27th International Congress on Pediatrics

&

13th Congress on Pediatrics Nursing

Oct 8-11, 2015

Children’s Medical Center

Tehran- Iran

Organizer:

- Children’s Medical Center, Pediatrics Center of Excellence, Tehran, IR Iran
- Growth & Development Research Center, Tehran University of Medical Sciences, Tehran, IR Iran
- Department of Pediatrics, Tehran University of Medical Sciences, Tehran, IR Iran
President
Dr. Gholamreza Valizadeh
Tehran University of Medical Sciences

Scientific Director
Ali Rabbani, MD
Tehran University of Medical Sciences

Executive Director:
Reza Shervin Badv, MD
Language editing by: Qazvini Baqer

In collaboration with:
- Tehran University of Medical Sciences International Relations Office
- Tehran University of Medical Sciences Public Relations Office
- Iranian Pediatric Endocrinology Association

Scientific Committee:
- Abbasi Farzaneh; MD
- Aghaei Ehsan; MD
- Ashrafi, Mahmoud-Reza; MD
- Badv Reza-Shervin; MD
- Malekan Rad Elaheh; MD
- Modaresi Mohammad-Reza; MD
- Rabbani Ali; MD
- Valizadeh Gholamreza; MD
- Zainalo Ali-Akbar; MD
- Zamani Gholam-Reza; MD
- Ziaee Vahid; MD
Executive Committee:

1- Afshari, Kh  
2- Afzali, P  
3- Aiati, M  
4- Akhlaghi, AA  
5- Aliakbari, H  
6- Archang, M  
7- Atemad Saeed, S  
8- Badv, R-Sh  
9- Barakati, S H  
10- Behfar, A  
11- Dastmalchi, F  
12- Ekrami, Sh  
13- Ghahvechi, M  
14- Gharehzadeh Shirazi, A  
15- Hadipour, M  
16- Hosaini Node, S S  
17- Kalhor, P  
18- Karimi, H  
19- Karimi, R  
20- Khajehzadeh, M A  
21- Kianmehr, Sh  
22- Kouhnavard, M  
23- Malekzadeh, I  
24- Mirzaei, B.  
25- Mohammadi, M S  
26- Mohebbi, A  
27- Momeni, A  
28- Pour Hadi,N  
29- Qazvini, M B  
30- Rad,R  
31- Rahimi, S.  
32- Ramzi, SH  
33- Rastin, Gh  
34- Rostami, Y  
35- Sadrosadat, T  
36- Seifkhani, A.  
37- Tabrizi, M  
38- Tajedini, P  
39- Vafaei, N  
40- Yarali, B  
41- Yosefzadeegan, S  
42- Zainalo, A  
43- Ziaali, A
Study of congenital heart disease risk factors: a case-control study

Moslem Taheri Soodejani1, Kourosh Rajabkhah2 1. Shahid Sadoughi Medical University of Medical Science, 2. Tehran Medical University of Medical Science, Tehran, Iran

Background: Congenital heart defects are called to some situations which start from birth and affect the baby’s heart and its performance. Different types of these defects can range from mild (like a small hole between the heart chambers) to rigid (such as a flaw or weakness in a part of the heart)

Methods: This is a case-control study. Newborns which their echocardiogram screening tests were abnormal at birth and were born during years 2011 and 2012 were participated in the study. For control group, all the children who were not diagnosed with any disorders were participated in the study and were matched according to sex and living area. Statistical methods such as descriptive statistical analysis, T student, and logistic regression modeling were performed and SPSS 16 was used for analyzing the data. All tests were done with significance level of 0.05.

Findings: logistic regression modeling determined that history of stillbirth in pregnant women (OR=7.85), not taking a multivitamin before pregnancy (OR=4.38), maternal obesity during pregnancy (OR=3.02), and maternal weight gain during pregnancy (OR=2.09) were risk factors that had orderly the highest correlation with these disorders.

Conclusions: Various factors are involved in congenital malformations which are mainly related to maternal factors during pregnancy. So in order to prevent the birth of children with heart disorders, maternal care, particularly during pregnancy seems to be necessary.

Keywords: Risk Factors, Congenital Malformations of the Heart, Neonatal, logistic regression, case-control study

Mitochondrial Cytb Gene Mutation in Pediatric Patients with Congenital Heart Diseases

Nafiseh Karimian1, Mehri Khatami1, Mohammad Mehdi Heidari1, Mehdi Hadadzadeh2 1. Department of Biology, Faculty of Science, Yazd University, Yazd, Iran; 2. Department of Cardiac Surgery, Afshar Hospital, Shahid Sadoughi University of Medical Sciences, Yazd, Iran.

Background: Congenital heart disease (CHD) is the leading cause of birth defects, and accounts for 10% of all congenital anomalies. CHD is multifactorial due to both genetic predisposition and environmental influences. Mitochondrial dysfunction is a characteristic of heart failure. Mutations in mitochondrial DNA, particularly in MT-CYB, have been associated with several cardiac disorders. We hypothesized that MT-CYB mutations might play an important causal or modifying role in Iranian pediatric patients suffering congenital heart disease.

Methods: In the present study, cytochrome B gene was analyzed using PCR-SSCP technique and DNA sequencing to search for mutations in 35 patients with congenital heart diseases.

