (MRI) provide anatomical detail and can be used to assess skeletal muscle volume and measure the intra-abdominal fat depot. Dual energy X-ray Absorptiometry (DXA) measures the relative attenuation of two different energy X rays by the body. It derives a three-compartment model of body composition, FM, lean and bone mineral. Lean tissue measured by DXA includes body water, and changes in hydration will be reflected by DXA as change in lean tissue. Anthropometry is a simple technique. Skinfold thickness measurement allows estimation of body fat content. Body density and, thus, subcutaneous body fat can be estimated from the sum of skinfold thicknesses. Waist circumference-to-height ratio is now the most preferred anthropometric index due to its ability to estimate both total adiposity and fat distribution. Weight-stature indices provide surrogate measures for body composition measurements. The most widely used weight-stature index is Body Mass Index (BMI,  $\text{weight/height}^2$ ,  $\text{kg/m}^2$ ). BMI is a global index of nutritional status, but its relation with body composition *per se* is controversial. BMI is a poor predictor for individual subject in clinical setting, because is not able to disentangle FM Vs FFM. Bioimpedance analysis (BIA) is a potential field and clinical method for evaluating %fat and skeletal muscle mass (SM). It offers the advantages of portability, compactness, economy, and ease of operation. Total body water in turn can be used to derive fat and fat free mass. In conclusion, among several possible applications, we want to underline the validity of waist circumference and BIA in the day by day clinical practice as clear, simple and useful measurements of pediatric body composition.

## A7

### Lung sonography

Paolo Tomà

Department of Imaging, Bambino Gesù Children's Hospital, IRCCS, Piazza S.

Onofrio, 00165 Rome, Italy

E-mail: paolo.toma@opbg.net

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New extensive use of thoracic ultrasound (TUS) takes information also from physical acoustic phenomena that are not directly convertible into images of the human body [1]. This tendency also takes into account the classic patterns (based on the presence of an adequate acoustic window) emphasizing the role of TUS as an all-in-one approach in many conditions. We stated our perplexities [2] and we maintain that great caution is warranted when this procedure is used. The evidences on neonatology and paediatrics are based on few articles with different biases i.e.:

No evaluation on technical issues feasibly (the generation of artefacts is conditioned by the time gain compensation setting, the ratio of probe curvature to the curvature of the lung surface, and whether or not movement artefact suppression and tissue harmonics are used), no comparison of the results of TUS and those of a reference standard (i.e. CXR); Diagnostic access bias (few cases, small hospital), bias from reader/training experience on CXR (no paediatric radiologists);

Imaging analysis bias i.e. absence of preliminary definition of the methods for imaging (CXR) interpretation, selection bias (patients from birth to adult undergoing chest radiography for suspected community acquired pneumonia).

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## A8

### Skeletal dysplasias: approach to the clinical diagnosis and implication of appropriate diagnosis for knowledge and research studies in these rare diseases. Hereditary multiple Osteochondromas as example/paradigm

Maria Gnoli, Elena Pedrini, Marina Mordenti, Morena Tremosini, Luca Sangiorgi\*  
Department of Medical Genetics and Skeletal Rare Diseases, Rizzoli  
Orthopaedic Institute, Bologna, Italy

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The skeletal dysplasias are a large, heterogeneous group of genetic conditions characterized by abnormal development, growth and maintenance of the elements (bones) comprised in the human skeleton [1]. In the 2010 revision of nosology and classification of genetic skeletal disorders, 456 conditions were included and placed in 40 groups defined by molecular, biochemical, and/or radiographic criteria. Of these conditions, 316 were associated with mutations in one or more of 226 different genes [2], and are present in about 5% of children with birth defects [3]. About 100 skeletal dysplasias have prenatal onset [4] with ultrasound findings particularly in the second trimester [5]. The first step for an accurate diagnosis is a detailed clinical-radiographic evaluation [4]. In fact, because of clinical and genetic heterogeneity of these diseases, with partial clinical overlap, diagnosis is difficult with a consequent delay in specific follow-up and management. Clinical and molecular characterization of a large patients series is the first step that leads to an improvement in knowledge about natural history, epidemiology and pathogenesis of this disease. These advancements are promoted by expertise centres where patients can be followed-up by multidisciplinary teams, required for syndromic nature or different skeletal segments involvement in the most of cases. To improve skeletal disease knowledge, often lacking, it is essential to analyze and integrate available clinical and genetic data; to this purpose, the design and development of disease-specific registries [5] is essential, as well as the presence of a Biobank for the collection of biospecimens.

Our experience as Reference Centre for Skeletal Dysplasias led us to activate specific diseases registries, as Multiple Osteochondromas Registry (REM), using an HL7 compliant platform, GePhCARD (Genotype-Phenotype Correlation, Analyses and Research Database) that can encompass clinical, genetic, genealogical and imaging data [6]. This web-application is protected by an authentication system, a relief tool articulated in multilevel access profile for data legal protection and patients' privacy. GePhCARD will be soon interfaced with BIOGEN (Genetic Biobank) and together will contribute to improve diagnosis and clinical and molecular characterization of rare diseases, allowing to collect high quality biological materials of skeletal dysplasias patients [7].

In this presentation we focus on our experience on a specific skeletal disease, Hereditary Multiple Osteochondromas (MO), and demonstrate how a systematic integration of clinical and molecular data is focal to increase the knowledge on MO natural history and epidemiology [8], contributing also to define personalized and appropriate follow-up and to hypothesize research studies.

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## A9

### The management of extremely preterm infants

Fabio Mosca\*, Mariarosa Colnaghi, Monica Fumagalli  
NICU, Fondazione IRCCS Ca' Granda - Ospedale Maggiore Policlinico,  
Università degli Studi di Milano, Milan, Italy

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Extreme prematurity is associated with an increased risk of mortality, morbidities and long-term neurodevelopmental impairment. Optimizing prenatal, perinatal and postnatal care is essential to improve long-term outcome of extremely preterm babies.

Intrauterine growth restriction (IUGR) has been associated with a poorer neurological outcome and prenatal care should aim to an optimal balance between minimizing fetal injury or death versus the risks of iatrogenic preterm delivery. In the clinical phase of IUGR, hemodynamic adaptation occurs with blood flow redistribution preferentially to the brain, the so-called "brain sparing effect". However, controversy remains as to whether "brain-sparing" indicates a higher risk of brain injury or is a protective mechanism [1].

The mechanisms underlying fetal growth restriction, also determine a limitation of lung growth and maturation, thus making the lung more vulnerable to postnatal damage and increasing the risk of bronchopulmonary dysplasia (BPD), the most significant respiratory complication of prematurity [2]. Current evidence suggests that the risk of BPD may be partially related to other antenatal factors as chorioamnionitis (CA), which is a leading cause of very preterm delivery. Whereas CA promotes lung maturation, mediated by prenatal inflammation mechanisms, and thereby decreases the severity of respiratory distress syndrome, it also seems to increase the risk of BPD, making the lung more susceptible to subsequent postnatal insults [3].

Neonatal resuscitation and postnatal management during the first minutes of life also play an important role in determining early and long-term outcome of VLBW. A multifaceted intervention in respiratory care in the delivery room immediately after birth, including a sustained lung inflation procedure and a non invasive starting ventilation, seems to be effective in improving respiratory outcome [4].

Delayed umbilical cord clamping has received increasing attention in the management at birth of preterm infants as it seems to be beneficial and safe, being associated with less delivery room resuscitation interventions, improved haemodynamic stability and decreased rates of intraventricular hemorrhage [5].

Different pathophysiological mechanisms are involved in injuring the premature brain, in particular infection-inflammation, pre- and/or postnatal malnutrition, and abnormalities in systemic and cerebral haemodynamics and oxygen supply. Preventative measures are key to reduce neurological morbidities in an extremely preterm population and research projects are focusing on the possibility to stabilize cerebral oxygenation in the first days of life through the application of cerebral near-infrared spectroscopy oximetry and implementation of clinical treatment guidelines in order to reduce the risk of brain damage [6].

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#### A10

##### In situ simulation in pediatrics

Marco de Luca

Emergency Department, Meyer Children's Hospital, Florence, 50139, Italy

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The increasing speed of medicine updating required a change in the training system: the classic teaching mode "see one-do one" is increasingly called into question for ethical reasons in order to protect patients from possible damages deriving from health personnel education [1]. This concept finds its implication in pediatric patients, where any unnecessary suffering must be avoided: for this reason new methodologies based on simulation have been introduced to prevent this type of problem.

Simulation is a technique that allows replacing or amplifying real experiences with guided experiences, artificially designed, which evoke or replicate substantial aspects of the real world in an entirely interactive situation. The growing interest in this approach is due to three main factors: the development of simulator technologies, the evolution of medical education understood as a set of competences in the field of knowledge, skills and behavior and finally the increasing attention to limit errors in team work.

Simulation in medicine is not only a training tool, but also a way of reducing clinical risk and of improving patient safety.

Pediatric emergencies are an example of how simulation can actually be a tool for health workers to achieve a performance improvement in medical assistance [2]. Pediatric emergency events are clinically rare, high-risk, and frequently involve the simultaneous presence of several subjects: pediatricians, nurses, specialists and the patient's family.

*In situ* simulation is a particular form of simulation, distinct from simulation conducted in a simulation center: it may be defined as, "Simulations that occur in the actual clinical environment and whose participants are on-duty clinical providers during their actual workday." The required materials can either be used specifically for simulation and taken each time to the site of the scenario, or they can be routinely available. *In situ* simulation training offers many advantages: it gives the opportunity to test systems, to reinforce training in authentic ways, and also to identify hazards and deficiencies in the clinical systems, in the environment and in the provider team [3].

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#### A11

##### Metabolomics in the diagnosis of sepsis

Vassilios Fanos<sup>1</sup>, Mauro Stronati<sup>2</sup>, Diego Gazzolo<sup>3</sup>, Giovanni Corsello<sup>4</sup>

<sup>1</sup>Neonatal Intensive Care Unit, Puericulture Institute and Neonatal Section, Azienda Ospedaliera Universitaria, University of Cagliari, Cagliari 09131, Italy;

<sup>2</sup>Neonatal Unit and Neonatal Intensive Care Unit, Maternal-Infant Department, Fondazione IRCCS Policlinico San Matteo, Pavia, Italy;

<sup>3</sup>Department of Maternal, Fetal and Neonatal Health, C. Arrigo Children's Hospital, Spalto Marengo 46, I- 15100 Alessandria, Italy; <sup>4</sup>Operative Unit of Pediatrics and Neonatal Intensive Therapy, Mother and Child Department, University of Palermo, Palermo, Italy

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**Introduction:** Sepsis is an important cause of mortality and morbidity for preterm and hospitalized newborn babies. Today, no single test satisfies the criteria as being the ideal marker for the early diagnosis of neonatal sepsis. Analysis of the entire metabolome is a promising method for determining metabolic variations correlated with sepsis [1-6].

**Metabolomics profiling and sepsis:** Works on metabolomics concerning sepsis conducted on animals and humans of different ages (newborn and adults) have recently been published and are presented in Table 1. In septic patients compared to controls (in plasma and urine) it is possible to observe an increase of metabolites which are part of the oxidative metabolism of fatty acids (such as hydroxybutyrate, acylcarnitines and acetoacetate). Briefly stated, alterations in the glucose metabolism in critical conditions can be seen as a redistribution of glucose consumption from the mitochondrial oxidative phosphorylation to other metabolic pathways, such as the production of lactate and the pentose phosphate pathway. In the study by Fanos et al. [7] a combined approach based on both nuclear magnetic resonance (1)H-NMR) and gas-chromatography-mass spectrometry (GC-MS)

**Table 1(Abstract A11) Metabolomic studies that have analyzed the metabolic profiles of septic patients and of experimental animals (From ref. 6, mod.)**

Author	Population study	Sample	Metabolomic analysis	Metabolite alterations
Fanos et al. 2014	9 septic newborns vs 16 control newborns	Urine	GC-MS 1H NMR	Lactate, glucose, maltose, ribitol, ribonic acid, pseudo-uridine, 2,3,4 trihydroxybutyric acid, 2-ketogluconic acid, 3,4 hydroxybutanoic acid, 3,4,5 trihydroxypentanoic acid <(GC-MS) Acetate, acetone, citrate, creatinine, glycine, lactate, lysine, glucose (1H-NMR)
Mickiewicz et al. 2013	60 septic shock vs 40 SIRS vs 40 control pediatric patients	Serum	1H-NMR	2-hydroxybutyrate, 2-hydroxyisovalerate, lactate, glucose, 2-oxoisocaproate, creatine, creatinine, histidine, and phenylalanine
Schmerler et al. 2012	74 SIRS vs 69 septic vs 16 control human adults	Blood	LC-MS/MS	Acylcarnitines and glycerophosphatidylcholines
Mickiewicz et al. 2014	39 septic shock adult patients vs 20 ICU control patients	Serum	1H-NMR	Isobutyrate, phenylalanine, 2 hydroxyisovalerate, myoinositol, acetylcarnitine, creatine, lactate, valine, arginine, methanol, glucose, glycine
Liu et al. 2010	40 septic vs control rats	Plasma	UPLC-Q-TOF-MS	Hypoxanthine, indoxyl sulfate, glucuronic acid, gluconic acid, proline, uracil, nitrotyrosine, uric acid and trihydroxy cholanoic acid
Lin et al. 2009	40 septic vs 20 control rats	Serum	1H NMR	Lactate, alanine, acetate, acetoacetate, hydroxybutyrate and formate
Izquierdo-García et al. 2011	14 septic vs 14 control rats	Lung tissue, BALF and serum	1H NMR	Alanine, creatine, phosphoethanolamine and myoinositol

techniques was used applied to neonatal infections. The study population included 25 neonates: 9 patients had a diagnosis of sepsis and 16 were healthy controls. This study showed a unique metabolic profile of the patients affected by sepsis compared to non-affected ones with a statistically significant difference between the two groups ( $p = 0.05$ ). Mickiewicz et al [8] examined serum samples collected from 60 patients with septic shock (by Gram- and/or Gram+), 40 patients with SIRS and 40 healthy children by nuclear magnetic resonance spectroscopy spectra. Some of the metabolite concentrations were able to separate between patient groups. The main messages from the published studies are as follows. a) Metabolomics is able to early diagnose the infection (in some cases in preclinical conditions). b) Metabolomics is able to predict the outcome in single individuals and the AUC values are close to 1. c) Metabolomics appears to be a promising and useful instrument also in the diagnosis of sepsis. d) In the next future some easy tools, like urinary dipsticks, with the discriminant metabolites will be available in clinical settings, bedside.

**Conclusions:** Present-day methods and procedures for the diagnosis of systemic neonatal infections are hindered by low sensitivity and long response times. Metabolomics is showing promise of becoming a most effective method, even in neonatology and paediatrics.

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#### A12

##### Immunity, gut microbiota and infection

Susanna Esposito  
 Pediatric Highly Intensive Care Unit, Department of Pathophysiology and Transplantation, Università degli Studi di Milano, Fondazione IRCCS Ca' Granda Ospedale Maggiore Policlinico, Milan, Italy  
*Italian Journal of Pediatrics* 2014, **40**(Suppl 1):A12

Recurrent infections are very common in children and a major challenge for pediatricians. They affect the children's quality of life, cause absences from school and lost parental working days, and repeated medical examinations, hospital admissions as well as antibiotic therapies lead to high costs for society. Given their prevalence and clinical importance, various prevention strategies have been developed.

Some of these strategies have considered that innate and adaptive immunity are strictly related with gut microbiota and interact in the modulation of host's reactivity to infections. Variations in gut microbiota related to age, nutrition or underlying disease may influence immune system defenses against viruses and bacteria. Furthermore, disturbed gut colonization patterns are proposed to be associated with the development of allergic disease. This explains why probiotics represent an important option in the prophylaxis and the management of recurrent infectious diseases as well as allergic diseases. However, the incomplete understanding of what constitutes a healthy gut microbiota that promotes tolerance, remains a challenge. Further understanding of gut microbial functions may pave the way for more effective prevention and treatment strategies.

Another approach used for prevention of recurrent infections is represented by the administration of immunostimulants: i.e. molecules of bacterial or synthetic origin that interact with immunological mechanisms *in vitro* and *in vivo*. Pidotimod (3-L-pyrroglutamyl-L-thiazolidine-4-carboxylic acid) is a synthetic dipeptide which stimulates the phagocytic activity of polymorphonuclear leukocytes, enhances the killing activity of human

alveolar macrophages, increases the cytotoxic activity of natural killer cells, and stimulates cytokine production. It also influences the maturation and activity of dendritic cells by increasing the expression of the key surface markers that help to trigger T cell activation and increased IL-12 production. The majority of studies have shown that the number of infections decreases after pidotimod treatment, but they are affected by some methodological weaknesses. Further studies are urgently needed to confirm the pediatric population that should have the greatest benefit with the administration of immunostimulants.

More studies are needed to identify the most promising probiotic strains and study populations, and to evaluate the mechanisms behind the possible effects of probiotics on OM.

#### A13

##### Bleeding management in pediatric patients

Paola Giordano\*, Giuseppe Lassandro  
Dipartimento di Scienze Biomediche e Oncologia Umana, Clinica Pediatrica  
F. Vecchio, Università degli Studi di Bari "Aldo Moro", Bari, Italy  
E-mail: paola.giordano@uniba.it  
*Italian Journal of Pediatrics* 2014, **40(Suppl 1):A13**

Bleeding has always been an alarming clinical symptom in all human societies, and physicians have had varying degrees of success in diagnosing and treating bleeding patients [1]. Because bleeding is part of the human experience, one of the most challenging tasks for a physician is to discriminate between "normal" and "pathologic" bleeding. Hemorrhages or bleeds may occur in every tissue and organ. Every bleeding symptom may, however, vary greatly in terms of magnitude: for instance bleeds in the subcutaneous tissues may present as small pinpoint lesions (petechiae) or large bruises, and epistaxis may range from some blood-streaked mucus to a massive hemorrhage [2]. Minor bleeding is a broad category encompassing a wide variety of symptoms that are, however, severe enough to interfere with the patient's everyday life. Major bleeding defines those episodes that may cause permanent damage to the patient or threaten his or her life. Major bleeding is defined as bleeding in a critical area (intracranial, intraspinal, intraocular, retroperitoneal, intra-articular or pericardial, or intramuscular with compartment syndrome) resulting in an hemoglobin fall > 2 g/dl or requiring transfusion [3]. Bleeding in a child can be a diagnostic challenge because of the wide range of causes, but making a specific diagnosis is clinically important in order to provide appropriate therapy. Bleeding disorders can be inherited or acquired, and include coagulation factor deficiencies or Von Willebrand diseases, platelet deficiencies and/or dysfunctions, blood vessel alterations [4]. The evaluation of a child presenting with bleeding should include a comprehensive medical and bleeding history, a complete family history, a detailed examination and selected laboratory tests. For immune thrombocytopenia, in newly diagnosed children, intravenous human immunoglobulin or corticosteroids can be used [5]. Individuals with platelet function defects should be managed by qualified specialist and platelet inhibitor medication should be avoided. First line drugs for patients with platelet dysfunction are: antifibrinolytics, desmopressin, platelet transfusions and recombinant factor VIIa [6,7]. Bleeding events still occur in patients with hemophilia despite prophylactic factor replacement. Adequate management depends not only upon the hemophilia subtype and site of bleeding, but also the episode severity, identifiable precipitants (e.g. trauma), the patients' age and current place of residence, adequacy of venous access, presence of inhibitors, previous history of bleeding, and the time of onset in relation to prophylactic therapy. For all diseases other important initial considerations include the applicability of first aid measures (e.g. ice, direct pressure, splinting) and appropriate analgesia [8].

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#### A14

##### The children with microcytosis

Bruno Nobili\*, Sofia MR Matarese, Francesca Rossi  
Department of Woman, Children and General and Specialistic Surgery,  
Second University of Naples, Naples, 80100, Italy  
*Italian Journal of Pediatrics* 2014, **40(Suppl 1):A14**

Microcytosis is decrease of red blood cells (RBCs) size. The RBCs size is measured by the mean corpuscular volume (MCV). In the children, MCV varies by age and sex and so it must be always compared with sex and age-based norms. A MCV less than the 5<sup>th</sup> percentile defines the children microcytosis. The most frequent causes of microcytosis are iron deficiency anaemia (IDA) and haemoglobinopathies [1].

IDA is the most prevalent acquired anemia, due to iron deficiency (ID), resulting from negative iron balance. Three are the commonest causes of ID: low dietary iron intake, malabsorption and blood losses. Children are particularly vulnerable to ID because of their increased iron requirements in the periods of rapid growth. IDA causes delay in cognitive development and poor motor and sensory system functioning. Therefore, it is very important to detect ID at its earliest stage in children and replenish the iron stores by proper supplementation [1].

Hemoglobinopathies constitute a major health problem worldwide with a high carrier frequency particularly in regions where malaria has been endemic.

These disorders are characterized by a clinical and hematological phenotypic heterogeneity. Differentiation between thalassemic and non thalassemic microcytosis has important clinical implications, because each has an entirely different pathogenesis, prognosis, and treatment [1-3].

Differential diagnosis requires hematological marker measurement (RBCs, MCV, RBC distribution width or RDW, which measures RBC size variance. An elevated RDW indicates RBCs of multiple sizes), quantification of HbA2 and HbF, detection of Hb variants by HPLC and valuation of iron status (measurement of ferritin, which reflects iron stores, and transferrin or total iron-binding capacity, which indicates the body's ability to transport iron for use in RBC production) [2,3].

In  $\beta$  thalassemia trait ( $\beta$ TT), the RBC count is generally higher than in IDA patients, whereas MCV value is lower. RDW is increased in anemic patients with respect to the healthy subjects, higher in IDA than  $\beta$ TT. The negative iron balance is a marker of ID, while the increase of HbA2 is a marker of  $\beta$ TT [2,3]. In recent years, the identification of new proteins involved in iron trafficking and regulation have led to the discovery of new forms of hereditary microcytosis, sharing features with the classic IDA. A careful patient history and evaluation of laboratory tests may enable these rare conditions to be distinguished from the more common IDA. Molecular studies allow distinction of the different types, a prerequisite for differentiated therapy [4,5].

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## A15

### Iodine deficiency and its consequences for cognitive and psychomotor development of children

Roberto Gastaldi<sup>1\*</sup>, Monica Muraca<sup>2</sup>, Agnese Beltramo<sup>2</sup>, Elena Poggi<sup>2</sup>  
<sup>1</sup>IRCCS Giannina Gaslini, Genoa, Italy; <sup>2</sup>Pediatric Department University of Genoa, Genoa, Italy

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Dietary iodine intake is required for the production of thyroid hormones. Thyroxine (T4) and triiodothyronine (T3) are essential for the development of the central nervous system, they regulate genes involved in myelination and neuronal/glia cell differentiation, and play an important role in axonal and dendrite growth in synaptogenesis and myelination. Deficiencies in thyroid hormones in pregnant women directly affect brain development of the fetus, resulting in neurological and neurocognitive disorders in infants with defects ranging from decrease in intelligence and lethargy to mental retardation. Thyroid deficiency at different stages of pregnancy affects different brain regions, for example, basal ganglia are affected by early thyroid hormone deficiency and cerebellar and hippocampal development is influenced by late thyroid dysfunction [1]. Therefore the consequences of the brain damage depend upon the timing and severity of the hypothyroxinaemia, when it appearing before or at 12 weeks of pregnancy it may affect cognitive development. The neurological cretinism, a disease characterized by severe neurological lesions without clinical hypothyroidism may be a consequence of severe hypothyroxinaemia in early pregnancy. Congenital hypothyroidism is one of the most common preventable causes of mental retardation in children and describes the deficiency in thyroid hormones as of prenatal onset of severe thyroid dysfunction. Newborn screening and thyroid therapy started within 2 weeks of age can normalize cognitive development. Mild reductions in maternal thyroid hormone levels in early pregnancy are associated with reduced IQ in offspring and prenatal exposure to maternal hypothyroxinaemia increased the risk of expressive language delay and nonverbal cognitive delay in preschool age children [2]. Mild iodine deficiency during pregnancy may lead to hypothyroxinaemia in the mother and/or elevated thyroid-stimulating hormone levels in the foetus, and these conditions have been found to be related to mild and subclinical cognitive and psychomotor deficits in neonates, infants and children [3]. Moderate cognitive impairments have been observed in children from the age of 3 weeks up to 5 years. Low maternal iodine status could be associated with an increased risk of suboptimal scores for verbal IQ at age 8 years and reading accuracy, comprehension and reading scores at age 9 years. Correction of mild-to-moderate iodine deficiency improves cognitive performance in school age children [4]. As iodine deficiency is still the most widespread cause of maternal hypothyroxinaemia, the birth of many children with learning disabilities may already be preventable by advising women to take iodine supplements as soon as pregnancy starts, or earlier if possible.

