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Non compaction left ventricle and dilated cardiomyopathy with lethal familial cardiac failure

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Background: Isolated non compaction left ventricle (NCLV) is a rare genetic disorder that involved in enhanced large ventricular trabeculation due to cardiac heart failure, arrhythmia and mural thrombosis. Echocardiography and cardiac MRI is the best diagnostic imaging for it.

Case Presentation: We reported 2 cases (single originated family): a 9 year-old girl and a 16 year old boy reported (family members) that after exercise and excitement were involved sudden cardiac Death. Both patients had a history of seizures and syncope (in the female case) that treated by anticonvulsant drugs. Following the occurrence of two patients’ sudden cardiac death in their family, other family members examined by echocardiography and then to be diagnosed as isolated NCLV in their family.

Findings: Seizure and syncope could be as the initial manifestations of NCLV, which is the embryonic origin of the myocardium. Syncope and seizure could be warning signs (risk factor) for these patients who are at risk of sudden death if they have exposed. Echocardiography which was done in patients with seizure attack, or syncope in the absence of underlying neurological disease, and especially in familial case of NCLV may be beneficial. Arrhythmias, SCD and thromboembolic events compared with cardiac heart failure were happen rarely, but the prognosis of medical treatment by anti-convulsion drugs extremely is poor in these patients after diagnosis was made.

Keywords: Cardiomyopathy, Left Ventricle Non Compaction, Failure, Death

Comparison of hospitalization in patients with severe coarctation that undergone surgery by lat, thoracotomy and mid-sternotomy

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Background: Today progression in cardiac surgery and anesthetic technique causes a good chance for small children who are undergone surgery due to coarctation of aorta (COA). On the other hand with increasing diagnosed cases of COA and concomitant intera cardiac lesion, then surgical options must be noted. Based of this fact, we tried to decrease complication and cost by shortening of duration of hospitalization. In this study we compared hospitalization time for different manners including L.thoracotomy and Midsternotomy in coarctation or coexisted with Ventricular Septal Defect (VSD) or another intracardiac anomaly. When coarctation coexists with a VSD in a neonate or infant with heart failure or VSD of membranous, outlet, or malalignment type one-stage repair of the coarctation (by end-to-end anastomosis) and the VSD through a mid-sternotomy is the procedure of choice for such situations. Two stages coarctation repair alone may be performed, with later VSD closure if it remains large or the infant has failure to thrive. Coarctation repair with concomitant banding of the pulmonary trunk can be performed, with later removal of the band and VSD closure.

Methods: Our study was a retrospective - descriptive study. Duration of study was from 2009 to 2012 at Children's Medical Center. Number of patients were 22 (L. Thoracotomy=8 cases, sternotomy=14 cases). Results were analyzed by using statistical software spss, 15.5 (assurance=95%). T-test descriptive (freq), Pearson (correlation age & Wt by admission time).

Findings: The minimum and maximum age of patients was 6-574 days. The minimum and maximum weights were 2.5-8.5 kg. Average stay in groups L. Thoracotomy and sternotomy were 33.62 and 32.42 days, respectively. According to the Pearson correlation (P<0.041) the relation between patient age or weight and duration of hospitalization was inversed and whatever the age and weight of patients increased, discharge time was decreased.

Conclusion: We advise prospective study and more cases for getting better results and comparison complications of coarctation surgery between Lat sternotomy and Thoracotomy.

Keywords: Hospitalization, Coarctation of Aorta Repair, Lat Thoracotomy, Mid Sternotomy

The bronchopulmonary dysplasia in infants with retinopathy of prematurity

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Background: For the last decade the bronchopulmonary dysplasia (BPD) became a growing problem in pediatrics and has taken a second place on frequency among chronic respiratory diseases after childhood asthma. In infants of early age the BPD is determined at 15-38% of cases, with the birth weight less than 1500g who need mechanical ventilation because of respiratory distress syndrome of newborn. With the development of nursing and respiratory therapy technologies in preterm infants the increase of BPD frequency has occurred with the reducing of mortality among children with birth weight less than 1000g and gestational age less than 30 weeks. The toxic effect of high concentrations of inspired oxygen and hyperoxia causes the damage not only for the lung tissue, but also non-infectious nonimmune inflammation with subsequent proliferation of endothelial and retinal neovascularization- retinopathy of prematurity (ROP). The objectives of the study were determining the frequency of bronchopulmonary dysplasia in infants with retinopathy of prematurity.

Methods: 263 preterm infants with retinopathy of prematurity were studied by chest radiography, and binocular indirect ophthalmoscopy.

Findings: Out of 263 children with ROP, 71.5% (188) were preterm infants with BPD, gestational age 27- 32
Acquired pericardial cyst with degenerative changes rare entity in Iranian Child: a case report

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Background: Pericardial cysts constitute two forms congenital and acquired. Second form is rare with unusual clinical presentations. Also keep minding, the second form had rare complications, such as spontaneous rupture and hemoptysis. Our article is based on the record of a boy with acquired pericardial cyst with degenerative changes and review of literatures.

Case presentation: A 9 year old boy presented with intermittent, non-exertional, retrosternal chest pain since 13 months ago. The pain radiated to the left arm. He denied constitutional symptoms, hemoptysis, chronic cough and sputum production. He also underwent surgery for lung hydatid cyst 1.5 years previously. On physical examination, the cardiopulmonary system was unremarkable. Chest X-ray revealed a rounded calcified cystic mass along the left cardiac border. A tricuspidal cyst was seen at thoracotomy. Histologic examination of excised lesion shown cystic wall including dense fibrous connectivetissues with calcification, focal ossification, aggregation of amorphous material containing cholesterol crystal and chronic inflammatory cells infiltration. Mesothelial cells lining was not found. He had uneventful postoperative period and was symptoms free when seen six months later in the clinic.

Conclusion: Acquired pericardial cyst has exceptional features in location of the cyst, macroscopic and microscopic pathology, and clinical presentations. Some of patients have past medical history of tuberculosis, hydatid cyst and other infectious diseases. Therefore, more investigations including serology for hydatid disease, search for specific granuloma, acid fast bacilli and … are highly mandatory.

Keywords: Acquired Pericardial Cyst, Children

Guidelines for sinusbradycardia in newborns

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Background: Heart rate monitoring has become a ubiquitous part of fetal and neonatal assessment, and has made detection of bradycardia in the fetal and neonatal periods a frequent occurrence. Evaluation of a fetus or neonate with bradycardia requires an understanding of the mechanisms of bradycardia as well as the cardiac and non-cardiac causes of bradycardia. The mechanisms of
Assessment of neonatal sepsis on myocardial function by tissue Doppler imaging

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Background: Neonatal period is the most critical stage in the life of newborn babies and neonatal sepsis is one of the most common causes for mortality at this age. Cardiovascular complications, myocyte damage and modification of cardiac blood flow induced by inflammatory mediators are among the consequences of neonatal sepsis on newborns. Nevertheless only a limited number of echocardiographic studies have been performed to investigate the pathologic effects of neonatal sepsis on heart. Tissue doppler echocardiography is a useful method for the assessment of regional heart systolic and diastolic functions. In this study, we aimed to determine the myocardial performance during neonatal sepsis by tissue echocardiography in term and preterm newborns.

Methods: This study was a descriptive research and the information was collected through questionnaires and sampling on 61 preterm and term neonates. Tissue echocardiography (TDE) and doppler echocardiography were performed. TDE was done at the level of mitral and tricuspid valve annulus and intraventricular septum level. The results were then statistically analyzed and evaluated.

Findings: In this study, 30 neonates were term and the rest were preterm with the age of 1 to 9 days. We have not observed significant difference between the tissue echocardiographic indices of mitral and tricuspid valves and interventricular septum between healthy and septic newborns. Our results, however, revealed significant difference in A&E waves at the level of tricuspid and mitral valves between sepsis and none sepsis group (p<0.001).

Conclusion: Doppler echocardiography is a useful tool to evaluate the myocardial function during neonatal sepsis. It is also possible to investigate the myocardial modifications during sepsis by Tissue Doppler Imaging (TDI).

Keywords: Sepsis, Neonate, Myocardial Function, Tissue, Doppler Echocardiography

Primary lung tumors in children

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Background: Primary lung tumors are rare in children; usually, single case reports appear in literature. The collective review cases are also limited, and there is much diversity in reported cases.

Methods: To review the literature on reports of primary lung tumors in children and adolescents, and to present personal experiences of cases with pediatric primary tumors.

Findings: Among the reported pediatric primary lung tumors, some are similar to adult lung pathology, while others are quite different and unique to the pediatric lung. The tumors are mostly endobronchial and the commonly reported cases are carcinoid and mucoepidermoid carcinoma. The pseudoneoplastic tumors, which are considered the most common benign primary lung tumors in children, are controversial in their nature.
**Conclusion:** Primary lung tumors in children are rare and histopathologically diverse. They mostly present as endobronchial and there is usually a delay in their diagnosis. For children with persistent respiratory symptoms and chronic wheezing who are not responding to conventional treatment, the possibility of an endobronchial lesion should also be considered in their differential diagnosis. The author’s personal cases highlight the issue. **Keywords:** Children, Lung Tumor, Endobronchial Tumor

**Pulmonary embolism in children: causes, presentations, diagnosis, treatment and prevention**

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**Background:** Although pulmonary embolism (PE) in children is not as common as in adults, but failure to diagnosis and treat can be fatal. During the pediatric age, it has a bimodal distribution in neonatal and adolescence periods. A prevalence of 0.7 to 4% has been reported in autopsies in children, whereas the reported incidence in medical registries is 0.9/100,000 admissions. This indicates that many cases of PE are undiagnosed. Predisposing factors include central venous catheters, prothrombotic disorders like factor V leiden mutation, prothrombin gene mutation, antithrombin III deficiency, protein C deficiency, protein S deficiency, elevated homocysteine, hyper osmolar state in diabetic patients, malignancy, infection, immobilization, nephritic syndrome, after surgery and in children with congenital heart disease following cardiac surgery using prosthetic valves or following Fontan operation. Clinical presentations are acute dyspnea, pleuritic chest pain, tachypnea, cough, tachycardia, hypoxemia, hypotension and sudden collapse. D-Dimer is elevated in more than 80% of cases. Normal D-dimer level has also been reported. Imaging modalities like pulmonary angiography by cardiac catheterization of CT angiography are sued for diagnosis. Treatment includes administration of thrombolytic agents. Successful endovascular thrombolytic therapy has also been in infants less than 24 month old. Deployment of IVC filters can prevent development of PE in high-risk patients and is also helpful for prevention of progression of PE in affected children. **Keywords:** Pulmonary, Embolism, Children

**Cardiovascular complications in a burned child; a case report**

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**Background:** Infectious endocarditis is a rare complication in burned patients that causes mortality and morbidity. **Methods:** We report a 3-year old girl with prolonged fever after boiled water burning, two months ago. She experienced fourteen sections of surgical debridement and grafting. Huge vegetation on the tricuspid valve and mild pericardial effusion with some debris were detected during sepsis work up. At the same time multiple resistant pseudomonas aeruginosa was grown in blood culture which was only sensitive to cefotin. **Findings:** Inserting pericardial window, 20 ml pus was drained and huge mycotic aortic aneurysm was detected. Despite anti-microbial therapy, replacing tricuspid valve with a synthetic one, and aortic graft, she died with a severe endophthalmitis. **Conclusion:** Infectious endocarditis should be considered in febrile burned patients. Also ophthalmic evaluation seems necessary in a patient who has had a surgery for mycotic aneurysm. **Keywords:** Infectious Endocarditis, Burning, Mycotic Aneurysm, Endophthalmitis

**Congenital chylothorax treated with octreotide**

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**Background:** Congenital chylothorax is the accumulation of lymphatic fluid within the pleural space. Cases unresponsive to conservative management usually require surgery. Although octreotide has been used successfully to treat chylothorax, the exact mechanism of its action is uncertain and it is believed to reduce the lymphatic drainage through a direct action on splanchic lymph flow. **Case presentation:** We report two cases of congenital chylothorax who did not respond to supportive therapy (nutritional support and drainage) and where surgery service were avoided with the compassionate trial of octreotide. The first case was a 50-day-old infant with Noonan syndrome phenotype and another was an 18-day-old neonate with idiopathic congenital chylothorax. Treatment was associated with prompt respiratory improvement soon after starting the octreotide treatment and in the first and second cases the clinical symptoms resolved completely in 12 and 10 days after the onset of the treatment, respectively. Octreotide infusion was started at an initial dosage of 3 μg/kg/hour and increased daily by 1 μg/kg/hour to maximum 8 μg/kg/hour. The patients were well 2 months after the treatment. **Conclusion:** Octreotide infusion appears to have a good safety profile in newborns and remains a promising alternative to surgery for recalcitrant cases of chylothorax. Further studies are required to ascertain its true value in congenital chylothorax. **Keywords:** Congenital chylothorax, pleural effusion, respiratory distress syndrome

**Exogenous lipid pneumonia: a case report of dramatic clinical and radiological improvement after multiple segmental bronchoalveolar lavages**

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**Background:** Acute exogenous lipid pneumonia is a consequence of exposure to significant quantities of mineral oils. Accidental poisoning is the most common cause of the acute form in children. Chest x-ray findings include consolidation, nodular lesions, reticular and alveolar-interstitial patterns. Lower lobes or right middle...
lobe is more involved, but multifocal or bilateral patterns can be seen too. Lipid-laden macrophages, which cause fibrosis in the alveoli and interstitium can be removed by multiple bronchoalveolar lavages (BALs).

**Case presentation:** We present a rare case of exogenous lipoid pneumonia in a 2.5 year old boy presented with cyanosis, severe respiratory distress who admitted to the pediatric intensive care unit. A chest x-ray showed diffuse opacities with an alveolar pattern in both lungs. CT scans of the chest revealed bilateral diffuse severe pneumonitis with ground glass and alveolar appearance and crazy paving. Bronchoscopy was done. Mucopurulent secretions aspirated then rinsed with saline. Cytological analysis of BAL fluid stained with sudan III showed many lipid laden macrophage (grade IV/IV). After weekly multiple segmental BAL, BAL fluid was almost clear and the cell count became in normal range values. After the forth therapeutic BAL, CT scan was performed which revealed dramatic improvement and last cytological analysis of bronchoalveolar lavage fluid showed few lipid-laden macrophages, much less than previous specimen. After one month hospitalization the patient discharged and recommended to return to follow up. Conclusion: The present study indicates that therapeutic multiple BAL is an effective method to remove intra-alveolar mineral oil with significant improvement of clinical, radiological and laboratory findings.

**Keywords:** Lipoid Pneumonia, Bronchoalveolar Lavage, Computed Tomography, Mineral Oil
Endocrinology and Metabolic Disorders

Precocious puberty

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Precocious puberty refers to the appearance of physical and hormonal signs of pubertal development at an earlier age than is considered normal. The onset of puberty before 7 or 8 years for white girls and 9 years for boys is considered precocious. Early onset of puberty can cause several problems. The early growth spurt initially can cause tall stature, but rapid bone maturation can cause linear growth to cease too early and can result in short adult stature. The early appearance of breasts or menses in girls and increased libido in boys can cause emotional distress for some children. Premature pubarche and premature thelarche are 2 common, benign, normal variant conditions that can resemble precocious puberty but are nonprogressive or very slowly progressive. A thorough history, physical examination, and growth curve review can help distinguish these normal variants from true sexual precocity. If the history, physical examination, and laboratory data suggest that a child exhibits early and sustained evidence of pubertal maturation, the clinician must differentiate central precocious puberty (CPP) from precocious pseudo puberty. Central precocious puberty, which is gonadotropin-dependent, is the early maturation of the entire hypothalamic-pituitary-gonadal (HPG) axis, with the full spectrum of physical and hormonal changes of puberty. Precocious pseudo puberty is much less common and refers to conditions in which increased production of sex steroids is gonadotropin-independent (Precocious pseudo puberty). Correct diagnosis of the etiology of sexual precocity is critical, because evaluation and diagnosis of patients with precocious pseudo puberty is quite different than that for patients with central precocious puberty.

Keywords: Precocious Puberty, Central, Pseudo Puberty, Diagnosis, Treatment

The prevalence of diabetes in students arriving to school in Hahavand city in 1392-93

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Background: Diabetes is a metabolic complex disease in which such factors as intrinsic aptitude, environmental conditions like life style, weight, food patterns range and people's awareness have determining role in it's prevalence. This chronic disease is classified into 2 types.

Methods: This investigation was carried out through medical examining in the health measurement plane of the pre-elementary students (7 years) in Nahavand city, 2012-2013. Data was gathered through interview and examining; and it was analyzed through statistical testing of Chi-square, analysis and variance, t test, and coefficient correlation. P<0.05 was considered as a meaningful level.

Findings: 2223 students (1125 boys, 1098 girls) were investigated regarding diabetes signs from which 420 people were considered as patients. Out of this number, 27 people were confirmed to have diabetes (12 boys, 15 girls). There was not a meaningful differences between sex and the disease spread. 19 patients of 27 patients were informed of their own disease. A meaningful relationship was observed between the parents' education and the disease prevalence. A meaningful relationship was observed between the parents' education and awareness of the child disease. A significant relationship was not observed between the parents' age and the diabetes prevalence.

Conclusion: This study and similar research indicate that with increasing growth of the number of people involved with diabetes 1 (students are treated as a branch of this group) the necessity of early diagnosis and educational programs regarding diabetes are unavoidable in schools. Thus, it is suggested that essential teaching for teachers and tutors concerning the students affected with diabetes will improve educational achievement of the students.

Keywords: Prevalence, Diabetes, Student

The diagnostic dilemmas of skeletal dysplasia: classification, incidence and mode of inheritance of different type (a clinical and radiological overview)

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Skeletal dysplasia are a primary result of mutation genes that are expressed in chondro-osseous tissue. However, there are also some other forms of secondary dysplasia caused by abnormalities of extra-osseous factors with effects on the skeletal system like metabolic errors in hypophosphatemic rickets. International nosology and classification of genetic disorders of bone (published 2009) includes 37 groups; from 250 well defined disorders about 70 are often lethal in the perinatal period. Primarily the classification of skeletal dysplasia is based on radiological features of skeletal changes. However, the progress in molecular genetics especially in the last decade permit owing to distinctions at the DNA level refinements in classification resulting in an increasing number and subtypes of dysplasias. The similarity of morphological findings with or without the same inheritance as well as their rare incidence are all reasons for diagnostic dilemma in this field. The first step in the diagnosis of skeletal dysplasia is to observe a disproportionate skeletal development, unusual habitus with or without mental retardation as well as other clinical signs. The second step is more difficult and includes genetic counseling and interpretation of radiologic skeletal changes in affected patients and their families. The rare incidence of skeletal dysplasia on one hand and its morphologic diversities on the other hand require some experience and therefore, centralization of knowhow in this field is recommendable. In this manner medical geneticists and radiologists gain the ability and expertise in this field. The aim of this presentation is to give an overview of skeletal dysplasia; a large number of different types will be shown. Special attention should be paid to their inheritance, molecular pathology and incidence as well as the radiological findings.
**Effect of growth hormone deficiency on brain MRI findings among children with growth restrictions**

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**Background:** Growth hormone deficiency (GHD) is a major problem among children with short stature. In this study, the role of brain magnetic resonance imaging (MRI) in defining the underlying defects among short children with GHD was evaluated.

**Methods:** In a cross-sectional study, data of 158 children were evaluated. Growth hormone (GH) levels were measured using stimulating tests and brain MRI with gadolinium contrast was applied, as well.

**Findings:** Some 25.3% of patients had GHD with a mean age of 8.01±3.40 years. MRI results showed 35 patients as normal, four patients with pituitary hypoplasia, and one patient with microadenoma. The MRI results were significantly associated with GH levels and presence of other endocrine disorders. There was a significant association between prenataal disorders and patients’ bone age delay.

**Conclusion:** In patients with severe GHD and patients with multiple pituitary hormone deficiencies, MRI is more likely to be abnormal, and bone age is much delayed in patients with history of prenataal disorders.

**Keywords:** Bone Age, Growth Hormone Deficiency, Magnetic Resonance Imaging, Short Stature

**Pediatric metabolic bone diseases classification and an overview of clinical and radiological findings**

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From a practical point of view the metabolic bone diseases should be categorized etiologically because of an overlap of some endocrine bone diseases and skeletal dysplasia. The metabolic bone diseases affecting bone formation and mineralization may be classified as follows (4 groups): A. Disorders with insufficient mineralization of the organic matrix: The group includes different types of rickets following vitamin D abnormalities. The most frequent causes of vitamin D deficiency are nutritional, hepatic- and renal disorders. Also rickets of prematurity, renal tubular disease and hypophosphatemia should be classified in this group. Phosphopenic rickets is an autosomal dominant abnormality with phosphate wastage. The radiological findings in all different types of rickets are similar with some differences depending on severity and according to type with variable structural changes. B. Abnormalities of bone matrix formation: Osteogenesis imperfecta with decreased bone mineralization due to insufficient matrix formation and quantitative and qualitative defects in synthesis of type I collagen. The radiographic feature has a wide variation in 5 major types and subtypes. In general osteoporosis, retarded calvarial bone formation, rib- and tubular bone fractures and collapsed vertebral bodies are all characteristic findings as well as deformities of limbs. Menkes disease and scurvy: collagen synthesis requires ascorbine acid and copper dependant ascorbine acid oxidase. Therefore children with scurvy and Menkes disease (copper deficiency syndrome) are both affected in addition to other clinical signs and symptoms with skeletal changes as a result of osteoid deficiency. The characteristic changes are osteopenia, bone fragility, metaphyseal fractures and others. Abnormalities of increased or decreased bone resorption: The major cause of increased bone resorption is hyperparathyroidism in children with renal failure. Radiologically they manifest themselves as renal ostedyostrophy with subperiostal resorption along the phalanges, involving also the distal phalangeal tufts. Soft tissue calcification and fractures among others also occur. Hyperphosphatasia is a rare autosomal recessive disease and is also a condition with increased bone turnover, and elevated serum alkaline phosphatase level. It appears to be due to deficiency of osteoprotegerin. Radiological signs are expanded and bowed diaphyses. There is a marked deminerilization with a heterogeneous pattern and cortical thickening, and pathological fractures. Osteopetrosis is an example of decreased bone resorption with calcified cartilage and abnormality of the osteoclast function. The different types are divided into infantile and juvenile. The juvenile type is categorized in six sub groups. Mostly they are autosomal recessive herited with exception of autosomal dominant type I and II of the juvenile type. D. Pharmacologic and toxic changes in the skeleton. There are several pharmacologic and toxic materials affecting the resorption of bone and calcified cartilage, including side effects of biphosphonate and prostaglandine and toxic effects by hypervitaminosis A and D and heavy metal poisoning (lead). Also primary oxalosis due to oxalate overproduction should be classified in this group. Obvious radiologic changes are different and characteristic. E.g. increased bone density and opaque dense lines due to cyclic therapy will be found following biphosphonate treatment. Lead lines due to excessive calcified cartilage are observed in lead poisoning. Based on the above mentioned data, numerous types of metabolic bone disorders will be shown with special attention for etiology, typical structural and anatomic changes in different disorders.