Results: Mutation analysis of these patients revealed a homoplasmic nucleotide substitution in 14766C>T location of cytochrome b gene sequence that cause Thr to Ile alteration. We found this substitution in 4 patients that was not detected in healthy controls.

Conclusion: Our finding suggests that missense mutation in MT-CYB gene may be associated with severity in pediatric CHD patients. Larger studies in different ethnic groups are needed to establish the precise role of this mutation for congenital heart diseases patients.

Keywords: Congenital Heart Disease, Mutation, PCR-SSCP, Cytochrome b

Lethal infantile hypertrophic cardiomyopathy: A case report

Ali Dehghani Firoozabad1, 2, faye petropoulou2, Mohammad Tahva Vahdi Mehjardi1, Reza Maroofian2, Mohammad Reza Dehghani1, Yalda Jamshidi2 1)Yazd Cardiovascular Research Center, Shahid Sadoughi University of Medical Sciences, Yazd, Iran. 2)Human Genetics Research Centre, St George's University of London, London, UK.3).Biomedical and Clinical genetic research center, Shahid Sadoughi University of Medical Sciences, Yazd, Iran.

Background and aims: Mutations in glucosidase, alpha, acid (GAA) are one of many causes of lethal infantile hypertrophic cardiomyopathy. This is one of the symptoms of Glycogen storage disease type II (Pompe disease), a rare autosomal recessive disorder, which is often fatal for those with early onset. Here, a family is presented with two infants, both of whom suffered a cardiac arrest and subsequently died at a very young age. They were diagnosed with severe hypertrophic cardiomyopathy but both also presented with respiratory insufficiency.

Methods: Exome sequencing and subsequent filtering was performed prior to validation by Sanger sequencing and pathogenicity assessment by online prediction software.

Findings: A rare variant, c.1927G>A (rs28937909), was found to segregate within this pedigree was predicted to be pathogenic. The first finding was in 1993 however other findings have since occurred.

Conclusions: This pedigree supports the association between GAA and (Pompe disease). It is the first case of this variant in individuals of Iranian descent. An unusual aspect of this case is for this variant to cause such a severe early-infantile phenotype, as well as both affected infants being homoyzygous while previous infant cases were compound heterozygous. The fact Pompe disease was not diagnosed would have delayed genetic causative identification without exome sequencing, and may have led to related symptoms being missed. Therefore, awareness among clinicians is an area for development. The variant occurred in a region of the gene with unknown function thus this finding implicates the need for further research into the role of this region.

Keywords: Infantile Cardiomyopathy, GAA, Pompe, Glycogen storage disease
The comparison of the left coronary artery diameter ratio to aorta and coronary sinus in children with Kawasaki disease with healthy children

Iran Maleksadat1, Elaheh Malekn-Rad2, Vahid Ziaee2, R. Rajaee2, Z. Shabahi1
1. Children’s Medical Center, Tehran University of Medical Sciences, Tehran, Iran
2. Department of Pediatrics, Tehran University of Medical Sciences, Tehran, Iran
3. Pediatric Rheumatology Research Group, Rheumatology Research Center, Tehran University of Medical Sciences, Tehran, Iran

Background: Kawasaki, a vasculitis that affects medium arteries, is the most common cause of acquired heart disease in children. This inflammation, if left untreated, leads to dilation and aneurysm in coronary arteries in 25% of cases. In this study, we try to introduce a novel method for examining coronary affection in Kawasaki disease.

Methods: The study was done as a case-coronary study. The ratio of the diameter of the LMCA to the diameter of the aorta (LMCA/Aorta), and LMCA to the coronary sinus (LMCA/CS), and LMCA to the sinus to the aorta (LMCA/CS/Aorta) were calculated.

Findings: Thirty-eight children with Kawasaki were studied as the “case” group and thirty-one healthy children were studied as the “control” group. Eight children in the case group had a Z-score above 2.5 in the first echo and five children had such a Z-score in the second echo. The ratios of the internal diameters LMCA/CS, LMCA/Aorta, LMCA, and LMCA/CS/ Aorta had significant differences between the case and control groups, in both the first and second echos. Cut-off was calculated as 0.15 for LMCA/Aorta with sensitivity of 60% and specificity of 81%. The cut-off was calculated for LMCA/CS as 0.4 and for LMCA/CS/Aorta as 0.03.

Conclusion: Although our study population was small and it is almost a pilot study, there is a significant difference in all three new measures between the case and control groups, based on the gold standard method. Moreover, the independency of these measures of age, weight, and body surface is an important benefit. So, we may achieve quicker and maybe even more accurate measures than the current ones.

Keywords: Kawasaki disease, Echocardiography, Coronary Arteries, Aorta, Left Main Coronary Artery

Comparing the effects of intermittent positive pressure ventilation and spontaneous ventilation on cardiorespiratory parameters of pediatric patients less than 1 year old, undergoing angiography under general anesthesia

Behrouz Nooralishahi, Mehrdad Goodarzi, Arman nasiri
Children Medical Hospital, Tehran University of Medical Sciences, Tehran, Iran

Background: Maintaining the physiological state of hemodynamic and respiratory conditions during diagnostic angiography is of utmost importance. According to the necessity of performing this procedure under general anesthesia in most pediatric patients, identifying the most reliable method with minimal hemodynamic and respiratory effects seems to be an important priority. Considering the possible effects of anesthesia and respiratory support on hemodynamic parameters and consequently on diagnostic results of an angiogram, evaluating these effects can help distinguish the most appropriate method to a great extent. Aims: We aimed to compare the effects of Intermittent Positive Pressure Ventilation and Spontaneous Ventilation on cardiorespiratory parameters of pediatric patients less than 1 year old, undergoing Angiography under general anesthesia in Children’s Medical Center in 2014-2015.