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## A16

### Iodine deficiency in pregnancy

Alessandra Cassio  
Department of Medical and Surgical Sciences, Pediatric Unit, Program of Pediatric Endocrinology, University of Bologna, Bologna, Italy  
*Italian Journal of Pediatrics* 2014, **40**(Suppl 1):A16

Iodine is an essential micronutrient for thyroid hormone synthesis and thus is essential for the normal progression of pregnancy and for the fetus neuropsychological development. Iodine requirements increase during pregnancy for physiological changes in iodine metabolism (increased renal iodine losses, iodine transfer from maternal circulation to the fetoplacental unit and fetal iodine needs for thyroid hormone production).

Therefore the iodine intake should be increased by about 50% during pregnancy and lactation and the daily dose recommended by the WHO is at least 250  $\mu\text{g}$  / day corresponding to a Urinary Iodine Excretion (UIE) range of 150-249  $\mu\text{g}$  /L. Iodine supplementation is recommended in all pregnant women, even in iodine sufficient countries or in areas where universal salt iodization has been achieved (iodized salt consumed in more than 90% of households). These measures, in fact, ensure the daily iodine intake of 150  $\mu\text{g}$  / day recommended in all women of childbearing age but they are not sufficient to support the increased requirements imposed by pregnancy. Recently, even in developed countries, such as UK, USA, and Australia, moderate iodine deficiency has re-emerged as an important public health concern, likely due to a change in eating habits of the population.

There are recent data that highlight how even mild degrees of iodine deficiency may adversely affect the progression of the pregnancy or the neurocognitive outcome in children. It should be emphasized that national programs of health policy must provide epidemiological surveillance measures to ensure the maintenance of iodine sufficiency over time.

Because the fetal thyroid gland may be particularly sensitive to the inhibitory effect of high iodine concentrations, possible negative effects of maternal iodine supplementation during pregnancy have been hypothesized. The currently available data, however, appear to indicate a relatively low risk of these effects against the benefits derived from the adequate transfer of maternal T4 to the fetus in conditions of iodine sufficiency especially in the first trimester of gestation.

## A17

### Urinary tract infection

Claudio La Scola  
Paediatric Nephrology and Dialysis Unit, Department of Pediatrics, Sant'Orsola-Malpighi University Hospital, Bologna, 40138, Italy  
*Italian Journal of Pediatrics* 2014, **40**(Suppl 1):A17

Urinary Tract Infections (UTIs) are common in every day pediatric practice. During the first 6 years of life, circa 6 to 7% of girls and 2.5% of boys will develop a UTI. UTI fall into two categories: non febrile lower UTI or cystitis and febrile upper UTI or pyelonephritis. The latter is considered by many to be the most common serious bacterial illness in children [1]. If UTI is suspected, urinalysis and urine culture should be performed. Collection methods used in clinical practice can be either invasive or non invasive, yet all carry a risk of contamination by bacteria not present in the bladder. Clean voided methods are preferable as they are quite easy to perform and reliable, while invasive methods should be limited to children in poor general health [2]. In the presence of clinical signs and positive urine analysis, while awaiting the results of antimicrobial sensitivity testing, antibiotic treatment should be started as soon as possible, considering local resistance patterns. The route of administration, either parenterally or orally, depends on the condition of the child and the severity of the infection and does not affect the duration of fever, recurrence of UTI or the incidence of post infectious scars. The need for imaging after a first febrile UTI has long been debated. New insights have led us to consider less aggressive imaging strategies, given the high rate of spontaneous resolution of vesico ureteral reflux with age and the good renal outcome for patients with acquired scarring. Therefore, voiding cystography and renal dimercaptosuccinic acid scintigraphy are not routinely recommended [2-4]. It is known that there are several risk factors for the recurrence of



UTI. Some are not modifiable (age, gender, familiarity), while others (reflux, phimosis, bladder function, voiding habits, constipation and fluid intake) are modifiable thanks to behavioral changes and/or medical treatments. As regards preventive interventions, the most controversial is the use of antibiotic prophylaxis and currently none of the recently published guidelines recommend a routine use [2,4].

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#### A18

##### Urine with stones: nephrocalcinosis and vitamin D

Luisa Santangelo, Mario Giordano\*  
Department of Pediatrics, Unit of Paediatric Nephrology, University Hospital "Policlinico Consorziale - Giovanni XXIII", Bari, Italy  
*Italian Journal of Pediatrics* 2014, **40(Suppl 1)**:A18

**Background:** Nephrocalcinosis (NC) refers to diffuse, fine, renal parenchymal calcification, as assessed by radiology or ultrasonography. In medullary NC, the calcification tends to occur in the area of the renal pyramids. It is typically associated with elevated urinary calcium, phosphate, and oxalate, or it can occur with alkaline urine. Any disorder that can lead to hypercalcemia or hypercalciuria may be implicated. Instead of stone formation, smaller parenchymal calcifications are deposited in the medulla, usually bilateral and symmetric.

Vitamin D hormones play a central role in calcium homeostasis. Tight control of the vitamin D system requires inactivation of its active compound 1,25-dihydroxyvitamin D3 (1,25(OH)2D3) through 24-hydroxylation by means of the enzyme 24-hydroxylase (CYP24A1) and degradation to calcitroic acid [1]. Recently loss-of-function mutations of vitamin D-24 hydroxylase (gene CYP24A1) have been recognized as a cause of hypercalcaemia and NC/nephrolithiasis in infants and adults suffering from Idiopathic Infantile Hypercalcaemia (IIH) [2]. Prevalence and natural history of this condition are still to be defined also in pediatric patients. We investigated the prevalence of CYP24A1 mutations in a cohort of pediatric patients with NC referred to our outpatient service from January 2012 to December 2013.

**Materials and methods:** Nineteen children with NC were investigated, by measuring calcium, phosphate, and vitamin D, and by performing CYP24A1 mutation analysis.

**Results:** All patients were diagnosed with NC after ultrasonographic assessment at a mean age of 3,5 years and were analyzed with the diagnostic flowchart proposed by Ammenti [3] for detecting causes of NC. Sequence analysis of CYP24A1, which encodes 25-hydroxyvitamin D 24-hydroxylase, the key enzyme of 1,25-dihydroxyvitamin D(3) degradation, revealed mutations in three affected children, two were compound heterozygous for their specific mutations and one presented only one mutation in a single allele.

All the affected patients showed hypercalciuria, while only compound heterozygous patients showed chronic hypercalcaemia with depressed parathyroid hormone (PTH).

All the patients had normal renal function, despite high-grade NC, and were treated with potassium citrate to restore normal citraturia. Of note all patients suffered of recurrent urinary tract infections, which disappeared after normalization of calciuria due to potassium citrate supplementation.

**Conclusion:** The results of this study suggest a close relationship between CYP24A1 mutations, and onset of NC in pediatric patients. However further studies are needed to determine the prevalence of

CYP24A1 mutation in NC patients, to define natural history and to develop novel therapeutic approaches.

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#### A19

##### Red urine in children

Carmine Pecoraro\*, Ilaria Luongo  
Unit of Nephrology and Dialysis, Children's Hospital "Santobono" via M. Fiore,6 (80129) Naples, Italy  
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Red Urine (RU) is an uncommon finding in an unselected population of children; its prevalence was reported as 0.13 % based on a retrospective USA review of children. However, RU may signify disorders ranging from benign conditions such as medications and food coloring to more serious conditions such as urinary tract infection, hereditary cystic renal diseases and glomerulonephritis. In the clinical approach and management of these children, it is important to firstly differentiate true gross hematuria from non-gross hematuria, and subsequently dissecting out the different causes of gross hematuria (GH).

To determine the prevalence and the etiology of GH in an Italian general pediatric setting we undertook a prospective study of all patients with GH in pediatric emergency walk-in clinic for consecutive 15 months. Study protocol: all patients who complained of RU were referred to the pediatric nephrologist for complete evaluation until diagnosis was made. Over the 15-month period 82,934 children visited the Emergency Clinic at our Children's Hospital; 85 patients made visit in which they complained of RU (1.1/ 1,000 visits). Sex ratio showed a male preponderance (M 50/F 35); mean age was 7.06 ± 4.6 (range 0.5-16.1 years). Among diagnosis, glomerulopathies accounted for 59.5%: Post infectious acute glomerulonephritis (37), primary (7) and secondary to Schonlein-Henoch purpura (3) IgA nephropathy (10), SLE (1), Alport Syndrome (3). 14/85 patients with GH had UTI ( 12 documented bacterial UTI and 2 with a presuntive adenovirus cystitis); 16.5% had Idiopathic Hypercalciuria (6 out showed microcalculi at ultrasonography); 2 patients suffered from hyperuricosuria; 1 child was affected by haemolytic uremic syndrome, 2 myoglobinuria from rhabdomyolysis and in 1 case no diagnosis was made. Our prospective study indicates that in our country, according to the previous report, the prevalence of GH is relatively uncommon, but the most common causes are represented by hematuric glomerulonephritis (mainly PIAGN and IGAN) in boys, and not by proven (26%) and unproven (23%) UTI mainly in girls; just one child was categorized as "unknown etiology" vs 9% of USA children. Two main reasons may explain such a difference: 1) the health social assurance systems, namely the availability in Italy of the family pediatrician which avoid that children with GH from trivial causes such as hemorrhagic cystitis, will turn to ER and 2) the systematic evaluation of all cases of GH, in our experience, directly by the paediatric nephrologist who could assure a definite diagnosis.

#### A20

##### Cardiovascular risk prevention in children

Francesco Martino  
Department of Pediatrics and Child Neuropsychiatry, Sapienza University of Rome, Italy  
*Italian Journal of Pediatrics* 2014, **40(Suppl 1)**:A20

Cardiovascular disease (CVD) is the major cause of mortality in industrialized countries. Atherosclerosis is characterized by a long lag time between onset and clinical manifestations. There is evidence that

CVD initiate before birth by developmental changes in utero. Genetic and environmental factors may interact in specific periods of life (prenatal, perinatal, early childhood), giving rise to altered developmental plasticity and epigenetic modifications with abnormal phenotypic expression of genetic information without altering DNA sequence. Nutritional imbalances and other environmental clues may cause intrauterine growth delay, decreased gestational age, low birth weight and postnatal catch-up growth. Fetal exposure to maternal hypercholesterolemia has been associated with increased risk and progression of atherosclerosis. Oxidative stress seems to play a major role in the atherosclerotic process by activating both platelets and monocytes forming proinflammatory and proatherogenic substances, resulting in endothelial dysfunction, decreased flow-mediated dilation, and increased carotid intima-media thickness [1,2]. CVD primary prevention should be implemented early. High risk, apparently healthy [3] subjects should be identified, and treated early by healthy eating, physical activity encouragement, healthy lifestyle, the possible addition of nutraceuticals [4] and adding drugs if risk profile is not decreased. Restoration of endothelial function in the reversible phase of atherosclerosis is an essential step. Epigenetics might provide novel and early markers to identify those at risk, thereby developing innovative therapies.

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#### A21

##### Physical activity and kidney diseases

Silvio Maringhini

Pediatric Nephrology Unit. "G. Di Cristina" Children's Hospital. A.R.N.A.S. Palermo. Via Benedettine 2, 90134 Palermo, Italy  
*Italian Journal of Pediatrics* 2014, **40(Suppl 1)**:A21

Regular physical activity (PA) in the early school years is recommended by several scientific associations for primary prevention of cardiovascular disease. Long-term observational studies have shown that subjects who exercise regularly have significantly less coronary heart disease (CHD) and a reduced risk of cardiovascular disease (CVD). Exercise reduces serum triglycerides, increases serum high density lipoprotein-cholesterol, lowers the blood pressure in patients with primary hypertension [1]. Regular exercise reduces the production of atherogenic cytokines and increases production of atheroprotective cytokines. It should be mentioned, however, that other factors may be associated with physical activity (e.g. a healthy diet, avoiding cigarette smoking, regular medical care) and may contribute to the improved health. No long term studies have been produced on the effects of PA during childhood on CHD and CVD in adult life but indirect evidence suggests that it may produce benefit.

Regular exercise, on the other end, is associated with potential adverse effects (eg. musculoskeletal injuries, arrhythmias, myocardial infarction, and rhabdomyolysis); however, the absolute risk of kidney disease during exercise is low [2]. Consensus guidelines for the pre-participation physical evaluation (PPE) suggest a PPE for all children, even those who do not participate in organized sports, as an opportunity to promote health and fitness. PPE includes a medical and family history and a physical examination, with particular emphasis on the musculoskeletal and cardiovascular systems [3].

Exercise induces profound changes in the renal haemodynamics and in electrolyte and protein excretion. Proteinuria, hematuria and changes in serum electrolyte balance have been reported during intense PA; the increase in glomerular filtration may explain these transient alterations but the "nutcracker" compression on the renal vein may have a role. Haemoglobinuria and myoglobinuria may be observed under special exercise conditions [4].

PA does not worsen nor reverse kidney disease but may reduce cardiovascular risk in chronic renal disease [5]. Children on dialysis and after a renal transplantation performing aerobic physical exercise show improvements in exercise tolerance, in quality of health and uraemic symptom scores; they gain weight loss, cardiovascular reactivity, avoiding an increase in blood pressure medication [6].

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#### A22

##### Sedentary behaviour as a risk factor for cardiovascular diseases in pediatric age

Chiara Mamei, Valentina Fabiano, Gian Vincenzo Zuccotti\*  
Department of Pediatrics, L.Sacco Hospital, University of Milan, Italy  
*Italian Journal of Pediatrics* 2014, **40(Suppl 1)**:A22

The term "sedentary" has historically been used to refer to individuals with low physical activity levels or not meeting some criterion level of physical activity. According to the recent findings, sedentary behaviour has been defined not simply as the absence of moderate-to-vigorous physical activity, but as a class of behaviours (e.g. sitting, watching TV or playing video games) characterized by little physical movement and low energy expenditure ( $\leq 1.5$  metabolic equivalents -METs-) [1].

Recent evidences suggest that sedentary is a distinct risk factors for chronic diseases including cardiometabolic diseases [2].

In Western countries sedentary time increased dramatically since 1960s [3]. Schools, homes, and public spaces have been re-engineered minimizing physical activity and contributing to reduce children independent mobility. [4] Moreover the new media (eg. computer and videogames) occupy a high proportion of leisure-time of children and youth.

Sedentary behavior can be assessed using both indirect (questionnaires) and direct measurement tools (accelerometers). These methods assess different aspects of sedentary: while self- and proxy-report questionnaires mainly focused on common sedentary behaviours (eg. watching TV), accelerometers better estimate the individual's total sedentary profile. Therefore in clinical studies the concurrent use of both strategies should be encouraged [5].

In pediatric age, studies have demonstrated that spending excessive time engaging in sedentary behaviors, independent from overall physical activity levels, is adversely associated with adiposity and other cardiometabolic risk factors such as lipid profile, blood pressure and central obesity [5,6]. In particular, the self-reported screen time appears to be more strongly associated with these risk factors than the total sedentary time assessed by accelerometers. One potential explanation could be that certain forms of sedentary activities (TV viewing and more in general screen-based activities) promote the excess of food intake and thus a positive energy balance. Research community should continue to accrue knowledge about the impact of sedentary on health in the pediatric age. Efforts should be done to promote deliberate physical

activity and to decrease sedentary behaviours in order to maximize health benefits since childhood.

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#### A23

##### Which kind and how much physical activity?

Ugo Giordano

Ospedale Bambin Gesù, Roma, Italy

*Italian Journal of Pediatrics* 2014, **40(Suppl 1)**:A23

Physical fitness is important for the optimal development both in children and adolescents. Fitness programs for youth should be recommended and encouraged.

Prior to participation in sport program a detailed medical examination should be performed including an ECG at rest.

It's important that children should be exposed to a wide variety of sporting activities to discover which is the best for them to find out what they like most and to figure out which is best suited to their auxological condition. The rules and duration of physical activities should be appropriate to the age of the participant and excessively long distance (competitive) running events are not recommended for children prior to maturation.

Important is the difference between competitive sport and not competitive, both in terms of efforts as hours per week as involvement in physical stress. Sports activities are necessary also to prevent chronic diseases like obesity, arterial hypertension, cardiovascular disease, hypercholesterolemia, diabetes and anxiety.

Good health or good physical condition is guaranteed by a healthy lifestyle rather than a genetic inheritance. Few but very important are the factors that contribute to a good aging, including physical movement.

#### A24

##### A physical activity program dedicated to adolescents

Maria Cristina Maggio

Universitary Department Pro.S.A.M.I. "G. D'Alessandro", University of Palermo, Italy

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Adolescence is a life stage in which the development of individuality and self-identity occurs.

The recent study of the SIP evaluated the life style in adolescents, highlighting the low participation to continuative sports programs and the high incidence to sports drop out, interesting more than 30% of adolescents. These problems are partially linked to low compliance of adolescents to coaches training, partially to the request of a sport close to adolescents requirements.

However sports participation is beneficial for physical and psychological development of adolescents. Sports programs promote responsible social behaviours and greater academic success, confidence in personal physical abilities, appreciation of personal health and fitness and strong

social bonds with individuals and institutions. Adolescents involved in physical activities fare better academically, have higher relationship skills, are more team-oriented and are healthier as determined by fitness standards.

For these reasons the program of the physical activity dedicated to the adolescent needs a personal evaluation of own desires and personal abilities, with a daily promotion of an active life style, preferring group activities (to go to school and to come back home by foot or by bike with friends, to walk for familial needs, etc.). The sport choice must be carried out together with them, in order to guarantee a four times a week physical activity, in conformity with scholastic needs. Once a week they need to have special physical activities in accordance to familial requests.

The skills of this program are: the education to a maintained physical assignment; the prevention of social deviation and/or alcohol, smoke, drugs dependence; the education to roles and rules, responsibilities, loyalty, cooperation, friendship.

#### A25

##### Recurrent fever: diagnostic procedure

Elisabetta Cortis

Department of Pediatrics - USL 2 - 06034 Orvieto (TR), Italy

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Fever is defined periodic/recurrent in presence of three or more episodes of fever of unknown origin that occur in a period of six months and whose onset is at least one week apart from each other. The diagnosis, often complex, should exclude an infectious cause and an onco-haematological disease.

History and physical examination are essential to make a diagnostic hypothesis of a periodic fever. A recurrence of febrile episodes throughout the year including the summer period, intervals of complete wellbeing, fever associated with symptoms and similar clinical signs, length of episodes and similar intervals, spontaneous resolution of symptoms, would suggest a periodic fever. The inflammatory markers (ESR, CRP, SAA) and the number of WBC increase in the period of fever and tend to normalize with fever resolution [1]. PFAPA syndrome, the most common, is characterized by episodes of fever, that occur under 5 years of age, are accompanied by cervical lymphadenitis, pharyngitis, mouth aphtas; in the febrile phase, the child keeps good general conditions. Steroid therapy has proven effective in the resolution of fever, even if it has been demonstrated that the regular intake of cortisone at any fever onset may lead to a shortening of the intervals. In the differential diagnosis, due consideration should be given to cyclic neutropenia that is a genetic disease (mutation of ELA-2 gene) characterized by fever every 3-4 weeks (it appears with a decrease of neutrophils), pharyngotonsillitis and aphtas larger than those of PFAPA, and often severe bacterial infections. In such cases, a complete blood count performed every week for at least four weeks may be useful to make the proper diagnosis PFAPA does not require further diagnostic investigations and prognosis is benign with spontaneous resolution within 10 years of age. For the monogenic periodic fevers that are FMF, TRAPS and MKD, fever is accompanied by a systemic involvement such as rash, arthritis, serositis (table 1). In the febrile period the child has a general malaise. Ethnicity, age of fever onset, duration of fever vary between one form and another. In order to make easier the identification of a periodic monogenic fever a diagnostic score has been developed that can predict the risk that a pediatric patient may have one of these forms. The score takes into consideration the following variables: age of fever onset, abdominal pain, mouth aphtas, diarrhea, positive family history [2].

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**Table 1(Abstract A25) Main clinical manifestations during fever episodes in patients with periodic fever syndrome and genes responsible of hereditary periodic syndrome.**

	FMF	TRAPS	MKD/ HyperIgD	PFAPA
Abdominal pain	+++	+++	++	-
Diarrhea	+/-	-	+++	-
Arthritis	+++	-	+++	-
Lymphadenopathy	+/-	++	+++	+++
Rash	+/-	++	+++	-
Stomatitis	-	-	-	+++
Pharyngitis	-	-	-	+++
Splenomegaly	++	-	+++	+/-
Conjunctivitis	-	+++	-	-
Duration of fever	1-4 Days	Variable >1 week	2-5 Days	2-8 Days
Gene/chromosome	MEFV16p13.3	TNFRSF1A12p13	MVK 12q24	unknown
Protein	Pyrin	TNFR1	MVK	unknown
Inheritance	AR	AD	AR	none

autoinflammatory diseases: the Eurofever experience. *Ann Rheum Dis* 2012, **71**:1177-82.

#### A26

##### Biological therapies in monogenic autoinflammatory diseases: long-term efficacy and safety

Maria A Pelagatti\*, Alessandro Cattoni, Carmelo Rizzari  
 Department of Pediatrics, University of Milano-Bicocca, San Gerardo Hospital, 20900 Monza (MB), Italy  
 E-mail: marettafel@yahoo.it  
*Italian Journal of Pediatrics* 2014, **40(Suppl 1):A26**

**Introduction:** Autoinflammatory diseases (AIDs) represent a group of monogenic disorders, related to mutations in genes encoding proteins involved in the regulation of inflammatory responses. The aims of therapy in AIDs patients are controlling disease activity, improving patient's quality of life and preventing long-term complications. Biological agents have recently changed the natural history and the prognosis of AIDs. According to a pathogenetic criterium, they can be classified as:

- 1) Anti-TNF agents: i) Etanercept, a dimeric fusion protein of TNFR2 linked with the Fc region of human IgG1, ii) Adalimumab, a fully human monoclonal anti-TNF antibody, iii) Infliximab, a mouse/human chimeric monoclonal anti-TNF antibody [1,2].
- 2) Anti-IL-1 agents: i) Anakinra, a competitive IL-1 receptor antagonist, ii) Canakinumab, a fully humanized anti-IL-1 monoclonal antibody, iii) Riloncept, a fusion protein of IL-1 extracellular domains and Fc region of human IgG [1].
- 3) Anti-IL-6 agents: Tocilizumab, a monoclonal antibody binding and inhibiting both soluble and membrane IL-6 receptor [1,2].

**Clinical use of biological drugs in autoinflammatory disorders:** → **Familial Mediterranean Fever (FMF)** is the only autoinflammatory disease in which a "classical" immunomodulator (colchicine) still plays a central role [3]. Nonetheless, a relevant number of non complete responders has been recently observed. According to the pivotal role of IL-1 in the pathogenesis of FMF [4,5], anti-IL-1 treatment has been employed in colchicine-resistant patients, with good response in terms of control of disease activity and systemic impairment [6,7].

→ In **TRAPS** (TNF Receptor Associated Periodic Fever Syndrome) patients, the failure of Infliximab and Adalimumab in controlling inflammatory flares has been already described [8]. Etanercept does not completely normalize symptoms or acute phase reactants [9] and the progressive development of neutralizing antibodies may compromise its effectiveness [2]. On the other hand, recent papers have shown the dramatic and persistent efficacy of anti-IL-1 treatment in TRAPS patients [10,11].

→ In **CAPS** (Cryopirin Associated Periodic Syndromes) patients treated with Anakinra, the clinical improvement appears to be strictly dependent on continuous drug administration [12,13]. Moreover, preliminary results on Riloncept and Canakinumab seem to be promising [14-16].

→ In **Hyper-IgD** (Mevalonate Kinase Deficiency), Anakinra has shown to be markedly effective in different case reports, while Etanercept provided variable results [2].

**Conclusions:** Although biologic agents are related to frequent side effects, their administration has revolutionized the clinical approach to AIDs patients. Further trials are needed to definitively assess their safety and efficacy.

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## A27

### The use of ultrasonography (US) in non traumatic musculo-skeletal disorders in infants and children

Laura Tanturri de Horatio  
Department of Imaging, Bambino Gesù Children's Hospital, IRCCS,  
Rome, Italy

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Ultrasonography (US) is a powerful diagnostic imaging tool for evaluating musculo-skeletal disorders in children. US is relatively cheap, non invasive and does not require sedation thus it is generally well-tolerated. Lack of ionizing radiation and dynamic imaging capabilities are significant advantages compared to CT and MRI too.

US is ideally suited to the evaluation of the bone, cartilage and soft-tissue structures, including tendons and ligaments, joints and muscles [1].

The combined use of new generation high frequency transducers and of Power Doppler increases the diagnostic accuracy of US.

US is widely used in the diagnosis of hip disorders in children [2]. In developmental dysplasia, it is essential for establishing an early diagnosis in order to allow a prompt treatment. In transient synovitis and hip infections US is able to demonstrate joint effusion, synovial thickening and cartilage damage. In Perthes disease US is a reliable technique to identify hip effusion, femoral head cartilage thickening as well as irregular, fragmented and flattened femoral epiphysis. In epiphysiolysis US allows the assessment of the severity of epiphyseal slipping by measuring the width of the physeal step [1].

