The increasing use of ultrasound in the study of multiple parts of the body has profoundly changed the diagnostic approach in numerous childhood clinical scenarios. Some technical aspects, such as minor thickness to go through and the lower fat content of tissues, make children optimal subjects for diagnostic ultrasound; exams are also well tolerated by young patients and appreciated by their parents, not painful or invasive and they do not need annoying preparations and/or sedation. Due to its characteristics ultrasound appears as an ideal integration instrument of the medical examination, increasing accuracy and providing real-time answers to many of the clinical questions open, without interrupting direct communication with the child and his family. The examination can be performed at the bedside, both in the ward and in the office, but also in emergency situations, in the ED or in the operating room, promptly and with the possibility of seriate checks to appreciate the evolution of the pathology and response to treatment. The ultrasound fields of application are widening every day: it has always been excellent for soft tissues, and has proved to be useful for the skeleton and lung parenchyma, for long time considered technically not explorable by ultrasound. The risk is that such and many advantages lead to infinitely expand the indications; in recent years we have seen an exponential growth in demand for ultrasound examinations in pediatric patients, both in primary care and specialized pediatrics. This situation has caused significant organizational difficulties, due to the limited availability of experienced operators and good quality equipment. It therefore seems desirable, alongside an increase in the number of operators dedicated to pediatric ultrasound, an intense activity on the issue of the appropriateness of diagnostic tests. One answer to the problem can be represented by the diffusion of the specialization school would represent the ideal answer.
References


A3

Patient and family centered care: a useful tool to integrate with guidelines

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The relationship between the paediatrician, the child and his family is often “paternalistic”: “I’m the physician in charge, it’s just me that knows what’s the best for the child’s health and I take decisions for the child and his family”. This approach doesn’t consider the autonomy of the patient and his right to have an active role in the decision making process about himself and his health. Almost every Clinical Guideline recommends to actively involve the patient in the diagnostic-therapeutic work-up but seldom they suggest how to. The Patient and Family Centered Care (PFCC) points out priorities, aims, methods to carry out a clinical approach really focused on the patient and his family and not just on his disease [1]. The PFCC highlights the importance of the patient’s experience, which will be better if physicians integrate the disease management procedures with standardized “relational procedures” [2], focused on the patient and his family. According to PFCC the Paediatrician is not interested into taking decisions for the patient (paternalistic approach) but he wants to involve him and the family in every step of the diagnostic/therapeutic pathway, turning the relationship into a partnership. The paediatrician should not just treat the disease but also care for the patient and his families, working hard to understand their real needs, wishes and fears, building an empathic relationship in order to make the child and his parents feel accepted, understood and supported.

An increasing number of Scientific Societies recognise the PFCC as an essential element of quality health care [3]. Clinical studies show that the PFCC improves clinical outcomes, entails better compliance to therapies, more satisfaction for health care workers and patients, as well as reduction of costs and legal issues [4]. According to PFCC the health workers have to learn the communication and empathy abilities needed to recognize feelings and wishes of the patients in order to give them answers [5]. Communication is a technical skill, which can be improved through experience and a specific training that should start since the beginning of the medical school [6]. In summary the PFCC focuses on the patient, as a person, at 360°, through strategies that are feasible, repeatable, verifiable and continuously evolving, according to evidence.

References


A4

Genes, microbiome, diet and inflammatory bowel disease

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Background: The incidence of inflammatory bowel diseases (IBD), including Crohn’s disease (CD) and ulcerative colitis (UC) is increasing worldwide [1]. These diseases result in chronic, relapsing inflammation of the gastrointestinal tract. The pathogenesis of IBD is currently thought to involve an inappropriate and persistent inflammatory response to commensal gut microbes in genetically susceptible individuals. Advances in DNA sequencing technology have led to the association of > 163 genetic polymorphisms with risk for IBD. However, in total, these loci only account for about 13% of CD and 7% of UC disease variance. Therefore, it appears that environmental factors make the largest contributions to IBD risk. Among the environmental factors associated with IBD, diet and the intestinal microbiota are the most likely to be modifiable making them targets for prevention and treatment of IBD. While nutritional therapy has been shown to be efficacious in the treatment of CD, the mechanism of action has not been well characterized. Some hypotheses involve reduction in luminal antigens and food exclusion, a direct anti-inflammatory effect of the formula, improved nutrition, and changes in the gut microbiota [2-5]. The discovery that formula composition does not impact outcome somewhat opposes the hypothesis that enteral nutritional therapy is delivering a substance that is beneficial to the gastrointestinal tract. A recent study completed at our institution showed exclusive enteral nutrition was similar to anti-TNF therapy for induction of remission but partial enteral nutrition was inferior to these therapies. Our data suggest that EEN is likely effective based on exclusion of a “harmful” factor rather than through more effective delivery of a specific nutrient. Modulation of the gut microbiota composition is a proposed mechanism of action of enteral nutritional therapy, although the current data are sparse [6]. The available literature on this subject suggests that there is a profound change in the fecal microbiota following EEN therapy [3,6].
Conclusions: In summary, it is clear that enteral nutritional therapy is a safe and effective approach to the treatment of Crohn’s disease. Induction of remission and healing of the intestinal mucosa can be accomplished with enteral nutritional therapy. Enteral nutritional therapy may also be effective in maintaining remission and preventing post-operative recurrence of disease following resection. An enhanced understanding of the mechanism of action may allow for the development of less restrictive protocols which achieve the same effect. Additionally, mechanistic studies may help to identify patient populations who may be more likely to respond.

References

A5
Vaccination risks in children with rare diseases
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Infectious diseases are a major health problem worldwide, causing many serious complications, including autoimmune disorders in unvaccinated populations. The vaccinations represent the most effective method of preventing infectious diseases and cost benefit analysis of vaccination programmes demonstrated their utility. On the basis of Orphanet database, in Italy rare diseases affect almost 2 million people, and 70% of them are children. Among neurodegenerative disorders, children with spinal muscular atrophy should receive routine immunizations, including influenza and pneumococcal vaccines; for pediatric patients affected by Becker’s Muscular Dystrophy influenza and pneumococcal vaccines are indicated in case of severe weakness of respiratory muscles [1]. Immunocompromised patients are a unique group with special issues regarding immunization; in case of B and T cell immune deficiency, live attenuated vaccines (Oral polio vaccine, Measles, mumps, rubella, Bacillus Calmette-Guerin, Varicella Zoster) should be avoided, as the major risk of vaccine induced diseases [1]. Immunocompromised individuals have suboptimal response to Hepatitis B vaccine, therefore they should receive a double dose. The administration of live vaccines is contraindicated in patients with agammaglobulinemia including X-linked agammaglobulinemia, hyper IgM Syndrome, and common variable immunodeficiency and in patients with Wiskott-Aldrich syndrome. Subjects with mitochondrial diseases should be vaccinated following the routine immunization schedule [1]. Vaccination with live attenuated vaccines should be avoided in patients with rheumatologic disorders only if they are receiving high dose of immunosuppressive drugs [2]. Measles-mumps-rubella immunization is contraindicated in subjects with autoimmune pure red cell aplasia as the greater risk of thrombocytopenia [3]. A special mention is reserved to premature infants. It is wrongly believed that in premature, infants affected by autoimmune pure red cell aplasia may present IgG deficit, as immunoglobulin cross placenta barrier mainly during the thirt trimester of pregnancy. However, it has been demonstrated that premature infants have a proper immune response to vaccinations received in the first year of life and side effects do not exceed that recorded for terms infants. Therefore, preterm babies should be vaccinated according to the recommended schedule for term infants, without correction for gestational age. In conclusion, immunocompromised subjects and premature infants do not have a higher incidence of adverse reactions following immunization if all preventive measures and recommendations are adopted [4].

References

A6
Familial Hypercholesterolemia: new therapeutic approaches
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Familial Hypercholesterolemia (FH) is caused by a mutation in the gene that encodes the low-density lipoprotein (LDL) receptor, resulting in very high levels of circulating LDL-cholesterol and endothelial damage during time. Apoprotein B, activated by LDL receptor, and PCSK9 (proprotein convertase subtilisin / kexin type 9 serine Protease), that impairs the clearance of LDL receptors, are important additional mechanisms that lead to increase LDL-cholesterol. The prevalence of Homozigous form (HoFH) is estimated to be 1 per 1 million individuals and, if untreated, proximal coronary disease or aortic valve disease frequently occurs during childhood and most affected patients suffer from fatal coronary disease before the age of 30. Heterozygous condition (HeFH) occurs in about 1/380-500 individuals. The levels of LDL-cholesterol in this form may greatly vary but the disease is always asymptomatic until adult age where there is a high risk of early cardiovascular disease. Lifestyle intervention and maximal statin therapy are the mainstays of treatment. In patients that do not achieve LDL-C targets, primarily HoFH patients, adjunctive lipoprotein apheresis is recommended where available. Biological agents represents to date a new therapeutic approach for this not so rare condition. The PCSK9 pathway is one of the best examples of how genetics has led to identification of a new target for cholesterol management. Different PCSK9 inhibitors (alirocumab, evolocumab) are now available [1]. Mipomersen is an antisense oligonucleotide (ASO) that targets apolipoprotein B-100 mRNA and disrupts its function; it is distributed mainly to the liver where it silences apoB mRNA, thereby reducing hepatic apoB-100 and giving rise to reductions in plasma total cholesterol, LDL-cholesterol, and apoB concentrations [2]. Lomitapide is a microsomal triglyceride transfer protein (MTP) inhibitor. MTP resides in the lumen of the endoplasmic reticulum, thereby preventing the assembly of apo B-containing lipoproteins in enterocytes and hepatocytes. This inhibition leads to a reduction in the synthesis of chylomicrons and very low-density lipoprotein, resulting in a reduction in plasma LDL levels [3].

References
Growing clinical evidences indicate the benefits of human milk (HM) for appropriate growth and development of a newborn: the particular composition make it a unique and inimitable nutrient [1]. Mother’s own milk is the first choice for all neonates including preterm infants, when it is unavailable or in short supply, donor milk (DM) is an important alternative. DM should be provided from an established Human Milk Bank (HMB), which follows specific safety guidelines [2]. When HMB is available, significantly less neonatal intensive care unit (NICU) neonates receive formula milk in the first weeks of life. In Italy, data from NICU patients show that exclusive breastfeeding at discharge is achieved for nearly 30% of neonates when banked milk is available during hospitalization and only for 16% of neonates when it is not [3]. DM banking should be promoted, protected, and supported as an extension of national breastfeeding policies, in this regard Italian National Guidelines were published on “Gazzetta Ufficiale della Repubblica Italiana” in 2014 [4]. Nonetheless storage and processing of human milk may reduce some biological components, which may diminish its health benefits but when donor milk is used instead of formula, it is demonstrated a reduction in the incidence of necrotizing enterocolitis and an enhanced feeding tolerance [5]. Pasteurization of the milk is necessary for minimize the risk of disease transmission, inactivating most of the viral and bacterial contaminants. Holder pasteurization (62.5°C for 30 minutes) is the most commonly used method and actually allows a good compromise between microbiological safety and nutritional and biological quality of the milk [2,4]. It results in the loss of the quantity and/or activity of some biologically functional milk components to varying degrees. The optimal pasteurization process should be optimized to maintain microbiological safety while preserving the highest amount and activity of the bioactive milk components. New methods to improve the biological quality and safety of DM are under investigation. High-temperature short-term pasteurization (flash pasteurization, 72°C for 5-15 seconds) and its homemade low-tech variant for developing countries (flash-heat treatment), thermoultrasonic treatment, high-pressure processing are the alternative methods on which present studies are focused.

Future research should focus on the improvement of milk processing in HMB, particularly of heat treatment on the optimization of DM fortification and on further evaluation of the potential clinical benefits of processed and fortified DM.

References

A7 Donor human milk: actuality and perspectives
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A8 Fever in the first month of life
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Background: Fever is a common presenting sign in children and infants. However, the management of febrile neonates (< 30 days) could be particularly challenging, mainly due to the paucity of specific signs and symptoms to discriminate “simple, self-limited infections”, mostly of viral origin, from serious, life-threatening infections, mostly of bacterial aetiology. Despite several strategies and protocols have been proposed in the medical literature, management of fever do remain a complex issue in the neonatal patient.

Contents: Evaluation of febrile newborns is primarily based on the clinical assessment, even though a combination of history, physical examination findings, and diagnostic screening tests is often required to exclude a serious illness reliably. Yet, in this high-risk group of patients, an adequate balance should be maintained, in order to appropriately identify and treat all sick newborns, while minimizing the risks associated with unnecessary invasive testing, hospitalization, and antibiotic treatment. Of note, practice guidelines for febrile neonates may differ substantially from centre to centre, in relation to execution and type of testing, intensity and level of treatment, and threshold for admission to the hospital.

According to several clinical practice guidelines, all febrile neonates should undergo a full sepsis evaluation and receive empirical antibiotics. Differently from older infants, in whom other risk factors must be taken into account, lumbar puncture is still widely recommended for any febrile newborn. Empiric antibiotic therapy usually consists in a combination of ampicillin and gentamicin, pending cultures results. In case of suspected herpes virus infection, i.e. acyclovir should be started immediately.

Finally, febrile newborns should be hospitalized or monitored in temporary observation units. Yet, consensus on the time of inpatient observation while awaiting culture results is lacking. In fact, despite a period of about 48-hour observation is generally accepted, recent data suggest that 24 hours could be adequate to detect most clinically significant bacteremia [1]. Interestingly, an observation period beyond 24 hours would capture just one additional bacteremic infant for every 556 to 1235 febrile infants evaluated [1]. Indeed, by using this cutoff, the number of nights spent in the hospital for these infants and caregivers, as well as related costs, would be markedly reduced.

Conclusions: Management of febrile newborns should include a full sepsis work-up, empirical antibiotics and strict monitoring in a hospital setting. However, recommended strategies are quite variable in the literature, and no protocol has been universally adopted as yet. A wide international consensus is still highly needed.

Reference

A9 Meningococcus B
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Between 13 different serogroups of N. meningitidis, identified according to the capsular polysaccharide’s antigenic structure, only 5 (A, B, C, W-135 and Y) are clinically relevant and responsible for 90% cases of meningococcal invasive disease. In Italy, serogroup B is the principal cause of invasive meningococcal disease [1].

There are two types of tetravalent vaccines for serogroups A, C, W-135 and Y, represented by polysaccharide and conjugate vaccines. However, only recently it has been possible to develop a vaccine against meningitis B with the “Reverse vaccinology” technique: after identifying more than 600 proteins as immunologically essential targets, 91 have been selected (expressed on the outer capsular). Only 28 of them could induce a bactericidal activity and, among these, 3 have been selected: fHbp, NHBA (expressed on the outer capsule). On 28 of them could induce a bactericidal activity and, among these, 3 have been selected: fHbp, NHBA and NaxA, which were able to stimulate an antibody protection, in addition to proteins DM (4CMenB)[2].

In November 2012, the multicomponent vaccine 4CMenB was approved from the European Medicines Agency (EMA). In January 2013, the
European Commission authorized the marketing of the new vaccine 4CMenB (Bexsero®), addressed to immunization from two months of age [3,4]. In Italy this authorization is implemented by AIFA determines of May 27, 2013. Moreover, in 2014 in the US, a new anti-meningococcal bivalent vaccine was approved: Trumenba® (Pfizer). Currently, 4CMenB has been authorized in 12 countries worldwide, the largest of which are Australia and Canada. Already 151,800 subjects have received at least one dose of vaccine in 18 countries worldwide.

In Italy, Basilicata has been the first region that implemented 4CMenB in its vaccination schedule, with resolution in 24 February 2014. A few months later also Puglia, Liguria, Tuscany, Veneto, Sicily, Friuli, Bolzano and Calabria have introduced the meningococcal B vaccine, and other regions are ready to follow them.

In the new Italian vaccination calendar, proposed by SLTL, FIMP and SIP, 4CMenB has been prepared in a 4-doses schedule, in consideration of the higher incidence of the infection among the first 4-6 months of life (first cycle based on three doses in the first year, starting from the 75th day of life, and a forth dose at 13-15 months of age) [1].

References

A10
Pediatric andrology: the andrological patient from infancy to adulthood. Prevention interventions: how and when
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Sonography, a non-invasive, quick method without biological impact on the gonad, has greatly improved the diagnostic possibilities; it is the gold standard for the male genital system (gonads and genito-urinary tract) and in fact has orientated the prevention interventions [1]. It recognizes almost 100% of scrotal lesions and their solid, liquid or complex nature. It has allowed to overcome the limits of traditional diagnostic methods for gonads like palpation, transillumination, and the Prader orchidometer for volumetric assessment. In fact, compared to this, sonography allows a more accurate volume measurement, especially in small testicles; a correlation between measurements made with the orchidometer and those made with ultrasounds was found only for testicular volumes over 4 cc. [2].

Measurement of testicular volume is a fundamental element in pediatric andrology for evaluating puberty onset and progression. Furthermore, the sonographic exam is one of the best tools for assessing testicular pathologies like torsion, undescended testis, varicocele and in general in pathologies involving male genitalia; these cases can show significant variations of testicular volume. Testicular growth restriction may have relevant clinical implications for future testicular function [3].