Methods: A total of 60 pediatric patients less than 1 year old, candidate for diagnostic angiography were randomly assigned to two groups of IPPV and Spontaneous ventilation. Induction with ketamine, intubation and maintenance of anesthesia with sevoflurane gas were performed. The first group had respiratory support via IPPV and the second group breathed spontaneously. The hemodynamic and respiratory parameters were evaluated through different steps of the procedure and then were analyzed via SPSS software version 20.

Findings: The mean age of the study population was 6.63±3.40 months with a minimum of 14 days and a maximum of 12 months. The comparison between the two groups of IPPV and spontaneous ventilation regarding the evaluated parameters showed that only the differences in the SPO2 before anesthesia (P=0.022), SPO2 after anesthesia (P=0.019) and PCO2 during angiography (P=0.02) were statistically significant.

Conclusion: The minor differences in respiratory parameters between the two groups suggested that these two methods can be interchangeably applied in pediatric patients undergoing general anesthesia for diagnostic angiography.

Keywords: Cardiac Angiography, IPPV, Spontaneous Ventilation, Pediatrics

Evaluation of the surgical outcome of the congenital heart disease patients in Children’s Medical Center: a two years survey

Salehi AbarghuiieForud1, MohamadReza Mirzaaghbayan2, Sara Baghery1, Hamid-Reza Riasi2
1. Atherosclerosis and Coronary Artery Research Center, Birjand University of Medical Sciences, Birjand; 2. Tehran University of Medical Sciences, Tehran; 3. Birjand University of Medical Sciences Birjand, Iran

Background: Today, with advances occurring was happen in cardiopulmonary bypass(CPB) to improve care processes, and improved processes of anesthesia care, including intubation, early surgical intervention for total correction of anatomic abnormalities in patients with congenital heart disease was possible. Today, with advances occurring in cardiopulmonary bypass (heart-Lung- pump) including improved anesthesia intubation and early surgical intervention to correct life-threatening congenital heart anatomic abnormalities in patients ages is possible

Methods: Because there is limited information in this field in our country. Our study aims was planned to collect information from one of the most important centers of pediatric cardiac surgery in Iran. In this study other methods of pediatric cardiac surgery and mortality was studied for two years. This retrospective descriptive study
was carried out on 789 patients made within two years. Information related to kinds of surgical operation and deaths were collected and analyzed.

**Findings:** The age range of patients varied from three days to 18 years. The male/female ratio was 53.4%. CPB was not applied for 21.1% of patients. The frequency of operations included PDA (Patent Ductus Arteriosus) [D&L] (4.7%), total correction for TF (Tetralogy of Fallot) [4.8%], systemic to pulmonary shunt (8.7%), ASD (Atrial Septal Defect) & VSD (Ventricular Septal Defect) closure (%2.7) and (%16.3), respectively. Total mortality rate was 8.6%. Analysis of operated population showed that outcome of patients was improved in recent years.

**Conclusion:** The results show that children's congenital heart surgery is on the rise.

**Keywords:** congenital heart diseases, cardiac surgery, mortality, children

---

**Comparison between two different methods for determination of the myocardial performance index (MPI or Tei index) to assess RV global function**

Ehsan Aghaei-Moghadam, Arman Kucharian, Mohammad-Reza Mirzaaghayan, Mohammad Rezaeiz, Maryam Nikofar

Children Medical Center, Tehran University of Medical Sciences, Tehran, Iran; and Department of Pediatrics, Tehran University of Medical Sciences, Tehran, Iran

Tetralogy of Fallot is the most common cyanotic disease of the childhood. Regarding to it's surgical treatment, pulmonary regurgitation is inevitable. In this study, clinical follow up of the fragile right ventricle of these children, is critically important for proper timing of interventional or surgical pulmonary valve replacement. This article was aimed to make a comparison between two different methods for determination of the myocardial performance index (MPI or Tei index) as reliable, repeatable, and easy way to assess RV global function.

**Keywords:** tetralogy of Fallot, pulsed doppler echocardiography, Tei index

---

**Red Flags for Chest Pain of Cardiac Origin in Children**

Elaheh Malakan Rad, MD, FACC; Children Medical Center, Tehran University of Medical Sciences, Tehran, Iran; and Department of Pediatrics, Tehran University of Medical Sciences, Tehran, Iran

**Background:** Chest pain is one of the common complaints in children and adolescents. However, a cardiac origin is usually rare. The aim of this study is to review the cardiac causes in this age group.

**Methods:** Using a combination of the Key words of chest pain, children, adolescents, pediatrics and cardiac; a literature review was performed. Papers with non-cardiac causes of chest pain were excluded. Those on cardiac causes were studied and red flags were extracted.