Besides hip's disorders, US can be employed in many other pathological conditions of the pediatric musculoskeletal system, including congenital, infectious, neoplastic and inflammatory disorders [3]. Particularly, in juvenile idiopathic arthritis (JIA) US is crucial for the diagnosis of the disease, the assessment of its severity and prognosis, the monitoring of disease progression and treatment response and the evaluation of complications related to the disease or its therapy. The US assessment of disease activity has been proven to be more informative than clinical examination. Moreover, multiple locations can be assessed during the same ultrasonographic session. US can guide intra-articular steroid injections for therapeutic purposes too.

In acute osteomyelitis US is able to identify early abnormalities in the soft tissues overlying the bone just few days after the onset of symptoms before the appearance of radiographic signs. As infection progresses, US can depict interruption of the cortical profile related to bone destruction and diffuse involvement of subcutaneous tissue with formation of abscesses.

Due to the peculiarities of the growing skeleton, the knowledge of musculoskeletal pediatric anatomy and especially of growth-related changes in healthy children is extremely important in establishing whether the US findings reflect pathology or are part of normal development [4].

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## A28

### Ultrasound in children and adolescents with inflammatory bowel disease

Francesco La Seta<sup>1\*</sup>, Michele Citrano<sup>2</sup>, Lorenzo Tesè<sup>1</sup>, Lucrezia Bruno<sup>3</sup>  
<sup>1</sup>Dept. of Radiology, 'V. Cervello' Hospital - A.O.R. 'Villa Sofia - Cervello', Palermo, Italy; <sup>2</sup>Dept. of Pediatrics, 'V. Cervello' Hospital - A.O.R. 'Villa Sofia - Cervello', Palermo, Italy; <sup>3</sup>Specialty in Pediatrics, University of Palermo, Italy  
E-mail: flaset@tin.it

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The term *Inflammatory Bowel Disease (IBD)* is used to describe several idiopathic gastrointestinal disorders; among these diseases, the most commonly encountered in clinical practice are Crohn's Disease (CD) and Ulcerative Colitis (UC).

CD and UC are different both in distribution of gastrointestinal tract involvement and depth of inflammation. 25% of patients reveals IBD during childhood or adolescence [1].

No consensus exists regarding the optimal techniques and imaging modalities when evaluating IBD, especially in the paediatric age. [2] The correct imaging choice often depends on clinical presentation of patients. The most common situations are: the initial diagnosis of suspected IBD; patients with known IBD presenting with acute symptoms; follow-up of patients with or without new symptoms [2].

Multiple methods can be used to diagnose and examine paediatric patients with IBD, including Computed Tomography (CT), Magnetic Resonance (MR), Small Bowel Follow-through radiographic examination (SBFT), Endoscopy, and Ultrasound (US) [3,4].

While UC is mainly clinically managed with endoscopy and US, CD very often needs cross-sectional imaging to assess the exact distribution and activity of the disease, and to detect extraluminal complications (phlegmon, abscess, fistula, bowel perforation).

Due to early onset of disease, paediatric patients obviously need additional attention to minimize radiation exposure: it is mandatory to avoid unnecessary ionizing radiation techniques in IBD imaging [2-4].

Consequence of the foregoing statements, US and (more recently) MR-enterography have become the preferred imaging modalities in management of young patients with IBD [2-6].

In clinical practice, in Europe, US is currently the first examination requested to evaluate patients with abdominal symptoms. US is accurate in diagnosis of several paediatric (intestinal and non-intestinal) diseases; US may also help when selecting patients needing additional caring among children suffering from gastroenteric tract diseases [5]. Furthermore, US may offer information that are difficult to obtain with other diagnostic modalities, and in fact it is the only technique which could possibly investigate in real time all the followings: the morphological and functional aspects of the bowel wall, the vascularisation (with color-Doppler) of thickened bowel, the stiffness of intestinal loops, and perivisceral spaces.[5].

The aim of this review is to elucidate the current role of US in diagnosis and follow-up of paediatric patients with UC and CD, and to discuss the advantages and disadvantages of this radiation-free imaging methods in

terms of cost, safe, diagnostic accuracy and accessibility compared with other diagnostic modalities.

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#### A29

##### Ultrasonography in pediatric inflammatory bowel diseases

Mauro Massimetti<sup>1</sup>, Francesca Mangianti, Rino Agostiniani  
Pediatric Department, S. Jacopo Hospital, Pistoia 51100, Italy  
*Italian Journal of Pediatrics* 2014, **40**(Suppl 1):A29

Inflammatory bowel diseases (IBD) are a heterogeneous group of chronic disorders of intestinal inflammation characterised by periods of remission and exacerbation. Crohn's disease (CD) and ulcerative colitis (UC) are the two major clinical subtypes of IBD [1]. CD is characterised by transmural inflammation in a non contiguous pattern anywhere from the mouth to the anus. Ileocolonic region is the most common location of disease in pediatric CD. Classically, UC involves disease that extends proximally for a variable distance from the rectum, with involvement of the superficial layers of the colonic mucosa. Pancolitis is the most frequent presentation of UC in childhood. Disease courses are different not only in childhood from adult life but also in the different ages of pediatric patients [2]. Definitive diagnosis of IBD relies on endoscopic and histologic findings often supported by radiologic imaging. Ultrasound scanning as innocuous and ubiquitous imaging technique can be used both as screening diagnostic tool in patients with suspected IBD than in the clinical management of patients with proven IBD in the effort of detect extension, grade of activity and early individuation of complications in the follow-up. Ultrasound can be performed as a standard examination without preparation called transabdominal ultrasonography (TUS) or associated with previous ingestion of an oral contrast solution that produce an osmotic fluid distension of intestinal lumen for a more sensitive and detailed valuation of sonographic aspects of the bowel wall called small intestine contrast ultrasonography (SICUS). In Crohn disease recent data in literature show that SICUS improves sensitivity in detecting small bowel lesions both in previous undiagnosed patients from 75% to 100% than in patients with proven CD from 76% to 100% compared to TUS [3]. The execution of ultrasound in the evaluation of pediatric intestinal tract requires time, good technique and experience of the sonographer. The main goal of this presentation is to show the most important technical aspects of the execution of ultrasound examination in pediatric IBD.

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#### A30

##### Giorgio Maggioni master of pediatrics and the SIP's group of History of Pediatrics

Maria G Gregorio<sup>1\*</sup>, Francesco S Biagiarelli<sup>2</sup>, Luigi Cataldi<sup>3</sup>  
<sup>1</sup>UOCC ASL 8, Cagliari, 09100, Italy; <sup>2</sup>La Sapienza University, Rome, 00100, Italy; <sup>3</sup>Dept. of Mother and Child, Division of Paediatrics, Catholic University of the Sacred Heart, Rome, 00168, Italy  
E-mail: giusi.gregorio@tiscali.it  
*Italian Journal of Pediatrics* 2014, **40**(Suppl 1):A30

It is limited for us, the old group of History of Pediatrics (Figure 1) (founded in 1998 in Turin thanks to Carlo Montinaro's tenacity), remember the birth, death, prizes and scientific publications of prof. Giorgio Maggioni.

Is not a coincidence that the poster we presented at SIP's congress in Palermo, has a title and different subtitles. This is because on the one hand we like to remember the prof. Maggioni's figure, recently deceased (1/5/2014), and there you will find a short curriculum, and on the other hand we want to highlight the different sides of his personality.

Although it is difficult to summarize in 400 words the Giorgio Maggioni's scientific and cultural heritage, authors refer particularly to the Master's commitment since 60th SIP's congress in Turin (1998) in supporting our "newborn" group of History of Pediatrics.

Giorgio Maggioni had never been in the executive committee board, but he was the soul, the historical memory(1), the Honorary President, the Example.

"We must remind... people exist only in the memory", he said. And consequently, the interest, attachment for books so much to imagine the Paradise, as a big library with Mozart's music in the background, books that kept him company and that he preferred on the television. We will always remember his intimate and tender side, in the role of father, example of intellectual honesty, curiosity for knowledge, but also liberal in letting choose the right way to each son, according to personal interests, or loving grandpa, that accepting grandchildren on his knees, invited them not to waste time, and to protect neurons from the decay.

And still "What you know no one can ever take away, remember that!"; "Knowledge is power" he added. He was a very tenderhearted person, always ready to hear you and give good advices, he never judged you. As Francesco recalls, his grandson pediatrician: the day of my graduation, he, seated and serious...but happy said: "I could not miss this historic event!" He said: "I wish you to be loved by your sons as I feel loved by mine". In his memory, we want underline his clarity, childlike curiosity like he saw the world, medicine and history.

At last he always said "I hope to see you soon".

"Goodbye Master"

#### A31

##### Humanization in the management of hospitalized children and adolescents

Gianluigi Gargantini  
Department of Pediatrics, Lodi, 26900, Italy  
*Italian Journal of Pediatrics* 2014, **40**(Suppl 1):A31

The rights of infants and children in hospital, which are universally accepted from an ideological point of view, result as being only partially applied and in a non-homogenous way in the different realities [1-3]. Many countries have set standards which would allow the translation of these principles into clinical practice. In 2008 the Italian Pediatric Society (SIP) and the Foundation for Children in Hospital, with the support of Progea & Joint Commission International, drafted a specific manual in which the requirements to be satisfied are clearly expressed and in which they also started up a voluntary accreditation programme of paediatric departments. The humanization of hospitals is a continually evolving process in relation to the changing needs of children and to the context in general. The current reference model is the Patient and Family Centered Care (PFCC) [4]. Intervention is required at all levels.

Training: this represents the foundation of the process and should examine the numerous aspects such as communication, management of relationships, cultural competence and teamwork.

Organization: activation of alternative assistance models, or models integrated to hospital admission; admission into spaces which are



**Figure 1(abtract A30)** Group of History of Pediatrics, from left to right, Cataldi; Maggioni; Paladini; Gregorio; Fasani (26/2/2011).

separated from those of adults (it occurs in 72.2% of subjects between 0-17 years of age and in 16% of those between 15 and 17); dedicated paths and guaranteed paediatric treatment in all areas of assistance, including the emergency area; family centered round care which represents the hospital paradigm of the PFCC concept in which the multidisciplinary discussion is carried out within the room, in the presence of the child and the family, integrating their outlook in the decision process; organizational flexibility and in the procedures [5].  
 Structure: the wards should be designed with the concept of a "paediatric hospital" within a general hospital where minors are cared for in

dedicated areas and with sufficient flexibility to respond to the various needs. The optimal number of single rooms is a subject of debate. For adolescents, separated areas which safeguard privacy and independence are essential. Spaces for support activities and family members' comfort must be provided for (table 1).

Information and education: aimed at actively involving the children and the families in the decisions and in the treatment, but also at understanding one's rights.

It is fundamental to have a strong local leadership that drives the change with group work, alliances with institutions and involving children and families. Validated measures of process and outcome are necessary.

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**Table 1(Abtract A32) SIP SURVEY OF 237 PEDIATRIC WARDS – 2010**

ASPECTS OF HOSPITALITY	%
NO SINGLE ROOM	23
ADOLESCENT HOSPITALIZATION AREA	26
GAMES ROOM	91
ADOLESCENT ENTERTAINMENT AREA	23
SCHOOL	53
VOLUNTEER ASSOCIATION	61
AVAILABILITY OF A CULTURAL MEDIATOR	74
TRANSLATION TELEPHONE SERVICE	19

### A32

#### Child abuse and neglect: psychiatric and neuro-biological consequences

Pietro Ferrara

Institute of Pediatrics, Catholic University of Sacred Heart, Rome, Italy and

Campus Bio-Medico University, Rome, Italy

E-mail: pferrara@rm.unicatt.it

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Child abuse and neglect is a common problem that is potentially damaging to long-term physical and psychological health of children. Over the past, researchers have documented this relationship and have identified two possible mechanisms that can explain the increased incidence of childhood stress and consequent adult somatic disease: the increased incidence of health harming behaviors and causes epigenetic and other changes that predispose individuals to disease through a raised non-specific inflammatory profile [1]. Abuse survivors, as well as persons who have experienced other types of childhood adversities, are more likely to participate in high-risk behaviors [2]. Possible etiologic factors in survivors' health problems include abuse-related alterations in brain functioning that can increase vulnerability to stress and decrease immune function. Adult survivors are also more likely to participate in risky behaviors that undermine health or to have cognitions and beliefs that amplify health problems [2].

Childhood abuse and early life stress may become hard-coded into the genome, creating an epigenetic memory of events that leads to impaired health at a later date [3].

Chronic early life stress results in long-term changes in HPA (hypothalamic-pituitary-adrenal) axis function and regulation typified by hypersecretion of CRH and ACTH. The initial hypersecretion of cortisol may over time lead to blunting of the cortisol response to CRH and ACTH and relative glucocorticoid resistance. A decrease in glucocorticoid levels or impaired glucocorticoid receptor function might then lead to increased stress responsiveness [2].

Childhood abuse and neglect is also associated with reduced adult hippocampal volume, particularly on the left side, and these findings support the hypothesis that exposure to early stress in humans, as in other animals, affects hippocampal subfield development [4].

Another recent study demonstrate that children who experienced two or more types of violence exposure showed significantly accelerated telomere erosion from age-5 baseline to age-10 follow-up measurement compared with children who had one type of violence exposure or who were not exposed to violence [5].

People experience and interpret physical and emotional insults in diverse ways and many contextual factors affect the phenomenology of abuse and neglect, and in Italy there are many children victims of various types of maltreatment particularly within their own families. But we must not forget the most vulnerable children who may have even more serious consequences: those who live in foster care [6] or abandoned babies [7] that are actually little-known but that equally needs of social interventions, health and human rights.

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### A33

#### Family-centered care

Filippo Festini

Department of Health Sciences, University of Florence, Nursing Research

Unit, Meyer Children Hospital, Florence, 50139, Italy

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Family-Centered Care (FCC) is a way to provide health care that recognizes the importance of family for a hospitalized child. According to it, it is essential to involve families in their children's recovery plan [1].

FCC was born in the Fifties of 20th century as a reaction to the leading idea in Pediatric care that emphasized physical care, technical abilities and emotional detachment and considered the family as a potential threat to a child's path to recovery. Thus, families -particularly mothers- were physically separated from their sick children, being "substituted" by healthcare professionals, who took on parents' caring functions.

FCC is aimed to maintain and strengthen family roles and the natural bond the family has with the child, in order to promote a healthy family functioning, even through sickness. The physical presence of the family at bedside was FCC's first achievement [2].

Some Authors define FCC as: "the professional support to the child and its family, through participation, engagement and sharing, in a contest of empowerment and negotiation" [1].

According to some Authors, there are eight core concepts that accomplish FCC in everyday clinical practice: 1- to consider family to be the centerpiece of a child's life, remembering that healthcare professionals and healthcare delivery structures should be temporary entities, thus they have to "enter on tiptoe" in a little patient's own world; 2- to promote cooperation between families and healthcare professionals; 3- to facilitate communication and a bidirectional exchange of information between families and healthcare professionals, with no omissions, nor falsifications; 4- to recognize and respect each family's peculiarities (cultural, ethnic, spiritual etc) and its strengths; 5- to recognize and respect each family's coping strategies, to value them and to include these strategies in every recovery plan; 6- to encourage support between families; 7- to assure the best and highest grade of flexibility and accessibility when providing health care services, particularly for those families whose children need specialized and chronic therapies; 8- To always care about emotions, fears and desires of the family [3].

Even though FCC is the common gold standard philosophy of health care in the pediatric departments of Western industrialized countries, it has recently been introduced in Italy, where FCC is still uncommon.

In order to evaluate to what extent FCC is implemented in a hospital, a FCC questionnaire is available [4].

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### A34

#### Parental alienation diagnosis. A modern and effective subtype of domestic violence, endemic in Italian courts

Maria Serenella Pignotti

Dipartimento Feto-neonatale, AOU Meyer-University of Firenze, Viale

Pieraccini 50100 Firenze Italy

E-mail: m.pignotti@meyer.it

Italian Journal of Pediatrics 2014, 40(Suppl 1):A34

In 1985 Gardner, publishing on sexual abuse [1], described a clinical picture, Parental Alienation Syndrome (PAS) as a "disorder that arises primarily in the context of child-custody disputes. Its primary manifestation is the child's campaign of denigration against a good, loving parent, a campaign that has no justification." [2]. Originally Gardner defined PAS in gender specific terms, mothers as alienators and fathers as victims. Later, he claimed that either parent could be an alienator [2]. However, almost always, PAS affects women. Gardner specified "when true parental abuse and/or neglect is



present the child's animosity may be justified, and so the parental alienation syndrome diagnosis is not applicable" [2] but never clarified how to make such differential diagnosis [3-5]. In Gardner's mind, women involved in divorce trial become psychopathic in the sphere of life related to parenting [6], although PAS "is not only simply a matter of brainwashing or programming in that the children contribute their own elements" [4]. It is based on three main principles: a. the children are liars; b. the father's alienation is a mother's responsibility; c. the allegations of maltreatment are false. To a large extent, PAS tautologically presumes lack of justification for the refusal, maternal programming and absence of children's credibility. PAS' treatment, based on legal coercion through court-ordered threats of deprivation of custody to force mothers and children to act with affection toward the father, violates the principles of good medical practices [7]. PAS's theoretical roots lie on Gardner's theory of human sexuality that justifies adult-child sexual contact and gender violence as beneficial to the reproduction of the species and PAS becomes "a defence strategy for abusive fathers, facilitating these men's projection of blame for their children's refusal onto mothers as a counter-claim to, and evidentiary shield against, allegations of violence" in domestic violence cases [7]. PAS is detrimental to children, women and honest men' civil rights and to Justice! It is a tool aimed to punish women and children reacting to a patriarchal system that presumes all reports of male violence are false and punishes protective mothers. According to American Academy of Paediatrics family violence is a paediatric issue[8]. PAS-ideology's influence on family and criminal Court could lead to wrong decisions with foreseeable emotional upset and trauma to the children and consequently to stark outcomes, including murder and suicide, as in the past [6].

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#### A35

##### Performance evaluation in healthcare: the experience of maternity pathway from Tuscany to the Italian network of regions

Manila Bonciani\*, Barbara Lupi, Sabina Nuti

Laboratorio Management e Sanità, Istituto di Management, Scuola Superiore Sant'Anna Pisa, Italy

*Italian Journal of Pediatrics* 2014, **40(Suppl 1)**:A35

**Background:** Considering that performance measurement is largely recognised as a key tool for quality improvement, which kind of performance evaluation can be useful for healthcare professional, general managers and regional policy makers to improve the mother and child healthcare services? The presentation of the experience in assessing the maternity pathway (MP), made initially by Tuscany and successively shared within a network of Italian regions, will contribute to answer this question.

**Materials and methods:** Since 2004, within the general framework of the multidimensional performance evaluation system introduced in Tuscany to assess and monitor local health authorities and teaching hospitals [1], a specific MP performance measurement system (PMSMP) was adopted in order to evaluate the MP process and outcomes [2]. It was based on several indicators grouping in six dimensions: population's state of health, compliance with regional guidelines, efficiency and financial performance, clinical and health assessment, patient satisfaction (assessing the women's experience of MP care), employees' satisfaction (measuring the MP organizational climate). Starting from 2008 this evaluation system has been

implemented also by a network of Italian regions (Basilicata, Autonomous Provinces of Bolzano and Trento, Liguria, Marche, , Umbria, Veneto and recently also Emilia Romagna and Friuli Venezia Giulia) [3].

**Results:** The PMSMP allowed to monitor the progressive changes in the Tuscan and other regional MP. Its implementation during these years have showed that the benchmarking approach encouraged to learn from other experiences, by identifying best practices in mother and child healthcare services. The mechanism of scoring the evaluation indicators and displaying them on a spider diagram allowed to identify immediately the performance dimensions with weak results requiring specific strategies for improvement. Actually, the PMSMP is developing in order to answer the regional and local health authorities managerial needs. A regional professional group is discussing on the introduction of specific indicators on healthcare process and results of new-born infants, and at the level of the Italian regions network, new indicators are being identified in order to evaluate specifically the paediatric care at hospital level.

**Conclusions:** The experience developed in Tuscany and in the network of the Italian regions represents a good example of performance evaluation in healthcare, also for the involvement of health professionals in identifying and defining evaluation indicators. It is an useful tool to evaluate all the phases of MP (pregnancy, delivery and postpartum) with managerial purposes, by identifying the organisational determinants of the MP process and outcomes.

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#### A36

##### Quality of care in children with chronic diseases

Paola Rucci\*, Elisa Zanello, Davide Roccaro, Simona Calugi, Giulia Pieri, Maria Pia Fantini

Department of Biomedical and Neuromotor Sciences, University of Bologna, Bologna, Italy

E-mail: paola.rucci2@unibo.it

*Italian Journal of Pediatrics* 2014, **40(Suppl 1)**:A36

**Background:** Improving health care for chronic health conditions is a major goal of contemporary health service delivery systems. To date, the main research focus has been on adults and elderly, while fewer attention has been focused on newborns and children with chronic health conditions. To address the challenges related to the provision of integrated care to children with special health care needs, the Special Needs Kids (SpeNK) project was carried out in Emilia-Romagna Region. The specific aims of this study were: to review the ongoing sheltered discharge procedures, to develop and test instruments to assess the families' perspective on the continuity of care and to estimate the time devoted by the family pediatrician to care coordination activities.

**Materials and methods:** The SpeNK project is still ongoing. After reviewing the procedures implemented in the Local Health Authorities of the study area, 10 face-to-face, 3 telephone semi-structured interviews and a focus group with the families at 1-6 months from discharge were conducted by a psychologist. A 20-item questionnaire on continuity of care was then developed, based on the contents of the interview and on Haggerty's constructs of informational, management and relational continuity. The questionnaire was validated on 102 parents of preterm newborns and then administered by phone to the families of children enrolled in the SpeNK study after 9 months from discharge.

**Results:** A qualitative analysis of the contents of the semi-structured interviews revealed that families underscored the importance of informational continuity among hospital clinicians and across health care services, set a high value on the information/training received during the hospital stay of children and at discharge, and exhibited a mixed attitude towards involvement in the health care decisions.

Examination of the structure of the questionnaire using factor analysis with oblique rotation identified 5 factors accounting for 61.1% of the

variance of items. The factors identified can be interpretable as 'management continuity', 'informational continuity' 'trustful relation with the family pediatrician', 'information provided to families' and 'family empowerment'.

**Conclusions:** The preliminary results of the qualitative analysis of the semi-structured interviews suggest the relevance to the families of the hospital experience and some criticalities in the informational continuity among professionals. The questionnaire on continuity of care has a well-defined structure and can be a useful tool to capture problems related to informational continuity and to the interaction of families with the professionals involved in the care of children with special care needs.

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### A37

#### Quality of care in pediatrics: the organization of a children's hospital

Massimiliano Raponi

Medical Director of Bambino Gesù Children's Hospital, Rome, 00165, Italy

E-mail: massimiliano.raponi@opbg.net

Italian Journal of Pediatrics 2014, 40(Suppl 1):A37

The Bambino Gesù Children's Hospital (OPBG) is a Research and Health Care Institute (IRCCS), organized in four sites located in Lazio Region (Roma Gianicolo, Roma S. Paolo, Palidoro, S. Marinella) and three sites located in Calabria, Basilicata and Sicily respectively. The OPBG as IRCCS, must always continue and reinforce its commitment in the integration of research and clinical practice, and transfer into the clinical practices of the knowledge produced by scientific research.

The OPBG Medical Direction is organized according to a medical and nursing multidisciplinary model and involves the interaction of a variety of professionals who work for the achievement of organizational and healthcare goals. This organization has allowed the implementation of multiple activities aimed to fully empower the staff, promote professionalism, develop awareness in relation to the needs of patients and their families and increase the focus on the continuous improvement of quality of care.

The IRCCS Bambino Gesù Children's Hospital has long since launched a series of initiatives aimed at the continuous improving in quality of care, organized according to a model of integrated clinical governance, which focuses on the whole patient and the care plan family. This approach integrates the treatment options with the patient/family's expectations in a model that imagine doctor and patient/family as cooperating partners (Family - Centered Care). In this context it is also ensured the empowerment of the patient and his family, who become a central point in decision-making and treatment processes.

Operationally, the Hospital every year prepare a Corporate Program for Continuous Improvement of Healthcare Quality, which identifies the objectives, even in terms of quantities, to be achieved in the year to ensure the continuous improvement of quality of care and monitors the achievement of targets set for the previous year.

The initiatives that are most representative of continuous promotion of healthcare quality carried out in the Hospital include: surveillance, control and prevention of adverse events; evaluating of the clinical and organizational appropriateness; production, dissemination and implementation of clinical protocols and evidence-based clinical pathway; surveillance, control and prevention of healthcare associated infections; design of clinical records that favor appropriateness and patient safety.