The limits of traditional tools, like the orchidometer or the ruler for the measurement of pre-puberal testicle, are easy to identify:
- The smallest pearl in Prader orchidometer is 1 ml, while sonography allows to make measurements in the first years of life (0,44 ml ± 0,03);
- Orchidometer and ruler are known for overestimating testicular volume because they measure not only the didymus but also the epididymis (which in the first childhood is relatively big, compared to total testicular volume) and the scrotal tissues.

Testicular volume is related to many reproductive endocrine parameters; therefore, a measurement of testicular volumes with a reliable method is appropriate, because any scrotal anomaly can influence testicular growth, and should be detected and treated as soon as possible.

Moreover, andrological sonography is a first choice exam for male genitalia pathologies like acute scrotum, where diagnosis and clinical evaluations are particularly difficult. For example, during a suspected testicular torsion, a measurement made with a high resolution ultrasound machine by an expert examiner allows to reach a sensibility of 98% and a specificity of 99%, like scintigraphy and MRI with contrast medium.

The sonographic study of the testicular region has absolute indications and relative indications. Absolute indications: difficult/inadequate objective clinical examination (sore and/or swelling testis, suggesting an acute scrotum; testicular torsion, trauma, undescended testis) or suspected testicular mass [4].

References

A11
Neonatal Expanded Screening toward lysosomal storage disorders
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Newborn Screening (NBS) is a public health program aimed at identifying treatable conditions in pre-symptomatic newborns to avoid premature mortality, morbidity and disability. The advent of tandem mass spectrometry (MS/MS) has enabled the interrogation of multiple disorders using a single, multianalyte assay changing the origin scenario of one screening, one disease. For example, even if a disorder was extremely rare, if it could have been detected and there were an effective intervention the minimal cost of adding it to a MS/MS panel might be cost effective. Similarly, if one could add a disorder which there was no accepted effective treatment, it might be cost effective to add it based upon minimizing diagnostic testing to determine the cause of the phenotype and being able to counsel parents about their reproductive options.

This new based-technology prevention program, aimed at identifying an increasing number of conditions, fits for some lysosomal disorders (LSDs) such as Gaucher, Pompe, Fabry, MPSI, krabbe and Niemann-Pick diseases that have been proposed for inclusion in newborn expanded screening programs. In different Countries, pilot studies including all the above diseases or more selected disorders have already found the opportunity to validate the effectiveness of different methods, define the cut-offs for detection of the LSDs and alert the entire system of urgent referral, follow-up confirmation, treatment and screening program communication.

A12
Protocol for identification and for age assessment of unaccompanied minors
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Background: The national technical board, established in 2008 on request of the Marche Region Health Department, is coordinated by the Observatory on Health Inequalities and composed of members of Regional Health Departments, Health Ministry technicians and experts from the Italian Society of Medicine of Migrations; it guarantees a constant dialogue and cooperation between the Regions and the national level about issues and policies for immigrant's health care. Its aim is to encourage policy makers to fight immigrants’ health inequalities and to achieve geographical uniformity and fairness in the access to health care by the immigrants.

Materials and methods: In 2012-2014 the board developed the "Protocol for identification and for age assessment of unaccompanied minors (UM)". The document, issued with the contribution of Ministries of the Interior and Justice, Save The Children, UNHCR and SIP, contains the philosophy of the Protocol:

1) the implementation of a holistic and multidisciplinary age assessment of the presumed minor so replacing the medical/radiological evaluation; it is assumed that both methodologies have some degree of uncertainty but the holistic one is preferred for its complex and multidisciplinary approach;
2) it is a "unitary" document containing the operating procedures to be followed by all the actors involved in the identification procedures and age assessment of UM; its implementation will lead all the different subjects involved, such as Regional Health workers and operators of the Interior and Justice Administrations, to speak a common language and attend the same practices.

Results: The Protocol provides for:
- description of the steps carried out by the Police for the "correct" identification of the presumed minor;
- procedures to ensure the legal protection and informed consent of the presumed minor;
- holistic and multidisciplinary age assessment - suggested only when a serious doubt remains after the identification steps and in extrema ratio - made in a public health service by a team composed of pediatrician, social worker, intercultural mediator, psychologist, pediatric neurologist; - the pediatrician, together with the multidisciplinary team, will decide which tests require, using the least invasive ones.

Conclusions: The Protocol is coherent with European Directives [1]; it follows the national and international scientific recommendations about age assessment in minors [2,3]; its application can help Italy to get out of the situation of of age assessment procedures. The Protocol is currently being evaluated by the Presidency of the Council of Ministers to be converted into a specific national law. Application of the Protocol requires training for health professionals to overcome the "old practice" of using radiological examinations.

References

A13 Lung Ultrasound (LUS) and neonatal respiratory distress
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Introduction: Neonatal lung diseases are often a diagnostic dilemma for the clinician due to the low sensitivity and specificity of clinical signs and symptoms. In the last decade of the previous century the use of ultrasound in the diagnostic work-up of adult respiratory diseases became widely used [1,2]. The purpose of this paper is to update the knowledges on LUS in the most common neonatal respiratory diseases [3,4].

Materials and methods: A high resolution linear probe 10 MHz or more is used for lung examination. Longitudinal and transversal sections of the anterior, lateral and posterior wall are obtained. In a normal lung the pleura appears as a regular echogenic line moving during respiration. Beyond the pleura the change in acoustic impedance at the pleura-lung interface results in horizontal artifacts, defined as A-lines [5]. Vertically oriented artifacts, called B-lines, indicate an abnormality amount of fluid in the interstitial or alveolar compartment [1].

Results: Respiratory Distress Syndrome (RDS).
RDS diagnosis is based on the presence of echographic white lung without spared areas, thickened pleural line [4]. The LUS appearance immediately after administration of exogenous surfactant does not change [6].

Transient Tachypnea of the Newborn (TTN).
TTN has normal pleural line and pleural sliding, with compact B-lines in the inferior pulmonary fields and few B-lines in the superior fields [3], or bilateral "wet lung" defined as presence of numerous non-compact B-lines.

Meconium Aspiration Syndrome (MAS).
MAS shows a picture of coalescent B-lines and subpleural consolidations along with few spared areas. Subpleural consolidation distribution is irregular and may be more evident in one side.

Pneumothorax.
LUS signs of pneumothorax are absence of lung sliding, absence of B-lines and evidence of "lung point". Air between parietal and visceral pleura does not allow to see the movement of the visceral pleura on the radiological evaluation; the B-lines that originate from visceral pleura. Lung point when present has a sensitivity and specificity of 100% [2]. Can be seen when the partially collapsed lung inflates and parietal and visceral pleura are in contact and lung sliding is again evident.

Conclusions: In neonatal age the use of LUS is becoming a new and reliable tool in the hand of the clinician.
LUS does not substitute chest X-ray, but can reduce its use with benefits in terms of irradiation risk [7]. The use of LUS in the clinical practice is a promising and already well established entity in neonatal age.

References

A14 The referral centers for the diagnosis and treatment of hypertension in adolescents
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Primary hypertension in adolescence was felt to be quite rare. However, the worldwide childhood obesity epidemic has had a profound impact on the frequency of high blood pressure (BP) with the result that primary hypertension should now be viewed as one of the most common health conditions in the young (estimated prevalence 1–9%). Therefore, current guidelines recommend that all children and adolescents seen in a medical setting should have their BP measured. The availability of BP tables with normal BP percentiles for age, sex and height has improved BP values classification.
Studies conducted at referral clinics for evaluation of hypertension have indicated that as many as 30 to 40% of adolescents may actually have in a clinical setting white-coat hypertension. This may lead to a misdiagnosis of "true" hypertension in a considerable number of cases. The usefulness of out-of-office BP evaluation using ambulatory or home monitoring is well established. These measurements allow the detection of the white-coat and masked hypertension, the opposite of white-coat hypertension, and are more closely associated with organ damage and cardiovascular risk than office measurements. A thorough familial and personal history is of primary importance as well as the physical examination that should be focused on the search for signs suggestive of an underlying cause and/or for the severity of hypertension.

Following investigations must be tailored to the child's age, anamnesis and clinical examination and to the severity of BP elevation, in order to investigate not only the possible cause of hypertension, but also associated diseases and target organs damage. Therapeutic approach should firstly include non-pharmacological measures, and the use of medications when indicated.

A key role in the management of the adolescents with hypertension may be attributed to the hypertension referral centers (table 1).

Table 1(abstract A14) What is required at the referral centers in the management of hypertension in adolescence

- To guarantee a multidisciplinary approach to the hypertension problem in adolescents
- To provide pediatric, cardiologic, nephrologic, endocrinologic, dietary and in some cases psychological expertise
- To obtain ample experience in the evaluation of organ damage, interpretation of 24 hour ambulatory blood pressure monitoring and self measurement of blood pressure at home
- To have access to laboratory techniques and instruments necessary for the diagnosis of different forms of secondary hypertension
- To build communication channels between pediatricians and family doctors with the aim of outlining the therapy and monitoring the adolescent with hypertension

The cranial ultrasound plays an important role in the study of the brain in the newborn and infant, therefore it represents the first choice technique to evaluate many diseases. In addition to the anterior fontanelle which serves as an acoustic window, the posterior fontanelle allows a more detailed study of the posterior fossa and occipital lobes [1,2]. Ultrasound is an irreplaceable, but not exclusive diagnostic tool; it is relatively simple to use in the majority of birth time points.

The ultrasound method has a high diagnostic value in the evaluation of hemorrhagic lesions, in ventriculomegaly, and in the form of cystic periventricular leukomalacia. Many lesions, however, are at risk of resulting as false positives for non-cystic periventricular leukomalacia. These are linked to the experience of the operator and the resolution of the equipment, as well as to white matter abnormalities. In these cases it is necessary to resort to magnetic resonance and, in particular, the new functional resonance techniques.

Neonatal hypoxic-ischemic encephalopathy, especially in the early stages, may result within normal range or exhibit only a diffuse hypeerechoic picture, while more serious injuries may be recognized only later. Failure to visualize the lateral ventricles, the disappearance of the groove impressions and cerebral convolutions may be considered indirect signs of diffuse edema that may, however, be overestimated or underestimated [3].

In congenital infections, ultrasound can easily identify some brain injuries, such as calcifications, germinal matrix cysts or ventriculomegaly; while alterations of the posterior intracranial fossa are often underestimated [4], and although ultrasound is possible in disorders of neuronal migration they are inadequately diagnosed (Figure 1). In the case of neonatal meningitis - ultrasound can reveal complications such as ventriculitis, hydrocephaus and intraparenchymal abscesses, which fundamentally impact the choice of therapy.

Although technological advancements have permitted the production of economical ultrasound equipment with an acceptable level of quality, the gap in image quality and thus diagnostic power between top of the line and economical devices is generally abysmal. Currently, some ultrasound devices allow, under certain conditions, revelation of anatomical details that are almost invisible to the naked eye [5]. However, the use of proper methodology is fundamental, which necessitates the need for both accurate diagnostic and prognostic data, in order to avoid repetitive testing, and negative influences on future diagnostic - therapeutic decisions. It should be emphasized, however, that many ultrasound limitations are actually limitations associated with operator training and experience.

References


A15 Cranial ultrasound: and the risk of tunnel vision?
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"One man's one eyed perspective".
H.H. Fudenberg.

Gestalt identification is the process by which healthcare practitioners actively organize clinical perceptions into specific diagnostic ideas. This implies that clinicians, in particular geneticists, have the ability to quickly generate diagnostic hypotheses in absence of complete information, simply on the basis of combination of specific facial dysmorphisms or particular elements of the clinical history. This is particularly frequent in clinical genetics because the “facial dysmorphisms” of the patient represent often the most specific clinical criteria of the syndrome itself. Down [1], Cornelia de Lange [2], Wolf-Hirshhorn [3], Noonan [4], Rubinstein-Taybi [5] Kabuki [6], Treacher Collins [7] and Williams syndrome [8] are examples well known to pediatricians but this is true for the great majority of genetic syndromes. In these situations facial dysmorphisms can have variable expression between different patients (also in the same family) or can become more or less evident over time. Moreover the “gestalt process” can also be applied in case of specific auloxic and/or neurologic evolution. In patients with Prader-Willi syndrome (PWS) in the first months of life hypotonia is evident and tends to improve between 8 and 11 months of age; poor growth and feeding problems are frequent too. Later on hypotonia improves and feeding difficulties are replaced by
hyperphagia and obesity. This typical evolution can permit to suspect in every phase PWS diagnosis and to perform methylation test for 15q11.2 region [9]. Some neuro pediatricians suggest that Angelman syndrome’s EEG pattern is typical enough to be recognized by sensitized professionals [10]. A specific behavioral phenotype, including significant sleep disturbance, stereotypes and oppositional behavior, is pretty characteristic of Smith-Magenis syndrome [11]. Again, PTEN gene testing may be considered for patients with extreme macrocephaly associated with intellectual disability/autistic behaviour [12]. Within overgrowth syndrome, Beckwith-Wiedemann syndrome (BWS) can be easily recognized thanks to the association between hemi-hypertrophy, macroglossia, abdominal wall defects and frontal nevus flammeus [13]. Finally within patients with short stature the evidence of disproportion between crown-rump and overall lengths, or between limbs and trunk univocally direct the diagnosis toward the skeletal dysplasias [14].

In conclusion the knowledge of facial features and/or natural history (auxological, neurological behavioural etc) of the most common genetic syndromes can help pediatricians in hypothesizing them through a “gestalt process” in order to quickly confirm the clinical diagnosis with the specific genetic tests.

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References


A17

Lyosomal storage disorders for the pediatric rheumatologist: the example of mucopolysaccharidoses

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The Mucopolysaccharidoses (MPS) are a group of diseases caused by complete or partial deficiency of lysosomal enzymes responsible for glycosaminoglycans catabolism. Their accumulation within the lysosome leads to cellular damage and organ failure [1,2]. The musculoskeletal system is the most frequently affected one. Joint stiffness, contractures (claw hand), dysostosis multiplex, and carpal tunnel syndrome are some of the most frequent features [3-9]. Often the joint symptoms may be confused with inflammatory arthritides such as Juvenile Idiopathic Arthritis [10,11]. A prompt differential diagnoses is a fundamental step: in MPS patients there are no signs of local inflammation such as swelling, warmth and tenderness, and lack of fever and increased inflammatory markers. In addition, patients with MPS do not respond to anti-inflammatory therapy [12,13]. In addition, characteristic facies, cognitive impairment, short stature, recurrent otitis media, sleep apnea, hearing, vision and heart problems can be present [14,15]. Because of a wide variety of clinical presentation, diagnosis of MPS disorders is often delayed, especially in patients with mild forms and without neurocognitive impairment such as Scheie Syndrome.

When clinical features are suggestive for MPS, the diagnosis is confirmed with the assay of urinary GAG concentration, which is a sensitive but not specific method [16] and, as the gold standard with determination of the specific enzyme in cultured fibroblasts, leukocytes, plasma or serum [17]. The genetic sequencing could be used to identify the disease-causing mutation.

The management of MPS disorders requires a multidisciplinary evaluation for multi-organ involvement. The new therapeutic approaches to MPS have drastically changed the natural history of disease. Transplantation of hematopoietic stem cells from bone marrow or umbilical cord can achieve stable engraftment without the development of graft-vs-host disease [18,20]. In early stages of disease, enzyme replacement therapy can benefit musculoskeletal symptoms and lung function [21-23]. This therapy, however, does not cross the blood-brain barrier and has not shown neurocognitive benefit.

In conclusion, the goal is the prompt identification of MPS disorders since an early diagnosis could allow early treatment: in this regard, particular
attention should be given to an accurate differential diagnosis with chronic inflammatory arthropathies.

References

A18
Why does my son have a genetic disease?
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The question "why" is a typical question of human beings, who not only know the world and react to it, but also wish to understand its meaning. From the classical reflection of Aristotle which was conducted in physics as well as in metaphysics on the fourfold dimension of causality, important observations are still deduced. The cause is always material, formal, efficient, final.

Every time, in fact, one asks the question "why", the person is making a complex question which contains others: why is he/she so, what is the reason, who is the person that has done this, what is the aim.

In front of the genetic disease of one’s son, the question "why" reveals one’s urgency and necessity, and at the same time imposes the awareness that it is necessary to avoid silence as well as non-response, or the arrogance of a definite answer.

The philosophic reflection proposes research outlines on the causes, on the subjects that can and should investigate them, on the relationship between fields of knowledge, above all in relation to notions connected to "life"[1]. Is life only a medical notion? Is it only philosophical? Or only religious? And should disease be studied only from a medical perspective? Or only philosophical? Or only religious?

The most efficient answers come from the composition of knowledge; only this is able to hold together the specificity of knowledge and the complexity of reality [2].