**Findings:** Cardiac causes range from relative benign causes like mitral valve prolapse to potentially fatal diagnoses like dissection of aorta and acute pulmonary embolism. Important cardiac causes of chest pain include: 1-Myocardial infarction either acquired or due to a congenital cardiac abnormality such as anomalous origin of left coronary artery from pulmonary artery. Astute attention to detection of abnormal Q waves on electrocardiogram in leads I, AVi, V5 and V6 is important. 2- Acute pulmonary embolism usually produces a sharp chest pain that may exacerbate with deep breathing. Attention to the predisposing factors for development of pulmonary embolism like deep vein thrombosis due to long-standing central venous catheter, right-sided bacterial endocarditis of the heart, sepsis is crucial. 3- Dissection of aorta is another cause of fatal chest pain in children. Children and adolescent with Marfan syndrome, bicuspid aortic valve with associated aortopathy and trauma are at higher risk for developing this complication. 4- Cardiac arrhythmias are easily diagnosed by physical and electrocardiographic examination. 5- Congenital heart diseases like aortic stenosis, hypertrophic cardiomyopathy and mitral valve prolapse can produce chest pain.

**Conclusion:** Chest pain is children and adolescents are rarely due to cardiac causes. However, when cardiac problems leads to chest pain, they are often life-threatening and need quick diagnosis and treatment.

**Keywords:** Chest pain, children, cardiac

---

**Pneumopericardium as a cause of ST segment elevation after open heart surgery**

Mino Dadakhah; Mohammadreza Mirzaaghayan; Reza Shahbani

Children Medical Center, Tehran University of Medical Sciences, Tehran, Iran; and Department of Pediatrics, Tehran University of Medical Sciences, Tehran, Iran

**Background:** ST segment elevation is a significant finding in electrocardiography and can be a manifestation of myocardial ischemia, myocarditis, pericarditis, hyperkalemia and Brugada syndrome. Other conditions such as pneumopericardium, pneumomediastinum and pneumothorax can mimic ST elevation seen in myocardial ischemia.

**Case presentation:** A 12-year-old male patient presented with exertional dyspnea. Electrocardiography (ECG) showed normal sinus rhythm. Echocardiography revealed 18 mm secundum atrial septal defect (ASD). Because of deficient aortic rim the patient was scheduled for open heart surgery. On the first postoperative day, the patient had chest pain. ECG showed diffuse ST segment elevation in all leads. Echocardiography was normal; creatine kinase MB isoenzyme (CK-MB) and cardiac troponin were also in normal ranges. Chest radiography (CXR) showed pneumopericardium). Serial ECG on the following days revealed improving ST-T changes without evidence of abnormal Q wave or inverted T waves in favor of myocardial infarction. Subsequent CXR showed also resolution of pneumopericardium. In our patient, normal myocardial biomarkers ruled out the diagnosis of ischemia.

**Conclusion:** In critically ill patients it is important to pay attention to nonischemic etiologies of ST elevation such as pneumopericardium especially in the presence of abnormal CXR and negative myocardial ischemic markers.

**Keywords:** Pneumopericardium, pneumomediastinum, ST segment

---
Silibinin inhibit proliferation of acute myeloid leukemia cells and induce differentiation into monocyte

Vahid Lesan
Department of Biology, Faculty of Food Industry and Agriculture, Standard Research Institute (SRI), Karaj, Iran

Background: The main involved component of silymarin, a standardized extract of the milk thistle seeds is called Silibinin. In vitro anti-cancer impacts on human prostate adenocarcinoma cells, both small and nonsmall human lung carcinoma cells, human ectocervical carcinoma cells, estrogen-dependent and -independent human breast carcinoma cells, and also human colon cancer cells, silibinin has been indicated. Acute myeloid leukemia (AML) accounts for 15-20% of childhood leukemias. Though remission is achieved by following treatment with front-line chemotherapy, almost half of the patients are faced with disease relapse associated with chemoresistance. Consequently, therapies that could maintain the remission phase in pediatric AML are urgently needed.

Methods: Cell viability and apoptosis were assessed trough MTT assay and flow cytometry respectively. The activity of PKC was calculated by using a commercially kit and the protein levels of PKC isoforms were determined by western blotting. The differentiation of HL-60 cell to monocyte was estimated by the NBT reduction assay and morphologic studies.

Findings: In this study it was demonstrated that silibinin inhibits proliferation and induces apoptosis in AML cells in dose dependent manner. The issue that silibinin caused differentiation of HL-60 cells predominantly into monocytes is proved by cytofluorometric analysis and morphologic surveys. Silibinin improves protein levels of both PKCa and PKCb in HL-60 cells and protein kinase C (PKC) activity. PKC and extracellular signal-regulated kinase (ERK) restrains significantly restrained HL-60 cell differentiation caused by silibinin, showing that PKC and ERK might be involved in silibinin-induced HL-60 cell differentiation. Finally silibinin could be a potent anticancer agent for targeting acute myeloid leukemias.

Key words: Silibinin, Leukemia, PKC, ERK, Apoptosis

Assessment of hypocalcemia in febrile convulsion

Khatereh Khamenehpour1, Fatemeh Saffari2
1 Children Growth Research Center, Qazvin University of Medical Sciences, Qazvin, Iran; 2 Metabolic Diseases Research Center, Qazvin University of Medical Sciences, Qazvin, Iran

Background: Hypocalcemia is the most common electrolytic cause of seizure and febrile convulsion (FC) is a frequent cause of hospitalization in pediatric population. The aim of this study was to assess hypocalcemia in children hospitalized with febrile convulsion in Qazvin, Iran.