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### A38

#### The Italian Pediatric Network

Riccardo Longhi<sup>1\*</sup>, Maria T Ortisi<sup>1</sup>, Gianluca Gargantini<sup>2</sup>, Domenico Minasi<sup>3</sup>, Nicoletta Matera<sup>1</sup>, Luciana Parola<sup>4</sup>

<sup>1</sup>U.O di Pediatria, Ospedale Sant'Anna, S.Fermo della Battaglia, Como, 22020, Italy;

<sup>2</sup>U.O di Pediatria e Neonatologia, Ospedale di Lodi, Lodi, 26900, Italy;

<sup>3</sup>U.O di Pediatria, Ospedale Santa Maria degli Ungheresi, Polistena, 18061, Italy;

<sup>4</sup>U.O di Pediatria e Neonatologia, Ospedale Fornaroli, Az.Osp.Ospedale civile di Legnano, Magenta, 20013, Italy

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The Italian Pediatric Network is now 8 years old.

The number of pediatric Departments has increased from the 19 that in 2006 began a feasibility study of the project, to the 129 of today.

The objectives of the network are: scientific research, surveillance of rare events, monitoring of complex phenomena, standardization of diagnostic criteria and therapeutic processes, creation of a hospital network capable of allowing a fast and profitable exchange of informations, evaluation of the degree of implementation of clinical Guide Lines.

The Network was created by the Working Group for the Accreditation and Quality Improvement (GSAQ) of the Italian Society of Pediatrics Clinical. Date are recorded at the time of discharge by a single operator for each hospital and loaded into anonymous electronic case report forms, different for each pathology, prepared with the help of subspecialty Scientific Societies and available in the Pediatric Network website [http://networkpediatrico.sip.it].

It is not possible to report here all the results obtained from the analysis of the data concerning the first four pathologies investigated (idiopathic thrombocytopenic purpura, diabetes at onset, bacterial meningitis after the neonatal period, acute asthma after the second year of life).

We will present just a few data on the fifth pathology: acute gastroenteritis in children below 5 years of age. 31 centers filled 612 case report forms in 7 months.

It is interesting to compare the diagnostic and therapeutic policies of the participating units with the indications of the ESPGHAN Guide Lines. The appropriateness of admissions was rather low, being only 42,5% altogether, and minimal for patients admitted under pressure from the family, 2%. Appropriateness of treatments: only 2/3 of the patients have been correctly treated showing complete or almost complete adherence to the recommendations (no more than 2 major violations or one major and two minor). Major violations were considered those that might negatively affect the course of the disease, unnecessarily increase the cost of treatment or any violation to high grade recommendations, minor violations those that did not change the course of the disease, even if not appropriate and all interventions in contrast with low grade recommendations.

The most frequent violations were: microbiological studies, wrong diet prescriptions, use of unrecommended antidiarrhoeal drugs or of probiotics lacking evidence of efficacy, prescription of antibiotics.

Two other report forms, concerning bronchiolitis and ALTE, will be officially presented at this meeting and are ready to be launched in the network.

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### A39

#### Nutrition of very low birth-weight infants

Mario De Curtis<sup>1\*</sup>, Lucia Dito<sup>1</sup>, Renato Lucchini<sup>1</sup>, Gianluca Terrin<sup>2</sup>

<sup>1</sup>Department of Pediatrics and Neuropsychiatry, University of Rome La

Sapienza, Rome, Italy; <sup>2</sup>Department of Gynecology-Obstetrics and Perinatal Medicine, University of Rome La Sapienza, Rome, Italy

E-mail: mario.decurtis@uniroma1.it

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**Background:** Numerous studies have underlined the importance of early feeding on short- and long-term development of very low birth-weight (VLBW) neonates [1,2]. Nutrition of preterm infants may be divided in two subsequent periods: the early adaptive or "transition" period from birth to the second week of life followed by the "stable-growing" period up to discharge from the neonatal unit. Depending on birth weight and gestational age the transition period may be prolonged. Immediately after birth most VLBW infants are unable to start enteral feeding. Parenteral nutrition is presently proposed since the first days of life to limit malnutrition. The ideal of nutrients intake for VLBW neonates is still a matter of debate [1,2].

**Materials and methods:** We revised studies with high level of evidence (randomized clinical trials and meta-analysis) regarding enteral and parenteral nutrition in VLBW neonates. Database: MEDLINE and Pubmed. Search term: Nutrition (AND) Very Low Birth Weight. Limits: Randomized clinical trial (AND) Meta-analysis.

**Results:** We analyzed 66 manuscript (12 meta-analysis, 44 RCT) published from 1984 to 2014. Analysis of the best evidences revealed that, up to now, early nutritional strategies vary dramatically among centers and there are no definitive and well accepted regimens demonstrating long term benefits in VLBW infants. To reduce the temporary interruption of the transfer of nutrients, a so called "aggressive" nutrition has been proposed, consisting in a high protein supply (>2g amino acids/kg/d) and the use of

IV lipid (0.5-1.0 g lipid/kg/d), from the first day of life. Minimal enteral nutrition will be added as early as possible to provide a small quantity of milk to stimulate the enterocytes [3]. As the infant matures and the medical conditions stabilize parenteral nutrition can be slowly replaced by enteral nutrition. Nutrition of VLBW infants during the transition period should cover as much as possible the nutritional needs, to limit the inevitable cumulative nutritional deficit, induce a positive nitrogen balance and reinstate weight gain and longitudinal growth. During "stable-growth" in most preterm infants nutritional supplies can be provided by fortified mother's milk or preterm formulas.

**Conclusions:** Analysis of best evidences suggest that, in order to specifically enhance lean mass growth and improve the early catch up growth, the use of a high protein regimen (4.0-4.4 g of protein/kg/d) combined with a high protein energy density regimen (protein/energy ratio up to 3.3 g/100 kcal) early in life promotes growth without metabolic stress in VLBW infants.

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#### A40

##### The Importance of immunonutrition in children

Roberto Berni Canani<sup>1</sup>, Lorella Paparo, Rosita Aitoro, Yelenia Maddalena, Simona Caprio, Antonio Amoroso, Carmen di Scala, Ludovica Leone, Rita Nocerino

Department of Translational Medical Sciences and European Laboratory for the Investigation of Food Induced Diseases, University of Naples "Federico II", Naples, Italy

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A healthy immune system is essential for prevention and recovery in many pediatric illnesses. During last decade, the role of nutrition beyond providing the calories and the macro- and micronutrients for body growth has been well established and clinically proven. Many nutrients have a tremendous potential to modulate directly or indirectly, through a regulation of gut microbiota composition, the development and function of innate and acquired immunity. The potential to modulate the activity of the immune system by interventions with specific nutrients is termed immunonutrition. When we prescribe a particular diet it is important to think that nutrients are not only factors able to influence body growth, but they are also a crucial driving force leading to body health through a regulation of immune system. Within the same category of nutrients it is possible to observe different effects on immune system. As example of this, comparing iso-energetic and iso-proteic doses of different mammalian milks (human milk, donkey milk and bovine milk) we have recently demonstrated, in an animal model, significantly different immunoregulatory and antioxidant properties. These effects are at least in part related to a modification of gut microbiota composition and function, and are able to modulate energy balance, glucose and lipid metabolism. We can modify the immunonutritional properties of a particular food. We have recently demonstrated the possibility through a dietary supplementation with fermented bovine milk with a selected probiotic strain (*Lactobacillus paracasei* CBA-L74) to significantly reduce the number of common winter infectious diseases in school-age children. This preventive effect derives from a complex network of different mechanisms of action: modulation of gut microbiota composition, stimulation of short chain fatty acids production, stimulation of innate immunity (alpha and beta-defensins, and cathelicidin LL-37 production), stimulation of acquired immunity (secretory IgA), modulation of gut permeability, modulation of epithelial cell growth and differentiation. Significant progress at identifying the gut microbiome has led to a better understanding of the interactions between them and our organs and tissues. Probiotics, while not considered a nutrient, are certainly became a even more frequent component of children diet. The roles that ingested organisms may play in atopic diseases are now potential targets of prevention and treatment.

#### A41

##### Nutrition and inborn errors of metabolism: challenges in Phenylketonuria

Elvira Verduci<sup>1</sup>, Valentina Rovelli, Francesca Moretti, Juri Zuvadelli, Elisabetta Salvatici  
Department of Pediatrics, San Paolo Hospital, Department of Health Sciences, University of Milan, 20142, Italy  
*Italian Journal of Pediatrics* 2014, **40**(Suppl 1):A41

Phenylketonuria (PKU) is caused by the deficiency of the phenylalanine hydroxylase enzyme, which converts phenylalanine (Phe) to tyrosine. If left untreated from birth, this deficiency results in high levels of Phe in the blood, neurotoxic to the brain [1]. The restriction of dietary Phe represents the mainstay of PKU management. PKU diet is mainly made up by variable amounts of vegetables and fruits (poor in Phe natural foods), minimal amounts of animal products (usually milk), low-protein foods (low-protein bread and pasta) and Phe-free protein substitutes, which provide mainly essential aminoacid and micronutrients, to reach the required amount of daily protein, minerals and vitamins [2,3]. This type of dietary regimen provide lower saturated and polyunsaturated fat, cholesterol as well as higher carbohydrates intake than healthy pediatric population. The PKU diet follows the norms of the so called "prudent" diet for the prevention of cardiovascular disorders. In particular saturated fats may be less than 7% and polyunsaturated higher than 5% total energy with a supply less than 50 mg cholesterol per day [2]. Indeed PKU children show lower plasma cholesterol levels as compared to healthy children, particularly low density lipoprotein particles. Nevertheless both dietary habits and genetic predisposition may interact in keeping low blood lipid levels in PKU population [2]. However a lower antioxidant status and higher homocysteine levels have been reported in PKU, suggesting a possibly increased risk for thrombosis, atherosclerosis and stroke [4,5].

Furthermore, even if PKU children are routinely long-term monitored for dietary intake some studies showed evidence for overweight in this population [6,7]. However more data on body composition in PKU individuals are needed. Further research should be necessary to better understand the nutritional quality of low-protein foods and Phe-free protein substitutes. This nutritional aspect is particularly interesting, in view of recent results about low-protein products: low-protein bread, pasta, flour and breakfast cereals appear to provide from 2 to 18% more energy than their protein-containing equivalent food [6] and some low-Phe pasta and crackers, commercially available, show a high glycemic index (data unpublished). Moreover an altered food behaviour, such as an irregular intake of higher fat food or low-protein foods, has been reported [8,9]. Few data are available on physical activity levels in PKU patients [8]. In conclusion given the growing population of adults with PKU, it could be important to investigate the non-communicable diseases risk in this population to better optimize nutritional treatment strategies.

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#### A42

##### Infant nutrition and risk of celiac disease

Carlo Catassi

Department of Pediatrics, Università Politecnica delle Marche, Ancona, Italy  
*Italian Journal of Pediatrics* 2014, **40(Suppl 1):A42**

Celiac disease (CD) is an immune-mediated enteropathy triggered by the ingestion of gluten in genetically susceptible individuals. Gluten is the major protein component of wheat, rye and barley. The major predisposing genes are the HLA-DQ2 and DQ8 genotypes found in at least 95 % of patients. CD is one of the most common lifelong disorders on a worldwide basis affecting 0.5-1 % of the general population in Western countries. CD is a multifactorial disorder that depends on both genetic and environmental factors for expression.

The analysis of factors associated with an epidemic of early-onset CD in Sweden during the 1980-90s indicated a role for infant nutrition, the disease risk being substantially lower in infants introducing small amount of gluten when still breast fed. The protective role of breast feeding has been supported by other case-control studies. As far as weaning, an increased risk of CD has been reported in infants introducing gluten containing food before the age of 4 months. Most of these data investigated only clinically detected cases of CD, leaving unanswered the question whether factors related to infant protect or simply delay the onset of disease. Ongoing European, prospective studies on at-risk infants followed on by serological screening are shedding light on the role of infant nutrition in the development of CD. The Italian Baby Study recently completed a 10-years period of follow-up. The results of this study suggest that genetically predisposed children tend to develop CD autoimmunity early in life, generally before the age of 5 years. Early dietary factors, particularly age at gluten introduction, seem to play a minor role in the development of CD, at least in infants at family risk, questioning the validity of the "window of exposure" hypothesis (lower risk in infants introducing the new antigen between 4 and 6 months). The possible preventive effect of delayed gluten introduction in infants with high HLA risk (double copy of DQB1\*02) is a puzzling finding deserving further investigation. In this study, breast-feeding in children at-risk of CD did not seem to exert any clear-cut effect on the disease risk. Another European, prospective and randomized study (PreventCD) will soon clarify whether small amount of gluten given during breast feeding do indeed protect from CD development.

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#### A43

##### The diet in children with diabetes mellitus (DM)

Dario Iafusco\*, Angela Zanfardino, Santino Confetto, Francesco Prisco  
Department of Pediatrics, Second University of Naples, Naples, Italy  
E-mail: dario.iafusco@unina2.it

*Italian Journal of Pediatrics* 2014, **40(Suppl 1):A43**

A proper nutritional plan is important in the treatment of children and adolescents with diabetes. Its main functions are a) maintaining a good metabolic control, reducing the risk of hyperglycemia and hypoglycemic crisis (induced by prolonged fasting or errors in insulin dose) and b) sustaining the "long term health", allowing a good general state of health, normal growth and pubertal development and reducing the risk of micro and macro vascular complications.

The common opinion is that the general characteristics of a diet for a child with diabetes do not differ from the recommended intakes for the general pediatric population (Italian term: LARN). However until a few years ago the adaptation to the recommendations was not entirely possible being necessary to modulate the intake of carbohydrates with the pharmacodynamics of the various types of insulins. The introduction of new insulins such as fast, slow analogs and, at the same time, the increased use of subcutaneous insulin infusion pumps have allowed a better adaptation of the dietary habits of the different ages of life not underestimating that the approach to food by children and adolescents contains emotional meanings (*premium, punishment, gratification, a sense of homology with peers, etc.*) that may affect the compliance to the therapy. In the basal-bolus insulin regimen it can be very useful to apply the "rule of carbohydrates" (adapting the dose of insulin to the amount of carbohydrates in the meal) and the *insulin index* (a parameter which takes into account the effect of amino acids and proteins on postprandial blood glucose levels). The choice of analog rapid or human

insulin as boluses in the treatment with multi injection therapy or the possibility to change the bolus type (*square-wave, double-wave, etc.*) in the case of the pump can take in consideration also the timing of absorption of carbohydrates delayed on the basis of the *glycemic index* of food [1].

However, in line with the global obesity epidemic, a raised weight gain has been described also in adolescents with type 1 diabetes mellitus (T1DM). We have recently shown high prevalence of abdominal adiposity (28,1%) as a sign of metabolic syndrome (MetSy) (9,5%) in adolescents with T1DM [2,3]. On the other hand, due to the prevalence of obesity in children, Type 2 diabetes mellitus (T2DM) in adolescents is becoming an increasingly important public health also in Italy. Diet and exercise are the therapy and prevention of Type 2 diabetes and obesity [4].

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#### A44

##### Research and young paediatricians

M Loredana Marcovecchio

Department of Paediatrics, University of Chieti, 66100, Italy  
*Italian Journal of Pediatrics* 2014, **40(Suppl 1):A44**

The fundamental role played by medical research in the diagnosis and treatment of diseases is well recognized. In the paediatric field, research is crucial for the health of future generations. For a young paediatrician, being engaged in research requires curiosity and enthusiasm, strong commitment and constant dedication, but represents a unique opportunity to contribute to the progress of medical knowledge as well as to the overall progress of the society.

Education and training in research should be part of every level of the paediatric training, from premedical to postgraduate education. This will allow the development of research skills, which are useful not only for those who aim to pursue an academic career, but for the daily activity of every doctor. Research training can allow clinicians to keep up with the growing medical literature as well as improve their skills in finding the best evidence to answer questions arising from their daily clinical activity.

Over the last years, national policies have affected the role of scientific research and employment opportunities for young researchers. Italy has a number of PhD students lower than in other European countries. Furthermore, Italian PhD students' salary, even after adjusting for the cost of living, is lower than that of their colleagues from Western European countries. In recent years, post-doctoral programs have also undergone considerable downsizing in terms of funds and availability.

Young people and their enthusiasm are essential for the advancement of research. Therefore, it is essential to promote and boost research in our country, through investments in scientific education of young doctors and by increasing the opportunities for those who wish to engage in medical research.

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#### A45

##### Impact in Italy of research in pediatrics

Generoso Andria

Department of Translational Medicine – Section of Pediatrics, University of Naples Federico II, Naples, Italy  
*Italian Journal of Pediatrics* 2014, **40(Suppl 1):A45**

We examined several parameters related to pediatric research in Italy over the past few years. We distinguished the research carried out by the scientific community as a whole on topics of pediatric interest from the research carried out by pediatricians. We considered only studies in which an Italian institution played a leading role and excluded studies with Italian collaborators, but coordinated by foreign researchers. Data relating to Italian pediatric research were retrieved from national and international sources and databases, such as PubMed, and compared with those obtained with the same methodology for the following European countries: France, Germany, United Kingdom, the Netherlands, Spain, Greece. The results obtained were then corrected for the gross domestic product at purchasing power parity per capita of any nation, which takes into account the number of inhabitants and the purchasing power of resources related to that country. From these data we can tentatively conclude that the Italian pediatric research, despite fewer resources invested, is at the same level, of European countries with comparable population and much higher investments devoted to scientific research. Another "Italian miracle" or a case of "submerged economy", even in research?

#### A46

##### Probiotics: current evidences and new perspectives

Valentina Fabiano, Chiara Mameli, Gian Vincenzo Zuccotti\*  
Università degli Studi di Milano, Department of Pediatrics, Luigi Sacco Hospital, 20157 Milan, Italy  
E-mail: gianvincenzo.zuccotti@unimi.it  
Italian Journal of Pediatrics 2014, 40(Suppl 1):A46

Probiotics are viable microorganisms that can exert potential benefits to human health. Use of probiotics in different human diseases and clinical conditions has gained much attention in the last decades. Several data exist about the potential benefits of probiotics in pediatric age for the management of some clinical conditions, with different and sometimes contrasting results. Currently, strong evidences support the use of probiotics for treatment of acute infectious diarrhoea and prevention of antibiotic-associated diarrhoea: bacteria belonging to the genus of lactobacilli and *Saccharomyces boulardii* showed the stronger evidences of benefits in different clinical trials and meta-analysis [1,2]. For other pediatric diseases and clinical conditions, evidences of the benefits of a probiotic supplementation are still not so strong. The recommendation to administer a probiotics supplementation in preterm, very low birth weight neonates at risk of developing necrotizing enterocolitis has recently been criticized by a critical review of the literature that conclude that well-designed clinical trials are still needed for suggesting its routinely use in these neonates [3]. Discussion is still ongoing about the benefits of probiotics in prevention or treatment of other pediatric diseases such as respiratory tract infections, urinary tract infections, *Helicobacter pylori* infection or allergic diseases. Clinical trials and meta-analysis about the use of probiotics in these pediatric clinical conditions have been published and still continued to be published; however, study designs are not always comparable and sometimes inadequate, species of probiotics used in the studies are different and the benefits demonstrated for one microorganism are not generalizable to other bacteria, results of the studies are still often not unequivocal or contrasting so that strong recommendations are still lacking. The use of probiotics has been suggested to be potentially beneficial for the management of gastrointestinal functional disorders, such as irritable bowel syndrome, again, with different results. These clinical conditions are supposed to be the result of a perturbation of the microbiota balance that leads to dysbiosis, responsible for the appearance of the characteristic symptoms. However, pathogenesis of gastrointestinal functional disorders appear to be much more complex and involve the nervous system and its bidirectional interactions with the intestine. Moreover, it has been hypothesized that intestinal microbial composition may itself influence end-points related to mood state, brain function, and mental outlook. A new fascinating perspective in studies about microbiota and probiotics is the characterization of the alterations of intestinal microbiome and their correlation with abnormalities in the bidirectional gut-brain interactions.

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#### A47

##### Undocumented children's health – key challenges and local responses

Michele LeVoy\*, Lilana Keith  
PICUM, Platform for International Cooperation on Undocumented Migrants, Brussels, 1000, Belgium  
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**Introduction:** Every European Union member state has legal obligations to protect the rights of the child. Nevertheless, there are children in Europe that face significant barriers to accessing even essential health services, due to their or their parent's migration or residence status. Undocumented children usually receive health care under the same conditions as adult undocumented migrants, with no extra protection, and are often unable to access the majority of the health care services they need.

**Access to health care in law and practice:** National laws regarding access to health care for undocumented children—what care children may access and under what conditions and costs—vary enormously across Europe. In many countries, undocumented children are only able to access emergency health care services, and in a few, even this many be subject to charging. In a number of countries, certain types of care that are considered 'urgent', 'essential' or 'medically necessary' are provided. In a number of others, all children are afforded equal or near equal access to health care services [1-3].

Numerous practical barriers can affect children's ability to access to care that they are entitled to. These include discretion on local level, varied interpretations of what care undocumented children are entitled to, lack of awareness of children's entitlements, and fear of detection/ denunciation [3-5]. Children's health may also suffer generally due to a lack of continuous care, lack of access to specialist and mental health care, and due to conditions related to living in an irregular migration situation, such as poor housing conditions and stress [6,7].

**Local responses:** Non-governmental organisations, individual health professionals and in some cases, local and regional hospitals and health authorities are striving to fill the gaps left in state service provision and meet urgent health needs in their communities despite limited resources, while also advocating for policy change [8-10].

**Conclusion:** It is the duty of governments to ensure non-discriminatory access to health care services. There is a growing body of evidence that indicates that universal health coverage is necessary and beneficial from the perspectives of individual health, development and social inclusion, public health (including functioning and financing of public health systems), human rights, medical ethics and humanitarian principles. Alongside the examples of inclusive practice from national, regional and local levels, this evidence base requires expeditious reform of discriminatory migration and health policies.

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#### A48

##### Metabolic syndrome: new therapeutic approaches

Claudia Della Corte, Valerio Nobili\*

Hepatometabolic Department, "Bambino Gesù" Children's Hospital, S. Onofrio Square, 4, 00165, Rome, Italy

E-mail: nobili66@yahoo.it

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In the last three decades in the United States the prevalence of overweight/obesity in pediatric population has more than tripled, causing the onset also in pediatric age of diseases previously considered exclusively of adults, such as metabolic syndrome (MetS) [1]. MetS represents a cluster of cardiometabolic abnormalities, including visceral obesity, dyslipidemia, hypertension and diabetes mellitus type 2 (T2DM) (Table 1). The prevalence of pediatric MetS ranged from 2% to 9% in the general population and from 12% to 44% in obese children, depending of definition used [1]. Several evidences suggest that the metabolic rearrangements observed in children may have a worrisome repercussion early on their health in adulthood [2,3].

Lifestyle modification, represented by the association among regular physical exercise and a balanced diet appropriate for age, is the most important therapeutic approach in children and adolescent with obesity and risk factors for MetS [4]. Behavioral intervention is mandatory but, in many cases, it is difficult to achieve or not sufficient and most pediatric patients require pharmacologic therapy early in their disease course. At the present time, the vast majority of drugs needed to treat insulin-resistance, hypercholesterolemia, hypertension are off-label in pediatric setting, although several studies demonstrated that pharmacological treatment for pediatric obesity and its related comorbidities are necessary [5].

Regarding dyslipidemia, the use of oral statins is reserved for children older than 10 years of age that, while on diet, continue to have dyslipidemia associated to family history for early cardiovascular disease (CVD) or additional risk factors.

Metformin is the only drug approved for treatment of impaired fasting glucose (IFG) or impaired glucose tolerance (IGT) in children. Several

studies demonstrated its efficacy in ameliorating gluco-insulinemic profile; moreover, it has been reported a moderately effect on body weight in obese children.

Anti-hypertensive drugs, such as angiotensin-converting-enzyme inhibitors (ACEIs), angiotensin-receptor blockers (ARBs), calcium channel blockers (CCBs), and beta-blocking agents are used in pediatric patients and the choice of drug class is made on the basis of clinical characteristics of the single patient.

The core of treatment of pediatric MetS is abdominal obesity. Currently no pharmacological approach to obesity is accepted for pediatric patients. Bariatric surgery has been considered a successful treatment for MetS in obese adults in term of weight loss and decrease of mortality rate. This procedure has been used also in carefully selected obese adolescents and the outcomes seem to be similar to those for adults. However, further studies are needed to better select the patients to surgically treat and define efficacy and safety of bariatric surgery in pediatric MetS.