**Keywords:** Pediatric, Metabolic, Bone, Classification, Clinical, Radiology

**Diabetic ketoacidosis and predisposing factors in diabetic patients**

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**Background:** Diabetic ketoacidosis is a final result of severe insulin deficiency in type 1 diabetes that occurs in 20-40% of children and it is a life-threatening complication. This phenomenon is a medical emergency and requires prompt diagnosis and treatment. The aim of this study was investigation of frequency and causes of diabetic ketoacidosis and its consequences to prevent complications.

**Methods:** This study was performed on 128 diabetic patients in Amirkola Children’s Hospital (1384-1392). Patient information was recorded in a predetermined questionnaire and data were then entered into SPSS version
Evaluation of the patients with Diabetes Mellitus in pediatric hospital of Khoramabad

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Background: Diabetic Mellitus is the most common endocrin-metabolic disorder of childhood and adolescence with important consequences for physical and emotional development. Morality and morbidity of acute metabolic derangements and long term complications that affect small and large vessels result in retinopathy, nephropathy, ischemic heart disease and arterial obstruction with gangrene of the extremities. The aim of this study was evaluation of patients with diabete mellitus in pediatric Hospital of Khoramabad in 2009-2013.

Methods: This study was done as a cross-sectional. Sampling was simple. All the patients that hospitalized with diagnosis of diabetic mellitus were evaluated. The studied variables included age, sex, clinical and paraclinical sign, duration of hospitalization and treatment. The data was collected by questioner and analyzed by SPSS software.

Findings: Out of 100 patients, 70% were females and 30% were males. 10% were <5 years old, 50% were 5-10 years old and 40% were >10 years old. 55% were urban and 45% were rural. 35% had polydipsia, polyuria and polyphagia. 70% were hospitalised with DKA. 25% had nocturnal enuresis and constipation. 20% had an infection disease. Duration of disease onset in 65% was 15-30 days. Mean of the BS was 384±96. 75% had glucosuria 3+. Duration of hospitalization in 70% was 6-9 days. One of the patients died.

Conclusion: In this study 70% of the patients were females. 70% were hospitalised with DKA. Considering the diabetes is the most common endocrine disorder, we suggest educational programs for parents.

Keywords: Diabetic Mellitus, DKA, Children

Genetics & public health: genetic prevention in the PKU program of Iran

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Background: A program for prevention and control of Hyperphenylalaninemia as the head part of a comprehensive programme for hereditary metabolic disorders was initiated by the ministry of health in 1997. The program mainly aim to provide patients with standardized care and their family and relatives with prevention services.

Methods: Thirty five designated children hospitals in the provinces are responsible to register PKU patients of the province. A clinical team including a metabolic-endocrine subspecialist (or general pediatrician), a nutritionist, a psychiatrist, welfare worker, lab expert and drug technician, work together to deliver services in the regular bases. Parents are referred to the genetic counseling services of the district by the clinician of the team. When patients screened receive their services. Parents in the sessions are informed of the risks and options to avoid them. Patient's relatives who are at risk of the diseases are also invited in following sessions and receive genetic counseling services.
Prevalence of vitamin D deficiency in healthy children in Kashan

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Background: The data on the prevalence of vitamin D deficiency in healthy children from Iran is limited. Therefore, the current study was performed to evaluate the level of vitamin D in healthy children in Kashan.

Methods: The cases was selected from the healthy children aged 1-6 years who referred to the health centers for checking their weight and height during 2013. The children who were suffering from liver, kidney or any endocrine diseases or were on supplementary diet for vitamin D were excluded from the study. Prior to the start of the protocol, 3 cc of venous blood was drawn from the children. Standard DIA source kit and ELISA method were used to measure the level of 25-hydroxy vitamin D. Based on the criterion set results of the recent studies, the levels of vitamin D below the 10 nanograms per milliliter is classified as severely low, between 10 to 30 as low, 30 to 100 as normal and above 100 is defined as the toxic level.

Findings: In total, 100 children (54 boys, 46 girls) with mean age of 2.6 years were enrolled. The percentages of children with severely low, low, normal and toxic levels of 25(OH)D were 32, 47, 21 and 0, respectively.

Conclusion: Level of vitamin D was remarkably low in the children in this study. Nevertheless, further studies are recommended in this regard.

Keywords: Vitamin D Deficiency, Healthy Children, Kashan

Gluaric aciduria Type II, clinical presentation and medical approach

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Background: Glutaric aciduria II is a rare autosomal recessive disorder. This results from mutation in at least 3 genes included ETFA, ETFB, and ETFDH.

Case presentation: The patient, a boy, was the first child of consanguineous healthy parents. He was referred for seizure. On examination, he had no dysmorphic feature. In the lab tests, he had hypoglycemia, metabolic acidosis without ketosis. Blood spot screening from the baby by tandem mass spectrometry showed increases in long chain and medium chain acyl carnitines. Gas-chromatography mass spectrometry revealed significant elevation of urinary glutaric acid, 2 Hydroxyglutaric acid and Isobuteric acid. The pattern accumulated metabolites was consistent with defect in activity of acyl–coA dehydrogenase. He was treated with riboflavin and carnitin. Finally, he developed liver function impairment and lactic acidosis. Then, he died at 5 month of age. The genetic analysis confirmed glutaricaciduria II, [mutation in the ETFDH gene G>c(p.Gly381 Arg)c,1141].

Conclusion: Glutaricaciduria II ,or multiple acyl-coA dehydrogenase (MADD), is a rare autosomal recessively inherited disorder of fatty acid, amino acid and choline metabolism. The disorder is due to a defect in either the alfa or beta–subunit of electron transfer flavoprotein (ETF A, ETF B, and ETFDH). Patients with this disorder have an unusual pattern in which a large number of organic acids are found in elevated amounts in plasma and urine. Lactic acid and glutaric acid are the most prominent. Some patients respond dramatically to pharmacological doses of riboflavin and carnitin.

Keywords: Glutaric Aciduria Type II, Multiple Acyl-CoA Ehydrogenase Deficiency

New medical approach to diabetes mellitus

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Regarding worldwide accelerating high incidence of diabetes mellitus, new hopeful medical horizons are appearing every day. Drug discovery and development to approval is a long risky road, about 10-15 years. For every 5000-10000 compounds in pipelines only one substance will be approved. Variety of hypoglycemic agents was presented to diabetic patients. Generally they were divided to 7 groups: -Insulins: ultrashort acting, short acting, intermediate acting, long acting.-Insulin sensitizers acting through liver: Biguanides. -Insulin sensitizers acting through peripheral tissues: thiazolidinediones. -Insulin secretagogues: sulfonylureas and meglitinids.- Carbohydrate absorption inhibitors: α-glucosidase inhibitors. -Incretin mimetics, affect as glucagon like peptide 1(GLP-1) or inhibit dipeptidyl peptidase-IV (DPP-IV), and Amylinomimetics that promote satiety and reduce appetite and rate of gastric emptying. The last group of medication which has been approved is sodium glucose co-transporter2 (SGLT-2) inhibitors which inhibit glucose re-absorption in kidneys. Canagliflozin and Dapagliflozin are two FDA approved medicines of them. They are applicable for type two diabetics with appropriate kidney function (GFR > 60 ml/min/1.75 m²). Inhaled insulin were collected from markets in 2007 in the USA, recently is produced in a safe and more convenient form. It is prepared in single dose cartridges and the delivery system is a whistle size device. Both the insulin and the powder are nearly completely cleared from the lungs of healthy individuals within 12 hours of inhalation; only 0.3 percent of the insulin and 0.4 percent of the powder concentration remain after 12 hours. Ultra-long acting Insulin (degulides) is multihexamer insulin, mixable with short acting insulins, activated in physiologic PH subcutaneously once a day or three times a
week. Artificial pancreas is composed from injecting insulin pump and a blood glucose monitoring device that inject appropriate dosage of insulin regarding blood glucose. It stops insulin infusion when blood glucose is in critical range.

**Keywords:** Diabetes Mellitus, Insulins, Inhaled Insulin, New Approach, Artificial Pancreas

**Physical activity and exercise in children with diabetes mellitus type 1**

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Doing exercise is one of the important components in diabetic children management. ADA recommended that all children with diabetes should be encouraged to do exercise at least 60 min of physical activity each day. The benefits of exercise are weight control, reduced cardiovascular risk; reduced Hb A1C, improved insulin sensitivity and improved sense of well-being. Due to increased insulin sensitivity, risk of post-exercise nocturnal hypoglycemia is high, if bedtime blood glucose level is < 125 mg/dL. Since hypoglycemia is common during the exercise, shortly after, or even up to 24 hours afterwards, these recommendations for diabetic patients which are approved by ISPAD and ADA must be taken into consideration: • Adjusting insulin regimen to activity • Reducing insulin dosage before exercise if necessary. • Discussing type and amount of carbohydrate required for specific activities • Having snacks and sugar in case of emergency • Avoiding any exercise if pre-exercise blood glucose levels are high (>250 mg/dL) with ketonuria/ketonemia. Pre –exercise Blood glucose between 100 up to 250 mg/dL is safe. • Drinking a lot of sugar-free fluids during exercise. • Not having injected insulin at a site that will be heavily involved during activity. The children with diabetes should be allowed to choose their favorite sports. Diabetes does not limit their ability to do exercise. They should record details of their activity, insulin, food and glucose results for good diabetes control during exercise. The children with diabetes should be encouraged to be treated soon.

Each new case of PKU should be tested for malignant types associated. BH4 deficiency, an Iranian experience

**Conclusion:** In any undiagnosed neurologic disease we should consider biogenic amines synthesis defects and recommend urine, CSF investigations to treat as early as possible to prevent brain damage and to improve outcome. Each new case of PKU should be tested for malignant types to be treated soon. Mild forms of PTPS deficiency can cause movement disorders in adults. Two adult cases were reported in this paper.

**Keywords:** Hyperphenylalaninemia, Malignant PKU, Biogenic Amine Synthesis Defect

**The phenotypic variability in tetrahydrobiopterin deficiency, an Iranian experience**

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**Background:** Tetrahydrobiopterin (BH4) activates phenylalanine hydroxylase as well as tyrosine 3 hydroxylase and tryptophan hydroxylase. BH4 deficiency causes biogenic amine synthesis defect and none specific neurologic abnormalities as well. BH4 deficiency, a mild form of HPA is mostly accompanied by a deficiency of the neurotransmitters such as dopamine and serotonin, so causes severe cases if not diagnosed. Elevated urine neopterin as well as hyperphenylalaninemia is most often in favor of diagnosis of 6-pruvoyl-tetrahydropterin synthase (PTPS) deficiency. Thus far, over 250 patients with PTPS were described with various degrees of phenotypic severity. Treatment with BH4, levodopa, 5HTP and diet low in phenylalanine, if it is high, is effective in the most of them. Dopamine agonist (pramipexol) may well result in facilitated clinical response to L-dopa therapy in BH4 deficiency. The Objective of the study was to study clinical, laboratory data, and outcome of patients with BH4 deficiency.

**Methods:** We studied 14 patients suspicious to have biogenic amine synthesis defect, among 257 patients of PKU group and 534 patients with non-specific neurologic manifestations without defined diagnoses.

**Findings:** 14 cases; (4 female, 10 male), age range: 26mo-27yr, onset; adulthood(3), juvenile(3). Clinical findings were mild psychomotor retardation in 1( 1/14), convulsion in 5 (5/14), hypersalivation in 5 (5/14), distal chorea 2 (2/14), temperature disturbances in 3 (3/14) autism 1 (1/14), oculogyric crises; 1, awkward walking 4, hypertonia (3) hypotonia (1) hyper/hypo (1). High urine neopterin and urine neopterin/creatinine in 14(14/14), high plasma phenylalanine; 3(3/10) and low urine VMA;6(6/10).

Tetrahydrobiopterin (BH4) activates BH4 deficiency, an Iranian experience

Treatment with levodopa-carbibido,5 hydroxy tryptophan and BH4 was effective in those which was started in early stages of symptomatic disease; onset; adulthood (3) juvenile (3), and childhood (3).

**Conclusion:** In any undiagnosed neurologic disease we should consider biogenic amines synthesis defects and recommend urine, CSF investigations to treat as early as possible to prevent brain damage and to improve outcome. Each new case of PKU should be tested for malignant types to be treated soon. Mild forms of PTPS deficiency can cause movement disorders in adults. Two adult cases were reported in this paper.

**Keywords:** Hyperphenylalaninemia, Malignant PKU, Biogenic Amine Synthesis Defect

**Association of 25-hydroxy vitamin D levels with indexes of generalized and abdominal obesity in Iranian adolescents: the CASPIAN-III Study**

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**Background:** This study aims to determine the association of serum 25-Hydroxy vitamin D (25(OH)D) levels with measures of generalized and abdominal obesity in Iranian adolescents.

**Methods:** This nationwide study was conducted among 1090 participants, aged 10-18 years, living in 27 provinces in Iran. Serum concentration of 25(OH)D was analyzed quantitatively by direct competitive immunossay chemiluminescence method. Body mass index (BMI) and waist-to-height ratio (WHtR) were considered as measures of generalized and abdominal obesity, respectively.

**Findings:** The mean age of participants was 14.7 ± 2.6 years; BMI 19.3 ± 4.2 kg/m2, the median serum 25(OH)D was 13.0ng/mL (interquartile range 6.8 – 27.4). A total of 40 % were vitamin D deﬁcient and 39 % were vitamin D insufﬁcient. Serum 25(OH)D level was not associated with BMI and WHtR.

**Conclusion:** We did not document signiﬁcant association between serum 25(OH) D level and anthropometric measures. This finding may be because of considerably high prevalence of hypovitaminosis D in the study population.
Keywords: Obesity, 25hydroxy Vitamin D, Adolescents

PKU: treatment standards in Europe

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The ESPKU is publishing this PKU:Closing the Gaps in Care report with one clear objective—to give PKU patients and their families a greater voice. The ESPKU envisages to initiate discussions and actions to enable a fairer deal for all PKU patients no matter where they live. The research and this report do not amount to scientific review of the management of PKU in Europe. The research on which this report is based to provide a useful insight into some of the issues affecting people with PKU and their families and gives an indication of some of the discrepancies in PKU care between some European countries. Phenylketonuria (PKU) is caused by a deficiency of phenylalanine hydroxylase, the enzyme converting the amino acid phenylalanine (Phe) into tyrosine. This causes the level of Phe in the blood to rise. Cases vary from mild to severe, with severe cases having very high blood Phe concentrations. A severe case, left untreated, will result in profound and irreversible mental disability. An abnormally high blood level of Phe is known as hyperphenylalaninaemia (HPA). This may be caused either by PKU or by a deficiency in one of the enzymes synthesizing or recycling the co-factor tetrahydrobiopterin (BH4), which stimulates the above called enzyme phenylalanine hydroxylase. Almost all European countries carry out neonatal screening programs that identify HPA cases at birth. Cases with HPA are referred to special clinics to identify whether the child has PKU or BH4 deficiency. Once identified, a child with PKU is placed on a low Phe diet which, if initiated soon after birth, will prevent most of the neurological complications. In practice, a low Phe diet consists of a diet low in natural protein combined with a high intake of a protein substitute which has all amino acids but Phe. Due to the restrictive nature of this diet, however, compliance tends to diminish as the child gets older. Drug treatment can allow some PKU patients to keep their Phe levels under control while on a less restrictive diet. However, data on long term drug treatment compliance on large scale of patient population is further needed.

Diagnose communication in Inborn Error of Metabolism

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Giving a diagnosis of an inborn error of metabolism is one of the most important moments for parents and patients with a significant laboratory result (after newborn screening or clinical presentation) as well as for the metabolic specialist. The given information will change the life of the family in a meaningful way. Parents and patients will remember the situation for the rest of their life. Likewise it will be the start of a close relation between the family and the metabolic team. The material of this modular system aims to support knowledge, communication and cooperation. It has been developed by specialists in the field with long-standing experience of treatment, care and research of inborn errors of metabolism. It has been repeatedly tested and modified and is based on the evidence that each patient asks the same questions: 1. What does the disorder mean? (terminology), 2. How was it discovered? (diagnostics), 3. How did I get it? (etiology), 4. What exactly is wrong? (nosology), 5. how does it show? (symptomatology), 6. Is it severe, dangerous, perilous, life-shortening? (outcome), 7. What can I expect? (prognosis), 8. What can be done (how, how often, how long, by whom)? (treatment/cure), 9. Will it recur? (development & transmission)

Case presentation about non ketotic hyperglycinemia

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You are called to the newborn nursery regarding a 8 hour old female infant who is listless and not interested in feeding. On exam, the baby is severely hypotonic and lethargic but no other obvious abnormalities are noted. Glucose shows normal glucose. Blood gas, complete metabolic profile, CBC, plasma ammonia, lactate and urinalysis all show normal results. Chest X-ray comes back normal. Along with other possibilities, you suspect a neuromuscular disorder and consult neurology. Maintenance IVFs are started. Pregnancy history is significant for decreased fetal movements. While awaiting neurology consult, the baby has apnea spells and develops myoclonic jerks, and is intubated. An EEG is performed and shows a “burst suppression” pattern.

Keywords: Non Ketotic, Hyperglycinemia, Convulsion

Is it not the time to do something for vitamin D deficiency?

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Background: Vitamin D deficiency (insufficiency) is a common nutritional disorder in our country. Vitamin D is important for the health of all human beings since birth. This study has been conducted in order to summarize epidemiologic researches regarding vitamin D deficiency in different parts of the country and to conclude if food fortification is necessary. Study designed in Thalassemia Research Center, Sari, Iran.

Methods: This was a narrative review about the current situation of vitamin D deficiency in Iran in December 2013. Related literature in Persian and English were explored since 10 years ago. Data source of this study was Medline, SID, PubMed, Scopus, Request, Web of knowledge, Springer, Ovid, Google scholar.

Findings: We found 27 cross-sectional researches regarding vitamin D status in Iran from 2003-2013. Also, there was a Meta analysis that has been done in 2008. 25(OH) D3 was measured mostly by radio immune assay (RIA) method. Most studies were done on adults. Definition of vitamin D deficiency was based on cut off of the kit in most studies; however, in some the serum parathryoid hormone (PTH) was measured and cut off for diagnosis was based on increased PTH. Maximum rate of severe vitamin D deficiency was reported as 47% in 2011.
Conclusion: Vitamin D deficiency is very frequent in Iran. Dealing with a national important nutritional problem is important. Fortification of a suitable food or edible product which in this case would be milk or cooking oil is one way. Increase public awareness about the problem and motivate people to do something in their expense is another option. In this case, taking vitamin D supplements as a regular basis daily, weekly or any other routines which could be available and cost effective may solve the problem. For all above one year old persons, our recommendation is to take 300,000 IU (as IM injection or oral dose) of vitamin D as a starting dose, then take 50,000 IU oral dose (as one pearl) every 3 months.

Keywords: Vitamin D2, Vitamin D3, Osteomalacia, Rickets, Prevention, Treatment, Fortification, Iodine, Iron

The influence of late treatment on oral communication skills in Farsi speaking children with Phenylketonuria

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Background: Screening programs have commenced for Iranian children with Phenylketonuria (PKU) in recent years. The aim of the study was to investigate oral communication skills in late treated children with PKU to find the influence of treatment onset on these skills.

Methods: In this cross-sectional study, 22 late-treated PKU and 8 early-treated PKU aged 4-6.5 evaluated by: a) Wechsler preschool and primary scale of intelligence (WPPSI) consist of verbal intelligence quotient (VIQ) and total intelligence quotient (TFQ) and by b) test of language development-third edition (TOLD-p:3) containing composite quotients of spoken language, listening, speaking, semantics, syntax, and organizing. Dependent on treatment onset, PKU children are divided into 3 groups: early-treated PKU (n=8) (patient from newborn screening program), late-treated PKU diagnosed before one year old (n=9), and late-treated PKU diagnosed after one year old (n=13). For all patients time of diagnosis and treatment onset was the same. The oral communication skills matched within PKU children and matched between PKU children and normal control. The relation between blood Phe level with spoken language and VIQ also computed.