Methods: This cross sectional study was conducted on 515 children (less than five years old) with febrile convulsion admitted at the Children hospital in Qazvin from March 2009 to March 2011. Age, personal and family history, temperature, symptoms and signs, duration and type of seizure, cause of fever, and the results of LP were recorded. Calcium and vitamin D levels were measured. Calcium level less than 8.2 mg/dl was defined as hypocalcemia. Data were analyzed using Mann-Whitney U test.

Findings: Mean age was 21.79±13.46 months. Of 515, 51.9% were female and 77 had complex febrile convolution. Hypocalcemia was present in 45 (8.7%) of the patients. Of 45, 17.8% had vitamin D deficiency, 57.8% had vitamin D insufficiency, and 4.44% had hypoparathyroidism. Mean duration of seizure was 6.38 minute in children with hypocalcemia. The duration of seizure was significantly higher in children with hypocalcemia than children without hypocalcemia (P<0.001). Age, temperature, type and number of seizures and length of hospitalization were not different between two groups.

Conclusion: Hypocalcemia was prevalent in children with febrile convulsion. Vitamin D deficiency was the most common cause of hypocalcemia. Vitamin D deficiency should be considered in the hypocalcemic FC work up.

Keywords: Febrile Convulsion, Hypocalcemia, Vitamin D Deficiency

First Determination of N-Acetylgalactosamine-6-Sulfate Sulfatase Activity on Leukocytes of Iranian MPS IV-A Patients

Sedigheh Shams1,2, Maliheh Barazandeh Tehrani1, Gabriel Civaliero1, Kosha Minokherad1, Roberto Giugliani3, Mohammad Taghi Haghi Ashtiani1,2, Neda Rezaei1, Heshmat Irani1
1. Pediatrics Center of Excellence, Children’s Medical Center, Tehran, Iran; 2. Department of Pathology, Tehran University of Medical Sciences, Tehran, Iran; 3. Pediatric Urology Research Center, Tehran University of Medical Sciences, Tehran, Iran; 4. Department of Medicinal chemistry, Faculty of Pharmacy and Pharmaceutical Sciences Research Center, Tehran University of Medical Sciences, Tehran, Iran; 5. Medical Genetics Service, Clinical Hospital of Porto Alegre, RS, Brazil

Background: Mucopolysaccharidosis type IV-A (Morquio A) is an inherited lysosomal storage disease caused by deficiency of N-acetylgalactosamine-6-sulfate sulfatase (GALNS). In the absence of the lysosomal enzyme, two major glycosaminoglycans (GAGs) such as keratan sulfate and chondroitin-6-sulfate are accumulated in the lysosomes leading to a tissue and organ dysfunctions. Skeletal and joint deformities, visual and auditory impairment, and tooth enamel defect, are common signs and symptoms of the disease. Cognitive and mental developments are normal. From February 2014, treatment of MPS IV-A with enzyme replacement therapy (VIMIZIM™, elsulfase alfa, BioMarin) was approved, being crucial an accurate and early diagnosis of the disorder to prevent long term damage and effectiveness of therapy.

Laboratory diagnosis of MPS IV-A includes qualitatively or quantitative determination of elevated GAGs in urine, measuring of GALNS activity and molecular study of defected gene. For the first time in IRAN, we set up a previously described protocol for testing GALNS activity
in leukocytes in Iranian Morquio A patients and healthy control groups. **Methods:** Leukocytes were obtained from heparinized blood of healthy individuals and patients with MPS IV-A whose diagnosis was previously confirmed with enzyme activity measurement or molecular study. Informed consent was obtained from the subjects or their parents prior to sample collection. GALNS activity was measured by a fluorometric method using 4-methylumbelliferyl-β-galactoside-6-sulfate as substrate, and appropriate buffer solutions and calibrators. **Findings:** The GALNS activities were undetectable in MPSIV-A patients and 72-182 μmol/17h/mg of protein in healthy individuals. Statistical significant differences in activity of enzyme were seen in all confirmed MPS IV-A patients compared to the healthy subjects. **Conclusions:** We have set up a fluorometric method for measuring GALNS activity in leukocytes samples to identify Iranian MPS IV-A patients. **Acknowledgment:** We would like to thank all physician and their patients who provided specimen for the study. **Keywords:** Mucopolysaccharidosis type IV-A, Morquio, glycosaminoglycan, Keratan sulfate

**Epidemiological and clinical data of diabetes type1 hospitalized patients, in Children’s Medical Center, Iran**

Fatemeh Sayarifard¹, Dorsay Sadeghian¹, Azadeh Sayarifard², Aria setoodeh³, Moloo Safari Rad¹

1. Growth and development research center, Children’s Medical Center, Tehran University of Medical Sciences, Tehran, Iran; 2. Preventive and Community Medicine, Tehran University of Medical Sciences, Tehran, Iran; 3. Children’s Medical Center, Tehran University of Medical Sciences, Tehran, Iran