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#### A49

##### Protein intake and nutritional programming: metabolic consequences

Elvira Verducci\*, Benedetta Mariani, Carlotta Lassandro, Alice ReDionigi  
 Department of Pediatrics, San Paolo Hospital, Department of Health Sciences, University of Milan, Milan, Italy

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According to recent epidemiological evidence, early protein intakes that exceeds metabolic requirements (>15% of energy) may increase weight gain during infancy and the risk of developing obesity in childhood: the

**Table 1(Abtract A48) Diagnostic criteria for metabolic syndrome in children and adolescents**

Age (years)	IDF criteria		
	6-9	10-15	>15 (adult criteria)
Waist circumference	≥90 <sup>th</sup> percentile for age (MS as entity is not diagnosed)	≥90 <sup>th</sup> percentile or adult cut-off if lower	≥94 cm for males, ≥80 cm for females
Blood pressure		Systolic ≥130 or diastolic ≥85 mmHg	Systolic ≥130 or diastolic ≥85 mmHg or treatment of previously diagnosed hypertension
Triglycerides		≥1.7mmol/L (≥150 mg/dL)	≥1.7mmol/l (≥150 mg/dL) or specific treatment for high triglycerides
HDL-C		< 1.03 mmol/L (<40 mg/dL)	< 1.03 mmol/L (<40 mg/dL) in male and 1.29 mmol/L (<50 mg/dL) in females or specific treatment for low HDL-C
Fasting glucose		5.6 mmol/l (100 mg/dL)	5.6 mmol/l (100 mg/dL) or known T2DM

IDF: International Diabetes Federation

HDL Cholesterol: High-density lipoprotein cholesterol

so-called "early protein hypothesis" [1]. A high protein intake, indeed, especially milk's protein [2,3], may enhance the secretion of insulin and insulin-like growth factor-I (IGF-I), associated with increased weight gain during the first 2 years of life, increase of adipocyte differentiation and adipogenic activity [4]. In a large number of studies increased weight gain in infancy and early adiposity rebound have been associated with the development of later obesity [4]. The presence of a positive strong association between early protein intake and increased weight gain in early childhood has been recently demonstrated in the European Childhood Obesity Project (CHOP): a multicenter, double-blind intervention trial involving both formula fed infants, randomly assigned to receive, during the first year of life, infant and follow-on formulas with different protein content (high or low), and breastfed infants as control group. This trial showed that both weight-for-length and BMI z-scores were significantly higher in the high protein (HP) compared with the low protein (LP) group at 12 and 24 months [5]. Moreover the body composition analysis at 6 months of life showed that weight gain velocity from baseline to 6 months was significantly associated with fat mass, proving that higher early protein intakes may influence adiposity [6]. Concerning metabolic data, HP group compared with LP group showed higher plasma concentrations of branched chain aminoacids, IGF-I and insulin at 6 months. Moreover, IGF-I concentrations have been associated with weight gain in the first 6 months of life [7]. Additionally IGF-I could partly mediate protein-induced kidney growth in healthy children [8]. This phenomenon was observed in the HP group: children at 6 months of life showed a significantly greater kidney volume compared with the LP group [9]. However the long-term consequences of these results should be further evaluated.

Lastly it has been recently published that protein content of infant formulas may influence not only the growth pattern in the first two years, but also the risk of developing obesity at school-age, showing a 2.43 increased risk of obesity at 6 years of age in the HP vs LP group [10].

In conclusion these results suggest that early nutritional intervention programs are needed to avoid negative long-term consequences on health, especially to prevent "non-communicable diseases" in adult age.

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#### A50

##### **Continuity of Care and Home Care / organization of a process to improve the integration with the territorial health care facilities and the involvement of the family for a safe discharge of chronic patients**

F Tondi

Nursing Management, Meyer Children's University Hospital, Florence, 50139, Italy

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**Objectives:** The increase in paediatric patients suffering from chronic disease, often involves the transfer between different care sectors. The transition from the protected hospital environment to the patient's home is often faced with difficulties and discomfort. Therefore it is essential to ensure proper management of the course of treatment and care of the child and his family, from admission to return to home, as a single longitudinal and transverse episode. The transition phase invests heavily on empowerment of the patient and the family, as a necessary condition for safe care in self-management.

**Materials and methods:** The early assessment of care needs in the early hours of admission allows signalling through a standardized form (within 48 hours of admission) to the territory of residence/domicile post-discharge care needs. The immediate activation of the territory allows you to organize the take-over without solutions of continuity. The preparation in particularly complex cases of a joint multi-professional assessment Hospital/Territory before discharge ensures total care of both patient and family. The investment in empowerment of patients and their families through a card intended to guide the informative/educational activities and assessment of learning outcomes ensures a safe discharge as well as a competent share of the care pathway for the patient and family.

**Results:** All children requiring home care have been taken into care before discharge and the first access at home was made within 2 days of discharge. Patients and family members have continued training activity even at home until reaching complete autonomy.

The detection of user satisfaction reported in 80% of cases who returned the questionnaire a good satisfaction.

**Conclusions/prospects for improvement:** The focus in 2014 will be placed primarily in the reporting of at least 90% of the cases that require territorial continuity of care within 48 hours of admission and the onset of training already at this stage. It should be also developed a greater integration between hospital school and schools of the area of residence of the child for activation when needed to address the educational activity.

#### A51

##### **PAlarm: a pediatric clinical worsening identification system**

Viviana Frigato<sup>1\*</sup>, Liliana Vagliano<sup>2</sup>, Maria Cristina Rossi<sup>3</sup>

<sup>1</sup>Città della Salute e della Scienza Hospital of Turin - Regina Margherita Children Department, Italy; <sup>2</sup>University of Turin - Pediatric Nursing Degree Course Italy; <sup>3</sup>Città della Salute e della Scienza Hospital of Turin - Management of In Hospital Emergency Working Group, Italy  
E-mail: vivi\_fri@hotmail.it

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Rapid Response System (RRS) is by now commonly used and internationally validate to manage in hospital emergency. Both its optimal management and early identification are essential to reduce mortality and neurological sequences of anoxia.

However, in the pediatric Italian population, this system is poorly employ, even if its effectiveness has already been demonstrated.

Therefore, in 2010 at Turin a study [1] was conducted in 5 units of a third level pediatric hospital; the aim was to experiment the only pediatric system completely validate in literature (the Pediatric early Warning Score System). In order to apply this system, it was necessary to adapt it at local

reality, analyzing relevant cases (PEWS  $\geq 3$ ); all nursing staff underwent an interview to analyze utility and effectiveness of the system.

The obtained results confirm the hypothesis that a PEWS system is suitable for general departments where respiratory pathologies (main cause of death in pediatric age) prevail.

Thanks to this pilot study, the Regina Margherita Children Hospital of Turin has developed the application of this system at the whole hospital, further adjusting it in order to improve the adaptation at local reality, based on preliminary results. The new system has been called PAlarm (Pediatric Alarm), to better connect with METal methodology broadly used for adult patient all over Italian country.

Moreover, specific educational courses has been developed, both for nurses and physicians, in order to correctly apply this system in journal clinical activity.

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#### A52

##### Which skills are necessary for territorial pediatric nurses?

Paolo Becherucci

Pediatra di Famiglia – Vicepresidente SICuPP, Firenze, Italy

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Territorial primary pediatric care is provided by Family Pediatricians. According to the ACN, Family Pediatricians (PdF) may work individually, in a association or in a group (sharing the same site of the office). The new structure, in accordance with the "Balduzzi Law", instead will include two forms: The Functional Territorial Association (AFT), in which the PdF operate in their studies but are connected functionally, and the UCCP, which instead, should provide services by pediatricians, nurses and other specialists in one office. In a study conducted in Tuscany in 2010, Pediatricians resulted as being equally divided into three organizational modes. The presence of nursing personnel in the offices was almost exclusively limited to working groups. Conversely, office assistants (AdS) were present in all three categories. When the two figures worked together in the same office, the work was divided; sometimes, when there was only one type present, there was a tendency to assign one's own work to the other person. It is therefore evident that in the prospective of radical change in regional pediatric care, the job description of the territorial pediatric nurse, what she should do in that particular job context, must be defined. Competences and a job description are the prerequisites to set a proper training process for this figure. A preventative approach to care represents one of the most important tasks of the family pediatrician. The implementation of routine health checkups (a series of regular assessments to obtain comprehensive health measures for each child) provided by the Childhood-Health project is the means for achieving this goal. The Nursing staff represents a valuable resource to assist family pediatricians in all of their activities. Nurses may do triage for accessing the study, run the self-help diagnostics (some quick tests in the office), offer vaccination counseling and perform vaccines, give advice on childcare, handle some pseudo-pathological situations and ease maternal anxiety. A document representing clear job guidelines, agreed upon by both doctors and nurses is necessary, also for legal purposes. In a time of reduced resources, there needs to be a certain degree of flexibility in the tasks assigned: obviously, for example, making appointments is a task that a nurse can do very well, but it is certainly not an "added value" for the structure in respect to the job of secretary.

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#### A53

##### Promoting children's pain alleviation in emergency department

BM Cantoni

SITRA Area Assistenziale della Pediatria Fondazione IRCCS Ca' Granda

Ospedale Maggiore Policlinico, 20122 Milan, Italy

E-mail: barbara.cantoni@policlinico.mi.it

*Italian Journal of Pediatrics* 2014, **40(Suppl 1)**:A53

**Background:** Pain is, by nature, a subjective experience influenced by social, psychological, and experiential factors.

Nowadays, pain relief is considered an imperative of the pediatric emergency nursing. Analgesia administration influences the entire child's medical experience and can have a lasting effect on patient and family's reaction to current and future medical cares [1].

Pain assessment represents the first step for analgesia management and it should be performed routinely by nurses in emergency department (ED) triage along with vital signs monitoring. Increasing evidence indicates that prompt pain evaluation improves the whole quality of subsequent care. Findings from studies investigating the use of scales for pain evaluation, in addition to the experience gained in clinical practice, suggest to treat all patients presenting a pain intensity major than 4 [2] (scale's range from 0 – no pain to 10 – the worst pain experienced). However, the myth that analgesic treatments would mask symptoms and delay diagnosis in some cases (i.e. children with acute abdominal disease) still frightens physicians and nurses. Therefore, pain reduction is often not considered as a primary task to be accomplished by the ED team [3].

**Materials and methods:** We retrospectively analyzed data regarding admission causes, pain assessment (Numeric Rate Scale, Face, Legs, Activity, Cry, Consolability scale) and management of 23677 children admitted to the pediatric ED of the "Ca"; Granda Ospedale Maggiore Hospital in Milan between January and December 2013.

**Results:** The first reason requiring medical attention was fever (37%). Pain was the second cause (25%). Moreover, children affected by other disease reported concomitant pain in 42% of cases.

Pain was assessed and recorded in 85% of children. A pain with an intensity major than 4 was detected in 31% of cases. Only 72% were treated with an analgesic drug. The non-treated children suffered from abdominal pain. Acetaminophen was the main drug administered (15 mg/kg/dose).

**Conclusions:** Our data indicate that pain is an important cause of concern for families that leads to medical consultation. However, pain assessment at triage is not performed in all patients, yet. Although several data show that analgesia does not mask signs of acute abdominal diseases, nurses still avoid the prompt treatment of abdominal pain. Although non-pharmacological interventions, such as distraction, positioning, sucrose, have been described to be useful in reducing abdominal pain [4] they are often unsatisfactory. In conclusion, we suggest that educational interventions and staff trainings should be addressed to promote children's pain alleviation at triage.

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#### A54

##### Pain in newborn

Lorenzo Giacchetti<sup>1</sup>, Monika Stablum<sup>2</sup>, Arianna De Martino<sup>3</sup>, Arturo Giustardi<sup>4\*</sup>

<sup>1</sup>Department of Pediatrics and Neonatology, Lugano, Switzerland;

<sup>2</sup>Associazione Italiana La Care in Perinatologia, Italy; <sup>3</sup>Department of

Neonatology, Mantova, Italy; <sup>4</sup>Department of Neonatology Merano, Italy

E-mail: artgiust@tin.it

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Recent scientific studies have added more and more consistent evidence that the newborn, even if preterm, is very sensitive to the nociceptive stimulus. The baby responds with a well known physiological, metabolic and hormonal reaction, that, if repeated, may lead to short and medium term negative effects on the newborn. This vulnerability to painful stimulus, especially in preterm infants, can be explained by neurophysiological mechanisms. In spite of a well developed sensory apparatus for nociception,



the descending inhibitory systems and their neurotransmitters responsible for nociceptive afferents, are deficient and immature until after the term birth. Behavioral changes and reductions in the volume of some sensitive brain areas were observed in ex preterm infants admitted to the neonatal intensive care unit; the modifications were very similar to those seen in experimental model of rat with same gestational age, exposed to early and repeated painful stimulus [1]. We consider as environmental interventions all steps that can reduce stress in the baby during a painful procedure. A variety of non pharmacologic pain-prevention and relief techniques have been shown to effectively reduce pain from minor procedures in neonates. These include use of oral sucrose/glucose, breastfeeding, non-nutritive sucking, kangaroo care, facilitated touch (holding the arms and legs in a flexed position), swaddling, and developmental care [2]. The involvement of the mother is recommended through skin-to-skin or breast-feeding during a single sampling. Several studies have shown how breastfeeding during a painful procedure reduces the stress in newborn [3].

The prevention of pain in neonates should be the goal of all caregivers, because repeated painful exposures can have deleterious consequences. Neonates at greatest risk of neurodevelopmental impairment as a result of preterm birth are also those most likely to be exposed to the greatest number of painful stimulus in the NICU. We have to improve strategies for routinely assessing pain, minimizing the number of painful procedures performed, using pharmacologic and non pharmacologic therapies for the prevention of pain associated with routine minor procedures, and eliminating pain associated with major procedures.

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#### A55

##### Essential of audiology: screening and post-screening

Francesco Martines<sup>1\*</sup>, Pietro Salvago<sup>1</sup>, Salvatore Cocuzza<sup>2</sup>, Eleonora La Mattina<sup>3</sup>, Martinelli Stefano<sup>4</sup>, Marianna Mucia<sup>3</sup>, Enrico Martines<sup>3</sup>  
<sup>1</sup>Università degli Studi di Palermo, Dipartimento di Biomedicina Sperimentale e Neuroscienze Cliniche, (BioNeC), Sezione di Otorinolaringoiatria. Via del Vespro, 129 – 90127 Palermo, Italy; <sup>2</sup>Università degli Studi di Catania, Dipartimento di Specialità Medico-Chirurgiche, Clinica Otorinolaringoiatrica, Via Santa Sofia, 68, 95125 Catania, Italy; <sup>3</sup>Università degli Studi di Palermo, Dipartimento di Biotologia e Biotecnologie Mediche e Forensi (Di.Bi.Me.F.), Sezione di Audiologia. Via del Vespro, 129 – 90127 Palermo, Italy; <sup>4</sup>Azienda Ospedaliera 'Ospedale Niguarda Cà Granda' – U.O.C. Neonatologia e Terapia Intensiva Neonatale. Piazza Ospedale Maggiore, 3 - 20162 Milano, Italy  
E-mail: francesco.martines@unipa.it

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Newborn hearing screening is a type of screening test for the early detection of hearing loss. It can recognize with good accuracy newborns affected by hearing impairment allowing an early diagnosis and intervention and avoiding cognitive and linguistic deficits [1-6].

The incidence of bilateral sensorineural hearing loss (SNHL) in Sicily is 2.35 cases per 1000 newborns; this value increases to 2.95 if we consider also unilateral SNHL [2,3] and to 10 cases per 1000 births among infants at risk [7-9].

A correct newborn hearing screening programme is based on different protocols depending on the presence/absence of audiological risk factors:

- Newborns without risk factors: [1-3]

**Initial Hearing Screening (Step I):** The initial screening should be performed using Transient-Evoked Otoacoustic Emissions (TEOAEs) in the birth centers as close to discharge as possible, preferably 12 hours or more after birth. It is recommended that an infant be referred for a re-screening (step 2) if s/he does not pass the initial screening or results cannot be obtained in one or both ears.

**Re-screening (Step II):** The re-screening should be performed in a second level center using TEOAEs and Automated Auditory Brainstem Response (AABR). If an infant does not pass the re-screening or if results cannot be obtained in one or both ears, s/he shall be referred to the regional third level center for diagnostic audiological evaluation.

- Newborns with risk factors (JCIH 2007) [7-10]

**Initial Hearing Screening (Step I):** The Initial Hearing Screening should be performed in a second level center using TEOAEs and AABR. If an infant does not pass the initial screening or if results cannot be obtained in one or both ears, s/he shall be referred to the regional third level center for diagnostic audiological evaluation.

**Screening variables:** Actually TEOAEs have a sensitivity of 100% and a specificity of about 70-95%. A higher TEOAEs specificity value depends on [1,2,11]:

- Timing of TEOAEs recording
- Trained and qualified personnel
- PASS/REFER criteria

**Limitations of screening:** Audiologic screening does not identify:

- Post-natal SNHL (prelingual or perilingual), mainly related to genetic causes [12,13]
- ANSD (Auditory Neuropathy Spectrum Disorder): the main risk factors associated to ANSD are severe jaundice, prematurity, respiratory distress, ototoxic drugs (used to treat neonatal infections), genetic mutations (e.g. OTOF gene). The diagnosis of ANSD is usually based on the combination of absent or abnormal ABR with normal TEOAEs and/or cochlear microphonics (CM) [14].

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#### A56

##### **Late-onset, acquired and progressive hearing loss: surveillance and the role of the pediatrician**

Giovanni Lenzi<sup>1\*</sup>, Stefano Berrettini<sup>2</sup>, Jodi M. Cutler<sup>3</sup>

<sup>1</sup>FIMP (Italian Pediatric Federation), Grosseto, Italy; <sup>2</sup>University of Pisa, U.O.

Otorinolaringoiatria Audiologia e Foniatria Universitaria, Pisa, Italy;

<sup>3</sup>Affrontiamo la Sordità' Insieme, Grosseto, Italy

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A survey related to universal newborn hearing screening was created with permission granted, based on the following study and adapted to the Italian situation: Primary Care Physicians' Knowledge, Attitudes, and Practices Related to Newborn Hearing Screening (Mary Pat Moeller, PhD, Karl R. White, PhD and Lenore Shisler, MS).

First distributed in October, 2008, the survey was given to 300 pediatricians of Tuscany on a regional level, and was then adapted as an online questionnaire made available to the members of the Italian Pediatric Federation (FIMP) in April, 2009. The purpose of the survey was to verify the attitudes, experience and knowledge of family pediatricians, in regard to newborn hearing screening, hearing loss and other related arguments.

The principle objectives of the study were (1) to provide an overview of the current situation of pediatricians in regard to newborn hearing screening and follow-up; (2) to evaluate the knowledge and experience of pediatricians regarding newborn hearing screening programs and (3) to identify the information gaps in order to evaluate the most effective methods of providing the resources and information necessary to assist the families whose children are diagnosed with hearing loss.

The results indicated that there are knowledge gaps as to how to properly manage cases of children with hearing loss (for example, when and where to send newborns to specialists for necessary follow-up care, genetics and deafness, the appropriate surveillance for late-onset congenital hearing loss, as well as information on the cochlear implant and the criteria for candidacy).

In the past five years, the FIMP Audiology Network has performed Refresher courses on Newborn Hearing Screening for over 5000 pediatricians [1].

A principle concentration on Late Onset, Acquired and Progressive Hearing Loss is now a fundamental objective.

If Pediatricians do not intervene on a national level to identify late onset hearing loss (congenital) and do not perform that which is defined as "audiological surveillance", the screening thereby fails in its essence of being "universal" [2]. In fact, the Newborn Hearing Screening programs are not able to identify acquired or progressive hearing loss that may occur after the first months or years of life. The cases that represent an important percentage (20-30% of all cases of infant hearing loss) may only be identified through effective monitoring programs [3].

Italian Pediatricians are in the position to guarantee this level of surveillance.

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#### A57

##### **An effective collaboration between parent associations and medical professionals: The Italian Pediatric Federation Audiology Network**

Jodi M. Cutler<sup>1</sup>, Deborah Renda<sup>2\*</sup>, Giovanni Lenzi<sup>3</sup>, Stefano Berrettini<sup>4</sup>

<sup>1</sup>Affrontiamo la Sordità' Insieme, Grosseto, Italy; <sup>2</sup>Associazione Io Sento, Palermo, Italy;

<sup>3</sup>Pediatrician, FIMP (Italian Pediatric Federation), Grosseto, Italy;

<sup>4</sup>University of Pisa, U.O. Otorinolaringoiatria Audiologia e Foniatria

Universitaria- Pisa, Italy

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In the absence of a national newborn hearing screening program in Italy, parent associations have been working with the Italian Paediatric Federation (FIMP), the Society of Neonatology (SIN) and members of the Italian Society of Audiology and Phoniatrics (SIAP) to promote guidelines, best practice and training courses in early hearing detection intervention that incorporate sensitivity training for professionals working with families of deaf children. The establishment of the Italian Paediatric Federation's Audiology Network is the result of an international collaboration between parents and medical professionals designed to promote an effective model in developing Early Hearing Detection Intervention Programs (EHDI) that recognize the role of parents as partners in the process. Among other factors, one important component frequently underestimated in most early intervention programs, both in the USA and in other countries, involves the role of parental involvement within the EHDI process. [1] From screening to identification and intervention, families must navigate through medical institutions, government and private agencies, their own family construct and community and other dynamics when raising a child who is affected by hearing loss. Family support is the "map" that keeps this process moving effectively, with sensitivity to the social and emotional needs a family will have as it adjusts to its baby's diagnosis. The family is the social context into which children who are deaf/hard of hearing are born. The impact of a child's hearing loss affects not only the child, but the parents, siblings, extended family and community as well.

After five years of working region by region in Italy, the network recently held its first National Pediatric Course of Audiology where the parental voice was fundamental in providing sensitivity training and in recruiting the participation of pediatricians with patients diagnosed with hearing loss.

In the absence of a Regional Protocol for Newborn Hearing Screening in Sicily, the Association Io Sento offers assistance to families of children with hearing loss. The Association collaborates with health structures and agencies to offer resources that inform, educate and support families regarding diagnosis, school services, the cochlear implant, speech habilitation by using a network created by information technology.

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#### A58

##### **The diabetic ketoacidosis**

Francesca Cardella

Pediatrics Diabetology Unit Children Hospital "G. Di Cristina", Department of Pediatrics, University of Palermo, Palermo, Italy

E-mail: cardellafrancesca@gmail.com

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The Diabetic Ketoacidosis (DKA) is still today a medical emergency in pediatrics. Despite the latest great sensibilization of the population and the doctors, the risk of DKA has not yet been eliminated and this pathology is still occurring in 25 to 40% of diabetes onset cases, in already diagnosed patients with poor compliance (10%), in patients undergoing acute medical or surgical events or in patients in Continuous Subcutaneous Insulin Therapy (CSII). In toddlers (0-3 years) it is twice more frequent than in the following ages and is characterized by the presence of more serious clinical dehydration (>10%) and neurological signs (obnubilation 40%). The other category at risk is represented by teen-agers, who may suffer from DKA at diabetes onset (scarce vigilance or reticence on the problems), or in diabetes treatment when there is poor compliance. In affected patients, missed recognition can influence morbidity and mortality rates. Despite the improvement in DKA management and therapy, a lot of controversies have been encountered in literature. For the insulin therapy a wide consent exists on the need to use small doses of regular insulin for continuous intravenous administration (0.05-0.1 U/Kg/h). For children hydration the most recent recommendations are not to overcome 5-10 ml/Kg/h in the first two hours (max 250 ml/h: ISPAD-IDF 2011, ADA 2013) and to continue hydration slowly calculating the body surface area so as not to exceed 3 lt/mq/day (average 2000-2500 ml/mq/day). The careful controls of plasmatic electrolytes (opportune integrations particularly of potassium deficit: 20-40 mEq/lt, 50% of KCl + 50% of KPO4) and of glycemia are suggested (to avoid too rapid falls: when glycemia <250-300 mg/dl replace the sol. NaCl 0.9% N

with mixed sol. constituted by 50% of Glucose 10% sol. and 50% of NaCl 0.9% N sol.). The follow-up of clinical patient conditions and the EKG evaluation prevent rapid falls of kaliemia with well-known cardiac consequences.

The success of the treatment is nevertheless tightly connected to a correct management of rehydration, of metabolic acidosis and of electrolyte deficit replacement more than on insulin therapy, aimed at avoiding the most dangerous complication of DKA: cerebral oedema, that seems to be more frequent in patients with more severe onsets, particularly in those with low paCO<sub>2</sub> and high levels of urea nitrogen, but seems to be correlated also to the rapid administration of fluids and to the inadequate use of NaHCO<sub>3</sub>.