References

A19
Blood pressure measurement in children
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In pediatric age diagnosis of hypertension is defined using a statistical criterion, the limit being the 95th percentile of the distribution of the blood pressure values, according to gender, age and height [1]. Pre-hypertension is defined as blood pressure (BP) values consistently above or equal to the 90th percentile, but lower than the 95th [Table 1]. The "gold standard" method to measure BP in children is auscultatory, using an aneroid non-mercury manometer (proscribed due to their toxicity). The aneroid devices need to be calibrated every six months. At least three measurements performed on different occasions are necessary for the diagnosis of hypertension [2]. Children above 3 years of age should have their blood pressure measured every year. In all children including the younger ones blood pressure should be measured under special circumstances that increase the risk for hypertension: intensive neonatal care, renal disease, treatment with drugs known to increase blood pressure, evidence of elevated intracranial pressure. The cuff should be of the appropriate size for the children’s upper arm. Small cuffs tend to overestimate while large cuffs underestimate. The width of the cuff should be 40% of the arm circumference at a point midway between the olecranon and the acromion. The children should be calm and relaxed, seated with their right arm resting under the elbow. Systolic blood pressure is defined by the first Korotkoff sound elevated intracranial pressure. The cuff should be of the appropriate size for the children’s upper arm. Small cuffs tend to overestimate while large cuffs underestimate. The width of the cuff should be 40% of the arm circumference at a point midway between the olecranon and the acromion. The children should be calm and relaxed, seated with their right arm resting under the elbow. Systolic blood pressure is defined by the first Korotkoff sound (K1) whereas diastolic blood pressure coincides with the disappearance of the pulse (K5). The use of oscillometric devices is increased in the last years. At least three measurements performed on different occasions are necessary for the diagnosis of hypertension [2]. Children above 3 years of age should have their blood pressure measured every year. In all children including the younger ones blood pressure should be measured under special circumstances that increase the risk for hypertension: intensive neonatal care, renal disease, treatment with drugs known to increase blood pressure, evidence of elevated intracranial pressure. The cuff should be of the appropriate size for the children’s upper arm. Small cuffs tend to overestimate while large cuffs underestimate. The width of the cuff should be 40% of the arm circumference at a point midway between the olecranon and the acromion. The children should be calm and relaxed, seated with their right arm resting under the elbow. Systolic blood pressure is defined by the first Korotkoff sound (K1) whereas diastolic blood pressure coincides with the disappearance of the pulse (K5). The use of oscillometric devices is increased in the last years.
children to measure BP values: ambulatory blood pressure monitoring (ABPM) and home blood pressure measurement. In children the use of ABPM has significant limitations due to the lack of reference values. It allows to identify “white coat hypertension” (elevated office BP values and normal ABPM values), “masked hypertension” (normal office BP values and elevated ABPM values) and subjects with or without reduced physiological day-night blood pressure variations (dipping) [3]. A new method is represented by the self-measurement of blood pressure at home [4]. Even in this case available data from children are scanty. Correct self-measurement requires two measurements within a few minutes, performed in the morning and in the evening for 3 consecutive days.

References

A20
Vasculitis and Systemic Lupus Erythematous (SLE)
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Vasculitis are a heterogeneous group of disorders characterized by inflammation of the blood vessels of different caliber and sometimes fibrinoid necrosis with vessel wall destruction [1].

Vasculitis are divided into cutaneous and systemic forms, primary and secondary to hypertension, immunosuppression therapy, metabolic complications. The classification is based on the affected vessel size (Table 1). They may have neurological manifestations at the onset and during the disease development. These are more common in systemic forms such as SLE and Nodose Polyarteritis (PAN).

The Schonlein-Henoch purpura and Kawasaki disease, the most frequent vasculitis in childhood, rarely can have neurological disorders. There are forms of mild to moderate intensity like headache, irritability, mood disorders and behavioral and forms of severe as seizures and sensory disturbance up to coma.

In the course of SLE, neuropsychiatric manifestations, headache and chorea are common, with an incidence of 20-40% (also 80% with cognitive disorders and asymptomatic alterations RMN). The neuropsychiatric manifestations involve 40-56% of children; headache the 22-64%; convulsions the 20-31%; chorea 4-10%; peripheral neuropathy 5-6%; myelopathy 1%. Heterogeneity in their neurological symptoms are important for prognostic purposes.

Vasculitis in large calibre vessels prevalence
• Takayasu arteritis
• Vasculitis in medium caliber vessels prevalence
• Nodose Polyarteritis
• Cutaneous polyarteritis
• Kawasaki disease
Vasculitis in small calibre vessels
A. Granulomatous
• Wegener Granulomatosis
• Churg-Strauss syndrome
B. Non granulomatous
• Microscopic polyangitis
• Schönlein-Henoch syndrome
• LVC isolated
• Urticarial vasculitis ipocomplementemica
Other forms
• Behcet disease
• Vasculitis secondary to infection (including nodose polyarteritis associated with hepatitis B), in tumor sand infections, including hypersensitivity vasculitis)
• Connective tissue diseases associated vasculitis
• Isolated SNC vasculitis
• Cogan syndrome
• Not classified vasculitis

Table 1.(abstract A19) Definition and classification of hypertension in children and adolescents

<table>
<thead>
<tr>
<th>Category</th>
<th>Systolic or diastolic blood pressure percentile</th>
</tr>
</thead>
<tbody>
<tr>
<td>Normal</td>
<td>&lt;$90^{th}$</td>
</tr>
<tr>
<td>Pre-hypertension</td>
<td>$90^{th}$ and &lt;$95^{th}$</td>
</tr>
<tr>
<td>Stage 1 hypertension</td>
<td>$95^{th}$ and &lt;$99^{th}$ + 5 mmHg</td>
</tr>
<tr>
<td>Stage 2 hypertension</td>
<td>$99^{th}$ + 5 mmHg</td>
</tr>
</tbody>
</table>

In antiphospholipid syndrome, primary or secondary, the following are common: transient cerebral ischemia and ischemic stroke, memory loss, chorea, seizures, vision problems [2].

The PAN is a necrotizing vasculitis histological examination, rarely aneurysm, stenosis or occlusion (not caused by fibro-muscular dysplasia, by other causes not inflammatory) artery of small and medium caliber. In addition, at least one of the following signs/symptoms: skin involvement (livedo, nodules or heart attacks); myalgia; hypertension; peripheral neuropathy (sensory or motor); renal involvement (proteinuria, haematuria, renal impairment).

The primary central nervous system vasculitis is a brain vessels inflammation not associated with vasculitis of other organs. The classification is based on the vessel size: small (with normal angiography) and medium-large (progressive and non-progressive). This form, responsible for 40-60% of arterial ischemic stroke, affects 3-8/100,000 children/year. Symptoms are characterized by acute severe headache (80%), focal neurological deficit (78%), motor deficit (62%), cognitive disorders (54%), cranial nerve involvement (59%), seizures (small vessel vasculitis).

Peripheral neuropathies are characteristic of the Churg-Strauss disease.

The clinical diagnosis is often difficult (Table 2). Neurological complications are diagnosed early because the treatment must be immediate and aggressive.

References
Feeding difficulties during the neonatal period

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Feeding difficulties (FD) are a major issue in neonatology, as they could hamper the assessment of an adequate enteral nutrition, delay hospital discharge and lead to breastfeeding failure. Functional and anatomical maturation of the gastrointestinal tract is strictly related to gestational age (GA); hence, premature infants are more prone to develop feeding intolerance (FI) as FI is very common among preterm infants; clinical symptoms of FI (e.g. abdominal distension, vomiting, bilious gastric residuals, occult or gross bloody stools) are observed in nearly 29% of such neonates [1]. FI could represent an early sign of necrotizing enterocolitis (NEC), which is the most feared gastrointestinal complication of prematurity. Hence, FI often brings clinicians to withhold, decrease or discontinue enteral feeds, thus hampering the establishment of an adequate enteral nutrition and leading to a prolonged duration of both parenteral nutrition (PN) and central lines, with increased risks of such complications as liver cholestasis or sepsis[2].

The coordination between sucking, swallowing and breathing is usually achieved at 34-36 weeks GA; hence, preterm infants are usually fed via an intragastric tube, through intermittent boluses or continuously. Poor sucking and sucking-swallowing incoordination are the major causes of FD and breastfeeding failure among late preterm infants (GA 34-36[2/7] weeks), with an increased risk of hypoglycemia, excessive weight loss, hyperbilirubinemia, dehydration[3]. Due to FD, up to 27% of all late preterm infants need to be initially supplemented with intravenous fluids[4]; moreover, tube feeding is frequently required for feeding administration in the first days of life.

The abovementioned problems are infrequent in healthy term newborns. Term neonates developing FD such as poor sucking and/or vomiting need to be evaluated for pathological causes. Physical examination could aid to identify anatomical malformations possibly responsible for FD (e.g. cleft palate). FD and sleepiness can be due to hyperbilirubinemia, hypoglycemia or electrolyte disturbances[5], but could also subend an underlying metabolic disease, such as hypothyroidism. FD, lethargy and/or other clinical neurological signs (e.g. seizures, focal neurological signs, hypo- or hypertonia, bulging fontanel, central apnoea) could address for central nervous system diseases (i.e. subarachnoid haemorrhage, ischaemic stroke, metabolic encephalopathy etc.). FD and lethargy could also represent a warning sign for invasive infections, especially if associated with respiratory distress, apnoea and bradycardia, temperature instability and increased capillary refill time[5]. Blood tests (including blood cells count, C-reactive protein, glucose, bilirubin, electrolytes, blood gas analysis) and cerebral ultrasound scan are useful tools to aid neonatologists in the differential diagnosis.

References
Uncontrolled asthma defined as at least one of the following: 1) Poor symptom control: ACQ consistently >1.5, ACT <20 (or “not well controlled” by NAEPP/GINA guidelines); 2) Frequent severe exacerbations: two or more bursts of systemic CS (>3 days each) in the previous year; 3) Serious exacerbations: at least one hospitalisation, ICU stay or mechanical ventilation in the previous year; 4) Airflow limitation: after appropriate bronchodilator withdrawal FEV1 <80% predicted (in the face of reduced FEV1/FVC defined as less than the lower limit of normal); 5) Controlled asthma that worsens on tapering of these high doses of ICS or systemic CS (or additional biologics).

Inherent in the definition of severe asthma is the exclusion of individuals who present with “difficult asthma” in whom appropriate diagnosis and/or treatment of confounders (comorbidity, adherence, psychosocial problems, etc.) improves their current condition. Therefore, it is recommended that patients presenting with “difficult asthma” have their asthma diagnosis confirmed and be evaluated by an asthma specialist for more than 3 months. Patients with confirmed severe asthma should receive an individualised treatment plan after a detailed and invasive protocol of investigations. Therapeutic options can be divided into medications used in lower doses for treatment of confounders (comorbidity, adherence, psychosocial problems, etc.) improves their current condition. Therefore, it is recommended that patients presenting with “difficult asthma” have their asthma diagnosis confirmed and be evaluated by an asthma specialist for more than 3 months. Patients with confirmed severe asthma should receive an individualised treatment plan after a detailed and invasive protocol of investigations. Therapeutic options can be divided into medications used in lower doses for children with less severe asthma, and those used in other paediatric diseases but not for asthma. Most treatments are unlicensed and the evidence base is poor. International collaborations, using standard protocols of investigation, will be essential if the mechanisms of severe therapy resistant asthma are to be understood, and evidence-based treatment delivered.

Reference

A24 Bioethics problems with extremely preterm infants (EPI) at birth
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Background: Each year, about 500,000 children are born in Italy and less than a thousand with a gestational age less than 26 weeks. In the past, they would very often be considered miscarriages or stillbirths. In recent years there has been a significant increase in their survival and the threshold of viability has gradually lowered to 22 weeks. The first bioethical issue on the health care of these infants involved the decision to start active care at birth. In Europe, about 19 scientific societies have published national guidelines on the care of infants < 26 weeks at birth.

Materials and methods: The guidelines of 19 European countries on the resuscitation of EPI at birth were evaluated and some considerations were made regarding the Italian situation. The diagnosis is complex, especially in early adolescence (8-12 years), because of the extreme heterogeneity of symptomatic expressions, which doesn’t allow a precise nosographic assignment [3,7-9]. The consequent diagnostic delay has a negative influence on the course of treatment and prognosis, making recoveries less and less frequent [10-12].

The role of the Family Pediatrics is, therefore, essential to intercept, through simple diagnostic tests (such as EAT-26) the first signs of these conditions, because from this depends on the subsequent diagnosis, therapy and prognosis [13-18] (Table 1).

The first task is to suspect a FED and to assess the differential diagnosis or comorbidity with other organic or mental diseases [2,3,6,19-21] (Table 2). The second task is to assess the severity of the problem for both organic and psychic aspects, in order to formulate an operational program of diagnosis, the emergency.

We propose to distinguish three steps of increasing severity, with which the diagnosis, the emergency. The suspect, includes those patients who have just embarked on dangerous or insane practices to lose weight without falling in any of the diagnostic categories of DMS-5 [1]. These patients need an educational intervention that can be done by the pediatrician (Table 3).

The diagnosis, includes cases that fully meet the diagnostic criteria of DMS-5 [1], without showing signs of serious and immediate biological or psychological risk. Such patients can be initially helped through the motivational interviewing [23] and subsequently entrusted to a multidisciplinary team, which also takes care of the family, promoting inter and intra-family relationship [11,24]. The emergency, includes patients in serious condition for which is indicated urgently indicated a taking in charge by a multi-professional team, possibly with an ICU admission, inpatient or outpatient (Table 4).

Table 1(abstract A25) The most significant questions in suspicion of FED among adolescent. Traits items of EAT-26 which correlate with positive total scores

<table>
<thead>
<tr>
<th>Question</th>
<th>Positive Total Score</th>
</tr>
</thead>
<tbody>
<tr>
<td>How many diets have you begun in last year?</td>
<td></td>
</tr>
<tr>
<td>Do you think, you should be on diet?</td>
<td></td>
</tr>
<tr>
<td>Do you feel dissatisfied of the weight of your body?</td>
<td></td>
</tr>
<tr>
<td>The weight influences the idea that you have of yourself?</td>
<td></td>
</tr>
</tbody>
</table>
Table 2(abstract A25) Differential diagnosis with other organic diseases

Endocrine: hyperthyroidism, diabetes mellitus, Addison’s disease, Simmonds syndrome
Gastrointestinal: achalasia, celiac disease, chronic inflammatory bowel disease, giardiasis and other malabsorption
Gynecological: pregnancy, other causes of amenorrhea
Infectious: AIDS, fungal infections, tuberculosis, subacute bacterial endocarditis
Neoplastic: meningiomas and any type of malignant tumor
Drugs: amphetamine, thyroid hormones, antidepressants, tricyclic, neuroleptics, lithium

Table 3(abstract A25) Educational Intervention by the family pediatrician (Health budget for FED). Valuations/informations relative to the following items

Balanced nutrition and health
Caloric needs
Satisfaction of the body image
"Necessary" and "dangerous" food
Using compensation mechanisms to bingeing (vomiting, compulsive motor activity),
Use of drugs
Family, social, emotional relations

Table 4(abstract A25) The indications of hospitalization

I. Biological decompensation (includes all the serious organic conditions)
   a. Serious weight loss (25-40%)
   b. Rapidly evolutive weight loss
c. Total refusal of food
d. Serious complications of malnutrition as syncopations, convulsions, cardiac arrhythmias or congestive heart failure, dehydration, acrocyanosis, instability of physiological parameters (Systolic Blood Pressure ≤ 90 mmHg, Heart Rate ≤ 40 / min, body temperature ≤36°C)

II. Psychological decompensation (includes all high risk situations and the psychiatric comorbidities)
   a. Suicide attempts
   b. Self-mutilations
   c. Abuse of drugs or other substances
   d. Severe depression
e. Anxiety
f. Obsessive-compulsive personality disorder Borderline personality disorder
g. Sexual or physical abuse

III. Other situations
   a. Failure of outpatient treatment, after attempt of 2-3 months without any modifications in the clinical picture
   b. Problematic family situation
c. Request from the patient or from his family

References
The epidemic spread of obesity in the last twenty years has led in pediatric setting to the appearance of diseases previously considered a prerogative of adulthood, such as metabolic syndrome (MetS). The Metabolic Syndrome is characterized by a cluster of cardiometabolic abnormalities, including visceral obesity, dyslipidemia, hypertension and diabetes mellitus type 2, that directly increase the risk of develop cardiovascular disease and diabetes [1]. Although the pathophysiological mechanism underlying the development of MetS is still only partially understood, the most widely accepted hypothesis identify in insulin-resistance and excessive production of free fatty acids (FFAs) the key components in the development of this disease [2]. Currently, several definition of Metabolic Syndrome are available in pediatric setting, causing confusion and discrepancy in the identification of these patients. Several studies have clearly demonstrated that the prevalence of MetS in the pediatric age may widely vary using different definitions, ranging from 2.2% to 52.1% among different studies [3]. Moreover, in the last years, several other co-morbidities, besides those traditionally used to define Metabolic Syndrome, that are also linked to the disease, have been identified, making its definition even more difficult. Among these, mainly non-alcoholic fatty liver disease (NAFLD) and obstructive sleep disorders (OSAS) have been strictly linked to Metabolic Syndrome. Lifestyle modification, based on regular physical exercise and a balanced diet appropriate for age, is the mainstay of therapeutic approach in children and adolescent with obesity and risk factors for MetS [4]. Behavioral intervention is often difficult to achieve and maintain and most pediatric patients require pharmacologic therapy early in their disease course. Given the relatively recent occurrence of MS in childhood, long-term follow-up studies are not available yet. However, it is reasonable to think that the metabolic derangement observed in obese children will have dramatic repercussions on their health earlier than that observed in adults, with a consequent worsening of the prognosis in terms of morbidity and mortality when they are still youth.

References

Disk battery ingestion: high clinic risk
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Over the last ten years disk battery (DB) ingestion have been increasing in children with serious consequences due to the diffusion of lithium battery (LB) that may cause catastrophic damages when lodged in the esophagus. The severity of injury depends on cell type, size, voltage, location and time of contact with the mucosa because electrical generation of hydroxide ions at the negative pole, leakage of alkaline content in stomach and mechanical pressure.