**Background:** Diabetes is one of the most important endocrine and metabolic disorders. Regarding the incongruous reports on epidemiologic aspects of this disease from different parts of the world, it is necessary to assess this problem in our country. We decide to evaluate epidemiologic and clinical data in diabetic patients referring to Children’s Medical Center in Tehran, during ten years period. **Methods:** In this cross-sectional survey conducted in 2013-2014, data on demographics characteristics and medical history of the diabetic patients referring to Children’s Medical Center in Tehran during 10 years period from 2004 to 2014 were gathered from the patients’ records. Data were entered into SPSS v.16 software and then were analyzed. **Findings:** In this study, 1303 diabetic children were included, comprised of 540 (41.4%) boys and 763 (58.6%) girls. The mean age of patients was 8 yrs. The first presentation of patients was polyuria and polydipsia (48%), diabetic ketoacidosis (42.3%) and nonspecific (9.7 %). Most of patients were born in summer. There was no difference in seasonality of birth between diabetic patient and other patients in this center. The average number of admissions were higher in children with family history of diabetes type1 and history of other autoimmune disorders than the other patients (p value 0.019 & 0.001 respectively). The mean age of onset of disease in these groups of patients were significantly lower compared to the rest of the patients (p value<0.001). **Conclusion:** It seems that Family history and history of autoimmune disorders, have some influence on earlier onset of disease, increasing of incidence of DKA and number of hospitalizations. **Key words:** Children, Diabetes, Epidemiology, Autoimmune

**Case Report: Congenital generalized lipodystrophy (Berardinelli Seip syndrome)**

Azad A. Haleem Al Mezori¹, Hasan Balik², Farhad Salih Hussein³

1. University of Duhok/Kurdistan/Iraq; 2. Pediatric specialist in Diyarbakir/Turkey; 3. Heevi Pediatrics Teaching Hospital, Duhok/Kurdistan/Iraq

Congenital generalized lipodystrophy (CGL, Berardinelli-Seip syndrome) is a rare autosomal recessive disorder with a primary genetic alterations in fat development resulted in clinical presentation of paucity of adipose tissue (lipodystrophy), muscular hypertrophy, organomegaly, dyslipidemia, insulin-resistant diabetes and hepatic steatosis. A 15 - month-old Kurdish female her clinical features, serum biochemistry and liver radiology were compatible with the diagnosis of CGL. We report a case of CGL which, to the best of our knowledge, is the first Kurdish child reported with this rare Syndrome. **Keywords:** lipodystrophy, dyslipidemia, Berardinelli Seip syndrome

**Clinical, laboratory data, molecular features and outcome of 16 Iranian citrulinemia type 1 patients**

SH. Moarefian¹, A. Rahmanifar¹, B. Belnam⁴, T. Zaman¹,²

1. Iranian National Society for Study on Inborn Metabolic Diseases, Tehran, Iran; 2. Metabolism.Department of Inborn Metabolic Disease, Faculty of Medicine, Tehran University of Medical Sciences, Tehran, Iran; 3. Department of Medical Genetics and Molecular Biology, Faculty of Medicine, Iran University of Medical Sciences, Tehran, Iran

**Background:** Citrulinemia type1 (CTLN1) is a rare autosomal recessive disorder caused by defect in ASS1 gene which varies among ethnic groups and shows very heterogeneous manifestations. The phenotypic diversity is explained partly by different residual enzyme activity due to different mutations, however most phenotypic variations remains unexplained. We aimed to determine the clinical, molecular characteristics and outcome of citrulinemia type1 in Iranian patients. **Methods:** This is a descriptive study on 16 CTLN1 patients (2008-2014). Sequence analysis of ASS1 gene confirmed the diagnosis. Followe up after standard & emergency treatment for CTLN1 was done. Results:16 patients:9 boys &7 girls:9 **Findings:** 7 consanguineous.13/16 (78%)early onset neonatal; 2/16(14%) late onset infantile; 1/16(7%) asymptomatic. Neonatal forms:8/13(54%) died at mean age of 6.5±7.8 days;2/13(18%) died in a hyperammonemic attack at mean age of 6.75±4.5 months of infancy;3/13(27%) are now well after mean follow up of 3.26±1.4 years with mild speech impairment. Two siblings one died at 17 years old without therapy with seizure and developmental delay; the other treated for 3 years is now well but mild speech delay. One of the cases is still asymptomatic after 6 years follow up. We have reported 11 mutations in 32 mutant alleles that except 2 previously reported the others were novel in Iranian population.
Conclusion: Except two, the other ASS1 gene mutations were novel in Iranian patients and prognosis is determined by mutation type, early diagnosis and ammonia control but not citrulline level.

Keywords: citrulinemia, hyperammonemia, Gentic study

Adrenocortical carcinoma: A case report

F. Tabatabaei, MD; MR Sharif, MD
1. Isfahan Endocrine & Metabolism Research Center, Isfahan University of Medical Sciences, Isfahan, Iran; 2. Trauma Research Center, Kashan University of Medical Sciences, Kashan, Iran.

Background: Adrenocortical carcinoma (ACC) is a rare neoplasm with a slight predilection for female patients, with an incidence of about one case per million population. They have a bimodal peak; the first one is in the fourth and fifth decades of life and the second one in the first decade. About 60% are functional tumors that secrete hormones and present with clinical features like Cushing syndrome due to cortisol, virilizing tumor due to androgens, or feminizing tumor due to estrogens. ACC in children appears to behave differently than that in the adult patients. Virilization is more frequently seen and has a better prognosis after complete resection than in adults. The overall 5-year survival rate ranges from 16% to 38%. Recurrence, even after seemingly complete resection, is common, occurring in 23% to 85% of patients. Death usually occurs in the first 2 years. Here in, we report a case of ACC.