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#### A59

##### Acute enteritis: dehydration and rehydration in pediatric patients

Piera Dones

Pediatric Infective Diseases U.O.C., G. Di Cristina Hospital, Palermo, Italy

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Acute infectious enteritis are widespread globally. In developing countries, they are one of the leading causes of infant mortality. In developed countries, they account for a high number of outpatient visits and hospitalizations, with an economic impact on national health systems and, considering indirect costs, on communities. In roughly 70% of cases, the infections are caused by viral agents, principally rotavirus. A study conducted in collaboration with the Regional Referral Center for the Monitoring of Rotavirus Infection found that the serotypes isolated in all rotavirus-positive samples in the province of Palermo were 68% G1P8, and in 12.9% G2P4. In 20% of the cases, infectious gastroenteritis was caused by bacterial agents. At our hospital, the prevalent agents are salmonella spp. and campylobacter. For viral infections, there are no epidemiologic differences between developed countries and developing countries, though in the latter there is a higher mortality rate, associated with more difficult access to health care, malnutrition, and underlying diseases. Rehydration is first line treatment for pediatric enteritis. Indications for parenteral rehydration are generally quite restricted, and reserved for cases of severe dehydration, or cases in which oral rehydration solutions cannot be given either because of uncontrollable vomiting or because they cannot sufficiently restore electrolytes. A recent meta-analysis published in *Pediatrics* found that hypotonic pediatric solutions are the most widely used in hospitalized children. Use of these low-sodium solutions can lead to hyponatremia. Therefore, pediatric patients should always be rehydrated with isotonic solutions (0.9% NSS). For patients with hyponatremia, treatment should be slowly extended over a period of 24 hours, and sodium gradually supplemented in relation to total body water in order to avoid pontine myelinolysis, which is most frequent in dehydrated patients with severe underlying disease. However, it has also been found in otherwise healthy patients in whom treatment of hyponatremia was administered too rapidly. In fact, too rapid treatment can also lead to cerebral edema and/or seizures. Treatment should, in fact, be administered gradually over a period of 48-72 hours. Hypernatremic patients with hypovolemia clear more free water than solutes, so the total concentration of corporeal sodium is likely lower than that found in testing; therefore, treatment should also include isotonic fluids.

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#### A60

##### Management of red codes in the paediatric emergency room

Paolo Biban\*, Simona Spada, Davide Silvagni, Monica Benedetti, Chiara Ghizzi  
Paediatric Emergency Room, Department of Paediatrics, Azienda Ospedaliera  
Universitaria Integrata Verona, Italy

E-mail: paolo.biban@ospedaleuniverona.it

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**Background:** Paediatric triage is a process to evaluate the conditions of children on their arrival in the Emergency Department. The goal of triage is to rapidly identify patients who are compromised or at risk of deterioration, assigning priorities of intervention and providing immediate assistance. The "red code" is attributed to any patient with potential or imminent risk of death, for whom evaluation and treatment should be immediately provided.

**Contents:** Red codes are rarely assigned in the paediatric age. The main objective in these cases is to prevent death or brain damage in any patient with severe impairment of consciousness, breathing and/or circulation. Usually the patient is immediately received in a "shock room", fully equipped with advanced monitoring systems, devices for airway and circulation management, drugs, manual defibrillator [1]. Initial actions are based on the ABCDE approach, i.e. Airway, Breathing, Circulation, Disability and Exposure. State of consciousness, airway's patency, presence of respiratory arrest or insufficiency, arrhythmias or cardiac arrest must be quickly evaluated. In case of absence of vital signs, cardiopulmonary resuscitation (CPR) should be performed immediately by the attending healthcare providers, while seeking for additional help. CPR includes 100% oxygen, orotracheal intubation, chest compressions, early monitoring and defibrillation, intravascular or intraosseous access, drugs, fluids administration and temperature control. In case vital signs are present, every effort should be performed to avoid the progression toward cardiopulmonary arrest (CA). At the same time, the medical emergency team (MET) should be activated. The subsequent phase follows the algorithms for advanced management of the critically ill child, as recommended by international guidelines. Crucial steps are the recognition and treatment of potentially reversible causes of the CA, summarized in the "4 I": hypoxia, hypovolemia, hypothermia, hypo-hyperkalemia and the "4 T": cardiac tamponade, thromboembolism, toxic, tension pneumothorax. Fast-ultrasound may be useful for quantitative diagnosis of some of these causes. After the initial stabilization the child should be transferred to other units for secondary care. A mention should be given to the presence of parents during invasive manoeuvres and CPR. In fact, a number of studies demonstrate that their presence during resuscitation efforts, may be helpful for them, particularly in case of poor outcome.

**Conclusions:** Red codes to triage are attributed to children in cardiac arrest or imminent risk of death. Prompt management includes early recognition and treatment of cardiorespiratory insufficiency or cardiac arrest. Timely interventions and optimal quality of care may positively affect outcome.

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#### A61

##### Sonography only in emergency? The black and the white

Paolo Adamoli

Pediatric Department, Moriggia Pelascini General Hospital, Gravedona ed Uniti (Como), Italy

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The ultrasound transducer generates ultrasounds. It emits them, collects the return echo and allows the ultrasound system to re-create an image that (few people) know how to interpret.

The probe and the ultrasound system work together by measuring the liquid component of a tissue, as ultrasounds propagate easily through the water.

On sonography imaging liquids appear black because they are "anechoic". It means that the ultrasound wave goes through them without emitting any return echo.

Tissues containing a lot of water appear dark, or "hypoechoic", because the larger part of the ultrasound waves go through them while a small part of them is reflected by the tissues and returns to transducer.

Otherwise less hydrated tissues appear lighter, or "hyperechoic", because they reflect the largest amount of ultrasound.

On sonography, a solid tissue appears white with a black shadow below, as it reflects all the ultrasounds.

The air does not reflect ultrasound but scatters them back toward the probe. The return echo signals are interpreted as dispersion artifacts.

With these simple concepts, it is easier to interpret many ultrasound images.

The search of liquid effusions, anechoic, in the context of serous cavities after a trauma, in well known as FAST sequence and its evolution EFAST. This sequence is a versatile and valuable tool, also available for those physicians inexperienced in sonography.

**Table 1(Abtract A61)**

Pathological aspects in thorax ecography
1 Pleural effusion
2 Bronchopneumonia thickening
3 Pneumothorax
4 Complex pathological aspects (thymic ectopia, neonatal respiratory distress)
Pathological aspects in abdominal ecography with anechoic areas (serous effusions)
1 Ascitis
2 haemoperitoneum
3 hydronephrosis
4 hydrocele
5 Complex pathological aspects (renal cysts, splenic cysts, hepatic cysts, ovarian cysts, cystic duplications)
Pathologic abdominal sets with echogenic areas and acoustic shadow
1 Cholelithiasis
2 Hepatic calcification
3 Nephrolithiasis
4 Appendicolith

The FAST sequence is feasible anywhere (point of care) and without mobilizing the inpatient (bedside) or during CPR .

The versatility of this sequence has allowed many physicians without specific training or background to approach clinical sonography.

Ultrasound focused lung sequence (LUS) was proposed for respiratory disease, very frequent in pediatrics.

The presence of typical artifacts allow to detect a regular air intake in lung parenchyma near next to the chest wall. Their absence is a sure sign of respiratory disease.

A question naturally arises: why should we use ultrasound just in urgency?

There is no unambiguous answer, but after the last decade, the US scanner is becoming a valuable tool to further investigate the physical examination, especially in emergency department.

In Table 1 it summarizes that a basic sonography could be the chest examination in order to evaluate the air intake Simultaneously, a further application could be addressed to the research of anechoic areas and/or hyperechoic pathological patterns of abdomen, not just after trauma.

During the last year an interesting application was referred to the detection of skull fractures.

This easy and bedside procedure presents good sensitivity (88%) and high specificity (97%).

All of these procedures appear useful tools for the optimization of waiting times of diagnostic imaging in emergency (golden hour!) and for reduction of ionizing radiations use in pediatric patients.

## A62

### Radiation protection in paediatrics age

Sergio Salerno\*, Claudia Geraci

Dipartimento di Biopatologia e Biotecnologie Mediche e Forensi, Università di Palermo, Italy

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Radiological protection in paediatric population from medical imaging is a subject promoted by various international associations and it is becoming a main field of interest. Paediatric patients have a significant risk from ionizing radiation (IR) following X-ray examination (chest, abdomen and skeletal segments), multidetector CT (MDCT) and PET imaging [1]. Their greater damage's risk is due to: growing tissues with elevated turnover and high radio sensitivity cellular, the high water content which amplifies the damage, the small body size that involves the exposure of large areas

associated with the expectation of long- life makes possible the development of diseases resulting from genetic damage [1]. Also there is a huge increase of exposure due to imaging is recorded in many country also in Italy, in emergency and is mainly performed in "non paediatric hospitals" often with adult setting of the machine [2]. The first type of radioprotection is the use of alternatives imaging techniques; ETG and MR offer, in expert hands, a vital potential diagnostic in complete safety radio protectionist [3]. The radiologist who elects to perform the X-ray examination must be certain that this is justified and run with minimal doses for the same diagnostic benefit (As Low As Reasonably Achievable: ALARA). In these patients the X-ray studies is characterized by repeated examinations and this results in high doses even if only one exam does not deliver high levels of IR. MDCT examinations when strictly necessary, should be optimized "patient fit" in terms of scanning parameters mAs and Kw (according to age and weight of the patient the paediatric patient range from 700gr to 90 kgs), well collimation and only the region of interest should be examined and multiple sequence should be avoided [4]. Is a common idea that using paediatric protocols and alternative imaging techniques are important for reduce dose [4] and many authors suggested that the Size-Specific Dose Estimates (SSDE) received by the patients should be included in the patient electronic medical record; paediatric radiologist and clinicians can use the SSDE when assessing risk versus benefit for the child prior to performance of a scan MDTC. The extensive use of ionizing radiation in paediatric age needs a profound balance between possible risks and clinical advantage by referring clinicians and a patient fit protocols adjusted to age and BMI of the patient by the radiologist.

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## A63

### The metabolic screening today

Giovanni Sorge

Dipartimento di Scienze Mediche e Pediatriche, Unità Operativa Complessa di Clinica Pediatrica, Centro di Riferimento Regionale per la prevenzione, la diagnosi e la cura delle malattie metaboliche congenite dell'infanzia, Università di Catania, Italy

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Newborn screening is the process of testing newborn babies for inborn errors of metabolism that include disorders of amino acids, organic acids, fatty acids and urea cycle disorders.

Affected baby are usually normal at birth. Symptoms develop after a latent period ranging from few hours to many years. Many of these disease are potentially fatal conditions and some are extremely rare, but the delay or the lack of diagnosis and treatment can lead to developmental disability, mental retardation and premature death [1,2]

A significant percentage of these conditions (approximately 30-40%) is susceptible to dietary therapy, while in the remaining cases have been proposed various treatment strategies such as enzyme replacement therapy (ERT), vitamin supplementation, organ transplants, in addition to the use of new drugs with specific action.

The neonatal screening by tandem mass spectrometry (expanded newborn screening) allows the early detection of about 50 of these congenital disorders.

The advantages of this method consist in the speed of execution (1-2 minutes for each analysis), the ability to examine numerous analytes in a single spot of blood, the ability to recognize variants "mild" of numerous diseases and then to have updated epidemiological data, a favourable cost effectiveness ratio.

Among the disadvantages we must mention the false positive and false negatives. The first become negligible if the sampling is performed correctly and if the cut-off values of various analytes are re-evaluated

periodically, the latter can be attributed to technical errors (sampling time, cut-off or laboratory errors) or to the fact that some of these diseases can not yet be recognized by the tandem mass spectrometry [3]. Technological and therapeutic advances during the past 5-10 years have made possible a great expansion of neonatal screening, however, views about which disorder should be included ranging from the five disorders screened in UK to more than 50 recommended in USA [4].

In Sicily, a Regional project, has allowed us to begin the expanded newborn screening, dividing the Region into two areas, east and west, the first assigned to the "Centre for the prevention, diagnosis and treatment of congenital metabolic diseases" of Catania and, the other, to the "Screening Centre" of Palermo. The project includes screening for 30 diseases.

There is a large debate in the scientific literature on the opportunity to introduce neonatal screening for other congenital diseases such as, for example, the lysosomal storage diseases [4-7]

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#### A64

##### Enzyme replacement therapy (ERT) in pompe disease

Agata Fiumara

Centro Riferimento Regionale per le Malattie Metaboliche, UO Clinica Pediatrica, Dip. Scienze Mediche e Pediatriche, Università di Catania, Italy  
*Italian Journal of Pediatrics* 2014, **40(Suppl 1)**:A64

Pompe disease (OMIM 232300) is an AR glycogenosis due to deficiency of the lysosomal enzyme alpha-glucosidase (GAA). As a result, glycogen storage occurs in muscles and patients present a wide clinical spectrum ranging from early onset severe cardiomyopathy (EOPD) to adult onset forms (LOPD). Severe loss of GAA activity correlates with early onset and severe phenotypes. Residual enzyme activity warrants later onset and a slowly progressive course, although a strict correlation is not always observed. Patients with Classic infantile Pompe disease show hypotonia, macroglossia, respiratory insufficiency and early death for cardiorespiratory failure.

The availability of enzyme replacement therapy (ERT) with alglucosidase alfa (human recombinant GAA) has changed the natural history of the disease allowing most children with EOPD a longer survival and a longer preserved muscle performance with increased quality of life in LOPD.

The experience of the Regional referral Center for Metabolic Diseases, University of Catania, concerns 11 patients (9 months to 65 years at the time of the diagnosis). Their clinical and laboratory follow-up included eye tracking and Muscle MRI.

EOPD, with a severe clinical picture since the first weeks of life and cardiomyopathy was observed in a newborn who died at age 4 months, despite early started and high dosage ERT. Another girl with EOPD is on ERT since the age of 9 months and, today, after 7 years of treatment shows only mild signs of muscle weakness and no cardiac involvement.

Five more LOPD patients on ERT had a significant improvement of blood parameters including transaminases, LDH and CPK. No progression of muscle impairment was seen after 5 to 8 years of treatment, except for one 35 years old man, who was already severely compromised and did not show any benefit in terms of respiratory ventilation.

In a recently performed Italian survey of classical Pompe disease treated with ERT, a substantial improvement in cardiac parameters and gross motor function was observed, even if residual muscle weakness still exist and gross motor function remain below age-appropriate levels.

The different response to ERT depends on several factors mainly on timing of treatment (worse outcome if started later than 5 months of age) and cross reactive immune material (CRIM-negative status associated with less good response to ERT).

Although some residual mild weakness is present in treated patients, ERT has demonstrated its effects on respiration, lowering nocturnal apneas and allowing patients a better quality of life.

#### A65

##### Identification of risk and health budget

Antonina Lo Cascio

Pediatra di Famiglia Palermo, Docente Scuola Specializzazione Pediatria Università di Palermo, Italy

E-mail: nettilocascio@aliceposta.it

*Italian Journal of Pediatrics* 2014, **40(Suppl 1)**:A65

The Language is a specific competence of human species that develops in a relatively short period of the life. Indeed, most of the linguistic rules are learned by the child within 4-5 years of life. In his firsts months of life, through the interaction with adults, the baby acquires the first signs of communication through the eye, sounds, facial expression, and response to the environmental stimulus that surrounds him.

Assessment of health status ("Bilancio di Salute"), regularly carried out by the primary care Pediatricians, are used to carry out surveillance longitudinal neuro- evolutionary development of the child through the integration of its communicative and linguistic, auditory, motor and visual-spatial skills. Since the sixth month of the child's life is possible to detect the risk indicators that deserve a very early assessment, as long as it has been excluded, through screening with otoacoustic emissions effected in birth centers, a hearing disorder.

In the first three years of life, the presence of an atypical development of language should be underlined by Pediatricians and parents, because it could not be just a temporary condition related to individual variability.

The "wait and see" approach can strongly affect the social life and learning at school.

The age of three is a sort of watershed between the so-called "late talkers" and children with a specific language disorder, provided that the child understands the language of the adult. Lowering the age of first consultation is recommended for increase the possibility of early detection, and then an early rehabilitative intervention, leading to an improvement in the prognosis of the pathology [1]. A delay in the acquisition of language can be an indicator of mild intellectual disability, that is often recognized only after the beginning of school activities.

An article published in May 2014 [2] aims to evaluate the association between the pragmatic skills of language and behavioral problems in a group of 40 adolescents who had experienced childhood speech problems. Longitudinally evaluation at the age of 7-9 years (T1) and at the age of 12-15 years (T2) was made with a control group (37) of typically developing.

The results obtained prove that the atypical group shows a deficit of linguistic, and that emotional problems, language and difficulties with peers at 7-9 years are closely related with the pragmatic difficulties in adolescence, and that they affect in a negative way social relations.

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## A66

### Language and movement disorders: early risk identification

Francesca Pardo\*, Elena Vanadia

"Io Comunico" Rehabilitation Center, Partinico (PA), Italy  
Italian Journal of Pediatrics 2014, **40(Suppl 1):A66**

Psychomotor development is a maturational process that in the early years of life enables the child to acquire postural, motor, cognitive, communicative and relational skills and abilities. It is a continuous progress, essentially dependent on the maturation of the central nervous system (CNS), with variable timing and conditions for each child, but where is possible to identify the "stages" that are achieved in a similar sequence [1-6]. Knowledge of this "normal" development is needed to identify the early indicators of risk and to enable a project of "Care" which has as its objective the support of the child and its potential. What was said can only occur in a network that sees Care and Rehabilitation Centers, Hospital and Territory, Pediatricians, School and Parents in close collaboration.

The therapeutic alliance Pediatrician-Child neurologist and psychiatrist-Parents is a key element for the evaluation and monitoring of the neurobehavioral development of the child and for the opportunity to do prevention. Through early diagnosis passes the opportunity to identify action habilitation / rehabilitation plans involving effective child and family, improving the knowledge of the natural history of specific neurodevelopmental disorders; ultimate goal is to guide the institutions on health policy interventions that enable an effective and efficient operation of the network [2].

The pathologies of language and movement are a "range of possibilities" with different gradations in the expressiveness, mirror of developmental system and brain mapping that underlies it, as well as the genetic control, we often see in comorbidity in different "individual trajectories". Therefore about Autism Spectrum Disorders, specific learning disorders or dyspraxia, is to identify a large chapter with transitional symptom patterns that change in attitude and somehow coexist with variable expressivity in relation to the multigenic neurodevelopmental and environmental control that modulates them [3,4]

Is appropriate a reflection on broad spectrum screening models that, from the identification of early indicators of risk in the first years of life, allowing us to change the traditional model of rehabilitation of the child in relation to neurobiological model and individual evolutionary characteristics whose language and movement are an expression [5].

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## A67

### The autism spectrum disorders (ASD)

G Gambino

Head of Surgery HUB 2nd level for Intensive Early Diagnosis and Treatment of Autistic Syndromes, UOC NPIA ASP Palermo, Italy  
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Screening within the 2nd year of life ( primarily between 12 and 18 months ) is a tool for early detection of the risk of a neurobehavioral

developmental disorder and is a chance to change the term prognosis of ASD.

The Analysis of the interactions of functional and dysfunctional interactive communicative signals from the very early ages of life, as well as of motor patterns , allows us to capture the significant elements of the neuro-behavioral development in children at risk of developmental ASD.

The ASD as the primary difficulties of social orientation and inter-subjectivity, which leads to not meeting the needs of dyadic relationship becomes even cognitive deficits, given that the abnormal behavior and the failure to derive anomalies early experiences of the process of neuronal growth.

The diagnostic label of ASD in the transition from DSM-IV TR DSM 5: basically goes from one classification categorical one dimensional paintings that defined and distinct switching to the concept of spectrum as a continuum of clinical features (anomalies qualitative different degree of movement and the axis linguistic communicative) until the extension to the poles towards frameworks neurodiversity.

The spectrum autism disorders in relation to the clinical expression of the core autism ( social and communicative disorders and restricted interests and repetitive, stereotyped ) occurs at three levels of severity , in line with the possibilities prognostic constitute the consideration for the needs and welfare of the degree of complexity in the "care" the person affected by ASD and his family.

There is no genetic marker of autism but there is a multi- gene and environmental control of the clinical expression of the responsible individual developmental trajectories of endophenotypes.

Developments in neuroscience constitute the antecedent of the models of cognitive behavioral treatment oriented, validated for the effectiveness in accordance with the Guidelines ISS. If activated at an early stage, it is more effective than subsequent treatments, resulting in significant improvements in language, IQ, and adaptive behaviors.

## A68

### Tourette syndrome: current data, comorbidities, and therapeutic approach in children

R Rizzo<sup>1,2</sup>

<sup>1</sup>Child and Adolescent Neurology and Psychiatry, Department of Medical and Pediatric Science; <sup>2</sup>Catania University, Via S.Sofia 78 Catania, 95123 Italy  
E-mail: [rerizzo@unict.it](mailto:rerizzo@unict.it)

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Tourette syndrome (TS) is the primary tic disorder with an estimated prevalence close to 1% between 5 and 18 years of age [1]. Motor and phonic tics are the core features of TS [2]. In addition to their well-characterized phenomenology, tics display a peculiar variability over time, which is strongly influenced by a variety of contextual factors. A relevant proportion of patients with TS display complex, tic-like, repetitive behaviors that include echophenomena, coprophenomena, and nonobscene socially inappropriate behaviors (NOSIBs). Co-morbid conditions are attention deficit hyperactivity disorder (ADHD), obsessive compulsive behaviours/disorder (OCB/D) and autistic spectrum disorder (ASD); co-existent psychopathologies include depression, anxiety, oppositional defiant disorder (ODD), conduct disorder (CD) and personality disorders (PDs) [3]. The complexity of the Tourette spectrum has been confirmed by cluster and factor analytical approaches [4]. It is suggested that TS is not a unitary condition and that one phenotype ("Pure TS" [tics only]) occurs in about 10-14 % [5]. The presence of comorbid attention deficit hyperactivity disorder (ADHD) is the main determinant of cognitive dysfunction in TS patients and influences heavily also the risk of developing disruptive behaviors [6]. The burden of behavioral comorbidities is very important in determining significant impairment, poor self-esteem, and a low quality of life [7,8]. While the evidence for a genetic contribution is strong, several genes, including *SLITRK1*, *LIM homeobox (LHX6, LHX8)*, and *HDC* have been suggested to be responsible for the different clinical phenotypes [9,10]. However its exact nature has yet to be clarified fully. Aetiological factors include genetic vulnerability pre- and peri-natal difficulties (PNDs), and probably neuro-immunological factors. Neuro-imaging are helpful to exclude other conditions, and although abnormalities are described, in an individual patient, they are not diagnostic. Treatment includes psycho-education and reassurance, medications, target-specific botulinum toxin injections [11] and in a few

severe refractory adult cases, deep brain stimulation life [12]. This review will summarise and highlight selected main findings from the author's clinic.

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#### A69

##### Early infantile epileptic encephalopathies

Alberto Fois  
Siena, Italy

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Epileptiform abnormalities contribute to progressive deterioration of cerebral function. Considered: Ohtahara Syndrome; Early myoclonic epileptic encephalopathy; West Syndrome; Dravet Syndrome; Myoclonic status in not progressive encephalopathies; CDKL5 encephalopathy.

**Ohtahara syndrome (OS)** early infantile encephalopathy (EIEE). Most cases linked to cerebral malformations or very occasionally to metabolic disorders. Main seizures: tonic spasms, tonic-clonic, myoclonic and atonic and partial spasms. Treatment Vigabatrin, Topamax, Zonigran, Steroids, ACTH and Ketogenic diet, vagal stimulation and more invasive surgery. Mutations of the gene STBX1 (Sintaxin binding protein) Mutations of SPTA gene and GC1 have been reported. Prognosis is poor.

**Early Myoclonic Encephalopathy (EME)**: Onset: neonatal period or first months of life. Seizures mainly erratic, myoclonic or partial, tonic spasms. EEG burst suppression. Etiology mostly unknown. Inborn errors of metabolism as nonketotic hyperglycinemia, or propionic acidemia reported. Prognosis poor. Therapy similar to that for OS.

**West Syndrome (WS)** aka as infantile spasms with hypsarrhythmia, EEG abnormality with asynchronous very high amplitude, irregular, continuous multifocal spike in slow wave discharges. In 80% of cases a cause can be found or suspected (symptomatic or cryptogenetic) In about 20% of

patients no cause can be found (idiopathic). 1:3200/1:3500 newborns. Causes: infections, cerebral malformations, hypoxic ischemic injuries, metabolic and genetic disorders, tuberous sclerosis. Corticotropin considered effective. This finding later confirmed. No definite therapeutic scheme. High dose and long term treatment associated with hypertension, cushingoid features and ipokalemia. Vigabatrin and Topiramate useful. Zonisamide, Lamotrigine and Levetiracetam can be used. Prognosis much better in idiopathic cases when treatment is started within the first month from the appearance of the symptoms.

**Myoclonic status in nonprogressive encephalopathies**. Rare, onset in the first years of life. Partial motor seizures, myoclonic absences or massive myoclonias sometimes with startles. Interictal EEG epileptiform discharges and background slowing. Described in genetic conditions like Angelman and 4p – syndromes. Prognosis poor.