Figure 1 (abstract A27) Algorithms for the management of ingested Disk Batteries in children (Lithium Batteries or Alkaline Batteries). 1. The follow-up, above all in case of esophageal lesion, should monitor possible late onset esophageal perforation or vessel fistula. 2. Consider all symptoms, excluding bleeding. 3. Endoscopy can be postponed within 48 hrs in not passed cells; reduce the waiting time in case of alkaline battery, very young age or not witness ingestion. DB: disk battery; GI: gastrointestinal, CV: cardio-vascular; FBC: full blood count; OR: operating room equipped for cardio-vascular surgery; CT angiogram computed tomography angiogram; F-up: follow-up
Because LB are larger (> 20 mm), flatter and have an higher voltage (3V) than alkaline DB (1.5 V) in small children their ingestion increases the risk of esophageal lodgment and tissue damage in just two hours [1,2]. DB ingestion is not witnessed in 92% of fatal outcomes and 56% of major complications; 36% of patients with esophageal lodgment are initially asymptomatic [3]. Clinical presentation can be variable from absence of symptoms to drooling, dysphagia, vomiting, chest pain, or dyspnea, fever, abdominal pain, irritability and feeding refusal and sudden fatal exanguination for a fistula between esophagus and mediastinic vessels [3,4]. Other complications are trachea-esophageal fistula, laryngeal/esophageal stenosis, esophageal perforation, vocal cord paralysis, tracheomalacia, aspiration pneumonia, empyema, lung abscess, and spondylodiscitis [2]. Complications can be delayed, as the mucosal lesions may worsen also after DB removal. Plain chest and abdomen X-ray have a primary role to address the diagnosis and locate DB, revealed by the double ring or “halo” effect. A “sentinel bleed”, isolated hematemesis/melena occurring hours or days before a fatal hemorrhage, is another atypical presenting symptom [4]. Exsanguination can occur with the DB still in the GI tract or until 28 days after its removal [1,2]. We propose a new protocol for DB ingestion management in children and stress the necessity of prevention with public awareness campaigns promoted by scientific Societies and preventive information addressed to parents and caregivers [3].

References

A28 Congenital heart defects in genetics syndromes
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The majority of congenital heart defects (CHDs) occur as isolated malformations, while approximately 25-30% of them are associated with extracardiac anomalies, in the setting of large or submicroscopic chromosomal anomalies, mendelian disorders, and malformation associations. Some types of CHD, such as atrioventricular canal defect and interrupted aortic arch, are more frequently found in association with genetic syndromes, whereas other types are prevalently isolated defects (tricuspid atresia, transposition of the great arteries, pulmonary atresia).

Epidemiological studies, clinical observations and recent advances in molecular genetics are all contributing to the understanding of their etiology and pathogenesis. Several phenotype-genotype correlation studies suggest that specific morphogenetic mechanisms put in motion by genes involved in the etiology of the CHD, and mutations of this isolated gene is often manifesting as non-syndromic malformation.

These observations have several clinical implications. In fact, distinct cardiac anatomic subtype may help in suggesting accurate diagnoses, which can be confirmed by molecular testing. In addition, a multidisciplinary approach, checked for the risk factors related to specific genetic syndromes, can be used in the patients' follow-up and treatment. Different surgical prognoses have been found in patients with CHD and some genetic syndromes, as in patients with non-syndromic CHDs.

A29 Cow’s milk proteins allergy: the latest on therapy
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The progressive increase in incidence in cow’s milk proteins allergy (CMPA) in the past decades required primary prevention strategies for children at high-risk. Evidence of the role of gut microbiota in promoting the maturation of the immune system during early life encouraged supplementing the diet with probiotics in order to facilitate tolerance and delay or prevent sensitization. The efficacy of this strategy has not been consistently proven [1]. Breastfeeding is the most common therapeutic approach to CMPA in infants, although recent data showed that human milk has no effect on the development of allergy [2]. Use of special formulas is recommended in infants who are allergic or at high risk for CMPA. Extensively hydrolysed formulas (eHF) are the first therapeutic option. Amino acid-based formulas (AAFs) are recommended in infants who fail to respond to eHF, or have poor growth and IgE-mediated gastrointestinal disorders, or severe atopic eczema [3]. Hydrolysed formulas that do not originate from CMP are tolerated in 90% of children with CMPA. Use of soy milk is contraindicated in the first six months of life because of allergenic proteins and the presence of phytates and phytoestrogens [4]. The primary therapy for CMPA remains a strict avoidance of CMP, which promotes natural acquisition of tolerance in 80% of cases within the first 3 years of life. When the reintroduction of food causes severe clinical manifestations, diet restriction cannot be considered a solution, but instead exposes the infant to the constant danger of accidental exposure. Oral Immunotherapy (OIT) in food allergy requires the oral administration of increasing amounts of food up to a target dose, with the aim of reaching tolerance acquisition, which is considered complete when it is fully dose-independent [5]. OIT guidelines are not yet available. In severe, IgE-mediated clinical reactions to CMP, the combined therapy with Omalizumab, before and during “rush” OIT, reduced the number of severe adverse reactions and the duration of therapy while enhancing the possibilities of tolerance acquisition [6]. The use of interferon-gamma (IFN-γ) administered subcutaneously, as adjuvant in oral immunotherapy (OIT + IFN-γ), is a recent treatment. Unlike Omalizumab, the efficacy of IFN-γ has been demonstrated in non-IgE-mediated food allergy. IFN-γ seems to play a key role in the induction of tolerance but not in its maintenance. In studies on IgE-mediated CMPA, duration of treatment in patients with combined therapy (OIT + IFN-γ) is 2-3 months compared to 6-12 months for patients treated exclusively with OIT [7].

References
Influenza vaccination in pediatric age

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Influenza is a very common disease among infants and young children, with a considerable clinical and socioeconomic impact [1]. To reduce the direct and indirect effects of pediatric influenza virus infection, influenza vaccination is recommended worldwide for children considered at risk due to a severe underlying disease. In healthy children influenza vaccination is recommended only in a small number of countries, although guidelines vary regarding the minimum age [2]. In USA universal influenza vaccination in all the age groups is recommended, including all the children until 17 years of age. In UK, influenza vaccination is recommended in the age group 4-17 years, although the program has been activated only recently. Finally, in Canada and other European countries in which health authorities recommend the vaccine also in the healthy pediatric population, school-age children and adolescents are excluded. However, a large number of European health authorities is still reluctant to include influenza vaccination in their national vaccination programs [2]. The reasons for this reluctance include the fact that the protection offered by the currently available vaccines is considered poor, particularly in younger children. Regarding immunogenicity, younger children are quite similar to the elderly, who, because of the senescence of their immune system, respond poorly to immune stimulation [3]. In both these groups of subjects, both the innate and adaptive immune system are poorly functioning. In particular, B-cells, that are essential for antibody production and immune memory, have limited responses. To increase the immune response of children to inactivated vaccines, a number of measures that have been tested and found to be effective in adults and in the elderly have been studied [3]. The use of adjuvanted vaccines, intradermal (ID) injection, the administration of an increased dose of antigens and the live attenuated influenza vaccine (LAIV) have been evaluated in controlled clinical trials, with good results [3]. Moreover, the possibility of protecting young children through the use of a quadrivalent influenza vaccine (QIV) has been evaluated [4]. None of these measures has been definitively accepted because of the fear of an increased risk of adverse events and because in some instances data regarding immunogenicity and/or clinical efficacy are lacking or are not completely convincing. However, the preliminary data are very interesting in some cases and suggest that some of these measures must be further developed if the problem of the poor protection of young infants has to be solved.

References

A31
Which treatment for upper respiratory tract infections?

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Upper Respiratory Tract Infections (URTIs) include rhinosinusitis, acute otitis media (AOM), pharyngotonsillitis and laryngitis [1]. Viruses are responsible for the great majority of URTIs therefore antimicrobial treatment is not always required [2]. Paracetamol (7-15 mg/kg/dose) and ibuprofen (4-10 mg/kg/dose) are considered as the standard analgesics [3].

Regarding rhinosinusitis, clinicians should suspect a bacterial etiology when a child presents with persistent, worsening or severe illness. S. pneumoniae, H. influenzae and M. catarrhalis are the most common isolated bacteria. Amoxicillin (50 mg/kg/day) alone or with clavulenate is the first line antibiotic. Ceftriaxone (50 mg/kg/day) should be given to children who cannot take oral medications. Duration of the treatment varies from 10 to 28 days. The antibiotic may be changed if the symptoms get worse or do not improve within 72 hours (cefotaxime 8 mg/kg/day) [4-6].

AOM is an inflammatory disease of the middle ear involving the tympanic cavity frequently caused by S. pneumoniae, H. influenzae and M. catarrhalis. Diagnostic criteria for AOM are:

- moderate to severe bulging of TM or new onset of otorrhea not due to acute otitis externa;
- mild bulging of the TM and recent onset of earache or intense erythema of the TM.

Pediatricians should prescribe antibiotics in children aged <6 months with both severe and moderate presentations, in children aged between 6 and 24 months with severe presentation in both unilateral and bilateral AOM or in those with moderate presentation in bilateral AOM. Children aged >24 months need antibiotic therapy only when the presentation is severe [7,8]. Amoxicillin (50 mg/kg/day) alone or with clavulenate is the first choice in moderate and severe presentation respectively. Alternative initial antibiotics include cefaclor (40-50 mg/kg/day) and cefuroxime-axetil (30 mg/kg/day) or cefpodoxime-proxetil (8 mg/kg/day) respectively. Ceftriaxone (50 mg/kg/day) can be given to children who cannot take oral medications or when the symptoms do not improve within 72 hours [9]. The duration of the treatment may vary from 5 to 10 days [8].

Most pharyngitis episodes are caused by viruses. Antibiotic therapy is recommended in every child with microbiologically documented group A β-hemolytic streptococcus pharyngitis (37%). The first-line treatment is amoxicillin (50 mg/kg/day for 10 days). In non-compliant cases, cefaclor (40 mg/kg/day) or cefuroxime-axetil (20-30 mg/kg/day) may be administered [10].

Epiglottitis is a supraglottic laryngitis. It may be caused by S. pneumonia, S. aureus, β-hemolytic streptococcus and H. influenzae. The priority is airway management followed by antibiotic (ceftriaxone 50-75 mg/kg/day) treatment and steroids [11].

References


A32
The most common errors in the management of gastroesophageal reflux
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Gastro-Esophageal Reflux (GER) occurs in more than two-thirds of otherwise healthy children and is one of the major reasons for referral to pediatricians; in fact, a quarter of all paediatric visits in the first six months of life [1] and numerous accesses to the paediatric gastroenterologist services are secondary to GER. GER is defined as the passage of gastric contents into the esophagus and is different from gastroesophageal reflux disease (GERD), which includes all the symptoms and/or complications associated with GER [2].

GER is considered a physiological normal process which occurs several times a day in healthy infants, children and adults. GER is usually associated with transient lower esophageal sphincter relaxations independent from swallowing, which allow the passage of gastric contents into the esophagus. Episodes of GER in healthy adults tend to occur after meals, lasting less than three minutes and usually do not cause symptoms. Although we know less on the normal physiology of newborns and infants, regurgitation is the most visible symptom and tends to occur daily in 50% of all infants with a peak incidence between 4 and 6 months of life [3].

For the majority of paediatric patients (especially in infants), the clinical history and the objective appraisal, in the absence of danger signals, are sufficient to reliably diagnose a not complicated GER and initiate conservative treatment strategies; in general, the diagnostic tests are not always necessary. The reliability of the symptoms to make the clinical diagnosis of GERD is higher in children younger than 8 years, reporting heartburn: only in this case, the doctor can make a diagnosis of Gastro Esophageal Reflux Syndrome and give an indication for therapy [4]. To date, no single symptom or set of symptoms can be trusted, and then used to diagnose GERD in children or to predict which patients are more likely to respond to therapy [2]. The only exception is the child (age> 8y. o.) reporting a history of long-term heartburn with or without vomiting.

The new ESPGHAN/NASPGHAN guidelines [2] describe different treatment options for the treatment of infants / children with GER and GERD. In particular, they emphasized the changes in lifestyle, because it can effectively reduce the symptoms of both infants and children. They are based on a combination of changes of milk formulas and positional therapy. In infants with GER, it can be effective the changes to the mother’s diet if children are breastfed, change the formula in use, reducing the volume of the single meal associated with an increased frequency of feedings. In particular, the guidelines stress that the allergy to milk proteins may have a clinical presentation similar to GERD. A strategy of nutritional intervention involves the use of thickened formulas that are able to decrease the regurgitation. A recent meta-analysis has shown that the anti regurgitation formula have the following functions: a) increase the number of infants without regurgitation; b) reduce the number of episodes of regurgitation and vomiting daily; c) increase the weight recovery of the infant, d) although they do not change the 24 hours pH study [4].

References

A33
Cases of usual ultrasound
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Background: Ultrasound (US) can be decisive for diagnosis, in some situations, in others the contribution of the US is only complementary and must be integrated with other diagnostic tests. Finally, in some cases, the ultrasound imaging does not provide solutions, but offers questions. The clinic, experience, and knowledge can find appropriate solutions.

When an ultrasound is a diagnosis: A male newborn of 3700 g, born after 40 weeks’ gestation via a vaginal dystotic delivery, Apgar 8 and 10 to 1’and 5’, had a fracture of the right clavicle. After about 48 hours he was admitted to our department for jaundice and hematoma of the right hemiscrotum (Fig.1), with no pain, no other symptoms. Scrotal Doppler US showed a normal testicle size and shape, regularly vascularized, but modest collection of fluid, finely corpusculated and marked thickening of subcutaneous tissue. Abdomen US revealed a complex mass upper pole of the right kidney (2.8 cm), not vascularized, withinhomogeneous echogenicity (Fig.2). Ultrasonography had been sufficient to make the diagnosis of adrenal hemorrhage with hemorrhagic spreading at right hemiscrotum. Were not performed further investigations and a conservative treatment was chosen. Ultrasound monitoring showed progressive organization of the hematoma and complete resolution after about 40 days.

Figure 1(abstract A33) Hematoma of the right hemiscrotum
When the ultrasound helps in diagnosis: A boy, six years old, previously healthy, about 15 days ago was treated with oral antibiotics for UTI. An ultrasound is performed because urinary frequency, dysuria, urgency, gross hematuria reappeared. US revealed one large stone bladder (Fig.3); microscopy of the urine sediments showed cystine crystals. Stone, removed from the bladder endoscopically, were composed of cystine and calcium oxalate, when analysed.

When the ultrasound raises clinical questions: Routine fetal ultrasound screening reports dilatation of the urinary tract in about 1-2% of all pregnancies, most of these are mild or moderate, limited to the renal pelvis, calyceal and ureter is not seen, the bladder is normal, renal parenchyma have normal thickness and appearance. These children do not have an obstructive pathology, nor a predisposition to UTI, and in most cases the dilatation is transient and has no pathological significance. Only clinical experience and up to date knowledge avoids these children to undergo invasive, painful and expensive imaging techniques, or to strenuous follow-up. Written informed consent for publication of clinical details and clinical images was obtained from the parents of the patients.

A34
The follow-up of preterm infant after discharge: family pediatrician (FP) medical viewpoint
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Survival rates and outcomes of infant and preterm infants have radically improved thanks to the most recent techniques of resuscitation and intensive care. Premature babies are exposed to an higher risk of growth problems, delayed development or complex medical problems and, compared to other children, have a higher prevalence of severe disability [1]. In order to discharge premature infants from neonatal intensive care we have to consider some critical aspects [2]. The active and early involvement of family pediatrician (FP) is essential to ensure therapeutic success . These infants represent a small portion of births (0.3 % among the choices of a FP), but they’re becoming more frequent . The definition of an individualized and shared assistance and follow-up program requires the establishment of a effective flow of information that encourages a two-way path of information and patients . In low complexity and risk cases, child and family should be assigned to a team consisting of FP, child psychiatrist and other health professionals for rehabilitation in residence territory.

The primary objective is to promote especially the early management of infants with gestational age less than 28 weeks, discharged from neonatal intensive care, favoring, before being discharged from the hospital, family involvement, integration between hospital professionals and local services involved in determination and implementation of an Individual Support Plan (IAP), in order to follow the child in his first year of life. FP during the specific and agreed health statements can evaluate and closely monitor growth, development, neuro developmental, visual and hearing functions and any medical problems still unresolved. The preterm infant must be subjected, according to the chronological age, to all recommended vaccinations, considering too the influenza vaccination. The FP figure is crucial to promote, encourage and support vaccination and preventive route (SRV).

FP must improve specific skills to be able to welcome and assist the premature infant and his family avoiding their unnecessary trips to hospitals. It is also important to provide intra and interprofessional training processes to promote team work. A real integration between different levels of care and a definition of roles and professional tasks, might avoid efforts duplication and ensure continuity of care, reducing discomfort and uncertainty of families.