Case presentation: She was of a 9-year-old girl who presented with clitoromegaly. The clinical, biochemical, histological features along with differential diagnosis are discussed. This case is presented because of the rarity, and also to highlight the importance of differentiating ACC from an adenoma particularly in pediatric patients.

Conclusion: ACC is an extremely rare neoplasm and particularly if it occurs in children, it is essential to differentiate it from an adrenocortical adenoma by correlating with clinical, biochemical, imaging, and histological features, because their prognoses are different.

Keywords: Adrenocortical carcinoma, Cushing’s syndrome, clitoromegaly

Bone Densitometry in Children and Adolescents: Indications, Interpretation, and its Risks

Sina Izadyar, M.D.
Associate Professor of Nuclear Medicine-Imam Khomeini Hospital Complex-Tehran University of Medical Sciences-Tehran-IRAN

Background: The bone health of children and adolescents has become an increasingly important medical concern. Pediatric patients with genetic and acquired chronic diseases, immobility, and inadequate nutrition may fail to achieve the expected gains in bone size, mass, and strength, which leave them vulnerable to fracture. The role of densitometry in the management of children at risk of bone fragility is less certain. The aim of this review article is to summarize the current knowledge about bone densitometry in the pediatric population, including indications for its use, interpretation, and its risks.

Methods: Data were collected using the PubMed database, as well as the American Academy of Pediatrics, The National Osteoporosis Foundation and the International Society for Clinical Densitometry publications.

Findings: Dual-energy X-ray absorptiometry (DXA) is the preferred method for the assessment of bone mineral density in children and adolescents. Indications for DXA in children: Primary bone disorders, potential secondary bone diseases, chronic immobilization, endocrine disturbance, cancer and therapies with adverse effects on bone health, post-transplantation, hematologic disorders, exposure to bone-toxic drugs (glucocorticoids, anticonvulsants), recurrent low-trauma fractures, and apparent osteopenia on radiographs. The preferred skeletal sites for DXA measurements in children are the lumbar spine and whole body. The hip is not a preferred measurement site in growing children due to variability in skeletal development. Bone mass, as measured by DXA, is reported as bone mineral content (BMC) (g) or areal BMD (g/cm²). These values are compared with reference values from healthy youth of similar age, gender, and if possible, race to calculate a Z-score, the number of SDs from the expected mean. ’Low bone mineral mass or bone mineral density’ is the term for pediatric DXA reports when BMC or areal BMD Z-scores are less than or equal to -2.0 SD. The diagnosis of osteoporosis in children is made only when both Z-scores of less than -2 and a clinically significant fracture history are present. If a follow-up DXA scan is indicated, the minimum interval between scans is 6-12 months. Exposure to ionizing radiation with DXA poses no known health risk. The estimated 5 to 6 microsieverts of radiation exposure from a spine and whole-body DXA scan is very low.

Conclusion: In the children or adolescent with a potential for fractures because of an underlying cause or a significant fracture history, clinicians now have guidelines and normative data to better focus their evaluation. Likewise, researchers can use this information to improve clinical trial design and interpretation of results.

Keywords: pediatrics, DXA, bone densitometry, osteoporosis

Berardinelli-Seip Syndrome - Case Report

Azad A. Haleem (Lecturer in University of Duhok, College of Medicine/Duhok/Kurdistan/Iraq)

We have reported a case of Berardinelli-syndrome in a female child now 15 months old born from non consanguineous parents. presented to us during neonatal periods with lethargy, poor feeding and failure of gaining weight. All investigations regarding septic screening were normal. The clue for diagnosis was turbid serum, which revealed hyperlipidemia (Serum cholesterol 596 mg/dl, triglycerides 2100 mg/dl) treated by Omega 3, plasma and skimmed milk. On follow up the infant have abnormal facial features and gradually increasing dark discoloration of skin, coarse facies, Generalized loss of subcutaneous fat, prominent musculatures over face and limbs, abdominal distension, hepatosplenomegaly, ultra sound of abdomen revealed fatty liver, now she is on Multivitamin, nutritional follow up and Lipid lowering agent. Congenital generalized lipodystrophy (Berardinelli-Seip syndrome) is a rare autosomal recessive disorder with primary genetic alterations in fat development resulted in lipodystrophy, dyslipidemia, diabetes and hepatic steatosis.

Keywords: Berardinelli, Seip Syndrome, hyperlipidemia
**Pyrimethamine for infantile GM2 gangliosidosis**

S. Salehpour,1 SH. Tonekaboni,1 F. Pazhouhandeh,1 M. Houshmand,2 O. Aryani3, VR Yasaee1, S. Setavand1, P. Karimizadeh4, AR Rezaei5, N. Montazmamesh6, S. Setavand, E. Sakhueian5

Genomic Research Center & Mofid Children Hospital, Loghman Hakim Medical Center, Shahid Beheshti University of Medical Sciences, Tehran, Iran; 2. National Institute of Genetic Engineering & Biotechnology, Tehran, Iran