**Dravet syndrome (Severe myoclonic infantile epilepsy or SMEI)**. 1/500 cases of infantile epilepsy. Febrile convulsions in the first year. Subsequently myoclonic seizure with fever. Photosensitivity. In 70% of cases mutations of SCN1A gene. Difficult to treat. Topiramate, Clobazam, Stiripentol.

**CDKL5 Encephalopathy**. Mutations in the human X linked cyclin-dependent kinase like 5 (CDKL5), infantile spasms and Rett (RTT) like phenotype. Initially normal EEG and severe hypotonia. Acquired microcephaly and hand stereotypes. Identified with early infantile epileptic encephalopathy 2. Prognosis severe.

#### A70

##### Complementary medicine and chronic disease: preliminary results

Gianfranco Trapani<sup>1</sup>, Luisella Zanino<sup>1</sup>, Gianna Gabbanelli<sup>1</sup>, Stefano Gandus<sup>1</sup>, Maurizio Annibali<sup>1</sup>, Pietro Prandi<sup>1</sup>, Gaetano M Micciché<sup>1</sup>, Maurizio Calzavara<sup>1</sup>, Domenico Careddu<sup>2</sup>, Sara Griseri<sup>2</sup>, Mariuccia Ventura<sup>2</sup>, Isabella Villa<sup>2</sup>, Illyria Sbizzera<sup>3</sup>, Gianpaolo Salvioli<sup>4</sup>

<sup>1</sup>Pediatra - Società di Medicina Bioterapica - SMB Italia, Roma, Italy; <sup>2</sup>Pediatra di Libera scelta, Italy; <sup>3</sup>Dottore in Scienze Statistiche, Verona, Italy; <sup>4</sup>Professore Emerito Di Pediatria all'Università di Bologna, Bologna, Italy

E-mail: gf.trapani@tin.it

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**Background:** The Study Group in Complementary and Alternative Medicines of the Italian Society of Pediatrics (SIP) has an ongoing study to assess how the relationship between families and pediatricians, who also prescribe Complementary and Alternative Medicine (CAM) affect on therapies of children with Chronic Diseases (CD) The study, which began in January 2014, will end in July 2014.

**Materials and methods:** Pediatricians administered a questionnaire to families with children affected by CD when they go into ambulatory. The questionnaire assesses the role of CAM requested by the family with regard to the natural history of the disease and the possible Adverse Effect (EA).

They came to our observation (March 2014) 121 Questionnaires. Among these, 109 (90.1%) were found to be suitable for the study. The diseases most frequently identified were: diabetes, inflammatory bowel disease (IBD), cancers, celiac disease, rheumatic diseases, disorders of conduct (DC) such as ADHD, autism, and other DC, cystic fibrosis, and epilepsy.

In addition, 27 families had 36 children (33.0% of cases) affected by other diseases, among them S. Peutz Jeghers, S. Kartegener, primary TBC complex, visually impaired ocular albinism (X gene locus 22), acute lymphoblastic leukemia with removal of colon and rectal anastomosis ileum, Beta sarcoglycanopathy, May Hegglin syndrome (hereditary thrombocytopenia).

**Results:** The families who consulted a pediatrician expert in CAM and conventional therapies (CT) have associated the two therapeutic techniques in 80, 7% (88 of 109 cases). 19.3% (21 of 109 cases) decided to leave temporarily or permanently therapies that followed, replacing them with the CAM. Of the 21 families who have abandoned CT, 13 (61.9%) had children with ADHD, autism and other DC, others have declared that have not been proposed therapies, but recommended "watchful waiting" (IBD, autoimmune inflammation of the thyroid, thrombocytopenia).

Of the 109 cases, 31.1% of parents [1] used CAM to enhance the effect of conventional therapy on the disease. 30.2% were not satisfied with the results obtained with the TC on the disease. 37.6% preferred to use CAM during acute illnesses not to take too many drugs.

**Conclusion:** Unlike the literature [2,3] No EA was observed for CAM use in 109 cases observed

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**A71**

**Fluoride therapy in the prevention of dental caries**

Laura Strohmer

Department Health's Science, H.San Paolo, Milan, Italy

E-mail: laura.strohmer@unimi.it

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The document "National Guidelines for the prevention and oral health promotion in childhood", 2013, is an act of address for those involved in the management of oral health and in particular to the pediatrician, because the figure of the pediatrician in the prevention of oral health is absolutely critical, as confirmed by the entire international literature in recent years. Correct attitudes and behaviors adopted since childhood that will allow the child to protect his health.

The fluoride is the cornerstone of prevention of tooth decay and is required for all individuals.

Over the years, have been developed different means of administration of fluoride, each with different strengths, dosages and frequency of use [1-5]. Fluoride supplements should be prescribed by the pediatrician in cases of real difficulty for topical administration of fluoride through toothpaste or fluoride added as a method of in subjects at risk of tooth decay [6].

The decline of caries in our country it is highly likely also due to the pediatrician who, using the national guidelines, can inform parents and families induce the acquisition of preventive behaviors currently defined by scientific research [7].

To this end, again, the ministerial guidelines on the subject that were reviewed by a team of experts representative of the Italian research in this area.

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**A72**

**First aid in dental trauma in pediatric age**

Stefania Zampogna<sup>1\*</sup>, Salvatore De Filippo<sup>1</sup>, Valentina Talarico<sup>2</sup>, Monica Aloe<sup>2</sup>, Nadia Severini<sup>2</sup>, Sivia Pizzi<sup>3</sup>, Maria De Filippo<sup>4</sup>, Antonella Polimeni<sup>4</sup>

<sup>1</sup>Azienda Ospedaliera "Pugliese-Ciaccio", Catanzaro, Italy; <sup>2</sup>Università "Magna Graecia" di Catanzaro, Catanzaro, Italy; <sup>3</sup>Università degli Studi di Parma, Parma, Italy; <sup>4</sup>Università degli Studi Roma La Sapienza, Rome, Italy

E-mail: stefaniazampogna@teletu.it

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The knowledge of the right management of dental traumas is very important for the pediatrician due both to their high incidence in this age group and to prevent further pathologic events related.

Several studies indicate that in industrialized countries, about one in five children have had a traumatic dental injury to permanent teeth before leaving school. Prevalence of injured teeth presented in the literature varies from 10 to 51% [1]. The pediatrician, more than any other health professionals, should have the necessary knowledge to ensure correct and professional advices for all issues concerning the child's health [2]. Most of the available literature emphasizes that awareness of the correct procedure following dental trauma is unsatisfactory. It is recognized that the prognosis of traumatic dental injuries is dependent on the time between the injury and the initiation of treatment.

Emergency dental treatment by a physician is sometimes required when a dentist is unavailable, so the first physician that comes to managing dental trauma is often the emergency room doctor or the family or hospital pediatrician. Some studies' findings suggest, however, that only 4% of physicians would provide an appropriate initial treatment that could help to save an avulsed tooth [1]. Even in medical courses and first-aid training, management of dental trauma is seldom covered. To ensure and facilitate the approach to this problem, we proposed an evaluation form of dental trauma (figure 1) in order to clearly identify the points that need more attention in presence of a dental trauma and the next steps to be performed for a complete and correct clinical management [3]. First step: initial evaluation of the child, subjective information (interview, where, when, how), evaluation of vital parameters and following attribution of a color code for the priority of access to the medical examination. Next steps are the medical examination, the identification of any cranial-cervical trauma and/or signs of abuse and finally the pulpal tests.

We have proposed a multicenter study in order to evaluate the adhesion and the application of the National Guidelines for the Prevention and Clinical Management of Dental Trauma in children, published in

**EVALUATION FORM OF DENTAL TRAUMA IN PEDIATRIC AGE**  
**FIRST AID**

Date   /  /  

Surname                      First name                      Date of Birth   /  /  

Sex: M F

• Place where trauma happened:  Home  School  Park  
 Other                     

• Trauma dynamics:  Fall  Game  Sports Other                     

• Time passed by trauma:  <30 min  30 min-1 hour  1hr - 2hrs  >2 hrs

• Symptoms after trauma:  Pain  Limitation in opening/closing mouth  
 Bleeding Other                     

• Has any tooth been interested?  Primary tooth  Permanent tooth

**UPPER TEETH**

**RIGHT**      **LEFT**

**LOWER TEETH**

Figure 1(abstract A72) Evaluation form of dental trauma



November 2012 by the Ministry of Health. Our goal is to verify and analyze the level of knowledge of various professionals (doctors of emergency room, family pediatricians, hospital pediatricians and dentists) about the rules to prevention of dental trauma and health education, the first aid of dental trauma and recognition of dental trauma in the child abuse.

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#### A73

##### Gastroesophageal reflux disease and oral manifestations

Claudio Romano<sup>1</sup>, Sabrina Cardile

Department of Pediatrics, University of Messina, Messina, Italy

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Gastroesophageal Reflux (GER) is a common condition in childhood characterized by the rise of gastric contents into the esophagus. According to the International Consensus of the Montreal, gastroesophageal reflux disease (GERD) is defined as "the condition that develops when the retrograde passage of gastric contents causes troublesome symptoms and/or complications that result in an impairment of the quality of life of these patients" [1]. In pediatric population, there are conditions at risk of developing GERD, like neurological impairment, history of esophageal atresia repaired and obesity [2]. The typical symptoms can include heartburn with or without regurgitation. For extra-esophageal syndromes, only dental erosions and Sandifer syndrome are considered conditions related to GERD. Acid reflux at the level of the oral cavity, in fact, can cause the dissolution of the tooth enamel, especially at the level of the palatal surfaces of the back teeth, with a reported prevalence of up to 42% [3]. In a pediatric cross sectional study, in 112 children was found a significant incidence for dental erosion in patients with GERD respect to control group, both in primary and permanent teeth [4]. In general, however, oral manifestations of GERD are reported mainly. In pediatric population, the dental erosion are not considered primary extra-esophageal manifestation of GERD because, when present, can be associated at multiple factors [5]. The typical manifestations can be considered dental caries, dry mouth, feeling at oral acid/burning sensation, halitosis, erythema of the palatal mucosa and uvula. For diagnosis is mandatory exclude other causes, like dietary factors, drugs, poor oral hygiene, eating behavior disorders, genetic and racial factors. The esophageal pH monitoring and/or endoscopy are usually necessary just to confirm the diagnosis of GERD. In this group of patients it's possible to start a pharmacological therapy in association with modifications of the diet (quantity and frequency of intake of foods and reduction of the beverages that contain fat, sugars and acids) to decrease the time of exposure at gastric acid and secretions. Therapeutic options are based on the medical treatment or surgery in severe cases [6], although there are few studies to evaluate the efficacy of the treatment of GERD to prevent oral cavity lesions.

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#### A74

##### Orthodontic problems in pediatric and growing subjects

Paola Cozza<sup>1</sup>, Roberta Lione

Department of Clinical Sciences and Translation Medicine, University

"Tor Vergata", Rome, 00133, Italy

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**Background:** In literature, the most effective timing for intercepting skeletal malocclusion is still widely debated. The aim of this study was to analyze the main orthodontic and occlusal problems needing correction in growing patients.

**Materials and methods:** Crossbite, open bite and Class III malocclusion were observed and compared each one with their own control groups with the same age and dentoskeletal characteristics. The statistical comparisons between the test and control groups were performed with independent sample t tests and chi-square tests (P <.05).

**Results:** The presence of posterior crossbite was significantly greater in growing subjects with oral breathing thus confirming the influence of these factors on skeletal development with constriction of the whole palate. Unilateral posterior crossbite is often associated with mandibular lateral displacement that is clinically characterized by deviation of the chin, facial asymmetry, dental midline discrepancy, and high prevalence of internal derangement of the temporomandibular joint. Children with anterior openbite presented with a greater prevalence rate of sucking habits. Hyperdivergency is a risk factors for negative overbite in mixed dentition subjects. The test group had significantly smaller maxillary intermolar and intercanine widths and greater posterior transverse discrepancy. Class III malocclusion is a complex clinical entity that entail the contraction of the maxilla and a narrowing of the base of the nose in addition to an increased mandibular total length.

**Conclusions:** Treatment of orthodontic problems that do not improve with age may be started earlier to avoid worsening of the condition in permanent dentition. The objective of any treatment in pediatric subjects before eruption of all permanent teeth are to correct the skeletal discrepancy between the jaws and improve function and facial esthetics by allowing them to develop normally, to create an ideal overbite and overjet relationship, to align the anterior permanent teeth and reduce the chance of trauma to these teeth, to improve the width of the dental arches and to reduce the risk for extraction of permanent teeth and for surgery in severe cases.

Morphologic and functional characteristics of unilateral posterior crossbite with mandibular lateral deviation should be clarified to correct and prevent this malocclusion. Children with mouth-breathing pattern and sucking habits showed a significant constriction of the maxillary arch and an increased palatal height when compared with control group. Early treatment of Class III malocclusion is able to produce significant and favorable long-term skeletal shape changes characterized by an anterior morphogenetic rotation of the mandible.

#### A75

##### Asthma and air pollution

Giuliana Ferrante<sup>1</sup>, Roberta Antona<sup>2</sup>, Velia Malizia<sup>2</sup>, Laura Montalbano<sup>2</sup>, Stefania La Grutta<sup>1,2\*</sup>

<sup>1</sup>Dipartimento di Scienze per la Promozione della Salute e Materno-Infantile "G. D'Alessandro", Università degli Studi di Palermo, Palermo, 90133, Italy;

<sup>2</sup>Istituto di Biomedicina e Immunologia Molecolare- IBIM "A. Monroy",

Consiglio Nazionale delle Ricerche, Palermo, 90146, Italy

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During the last decades research all over the world has highlighted the deleterious effects of pollution on respiratory health of adults and children. Nevertheless, air pollution still represents a significant threat to health. Children are more sensitive than adults to pollutants for several factors: increased respiration relative to body size; physiologic immaturity of respiratory and immunologic systems; low metabolic capacity; longer life expectancy.

Several studies demonstrated an association between exposure to outdoor pollutants and respiratory diseases in childhood. Outdoor

pollutants, such as nitrogen oxides (NO<sub>x</sub>), particulate (PM), carbon monoxide (CO), carbon dioxide (CO<sub>2</sub>), ozone (O<sub>3</sub>), sulfur dioxide (SO<sub>2</sub>), may provoke cytotoxic and functional damages in the airways through oxidative stress and inflammation. During the last decades the amount of pollutants from vehicular traffic has significantly increased, especially in urban areas. Recent epidemiological studies have shown that vehicular traffic represents the main source of outdoor pollutants and that it may increase the risk of respiratory outcomes (cough, phlegm, wheeze, asthma) in children through short-term and long-term effects on airways, lung function and allergic sensitization [1].

Indoor pollution is also particularly dangerous, mainly for children and adolescents, that typically spend most of the time in confined spaces (home, school and public spaces). Indoor pollutants concentration depends on external environmental pollutants filtered inside buildings, pollutants generated inside buildings (domestic work) and pollutants generated by personal activities. Combustion products (tobacco smoke and wood burning), CO, CO<sub>2</sub>, volatile organic compounds (VOC), microbial agents (fungi and bacterial endotoxins), organic products (pet derived and mite allergens, dampness, mold derived components) are the most important indoor pollutants. There is growing epidemiological evidence that indoor allergen exposure may contribute to the development of allergic respiratory symptoms, such as wheezing, coughing and asthma in children [2].

Tobacco smoke is one of the environmental pollutants influencing morbidity and death rate in childhood as it is responsible for adverse health effects in both prenatal and postnatal life. Homes remain a site where children are dangerously exposed to environmental tobacco smoke (ETS). The combination of tobacco smoke pollutants which remain in an indoor environment, the so-called 'third-hand smoke' (THS), represents a new concept in the field of tobacco control [3].

Children still need to be protected with strict air quality standards, in order to improve their respiratory health. Therefore, policies that ensure better air quality are strongly desirable all over the world.

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#### A76

##### Mycoplasma infections in children: an update

Anna R Cappiello<sup>1</sup>, Maria F Mastrototaro<sup>1</sup>, Mariacristina Pignatelli<sup>2</sup>, Gianluca Piccolo<sup>2</sup>, Iolanda Chinellato<sup>3</sup>, Fabio Cardinale<sup>4</sup>

<sup>1</sup>Scuola di Specializzazione in Pediatria, Università degli Studi di Bari "A. Moro", Bari, 70121, Italy; <sup>2</sup>Università degli Studi di Bari "A. Moro", Bari, 70121, Italy; <sup>3</sup>Unità Operativa di Pediatria e Fibrosi Cistica, Ospedale "Tatarella", Cerignola (Foggia), 71042, Italy; <sup>4</sup>Unità Operativa Complessa di Pediatria, Servizio di Pneumologia e Immuno-allergologia Pediatrica, Azienda Ospedaliero-Universitaria "Policlinico-Giovanni XXIII", Bari, 70126, Italy  
*Italian Journal of Pediatrics* 2014, **40(Suppl 1)**:A76

*Mycoplasma pneumoniae* (MP) is a human bacterium that lacks a cell wall and is adapted to life as an obligate pathogen in the respiratory tract [1]. MP is responsible of 20-40% of all community acquired pneumonia (CAP) and is often associated with other airway disorders. Extrapulmonary manifestations are supposed to be sequelae of primary MP infections [2]. MP is believed to first act as an extracellular parasite. However the microbial factors responsible for the observed host cell injury have not been satisfactorily determined [3]. In 2006, a 68-kDa protein CARDS-Tx has been identified [2] from Kannan et al. demonstrates that CARDS-Tx concentrations in BAL of mice inoculated intranasally with three different MP strains, was directly linked to the ability of specific MP strains to colonize, replicate, persist and elicit lung histopathology damage [3]. Furthermore Medina et al. found that CARDS-Tx can induce increased

expression of IL4, IL13 and Th-2 chemokines, causing cellular inflammatory response, mucus metaplasia and increase in airway hyperreactivity [4].

On the basis of this observation some authors suggest the use of systemic steroids in order to diminish the host response in MP infection. Observational data, also, indicate that the addition of systemic steroids to antibiotics may improve the outcome of severe MP pneumonia.

Tagliabue et al. demonstrates, in mice, that combination therapy with clarithromycin and dexamethasone is more effective in reducing MP induced pulmonary inflammation than either clarithromycin alone or dexamethasone alone. The authors, however, conclude that more controlled clinical studies in humans are necessary [5].

Macrolide (ML) are recognized as first-choice agents for MP infections. In 2000, however, MP showing resistance to macrolides was isolated from clinical samples obtained from Japanese children with CAP. Since then, prevalence of ML resistant MP isolates in pediatric patients has increased rapidly worldwide.

ML inhibit MP protein synthesis by binding to domain V of 23S rRNA at nucleotide positions 2063 and 2064. Mutations at A2063 or A2064 confer the highest resistance to these antimicrobials [6].

On the basis of studies of the prevalence of ML resistance Principi et al suggest that in countries with low incidence of ML-resistant MP strains no change in ML prescription is initially needed. Nevertheless, in countries in which ML-resistant MP strains are very common, replacement of a ML with a tetracyclines or fluoroquinolones should be considered also based on the severity of the disease [7].

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#### A77

##### Risk factors for asthma

Michele Miraglia del Giudice<sup>1\*</sup>, Annalisa Allegorico<sup>1</sup>, Giuseppe Parisi<sup>2</sup>, Francesca Galdo<sup>1</sup>, Emilia Alterio<sup>1</sup>, Amalia Coronella<sup>1</sup>, Giuseppina Campana<sup>1</sup>, Cristiana Indolfi<sup>1</sup>, Natja Valenti<sup>1</sup>, Sonia Di Prisco<sup>1</sup>, Serena Caggiano<sup>1</sup>, Nunzia Maiello<sup>1</sup>

<sup>1</sup>Department of women and child and general and specialized surgery, Second University of Naples, Italy; <sup>2</sup>Rizzoli Hospital of Ischia, Naples, Italy  
E-mail: michele.miraglia@unina2.it

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Asthma is the most common chronic respiratory disease of childhood, and even if there have been many advances in the understanding pathogenesis of the disease, many aspects remain to be clarified.

In the pathogenesis of asthma are involved both "protective" and "predisposing" factors as a result of the complex interactions that occur between genetic predisposition and environmental exposure.

From the genetic point of view, the identified genes responsible are more than 100, and many polymorphisms have been shown to be associated to the onset of asthma, although none of these, alone or in combination, is able to predict the occurrence of disease.

The environmental factors most involved in the onset of asthma in children are represented by allergens, tobacco smoke, respiratory infections and air pollution.

Indoor allergens (dust mites, mold and animal dander) and outdoor (pollens and molds) are able to induce sensitization by prolonged exposure and trigger acute asthma. Allergic sensitization, in the concept of atopic march, represents a major risk factor for the development of asthma. In particular, the subjects polysensitized and with food allergy may present more severe asthma [1].

The exposure to cigarette smoke in both prenatal and postnatal increases the risk of the child becoming asthmatic and the asthma severity.

It has also noted recently that obesity is a risk factor for asthma because it causes an increase of leptin, TNF- $\alpha$ , and IL-6, which exert a pro-inflammatory non-eosinophil action [2]. In addition, the lack of physical activity, for weight gain, contributes to the determinism of the disease [3]. Vitamin D is involved in the processes of development and fetal lung maturation; the levels of 25-OH vitamin D from umbilical cord blood are inversely correlated with the risk of respiratory infections and wheezing in childhood [4]. The vitamin D has immunomodulatory properties exerting an action of inhibiting the production of pro-inflammatory cytokines and induction of the synthesis of antimicrobial peptide on cells of the innate immune system [5]. The vitamin D modulates the effects of glucocorticoids and also has a role in bronchial remodeling, as it regulates the expression of genes of bronchial smooth muscle.

Infections early in life may play a role of "induction" of wheezing or "protection" against the development of allergic diseases (according to the hygiene hypothesis)[6-8]. In infants at risk viral respiratory infections can cause wheezing, which in turn can evolve later in asthma particularly in individuals with atopic predisposition.

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#### A78

##### Molecular allergology

Roberto Bernardini\*, Vincenzo Ragazzo, Cinzia Cioni, Rosa Cervone  
Pediatric Unit, San Giuseppe Hospital, Empoli, Italy  
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**Background:** Allergy diagnosis is based on the history, clinical symptoms and physical examination of the patient in combination with results of in vivo and in vitro test. Today the allergy diagnosis can be refined using

molecular allergology, which allows the quantification of allergen specific IgE antibodies to a single, pure allergen molecule (component). This contributes to a more exact diagnosis, and thereby enables a more accurate disease prognosis in terms of tolerance development or risk assessment leading to an improved patient management. Depending on how unique the components are in distribution and structure, they are classified as Specific or Cross-reactive. Specific allergen components are more or less unique for their source, and are found only in a rather limited number of very closely related species. In each allergen sources there are one or more specific allergen components and sensitization to these indicate a true/genuine sensitization, meaning that the corresponding allergen source is the primary cause of the clinical symptoms. Cross reactive allergens on the other hand, are widely distributed (also in distantly related species) and due to their high degree of structural similarity may cause IgE antibody cross-reactivity. Components can be either labile or stable depending on their structure. Stable proteins are not easily broken down by cooking, processing or by enzymes in the saliva and in the gut. Stable proteins will reach the circulation in a more or less intact form and therefore potentially give rise to systemic reactions. Labile proteins that are easily broken down by processing, cooking or by enzymes in the saliva or gut will mainly give rise to local reactions, such as Oral Allergy Syndrome.

**Conclusions:** The Molecular Allergology allows: 1) To identify the right patients for Specific Immunotherapy: sensitization to specific allergen components is essential for successful Specific Immunotherapy. By matching patients having a genuine sensitization with an extract from the relevant source, treatment outcome is improved. 2) To explain symptoms due to cross-reactivity: symptoms elicited by cross-reacting antibodies can be distinguished from those caused by genuine sensitization which is important for patient management and for giving adequate avoidance advice. In cases where only cross-reactive sensitization is identified, further testing to find the primary sensitizer should be undertaken. 3) To assess the clinical risk for reaction: sensitization to allergen components that are stable may elicit systemic reactions, as well as local reactions, while sensitization to labile components is connected mainly with local reactions.