References

A35
Arterial blood pressure monitoring in children
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In 2008 the first set of consensus recommendations for performance and interpretation of 24 hours Arterial Blood Pressure Monitoring (ABPM) in children and adolescents have been published [1]. Since then, ABPM has found increasing use in pediatrics. These recommendations have been updated in 2014 [2]. For this reason the Group of Hypertension Study of the Italian Society of Pediatrics (GISPER) has felt the need to perform an update of the Italian recommendations on this topic. The ABPM should be used by experts who know how to run it and interpret it. Proper execution is in fact necessary and only trained staff can guarantee it. Children and parents should be educated on the significance of the examination and care should be taken in selection of the appropriate size cuff according to the size of the child’s arm. For the interpretation of the ABPM data, the age- and sex-specific percentiles of Wühl et al [3] are the preferred reference nomograms. Table 1 shows the suggesting schema for interpretation of ABPM values, in defining Blood Pressure categories. On the contrary that in the adult, the ABPM in children cannot be considered the gold standard for the diagnosis of high blood pressure, which must be done by measuring office Blood Pressure values, according to the criteria established by the National High Blood Pressure Education Program Working Group on High Blood Pressure in Children and Adolescents [4]. Other important differences compared to adults are using the pressure load (defined as pathological in the presence of a number of measurements of systolic or diastolic Blood Pressure values >25% of total) to define the different blood pressure categories and the presence of Pre-Hypertension among Blood Pressure categories. The ABPM also allows to identify individuals with White Coat or Masked Hypertension, clinical situations that, in children as in adults, suggest the need for careful follow-up. It was shown that both of these conditions can be associated with the presence of early organ damage, such as left ventricular hypertrophy in children. Finally, ABPM can give important information about Blood Pressure variability, distinguishing subjects with normal nocturnal Blood Pressure dip (>10% compared to the day, dipping), from non-dipping children.

References

Table 1(abstract A35) Suggesting schema for Ambulatory Blood Pressure levels interpretation in children (modified by ref.2)

<table>
<thead>
<tr>
<th>CLASSIFICATION</th>
<th>OFFICE BLOOD PRESSURE: &lt;90th percentile</th>
<th>AMBULATORY SYSTOLIC OR DIASTOLIC BLOOD PRESSURE: &gt;95th percentile</th>
<th>SYSTOLIC OR DIASTOLIC LOAD: &lt;25%</th>
</tr>
</thead>
<tbody>
<tr>
<td>Normal Blood Pressure</td>
<td>&lt;90th percentile</td>
<td>&gt;95th percentile</td>
<td>&lt;25%</td>
</tr>
<tr>
<td>White Coat Hypertension</td>
<td>&gt;95th percentile</td>
<td>&gt;95th percentile</td>
<td>&lt;25%</td>
</tr>
<tr>
<td>Pre-Hypertension</td>
<td>&gt;90th percentile or &gt;120/80 mmHg</td>
<td>&gt;95th percentile</td>
<td>&gt;25%</td>
</tr>
<tr>
<td>Masked Hypertension</td>
<td>&lt;90th percentile</td>
<td>&gt;95th percentile</td>
<td>&gt;25%</td>
</tr>
<tr>
<td>Ambulatory Hypertension</td>
<td>&gt;95th percentile</td>
<td>&gt;95th percentile</td>
<td>25-50%</td>
</tr>
<tr>
<td>Severe ambulatory Hypertension</td>
<td>&lt;95th percentile</td>
<td>&gt;95th percentile</td>
<td>&gt;50%</td>
</tr>
</tbody>
</table>
Arterial hypertension and sport. Related aspects to certification for physical activity and contraindications to sports practice in hypertensive child and adolescent

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The control of blood pressure during exercise is a complex process as it involves increases in stroke volume and heart rate, changes in peripheral vascular resistance and in sympathetic tone; it is related to the type of exercise practiced as its intensity and duration, ratio of lean mass/fat mass, depending on the sex. We distinguish two main types of exercise, aerobic or dynamic and isometric or static. The first is a type of exercise that involves an increase in cardiac output associated with rapid increases in heart rate and systolic BP (with a minimum decrease in diastolic BP) but with a significant decrease in peripheral vascular resistance. The isometric exercise, or static, involves a sharp increase in both systolic and diastolic blood pressure, a modest increase in heart rate with a range stable or slightly reduced but no decrease in peripheral vascular resistance. In Figure 1 are shown the modifications and interactions between the various parameters [1].

From these premises it emerges therefore the need to start young people suffering for arterial hypertension to play a type of sport that is suitable for the individual cardiovascular status.

In Italy, compared to the US, to practice a sport activity is required medical certification involving a civil and criminal liability on the part of the physician certification and which is regulated by specific decrees, in particular the latest amendments to the Balduzzi Decree [2]. In the case of healthy children, or apparently healthy, it is scheduled to perform at least once a 12-lead ECG at rest and blood pressure measurement (as specifically stated on the label of certification then signed at the bottom by a doctor). In the case of children and adolescents with arterial hypertension, both for primary and secondary hypertension, you need to pay more attention and care, both in relation to cardiovascular stresses that this entails as previously exposed, both in relation to the importance of implementing the changes in lifestyle who are always the first step, no drugs, which will contribute to the maintenance of blood pressure in the normal range. For what concerns instead the practice of competitive sports activities, please refer to specialists in Sports Medicine, which is reserved for the certification.

Acknowledgements: This is a contribution to the joint recommendations of the Italian Society of Pediatrics and the Italian Society of Hypertension. Arterial hypertension and physical activity: aspects of prevention, treatment and certification. GSIPER. Update 2015

References

A38
Intellectual disability in developmental age
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Intellectual disability (ID) is a neurodevelopmental disorder characterized by deficits in intellectual and adaptive functioning that present before 18 years of age [1]. ID is heterogeneous in etiology and encompasses a broad spectrum of functioning, disability, needs and strengths. Originally formulated in strictly psychometric terms as performance greater than 2.5 SDs below the mean on intelligence testing, the conceptualisation of ID has been extended to include defects in adaptive behaviours [2]. The term global developmental delay (GDD) is usually used to describe children younger than 5-years of age who fail to meet expected developmental milestones in multiple areas of intellectual functioning [1]. In both conditions the symptoms must be present in the early developmental period, but they may not become fully manifest until social demands exceed patients’ capacities.

ID affects 1.5 to 2% of the population in Western countries and represents an important health burden [3]. During the past decade, advances in genetic research have enabled genomewide discovery of chromosomal copy-number and single-nucleotide changes in patients with ID and autism as well as in those with other neurodevelopmental disorders. These technological advances—which include array comparative genomic hybridization (CGH), single nucleotide polymorphism genotyping arrays and massively parallel sequencing—have transformed the approach to the identification of etiologic genes and genomic rearrangements in the research laboratory and are now being applied in the clinical diagnostic arena [4]. In this view, the American Academy of Pediatrics recently released a guidance for the clinician in rendering Pediatric Care [5]. The suggested clinical approach to the patient should be conducted closely with a geneticist and includes the child’s medical history, the family history, the physical and neurologic examinations (emphasizing the dysmorphology examination) and the examination for neurologic or behavioral signs that might suggest a specific recognizable syndrome or diagnosis. After this clinical evaluation, focused use of genetic laboratory tests, imaging and other consultations are critical in establishing the right diagnosis, its pattern of inheritance and the subsequent follow-up.

Finally, this guidance highlights a renewed emphasis on array CGH, that is now considered the first-line diagnostic test for children who present with GDD/ID of unknown cause, and on the identification of treatable-causes of GDD/ID with the recommendation to consider screening for inborn errors of metabolism [5]. The future use of whole-genome or next generation sequencing offers promises and challenges needing to be yet addressed before their regular implementation in the clinic.

References

A39
Staff management in Paediatric Primary care setting
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In the last 6-7 years there has been a progressive increase in the use of nurses or secretaries in Paediatric Primary care setting.

Regional incentives (few) and the rising (remarkable) of the workload and patients’ requests, convinced about the 40% of the Italian Family Paediatricians (FP) to hire new staff. In the 70% of cases the employers were organized in working group in one private practice that, thanks to the costs’ sharing, frequently had nurses in their staff (the data represent the situation at the end of 2013 and were obtained asking directly FP’s professional organizations). Solo Paediatricians and Associated Paediatricians (that work in several private practices in coordination each other), on the other hand, hired more frequently secretaries with a working time generally reduced if compared to working groups’ colleagues.

Economic factors (the fear to be unable to fulfill the expenses related to hiring) and those concerning organization and management (the not perfect knowledge of how best to use the staff) are the main causes that prevent most of Paediatricians to employ [1]. Who decided to use co-workers, did it to delegate certain duties, to filter patients’ requirements, to share work experience [1].

Secretaries can officiated the following tasks: sorting incoming calls, appointments, advise on admission to medical consultation, set up patient’s recording (1st consultation), illustration and delivery of the

Figure 1 (abstract A37) Cardiovascular response to physical exercise Legend: (A) Response to dynamic exercise gradually to the increase in workload to the maximum oxygen consumption; (B) Response to static exercise (handgrip at 30% of maximum voluntary contraction); VO2 (ml/min/kg); HR (bpm); Stroke volume; ABP (mmHg): systolic, diastolic and mean arterial pressure; TPR (PRU) peripheral vascular resistances (expressed in peripheral resistance Unit).
services charter, delivery questionnaires, registration of diagnostic services, recurrent prescriptions, preparation of medical certificates, patients’ active call for programmed medical examinations, billing, payment deadlines and commitments, check of the monthly payment slips obtained by the Health Service, registration deadlines of drugs and medical devices, suppliers list, maintenance men and contracts list, health services summaries, statistics, chart statistic diagrams, waiting room monitoring (games, cartoons, magazines, books, etc.), appointment calendar update.

Nurses can perform the following functions: Telephone Triage; Telephone advices based on defined protocols that are recorded in the electronic patient health card, axiological recordings, performing diagnostic tests and screening (urine examination, rapid Strep test, RCP, Blood Count, Prick Test, oximetry, spirometry, eye test, Lang test, Boel test, Plicometry, etc.), infant care, clinical and epidemiological research, health education messages in the waiting room, drugs and medical devices deadlines monitoring, provisions supervision, medical equipment efficiency preservation, health care protocols update and recording, check office’s e-mails.

In the near future the decrease of FP’s number, the need to take care of a larger number of patients and to organize work in some way of collaboration, will push more and more colleagues to hire staff.

In the near future the decrease of FP’s number, the need to take care of a larger number of patients and to organize work in some way of collaboration, will push more and more colleagues to hire staff.

Reference
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A40
The Convention on the rights of persons with disabilities and newborns/children
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The Convention on the rights of persons with disabilities of United Nations (CRPD 2006) is an international standard, but not enough know in Italy. The CRPD introduce a strong change of paradigm in the approach on persons with disabilities. Based on the respect of human rights, is innovative regard the Convention on the rights of the child (1989). Redefining the concept of disability, stress the importance of the social and relationship components. For the minors with disabilities means a new responsibility of the pediatricians on the social components that can determine factors of impoverishment or empowerment of capacities and competences. New concepts (related to new treatment) are stressed: from the empowerment to habilitation and to capability. The relationship with the family too from the curative attention, often totally transfer to specialism, to educational responsibility, guiding to the empowerment of capacities and competences. New concepts (related to the Convention on the rights of persons with disabilities) that are part of the human kind.

A41
Professional training and research: which resources?
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The National Plan of Prevention (PNP 2014-2018) [1], which is the key document for the Italian Public Health, underlines the need to realize for all people an empowerment level able to increase or maintain the health control. At international level, the Ljubljana Charter [2] contains the sanitary systems commitment to rearrange, in the aim to improve continually the quality, with reference to, in particular, the rate cost /efficacy of the system. The manual for medical practitioners and pediatricians (Ministry of Heath, 2010) [3] acknowledges the seven fields of skills for operators, as defined by the Canadian Patient Safety Institute, the CPSI. Continuous training for health professionals started as a national program in Italy in 2002; some years later (2007) administrative control had been transferred from the Ministry of Health to the National Agency for Health Services, Agenas. At distance training and online updating are now becoming very common, close to traditional kind of events for training, always in respect of the Evidence Based Medicine (EBM).

We can identify, in the continuous sanitary training system, different roles and various kinds of providers. Relevant organisations include the National Committee for Training, in the Ministry of Health, the Warranty Committee, and the National Observatory about Quality of Training.

We can identify, in the continuous sanitary training system, different roles and various kinds of providers. Relevant organisations include the National Committee for Training, in the Ministry of Health, the Warranty Committee, and the National Observatory about Quality of Training.

Regarding research in the field of health, we can consider it as the architrave of the National Health Service; the Report on Health Status in Italy 2012-13 [4] defines some essential aspects of it, such as the improvement of criteria of selection of the projects, to simplify bureaucratic procedures, evaluation criteria and the rapid spread of the results.

The National Committee for Sanitarian Research defines the program and initiatives, monitors and evaluates the results. Research institutes involved in Italy must accept different challenges, according with the Singapore Declaration [5], which defined some criteria about loyalty and professional honesty.

The European Union invested 80M Euro in research with Horizon 2020, in the aim of destroying barriers and realising, all over the world, a common environment concerning knowledge, research and innovation.

A recent piece of research related to the food habits in the age of complementary feeding is Nutritake 6/36 [6], the first Italian study which confirms some excesses and deficiencies in the diet. It suggests the opportunity of the definition of specific nutritional guidelines.

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A42
Early discharge of term neonates: we can do it safely
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The National Committee for Training, in the Ministry of Health, the Warranty Committee, and the National Observatory about Quality of Training.

The recent pronouncements of scientific societies [10] and the World Health Organization (2013) [11] together with legislation produced in our country (Progetto Obiettivo Materno Infantile 2000; Piano Sanitario Nazionale 2006 - 2008; Conferenza Unificata Stato-Regioni 2010, Raccomandazione n° 16 del Ministero della Salute 2014), can help identify criteria for appropriate and safe discharge of the mother-infant dyad. All efforts should be made to promote simultaneous mother-neonate discharge and the length of hospital stay should be based on the unique characteristics of each mother-infant dyad, including not only the health of the mother and the neonate but also the ability and confidence of the mother to care for her infant, the adequacy of support systems at home, and the access to appropriate follow-up care (Table 1).
Table 1 (abstract A42) Criteria to be met before discharge of a term neonate (modified from American Academy of Pediatrics 2015)

A) Neonatal health
1. Clinical course and physical examination at discharge have not revealed abnormalities that require continued hospitalization
2. Infant’s vital signs within normal ranges and stable for the 12 hours preceding discharge
3. The infant has urinated regularly and passed at least 1 stool spontaneously
4. The infant is able to coordinate sucking, swallowing, and breathing while feeding
5. The clinical risk of development of subsequent hyperbilirubinemia has been assessed, and appropriate management and/or follow-up plans have been instituted as recommended in guidelines for management of hyperbilirubinemia
6. The infant has been adequately evaluated and monitored for sepsis on the basis of maternal risk factors and in accordance with current guidelines for prevention of perinatal group B streptococcal disease
7. Availability and evaluation of maternal screening results for syphilis, hepatitis B, HIV and appropriate treatment instituted when needed
8. Newborn metabolic and hearing screenings completed

B) Maternal competency
1. Breastfeeding (positions, latch-on, efficacy of swallowing, importance and benefits) or bottle feeding
2. Appropriate urination and defecation frequency for the infant
3. Cord, skin, and genital care for the infant
4. Infant safety
5. The ability to recognize signs of illness and common infant problems, particularly jaundice

C) Assessment of family, environmental, and social risk factors and discussions with social services when plan to safeguard the infant is needed
1. Untreated parental substance abuse or positive urine toxicology results in the mother or newborn
2. History of child abuse or neglect or history of domestic violence
3. Mental illness in a parent who is in the home
4. Lack of social support, particularly for adolescent mother or single mother who live in a shelter, a rehabilitation home, or on the street
5. Communicable illness in a parent or other members of the household
6. Assessment of barriers to adequate follow-up care for the newborn, such as lack of transportation to medical care services or language barriers to make suitable arrangements to address the family

D) Plan for continuing medical care
1. Identification of medical services for postnatal checks
2. Date of first appointment after discharge
3. Planning bilirubin check or other individualized controls when needed

References

A43
Conventional medicine and complementary medicine: more similarities than contradictions
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Ilkka Tuomi said that it is impossible to divide knowledge into two clearly separated fields, one for expressed knowledge and the other for unexpressed knowledge [1] and this concept leads us to consider that the
differences between Complementary Medicines (CAM) and Conventional Medicine (CM) are often mistakenly given as true, so we will deepen this theme searching for common elements between CM and the most widespread CAM: herbal medicine, acupuncture and homeopathy. For herbal medicine concepts are simple: the herbal products are in practice normal drugs, with their therapeutic effects and their side effects, and their use follows the rules of pharmacology. For acupuncture we have now a modern theory to explain its mechanism of action, because recent works indicate that the insertion and rotation of the needles in the subcutaneous tissue is able to promote the production of cytokines with various biological effects, showing in a pathophysiological way the action of this practice [2].

Regarding homeopathy the speech is more complicated, possibly due to the theoretical structure at the base of it. Francois Laplantine [3] succeeded in his book, “Anthropology of Illness”, in envisaging a key of lecture surely original and interesting, showing that the transition between the homeopathic and allopathic therapeutic models is allowed.

In practice, the MC and homeopathic medicine are only in apparent contradiction. Here’s an example: behind the constitutional classification of individuals in Carbonic, Sulphuric and Phosphoric, established in Homeopathy and based on biomorphological parameters, there is a clear relationship with genetics, seen as individual predisposition to developing specific diseases, therefore mesoblasti (ed Sulphuric) are, in example, at increased risk of cardiovascular disease and endoblasti (ed Carbonic) are at greater risk of metabolic diseases [4,5].