Findings: Normal groups performed significantly better than early-treated children in all composite quotients of TOLD-p:3 and VIQ (p<0.002). Early-treated PKU children significantly performed better than late-treated children in spoken language (P<0.01), speaking (p<0.04), syntax (p<0.02) and VIQ (p<0.002). There were negative relation between Phe and VIQ (r=-0.79) in early-treated and negative relation between Phe and spoken language (r=-0.71), organizing (r=-0.82), and semantics (r=-0.82) in late-treated PKU children that had been detected before one year old.

Conclusion: We discovered that postponement in diagnosis and treatment onset affects children with PKU suffer more oral communication impairment. The sooner therapy is initiated; the more recovery in oral communication skills is obtained so that late-treated children before one year old compared to after it had superior performance in oral communication and cognitive function.

Keywords: Phenylalanine, treatment onset, oral communication

Association of serum 25-hydroxy vitamin D levels and liver enzymes in a nationally representative sample of Iranian adolescents: the CASPIAN III Study

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Background: Hypovitaminosis D is highly prevalent, and has several adverse health effects. This study aimed to assess the relationship of serum concentrations of 25-hydroxy vitamin D [25(OH)D] and liver enzymes in adolescents.

Methods: This population-based cross-sectional survey was conducted among a nationally representative multi-stage sample of 1095 adolescents (52% boys), aged 10-18 years, living in different provinces of Iran. Serum 25(OH)D concentration<30ng/mL was considered as hypovitaminosis D, and liver enzymes (alanine aminotransaminase, ALT and aspartate aminotransaminase, AST) of > 40 U/L as high level. To determine the association between serum 25(OH)D categories and elevated levels of liver enzymes, multiple regression models and linear regression analysis were applied, after adjustment for potential confounders. Odds ratio (OR) 95% confidence interval (CI) of serum 25(OH)D and elevated liver enzymes were assessed by logistic regression analysis.

Findings: Higher rates of vitamin D deficiency were documented among individuals with increased levels of liver enzymes. Compared to boys, median of 25(OH)D was lower in girls with elevated levels of liver function tests (12.75 vs. 25.60 ng/mL for ALT, and 13 vs. 14.10 ng/mL for AST), with marginally significant gender differences regarding AST.

Conclusion: We found a relatively high frequency of hypovitaminosis D among adolescents with abnormal liver function. Further prospective studies are needed to examine these associations from early life.

Keywords: Vitamin D, liver function tests, adolescents

Dental problems in hypophosphatemic rickets

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Background: Hypophosphatemic Rickets is an uncommon metabolic bone disorder which affects all ages and both sex. It is characterized by low concentration of serum levels, impairment of mineralization of bone matrix and teeth with variable etiology. Dental problems have not been described well in previous studies.

Methods: All hypophosphatemic rickets patients who came to referral clinic during 2008-2010 enrolled in this study. All patients had low phosphorous level and high ALP & normal PTH & 25-hydroxy vit D and normal or low serum calcium. After diagnosis by a all patients were examined by dentist for enamel hypoplasia, taurodontism, dental abscesses, dental caries and dentition delay.

Findings: Nineteen patients were enrolled in this study. The average age of patients was 10 years (range 3-17). 79% patients had regular follow up after diagnosis of background disease. Dental caries and delay in dentition were most prevalent each one(47.7%) followed by enamel hypoplasia in 42.1% patients. Other problems were...
taurontism in 15.8% patients, dental abscess and gingivitis in 10.9%.

**Conclusion:** Hypophosphatemic rickets is a disease with different clinical features that one of them is dental blem. Dental caries is the most common problem.

**Keywords:** Dental Caries, Hypophatemia, Rickets, Enamel Hypoplasia

**Approach to hypocalcemia with three presentation**

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**Case Presentation:** Case 1: A 2-year-old girl was referred to our hospital because of failure to walk and absent teeth development. The patient appeared to be well nourished and content. His body mass index, height and weight were 20.1 kg/m² (90th percentile), 87 cm long (25th percentile) and 14 kg (75th percentile) respectively. Palpation of the patient's extremities revealed prominent, widening of wrist and ankle. The result of a total serum calcium test was 7 mg/dl. Case 2: A 7-year-old boy had short stature (< 3rd percentile) which presented with trunkal hard, nodular skin lesions, and he had mild obesity and developmental delays. Because a skin biopsy demonstrated subcutaneous calcification, his total serum calcium level was measured and found to be 7.5 mg/dl. Case 3: A 14-year-old girl presented with intermittent numbness of her extremities, and “lost control” of her right leg and falling. A CT scan showed the basal ganglia calcification. Her total serum calcium and phosphorus level was 6 and 7 mg/dl respectively.

**Keywords:** Hypocalcemia, Calcium, Short stature
**Gastroenterology & Nutrition Abstracts**

**Does low birth weight predict hypertension and obesity in schoolchildren?**

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**Background:** Birth weight appears to play a role in determining high blood pressure and obesity during childhood. The purpose of this study was to investigate the association between birth weight (BW) and later obesity and hypertension among 10 to 13-year-old schoolchildren.

**Methods:** A total of 1184 primary school students were selected from 20 randomized schools between 2011 and 2012 in Iran. Height, weight, waist circumference (WC) and blood pressure (BP) were measured using standard instruments. Data were analyzed using Stepwise regression and Logistic regression models.

**Findings:** 13.5% of children had a history of Low Birth Weight (LBW). First-degree family history of obesity, excessive gestational weight gain and birth weight were significantly correlated with obesity/overweight and abdominal obesity (p<0.001), whereas only birth weight was associated with high blood pressure (p<0.001). An inverse correlation was found between WC and SBP/DBP. Duration of breastfeeding in children with LBW was inversely correlated with obesity/overweight, abdominal obesity and hypertension.

**Conclusion:** The results suggests that BW is inversely associated with blood pressure and also with obesity and abdominal obesity. Duration of having been breast-fed could prevent any later hypertension, obesity and abdominal obesity. Further studies are needed to test these correlations as well as diagnosing early life factors to prevent young adult obesity, overweight or hypertension.

**Keywords:** Low Birth Weight, Children, Obesity, Hypertension

**Comparison of sequential and standard triple therapy for eradication of Helicobacter pylori in children**

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**Background:** Purpose of this study was comparing of sequential and conventional therapy regimens in eradication of Helicobacter pylori (H. pylori) infection in children.

**Methods:** Children with gastrointestinal complaints undergoing diagnostic endoscopy in Children's Medical Center Hospital between 2013 and 2014 which were 2 to 14 years old were enrolled in this study and 2 gastric antral wall biopsy specimens were taken for histologic investigation and rapid urease test (RUT). If Helicobacter pylori infection was documented by histology or RUT then the patient was included in intervention. Patients were divided randomly to two intervention groups. One group received conventional triple treatment including omeprazole 1 mg/kg/day for 30 days and a 10-day course of combination therapy with amoxicillin 50 mg/kg/day and metronidazole 20 mg/kg/day. Another group received sequential therapy including omeprazole 1 mg/kg/day for 30 days and a first 5-day course of amoxicillin 50 mg/kg/day followed by a second 5-day course of combination therapy with clarithromycin 15 mg/kg/day and metronidazole 20 mg/kg/day. Patients received all drugs with divided doses two times a day. All patients were reinvestigated by stool antigen test for H. pylori eradication 1 month after finishing the accomplished treatment.

**Findings:** At all 87 children recruited for this study. Thirteen patients didn’t complete the treatment or follow-up test. Sixty four patients enrolled in analysis which had a mean age of 9.13± 3.13 (range: 2-14) years. There were 28 male (43.75%) and 36 female (56.25%). Totally treatment was successful in 71.9% of patients. The sequential therapy-treated group showed more eradication rate than conventional therapy-treated group (83.9% versus 60.6%) that was statistically significant (P<0.039). Treatment side effects (abdominal bloating, nausea and vomiting) were not different significantly between 2 groups, but diarrhea was less frequent during sequential therapy (p<0.003). The most common clinical presentation was abdominal pain and the most common endoscopic finding was gastric nodularity.

**Conclusion:** Our study demonstrated that sequential therapy is greatly effective in eradicating H. pylori in children. This therapy regimen would be useful for the first-line option for H. pylori eradication.

**Keywords:** Helicobacter Pylori, Sequential Therapy, Standard Triple Therapy

**Extra intestinal manifestation of celiac disease**

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**Background:** Celiac is a common multisystem autoimmune disease with prevalence of about 1%. For every recognized case of celiac disease, 8 more remain undiagnosed. Asymptomatic or minimally symptomatic celiac disease is probably the most common form of the disease, especially in older children and adults. Celiac disease is no longer a disorder limited to childhood and adolescence; it has even been diagnosed for the first time in elderly patients. Infants and young children typically present with chronic diarrhea, anorexia, abdominal distension, abdominal pain, poor weight gain or weight loss, and vomiting. Severe malnutrition can occur if the diagnosis is delayed. Behavioral changes are common and include irritability and an introverted attitude. Rarely, severely affected infants present with a celiac crisis, which is characterized by explosive watery diarrhea, marked abdominal distension, dehydration, hypotension, and lethargy, often with profound electrolyte abnormalities, including severe hypokalemia. Older children with celiac disease who present with GI manifestations may have onset of symptoms at any age. GI symptoms in older children are typically less evident and include nausea, recurrent abdominal pain, bloating, constipation and intermittent diarrhea. Because of the myriad and frequently enigmatic presentations of celiac disease, the challenge of diagnosis falls squarely on the shoulders of primary care practitioners. In infants and toddlers, GI symptoms and
natural history, characteristics and complications of FR.

**Keywords:** Food, Regurgitation, Iran, Infant, Prevalence, Risk Factor

### Management of GI compromised children

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Gastrointestinal function can be compromised in children with a variety of disorders either directly as in Crohn's disease and short bowel syndrome, or indirectly as in cerebral palsy. Whey-based diets have been shown to reduce the degree of regurgitation, gastric emptying times, and gagging in neurologically impaired children. Even an energy-dense (1.5 kcal/mL) whey-based formula showed equally good tolerance and gastric emptying as compared with a regular strength (1 kcal/mL) formula. These diets are well-tolerated and provide nutrition to maintain and achieve growth. Gastrointestinal disorders should be to meet their nutritional needs and promote growth without compromising tolerance. Peptide-based enteral diets are often used as the basis of nutritional therapy for some children with compromised gastrointestinal function. A 100% whey, peptide–based diet containing insoluble and prebiotic fiber was as well tolerated as a commercially available, fiber-free control diet in a small and heterogeneous population of children with impaired gastrointestinal function and was associated with a substantial improvement in stool consistency, especially in neurologically impaired children.

**Keywords:** Gastrointestinal Compromised, Children, Management

### Assessment quality of life in Iranian food allergic patients' families

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**Background:** Food allergy is common disorder especially in the first years of life (Incidence=6-8%). Food avoidance is the only effective method for controlling signs symptoms of this disorder up to now. But strict avoidance need continuous vigilance of caregivers about feeding related matters and activities that causes persistent stress and decreases their quality of life. We decided to assess burden of food allergy on parents in Iranian patients. For this aim we provided and approved first Persian food allergy specific quality of life questionnaire.

**Methods:** We chose FAQL-PB questionnaire and after translation approved its Reliability and Validity. Then in a cross-sectional study we assessed quality of life in 90 Iranian parents of under avoidance food allergy patients in Tehran children hospital with that questionnaire during 2012.

**Findings:** Our questionnaire reliability was approved (ICC=0.75% & CRONBACHα=0.09). The most common allergens were wheat (60%) and cow milk (42%). The greatest burden of food allergy on quality of life observed in emotional domain. In this domain, half of parents had extremely affected quality of life. Quality of life was obviously affected in parents of female patients than male patients (P<0.002). History of anaphylaxis had not burden on each domains.
Liver function tests in children

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Liver, the largest internal organ in the body has many complex functions. It acts as a filter of the blood received from the GI tract through the portal vein. It synthesizes proteins that are involved in vital functions and is an important site of carbohydrate, protein and lipid metabolism. It detoxifies toxins, metabolizes drugs and hormones, conjugates bilirubin and excretes it through the bile. Liver dysfunction, therefore, has catastrophic consequences on the body. Liver function tests comprise of a battery of tests that are used: a. as a tool for screening and documenting liver injury, to provide vital clues to the etiology of liver disease, to monitor the disease progression and response to treatment and to assess prognosis in children with liver failure. The liver function tests can be categorized into five: A. Tests to detect hepatocyte injury B. Tests to detect cholestasis or impaired bile flow C. Tests that assess synthetic function of liver D. Tests that assess metabolic and excretory function of liver E. Tests that assess liver fibrosis. Appropriate utilization of the tests requires knowledge of the injury patterns in liver diseases. 1. Liver function tests must be interpreted in the context of a clinical diagnosis 2. These tests lack sensitivity and so normal results do not confirm absence of disease. 3. Liver function tests are useful in identifying the pattern of liver disease rather than arriving at an etiological diagnosis. 4. Age specific normal values have to be employed when interpreting lab tests like alkaline phosphatase, GGT and prothrombin time. 5. Gross elevation of transaminases occurs in primary hepatocellular diseases while ALP and GGT are very high in cholestatic diseases. 6. Increases gi bilirubin with falling transaminases denotes poor prognosis in acute severe failure.

Keywords: Liver Diseases, Liver Function Test, Children

The effect of adjuvant probiotic therapy on recovery from acute gastroenteritis in outpatient children over 2 years: a double-blind randomized controlled trial

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Background: Acute gastroenteritis is one of the important causes of mortality in infants and children and one of the six factors of death among children. The aim of this study was to compare the effect of Zinc Sulfate and Saccharomyces boulardii probiotic (Yomogi) on children over 2 years with acute gastroenteritis admitted to the pediatric clinic during 2011 to 2013.

Methods: In this clinical trial children were studied in two groups of case and control. The control group received standard treatment of diarrhea (fluid therapy, continuing feeding and Zinc Sulfate syrup 5 cc QID) and case group received Saccharomyces boulardii probiotic capsule with trade name of Yomogi (250 mg daily) in addition to standard treatment. Patient assigned to placebo received the same capsules as Saccharomyces boulardii probiotic capsule but without Saccharomyces boulardii probiotic. Demographic information, diarrhea status, appetite recovery and mean frequency of diarrhea before and during the treatment period were recorded and then data were analyzed.

Findings: There were 100 children in each group. Average age of children was 35.9±6.18 months in the control group and 37.6±3.64 months in case group. Mean duration of symptoms and times of diarrhea after treatment in the two groups had a significant difference. In addition, the number of days until recovery, patient’s appetite and satisfaction between two groups had a significant difference. But Age, gender and the number of times of diarrhea before starting treatment did not have any significant difference and before treatment, both groups were similar in terms of severity of disease.

Conclusion: According to this study, a combination of zinc sulfate and saccharomycesboulardii probiotic (yomogi) is recommended in children with acute gastroenteritis.

Keywords: Zinc Sulfate, Yomogi, Gastroenteritis, Children

Prevalence of malnutrition based on three nutritional risk scores in an eastern Iranian pediatric hospital

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Background: Malnutrition is a major health problem in hospitalized paediatric patients. It is reported that the number of malnourished paediatric patients varies between 21% and 80% according to the level of the country’s development. It is essential that patients who are malnourished or at risk of malnutrition be identified as soon as they are admitted to the hospital. A recent study applied three of nutritional risk screening tools (STRONGkids, STAMP and PYMS) to children admitted to a tertiary children’s hospital in Iran. This study aimed to evaluate the nutritional status of hospitalized children in a tertiary paediatric hospital in Mashhad-Iran and compare the validity, ease of use, and the varying prevalence of malnutrition according to these three nutritional risk screening tools.

Methods: Three nutritional risk score tools were applied to all patients and classified into low, medium and high-risk groups. The anthropometry of hospitalized children was determined and classified using standard criteria. The validity and the ease of use of the tools was assessed.

Findings: Of children classified, 30.6% were found to be undernourished based on their WFH z-score and the prevalence of moderate and severe malnutrition was 22.8% according to the HFA. PYMS identified 23.5% in the medium-risk group and 52.2% in the high-risk group. STAMP identified 20.9% in the medium-risk group and 69.6% in the high-risk group. STRONGkids classified 71.3% of children as medium and just 7.8% as high-risk.
STAMP detected more malnourished children (21/21) compared to PYMS (2021) and STRONGkids (17/21).

Conclusion: NRS tools were able to detect children at a higher risk of nutrition deterioration; however, variable utility was observed. Further assessment of NRS tools in developing countries is required. In these countries, PYMS was the most reliable tool.

Keywords: Malnutrition, Paediatric, Nutrition Screening Tool, Iran

Congenital gastrointestinal anomalies in pediatric patients and their nutritional management

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Studies of other defects associated with specific congenital anomalies may be helpful to identify the causes of congenital anomalies, determine recurrence risks, and guide expectations for the efficacy of prevention strategies. Infants born with congenital anomalies demand individualized nutritional evaluations and recommendations. The anatomical changes of neonatal surgical diseases create specific physiological constraints. Patients with different congenital anomalies have different nutritional support needs. It is essential to know the exact physiology of these anomalies in order to be able to manage and provide them with appropriate and suitable nutritional supports. This article reviews several nutrition-centered options to aid the medical provider caring for babies with common surgical diseases.

Methods: Medline searches were performed using the keywords congenital anomalies, gastroesophageal reflux, nutrition and the text word nutrition in congenital GI anomalies. Bibliographies of recent review articles and relevant primary research reports, as well as current contents of ASPEN, ESPGHAN guidelines were reviewed for additional relevant citations.

Findings: Several commonly encountered surgical diseases of infants involved foregut and midgut anomalies, pulmonary hypoplasia, congenital diaphragmatic hernia, abdominal wall defects and also diseases treated with enterostomy.

Conclusion: Just within general and thoracic pediatric surgery, diseases include congenital anomalies in the infant, common acquired conditions such as pyloric stenosis and necrotizing enterocolitis, trauma, feeding tube replacement, and organ transplantation. To successfully support these patients, the dietitian must understand not only the essential details of these diseases, but also the particular ways that the diseases and their surgical treatments impose demands and constraints on nutritional support.

Keywords: Congenital Anomalies, Motility, Nutrition, Gastroesophageal Reflux

Pediatric gastrointestinal emergencies: priorities in use of different modalities

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There are a large number of gastro-intestinal diseases which manifest themselves as a life threatening emergency needing immediate diagnosis and treatment. An accurate history and data of physical and laboratory findings are necessary for an adequate imaging procedure. From a practical point of view the gastro-intestinal diseases should be divided into two groups. The first group includes neonates and infants under the age of three months. The congenital malformation is the most frequent finding in this period of life in the form of different types of atresia, stenosis or other functional and anatomic GI disorders. However acquired GI abnormality in this age group is not uncommon such as necrotizing enterocolitis or pyloric stenosis. The second group consists of infants and older children over the age of 3 months who suffer mostly from acquired gastro-intestinal disorders like appendicitis, intussusception and others. Congenital abnormalities are not frequent in this age group. Diagnostic imaging: The abdominal plain film is the first step in the evaluation of an acute abdomen. Look for pathological gas distribution, fluid levels, pneumoperitoneum or pathological masses. Sonography is an additional modality to search for inflammatory changes, free fluid, thickening of bowel wall and motility disorders of the intestine as well as cystic or solid masses. MR enteroclyses and endoscopy have high priority in some gastro-intestinal disorders like Crohn’s disease. CT sometimes is a valuable modality especially in abdominal trauma and suspicion of surgical complications. Nuclear scanning is useful in the detection of Meckel’s diverticulum or intestinal duplication. Contrast studies of the GI tract are necessary in some emergency cases. The aim of this workshop is to give an overview of pediatric gastrointestinal abnormalities with special attention to emergency cases. The priorities in the use of different modalities will be interactively discussed.

Keywords: Emergency, Priority, Imaging, Children, Gastrointestinal

Evaluation serum selenium level in children with acute gastroenteritis admitted in Mofid Children’s Hospital

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Background: Gastroenteritis refers to the passage of three or more loose or watery stools per day. Selenium (Se) is suggested that like other trace mineral and anti-oxidant enzymes is one of the important immune factor that neutralized free oxygen radicals. Selenium deficiency affects the occurrence, virulence, or disease progression of some viral infections. Recent research has suggested that selenium deficiency has role in gastroenteritis, therefore we designed a study for relation between serum selenium level and acute gastroenteritis.

Methods: This is a case-control study which performed on 80 infants and children in age of 6-36 months. In case group were 35 children with acute gastroenteritis and in control group were 45 healthy children. We measured serum selenium level by atomic absorption spectrophotometry method in groups. In control group performed one time and in case group two times during admission and recovery phase 7-10 days after disease. Data entered in SPSS software version 18 and analyzed by statistical tests.
Findings: Mean serum selenium level in acute phase is 80.8±18.2 µg/L and after 7-10 days in recovery phase is 105.6±18.4 µg/L. Mean serum selenium in control group is 94.5±15.3 µg/L. Therefore mean serum selenium level in acute phase was less than control group and recovery phase. Serum selenium level was decrease significantly in severe dehydration than mild and moderate dehydration (P<0.001). There is no relation between selenium level with age, weight, sex, breastfeeding, socioeconomic and underlying disease. No correlation was detected between serum selenium levels and the parameters above.

Conclusion: The results of this study indicate that decrease selenium level in acute phase of gastroenteritis compare to control group. In recovery phase, selenium level has significantly increased. We suggest for treatment of gastroenteritis pay attention to selenium deficiency and further studies are required for more result.