#### A79

##### Allergen immunotherapy

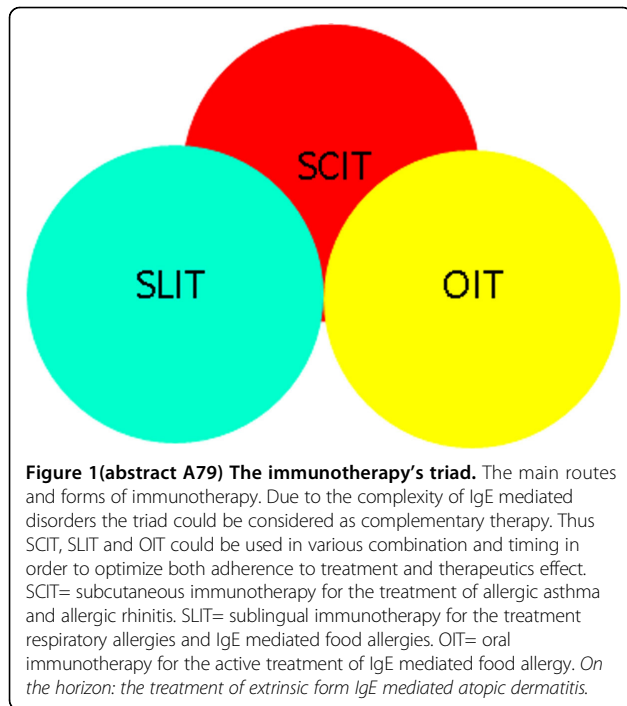
Giuseppe Crisafulli, Lucia Caminiti, Fernanda Chiera, Vincenzo Ramistella, Stefania Arasi, Giovanni Battista Pajno\*  
Department of Pediatrics, Allergy Unit- University of Messina- Italy  
*Italian Journal of Pediatrics* 2014, **40(Suppl 1)**:A79

Allergen Immunotherapy (AIT), is effective in reducing the clinical symptoms associated with allergic rhinitis, asthma and venom induced anaphylaxis [1]. Subcutaneous (SCIT) and Sublingual (SLIT) with unmodified allergen extracts are the two routes of administration of allergen vaccines.

In addition, AIT has been positioned as the only treatment that may alter the natural course of allergic disease [2]. However both SCIT and SLIT require that the treatment is taken regularly over several years e.g. monthly in a supervised medical setting with SCIT and at lowest three times a week with SLIT. Emerging evidence suggests that specific allergen immunotherapy may be effective in other allergic conditions such as IgE mediated food allergy [3,4] and extrinsic form IgE mediated Atopic Dermatitis [5]. On all these fields, the immunotherapy's triad can be an effective tool (Figure 1).

Moreover due to the complexity of IgE mediated disorders, each component of triad: SCIT, SLIT or Oral Immunotherapy (OIT) could be considered as complementary or synergic therapy. Currently, in paediatrics, the challenge is represented by the possibility of defeat the reluctance to encourage the implementation of early intervention in IgE mediated allergic diseases, with the goal of achieving either secondary prevention or long lasting benefit through immunotherapy(ies) which is the only antigen specific immunomodulatory treatment routinely available. These effects are of particular relevance in paediatric population with the aim of impairing the natural history of allergic diseases.

The mechanisms of action of AIT have been elucidated: a diminished allergen specific T-cell proliferation and suppressed secretion of TH2 cell responses are the characteristic hallmarks. In addition, T regulatory (Treg) cells inhibit the development of allergen specific TH2 and TH1 cell responses and therefore exert key roles in healthy immune response to allergens. Treg cells potently suppress IgE production and directly or



indirectly control the activity of effectors cells of allergic inflammation, such as eosinophils, basophils, and mast-cells [6]. Therefore, AIT in different forms represent an effective therapeutic approach in children with IgE mediated respiratory disorders. Moreover, in other allergic disorders that are no part of the respiratory disease spectrum, the evidence is beginning to emerge that these diseases also will respond to allergen specific immunotherapy.

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#### A80

##### Good practices in bilingual children

Francesca Cucinotta, Roberta Maggio, Matilde Gagliano, Maria Venuti, Antonella Gagliano  
UOC di Neuropsichiatria Infantile, Policlinico Universitario G. Martino, Messina, Italy  
E-mail: agagliano@unime.it  
*Italian Journal of Pediatrics* 2014, **40**(Suppl 1):A80

Bilingualism is the regular use of more than one language in everyday life [1,2]. In our country, 47.2% of students were born from non-Italian

parents [3], and in Europe, bilingualism is even more prevalent: 56% of the population across all European Union countries is reported being functionally bilingual [4]. So it is critical to understand risks and protective factors specific to the development of bilingual children for clinical and educational reasons.

It is true that linguistic tasks are often performed more poorly by bilingual children than monolinguals [5,6], especially as regards assessment of vocabulary [7,8], picture-naming tasks [9,10], comprehending and producing words [11,12]. It is also common among bilingual children who learn Italian as a second language, to have lower skill levels than those of monolinguals, in relation to reading and writing or understanding of texts [13]; but these results cannot be described as a pathological outcome.

In contrast to this pattern, bilinguals at all ages demonstrate better executive control than monolinguals matched in age and other background factors.

In the last decades, many studies showed that the experience of early exposure to two languages, and the constant practice of selecting the target language avoiding intrusions of the non-target language, can improve skills such as selective attention, inhibition and cognitive control with respect to non-verbal tasks. Other benefits also include the early development of metalinguistic ability and better achievements in working memory tasks.

These effects have been found at all stages across the life span, beginning from infancy and toddlerhood, continuing through young adulthood and up to older age [14-17].

Thus, it is a matter of considerable concern with the large and growing dual language population, how to properly recognize normal and abnormal dual language development, and several important implications can be derived from extant developmental and clinical research: a language disorder should be suspected in a dual language child, when the child is reported to be significantly behind in the understanding of both languages, although there has been significant exposure to both languages, and when there are language-based learning problems [18]. While it has been clearly documented that bilingualism does not cause language delay or language disorders [19], the latter are certainly possible in bilingual children. Such possibility should not be easily dismissed and slight delays should not instead be misattributed to the child's bilingual condition even though a more severe delay must be assessed.

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## A81

### Project over: nocturnal enuresis and urinary disorders

Giuseppe Ragnatela<sup>1\*</sup>, Marina Picca<sup>2</sup>, Lorenzo Cresta<sup>3</sup>, Mario Fama<sup>4</sup>, Carmela Lo Giudice<sup>5</sup>, Angela Pasinato<sup>6</sup>, Marco Sequi<sup>8</sup>, Pier Luigi Tucci<sup>7</sup>  
<sup>1</sup>ASL BAT Barletta, Italy; <sup>2</sup>ASL Milano, Italy; <sup>3</sup>ASL Genova, Italy; <sup>4</sup>ASL 17 Monselice-Este, Italy; <sup>5</sup>ASP 6 Palermo, Italy; <sup>6</sup>ASL Vicenza, Italy; <sup>7</sup>ASL 10 Firenze, Italy; <sup>8</sup>Mario Negri Institute for Pharmacological Research, Milan, Italy  
*Italian Journal of Pediatrics* 2014, **40**(Suppl 1):A81

**Background:** "Enuresis " also known as *bedwetting* or night-time incontinence is the inability to control urination during sleep [1,2]. Mono-symptomatic enuresis (MNE) is the only night-time incontinence and "not mono-symptomatic enuresis" (NMNE) is also the daytime urinary disorders. Bedwetting is a disorder that affects the development of children personality and interferes with social relationships of children and their family.

**Aim:** Primary endpoint is to estimate the prevalence of enuresis in patients aged 5 to 14 years, to evaluate the awareness of the problem by the family [3], to assess the discomfort experienced by patients and then to look for the presence of co-factors.

**Methods:** The study, promoted by SICuPP (Italian Society of Pediatric Primary Care), was based on a questionnaire filled by the parents of children enrolled by 75 pediatricians from three different regions (Veneto, Tuscany and Puglia).

**Results:** We evaluated 3165 children (1618 males and 1547 females). The enuresis is present in 262 children (8.2 % of the study); the prevalence was 16.5% at 5 years 10.5% and 6% NMNE/MNE) and decreases with age up to 4.6% (2.6% and 2% NMNE/ MNE) at the age of 13 years; these data agree with other epidemiological studies . Familiarity and sleep disorders are potential risk factors. Only 33% of parents tell the problem to the pediatrician: 67.8% of the parents don't speak of it because they consider it unimportant, 2.7% because they are "ashamed", 29.5% for "other reasons". Only 32 patients with enuresis (12.2%) were in treatment. 21.8% of parents think that enuresis "very emotionally involved" their child, 8.5% think that the child is "very limited" in its activities with classmates, 8.7% think that the child is "very concerned" about his health. 33.3% consider bedwetting problem "very important" for the family organization, 24.7% consider that it restricts greatly the activity of the child with classmates, 17.3% is "very concerned" about the health of the child, and finally 11% think that bedwetting is something to be "very ashamed" of.

**Conclusions:** Preliminary data show that bedwetting is a "masked" disease. It is important to look for and recognize the problem, to improve the quality of life of patients and their families

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## A82

### Diagnosis and management of headache in children and adolescents: an overview

Vincenzo Belcastro  
Neurology Unit, S. Anna Hospital, Como, Italy  
E-mail: vincenzobelcastro@libero.it  
*Italian Journal of Pediatrics* 2014, **40**(Suppl 1):A82

**Background:** Headache is a common complaint in childhood with up to 75% of children reporting a notable headache by the age of 15 years.

Pediatric migraine is the most frequent recurrent headache, occurring in up to 28% of older teenagers. Migraine can have a substantial effect on the life of the child, as well as their family, leading to lost school days and withdrawal from social interactions. The prevalence of non-migrainous headache is 10-25% in childhood and adolescence.

**Materials and methods:** The distinction of tension-type headache from migraine can be difficult. Although the International Classification of Headache Disorders criteria help, these criteria might be too restrictive to differentiate tension-type headache from migraine without aura in children. A headache diary is a useful method for the differentiation of headache types. The diagnosis of primary headache requires exclusion of secondary headache since many organic disorders can present as tension-type headache because disease-specific features are absent. Fever is the most common cause of benign paroxysmal headache. The basis of diagnosis is a systematic headache history, careful physical and neurological examination, including fundoscopy, and follow-up using a headache diary. If the results of neurological examination are normal in children with frequent headache neuroimaging is not routinely done.

**Results:** Several conditions have a comorbid relationship with migraine, such as asthma and allergic disorders, obesity, epilepsy, sleep disorders, and psychological or emotional disorders. The mechanisms by which these comorbid conditions alter the underlying pathophysiology and thus affect the manifestation of migraine are largely unknown. At the biological level, the comorbid disorders might have a common neuropathological pathway, whereas from a behavioural perspective, the difficulty in coping with multiple illnesses might alter the manifestation of the headache. In children, a connection seems possible between tension-type headache and psychosocial stress, psychiatric disorders, muscular stress, or oromandibular dysfunction.

**Conclusions:** Most tension-type headache is best managed by primary care. Episodic tension-type headache is self-limiting, but children and their parents generally consult doctors when headache become frequent and are no longer responsive to analgesic. Medication overuse headache can also be a common problem in patients with frequent headache. The treatment of migraine and tension-type headache overlap. Both require acute treatment, either behavioural or pharmaceutical. Preventive pharmaceutical treatment is needed for frequent tension-type headache and migraine.

## A83

### Headache and comorbidity in pediatric age

Alberto Verrotti<sup>1</sup>, Ilaria Bizzarri, Giulia Cecconi, Lorenza Di Genova, Manuela Vultaggio, Marta Cofini  
Department of Pediatrics, University of Perugia, Perugia, 06132, Italy  
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**Background:** Frequently headache is associated with numerous comorbidities, but data are incomplete and sometimes conflicting.

The aim of this review is to highlight the relation between headache and associated disorders.

**Materials and methods:** We identified studies via PubMed search indexed for MEDLINE using "headache comorbidity and children" as key words. The time period covered was approximately 10 years. Only English language articles were reviewed.

**Results:** The findings of our research demonstrate that epilepsy, anxiety/depression, attention deficit/hyperactivity disorder (ADHD), obesity and childhood abuse are the most frequent and investigated headache-comorbidities.

Headache and epilepsy is the most frequent comorbidity in pediatric age [1-3]. The major association is with focal epilepsies, in particular cryptogenetic ones [4,5]. The real effect of headache on seizure-related-headache is still unclear. Peri-ictal headache is a common feature of epileptic seizures: it can occur before, during or after seizures [6]. Although family history of headache and/or epilepsy is referred, there is not a clear association with specific type of both disorders [5].

Moreover, migraineurs have a higher prevalence of anxiety/depression than controls [7,8]. Severity of anxiety/depression is linked with severity and frequency of migraine [9]. Therefore, parents and relatives of migraineurs are affected by both headache and psychiatric disorders [10]. Another important comorbidity is headache and ADHD [11]. Headache is not associated with ADHD overall but with hyperactivity/impulsivity symptoms [12]. While the relation with inactivity symptoms is controversial, differences

between headache types and ADHD have been not found [13]. Structural and functional abnormalities in the brain networks seem to be central in both headache and ADHD pathophysiology [14].

It has been demonstrated an association between migraine and obesity, but the real link is still matter of debate. Psychological conditions and inflammatory mediators may be involved, as a common pathophysiologic mechanism [15]. Body mass index seems to be related to high frequency, degree of migraine attacks [16-18], and chronic migraine [19]. Many studies evidence the relationship between migraine and childhood abuse. The emotional abuses' prevalence in migraineurs is higher than controls and it is more common in women [20]. Childhood maltreatment appears to be related to chronic and disabling headache [21]. This pattern leads to an early life stress that influences the neurobiological physiology [22].

**Conclusions:** Further studies are needed to obtain detailed epidemiological data and to understand whether common mechanism(s) below these conditions exists. Physicians should consider and investigate the possible co-occurrence of these disorders in patients with headache.

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#### A84

##### Pharmacological and non-pharmacological treatment of pediatric primary headaches

Massimiliano Valeriani<sup>1,2</sup>

<sup>1</sup>Headache Center, Neurology, Ospedale Pediatrico Bambino Gesù, IRCCS, Rome, Italy; <sup>2</sup>Center for Sensory-Motor Interaction, Aalborg University, Aalborg, Denmark

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Primary headaches represent highly prevalent diseases in childhood and adolescence. Unlike in adulthood, in children the most common primary headache is migraine, while tension-type headache (TTH) is less diffused, especially up to 10 years of age. Cluster headache is to be considered exceptional, showing a prevalence of 0.1%.

While in TTH painful attacks are often of weak intensity and do not require particular treatment, migraine attacks often impair child's quality of life and need to be interrupted. The first choice drug for the migrainous attack treatment should be ibuprofen. In case of failure, paracetamol and other NSAIDs should be considered. Triptans, which are the most effective drugs for the migraine attack treatment in adults, are not authorized in children, with the exclusion of sumatriptan 10 mg spray.

Whether migraine attacks become too frequent or they do not respond to symptomatic treatment, a prophylactic therapy should be considered. Among drugs used for migraine prophylaxis, only flunarizine and topiramate have solid evidence of effectiveness, although there are also some data in favor of valproate and amitriptyline. It should be underlined that the practice to use minerals or herbs for migraine prophylaxis does not have any scientific support.

Non-pharmacological treatments have also been proposed for pediatric migraine. Cognitive treatment, associated with amitriptyline, has proved useful in chronic migraine. In different painful syndrome, acupuncture was demonstrated to have an analgesic effect. Although a specific demonstration of acupuncture efficacy in pediatric migraine is lacking, this non-pharmacological intervention should be considered, especially in drug resistant cases.

#### A85

##### Path Diagnostic Therapeutic Care (PDTA) in children and adolescents with headache

Vincenzo Raieli<sup>1</sup>, Angelo Vecchio<sup>2</sup>, Flavia Consolo<sup>1</sup>, Giuseppe Santangelo<sup>1</sup>, Renata Pitino<sup>1</sup>, Giuseppe Porrello<sup>2</sup>, Francesca Vanadia<sup>1\*</sup>

<sup>1</sup>Department of Childhood and Adolescent Neuropsychiatry of ASP6, Palermo, 90100, Italy; <sup>2</sup>Department of Childhood and Adolescent Neuropsychiatry Di Cristina ARNAS Civico, Palermo, 90100, Italy  
Italian Journal of Pediatrics 2014, **40**(Suppl 1):A85

**Background:** Headache is a common and very disabling disease in pediatric population and also its management in ambulatory and emergency pediatric unit has a significant economic and social impact.

For these reasons, it seemed appropriate to identify care pathways involving different specialists of hospital and local healthcare services that collaborate together to improve diagnosis and management of headache in young people. "Path Diagnostic Therapeutic Care (PDTA) in children and adolescent with headache" is a (appropriate and individualized) care pathway involving a network of different medical structures specialized for neuropsychiatric disease in childhood.

**Methods:** The purpose of the PDTA in children and adolescents with headache is to reinforce collaboration between hospital and territorial neuropsychiatric services, through the creation of dedicated and appropriate paths; to encourage patients to take a proactive approach to the management of their headache in to the territorial healthcare service, already in the hospital, implementing shared diagnostic and therapeutic project. PDTA targets are patients with acute headache (primary or secondary), which require hospitalization at the Department of pediatric neuropsychiatry (NPI) ARNAS "Di Cristina" Palermo and patients which require NPI territorial healthcare services, through shared diagnostic and therapeutic pathways.

Established paths:

- 1) Inside the territorial services: path from level I territorial surgery to Level II specialized department of "Diagnosis and treatment of headaches in children and adolescents";
- 2) From territorial services to hospital: path from level II specialized department "Diagnosis and treatment of headaches in children and adolescents" to Department of pediatric neuropsychiatry (NPI) ARNAS "Di Cristina" Palermo;
- 3) From hospital to territorial services: path from Department of pediatric neuropsychiatry (NPI) ARNAS "Di Cristina" Palermo to "Level II specialized territorial department of Diagnosis and treatment of headaches in children and adolescents";
- 4) Inside the territorial services: path from "Level II specialized department: Diagnosis and treatment of headaches in children and adolescents" to "level I NPIA territorial surgery"

**Results:** According to data collected, PDTA implementation will permit to reduce by 20% of inappropriate accesses to Emergency Pediatric Unit (roughly 50% admitted primary headaches) and by 15% reduction of neuroradiological examinations performed in the emergency department.

**Conclusions:** PDTA propose to improve sanitary assistance in children and adolescent with headache and his parents, through a appropriated and dedicated diagnostic and therapeutic paths. Furthermore, the main expected result is improving patient compliance to diagnosis and treatment of headache in pediatric population

## A86

### Quality of clinical guidelines in pediatric headache

Pasquale Parisi

NESMOS Department, Chair of Paediatrics, Pediatric Headache Centre, Paediatric Sleep Centre & Child Neurology, Faculty of Medicine & Psychology, "Sapienza University", Sant'Andrea Hospital, Via di Grottarossa, 1035-1039, Rome, Italy

E-mail: pasquale.paris@uniroma1.it

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**Background:** Headache is a very common complaint in children, and can have a profound impact on school performance [1], being the major cause of absence from school [2], and interfering with other daily activities [3]. The studies based on parental reports may be an unreliable source of information on the frequency of headache in young children; in fact, it has been suggested that almost 36% of the parents of children with headache are unaware of the headache [4]. In any case, the increased incidence over the last 30 years probably reflects the significant changes in children's lifestyles.

Given the elevated prevalence and the associated high degree of disability, it is not surprising that headache represents an important public health issue with considerable costs for the National Health Care System (NHCS), although, as children are not directly involved in the productivity process, it is not so easy to quantify the enormous, both, direct and indirect NHCS costs in this population [5].

**Methods:** To assess the appropriateness and uniformity of application of the available pediatric clinical guidelines (CGs) for the diagnosis and treatment of headache in children, It has been conducted a systematic

literature search using the following terms: headache, cephalalgia, guidelines and children (MESH or text words). Six CGs containing informations on the diagnosis and management of headache with specific recommendations for children were selected [6-11]. Eleven neuropsychiatric centers evaluated, by means of the AGREE II instrument, the quality and the appropriateness of available CGs.

**Results:** NICE CGs resulted "strongly recommended", while the French and Danish CGs were mainly "not recommended". The comparison between the overall quality score of the French and NICE CGs was statistically significant ( $6.54 \pm 0.69$  vs  $4.18 \pm 1.08$ ;  $p = 0.001$ ). A correlation analysis showed a significant association only for the "editorial independence" domain ( $r = 0.842$   $p = 0.035$ ). The intra-class coefficients showed that the higher agreement between 11 reviewers was present for the Lewis CGs ( $r = 0.857$ ) while the lower one for the NICE CGs ( $r = 0.656$ ).

**Conclusions:** CGs are definitely scarce and non "homogeneous". A major efforts to update the existing CGs according to principles of the evidence based medicine are needed.

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## A87

### Pediatric claims in Italy during a 8-years survey

Rino Agostiniani, Antonio Correr, Paolo D'Agostino, Ernesto D'Aloja, Luigi Greco, Paolo Tagliabue, Vassilios Fanos, The Risk Management Commission of the Italian Society of Pediatrics Neonatal Intensive Care Unit, Puericulture Institute and Neonatal Section, AOU and University of Cagliari, Italy  
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Very few data are available on pediatric malpractice claims. We report the first data obtained in Italy on pediatric malpractice regarding a wide population study during a 8 years survey. Data concerning 164 claims are presented

**Table 1(Abstract A87) Main area of claims in the 8 years-survey**

Main area of claims	Number of claims
Surgical pathology	26
Infections	20
Gastrointestinal medical problem (vomiting, diarrhea) with subsequent dehydration	14
Iatrogenic pathology (drugs, catheters...)	5
Undiagnosed tumor	4
Undiagnosed congenital cardiopathy	3
Post vaccinal encephalitis	3
Undiagnosed urinary disease	2
Undiagnosed rare disease	2
Undiagnosed deafness	2

and discussed. Our data suggest how big is the problem and they may be helpful to face it.

**Introduction:** Pediatrics is not a high-risk specialty in terms of the number of claims, although some of the largest financial payouts have been for multiple disabled children with perinatal injuries and long life expectancy [1-5]. We report the first data obtained in Italy on pediatrics regarding a wide population study.

**Materials and methods:** We conducted a retrospective, descriptive analysis of a nation-wide database on pediatric malpractice claims, in which patients alleged a permanent impairment related to a medical misconduct. The Italian Society of Pediatrics (Società Italiana di Pediatria; SIP) has developed a link-thorough insurance broker Willis Italian SpA—with an insurance company (CARIGE Assicurazioni SpA) that insures a wide proportion of Italian pediatricians (nearly 60% out of 8000 physicians). We asked Willis to perform a query of its database, looking at malpractice claims reported between January, 1<sup>st</sup> 2005 and December, 31<sup>st</sup> 2012 involving pediatrics while avoiding neonatology. Definitions are used as previously reported by ours [6].

**Results:** We found 164 claims, the majority of which were reported in the last two years (year 2011: n=65; year 2012: n=35), covering more than 2/3 of the total number of claims. 89 were from South Italy, 43 from the north and 32 from Central Italy. 141 involved the public health system, 13 the private health system and 8 family pediatricians. 102 were criminal actions, 53 civil actions, 5 mixed actions and 4 cautelative claims. We found 89 death claims and 65 claims for permanent impairment. Each claim interested one or more physicians. Main areas of class are presented in table 1. Among surgical pathologies, 6 were gastrointestinal and 6 involved testis. Among infections, 6 were pleura-pulmonary diseases, 5 fulminant sepsis (2 meningococcal), 3 tuberculosis (2 meningal, 1 pulmonary), 1 *Salmonellosis*, 1 *Candidosis*, 1 chicken pox, 1 scarlet fever.

**Conclusions:** Malpractice data can be used to identify problem-prone clinical processes and suggest interventions that may reduce errors. Continual medical education should be oriented in areas of claims also improving physician communication skills [7].

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#### A88

##### The right to health-care and the regionalization of the health-care system

Stefano Semplici  
Department of Business Government Philosophy Studies, University of Rome "Tor Vergata", Rome, 00173, Italy  
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In Italy, the process of decentralization of the health-care organization was given a strong boost during the last decade of the twentieth century, according to some inspiring lines of the Law itself, which established in 1978 the National Healthcare Service. Regions were given more and more autonomy and legislative powers in this field, so that they gained a substantial role in programming, organizing and managing health care services. The new Article 117 of the Constitution, adopted in 2001, made it explicit that health protection is a matter of concurring legislation, with the clarification that in such matters the State, which maintains exclusive powers with regard to the determination of the basic level of benefits relating to civil and social entitlements, is given the role to lay down only the fundamental principles. As a consequence, the role of the State—as it is affirmed on the website of the Italian Ministry of Health—has changed into a function of warrantor of fairness, building on the concept of the essential levels of assistance which ought to be guaranteed throughout the national territory.

The major risks that deserve a special attention are those concerning the possibility of a too deep fault of inequality as to what is offered in different regions beyond the threshold of what is established as essential at the national level and a too broad discretion in defining standards, procedures, guidelines, not to mention the burning issues of costs and quality. Paediatrics, far from being an exception, offers telling examples of the asymmetries and gaps that we should fill in, in order not to miss the obligation entailed in the recognition of the right to health-care as a constitutional essential. The neonatal screening programmes on the one side and the management of vaccination on the other side underline the difficulty of drawing the line of what is essential as well as acknowledging the limits of regional autonomy in applying fundamental principles. The situation of neonatal intensive care units underlines faults of inequality that look hard to reconcile with the idea of one and the same dignity of all citizens. Needless to say, starting with the newborn.

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