Obviously it becomes indispensable to check if it’s possible that the assertion that behind one aspect of “morphological” type there is a corresponding aspect of the functional or, anyway, pathophysiological type (concept of Biomorphology). In example, the ratio between the length of the 2nd and 4th finger of the hand leads to differentiate individuals and their susceptibility to specific diseases [6].

We can comment that the same types of knowledge, ideas, innovations, are found among intellectuals, among the peasants in the villages, in the forests between the tribesmen and even in universities among scientists [7].

References

A44

Overweight and obesity: prevention in the first 1,000 days of life
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More than 30% of 9-10-year old Italian children are overweight or obese. The American Academy of Pediatrics as well as the Italian Society of Pediatrics proposed some specific targets for both prevention and treatment of childhood obesity. However, consistent evidence suggests that nutrition is the most promising among them. Nutritional intervention may be efficacious at every age but the most sensitive age window is the first 1,000 days of life, i.e., intrauterine period and the first two years after birth. In early life, the delicate phase of anatomic growth and functional development and specialization of the neuroendocrine system that regulates energy balance and body composition may be affected by nutrition. Unfavorable nutrition conditions, lead to a non-physiologic “metabolic programming”, i.e., the maturation of the biochemical pathways regulating metabolism and adiposity, also in a long term perspective. The epigenetic consequences induced by inadequate nutritional exposure makes individuals less resistant to the environmental obesogenic pressure, leading to a morbidity-prone phenotype. What are the nutritional targets in the first 2 years after birth? Prolonged breast feeding and, at weaning, provide complimentary food that are nutritionally adequate and safe. Diet composition should guarantee all macro- and micro-nutrients recommended for optimal growth. In particular, it should be avoided the excess of energy, protein and sugar in respect to requirement as well as the shortage of iron, vitamin D, and lipids, especially long-chain polyunsaturated fatty acids. Moreover, an accurate monitoring of weight and length growth should be always provided, to avoid undesirable acceleration of weight growth velocity in respect to length growth velocity, an important risk factor of obesity. Cow milk should be avoided at least in the first 12 months of life. Parents have a high responsibility in educating their children to correct eating behavior and the role of pediatricians to inform and guide parents in this delicate work is crucial.

References
Acute kidney injury (AKI), previously called acute renal failure, is characterized by an abrupt increase in the concentration of serum creatinine (SCr) and nitrogenous waste products and by the inability of the kidney to appropriately regulate fluid and electrolyte homeostasis. The incidence of AKI in children has increased over the last decades, and the etiology of AKI has evolved from primary renal disease to multifactorial causes due to major advances in the medical management of critical illnesses, such as solid organ and stem cell transplantation, corrective surgery for congenital heart disease, sepsis, and septic shock. AKI is not just a marker of illness severity in children, but has a direct association with poor outcomes. Even when the definitions and characterization of AKI in children have advanced significantly over the past two decades, the diagnosis of AKI is still made with surrogate markers of glomerular filtration rate, such as SCr and urine output [1, 2]. There are improved consensus multidimensional AKI definitions, namely the pRIFLE, AKIN and The KDIGO AKI definition and staging criteria being used worldwide, but with significant limitations. SCr continues to be quite limited as a marker of kidney dysfunction: it is often inaccurate in patients with low muscle mass, fluid overload or prior chronic kidney disease. Furthermore, SCr shows a demonstrable rise in concentration many hours to days after insult to the kidney, making it an insensitive, late and unmodifiable functional AKI marker. Thus, creatinine-based AKI diagnosis is often delayed, rendering the adverse effects of AKI until renal function recovery occurs [3, 4]. Some of these promising serum and urine biomarkers are CyC and NGAL, IL-18, KIM-1, and LFPAB respectively [5]. All of them have shown considerable promise diagnosing AKI earlier than SCr. Given that AKI is a complex and heterogeneous disease, it will probably be best diagnosed by a panel of biomarkers, rather than a single one. In the future, once these biomarker panels have been used widely in clinical practice and linked to clinical outcomes, it is likely that pediatric AKI will become defined by biomarker elevations and not by SCr [6].

References
Cardiovascular diseases (CVD), namely coronary artery disease and cerebrovascular accidents, are the main cause of mortality and morbidity worldwide [1]. Risk factors for CVD in adults are: hypertension, diabetes, hyperlipidemia, hyperuricemia, microalbuminuria, obesity and weight; over-eating, physical inactivity, tobacco smoking, exaggerated alcohol consumption, represent risk habits [2]. The WHO has promoted a program of prevention of CVD in adults [3]. It has been proved that risk factors for CVD are already present in childhood [4]; genetic and environment factors play a role and early intervention may produce a benefit [5]. Hypertension is present in a consistent percentage of adults and is related to high mortality rate. Criteria used for diagnosis of hypertension in children are more restrictive but it has been proved that adults with hypertension have high values of blood pressure in childhood [6]. Blood pressure measurement in children should be done according to standard procedures; ambulatory blood pressure (ABP) may be a better predictor of adverse cardiovascular events than office BP (OBP) [7]. Vascular biomarkers, which are parameters of subclinical cardiovascular disease, could increase the estimation of the individual cardiovascular risk and can be measured by non-invasive techniques [8]. Familial hypertension, low birth weight, high BMI; high sodium consumption, reduced physical activity in childhood are risk factors for hypertension in adulthood. Renal insufficiency is often associated with hypertension and it is well known that uremia represents an increased risk for CVD, but congenital renal disease may produce hypertension and CVD before glomerular filtration rate is reduced [9]. Pediatricians have a crucial role in preventing hypertension and CVD [10].

References
Taking an accurate three-generation family history is important when a genetic syndrome is suspected. Important elements include the age and sex of family members; when family members were affected by disease or when they died; the ethnic background; and if there is consanguinity. Every effort should be made to obtain complete information about the gestation leading to the birth of the affected child. Events surrounding the birth of the child may be critical: antepartum status, length of labor, mode of delivery, baby’s condition at birth, measurements. Finally, information is obtained about the child’s course since infancy along the lines of a standard pediatric history, but with a few special emphases: general health, growth, developmental progress, behavior, special testing or therapy.

The physical examination is of enormous importance: the basic techniques of physical diagnosis are used, but the examination is fine tuned to promote detection of many subtle physical clues that might otherwise be overlooked. Selected measurements of physical features can be extremely useful in confirming a clinical impression of abnormality. The diagnosis of a particular disorder can be based on clinical features, laboratory data, or a combination of both.

The provision of an accurate diagnosis is only the first step in the long process of care for a child with genetic syndrome and the pediatrician may have a continuing role in working with the patient and the family.

**A51**

**Informed consent and minors**

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The provision of informed consent by a patient should be the end point of a process of engagement in which one or more health practitioners have supported the patient to come to an informed decision to agree to the healthcare offered. Informed consent is a means to comply with the principle of respect for the person in a healthcare context. This act can be alternatively interpreted according to a libertarian perspective or according to a more solidaristic one [1]. According to the former perspective, informed consent is tightly linked to the idea that every person is the proprietor of their own body with which they have the absolute right to decide whatever they want done to it. This view leads to the construction of the patient physician relationship founded on a contractual basis. According to the latter perspective, respect for the person implies a special care and not, indifference, so that consent implies the due engagement of the person in the decision process as the very etymology of the word consent suggests. Respect for a person certainly means respect for his or her autonomy; but the former concept does not correspond necessarily with the latter, especially if autonomy is not interpreted within a framework of solidarity and the patient is not sufficiently mature. Otherwise respect would slide into indifference.

As for minors, the Oviedo convention prefers to use the word *authorisation* instead of the common expression *consent on behalf of the child*. In fact, authorisation relates to the concept of a third authority, such as parental responsibility, which implies a different framework than agreeing on any given issue. This is the concept of the child’s best interests as the fundamental criterion of making decisions regarding children. Since informed consent can be seen as an expression of personal choice, it can only be given by the person who is to be provided with health care. Moreover, the Oviedo convention requires that the opinion of the minor shall be taken into consideration. As a result, the decision-making process involves three categories of subjects: the physician who proposes the therapy, the parents who give authorisation and the minor whose opinion “is an increasingly determining factor in proportion of his or her degree of maturity” [2-5]. Further problems can derive from the fragile condition of modern-day families and the related difficulty in parents agreeing on any given issue.

**References**


**A52**

**The charter on the rights of hospitalized children with disability**

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As the United Nation Convention on the Rights of Persons with Disabilities states, People with disabilities have the same general health care needs as other patients, but they are more likely to find health care providers’ skill and facilities inadequate, to be denied health care, and to be treated badly in the health care systems [1]. In the United Kingdom several inquires report deaths of people with a learning disability, as consequences of healthcare inequalities [see https://www.mencap.org.uk/get-involved/campaigns/successes/our-fight-equal-healthcare].

Spes contra spem, a non profit organization, processed the charter of rights of people with disabilities in hospital [2]. The Charter shapes the rights set down in the European Charter of patients’ rights for people with disabilities.

The purpose was to have a tool to overcome the “sanitary barriers”: architectural, organizational and cultural. It based on the principle that people with disability do not have special rights but equal rights as the others people have. Only The ways of benefit of these rights are different, and it is a duty of society to put everyone in a position to be able to benefit, by removing the barriers that stand in the way. This is the application of the principles of equality and non-discrimination of United Nation Convention [3].

Recently, we evaluated the need for a specific charter of the rights of children with disabilities in hospital. Children with disabilities have some distinctive features compared to their peers without disability that should be highlighted.

As well as should be considered the cultural barriers about the disability, present in the care of children too. It is important that health professionals understand that disability in itself is not sufficient to deny a treatment or a cure. Frequently, inadequate medical treatment of children with disability is related to the belief that they have a poor quality of life [4]. So we decided to promote the charter on the rights of hospitalized children with disability. We decided to use an existing charter, the *Carta dei diritti del bambino in Ospedale* drawn up by “Fondazione per il Bambino in ospedale (ABIO)” and Italian Society of Pediatrics. The ten articles, unchanged in their formulation, should be declined taking due account the needs of children with disabilities and their parents.

We propose a road map for a development of the Charter that should be the result of the broadest possible sharing between pediatricians, nurses, the associations of disabled people and their families.

**References**


**A53**

**Children with rare diseases: do they really have an increased risk of developing epilepsy?**

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Symptoms involving the central nervous system (CNS) are frequently part of the variable clinical picture of genetic-dysmorphic syndromes. Intellectual disability represent the most common associated finding, but epilepsy is very frequently reported, for example in chromosomal abnormalities [1-3]. Seizure risk has been pointed out to be significantly higher in those with chromosomal abnormalities with respect to general population [4-6], and represent an important element to recognize and define for both diagnostic and therapeutic purposes. Seizures’ phenomenology, severity and frequency can be highly variable in different syndromes, depending on the type of genetic defect, but even amongst subjects with the same genotype there can be a great variability in clinical manifestations, even intramurally. Therefore, within the association between genetic syndromes and epilepsy, we should consider two possible situations:

1) Syndromes in which epilepsy could represent a “specific marker”. In certain conditions the recognition of a peculiar electroclinical phenotype could give a significant contribution to diagnostic definition of the syndrome. The best known example is Angelman syndrome [7,8], in which the electroencephalographic (EEG) pattern could suggest the suspected diagnosis even in infants and toddlers, i.e. before the “classical” clinical and behavioral phenotype becomes evident. But epilepsy can represent a significant clinical marker in other conditions as well, such as Rett syndrome [9], Wolf-Hirschhorn syndrome [10], and other recently defined deletion or duplication syndromes.

2) Syndromes in which epilepsy is reported occasionally. In these conditions the diagnostic value of the electroclinical phenotype is limited, but the correct identification and treatment of the symptom still has an important effect on individual and familial health and quality of life.

Finally in genetic syndromes some special considerations should be made regarding the role of interictal EEG abnormalities and discussing clinical indications of antiepileptic treatment. References:


There is growing awareness that in children also “essential” hypertension is the most prevalent form of blood pressure (BP) elevation, and that it may be associated with target organ damage. Evidence is also available that suggests that a child with hypertension has a higher probability of being hypertensive at an adult age than a child with normal BP levels. This suggests that pathophysiological mechanisms contributing to the development of hypertension in childhood might contribute to consolidate such a condition at an adult age. As in adults, in children being overweight or obese have become an epidemic problem associated with elevated BP. This is also the case for lack of physical exercise and excessive intake of sodium with food. It has to be emphasized that in children it is not only weight excess that is associated with increased BP, but the distribution of adipose tissue also plays a role. Among young subjects with similar body weight, those with a greater waist circumference are at a greater risk of having high BP, possibly due to humoral factors released by visceral fat. Indeed, increased insulin resistance (assessed through the HOMA index), increased leptin and uric acid levels are positively and independently associated with increased BP values in children and adolescents, while an inverse correlation has been found with plasma adiponectin levels. Another factor which might contribute to hypertension in childhood is a dysfunction of autonomic neural cardiovascular modulation, characterized by an increase in autonomic cardiovascular sympathetic and a reduction in cardiac vagal modulation, documented by a reduction of cardiac arterial baroreflex sensitivity, remaining significant even after adjustment for increased body weight. These considerations underscore the importance of using techniques for 24h ambulatory BP and heart rate monitoring in the diagnostic and therapeutic management of children and adolescents with hypertension aimed at assessing their hemodynamic reactivity and the related pattern of cardiovascular autonomic nervous modulation, in daily life conditions. Given the young age of the subjects being addressed, it is possible that pathophysiological factors contributing to hypertension might still be modifiable and that no structural cardiovascular changes have yet occurred in this age range, the appearance of which might make hypertension an irreversible condition. The hypertensive child therefore represents a challenge to the medical community for implementing interventions able to achieve true early cardiovascular disease prevention, besides being a potential model for a better understanding of the pathophysiological mechanisms underlying essential hypertension in adults.

A55

Asthma and hypoxia

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More or less severe asthma attacks can damage the CNS and peripheral system in adults and children, both directly causing hypoxia and indirectly with the production of autoimmune antibodies. The pathogenic mechanism is still unknown: cytokines, TNFs, interleukins, free radicals and nitric oxide are often involved as causative factors for the disorders [1,2].

In this review we focus our attention on the relationship between atopy and CNS (central) or PNS (peripheral system) involvement. The main disorders associated with atopic disease are: Atopic myelitis. Hopkins syndrome. Hirayama disease. Spinal progressive muscular atrophy (SPMA).

The occurrence of myelitis in patients with atopic diathesis (atopic myelitis) affecting young adults has been widely reported especially in Japan[2,3]. In general, the disorders affect the posterior column of the spinal cord, as shown clinically and by an MRI, and are characterized by hyperreflexia and the presence of light antigen-specific IgE. Another disease that has been reported in association with atopic disorders is Hirayama disease, a juvenile muscular atrophy of the distal upper extremity, which is also associated with airway allergies such as allergic rhinitis and atopic asthma.
Our group reported on a 13-year-old girl who had recurrent acute episodes of myelitis after asthma attacks; the patient showed to be affected by Hopkins syndrome [4] (HS); a flaccid paralysis affecting one or more limbs after an asthma attack. It is a poliomielitis-like illness, with frequent recurrences in which a possible link between the disease and atopic myelitis was reported, but until now the precise etiopathogenetic mechanism still remains unknown.

Prospective study on the history of allergic disorders in known neurological diseases, an association between spinal progressive muscular atrophy and asthma as well as between myelitis and atopic dermatitis has been demonstrated.

Central nervous system damage associated with atopic diathesis may be classified into two categories: eosinophilic myelitis mainly affecting the cervical spinal cord and those lower motor neuron, such as Hopkins syndrome, Hirayama disease and SPMA [2,4]. Recently we reported children with epileptic crisis related to cow’s milk protein allergy; this disease can manifest itself with skin reactions, failure to thrive, and anaphylaxis as well as gastrointestinal and respiratory disorders [5].

In conclusion, the aim of this report is to analyse current data on the link between asthma and CNS or PNS involvement and also, cow’s milk protein allergy and epileptic events, highlighting the presence and scientific evidence for any potential pathogenetic mechanism.

References

A56
Prevention of Chronic kidney disease (CKD) in children
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CKD is recognized worldwide as an important problem in children. The overt stage of CKD is end-stage renal disease, which is merely the tip of the iceberg of a large number of “covert less severe diseases.” CKD represents a developing process that is initiated by various causes, all with the common end result of persistent and usually progressive damage of varying severity to the kidneys. It is characterized by the common histopathological end point of glomerulosclerosis, tubulointerstitial fibrosis and tubular atrophy, irrespective of the underlying etiology of kidney disease. The aim of the presentation is to identify the clinical and pathological conditions that cause renal damage and suggest measures for its prevention. CKD is defined by the following criteria: a) Kidney damage for >3 months, as defined by structural or functional abnormalities of the kidney with or without decreased glomerular filtration rate (GFR) manifested by one or more of the following features: abnormalities in the composition of the blood or urine, abnormalities in imaging tests, abnormalities on kidney biopsy; b) GFR <60 ml/min/1.73 m² for >3 months with or without the other above mentioned signs of kidney damage. CKD has been classified into five stages for the purpose of prevention, early identification of renal damage and institution and preventive measures for progression of primary damage and appropriate guidelines for instituting management for prevention of complications in severe CKD [1]. Pediatricians have the opportunity to screen at-risk patients, identify affected patients, prevent renal damage and ameliorate the impact of CKD by initiating early therapy and monitoring disease progression. The prevention of CKD constitutes three important aspects. Primary prevention aims to eliminate or reduce exposure to factors that cause renal disease. Secondary prevention in which the prevention of the progression of renal damage from stage 1 to stage 5 is carried out by introducing appropriate measures at various stages of CKD. Tertiary prevention strategies are focussed on the reduction or delay of long-term complications, impairments or disabilities in established disease, requiring renal replacement therapy (RRT). Meanwhile, in adults, the primary preventative measures are limited to few conditions, such as diabetes mellitus, atherosclerosis and hypertension, in children there is a wide range of conditions that can start even before birth; if not recognized earlier, these conditions can cause renal damage and progress to CKD in childhood, adolescence or later in the adult life.