Keywords: Selenium, Children, Acute Gastroenteritis

Health effects of probiotics in infancy

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Based upon a wide range of putative modes of action involving modification of the gut microbiome probiotics have been the target of intensive research efforts. Consequently, there is now a massive literature on the health effects of probiotics with >800 completed randomized controlled trials and >100 meta-analyses. Many of these relate to infants and have been assessed by ESPGHAN and AAP to form the basis of their recommendations on the topic. It is important to appreciate that any health effects may vary according to the genera, species and strain of probiotic bacteria. They may also vary by dosage, timing, duration and the matrix with which they are administered. Thus health practitioners should take care to review the evidence carefully before prescribing. Both ESPGHAN and AAP conclude that probiotics in infant formula are safe and may have benefit in reducing GI infections. Probiotics administered separately from formula, and including to breast-fed babies, show strong (but sometimes species/strain specific) protection against acute gastroenteritis, antibiotic associated diarrhea, and nosocomial diarrhea and respiratory tract infections. There is evidence for protection against infant colic and against later allergy, asthma and eczema but further studies are needed. ESPGHAN, AAP and ASPEN all conclude that the evidence that probiotics protect premature babies against NEC is insecure.

Keywords: Probiotic, Gastroenteritis, Asthma, Allergy

Infant feeding and later risk of obesity: the role of protein

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Evolution has determined that human infants should grow very slowly compared to other species. This is to allow ample time for the development and training of a complex brain. A consequence of this evolutionary choice is that, after the first 3m of life, an infant’s nutrient needs for growth are very low. This is reflected in the composition of human milk where, in particular, protein levels are very low. Mature breastmilk milk contains ~0.8g protein per 100ml and ~0.25g non-protein nitrogen; about one quarter of the concentration in cow’s milk. Therefore old-fashioned milk formulas used to have a much higher protein content than babies are ‘designed’ to consume. There are competing hypotheses about what effect an excess of protein may have on infant growth and later adiposity. One view holds that high protein is obesogenic, whilst the alternative ‘protein leverage’ hypothesis would postulate the reverse. The former view was based largely on uncontrolled epidemiological association studies and the latter view has not been examined in children. Fresh light has been cast on the issue by the very recent publication of a controlled trial in which infants were randomized to receive formula containing two different concentrations of protein (selected as being at each end of the range permitted by CODEX regulations). This gives strong evidence that babies consuming the higher protein formula grow faster and fatter than those consuming the lower protein formula. This has implications for the continued trend towards humanizing infant formula milks.

Keywords: Obesity, Milk Formula, Infant Feeding
Health and Miscellaneous Abstracts

Effect of Urbanization on Child safety

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Background: The process whereby a society changes from a rural to an urban way of life is called urbanization. The world’s population living in urban areas has grown up from 14% to over 50%. It is associated with many changes including greater access to transportation and clear implications for determining the disease and injury pattern. This study was designed aiming to compare injuries related mortality and morbidity in urban vs rural settings among children in Iran.

Methods: Data was gathered from a household survey. A t-test was used to analyze the relationship between outcomes.

Findings: Injury in all ages accounts for 17% of all deaths in Iran. 20% of deaths in children is due to injuries. Traffic related injuries account for 42%. Airway obstruction and drowning are considered to be the second and third causes of death from injury among children respectively. Children in urban settings sustain injuries due to traffic accident, airway blockage and fall from height more than rural children. Drowning is more prevalent in rural areas than in urban (p<0.05). About 63.5% of morbidity related to injuries occurs in private homes or in residential areas e.g. yards and compounds. In these injuries no significant statistically difference was found between rural vs. urban setting (p<0.05).

Conclusion: Beside area level measures as an index for considering urbanization, other elements including quality of roads, distances from markets, types of markets available, transportation options accessibility of health services and so on should be considered as well. Safety for children is an important element to be provided prior to planning cities by urban developers. Expansion of the International Safe Community program is a potential solution.

Keywords: urbanization, child safety, safe community

Attention Bias Modification Therapy (ABMT) as a modern technique for obesity management in children

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Background: Pediatric obesity is an epidemic that challenges the health of children on all levels of health care system. During the past decade, pediatric psychologists have actively pursued an understanding of the psychosocial correlates of pediatric obesity and developed effective interventions based on this knowledge. Attention Bias Modification Therapy (ABMT) is an innovative intervention to increase children’s motivation for compliance of diet programs. It remains unknown if ABMT and related components following initial skills training into pediatric obesity interventions facilitate child behavior change following treatment cessation. It was concerning issue in this study.

Methods: ABMT uses the dot-probe task as a therapeutic tool by computer program. In this randomized clinical trial, 72 overweight/obese children were assigned to either self-directed or prescribed intervention for 8 cessions of ABMT or placebo. Anthropometric measurements from child at baseline, post-treatment, and 3-month follow-up were evaluated for change.

Findings: Participants who were randomized to receive ABMT reported better success in weight losing (p<0.043), adherence of self-management program for weight control (p<0.003), and shown better self-image and self-esteem (p<0.01) than those who received placebo.

Conclusion: The results of these studies show that there is potential in the application of ABMT to control weight, and a positive effect of ABMT on clinical outcomes suggests that this technique is worthy of future study as an intervention for obese children.

Keywords: Attention Bias Modification Therapy, Obesity, Management, Children

Quality of life of mothers of hard of hearing children

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Background: Hearing impairment in children can affect all the family members and mostly is associated with parental distress. This study was conducted to investigate the quality of life of mothers of hard of hearing children.

Methods: It was a cross-sectional study. The population comprised all the mothers of hard of hearing children in Karaj special primary schools for deaf. The study participants were selected through purposeful sampling, 30 mothers were volunteers for the study. Samples were been evaluated with the quality of life scale (WHO-QOL BREEF). Data was collected and processed through SPSS to calculate the mean score, standard deviation.

Findings: The results showed that the mean score of the quality of life mothers of hard of hearing children were significantly lower and under normal scores.

Conclusion: Quality of life mothers who have hard of hearing children is under normal range and appropriate interventional programs are necessary for them and they need special psychological support.

Keywords: Quality of Life, Mothers, Hard of Hearing Children

Evaluation of the experience of hearing and vision health control in children before entering school age

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Background: Joint Committee on Infant Hearing (JCH) guidelines axis based on the assumption that all infants before the age of one month of age should be encouraged to hearing evaluation by an electrophysiological test. On the other hand, one of the goals of the WHO’s 2020 program for vision is the increase the public awareness about eye disease and increased levels of care in childhood diseases...
such as amblyopia and refractive errors of eye. But whether these services are performed for the children in our country? And, if applicable, to what extent the target population covered by it?

Methods: Coincident with the national evaluation of school beginners in the city of Shiraz in 2012, a public survey was performed on the children. During the study, parents should be declare whether the senses health of hearing and vision of their child ever been evaluated? However, in case that a problem is confirmed, whether the parents were aware of the existence of this problem in children?

Findings: Statistics clearly show that over 92% of children at least once were assessed for health of vision. But the results were reversed for the hearing exams and nearly 92% of children has been denied of any hearing evaluations and nearly 80% of the children had hearing problems that their diseases were still remain unknown at these ages.

Conclusion: The statistical results indicate acceptable performance for evaluation of vision but these figures are very disappointing in the area of children hearing in the community. Perhaps it is due to the lack of awareness, management, or lack of access to equipment and personnel, as well as sometimes high cost of examinations and related equipment.

Keywords: Hearing, Vision, Screening, Preschool, Children

Improvement of potential hearing in children with bilateral sensorineural hearing loss

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Background: Sensorineural hearing loss is the most sensory deficit in children. Auditory function in these affected children is poor and their potential hearing may be influenced by some factors such as otitis media and cerumen. In this paper, our interest was to seek the effects of overlooked otitis media and cerumen on potential hearing of these children.

Methods: This was a quasi experimental, pretest post-test study that was conducted in 48 bilateral sensorineural hearing impaired children, between 2010-2013. Their age ranged 5-20 years, suffering from the mentioned ear diseases in the better ear, without serious ear symptoms. Modes of interventions were: cerumen removal, medical treatment of otitis media, and both of modes. Two indices including auditory threshold shift by means of (SRT) examined through pure tone audiometer, and scored by dB (HL). The data was analyzed using the nonparametric Fisher’s Exact Test and Kruskal-Wallis.

Findings: Potential hearing improved in about 80% of the cases. Auditory threshold decreased in 50% of them and in all three types of intervention, but with different frequency and different degree. It showed statistically significant relation between frequency, and degree of SRT changes as well as mode of intervention. Hearing ability improved in most of the cases after intervention, but the relation between these improvement and mode of intervention was not statistically significant.

Conclusion: This study showed that potential hearing improved in most of the cases after medical intervention. Although the effect of otitis media treatment on threshold shift was more prominent than cerumen removal, but cerumen had a considerable effect on hearing ability or potential hearing of hearing impaired children.

Keywords: Cerumen, Sensorineural Hearing Loss, Speech Reception Threshold, Otitis Media, Auditory Threshold

The prevalence of overweight and obesity and their relation with stunting among primary school children of Khorramabad city

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Background: Obesity is a medical condition that occurs after excessive accumulation of fat in the body. Stunting is one of the first signs of malnutrition in children, which could have irreversible effects on child development process in the future. This study aimed to investigate the prevalence of overweight, obesity, and stunting, and their relationship with each other and other environmental factors among primary school children in Khorramabad.

Methods: This descriptive-analytic study was conducted on 420 primary school students in Khorramabad in 2012. The samples were selected through cluster sampling. Weight was measured in kilograms and height in centimeters, and the percentiles in terms of age and sex were determined and compared with reference values.

Findings: Of 420 students participating in this study, 54 patients (12.8%) had short stature, 41 (9.7%) were overweight, and 23 (5.4%) were obese. Inverse associations were found between short stature and obesity as well as overweight.

Conclusion: The obtained indicators of child anthropometric indices were indistinguishable from those in other parts of the world, indicating a favorable condition of the anthropometric indicators. However, the findings of this study indicate that achieving the Millennium Development Goals requires further enhancements in living standards and improvements in diet quality.

Keywords: Stunting, Overweight, Obesity, Anthropometric Indices, School Children

The prevalence of consanguineous marriages and its demographic characteristics among couples referred to marriage counseling centers in Hamadan

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Background: Given that the high rate of consanguineous marriage and transferring of defective genes to the next generation in our society, genetic diseases have high prevalence. Given that the congenital malformations have been the most common causes of children death in Hamadan Province in the recent years and considering that the risk of genetic disorders is high in consanguinity marriage, so evaluation of this type of marriage and related
Impact on performance of hearing screening program through prevalence and diagnostic age evaluation in primary school students in Iran

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Background: Lack of a national neonatal screening protocol in Iran, displaces routine hearing screening at school entry. Early diagnosis of hearing loss (HL) leads to early intervention and improvement of developmental skills in children. We aimed to determine diagnostic age and causes of HL at school entry to evaluate this hearing screening performance and its efficacy as a guideline for deciding early diagnosis.

Methods: This cross-sectional study included 3295 children aged 6-7 years from primary schools of mashhad, Iran from 2010 to 2011. Inclusion criteria were students at school entry who were affected by HL. Exclusion criteria were healthy children at school entry. For data obtaining, we evaluated subjects’ medical archives, interviewed with their parents and hygiene teacher and took general physical examination, demographic information, birth history and hearing loss history. Audiologic assessment consisted of otoscopy, tympanometry and audiometry. The affection of students with history of HL diagnosis confirmed by auditory tests again.

Findings: Of total of 3295 students (2282 males and 1011 females) 44(1.33% ) of students had hearing impairment with statistically significantly higher prevalence in male students (p<0.001). In male group, prevalence of hearing loss in urban schools was significantly higher than rural schools, (p<0.02). Making diagnosis by physician was significantly more than by parents (p=0.041) with no diagnosis role of teachers. The common age of HL diagnostic was at school entry, at 6-7 years old (59%) (p<0.001). Use of hearing aids was significant (p<0.001) and (p<0.003) in male and female students respectively.

Conclusion: Higher prevalence of HL in urban schools shows enough perception in their family for diagnosis. The parents, teachers and population should have much greater awareness of HL diagnosis. Screening protocol at school entry in Iran suggested late diagnosis and poor outcome. Therefore, national screening are necessary in newborns, at school entry and every time we suspected of hearing impairments.

Keywords: Hearing Loss, Student, Primary School, Prevalence

An X-autosome translocation with dysmorphic features: a case report

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Background: Balanced X- autosome translocations are associated with a variety of phenotypes depending on X breakpoint position and replication behavior. The present study is an attempt to define the clinical features and diagnosis of a rare event.

Case presentation: We report a female infant who exhibited specific features such as IUGR (Intra uterine growth retardation), low-set ear, microcephaly, hypertelorism and micrognatia. She was born at 38 weeks' gestation, with 45.8cm in length, 2100g in weight, and with head circumference of 31.5cm. Cytogenetic analysis confirmed a karyotype of 45X, t(X:21)(p11.4;q11.2).

Conclusion: To our knowledge, this de novo X-autosome translocation has not been described yet and thus, the localization of its breakpoints may lead to a novel origin of translocation. This comparison of clinical and cytogenetical findings may also provide an opportunity to detect some phenotype/karyotype correlations.

Keywords: Chromosomal abnormality, dysmorphic feature, low-set ear, X-autosome translocation

Unusual presentation of Goltz syndrome with minimal ectodermal involvement in a 3-year-old Iranian girl

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Background: Goltz syndrome (MIM 305600) is a rare genetic disorder characterized by distinctive skin abnormalities and a range of defects affecting the eyes, teeth, limbs, skeletal, urinary, gastrointestinal, cardiovascular and central nervous system. It is inherited in an X-linked dominant mode with lethality in males. One of the main features in this syndrome is the skin changes which are usually present at birth. Ectodermal features include symmetric linear reticulated thin skin, linear hyperpigmentation, ulcerations, telangiectasias, inflammation, hermalike outpuchings of fatty tissue, and papillomas.

Case presentation: Here we report a 3-year-old girl, with asymmetric involvement, greater severity of findings on the right side. She had sparse hair, hyperkeratosis on 2/3 of the right side of the forehead, lacrimal duct stenosis,
hypoplastic alae nasi, hyperkeratosis of the nose, simple ear, narrow auditory canal and hypoplastic tragus., partial cleft of upper lip and pitting on lower lip and slight defect on tongue, hypoplastic nipple, ectrodactyly of hand and foot (on the right side). She has bilateral dysplastic nails on feet, scoliosis, syndactyly of third and 4th toes on the left side. We only had a chest X-ray from our patient which did not show striaed bones. Initially Ectrodactyly-Ectodermal dysplasia was suspected and genetic testing for TP73L did not reveal any pathogenic mutation. We then sequenced PORCN gene and identified c.611T>C (p.Leu204Pro) mutation.

Conclusion: we present a patient with Goltz syndrome with unusual findings. Our patient did not have any of the common ectodermal, skeletal or ocular findings seen in such patients.

Keywords: Goltz Syndrome, Genetic Disorder

Genetic testing reports: implications for pediatricians

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Background: Genetic testing is opening own venue in different branches of medicine, even in our country. There are many applications of testing including newborn screening, diagnostic testing, carrier testing, preimplantation genetic diagnosis, prenatal diagnosis, predictive and presymptomatic testing, pharmacogenomics and forensic and paternity testing. For children, there are some limitations about these tests. Several methods can be used once a person with a genetic disorder decides to proceed with genetic testing; chromosomal analysis and/or DNA testing and/or biochemical genetic tests may be applied to determine the cause of his/her disease. Genetic tests are performed on a sample of blood, hair, skin, amniotic fluid or other biological specimens. The laboratory reports the test results to the physician or genetic counselor. Discussing with a geneticist or genetic counselor could be helpful about the pros and cons of the test. In the other hand, knowing the basics of genetic reports is essential to physicians for communicating with the lab and geneticist and managing the disease.

Keywords: Genetic testing, molecular tests, chromosomal analysis

Studing of growth monitoring chart in 1 to 5 years old children in Tehran Boaali Hospital at year 2013

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Background: The scope of mortality in children under 5 in the developing countries is an important sign of health in the region and monitoring the children growth. Including regular monitoring of weight is a precise and important index for discovery and identification of threatening factors for children health. Monitoring children growth in every society can present a comprehensive plan of growth and health process of the children in that society and help the physicians and health employees in micro and macro policy - making for health of the society.

Methods: The research in descriptive analytical-sectional form through single random sampling has been done with 403 children, 206 girls and 197 boys. Measuring the weight during one year and also filling in a questionnaire for getting more information from every child are among tools for collecting information. The study variables included: parents education, their job, their age, and family relationship between them, number of children in a family, their nutrition habits and the scope of family income.

Conclusion: After analyzing the data, it appeared that significant relation exist between the child weight and the level of education of the father and mother and nutrition habits of the family, it also appeared that weight differences in our children in comparison with the standard weigh curve are more in the middle of the age spectrum and less in the two ends of the age spectrum. This can be a sign of lack of correct and suitable nutrition after ending the breastfeeding period as the main nutrition. It should be mentioned that the growth curve, due to its slight speciality, has the capacity to train mothers for more active participation in their child growth monitoring. This curve is very valuable tool for discovery of high risk children as well as malnutrition before appearing the clinical signs.

Keywords: Growth, Growth Curve, Child

The evaluation of serum level of zinc in children with anorexia refer to shohadaye kargar Hospital

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Background: Loss of appetite is one of the most common nutritional problems in children, especially during the complementary feeding, if it continues it can lead to deviations in patterns of child development and lead to a high incidence of hemodynamic, hematologic, endocrine and bone density disorders. One of the causes of loss of appetite is lack of micronutrients such as zinc which through different ways can lead to growth retardation. According to nutritional differences in different regions and the effects of anorexia in children’s health, determining the serum level of zinc in anorexic children looks essential.

Methods: This was a cross-sectional study. Samples of this study randomly selected from children admitted to the pediatric clinic of Shohada kargar hospital in 2013, and anorexic children (with the informed consent of parents) were enrolled. The questionnaire was structured in advance including demographic data of children that completed and the results added to Inventory. Finally, the data were analyzed.

Findings: This study was conducted on 81 anorexic children including 50 Girls (61.7%) and 31 boys (38.3%). The mean age was 3.6 ± 2.27 years. Children were divided into four age groups; Under 1 year, 1-3 years, 3-5 years and 5-10 years. In this study, an average of serum zinc was 79.24 ± 23.31 μg/dl. Besides, 66 children (81.48%) had normal serum level of zinc (60 μg/dl and higher) and 15 children (18.52%) had abnormal serum zinc level (less than 60 μg/dl).

Conclusion: In this study, the mean serum zinc concentration in the samples was higher than the minimum acceptable level of zinc. Given the low percentage of children with anorexia had abnormal serum zinc level might be justified because of high percentage of families with low socioeconomic level in our survey. Naturally a lot
of social problems involved, so the psychological problems of children with anorexia has become more prominent.

**Keywords:** Anorexia, Zinc, Children

**Reasons of children’s readmission in Motahari Hospital, Urmia, Iran**


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**Background:** In recent years, the rate of avoidable readmission as an indicator of the quality of hospital care is introduced. Readmission has some effects on the cost and quality of hospital care and it imposes additional burden for patients and their families. Also, reduction the costs is a priority for hospitals administrators and patients. The aim of this study was to identify the reasons of readmission for conducting the appropriate interventions.

**Methods:** In this descriptive study, 250 documents of children who were readmitted in 2013 were assessed. A checklist used for data collection. We used descriptive statistics to summarize the readmission rates and the causes.

**Findings:** Out of the 250 children who readmitted, 41.6% were female and 58.4% were male. The mean age was 2.8 years and the mean length of hospital stay was 4.37 days. The average cost of treatment for every patient was 1,361,814 Rails and the average number of readmission was 3.2 times. The most common reasons of readmission included 30.4% unrelated to the first hospitalization, recurrence of the disease (21.2%), follow up (16.8%), discharge without the physician's order (16.4%), treatment failure (4.3%), complications of surgery (4.1%), infection (3.9%), wrong diagnosis (1.8%) and other causes (1.1%).

**Conclusion:** The result of this study shows that readmission increase patients' length of stay as well as additional costs for patients and hospitals, and many causes of readmission are preventable. So, the rate of patient readmission can be minimized by implementing clinical governance guidelines and training of health care workers and patients.

**Keywords:** Children, Readmission, Reasons

**E-professionalism in pediatrics**

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**Background:** Now a days, the internet has created the opportunity to communicate and share information easily, and quickly among millions of people around the world. It also facilitates the same situation for medical students and physicians. They can participate in social networking and other internet opportunities. These professions should consider patient privacy and confidentiality, honesty, appropriate boundaries of the patient and physician relationship, separating personal and professional content online, public and private personae, attention to advertising by corporation and pharmaceutical companies, and trust in the field of medical professionalism. Professionalism breaks in virtual environment can have consequences not only for individuals but also for public trust especially in the field of pediatrics according to the vulnerability of children. Modeling and teaching this new aspect of professionalism or e-professionalism in digital era is very important.

**Conclusion:** Careful attention is needed to define the implications of virtual communication and use of social media on the traditional role of pediatricians as e-professionalism.

**Keywords:** E-Professionalism, Internet, Pediatrics, Professionalism, Social Media
Hematology & Oncology Abstracts

Radionuclide therapy in pediatric malignancies

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The following article aims to provide contemporary information on therapeutic nuclear medicine. Neuroblastoma is the most common malignancies among pediatric malignancies. Pediatric extra cranial solid cancer characterized by meta-iodobenzylguanidine (MBG) avidity in ≈90% of patients. There exists approximately a 30-year experience with I-131-MBG treatment. Neuroendocrine tumors (NETs) are very rare neoplasms in the paediatric population accounting for <1% of all pediatric malignancies. These neoplasms are characterized by the presence of neuroamine uptake mechanisms and/or peptide receptors at the cell membrane. These features constitute the basis of the clinical use of peptide receptor radionuclide therapy (PRRNT) using radiolabeled somatostatin analogues. Patients with chemoresistant CNS acute lymphoblastic leukemia (ALL) require carnosinal radiotherapy which is associated with major toxicities including growth and learning disorders in young children. Radioimmunotherapy with I131 labeled anti-CD10 and anti-CD19 mAbs administered intrathechally is associated with clearing of the cerebrospinal fluid lymphoblasts. Osteosarcoma is the most common primary bone tumor in children usually treated with chemotherapy and surgery. In palliative situations bone seeking radionuclide therapies (strontium-89 [Sr-89], rhenium-186 hydroxyethylene diphosphonate [Rh-186 HEDP] and Samarium-153-ethylene diamine tetramethylene phosphonic acid [Sm-153-EDTMP]) may be offered to patients with painful metastatic osteosarcoma or in case of recurrent bone sites inaccessible to local therapies (surgery, external irradiation). Thyroid cancer is a rare childhood malignancy but is the most frequent tumor of endocrine glands in children and adolescents. Management includes radiodine therapy, but there are some distinct differences in comparison to adult thyroid cancer management.

Keywords: Radionuclide Therapy, Pediatric Malignancies, Nuclear Medicine

The results of treatment of retinoblastoma in children

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Background: Retinoblastoma intraocular malignant tumor, which can occur at any age, but more often diagnosed in children under 2 years. Among all malignant tumours at children RB accounts for about 3% of cases. Overall survival for various literary data ranges from 90 to 95%. Patients with extensive-stage disease, including interstitial RB do not receive adequate treatment. Extremely poor prognosis in patients with extraocular the spread of the RB dictates the necessity of creation of adequate therapy. The aim of the study was to determine the effectiveness of therapy adapted accordingly risk in 17 patients with retinoblastoma.

Methods: The study included 12 primary patients with locally common, extraocular metastatic RB and 5 patients with relapses of the RB after the enucleation of the eyeball. The program of treatment depending on the risk groups include surgery, radiation therapy (RT), or intensive polychemotherapy. In 2013-2014, 17 children (boy-7, girls-10) aged to 6 years were observed.

Findings: After diagnostic study, which included clinical examination, x-ray, ultrasound, CT and MRI of orbit, one-sided retinoblastoma (ORB) was diagnosed in 16 (94%), and bilateral BR (BRB) in 1 (5%) patients. In the high-risk group, RB included 6 children: one of them with BRB and other five with ORB. In the group of medium risk RB included 10 patients, and one patient with standard RB risk. In the group of patients, in 2 patients (11%) retinoblastomas were detected at stage I, in 5 patients (29%) at stage II, in 6 patients (35%) at stage III, and in 4 patients (23%) at stage IV. Multiple metastases were observed in 35% of children in the form of a lesion in the bone marrow, central nervous system, parotid salivary glands, the skull bones or liver. Hardcore metastases were observed in the form of a lesion of the bone marrow. In 5 patients (29%) relapse occured. 14 patients received protocol therapy, and 3 patients refused treatment. On the scheme "chemotherapy (CT) + enucleation + CT" were 6 patients and "enucleation + CT" included 8 patients. 9 patients received Chemotherapy Protocol RBA-2003: two blocks of CT was received by 55% (5) patients, 25% (2) of children received 4 block of chemotherapy and 25% (2) received 5-6 blocks, CT and Protocol JCE/JOE were received by 5 patients: 60% (3) patients had 2 block of chemotherapy, 15% (1) had 3 block of chemotherapy, 15% (1) had 6 blocks of CT. In our work, all patients underwent surgical treatment of residual method enucleation and subperioveal one essentially in orbit. The possibility of disease progression in the central nervous system in patients with retrolental invasion of the optic nerve gave the right to recommend RT included 4 (23%) patients. Causes of disease progression in the central nervous system in 6 patients (35%) included tumor invasion of the optic nerve to the line resection and extraocular extension of tumor to retro-bulbar fibre and extraocular muscles. Among the complications of risk-adapted therapy for children with ORB after primary eye enucleation, it should be noted hematologic toxicity identified in 88.5% of cases, which required the application of an adequate supporting therapy.

Conclusion: Out of 14 patients, which received program chemotherapy, 64% (9) children are in remission. For 3 of them child-treatment continued, 1 child received autologous transplantation of bone marrow, 5 graduated from the PCT; the average duration of the observation was 10 months. 36% (5) of patients experienced the progression of the disease, included one death. Of late-diagnosed with retinoblastoma, 60% of children entered 3-4 stage of the process. High-dose chemotherapy is the only effective method of treatment of metastatic retinoblastoma, allowing to achieve long-term remission. Transplantation of autologous stem cells is effective for the treatment of children in the absence of the primary lesion of the Central nervous system.

Keywords: Retinoblastoma, Treatment, Chemotherapy
The efficacy of recombinant human erythropoietin in treatment of chemotherapy-induced anemia in children

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Background: Anemia is a frequent complaint during intensive chemotherapy in children with cancer. Recombinant human erythropoietin (rHuEPO) treatment can increase hemoglobin levels, decreased transfusion requirements, and improve quality of life in patients with cancer. The aim of this study posed a decrease in blood transfusions and an increase of hemoglobin levels in patients receiving rHuEPO for 12 weeks.

Methods: This was a randomized clinical trial. Participants were 60 patients aged <15 years with anemia and diagnosis of solid tumor between February 2013 and March 2014. 29 Patients received 150 IU/kg/dose rHuEPO subcutaneously, 3 times a week, for 12 weeks. All patients had blood tests every week to determine complete blood count. Number of patients receiving transfusion during the treatment period was compared in the following 12 weeks.

Findings: There was a significant decrease in transfusion requirements in the rHuEPO receiving group. 5 patients (17.2%) in the rHuEPO group needed a blood transfusion, whereas 15 patients (53.6%) needed a transfusion in the control group. Increase in hemoglobin levels, began from the 5th to 6th week, and continued to the end with an ascending trend in the rHuEPO group. There was no significant difference in hemoglobin levels before and after study in CDDP regimens and non-CDDP regimens in the both groups. There was no significant difference in mean hemoglobin before and after the study in the patients receiving G-CSF and non-G-CSF receiving in the participating patients. rHuEPO caused hypertension in one patient at 4th week that stopped the treatment.

Conclusion: rHuEPO (150 IU/kg/day, 3 times a week) is effective in increasing hemoglobin levels and also decreasing blood transfusion requirements in children with anemia following intensive chemotherapy. As a result, this study recommends using of rHuEPO in treatment of chemotherapy-induced anemia in children with solid tumor.

Keywords: Recombinant Human Erythropoietin, Anemia, Chemotherapy

Evaluation of soluble transferrin receptors in children with iron deficiency anemia

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Background: Iron deficiency anemia is one of the most common nutritional deficiencies and public health around the world. The growing children are one of groups that are at high risk for this problem. So early diagnosis and treatment can prevent great side effects. One of the diagnosis means is measuring of soluble transferrin receptor level (sTfR) in serum because level of this receptor increase in result of iron depletion but has no change in anemia of chronic disease or inflammation.

Methods: This was a case control study included 64 children with iron deficiency anemia (IDA) and healthy subjects. The study conducted in 2008-2010 in Children's Medical Center. Hb, MCV, Fe, Ferritin, TIBC and sTfR were measured in both groups.

Findings: Compared to the control group, serum sTfR mean level was significantly higher in children with IDA than control group (\(1.87 \text{ vs.} 1.06 \mu g/ml, P<0.002\)). sTfR showed negative correlations with Hb (r=0.629, p<0.001), MCV (r=0.649, p<0.001) and serum Ferritin (r=0.224, p<0.053), although it was not significant for Ferritin. There was no significant differences between cases and controls in sTfR mean level with regard to gender and age categories.

Conclusion: This study shows sTfR level can be an appropriate biomarker for diagnosis of IDA, particularly in patient with IDA coexisting with inflammation.

Keywords: Soluble Transferrin Receptor, Iron Deficiency Anemia, Children

The use of intravenous immunoglobulin in pediatric oncohematological practice

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Background: Presence of infectious complications in oncohematological patients significantly complicates treatment and worsens the prognosis of the underlying disease. In such situations immunoglobulin (IVIG) is applied. The aim of our study was to examine the efficiency of the use of IVIG in 67 children with oncohematological pathology.

Methods: IVIG for the treatment of purulent-septic complications (neutropenia) used in patients with ALL - 47%(32), AML - 40%(26), histiocytosis 3% - (2), AA - 6%(4), MDS -3% (2) and PID is -1 (1.4%), were administered a single dose of 0.2 g/kg/day, daily 1 per day, duration ranged from 1 to 7 days. The effectiveness of the drug in patients with oncohematological pathology with infectious complications was estimated on the normalization of body temperature, the results of the general analysis of peripheral blood, the general analysis of urine, triple culture of blood.

Findings: Patients receiving of IVIG due to the presence of infectious complications were at various stages of therapy. Patients with acute lymphoblast leukemia received therapy program ALL-BFM-2002; patients with c myeloblastic leukemia (CML) received therapy program AML-BFM-2004; patients with Langerhans LCH-III or aplastic anemia received immunosuppressive therapy. Infectious complications in the study patients included sepsis, which took place in the form of sepsis without localized foci of infection in 15.3%, septicemia in 2.6%, sepsis with septicemia in 10.2%, and septicemia in 78.6%. Localization pyocemia include bacterial carditis in 2.8%, ulceero-necrotic colitis in 8.5%, pneumonia in - 10.8%, and necrotic lesions disease in 20.8%. The extension of infection occurred at the background of deep cytopenia-neutropenia (leukocytes less than 1 × 109, granulocytes less than 0.8h). Microbial landscape of the patients was presented in 78.4% of the cases as gram-positive microorganisms and in 21.6% as gram-negative microorganisms. From blood of patients, gram-positive pathogens was seen in 55%. Among them the most
often organisms was Staphylococcus aureus in 71.2%, Streptococcus Epidermidis in 66.7%, and Streptococcus in 67.5%. Among fungal infection Candida (36.4%), Penicillium (31.8%), Mucor (22.7%), and Aspergillus (9.1%) were seen. Mushrooms more often were detected from the blood (72.6%) than from other sources of infection. The markers to the virus Abstain-Barr (EBV) were seropositive in 48.9% of children, of Herpes virus in 93.3% of children, and of cytomegalovirus (CMV) from 80.2%. The dynamics of the temperature reaction was the following: the initial temperature of 38.4±0.36 Degrees; it was normalized after 1 day from the beginning of introduction of IGW treatment, in 18 patients (26%), within 3 days from the introduction of IGW in 25 patients (39%), within 5-7 days in 17 patients (25%), and was inefficient in 7 (10%) cases among whom were patients with progression of the underlying disease, recurrence ALL (1 case) and AML (2 cases), one patient with myelodysplastic resistant to therapy with manifestations of fungal sepsis not having achieved remission.

**Conclusion:** Use of immunoglobulin in complex therapy of oncohematological patients, in the period of agranulocytosis has allowed to achieve improvement in 92.3% of cases, decreased the incidence and severity of infectious complications, achieved more rapid recovery of hemopoiesis, accelerated normalization of blood, and reducing interruptions in the treatment course.

**Keywords:** Immune globulin, Oncohematology, IV Ig

**Comparing the heart iron deposition status measured by MRI T2\* with the echocardiography findings in major β-thalassemia patients**

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**Background:** Excessive iron store of transfusion causes many complications in major β thalassemia patients. In this study, the relation among iron deposition status reported by the heart MRI T2* technique and ejection fraction and the heart structural changes in major β thalassemia patients is considered.

**Methods:** 52 patients with major β thalassemia, aged 7 to 29 years old, were studied. The heart MRI T2* scan and echocardiography were taken from the patients. Thereafter, the iron deposition status reported by the heart MRI T2* technique was compared with echocardiography results. The collected data were statistically analyzed by SPSS software.

**Findings:** 44.2% of the patients were females and 55.8% were males with averagely 17 years old. The mean of packed cell transfusion was between 150-180 cc/kg. Average of relaxation time and ejection fraction were 26.59 ms and 65%. 43 patients had normal echocardiography and 9 patients had some degrees of the cardiac muscle hypertrophy and dilatation. There is a statistically significant correlation between the ejection fraction and the iron deposition status reported by the heart MRI T2* technique (rs=0.282, P<0.043). But no statistically significant difference is observed in the iron overload reported by the heart MRI T2* technique for the group with normal and abnormal echocardiography group.

**Conclusion:** Due to the relation between the ejection fraction and the iron deposition status reported by the heart MRI T2* technique, the heart iron overload could be evaluated cheaper using ejection fraction level.

**Keywords:** Major Beta-Thalassemia, Iron Overload, Magnetic Resonance Imaging, Echocardiography

**Assessment of propranolol efficacy on pediatric haemangioma**

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**Background:** Hemangiomas are the most common vascular tumor in infancy. Systemic corticosteroids are the first choice of treatment although they have many side effects. Recent studies show Propranolol efficacy on hemangiomas treatment. It has rapid effect and few side effects. These reasons make us to review its effect.

**Methods:** This interventional study was done on 20 children refer to Bu-Ali hospital. Treatment indications were multiple hemangiomas, organ malfunction and enlarging hemangioma. Treatment on those was started by 1mg/kg/day and increased to 3 mg/kg/day and continued for 6 month. After treatment completion 6 month follow up was done.

**Findings:** Patients mean age was 23.15 ± 11.24 months. 65% of them were female and 35% was male. Mean size of lesion was 4.85 ± 3.26 Cm. 70% patients had acceptable respond (more than 50% decrease in size) and 30% had partial respond (less than 50%). This effect was similar to corticosteroid effect (about 84%), but with less side effects. In this study there was significant relationship between mean lesion size decrease and propranolol usage, but no significant relationship between size of lesion, age, sex and regression.

**Conclusion:** This study shows that propranolol has acceptable effect on hemangiomas regression and could be suggested as first choice of hemangiomas treatment.

**Keywords:** Hemangioma, Propranolol, Pediatric Clinic

**Aquired hemophilia**

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Acquired Hemophilia is a rare but serious bleeding disorder characterized by the development of autoantibodies (inhibitors) against plasma coagulation factors, mostly commonly FVIII. It classically presents with the sudden onset of bleeding symptoms in a patient with no past or family history of bleeding disorder. It is thought to be exceedingly rare in the pediatric population with an estimated annual incidence to be 0.045 per million. Pediatric acquired hemophilia has been described in association with autoimmune conditions, infection and antibiotics, most commonly penicillin- like antibiotics. Unlike classical hemophilia where hemarthrosis is the characteristic bleeding manifestation, most patients with acquired hemophilia present with bleeding into the skin, subcutaneous tissue and muscles, hematuria, hematemesis or melena and postoperative bleeding. Severe subcutaneous bleedings following venipuncture and intramuscular injections have been described.

**Keywords:** Acquired Hemophilia, Acquired Autoantibody, Eradication of Inhibitor
Distribution of ABO and Rhesus blood group system in Iranian glucose-6-phosphate dehydrogenase deficient newborns

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Background: Glucose-6-phosphate dehydrogenase (G6PD) deficiency is an inherited disorder which is common in Iran and may cause neonatal jaundice. As combination of G6PD and ABO or Rhesus incompatibility leads to higher risk, we aimed to evaluate the distribution of ABO and Rhesus blood groups in G6PD deficient newborns.

Methods: This cohort study was conducted on 150 icteric newborns who admitted to the NICU of educational hospitals in Azad University in North-East state of Iran, Mashhad. G6PD deficiency was evaluated and case and control groups were considered of 50 icteric newborns with G6PD deficiency and 100 icteric newborns with normal levels of enzyme respectively. Distribution of ABO and Rhesus blood group was considered in G6PD deficient newborns and compared with newborns with normal levels of the enzyme. The prevalence of hemolysis was compared in two groups as well.

Findings: Prevalence of hemolysis was 22% in case group and 19% in controls. There was no significant relationship between G6PD deficiency and hemolysis. There was no significant relationship between distribution of ABO and Rhesus blood groups in G6PD deficient newborns.

Conclusion: There was no significant relationship between distribution of ABO and Rhesus blood groups in G6PD deficient newborns in Iran unlike other similar studies.

Keywords: ABO Blood Group, Rhesus Blood Group, G6PD Deficiency, Neonatal Hyperbilirubinemia

Distribution of HBB gene mutations in Iran: a comprehensive review

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Beta thalassemia caused by HBB gene mutations is one of the most common single gene disorders worldwide. Nearly 80 to 90 million beta thalassemia minors are living and 60-70 thousands affected infants are born annually worldwide. A comprehensive search on English databases was performed fulfilling all English and Persian papers about mutation detection and frequency of beta thalassemia. The search was done using these keywords ‘gene mutation’ and ‘beta globin’ and/or ‘beta thalassemia’ and ‘Iran’. All papers in English and Persian reporting mutation frequency of beta thalassemia patients and premarital couples were selected to analyze the frequency of mutations in different regions and various ethnicities living in Iran. Twenty common mutations were selected for more analysis; the frequency of the mutations was searched among neighboring countries. Mutations of 2104 thalassemics were identified. About 90 beta globin mutations including 30 frameshift, 25 splice and intronic, 15 UTR and regulatory, 14 missense and 6 nonsense mutations is reported from Iranian groups. The following mutations IVS1-5 (G>C), IVS II-1 (G>A), IVS 1-10(G>A), CD36/37 (-T), Fsc 8/9, IVS I-1 (G>A), IVS I-25 bp, Fsc8 (-AA), Codon30 G>C (Monroe) and FSC44(-C) are responsible for about 80% of all mutant alleles throughout Iran. These mutations, even, are the common among neighboring of Iran so that responsible for 60 to 90% of mutations in these countries. Genetics of beta thalassemia mutations in Iran is extensively heterogeneous that neighboring countries. Some of common mutations have been arisen historically from Iran and moved to other populations due to population migrations and genetic drift affects them so that frequencies of some of them have been increased in small populations.

Keywords: Beta-thalassemia, HBB Mutations, Iran

Prevalence of anemia in Abhar city children and its relation with breast feeding and iron supplement intake

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Background: Anemia is a global, public health problem affecting both developing and developed countries, especially in childhood. Anemia is the result of wide variety of causes, but the most important cause is iron deficiency; so that iron deficiency anemia and anemia are often used synonymously and the prevalence of anemia has often been used as a proxy of anemia.

Methods: Over period of 1 year, about 200 samples were taken from the children in Abhar Omid Hospital. These data included level of hemoglobin, history of breast feeding, and iron supplement intake. Normal hemoglobin cut of point considered 11 mg/dl.

Findings: In statistical analysis with SPSS, prevalence of anemia in age group 6-24 month was 45% and in age group 2-12 year was 17%. This study revealed that, there is no significant relationship between anemia and breast feeding or iron supplement intake.

Keywords: Anemia, Breast Milk, Iron Drop

Comparision of therapeutic effect of osveral & desferal in patients with thalassemia (bahonar hospital in karaj 2012-2013)

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Background: This study was performed to compare the therapeutic effect of osveral and desferal in patients with major thalassemia attending to bahonar hospital of karaj 2012-2013.

Methods: In this randomized clinical trial 30 patients with major thalassemia attending to bahonar hospital of karaj in 2012-2013 were enrolled and underwent desferal or osferal group and the ferritin level was measured at baseline and also 1,3,6,9,12 months after drug prescription.

Findings: In this study, the mean ferritin level was alike between two groups except for third month follow up that was significantly higher in osveral group (p<0.03).

Conclusion: Totally, according to the obtained results in this study and comaparison with other studies it may be concluded that therapeutic effect of osveral and desferal in patients with major thalassemia are similar.

Keywords: Major Thalassemia, Osveral, Desferal, Treatment
Iranian national program for prevention of beta-thalassemia major: comparing genetics counseling result between definite carrier and final suspected couples

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Background: In the Iranian national program for prevention of beta-thalassemia, 1,000,000 couples who plan for marriage are on average screened annually. Results of the screening of all couples are evaluated by genetic counselors and based on national algorithm for the program, couples who are definitely carrier or highly suspicious of being so, are detected and take part in the specific sessions of genetic counseling.

Methods: The data of specific genetic counseling with 6028987 couples were evaluated to find out the "percentage of couples who withdraw the sessions and who choose not to marry as a estimate of compliance of definite carrier couples and highly suspicious of being carrier couples. The results were examined with Man-Whitney test.

Findings: According to the hematologic indices, 36% couples have been identified at first stage as suspected of being carrier of beta thalassemia (n= 2189245). Among them, 4.19% were found as definite carriers and highly suspicious of being carrier couples (that equal to1.52% of all screened couples). The proportion of definite carrier and highly suspicious couples were respectively 26 and 126 per 10000 screened couples (SE=1.08; CV=0.10 and SE=6.30; CV=0.12). The percentage of not-to-marry couples by definite carrier and highly suspicious couples were respectively 19 and 7 which differed significantly in these two groups (P<0.001). The study also showed that the percentage of withdrawal in definite carrier couples is 1.9 greater than highly suspicious couples (P<0.05).

Conclusion: Higher percentage of withdrawal in definite carrier couples can show that the higher level of certainty of being at risk of the disease increase compliance of the couples to genetic counseling. This finding can be examined by evaluation of the relevant genetic diagnostic test results of the couples.

Keywords: Beta Thalassemia, Genetics Counseling, Carrier Couples, Compliance
**Immunology, Allergy and Immunodeficiencies**

The role of infectious agents in acute asthma in children

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**Background:** The aim of the study was assessing the role of infectious agents as a cause of asthma in children and then evaluating this role in both types of asthma (allergic or no allergic).