Reference

A57
Adolescents and primary care
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From 1978 National Health System (NHS) primary care pediatricians (PdF) in Italy take care of infants and adolescents from birth until 14 (in selected cases 16) years of age [1]. They are not allowed to take care of their patients up to 18 years, so it’s not possible to follow the whole adolescence under the NHS. There is also increased awareness in families that the pediatrician is the natural medical reference of a teenager; their education and training allows the understanding of changes due to puberty and their experience and background permit thorough understanding not only of the physical but also of the psychological and social changes which belong to this period of transition [2]. However, pediatricians are increasing their knowledge and competence regarding adolescent care and are promoting new preventive programs for adolescent health, such as the “Adolescents Health Project” to strengthen the preventative and educative actions and improve the lifestyle of the young.

Another objective of primary care pediatricians will be to structure shared programs with general practitioners (MMG) to favour the shift of care from the primary care pediatrician to the family doctor. It should be necessary to always support a well planned care transition between Pdf and MMG (exchange of computerized or paper medical records and when possible direct contact). This will enable the MMG to receive the complete information on the health of the patient as well as on social and relational aspects.

References

A58
L’esperienza di uno specializzando
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Located in the central plateau area of Tanzania in East Africa, St Gaspar’s Hospital was founded in 1968 by the Society of the Precious Blood Catholic Missionaries. Elected to referral hospital at a regional level, with an approved capacity of 320 beds, it provides residential consultancy services in general medicine, surgery, obstetrics, gynecology and pediatrics for more than 100,000 people.
In 2013, a cooperation agreement between the "Bambino Gesù" Children's Hospital (OPBG) and St Gaspar's has been established for the creation of a "Clinical and Surgical Pediatric Centre". That led to the complete renovation of the Pediatric Ward, the construction of a new operating theatre and the installation of a telemedecine system for remote medical diagnosis and consultation.

In this setting, a periodic turnover of clinical and surgical missions is guaranteed in order to form the local healthcare providers and improve the quality of assistance. From 2014, the Residency School in Pediatrics of the University of Rome "La Sapienza", in agreement with OPBG, allows its residents to spend a 3-month period of work at the St Gaspar's Hospital's Pediatric Ward. Under the supervision of a local tutor, residents are involved in clinical activities including: morning work round with 90 to 150 patients visited per day; evaluation and management of new admissions for malaria, typhoid fever, meningitis, gastroenteritis, severe malnutrition and local herbs intoxication; weekly mobile clinic service, for antenatal visits, vaccinations and health education programs in rural areas around the hospital. Residents are also engaged in research activities and training courses for local health workers.

One year has passed from when I decided to join the OPBG project but memories and feelings of those days are still vivid in my mind. From the sorrow for the premature deaths that you know easily preventable in developed countries to the joy for the smile of the baby you manage to treat and send back home. In the end, what you receive from this kind of experience is so much more than what you give and that is why I am certain to say, "I want to do it again".

**References**


**A60**

Eosinophilic esophagitis: between reflux and allergy

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Eosinophilic esophagitis (EoE) is a chronic/recurrent inflammatory disease of the esophagus characterized by the association between symptoms of esophageal dysfunction and a prominent eosinophilic infiltrate (>15 HPF) of the esophageal epithelium [1]. This condition mostly affects males and is increasingly being reported in all age groups, with a prevalence currently estimated at around 1:1000 in the Western world. Many experimental and clinical observations suggest that EoE is indeed an allergic condition with a strong genetic predisposition [2]. The natural history is unknown, but several studies suggest the potential progression of untreated disease, such as escalation of symptoms from feeding difficulties in infants to strictures and food impaction in teenagers [2,3]. This evolutionary pattern is somehow similar to that of gastroesophageal reflux disease (GERD), and could be related to a histological remodelig of the esophageal wall characterized by increasing fibrosis [1,2]. Furthermore, cow’s milk protein allergy often causes GERD-like symptoms, and both GERD and cow’s milk allergy cause an increase eosinophil count in the esophageal mucosa of infants. Topical steroids as well as elemental or selective exclusion diet have proven effective in inducing clinical and histologic remission in EoE, but the long-term management of EoE is not yet defined [3,4]. In most pediatric studies, the only mechanism of outgrowing EoE appears to be the long-term avoidance of sensitizing food(s) rather than spontaneous remission [3]. An alternative explanation is that there are different phenotypes of EoE, including a proton pump inhibitor (PPI)-sensitive variant for which long-term PPI therapy may be indicated [5].

**References**


**A61**

Current findings in pediatric non organic feeding disorders (nofeds): the gastroenterologist point of view

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Several epidemiological studies have evidenced that children with underlying chronic conditions are at increased risk of pneumococcal infections with higher case-fatality rates than healthy subjects. Because pharyngeal pneumococcal carriage is a prerequisite for pneumococcal infection development, the high colonization rates found in these subjects is the most important reason for this phenomenon [1]. To overcome this problem, health authorities worldwide have recommended that children with severe chronic disease receive pneumococcal vaccine prophylaxis with the 13-valent conjugate preparation (PCV13) not only in the first years of life but also during school age and adolescence. Conjugate pneumococcal vaccines significantly reduce pneumococcal carriage of serotypes included in the vaccines and consequently limit development of pneumococcal infections. A dose of PCV13 has been added to the 23-valent polysaccharide vaccine in order to obtain a stronger protection at least against the 13 pneumococcal serotypes included in this vaccine. The choice of PCV13, already found extremely effective in reducing pneumococcal infections and pharyngeal carriage rates in both vaccinated healthy children and unvaccinated healthy individuals, was based on a large number of studies that have clearly evidenced that this vaccine can evoke a protective immune response in most of the subjects with underlying disease with a satisfactory profile of tolerability and safety [2].

Unfortunately, despite recommendations pneumococcal vaccination coverage remains in most of the children at increased risk of pneumococcal complications significantly lower than desired. Limited knowledge about the real importance of pneumococcal infections in children at increased risk by patients, parents and physicians themselves together with a not justified fear of poor safety and tolerability of the vaccine are the most important reasons for the low compliance with official recommendations [3]. Specific educational programs have to be planned if this problem has to be solved.

However, the use of a single dose of PCV13 could not definitively solve the problem of the protection of children at risk from pneumococcal infection. The duration of protection offered by this vaccine is not established and it is not defined whether one or more booster does have to be given. Moreover, with time it is possible that a new replacement phenomenon could take place and this could require the use of a vaccine able to cover a greater number of serotypes.

**References**


The process of taking in and swallowing food or drink involves three linked anatomical and physiological regions connected to each other both anatomically and neurologically. The non-nutritive suck is first seen in the fetus between 18 and 24 weeks. The second type of suck is the nutritive or “active feeding suck”, which is more mature and complex and is designed to deal with fluid. It is estimated that non-organic feeding disorders (NOFEDs) occur in 25% of healthy infants and 80% of young children with developmental disability [1]. Furthermore, infants born preterm and/or with a birth weight below the 10th percentile for gestational age are at high risk for developing NOFEDs. This implies that feeding disorders could be related to intrauterine growth retardation [2]. About 20-60% of parents report that their children are not eating optimally, that is, that they are picky, have food phobia, eat too little and have weight loss [3]. Several studies have suggested that only 16-30% of feeding disorders are organic and that up to 80% of cases referred to specialist pediatric services have a significant behavioral component [4]. NOFED is a formal diagnostic term used to indicate a condition in children younger than 6 years of age. It usually presents as food refusal, aversion to feeding, selective eating, low food intake or failure to thrive [5]. NOFEDs often result from multifactorial etiologies and the classification of disorders based on an organic versus non-organic dichotomy fails to provide a system that can represent the often complex interactions between medical problems, the family system and behavioral difficulties associated with feeding disorders. Some classifications are simplistic and do not help differentiate between NOFEDs and organic disorders, whereas others do not have a single set of criteria for diagnosis. The most serious complication of NOFED is failure to thrive (FTT) which requires a medical or nutritional approach (Figure 1). There is often confusion, misdiagnosis and under-diagnosis with regard to NOFED and the patient is often initially diagnosed with another medical condition [6]. Differential diagnosis can include gastroesophageal reflux disease (GERD), food allergy or swallowing disorders [7]. Medical assessment should include family, social, feeding, past and current medical histories and a complete physical examination. It is necessary to emphasize the difficulties in recognizing NOFED by primary care physicians. FTT is present in 40-50% of NOFED patients correlated with a delayed diagnosis.

References

A62
Global health training in paediatric residency programs: the Italian experience
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Osservatorio Nazionale Specializzandi Pediaitia (ONSP) is an Italian association of residents in paediatrics, and one of its interests is to support paediatric training in developing countries. In 2014, for this purpose, ONSP performed a survey with the aim of describing interest, participation, resources, and obstacles of residents who are involved in global health training within paediatric residency programs. Once the final data were known, an informative brochure was produced for publicizing the results of this survey and the projects that Italian Pediatric Schools have activated in developing countries. 35 of 38 paediatric residency schools (92%) participated in the survey. 67% of them offer an elective training program in global health and 42% have a formal program that is part of the curriculum of trainees. 47% of the paediatric residency schools have a collaborative program with developing country or non-governmental organization (NGO) and 17% had a program in the past years but not still ongoing. In most cases, 3rd, 4th and 5th year paediatric trainees were involved in “global health” training. The duration of training was less than 1 month in 13% of cases, 1-3 months in 39%, 3-6 months in 43%, and more than six months in 4%.

Figure 1 (abstract A61) Algorithm of the NOFED
Only 22% of residents who were involved in training in developing countries were evaluated before departure, and only 35% received training in global health before leaving; 39% participated in post elective debriefing meetings. 74% had the supervision of a local tutor (66% pediatricians, 34% other doctors) and 61% also had an Italian tutor for the whole project. Almost all of the collaborative programs were in Central or Southern Africa; only two of them were in Central America (Nicaragua). The training in global health provided child care (in 62% of cases), neonatal care (45%), malnutrition support (58%), HIV prevention and AIDS care (20%) and local staff training (42%). All residents were satisfied after their training in developing countries and only a few of them suffered for some minor reasons during that period.

In conclusion, more than 50% of Italian paediatric residency programs actually offer an elective program of global health program and most of the trainees consider it a great opportunity for professional growth.

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Conflict of interest and professional ethics
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A65

In general terms, we can face a conflict of interest every time that we have to deal with competing goals and responsibilities. In a more specific sense, however, such a conflict is understood as a situation where a personal self-interest could unduly interfere with professional responsibilities, particularly when a situation appears particularly blameworthy, when public interest and protection of fundamental individual rights are involved. According to the Italian Code of medical deontology, conflict of interest is itself a condition that a physician may well happen to experience and not ipso facto a violation of legal or moral rules. As a condition, it entails the necessity to make decisions which should always comply with an unquestionable priority: the best interest of the patient must always come first and professional behaviour can never be conditioned by improper benefits of economic or other nature. Full disclosure is required, whenever conflicts of interest may impinge upon the diagnostic or therapeutic prescriptions or the possible relationships with industries and institutions [1].

It is also worth observing that a physician is very often an employee. The employer’s and the patient’s interest may not coincide and this can generate a conflict of loyalties. The distinction between external and internal goods, proposed by the philosopher Alasdair Maclntyre, helps clarify this ethical challenge. By external goods, we understand those goods, such as money, power and fame, that can be achieved as a result of the practice, but are not a component of its standard of excellence and tend to be enjoyed exactly as a private property. Internal goods, on the contrary, correspond to the very fundamental goals and essence of the practice and tend to be a good for all those, who are involved in it [2].

In the present “market society”, physicians are frequently faced with possible conflicts of interest in their prescriptions, but paediatricians are particularly vulnerable when they must decide how their patients should be fed. In the case of breast-feeding, the standard on which the internal good of excellence relies has been already set by the WHO’s International Code of 1981: “Health workers should encourage and protect breast-feeding” and “no financial or material inducements” should be offered “to health workers or members of their families” nor accepted by them to promote the use of other products beyond their real need [3].

References

A66

Obstacles in access to care for children
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Background: Doctors of the World – Médecins du Monde (MdM) provides free frontline medical and social services for people facing barriers to the mainstream healthcare system. Child poverty rate has been soaring since 2008, especially in Spain and Greece. Restrictive laws across Europe often hinder any access to preventive and curative care, including for children whose parents are undocumented migrant (UDM) or lack health coverage. For example, in Germany, civil servants have to denounce children of UDM. In Spain, the legal restrictions on UDM negatively impacts their children rights. In the UK, if one of their parents is not registered at a GP, the children will not be accepted. In Greece, in 2014, around a third of the population has no more health coverage.

Materials and methods: Data collected by MdM teams with 23,040 people in 25 cities in 10 European countries with the use of common social and medical questionnaires administered during face to face social and medical consultations [1].

Results: Most of the 623 children for whom we have detailed data live in harsh conditions (slums, overcrowded and health-threatening housing, or even out in the streets). Less than half were immunised against tetanus (42.5%), pertussis (39.8%), hepatitis B (38.7%) and measles, mumps and rubella (34.5%). Also, 38.8% of the parents didn’t know where to go to get vaccination for their children. Children of UDM also fear their parents’ arrest, have to move from place to place hindering continuity in care and a stable social life. Migrants’ children are put at risk because of the barriers to antenatal care faced by their mothers. Even when children have theoretically full access to care, administrative barriers too often stop their access.

Conclusions: European States must offer universal public health systems built on solidarity, equality and equity (and not on profit rationale), open to everyone living in Europe. They should ensure that all children residing in Europe have full access to immunisation programs and paediatric care. All pregnant women must have access to abortion, antenatal and postnatal care, and safe delivery. Launched in April 2014, the Granada Declaration [2] calls for a better protection of migrants’ health and healthcare, including that of UDM. In accordance with the World Medical Association’s Declaration on the Rights of the Patient, we urge all health professionals to provide appropriate medical care to all without discrimination, and to refuse restrictive legal measures that alter medical ethics.

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A67

Mycoplasma pneumoniae infection with neurologic complications
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Mycoplasma pneumoniae is one of the important causes of upper and/or lower respiratory tract infections during childhood. Central nervous system (CNS) related findings and complications are most commonly seen and have been described in patients with M. pneumoniae infections [1,2]. Patients suffering M. pneumoniae infection may have varying degrees of neurological complications at a ratio of approximately 6-7% [1,2]. Neurological manifestations include encephalitis, transverse myelitis, acute disseminated encephalomyelitis (ADEM), Guillain-Barre syndrome, and thromboembolic stroke [2]. The time period between the onset of respiratory symptoms and neurological symptoms varies 2 to 14 days [3,4]. More than 80% of patients with CNS findings have concomitant respiratory infection [3].

Central nervous system complications have been reported with Mycoplasma infection. Cerebellar syndrome, polyradiculitis, cranial nerve palsies, aseptic meningitis, meningoencephalitis, acute disseminated encephalomyelitis, coma, optic neuritis, diplopia, mental confusion, and, acute psychosis secondary to encephalitis, cranial nerve palsy, brachial plexus neuropathy, ataxia, choreoathetosis, and ascending paralysis (Guillain-Barre syndrome) are neurologic complications seen with M. pneumoniae infection [1]. Neuroimaging may reveal normal findings or focal diffuse edema in cases of encephalitis or meningoencephalitis. Patchy asymmetric or diffuse signal change of gray and white matter may be seen in patients with ADEM with multifocal, asymmetric foci of high signal intensity on FLAIR and T2 weighted images. A focal infarction may be seen with M. pneumoniae related stroke [4].

Treatment of neurological complications of M. pneumoniae is controversial. Treatment may be adjusted according to infection mechanism such as antibiotics, corticosteroids, intravenous immunoglobulin [3-5]. Antimicrobial treatment, especially macrolides, may be sufficient for CNS involvement associated with M. pneumoniae, beside the beneficial effect of treatment with steroids this treatment must be considered with direct invasion of CNS by the organism when other causative agents have been excluded. Plasma exchange has also been reported and seemed to be beneficial.

References
Redox alterations of platelets and erythrocytes represent progression marker and pathogenetic determinants in Kawasaki disease

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Background: Kawasaki disease (KD) is a rare generalized systemic vasculitis of unknown etiology in which the main complication is the development of coronary artery abnormalities. Considering that an inflammation-associated systemic pro-oxidant status could play a critical pathogenetic role in KD progression [1], we evaluated some peripheral blood redox-associated parameters, including redox and aging features associated with red blood cells (RBCs) and platelets (PLT) integrity as possible pathogenetic determinants or progression markers in KD disease.

Materials and methods: The 18 KD patients, aged between 6 and 24 months, were recruited from the Bambino Gesù Hospital (BGH) of Rome (Italy) and studied, after obtaining the parent informed consent, before to start therapy with intravenous immunoglobulin and aspirin. Ten age-matched healthy donors (HD) were enrolled as controls. The study was approved by the BGH Institutional Review Board.