**Methods:** During the period from 01/12/2006 to 01/06/2007, we did a prospective research for the presence of infectious agents in children hospitalized for asthma in the pediatric department in the hospital of Mantes La Jolie, on the outskirts of Paris, all of these cases had been admitted in the emergency department, where they received three doses of aerosol of salbutamol which were not sufficient to allow the shipment to the house; we also studied the allergic situation of these patients according to specific criteria to classify the child in allergic or no allergic asthma.

**Findings:** During this period 44 children “in accordance” with the terms of our study had been hospitalized with a mean age of 36.4 months. 10 of them were infected by Mycoplasma pneumoniae and 8 by respiratory syncytial virus (RSV); the distribution of infectious agents in both types of asthma was similar.

**Conclusion:** This study shows that M. pneumoniae is the main infectious cause of an asthma attack, followed by RSV without any differences according to the allergic situation.

**Keywords:** Asthma, mycoplasma pneumoniae, Respiratory Syncytial Virus

Effect of education on spirometry in the patients referred to Childrens’ Medical Center between 2008-2009

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**Background:** Asthma is a common chronic disease that can cause disability in patients. Studies have shown that patient education plays an important role in asthma control and improving disease. The aim of this study was to investigate the effect of patient education on spirometry.

**Methods:** In this interventional study, 6-14 years old children with asthma referred to asthma and allergy clinic of Children’s Medical Center between 2008-2009 were studied. Patients were divided into two groups; Case and Control, and each group was treated with common and standard therapy of asthma. For case group besides general education, special education was conducted. For each patient two pulmonary function tests(spirometry) on first visit and one year after that have been done. The collected data were analyzed with SPSS software version 18.

**Findings:** A total of 104 patients were studied. The mean age of patients in case group was 8.01±1.62 and in control group was 8.42±2.08 years. 76.92% of patients had a history of referring to emergency department. The mean changes in FEV1/FVC ratio before training was -3.11±10.55 and after training was 1.52±7.28 that difference was statistically significant (P<0.018). In case group, mean FEV1/FVC ratio before training was 102.97±4.45 and after training was 104.07±9.76 and there was no significant difference (P<0.499). In control group, the mean changes in FEV1/FVC ratio at baseline was 102.85±8.97 and the mean at the end of study was 102.76±11.35, which was not significantly different (P<0.969). In addition, the mean changes in FEV1/FVC ratio at baseline was 0.59±4.37 and the mean at the end of the study was 2.26±13.55 and there was no significant difference (P<0.468).

**Conclusion:** Pulmonary status did not change much during the study period and the effect of education on patient behavior and lifestyle modification resulted in the prevention of severe attacks and sudden and short-term risky exacerbation episodes. However, a significant reduction in FEV1/FVC ratio changes after training is also indicating the effect of training on this index. Therefore, a training program for asthmatic patients and their families is recommended.

**Keywords:** Education, Asthma, Children, Spirometry

An etiologic survey of patients with chronic urticaria in Kurdistan provience

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**Background:** Urticaria, also known as hives, is a very common disorder and is thought to afflict up to 20% of the population at some point in time. Chronic urticaria (CU) generally is defined by the presence of urticaria on most days of the week, for a period of 6 weeks or longer. Approximately 40% of patients with CU have accompanying episodes of angioedema or deeper swelling of dermal or mucosal tissues, whereas 10% have angioedema as their main manifestation.

**Methods:** We gathered all data of patients with CU. Serum autologous skin test was performed for patients without any specific etiologies.

**Findings:** 185 Kurd patients with chronic urticarial, 80 male (43.2%) and 105 female (56.8%), who were referred to Kurdistan asthma and allergy clinic were evaluated for causes of CU. The mean (SD) age of the patients was 36.5 (12.8) years. The mean (SD) serum IgE level was 137.5 (151.8). 11 patients (5.9%) had infectious etiologies (Helicobacter pylori, GI fungal infections, chronic viral infections, ...). 7 patients (3.7%) had reumatologic diseases and physical triggers were defined in 34 patients (18.3%). 71.1% of patients did not have any etiologies which were classified as idiopathic or spontaneous chronic urticarial. Serum autologous test was performed for patients and was positive on 76 person (41%).

**Conclusion:** Approximately 20% of patients with CU have a reproducible physical trigger for their skin lesions; this form of the disorder is termed physical urticaria. In the remaining 80% of cases, no external allergic cause or contributing disease process can be identified; accordingly, the condition is termed chronic idiopathic urticaria (CIU). Some guidelines and experts identify a subset of patients
with CIU on the basis of serologic evidence of a presumed autoimmune etiology (observed in 30% to 40% of these patients) and call the condition chronic autoimmune urticaria (CAU).

**Keywords:** Chronic Urticaria, Serum Autologous Test, IgE Serum Level, Chronic Autoimmune Urticaria

### Pneumonia due to Acinetobacter Lwoffii in a patient with X-linked Agammaglobulinemia

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Acinetobacter Lwoffii is a nonfermentative aerobic Gram-negative bacillus. It is an opportunistic pathogen in immunocompromised patients and has a cause of nosocomial infections like septicemia, pneumonia, meningitis, urinary tract infections, skin and wound infections. Herein we described a case of pulmonary infection due to Acinetobacter Lwoffii in a patient with X-linked agammaglobulinemia.

**Case presentation:** A 5-year-old boy admitted to hospital with tachypnea, fever and cough of 3 day’s duration. He was the a known case of X-linked agammaglobulinemia from infancy. CXR showed infiltration in near heart border in the left lung. Ceftriaxone was started but response to therapy was poor. The culture of sputum obtained during bronchoscopy revealed growth of Acinetobacter Lwoffii. A. Lwoffii must be considered in severe infections especially in immunocompromised patients.

**Keywords:** X-Linked Agammaglobulinemia; Immuno-deficiency; Acinetobacter Lwoffii

### Late-onset familial hemophagocytic lymphohistiocytosis with STXBP2 mutations

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**Background:** Hemophagocytic lymphohistiocytosis (HLH) is a rare condition, clinically characterized by fever, hepatosplenomegaly, cytopenia, and widespread accumulation of lymphocytes and histiocytes, sometimes with hemophagocytosis, which can either occur sporadically or as part of a familial syndrome (primary HLH). Familial hemophagocytic lymphohistiocytosis (FHL) is a genetically heterogeneous inflammatory syndrome, caused by an uncontrolled proliferation and activation of T-lymphocytes, NK-cells, and macrophages that infiltrate multiple organs, coagulation abnormalities, and inflammatory CNS disease.

**Case presentation:** Herein, a male patient is presented who was well until 7 years old, when he was hospitalized because of fever and jaundice after 10 days of watery diarrhea. He had ascites and hepatosplenomegaly. Laboratory data revealed high bilirubin and liver enzymes. Investigations for viral hepatitis, Wilson were inconclusive. Bone marrow biopsy was normal, but liver biopsy revealed marked interface activity and bridging necrosis and no report of hemophagocytosis. Hence the diagnosis of autoimmune hepatitis was made for him and Azathioprine plus prednisolon were started. The patient was well for 6 months until he was admitted to the intensive care unit, because of pneumonia. One year later, he developed sudden onset of ataxia associated with diplopia. Physical examination revealed an atactic speech, nystagmus when gazing to the right side, asymmetric plantar reflexes, and abnormal cerebellum tests. MRI imaging revealed several ring enhancement lesions. fibrinogen, ferritin and triglyceride were normal. Sterotaxic biopsy showed mixed inflammatory cells within a loose necrotic fibrillary background. Immunohistochemical staining was compatible with a reactive inflammatory process. Soluble IL2 receptor was high and CD4+ and CD8+ T cells were highly activated. An NK cell activity assay showed absent degranulation after stimulation with target cells and no recovery of degranulation after stimulation with IL-2.

**Conclusion:** This case highlights that patients with FHL, especially late-onset ones should be visited in a multidisciplinary system by expert immunologists, hematologists, and neurologists.

**Keywords:** Familial Hemophagocytic Lymphohistiocytosis, STXBP2, Brain Lesions, Autoimmune Hepatitis

### Immunometabolism in obesity and its clinical relevance in paediatrics: when immune system meets mitochondria and cellular metabolism

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The human fatty tissue is not just a passive organ to save the excessive energy, but acts as a store for immune cells such as macrophages, neutrophils, T and B cells and produces as an active endocrine unit the biologically active substances, called Adipokine. The human fatty tissue contributes to the innate immune system. The adipocytes are able to detect foreign antigens via specific receptors on their cell surface and release proinflammatory cytokines of mitochondrial dysfunction, metabolic diseases and insulin resistance. This inflammatory state is reflected in increased circulating levels of pro-inflammatory cytokines and acute phase proteins like tumor necrosis factor α (TNF α), interleukin 6 (IL 6), C-reactive protein (CRP), plasminogen activator inhibitor-1 (PAI-1), vascular cell adhesion molecule-1 (VCAM-1), p-selectin, serum amyloid A3, fibrinogen and angiotsinsinogen. Obesity shares with most chronic diseases the presence of a permanent local and systemic inflammation which leads to the development of mitochondrial dysfunction, metabolic diseases and insulin resistance. This inflammatory state is reflected in increased circulating levels of pro-inflammatory cytokines, and it occurs not only in adults but also in adolescents and children. The chronic inflammatory response has its origin in the links existing between the adipose tissue (AT) and the immune system. Under physiological conditions, the proportion of CD14 and CD31 positive macrophages in stroma cells of fatty tissue amounts to 5-10%. In obese patients the amount of macrophages increases up to 60% in adipose tissue (AT). The number of macrophages in AT correlates positively with BMI and the size of adipocytes. Macrophages are prime players in the initiation of a chronic inflammatory state in obesity. In response to increases in free fatty acid release from obese adipose depots, M1-polarized macrophages infiltrate adipose tissues. These M1
macrophages trigger inflammatory signaling and stress responses within cells that signal through JNK or IKK β pathways, leading to insulin resistance. If overnutrition persists, mechanisms such as M2 macrophages and PPAR signaling that counteract inflammation are suppressed. Macrophages are the major origin for production of TNF-alpha and IL-6 which lead to the insulin resistance. The expression and secretion of TNF-alpha correlate with body weight, especially in visceral AT, compared to subcutaneous fatty tissue. TNF-alpha disturbs insulin signal and leads to insulin resistance by reduction of phosphorylation of insulin receptor substrate-1 (IRS-1) and disturbance of synthesis and translocation of glucose transporters type 4(GLUT-4). On the other hand the increased glucose levels and oxidated low density lipoprotein (LDL) activate the phagocytes and lead to local tissue damages by production and secretion of inflammatory and cytotoxic metabolites. In mouse model, it could be shown a positive correlation between increase of weight and enhancement of expression of mRNA transcript inflammatory genes in fatty tissue. The weight reduction leads significantly to decrease of systemic circulating inflammatory molecules in serum. Recently it could be demonstrated that RBP4, a retinol transporter, is upregulated in insulin resistance and contributes to increased diabetes risk. RBP4 activates macrophage and CD4 T cell. RBP4-overexpressing mice (RBP4-Ox) are insulin resistant and glucose intolerant and have increased AT macrophage and CD4 T cell infiltration. In RBP4-Ox, AT CD206+ macrophages express proinflammatory markers and activate CD4 T cells while maintaining alternatively activated macrophage markers. These effects result from direct activation of AT antigen-presenting cells (APCs) by RBP4 through a JNK-dependent pathway. Transfer of RBP4-activated APCs into normal mice is sufficient to induce AT inflammation, insulin resistance, and glucose intolerance. Thus, RBP4 causes insulin resistance, at least partly, by activating AT antigen-presenting cells.

Transfer of RBP4 through a JNK-dependent pathway. This article will try to shed a light on the significant importance of research in the field of immune system and metabolism. Research is necessary to develop therapies that prevent or cure diabetes.

Keywords: Metabolism, Adipose Tissue, Inflammation, Diabetes, Insulin Resistance

Approach to patients with phagocyte defects

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Defects of neutrophil function and/or differentiation, defects of motility, defects of respiratory burst, and Mendelian susceptibility to mycobacterial diseases could be classified as main diseases in the category of phagocytes defects. Severe congenital neutropenia, cyclic neutropenia, glycogen storage disease type 1b, p14 deficiency, Barth syndrome, Cohen syndrome, and poikilo daylight deficiency are primary immunodeficiency diseases with neutrophil function/differentiation defects. Leukocyte adhesion deficiency (LAD types I-II), Rac2 deficiency, β-actin deficiency, localized juvenile periodontitis, Papillon–Lefèvre syndrome, specific granule deficiency, and Shwachman–Diamond syndrome are classified in group of motility defects. Chronic granulomatous disease (CYBB, CYBA, NCF1, NCF2, NCF4) is the prototype of defects of respiratory burst. Mendelian susceptibility to mycobacterial diseases predispose individuals to mycobacterium. Mutations in several gene loci have been detected for MSMD, including IL-12RB1, IFNGR1, IFNGR2, IL-12B, STAT1, CYBB, IRF8, and ISG15. GATA2 deficiency, pulmonary alveolar proteinosis along with autosomal recessive form of IFR8 deficiency are other diseases that have been classified as phagocytes defects.

Keywords: Phagocyte Defects, Mycobacterial Infection, Chronic Granulomatous Disease, Leukocyte Adhesion Deficiency

Idiopathic CD4+ T lymphocytopenia

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Idiopathic CD4+ T lymphocytopenia (ICL) is a rare immune deficiency with heterogeneous clinical manifestations. This syndrome first described in 1992 and defined as absolute CD4+T-lymphocyte count <300/mm³ or less than 20% of total lymphocytes that confirmed at least twice during a period of 1 to 3 months in the absence of HIV-1 infection disease or any other cause of immunodeficiency. Patients with ICL often presents with opportunistic infections, malignancies, or autoimmune disorders and the major risk ICL is unexpected infections, including cryptococcus, atypical mycobacterial and pneumocystis pneumonia (PCP). In some patients also additional immunologic defects including CD8+ lymphocytopenia and low immunoglobulin levels was occur. This disease was seen in both children and adults and two genders. At present the etiology of disease is unknown and also it does not appear to be caused by a transmissible agent, such as a virus, but it is widely believed that there is more than one cause. Although in general, prognosis of disease is depends on absolute number of CD4, but in contrast to the CD4+ cell depletion caused by HIV, patients with idiopathic CD4 lymphocytopenia have a good prognosis. The decline in CD4+ T-cells in patients with ICL is generally slower than that seen in HIV-infected patients and in some cases this condition may also resolve on its own. We present 3 cases of ICL with different prognosis.

Keywords: T lymphocytopenia, CD4, Immunodeficiency

Nonhodgkins Lymphoma in a patient with Leukocyte Adhesion Deficiency Syndrome

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Background: Leukocyte adhesion deficiency syndrome type 1 (LAD-1) is a rare autosomal recessive primary immunodeficiency disorder of neutrophil phagocytic function characterized by the deficiency of one or several surface integrins which altered adhesion and cause recurrent infection. Some reports shows association of LADS with malignancy.
**Case presentation:** We describe a case of a 7-day-old boy who presented with an omphalitis, sepsis, icter and erythematous rashes and characteristic history of recurrent infections, marked leukocytosis and delayed separation of umbilical cord. The diagnosis is based primarily on flow cytometric analysis of neutrophils for the surface expression of CD11, CD18 and CD15s. He developed lymphadenopathy and abdominal mass with diagnosis Nonhodgkins lymphoma (NHL). Our patient represents the first clinically and histopathologically documented association between LADS and NHL. Data could support to the role of tumor genesis of β2 integrins in the human.

**Keywords:** B-cell non-Hodgkin’s lymphoma, Leukocyte adhesion defect, surface integrins, recurrent infections

**Vitamin D deficiency in chronic idiopathic urticaria**

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**Background:** Chronic urticaria is one of the most common skin diseases, characterized by as a chronic cutaneous condition which severely debilitates patients in several aspects of their everyday life. CU patients suffer from physical, social, and psychological morbidities resulting in a low quality of life. Vitamin D is known to exert several actions in the immune system and to influence function and differentiation of mast cells, central role players in the pathogenesis of chronic idiopathic urticaria. Recently, an increasing body of literature showed paradoxical relationships between vitamin D and allergic diseases like food allergy, rhinosinusitis, recurrent wheeze, asthma, atopic dermatitis and eczema. This study was performed to evaluate the relationship between vitamin D levels and susceptibility to chronic idiopathic urticaria.

**Methods:** One hundred and fourteen patients with chronic idiopathic urticaria were recruited in this study along with one hundred and eighty seven sex matched and age matched healthy volunteers as the control group. For each patient, urticarial activity score was calculated and autologous serum skin test was done. Vitamin D metabolic statue was measured as 25 hydroxyvitamin D using enzyme immunoassay method.

**Findings:** Patients with chronic idiopathic urticaria significantly had lower levels of vitamin D (p<0.01). Vitamin D deficiency was significantly associated with increased susceptibility to chronic idiopathic urticaria (p<0.01). There was a significant positive correlation between vitamin D levels and urticarial activity score (r=0.2, p<0.05).

**Conclusion:** This study showed patients with chronic idiopathic urticaria had reduced levels of vitamin D, while vitamin D deficiency could increase susceptibility to chronic idiopathic urticarial.

**Keywords:** Vitamin D Deficiency, Idiopathic Urticarial, Chronic Urticaria

**Phenotyping and follow up of forty seven Iranian common variable immune deficiency patients**


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Common variable immune deficiency (CVID) is a heterogeneous syndrome with infective, autoimmune and malignant manifestations. This study describes retrospectively the phenotyping and follow-up of the CVID patients in the allergy and clinical immunology department of Rasol E Akram Hospital of Iran University of Medical Sciences in Tehran until January 2014. The study included forty seven CVID patients with mean age at onset of symptoms and diagnosis of 11.2 and 20.2 years respectively. Phenotyping of our patients was: only infection (62%), cytopenia (26%) and PLI (19 %) and 94% of cases had only one phenotype. We did not find a significant relation between the clinical phenotypes and immunologic or demographic data. Rate of parental consanguinity in our cases was 47%. Parental consanguinity was related to lower age at onset, lower age at diagnosis and higher baseline IgG levels. Patients with malignancy and autoimmune had significantly higher age at onset. Our patients were followed for 6.9 years and the mortality rate during this time was 6%.

**Keywords:** Common Variable Immune Deficiency, CVID Phenotype, Iran.

**APRIL gene polymorphism and serum sAPRIL and Th1/Th2 cytokines level in children with SLE**

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**Background:** Systemic Lupus Erythematosus (SLE) is a prototypic systemic autoimmune disease diagnosed by the generation of autoantibodies against nuclear and cytoplasmic components that are originated from autoreactive B cells. The etiology of SLE includes immunological disturbances, genetic and environmental factors. Among these, APRIL and Th1/Th2 cytokines (IFN-γ, IL-4) have roles in the stimulation and antibody production in B cells. These cytokines were hypothesized to be associated with SLE. Therefore, the aim of this study was to evaluate the hypothesis by assessment of APRIL.
polymorphism particularly rs11552708 in addition to the serum levels of APRIL, IFN-γ and IL-4 in Iranian children with SLE.

Methods: A single nucleotide polymorphisms (SNP) for rs11552708, of APRIL gene were analyzed by Real-time PCR in 60 SLE Iranian children and 64 healthy controls. DNA samples of patients and healthy controls were extracted from peripheral blood leukocytes by phenol-chloroform. Serum samples obtained from 45 children with SLE and 45 healthy controls were assessed by Enzyme-linked immunosorbent assay (ELISA).

Findings: The G/G genotype (odds ratio (OR) 0.67, 95% confidence interval (CI) 0.25-2.56; P=0.89) frequencies of polymorphism at codon 67 (67G) not differ significantly in SLE patients compared with healthy controls. The serum APRIL levels in SLE patients (mean +/- SD=29.27 ng/ml +/- 20.77, range from 0 to 55.33 ng/ml) were significantly higher than in healthy controls (P=0.02). No significant differences in the serum levels of IFN-γ and IL-4 were observed between children with SLE and healthy controls.

Conclusion: Our results demonstrated that rs11552708 of the APRIL gene is not associated with SLE susceptibility in Iranian children. Likewise, these findings suggest that APRIL antagonists could be a potential therapeutic target to control of SLE in children.

Keywords: Systemic Lupus Erythematosus, SLE; APRIL

Interleukin-1 gene cluster polymorphisms in inflammatory bowel disease

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Background: Crohn’s disease (CD) and ulcerative colitis (UC) are two inflammatory bowel diseases (IBD) in which host-microbiota dysbiosis is expected in a genetically susceptible host. There are conflicting results on role of single nucleotide polymorphisms (SNPs) in IL-1 family members in IBD.

Methods: In this study, SNPs of IL-1 family were investigated in 74 patients with IBD (40 CD and 35 UC), using PCR-SSP method.

Findings: IL-1β -511 CC genotype was significantly less present in UC compared to controls, while IL-1RAMspa-II1100 CC was significantly associated with both CD and UC. IL-1u -898 TT genotype was more frequently associated with extraintestinal manifestations. A significant association was observed between IL-1β +3962 TT genotype and the disease activity in IBD. IL-1RA Mspa-II1100 CC significantly less frequent in CD patients who need immunosuppressive therapy. IL-1RA Mspa-II1100 CT was associated with earlier age of onset in IBD, while TT genotype was associated with higher age of onset in IBD.

Conclusion: IL-1 SNPs seem to be associated with IBD and could affect the disease severity as well.

Keywords: Inflammatory Bowel Diseases, Crohn’s Disease, Ulcerative Colitis, Interleukin-1

Association of Interleukin 4 Single Nucleotide Polymorphisms with Febrile Seizures

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Background: Interleukin-4 (IL-4) plays a critical role in forming the nature of immune responses. As of its importance in inhibiting the production of proinflammatory cytokines by monocytes and activated T cells, the IL-4 gene polymorphisms were investigated in a group of patients with febrile seizure (FS).

Methods: Ninety patients with febrile seizure were enrolled in this study and compared with 140 controls. The allele and genotype frequency of 3 single nucleotide polymorphisms (SNPs) within the IL-4 gene were determined.

Findings: The frequency of the IL-4 -590/C allele in the patient group was significantly higher than in the control group (p<0.001). The most frequent genotypes in patients with febrile seizure were IL-4 (-33) CC (p<0.01), IL-4 (-1098) GT (p<0.046), IL-4 (-590) CC (p<0.001) and IL-4(-33) TT (p<0.02). The frequency of the following genotypes was significantly lower in patients compared to controls: IL-4 (-590) TC (p<0.01) and IL-4 (-33) TC (p<0.001). The most frequent IL-4 haplotypes in the patient group, which were significantly higher than in the control group, were TCC (p<0.00), TCT (p<0.02), and GTC (p<0.02) haplotypes. In contrast, the frequencies of the following haplotypes in the patient group were significantly lower than the controls: GCC (p<0.01), TTT (p<0.009), and TTC (p<0.007).

Conclusion: Certain alleles, genotypes, and haplotypes in IL-4 gene were overrepresented in patients with febrile seizure, which possibly could predispose individuals to this disease.

Keywords: Febrile seizure, gene polymorphisms, interleukin-4, interleukin-4 receptor alpha

Diagnostic approach to primary immunodeficiency disorders

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Primary immunodeficiency disorders (PIDs) are a heterogeneous group of inherited disorders that affect different components of the immune system. During last years, advances in molecular genetics and immunology have resulted in the identification of a growing number of genes causing primary immunodeficiencies (PIDs) in human subjects. Since 1952, more than 220 different PID disorders which have been described. Despite progress in discovery of PIDs and understanding of pathogenesis of these disorders over the last 20 years, many patients remain undiagnosed. Recognition of PIDs may be difficult as
infections are common in young children in particular. Identifying different clinical manifestations of PID is the first most important step of diagnosis of PID. Clues to the diagnosis of PID may be found in history, physical examination. Different diagnostic tools have been developed for diagnosis of primary immunodeficiencies. The investigations are largely guided by the clinical presentation of the patient, the suspected immune defect and the results of initial laboratory evaluation. This review will focus on essential and necessary laboratory approach for diagnosis of suspected cases of PID.

**Keywords:** Primary Immunodeficiency, Diagnosis

**Successful Vp16 desensitization**

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**Background:** Adverse drug reactions are frequent and occur in 10% to 20% of hospitalized patients and approximately 7% of the general population. Reactions to drugs can be divided into two main categories; predictable (type A) and unpredictable drug reactions (type B). The latter one consists only 20% of drug reactions. Drugs are one of the most frequent causes of anaphylaxis, ranging between 8% and 62%. The clinical manifestations of anaphylaxis can involve any organ systems, mainly cutaneous, respiratory, GI, and cardiovascular systems.

**Case presentation:** A 10 years old girl was treating because of Anablastic T cell lymphoma. During 4th chemotherapy course, she developed urticaria, edema of lips and tongue, dyspnea, and cough (anaphylaxis reaction) while receiving VP16 injection. The medication was discontinued immediately and anaphylaxis treated. She was consulted to our department to receive VP16 in the rest of her chemotherapy courses. As it was necessary in the treatment courses, we decided to desensitize her. Skin tests for VP16 were done according to drug provocation tests protocols 1 month after the reaction. The intradermal test was positive in 1/1000 dilution. Then the desensitization process was started in accordance to the 12 step desensitization protocol. Finally she can tolerate all the VP16 during 6 to 7 hours. Rapid desensitization protocols are available to patients who present with IgE and non-IgE-dependent hypersensitivity to drugs. Temporary toleration which is achieved in hours can be maintained if the drug is administered at regular intervals.

**Keywords:** Drug Reaction, Vp16, Desensitization

**New protocol for Cow’s Milk desensitization based on SPT**

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Cow’s milk Allergy is one of the most common food allergies. It could be IgE mediated or Non-IgE mediates. As 25% of IgE mediated Food allergies persist up to second decade, avoidance is somehow impossible for most of these patients. Fourteen children over 3 year with history of anaphylaxis to cow’s milk, positive skin prick test, and positive OFC were recruited. SPT with Milk extract of Greer, full dilution of milk, 1/10, 1/25, 1/50, 1/100, 1/200, and or 1/500, depends on patient’s reaction was performed for all the patients. The dilution of milk which induced 3-5 mm of wheal was selected as a stating dilution. Then it started form 2 drop of the dilution and increased according the protocol. The dose was increased weekly to achieve an intake of 120 mL in approximately 4 months. All doses were administered under medical supervision in a clinical setting. After receiving the dose, children were carefully assessed for positive reactions. Pre and post sIgE to milk and repeated SPT after finishing the protocol were performed for all the patients. In the cases of anaphylaxis the dose returned to the last tolerated dose for one week, then it was increased slower. Finally Full tolerance (120 mL of milk) was achieved in 13 patients (92.8%). Only one child had partially desensitized (64 cc). Six of them had anaphylactic reactions during the protocol that were managed by epinephrine, steroid, and antihistamines, while 2 mild reactions managed only by antihistamines. Prick sizes were significantly decreased in patients after desensitization. Milk desensitization and OIT appear to be efficacious in the treatment of cow’s milk allergy. The side-effect profile appears acceptable but requires further study. The permanency of the induced tolerance is still unclear. We propose the second study to survey whether this effect results to OIT.

**Keywords:** Cow’s Milk Allergy, Desensitization

**Pregnancy in primary immunodeficiency disorders**

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**Background:** Primary immunodeficiency disorder (PID) refers to a heterogeneous group of disorders characterized by poor or absent function in one or more components of the immune system. More than 200 different disorders have been identified to date, with new disorders continually being recognized. Treatment in PID patients has reduced the mortality and morbidity. Thus, such patients often survive into adulthood. Although it is likely that more women with PID will wish to become pregnant in the future, only a few such cases have been reported to date. In this article pregnancy PID pregnant cases and their management has been evaluated.

**Methods:** The PID cases that became pregnant have evaluated and followed in clinical immunology clinic of Isfahan medical university between 1997 and 2014. Depend on variety of immunodeficiency, supportive and definitive treatment has been done for pregant patients.

**Findings:** Two patients had pregnancy and were followed and managed, the first patient with CGD who treated several times for recurrent infections such as empyema, pneumonia, TB lymphadenitis, cutaneous abscess, mouth ulcer and gingivitis. After normal vaginal delivery, she faced to severe low back pain, sacral fungal osteomyelitis and granulomatous all over lesion in her uterus and liver who has been treated with antifungal and interferon gamma. The second patient with CVID who treated several times for thrombocytopenia and recurrent infection of upper and lower respiratory system. During her pregnancy period and delivery, she has been managed for severe thrombocytopenia by IVIG and platelet infusion.

**Conclusion:** Ig replacement therapy, antifungal and antibiotic prophylaxis, cytokine replacement, vaccinations and bone marrow transplantation are different treatments for PID patients. Immune regulation switch put provide
more complication and disorders during pregnancy. According to the type of PID, management and treatment of PID patients could be different and future researchers in this area have to be done.

**Keywords:** Primary Immunodeficiency, Pregnancy, CVID, CGD

**The role of Bid pro apoptotic gene in Ataxia Telangiectasia**

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**Background:** Ataxia telangiectasia (AT) is an early onset genetic disorder characterized by progressive neurodegeneration, chromosomal instability and an extreme sensitivity to DNA damaging agents such as ionizing radiation. An autosomal recessive disorder with signs cerebellar atrophy as the disease progresses including oculocutaneous telangiectasias, immunodeficiency, as well as a predisposition to cancer, particularly lymphoid cancers. ATM plays a central role in DNA damage checkpoint activation that is responsible for arresting cell cycle in order to promote repair and genome stability. However, it is becoming apparent that this is not the only role of ATM in promoting cell survival. One of factor involved in ATM mediated cell survival is the pro apoptotic protein BID. BID is a member of the “BH3 only” factors in the BCL 2 family and is typically found in the cytosol as a full length, inactive form.. However, when phosphorylated, BID becomes resistant to caspase 8 cleavage in what is described as a regulatory event.

**Methods:** Allelic discrimination assays by Taqman PCR were run on a 7500 FAST Real time PCR Thermocycler. Clustering of three genotypes: common homozygotes, rare homozygous and heterozygous were evaluated for Bid proapoptotic gene between 50 Normal controls of Iranian population and 50 Ataxia Telangiectasia patients.

**Findings:** Distributon of genotypes between controls and cases were shown 8 patients were heterozygote and one family was homozygote for BID proapoptotic gene.

**Conclusion:** In summary, this study raises the novel possibility that the BH3-only BID protein, a molecule that was previously considered to be active only as a proapoptotic factor, may also play a prosurvival role. If BID is indeed playing both a proapoptotic and a prosurvival function in the DNA-damage pathway, then it is an excellent candidate to link DNA repair processes and apoptosis.

**Keywords:** Ataxia telangiectasia, Bid Proapoptotic Gene Repair

**Malignancy or immunodeficiency? A case presentation**

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**Background:** Chediak-Higashi Syndrome (CHS) is a rare primary immunodeficiency. The current treatment consists of hematopoietic stem cell transplantation (HSCT). Untreated patients may experience a lethal malignancy like condition “Hemophagocytic Lympho Histiocytosis (HLH)”

**Methods:** We present here the clinical and laboratorial features of an undiagnosed case of CHS until he presented with lymphoma like symptoms. Changes on his features in response to treatment were compared using linear regression.

**Findings:** The patient was a 3-year-old boy who presented with episodes of unexplained fever, pallor, bilateral cervical masses, and abdominal distention from ten days before admission. He had a history of perianal abscess and recurrent upper respiratory tract infections. On physical examination, he had silvery and metallic sheen hairs. Several cervical lymph nodes, liver and spleen were palpable. Initial laboratory investigations revealed hemoglobin of 9.1g/dl, total leucocyte count (TLC) 10600/μl [neutrophil=16% and lymphocyte=84%] and platelet count of 81000 /μl. CT scan of the abdomen showed hepatosplenomegaly and abdominal lymphadenopathy. The lymph node biopsy was highly suggestive of lymphoreticular neoplasm. The bone marrow biopsy also showed lymphoid cell infiltration and in granulocyte cell domain a few giant granules inside them were seen. In the peripheral blood smear giant granules inside the neutrophils was seen and diagnosis of CHS was confirmed. Regarding to the patient’s conditions, other laboratory measures indicative of accelerated phase in CHS were performed. The laboratory findings were hemoglobin=6.3 g/dl, TLC=3000/μl [neutrophil=1% and lymphocyte=90%], platelet =28000/μl, triglyceride=381, total cholesterol=466, ferritin>5000, high AST and ALT (>300) and low fibrinogen levels (fibrinogen=70). The patient fulfilled five out of eight diagnostic criteria of HLH. So, he was diagnosed with accelerated phase in CHS and was started on HLH-2004 protocol. After beginning treatment the patient’s conditions improved gradually. Improvement in most clinical and laboratory conditions was significant (p<0.05), but decrease in splenomegaly wasn’t significant statistically. The child now has given HSCT and his condition is good.

**Conclusion:** The diagnosis of CHS should be suspected in patients with lymphoma like symptoms and any history of Partial albinism, recurrent infections, bleeding tendency and neurodegeneration

**Keywords:** Chediak-Higashi syndrome, oculo-cutaneous, albinism, accelerated phase, giant lysosomal inclusions

**Griscelli syndrome in clinical practice**

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**Background:** Primary immunodeficiencies (PIDs) are a group of diseases based on congenital, genetically determined, functional disorders of the immune system. According to the European Society for Primary Immunodeficiency the epidemiology of PIDs is 1 case per 25,000-100,000 people, selective IgA deficiency occurs with a frequency of 1 case per 500-700 people. Combined deficiency of humoral and cell-mediated immunity is 20-25% of all primary immunodeficiencies. Griscelli syndrome is one of those diseases. As combined immunodeficiencies are rarely occurred in pediatric practice, we consider to describe the case of Griscelli syndrome that was in our department’s practice.

**Case presentation:** Feature of this case was the presence of bleaching eyelashes and hair on the head, persistant
fever, enteropathy, polymorphous exanthem, and lymphadenopathy. The data of laboratory examination includes blood test which showed anemia, mild leukocytosis, thrombocytosis, neutrophilia, accelerated sedimentation (67 mm/h); immunotests showed severe disbalance of subpopulations caused by deep decreasing in the relative number of T helper cells and increasing in the relative and absolute number of T cytotoxic lymphocytes; histology of inguinal lymph node showed the image of chronic granulomatous process; histology of the skin showed local lymphoid infiltration with clusters of histiocytes. Based on this data primary immunodeficiency Griscelli syndrome was diagnosed. We performed replacement therapy for emergencies by concentrated red blood cells, for passive immunization intravenous immunoglobulin G, antiviral, antifungal, antiplatelete, and antiulcer combination therapy. The only effective treatment for this disease is the transplantation of hematopoietic stem cells. Thus, as shown by this case, caution is needed for primary care pediatricians and pediatric oncohematologists for possible primary immunodeficiency diseases caused by rare genetic abnormalities.

Keywords: Primary Immunodeficiencies, Griscelli Syndrome, Albinism
**Infectious Diseases & Vaccination Abstracts**

**Frequency of type 1 fimbriae among E.coli subtypes isolated from patients with urinary tract infections**

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**Background:** The gut constitutes an important reservoir of bacteria causing extra intestinal infections such as urinary tract infection (UTI). The fecal Escherichia coli population structure may influence the occurrence and etiology of extra intestinal infection, but it is poorly understood. The structure of the fecal E. coli population may be important in the pathogenesis of extra intestinal infections.

**Methods:** Rectal swab specimens from children were collected using wide mouthed sterile plastic containers and transported immediately to the microbiology laboratory for analysis within two hours of collection. Also urine samples had been collected by supra pubic aspiration, catheterization, or use of urine bags. All bacterial isolates were microbiologically identified in the microbiology laboratory of CMC (Children’s Medical Center) using standard biochemical identification methods. DNA Extraction: To extract DNA a sweep of growth on a nutrient agar slant was boiled in 500 µL of sterile distilled water for 10 minutes. Then centrifugation was done at 13000 rpm for 5 minutes to pellet the cell debris.

**Conclusion:** In our study type 1 pili like other study has shown to be the most commonly expressed virulence factors in UPEC and also DEC. This pili is commonly found among UPEC as well as non-UPEC strains. In confirmation of other study our results show more than 80% of E. coli isolates from the urine and rectal swab samples had fimH gene. More than 90% of E. coli isolates from the gut of healthy Swedish adults have the genes for type 1 fimbriae.

**Keywords:** E. Coli Subtypes, Multiplex PCR, Type 1 Fimbriae

**Serum levels of zinc, copper, magnesium elements and vitamin B12 in children with Giardiasis and Entrobiosis in Kashan, Iran**

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**Background:** Giardia lamblia and Enterobius vermicularis are noted to be the most important intestinal parasites in Iran as similarly reported in the world. The association of giardiasis and entrobiosis with the malabsorption of zinc, copper,magnesium and vitamin B12 remains controversial. The aim of the present study was to investigate the changes in the serum zinc, copper, magnesium and vitamin B12 levels in children infected with Giardia intestinalis and Enterobius vermicularis in comparison to normal subjects inhabiting in Kashan, Center of Iran.

**Methods:** The case control study was conducted among 359 children from 6-12 years old in region study. The study participants were selected using multistage sampling method. Data were gathered through house-to-house survey. Examination of stool samples and cello-tape anal swabs were done by standard techniques and detect G.intestinalis and E.vermicularis infection. From the whole population 37 Giardia and 50 Enterobius positive were chosen for case group, and 30 age and sex matched healthy children without parasitic infection were chosen as the control group. Both groups had no record of serum samples were obtained for further laboratory examination. Zinc, copper and magnesium levels was measured by Zeitschm Diagnostics Kit and colorimetric endpoint method respectively. Vitamin B12 levels were measured by Radioimmunoassay technique. All data were analyzed using SPSS version 17.

**Findings:** Out of the total examined subjects, 124 (34.5%) were girls and the mean ago was 3.4 years. Streptococcus pneumonia was 15.7%, but this rate in other studies is different from 4.2% to 72%. Some of the difference among population may related to sampling or laboratory methodology.

**Keywords:** Streptococcus Pneumunine, Nasophryngeal Carriage, Antibistic Sensitivity
Nasal carriage and resistance pattern of multi-drug resistant staphylococcus aureus among healthy children in Kashan/Iran

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Background: Nasal carriage of Staphylococcus aureus (S.aureus) is a substantial source of human infections. Detection and treatment of nasal carriage in children with methicillin-resistant (MRSA) and multi-drug resistant (MDR) S.aureus may be an important modality in prevention of infections. This study determined the prevalence, antibiotic resistance patterns and risk factors for nasal carriage of MDR S.aureus among healthy children.

Methods: This cross-sectional study was carried out on 350 one month to 14-year-old healthy children in Kashan city, the center of Iran. Of total health-care centers, four centers were chosen by simple random sampling. Nasal samples were cultured in blood agar medium for S.aureus diffusion and antibiotic susceptibility profile was determined by disc diffusion and E-test. Risk factors for nasal carriage of MDR S.aureus also were determined.

Findings: A total of 92(26.3%) S.aureus isolates were obtained, of which 33(35.9%) was MRSA and 27(29.3%) was MDR. Of MRSA strains, 19(70.4%) were MDR. S.aureus isolates showed 52.2% resistance to cephalothin, 33.7% to co-trimoxazole, 26.1% to clindamycin, 26.1% to ciprofloxacin, 4.3% to vancomycin and 35.9% to oxacillin. Risk factors for nasal carriage of MDR S.aureus were antibiotic use during recent 3 months (P<0.006), family size more than 4 members (P=0.044) and parental smoking(P=0.045).

Conclusion: MDR S.aureus was not uncommon among healthy children in Kashan and prevention of its spread in population is judicious.

Keywords: Staphylococcus Aureus, Nasal, Carriers, MDR, Healthy, Children

Screening of syphilis in pregnant women referred to Valiasr Hospital, Emam Khomeini Hospital

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Background: Syphilis is a systemic communicable infection and two forms of syphils are encountered in children: Acquired and congenital form. Syphilis during pregnancy has a transmission rate approaching 100%. Fetal or perinatal death occurs in 40% of affected infants. But approximately two thirds of infected infants are asymptomatic at the time of birth. This study showed the prevalence of syphilis in pregnant women by screening of VDRL at the delivery time.

Methods: This study was a cross-sectional one and VDRL test was done on blood samples of all pregnant women that admitted for delivery in gynecology dept of Valiasr Hospital, Imam Khomeini Hospital and studied mother age, pregnancy age, previous abortions, sex and weight of infants and if the VDRL of mother was positive, the VDRL of infants and clinical manifestations of neonates was studied.

Findings: With a total number of 605 pregnant women that tested with VDRL method, in two cases the results were weakly reactive and the remaining were negative. The test has been repeated in the first case by VDRL and in second case by FTA-ABS, that were negative in this two cases. Therefore, all 605 cases were negative.

Conclusion: The results of the current study, like other studies in Iran (Yazd, Mashhad &...), showed the prevalence of syphilis in Iran is very low, but we suggest this screening should be done, because the sequel is high and the treatment is easy.

Keywords: Syphilis, Pregnancy, VDRL, FTA-ABS

Role of rotavirus genotype in causing gastroenteritis associated with fever and seizure; does the virus directly affect the central nervous system?

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Background: Acute diarrhea is a major cause of mortality in young infants across the developing countries and rotaviral agents are the main etiology of this disease. The accompaniment of seizure with gastroenteritis is rather common. We conducted a study to assess the rate of rotaviral infection in children with gastroenteritis, fever and seizure, and to find the viral genotypes correlated with this condition.

Methods: Children with fever and gastroenteritis and seizure presenting to a children hospital were evaluated regarding the presence of rotavirus in feces and cerebrospinal fluid (CSF). The patients affected by rotavirus were compared to other patients regarding demographical and clinical characteristics.
**Findings:** Thirty one children were included. Ten cases (32%) were diagnosed to have rotaviral infection. The most common genotype was G1P8 which was found in 30%. Other common genotypes included G1P4 in 20%, G1P and G2P in 10% each. Viral agents were not found in the CSF of any of the patients.

**Conclusion:** Rotavirus is an important cause of gastroenteritis in association with fever and seizure in young children. The central nervous system (CNS) is not directly affected by the virus. In addition to other reasons, prevention of seizures and their consequences is a strong reason to proceed to nation-wide immunization of children against retroviral infections.

**Keywords:** Acute Diarrhea, Seizure, Gastroenteritis, Rotavirus

**Pattern of leishmaniasis demonstration (CL) in children referred to the of the skin and the leishmaniasis (sedigheh Tahereh), Isfahan 1392**

Nilforoushzadeh MA, Sokhanvari F, Ansari N, Heydari A

**Background:** Cutaneous leishmaniasis from remote times has been in Iran. Today, our country is one of the most important area of this disease in the world. The number of patients with cutaneous form in our country is about 20 thousands per annum. Some believe that the actual figures of the disease is more than 4-5 times this number and this disease is the most important parasitic disease after malaria in Iran. So far, an effective vaccine for this disease has not yet been made. In spite of massive efforts and national and international investments, not only this disease hasn't been eradicated, but is becoming the new area in the corner and in the future the country will find the higher prevalence. One of the major complications of the disease is wound scar that especially in children and girls, creating concern in family and can be applied the economic and social problems. This study was accepted to evaluate the clinical manifestations of disease patterns and the results of the treatment of disease in children.