Morphological, biophysical, biochemical and flow cytometrical methods were used to evaluate: i) reactive oxidizing species (ROS) formation and oxidative stress-related biomarkers [3-nitrotyrosine, the endothelial nitric oxide synthase inhibitor, asymmetric dimethylarginine (ADMA), the pro-oxidative stress-related biomarkers 3-nitrotyrosine, the endothelial nitric oxide synthase inhibitor, asymmetric dimethylarginine (ADMA), the pro-oxidative stress-related biomarkers 3-nitrotyrosine, the endothelial nitric oxide synthase inhibitor, asymmetric dimethylarginine (ADMA), the pro-oxidative stress-related biomarkers 3-nitrotyrosine, the endothelial nitric oxide synthase inhibitor, asymmetric dimethylarginine (ADMA), the pro-oxidative stress-related biomarkers]; ii) oxidative stress in gastric and lung microflora with acid suppression; iii) coagulation status (annexin V positivity); and iv) changes in mitochondrial function of both platelets and RBCs, including mitochondrial-membrane potential and aging.

Results: With respect to HD, peripheral blood of KD patients showed increased levels of O2·-, ·NO, 3-nitrotyrosine and MPO, and decreased ADMA concentration (Figure 1). In RBCs, alterations of biomarkers correlated with cell aging and death (i.e., decreased glycoprotein A and CD47 expression, annexin V positivity).

Conclusions: These results lead us to hypothesize that the oxidative/nitrative stress occurring in KD inflamed blood vessels could alter both RBCs and PLT homeostasis, resulting in a sort of premature aging in these circulating cells that could lead to anemia and the formation of blood clots. These alterations could play a pathogenetic role in the cardiovascular complications often associated with KD but, in addition, the possible use of these data as real time biomarkers of progression cannot be ruled out.

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References
A71 Secondary prophylaxis in rheumatic fever: is it time to change? Andrea Taddio1,2*, A Pirrone2, Serena Pastore1, Loredana Lepore2, Caterina Di Battista3, Gabriele Simonini3, Lucia Breda3, Rolando Cimaz4 1Institute for Maternal and Child Health - ICCS “Burlo Garofolo” – Trieste, Italy; 2University of Trieste, Trieste, Italy; 3Department of Paediatrics, University of Chieti, Chieti, Italy; 4MOU Florence, Florence, Italy E-mail: andrea.taddio@burlo.trieste.it

Background: Carditis and rheumatic chronic heart disease are the most serious complications of Acute Rheumatic Fever. Nowadays prevention of recurrent episodes of group A β-hemolytic streptococcal pharyngitis is the most effective method to prevent the development of severe rheumatic heart disease. However the evidence of these guidelines are weak and result from studies conducted more than 50 years ago [1]. To detect rate of Carditis related to Acute Rheumatic Fever found at follow up and to find a relationship with clinical data at diagnosis and compliance to prophylaxis.

Material and methods: This is a multicentre retrospective study conducted among 117 pediatric patients admitted with diagnosis of rheumatic carditis. We analysed the presence of carditis at diagnosis and at follow up comparing it with the number of infection recurrences and with the level of compliance of the patients to antibiotic prophylaxis. Compliance to antibiotic prophylaxis was evaluated individually with a questionnaire. The association between recurrences and compliance to therapy and between recurrences of streptococcal infections and carditis at follow up was also analyzed. Data were analysed using Fisher exact test.

Results: We examined 117 pediatric patients with rheumatic fever carditis. The median age of the patients at diagnosis was 9 years (6-11 years). The median age at follow up was 15 years, and the data at follow up were taken at a median time of 6.8 years from diagnosis. The data show that carditis at follow up was associated with the presence of carditis at diagnosis (p<0.000) and not with the level of compliance to antibiotic prophylaxis (p=NS). Also there was no statistically significant association between recurrences of infections and good level of compliance to therapy (p=NS) and between number of recurrences and presence of carditis at follow up (p=NS).

Conclusions: We observed that the risk to develop carditis at follow up in pediatric patients with Acute Rheumatic Fever is independent from the compliance to antibiotic prophylaxis and the number of infection episodes caused by group A β-hemolytic Streptococcus while it seems related to the presence of carditis at time of diagnosis.

Reference

A72 Future directions in prevention and treatment of children obesity and eating disorders Rita Tanas1,2, Guido Caggese3, Simonetta Marucci4 1UO Pediatria, Azienda Ospedaliero Universitaria, Ferrara, Italy; 2Formazione Professionale, Azienda Ospedaliero Universitaria, Ferrara, Italy; 3Centro per il trattamento dei Disturbi del Comportamento Alimentare, Todi, Perugia, Italy E-mail: tanas.rita@tin.it


The childhood obesity epidemic has promoted many healthcare programs focused on nutritional education in order to teach correct nutrition and to promote early diagnosis and care of overweight/obese children including offering them a correct food supply. For almost 20 years, however, scientific literature has pointed out that eating disorders (ED), which typically onset during adolescence, are increasing, and that dieting (restrictive behaviors, adopted today by many adults and adolescents, mostly managed without the support of a professional format, to match the ideal of thinness proposed by the media and being or feeling overweight is likely to be the most avoidable risk factors [1-4]. Body stigma, shared by those with obesity and ED [5] and based on the attribution of a strong personal responsibility in the onset of these diseases, has been shown to increase stress, blood pressure, cortisol, oxidative stress, C-reactive protein and worse glycemic control, while decreasing motivation for physical activity (PA) encouraging a sedentary lifestyle. Stigma also increases psychosocial issues, such as depression, body-image distortion, loss of oral control and affects motivation and effectiveness of treatments. Primary care staff are increasingly asked to promote parents awareness on their children being overweight although this has been recognized as a trigger for the onset of an ED [6], but at the same time they are not provided with adequate means to run a truly effective therapeutic program (strategies to promote change, such as the patient-centered motivational interview) and the suffering related to family and professional weight-derision.

Children are then pushed to see themselves as fat, to dislike themselves and go on a diet, focusing dangerously on food, body and weight and with the result of being bullied in many life contexts such as home, school, medical services. All this leads to dysfunctional eating patterns, loss of control and increased BMI. It has been reported that obesity is the strongest predictive factor to the onset of an ED [7]. The pervasive and universal body weight-stigma enhances restrictive behavior and creates a vicious circle that leads to a real ED epidemic among young adults [8]. It’s time to think about obesity prevention and treatment in a different way (Tables 1 and 2), getting to know EDs [8,9] and using tools that do not favor them, protecting children against weight-stigma, especially ones coming from parents, educators and health workers [10].

References
At present, factors that have been recognized as being able to influence growth are nutritional, physical, chemical, psychological and genetic. The causes of short stature are numerous, with about 90% of cases classified as Idiopathic Short Stature and divided into two types: familial short stature and constitutional short stature. Part of the population with growth disorders are SGA newborns (10%) who don’t show recovery growth that physiologically should occur within 2-3 years in 90% of cases.
Salbutamol and around

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Short-acting beta agonists (SABAs) such as salbutamol are well known as the first-line therapy for the treatment of acute exacerbations, exercise-induced asthma and childhood intermittent asthma. As needed, SABAs with no controller should be considered only if symptoms are rare and there is no awakening due to asthma. Salbutamol, through its effects of relaxing airway smooth muscle and increasing airflow, provides rapid relief of acute asthma symptoms. Treatment’s effects begin in about 15 minutes and peak effect is usually within 30 minutes. Patient experience prompt benefit with reduction in coughing, wheezing, chest tightness and shortness of breath. The treatment dose includes 4-10 puffs by pMDI+spacer every 20 minutes for 1 hour [1]. The rapid relief provided by salbutamol may invite misuse and overuse, particularly in adolescents. SABAs should not be prescribed on a regular schedule as it may lead to a beta receptor downregulation. Moreover, frequent use of salbutamol is an indication of poor asthma control and an increased use of controller therapy should be considered. An heterogenous response to beta agonist therapy has been described and individual response is related to many factors, including: the relative contribution of bronchoconstriction versus airway inflammation and edema in producing the airway obstruction, different triggering mechanisms, the duration of symptoms, the patient’s age and the route of medication delivery. Salbutamol is normally delivered through the inhaled route while intravenous salbutamol is used when children are unresponsive to inhaled treatment [A]. Less common are the oral and subcutaneous routes. Inhaled SABAs, administered through a MDI with attached spacer device along with infant- or child-sized mask or by nebulization, have the advantages of smaller doses and fewer side effects [3]. Inhaled SABAs allow selectivity for beta-2 receptors on bronchial smooth muscle to achieve bronchodilation without significant tachycardia associated with activation of beta-1 receptors on cardiac muscle. The incidence of adverse effects is dependent upon age of patient, dose, and route of administration. Examples of possible mild or moderate side effects in children and adolescents, not very common at recommended doses, include: palpitations, tachycardia, excitement, hyperactivity, insomnia, nervousness, shakiness and unpleasant taste (inhalation site) [4].

References


Is organic diet really necessary for children?

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Pesticides are a wide class of phytochemicals commonly used in conventional agriculture. Most of them are characterized by being very toxic to humans and persistent in the environment, and represent long-term dangers as they biomagnify up the food-chain. Humans, and particularly breastfed babies, are at the top of the food-chain since they ingest more food and water per unit body weight than adults, so any exposure is greater in proportion to their size. For such a reason, dietary intake of pesticides represents the major source of exposure for infants and children. Since they are growing so quickly, infants and young children are more susceptible to the effects of pesticide exposure than adults [1]. Their internal organs are still developing and maturing and the enzymatic, metabolic, and immune systems are less efficient than those of an adult. Furthermore, it has been shown that there are “critical periods” in human development when exposure to a toxin can permanently alter the way in which an individual’s biological system operates. Children’s exposure to pesticides has been linked to a wide range of disease, including asthma, ADHD, autism and cancer [2]. A maximum residue level (MRL) is the highest level of a pesticide residue that is legally tolerated in/on food or feed when pesticides are applied correctly, so that the amounts of residues found in food must be safe for consumers and must be as low as possible possible. However, the toxicity was set in the adult population, not on children, and it does not take into account the multiresidue in food. Therefore is a priority for children health to reduce exposure to contaminated food, similarly to baby food, where no pesticide residues are admitted according to the European Directive. Interventional studies have shown that an organic diet reduces children’s exposure to pesticides, and when kids switched from a conventional to an organic diet, urinary pesticide metabolites dropped to almost undetectable levels [3]. Organic food also contributes to increasing nutritional quality: a recent meta-analysis reported that organic food is richer in vitamins and antioxidants compared to conventional ones [4]. The environmental benefits of organic agriculture to air, soil and water, consist of lowering the total toxic burden to our ecosystems. As demand for organic foods continues to grow, more farmers are likely to view organic methods as a viable and marketable option, helping to stabilize supply and price.

References


Is organic diet really necessary for children?

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References


Fructose consumption has been increasing over recent decades and is believed to play a role in the rising epidemic of metabolic disorders and hypertension (HT) in children [1,2]. This theory is supported by epidemiological and experimental studies in animals and humans. High-fructose diets upregulate sodium and chloride transporters, resulting in salt overload that increases blood pressure (BP) [3]. Moreover, excess fructose has also been found to deregulate vasconstrictors and vasodilators, and over-stimulate the sympathetic nervous system. Metabolism of fructose is mediated by fructokinase, which uses ATP as a phosphate donor. Unlike glucose, there is no feedback mechanism regulating fructokinase. As a result,
AMP is continuously involved in the production of uric acid (UA) [4]. In adolescents in the US, serum UA was shown to increase from the lowest to the highest category of fructose-sweetened beverage intake and this increment was paralleled by an increase in BP, even independently of body mass index [5]. These data suggest pathways other than obesity relating soft drinks to the development of HT. Epidemiological studies demonstrate an association between serum UA and both prevalence and new onset of essential HT in adolescents [6]. Recently, UA showed a strong independent relationship with BP values across different BP categories, from normal BP up to pre-and finally to established HT in children at relatively high cardiovascular risk [7]. Animal models support a two-phase mechanism for the development of hypertension (HT). Initially, UA induces vasoconstriction by activation of the renin-angiotensin system and reduction of nitric oxide. Over time, UA uptake into vascular smooth muscle cells (VSMC) causes cellular proliferation and arteriosclerosis that impair pressure natriuresis, causing sodium-sensitive HT [8]. Increased UA causes endothelial dysfunction by inhibiting oxidative stress once inside cells. UA internalization is mediated by URAT-1, and stimulates production of growth factors and chemokines inflicting oxidative stress once inside cells. UA internalization is mediated by URAT-1, and stimulates production of growth factors and chemokines. Recently, UA showed a strong independent relationship with BP values and both prevalence and new onset of essential HT in adolescents [6].

Interestingly, in both animal and human studies, allopurinol attenuates the development of fructose-induced HT by lowering UA. Indeed, lowering UA with either allopurinol or probenecid reduces BP in adolescents with HT or pre-HT [13,14]. While larger studies are needed, a fructose assumption and serum UA are emerging as potentially modifiable risk factors for the prevention and treatment of HT in children.

References

Results after one year of rotavirus universal mass vaccination in Sicily

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Background: Rotavirus (RV) vaccination is the best strategy to prevent hospitalizations due to rotavirus gastroenteritis (RVGE) and is strongly recommended by international health authority [1]. The Sicilian Health Department introduced rotavirus universal mass vaccination (RUMV) into regional immunizations schedule in 2013 (mean vaccination coverage = 31%). Intussusception is the invagination of one segment of the intestine within a more distal segment and even though the etiology is still unknown, in 1998, a relationship with a tetravalent rotavirus vaccine that was promptly withdrawn was suggested [2]. Post licensure surveillance studies have not confirmed previous findings and no increased risk of intussusception was found between vaccinated infants with both of the actually licensed rotavirus vaccines [3,4].

Aim of this study is to analyze the trend of RVGE hospitalizations and contextually to monitor the trend of intussusception in Sicily from 2009 to 2013 after one year of RUMV.

Material and methods: Were collected data from hospital discharge records occurred from 1st January 2009 to 31st December 2013 in Sicily. Cases of RVGE were defined as all hospitalizations with an ICD-9-CM diagnosis code of 008.61 on any position [5]. Furthermore, cases of intussusception were defined as all hospitalizations with an ICD-9-CM code of 560.0 on any discharge diagnoses.

Results: In 2013 the RVGE hospitalizations were 41% less in children aged 0-59 months and 43% less in children aged 0-23 months with respect to the mean number of cases observed from 2009 to 2012 in Sicily (figure 1).

Analyzing RVGE hospitalization rates per 100,000, was reported a significant reduction in both age classes in 2013 respect to mean incidence observed from 2009 to 2012 (0-59 months: from 395 to 242 cases/100,000; 0-23 months from 609 to 364 cases/100,000) (figure 2).

Finally, a significant increase in intussusception hospitalizations was not reported with respect to mean number of hospitalized children observed from 2009 to 2012 in age class 0 to 59 months. In particular, among children aged 0-23 months (directly exposed to vaccination in 2013) a slight reduction was observed with respect to mean number of cases reported in 2009-2012 (figure 3).

Conclusions: After one year of surveillance and despite reaching low vaccination coverage, our results demonstrated the high effectiveness of the
RUMV strategy on reduction of RV circulation. Similar data on RV vaccination efficacy on early vaccination campaign was reported in Belgium [6]. Moreover, the steadiness of intussusception hospitalizations after introduction of RV vaccination allows us to confirm the security profile of the available vaccine.

References

A78
From healing to taking care
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Disease is an absolute evil, the sick human being is an absolute good.
As doctors we are used to fighting disease to save the patient. But when disease and patient coincide, the picture changes. We must cure symptoms, prevent complications, reduce effects, but at same time we must help the family to accept their child. It is necessary to modify the approach moving from healing to caring.
Caring for the child is conditioned by recognition of his needs, that is identifying the concrete situation of the child and his family, in relation to his condition and its impact on their daily life.
The disabled child needs preventive and curative assistance, meaning a pediatric approach. In spite of big number of rare conditions, it is possible to sum up the pathologies which most frequently involve children with congenital but also acquired disabilities. The complexity of the assistance is due especially to the involvement of numerous strictly connected systems, therefore multidisciplinary and well coordinated approach is necessary.
The coexistence of medical problems together with psychological and social aspects obliges the pediatrician to have a multisectorial view. The family pediatrician should use health balance to reveal the clinical problems frequent in a specific disability in order to avoid the most common complications of that condition, and to verify if the care project developed by social and medical operators is sustainable for the child, the family and the Country. The role of the hospital pediatrician consists in coordinating the interventions of different specialists, managing the emergencies and developing the treatment strategies calibrated for every single child and family [1]. So the correct caring of the child with malformative syndrome and more general with disability, includes an effective and functional integration among the internal clinical aspects, rehabilitative and educative aspects and psychosocial elements [2].
The advantage of coordinated and integrated assistance is clear also in economically terms, family satisfaction, and the team’s cultural improvement.
The cost of an integrated assistance is compensated by a notable saving that the healthy system has for the cost reduction due to prevention of complications and to unnecessary exams [3].
Care means, first of all, giving high value to human relationships and taking into account the globality of the person, his needs and his problems. Always ensuring hope. The goal is to find equilibrium and sostenibility for the child and his family [4].

References