**Methods:** This study was a descriptive study that has been done retrospectively. The community study was the children with cutaneous leishmaniasis has been referred to the centre for treatment of skin and leishmaniasis in Isfahan in 1392. Information collected via electronic files of the patients and then analyzed by SPSS software.

**Findings:** The findings based on this study of total 428 number referred to the skin center, the children were given up were 40.6%. Of this number, 53.4% was girls and 46.6% was boys. In terms of the age group, 48.3% was in Group 0-4 years, 38.5% was in Group 5-9 years and 13.2% was in Group 10-14 years. In terms of the location of the lesion, lesions on the face and neck have been 24% which 43.8% has been in girls and 56.2% in boys. 53.4% of lesions were on the limbs which 52.7% has been in the girls and 47.3 in the boys. Lesions on the body have been 10.3% which 66.7% has been in the girls and 33.3% in the boys. Of the total patients treated, 54.6% have had complete recovery, 45.4% have had partial recovery or no recovery.

**Conclusion:** The results suggest that nearly half of the patients are children. These results comment that the special efforts on the preparation of new drugs in the treatment of cutaneous leishmaniasis is not done yet in developed countries. Considering that repeated efforts in the fight against mosquito and rodent control have not had high efficiency in the country, we must pay special attention to supply vaccines and methods of prevention and preparation of new and effective drugs in the treatment of this disease. This issue needs to apply comprehensive cooperation from all authorities and respected professors including pediatricians.

**Keywords:** Cutaneous leishmaniasis, demonstrations, children, Sedigheh Tahereh

**Asymptomatic herpes simplex virus infection in iranian mothers and their newborns**

Fallah FO, Monavari SH, Abedi Kiasari B

**Background:** Human herpesvirus infections are very common and prevalent in the population and establish latency in different tissues. The study aimed to determine the prevalence of congenital herpes simplex virus infection in a randomly selected pregnant women and their newborns in Tehran, Iran.

**Methods:** The sera of pregnant women (n=100) were analysed for the presence of the herpes simplex virus specific antibodies (IgG and IgM) with an enzyme linked immunosorbent assay (ELISA). Consecutive umbilical cord blood samples from their newborn (n=100) were analysed for the presence of herpes simplex virus (HSV)-1 and HSV-2 DNAs using real-time polymerase chain reaction (PCR).

**Findings:** HSV IgG and IgM antibodies were found in 97 (97%) and 2 (2%) of 100 pregnant women. Of the 100 cord blood specimens tested, 6 (6%) were positive for herpes simplex virus DNA (HSV-1 DNA [n=2]; HSV-2 DNA [n=4]). From six HSV DNA positive newborns, 2 cases (HSV-2 positive) were from mothers who had detectable IgM in their sera samples. All corresponding mothers of six HSV positive infants had detectable IgG antibodies in their serum specimens.

**Conclusion:** The presence of HSV-1 and HSV-2 DNAs in the cord blood of newborns could be a risk marker for transmission of the virus from asymptomatic pregnant women to the child. Screening of newborns may help identify asymptomatic or misdiagnosed cases of genital and neonatal HSV infection.

**Keywords:** Herpes Simplex Virus Infection, Cord Blood, Real Time PCR, Congenital Infections

**Palmoplantar desquamation and nail shedding follow to hand foot mouth disease**

**Methods:** This is a retrospective case study in 50 patients that referred to Madany children hospital in khorram Abad for rash, fever, unusual palmoplantar desquamation and

**Keywords:** Acute Diarrhea, Seizure, Gastroenteritis, Rotavirus

**Palmoplantar desquamation and nail shedding follow to hand foot mouth disease**

Tae N, Faraji Goudarzi M, Sherkatolabbasieh HR

**Background:** Hand foot mouth disease (HFMD) is a common childhood disease. Coxsackievirus A16 is the main cause of classic disease and Coxsackievirus A6 is the uncommon etiology of nonclassic and severe HFMD. HFMD is self-limited illness that presented after short incubation period with low grade fever, exanthema and anathema lesions. We report 50 patients who presented unusual palmoplantar desquamation and nail shedding follow to HFMD in last summer and fall.

**Methods:** This is a retrospective case study in 50 patients that referred to Madany children hospital in khorram Abad for rash, fever, unusual palmoplantar desquamation and
nail shedding. All patients had clinical criteria for HFMD.

**Findings:** Females and males were presented equally. The ages of the patients were under 5 years with median age 3 years (range between 6 months to 5 years). The most symptoms were fever 80%, pharyngitis 60%, cough 20%, vomiting 15%, and diarrhea 15%. No patients had seizure and neurologic complications. The illness was self limited in all patients and cutaneous lesions cured after 2–3 weeks. After 3–4 weeks of the first febrile symptom of HFMD, palmoplantar desquamation and nail shedding presented. For all patients CBC, ESR, calcium and thyroid function tests performed and all laboratory findings were normal. Palmoplantar desquamation resolve after 2–3 weeks and nail shedding resolve after 2-6 months.

**Conclusion:** Palmoplantar desquamation and nail shedding are rare complications of HFMD that presented after involving with coxsackivirus A6. It must be differentiated from other childhood disease such as Thyroid disease, Kawasaki, and chronic disease.

**Keywords:** Hand Foot Mouth Disease, Palmoplantar Desquamation, Nail Shedding

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**Fever of unknown origin in children aged 3 Months to 15 years**

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**Background:** Fever of unknown origin (FUO) is defined as the presence of fever for 8 or more days in a child in which a careful history and physical examination and preliminary laboratory results failed to reveal the probable cause of fever. The causes of FUO are different according to geographical regions. The aim of the study was to evaluate the common causes of childhood FUO in our region.

**Methods:** A 6 years retrospective study conducted on all admitted children aged from 3 months to 15 years and those with final diagnostic of FUO were entered to the study.

**Findings:** Numbers of eligible cases for the study were 1100 patients. The causes of FUO were infectious diseases (55.1%), collagen vascular (4.6%), neoplasm (6.7%), miscellaneous (23.3%) and undiagnosed (10.3%).

**Conclusion:** Most fever of unknown origin results from atypical presentation of common diseases like TB and Salmonellosis, Brucellosis and Pneumonia.

**Keywords:** FUO, Pediatrics, Infectious Diseases

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**Virulence factors in uropathogenic and commensal Escherichia coli isolates**

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**Background:** Urinary tract infections (UTIs) including cystitis and pyelonephritis are the most common infectious diseases in childhood. Escherichia coli (E. coli) accounts for as much as 90% of the community-acquired and also 50% of nosocomial UTIs. Therefore, identification of E. coli strains and antibiotic resistance patterns is important for both clinical and epidemiological implications.

**Methods:** To characterize uropathogenic strains of E. coli, we studied 50 E. coli strains recovered from urine samples of children aged less than 7 years with community-acquired UTIs and 50 E. coli recovered from stool sample of healthy children. We assessed virulence factors (VFs) and drug sensitivities of E. coli isolates.

**Findings:** Drug sensitivities of the isolates were: 94% (amikacin), 90% (nitrofurantoin), 66% (gentamicin), 56% (ceftriaxone), 40% (malidixic acid) and 28% (cotrimoxazol). Laboratory tests showed that the prevalence of virulence factors ranged from 18% for hemolysin and P-fimbriae 2% for type1-fimbriae.

**Conclusion:** Most drug resistance was to cotrimoxazole and amikacin was the lowest. P-Fimbriae and hemolysin in uropathogenic Escherichia coli was more frequent than non-pathogen type of E. coli.

**Keywords:** Escherichia Coli, Virulence Factor, Drug Resistance, Urinary Tract Infection

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**The effect of bacterial and yeast probiotics on acute watery diarrhea in children**

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**Background:** Diarrheal diseases are one of the main causes of mortality and morbidity in the developing countries. ORS (oral rehydration solution) has turned up to decrease diarrheal mortality, but has left no effect on the duration and severity of the disease. Different studies have focused on the use of probiotics as a supplementary treatment for acute diarrhea. This current study was to evaluate the effect of bacterial and yeast probiotics in children with acute diarrhea.

**Methods:** This clinical trial was conducted on 90 two-five year old children with acute watery diarrhea referred to pediatrics clinic in 2013 as outpatient cases. These children were divided randomly into three different groups of thirty. One group was under standard treatment with oral dehydration solution in addition to bacterial probiotics in form of Kidilact once a day. The second group, in addition to ORS, received yeast probiotics in form of Argopharm 250mg once a day. The third group received standard rehydration therapy along with placebo. Treatment with probiotics or placebo lasted for 5 days.

**Findings:** Our findings demonstrated no significant difference in the three groups in terms of age, sex, frequency, and duration of diarrhea before treatment. Mean durations of diarrhea from commence of treatment to recovery were 2.80, 3.17, and 4.43 days for the first (bacterial probiotic), the second (yeast probiotic), and the third (placebo) groups, respectively. A significant difference was revealed among the three groups in terms of diarrhea duration. The bacterial probiotic group had the best effect in reducing diarrheal length, while the placebo group had the least effect in this respect.

**Conclusion:** Findings of the current study and those of other studies confirm that application of probiotics in pediatric diarrheal diseases can be effective in minimizing course of diarrhea. According to our study, bacterial probiotic appeared more effective than the yeast probiotics in this regard.

**Keywords:** Diarrhea, Probiotic, Yeast, Children
**Bilateral parotiditis as the first presentation of HIV infection**

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**Background:** Parotiditis is prevalent in childhood. The etiology of parotiditis can be divided into infectious and non-infectious ones. Viral infections are the main causes of Mumps (post vaccinal or disease) parotiditis such as enteroviral and HIV. Apart from this viruses, EBV, CMV, influenza, para influenza 1, 2 infections as well as staph aureus can cause the same problem. Most of bacterial parotiditis are unilateral. There are also noninfectious cause for parotiditis; Obstruction of stensen duct, collagen vascular disease (sjogren disease, SLE,…) and tumors are among this types of causes.

**Case presentation:** A 2 year old girl admitted to hospital with bilateral parotiditis, having fever and chills for two weeks. Her vital signs were stable. Vaccination were carried out in due times. In physical examination of head and neck, both parotids were erythematous and tender. Liver and spleen had normal size, no lymph nodes were palpable. In lab test (Na=26%, L=70%, mono=74%); WBC: 9200, Hb=10.3, PLT=383000. ESR was raised upto 117. PPD test, IgM VCA and toxoplasma antibody (IgM, IgG) were negative. Serum amylase raised up to 250 unit/L. She tested for HIV infection and the test was positive. In paracinal study sonography were done and revealed a large hypoechoic mass with several necrotic cycts. In left side there were collected fluid. Chest radiography reported diffuse bilateral nodular infiltration. she underwent surgery for abssece fluid. Her parents were tested for HIV infection and the test result for both of them proved positive.

**Conclusion:** In childhood, bilateral infections with this presentation of HIV infection must be remembered.

**Keywords:** HIV, Infection, Parotiditis

**Antibiotic susceptibility survey of Group A streptococcus isolated from Iranian children with pharyngitis**

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**Background:** The aim of this study was to contemplate the antibiotic susceptibility of Group A streptococcus (GAS) to the antibiotics which are usually used in Iran for treatment of GAS pharyngitis in children.

**Methods:** From 2010 to 2013, children 3-15 years of age with acute tonsilopharyngitis who attended Mofid children’s hospital clinics and emergency ward and met the inclusion criteria were enrolled in a prospective study in a sequential manner. Throat culture was carried out in 200 children. The isolates strains were identified as GAS by colony morphology, gram staining, beta hemolysis on blood agar, sensitivity to bacitracin, a positive pyrrolidonylaminopeptidase (PYR) test result and the presence of Lancefield A antigen determined by agglutination test. Antimicrobial susceptibility was first identified by disk diffusion method using the disk of commonly used antibiotics in Iran and in accordance with Clinical and Laboratory Standards Institute (CLSI 2012) guideline. Susceptibility of the GAS strains to the described antibiotics was also checked by broth dilution method according to CLSI guideline recommendation.

**Findings:** Of the 200 children enrolled in this study, 59 (30%) cases were culture positive for GAS. All isolates were sensitive to penicillin G. The prevalence of Erythromycin, Azithromycin and Clarythromycin resistance was 33.9%, 57.6% and 33.9%, respectively. Surprisingly 8.4% of GAS strains were resistant to rifampin. In this study, 13.5% and 32.2% of the strains were resistant to clindamycin and ofloxacin respectively.

**Conclusion:** The high rate of resistance of GAS to some antibiotics in Iran should warn the physicians to use the antibiotics restrictedly and logically to prevent the rising of resistance rate in future. It also seems that continuous local surveillance is necessary to achieve the best therapeutic option for GAS treatment.

**Keywords:** Group A Beta-Hemolytic Streptococci, Pharyngo, Tonsillitis, Streptococcus

**Efficacy of zinc sulfate on common cold in children aging from 1 to 5 years**

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**Background:** Common cold is the most common acute upper respiratory tract disease in children. The efficacy of vitamins and minerals on duration, symptoms and complications of common cold were evaluated in several studies. This study was performed to determine the efficacy of zinc sulfate on common cold in children aging from 1 to 5 years attending to Javaheri hospital in 2012-2013.

**Methods:** In this clinical trial study, 112 children with common cold aging from 1 to 5 years attending to Javaheri hospital in 2012-2013 were enrolled. They are randomly assigned to receive either conventional treatment including Acetaminophen, pediatric cold and nasal drop or these drugs plus zinc sulfate for 5 days. Then the duration of symptoms and complications of common cold were compared across two groups.

**Findings:** The results demonstrated disease duration was significantly shorter in zinc group(p<0.03). Also the frequency of nasal discharge, sneezing, cough and sinusitis was significantly less in zinc group(p<0.05).

**Conclusion:** Totally according to the obtained results, it may be concluded that zinc sulfate is effective on duration of symptoms and complications of common cold in children aging from 1 to 5 years old.

**Keywords:** Zinc Sulfate, Common Cold, Children, Complications

**Evaluation of procalcitonin in diagnosis of renal parenchymal involvement in children with urinary track infection**

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**Background:** Febrile urinary tract infection (UTI) is a common problem among children and carries the risk of parenchymal damage and sequelae. The location of the infection within the urinary tract influences decisions regarding both therapeutics and follow-up. Because clinical features and laboratory markers of infection at an early age
A four-year evaluation of causative pathogens of recurrent urinary tract infection and their antibiotic resistance patterns among the children referred to Loghman Hospital in Iran

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Background: Recurrent urinary tract infections (RUTIs) are mostly caused by re-infection with the original bacteria or another different isolates in children. This study was conducted to detect the most common causative agents of RUTI as well as their antibiotic susceptibility pattern.

Methods: A retrospective study was conducted among patients who referred to Logman hospital in Tehran, Iran, from March 2008 until March 2011. The isolates from the urine cultures were subjected to antibiotic susceptibility test using disk diffusion method according to CLSI guidelines. Extended spectrum beta-lactamase (ESBL) and multi drug resistant (MDR)-ESBL patterns were defined based on CLSI definition, and CDC and ECDC guidelines.

Findings: Seventeen isolates of gram-negative bacteria from the patients with RUTI were identified among 1,112 patients who referred to the hospital in this period. Among the patients, 1,045 were boys and 67 were girls. Twelve patients had more than two episodes of UTI. The most frequently isolated organism was E. coli (45.6%), followed by Serratia marcescens (20%), Acinetobacter baumannii (16%), and Enterobacter species (12%). The antibiotic susceptibility of the isolates was determined by disk diffusion method according to CLSI guidelines. The susceptibility pattern of common isolates i.e. Serratia marcescens (45.6%), E. coli (20%), and Acinetobacter baumannii (16%) were mostly sensitive to gentamicin, amikacin, and tobramycin. However, S. marcescens showed more MDR patterns compared to E. coli and K. pneumoniae isolates. The Susceptibility pattern of common isolates i.e. Serratia marcescens (45.6%), E. coli (20%), and Acinetobacter baumannii (16%) were mostly sensitive to gentamicin, amikacin, and tobramycin. However, S. marcescens showed more MDR patterns compared to E. coli and K. pneumoniae isolates. The Susceptibility pattern of common isolates i.e. Serratia marcescens (45.6%), E. coli (20%), and Acinetobacter baumannii (16%) were mostly sensitive to gentamicin, amikacin, and tobramycin. However, S. marcescens showed more MDR patterns compared to E. coli and K. pneumoniae isolates.

Keywords: Recurrent UTI, Antibiotic Susceptibility Pattern

Antibiotic resistance patterns of gram-negative bacteria in two tertiary hospitals in Sanandaj, Iran

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Background: There is a growing concern about the rapid rise in the resistance of gram negative bacteria to antimicrobial agents. We conducted a study to assess the antibiotic susceptibility patterns of common gram-negative bacteria isolated from infections of normally sterile body sites at two tertiary hospitals in the Sanandaj city, Kurdistan Provenience, Iran.

Methods: From January 2011 to December 2011, all positive cultures from potentially sterile body fluids were gathered from two university hospitals in Sanandaj. The antibiotic susceptibility and minimum inhibitory concentration (MIC) were determined using the Kirby-Bauer method (disk diffusion technique). The results were interpreted as per Clinical and Laboratory Standards Institute (CLSI) guidelines against a panel of the antimicrobials.

Findings: Seventy isolates of gram negative bacteria from patients with infections were collected. Serratia marcescens was the most frequently isolated organism (38%) followed by Escherichia coli (19%), Klebsiella pneumoniae (19%), Acinetobacter baumannii (6%), Enterobacter species (6%), Serratia odorifera (4%) and Pseudomonas species (5%). The Susceptibility pattern of common isolates i.e. Serratia marcescens, E.coli, and K.pneumoniae for commonly used antibiotics were as follows: ampicillin 3.3%, 6.7%, 20%; gentamycin 73.3%, 73.3%, 46.7%; amikacin 76.7, 93.3%, 53.3%; cotrimoxazole 70%, 13.3%, 40%; cephalothin 3.3%, 40%, 33.3%; cefazidim 80%, 73.3%, 33.3%; piperacillin/tazobactam 90%, 66.7%, 86.7%; cefepime 80%.
Prevalence of different microorganisms in CSF cultures and their susceptibility pattern in Children's Medical Center hospital during 2009-2014, Tehran, Iran

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Background: Bacterial meningitis is associated with high mortality rate. So, it is important to define antimicrobial sensitivity of the causative microorganism for proper treatment. This study has designed to recognize organisms’ types in CSF cultures in a tertiary children’s hospital to determine the prevalence of different microorganisms and their antimicrobial sensitivity to detect antibiotics suitable for empirical treatment.

Methods: The data of CSF cultures from March 2009 till March 2014 were collected from hospital laboratory. All positive CSF cultures have been included except repeated ones with the same organisms and antibiogram. The antimicrobial sensitivity testing was performed on Muller-Hinton agar by the Kirby-Bauer disk diffusion method and if it was necessary, by determining MIC level using E-test method according to CLSI instruction.

Findings: 189 specimens were positive among 5578 CSF cultures received during this period. Most frequent organisms were enteric gram negative bacilli (21.9%), coagulase negative staphylococci (21.9%), pseudomonas spp (10.6%), acentobacter spp (10.1%), staph aureus (9%), streptococcus pneumonia (5.3%), other streptococcal spp (5.3%), haemophilus spp (4.8%), enterococcus (4.8%), candida spp (2.6%), micrococci (1.0%), salmonella spp (0.5%), neisseria meningitides (0.5%) and diphtheria spp (0.5%). Near half of enteric gram negative bacilli were resistant to all ephalosporins. This rate was less among Aminoglycosides. Among Staph aureus and pseudomonas which are main causes for nosocomial infections, resistancy to Imipenem and Tazobactam-Piperacillin have been reported frequently.

Conclusion: Our data shows that most of the organisms are resistant to first line medications and it is necessary to review our antibiotic administration protocols occasionally in order to treat bacterial meningitis and to avoid resistance spreading in surgical and ICU wards.

Keywords: CSF Culture, Antimicrobial Susceptibility, Bacterial Meningitis, Nosocomial Infection

Latest situation of new vaccine introduction in Iran, EMRO and in the World

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Immunization prevents illness, disability and death from vaccine-preventable diseases and averts an estimated 2 to 3 million deaths every year from diphtheria, tetanus, pertussis and measles. World health organization recommend that every child in the world should receive at least 11 antigen in their routine immunization which includes BCG, DTP, Measles, polio, hepatitis B, Hib, Pneumococcus, Rota and rubella. According to WHO annual report only 5% of children in the world receives base vaccination. During 2013, about 84% (112 million) of infants worldwide received 3 doses of DTP3 vaccine. Hib vaccine had been introduced in 189 countries by the end of 2013. Global coverage with 3 doses of Hib vaccine is estimated at 52%. Hepatitis B vaccine for infants had been introduced nationwide in 183 countries by the end of 2013. Global coverage with 3 doses of hepatitis B vaccine is estimated at 81%. Pneumococcal vaccine had been introduced in 103 countries by the end of 2013, and global coverage was estimated at 25%. Polio is a highly infectious viral disease that can cause irreversible paralysis. In 2013, 84% of infants around the world received 3 doses of polio vaccine. Targeted for global eradication, polio has been stopped in all countries except Afghanistan, Nigeria and Pakistan. Rotavirus vaccine was introduced in 52 countries by the end of 2013, and global coverage was estimated at 14%. Although there are enough data on vaccine preventable disease burden and cost benefit for new vaccine introduction, it is necessary to move forward and change our routine vaccination program and introduce Hib, Rota and pneumococcal vaccine as soon as possible in our national immunization schedule. It is due to lunch Hib vaccine before the end of this year.

Keywords: Immunization, Vaccination, New Vaccine