**Background:** GM2 gangliosidosis is due to β-hexosaminidase deficiency causing large accumulation of GM2 ganglioside in brain. No enzymatic replacement therapy is currently available. The inhibitors of glycosphingolipid biosynthesis for substrate reduction therapy are associated with serious side effects. Recently the chaperone effect of pyrimethamine on the recovery of β-hexosaminidase activity in cultured fibroblasts of late onset -Sandhoff patients was shown. The aim of this study is to evaluate the effect of oral pyrimethamine on infantile cases of GM2 gangliosidosis. Methods: Nine children with infantile GM2 gangliosidosis were treated with pyrimethamine, started with 6.25 mg once daily and gradually increased to 25 mg twice daily. Improvement was evaluated by developmental quotient (Bayley’s scale-version III), or β-hexosaminidase activity in leukocytes. Results: Hexosaminidase B activity in 5, and Hexosaminidase A and A-B in 3 patients increased (p<0.01). Six patients showed significant improvement in developmental quotient. Seizure rates increased in 3 patients either for pyrimethamine or the progression of disease itself. Conclusion: Pyrimethamine may preserve the neurodevelopment of children with infantile GM2 gangliosidosis by increasing the activity of hexosaminidases.

**Keywords:** pyrimethamine, Gm2 gangliosidosis

---

**The role of neonatal screening in the detection and treatment of thyroid disease**

Massomeh Shahangui, Fatemeh Askari, Somayeh Dab, Seyyed Reza Heidari, Azam Yazdai

Sarakhs Medical Center, Sarakhs, Iran

**Background:** Neonatal Hypothyroidism disease is condition of thyroid hormone deficiency can be treated Which if not diagnosed early or inappropriate treatment can cause severe retardation and impaired growth and a heavy burden on the family and society. With neonatal screening and identification and treatment of patient the serious disease complication not occur and with maintence of normal IQ, production and healthy person healthy to be delivered to the community . The incidence of hypothyroidism is 1 in every 3000 to 4000 live birth in the world . The aim of this study was to evaluate the neonatal screening program to detect and treat the disease in last 5 years from 2010 to 2014 in Serakhs city, Iran.

**Methods:** this is a descriptive study , from all children born since the beginning of 2010 up to the end of 2014 were enrolled in Serakhs city.

**Findings:** All neonatal in this study were 11559 that during the study with Serum test about 40 children were taken off treatment. Conclusion: Due to the high incidence of the disease in compare with world-wide rate it needs more study and the project has successful in identifying and controlling the patient treatment and prevention of complications.

**Keywords:** Hypothyroidism, screening, serum test

---

**Genetic Testing for Metabolic and Endocrine Disorders**

Nejat Mahdieh, PhD; Bahareh Rabhani, PhD

Heaet Genetic Research Center of Shahid Rajaei

Clinical molecular diagnostics has grown rapidly during the last decade. Endocrine and metabolic disorders are often quite complex, involving a mixed picture of hyposecretion and hypersecretion; because of the feedback mechanisms involved in the endocrine system. Some endocrine and metabolic disorders show familial patterns of inheritance, while others may occur due to sporadic mutations. In both (familial and sporadic) cases, finding the exact genetic cause of disease improves understanding of the underlying mechanism provides clear explanation to families about the disease, guide decisions about screening, prevention and/or treatment, and allows the physician to test other family members, who may be asymptomatic. For example, in the case of a mutation with potential for adverse outcome such as medullary thyroid cancer; Physician and patient can now consider prophylactic thyroidectomy, for harboring such a gene prior to actual presentation of clinical disease; an approach possible only with the endocrine “genetic” revolution. Knowledge of which genetic tests to order must be supported by a full understanding of the genetic information they provide. The health care team should include physicians and a geneticist/genetic counselor. Here, a clinically relevant review of important genetic tests currently in use for the diagnosis of endocrine and metabolic disorders is presented as well as practical information as to where these tests are performed.

**Keywords:** genetics, endocrine disorder, mutation, genetic testing

---

**Clinical application of next generation sequencing for pediatric disorders; focusing on metabolic and neurological disorders**

Nejat Mahdieh, PhD; Bahareh Rabhani, PhD

Heaet Genetic Research Center of Shahid Rajaei, Tehran, Iran

With advance in sequencing technology, our understanding of the genetic basis of diseases has increased rapidly over the past years. Identification of causal gene mutations may lead clinicians to confirm diagnosis and better management of patients. Genomic sequencing tests provide multiple analyses of genes and variants over short period and its cost benefits in comparison with classic sequencing methods. Here we provide an overview of NGS tests, results and reporting in pediatric genetic disorders; exploring diagnostic utility of genome sequencing and its use in translational medicine. Complexity of phenotypes and
heterogeneity of disease are of criteria of NGS utility. Of course, with the help of all these technologies, better management of the patients and families would be available if counseled properly.

**Keywords:** next generation sequencing, sequencing, pediatric disorders, mutation, clinic, diagnosis

**Motor development skills of 0-29 month’s children with phenylketonuria in Hamadan city**

Saeidh Sadat Mortazavi, Shakiba Gajfari
Hamadan University of Medical Science, Hamadan, Iran

**Background:** Phenylketonuria is one of the most common autosomal recessive inherited metabolic diseases. Though apparently normal birth, but gradually assorted children are developmental disorders. Since few studies in the field of children’s motor skills Phenylketonuria (PKU), We decided to research as compared with gross motor skills and fine children with phenylketonuria and normal children 29-0 months in Hamadan.

**Methods:** This study is cross-sectional analytic study on 10 children with PKU and 10 healthy children 29-0 month period in Hamadan. Children phenylketonuria census sampling and random sampling was performed in healthy children. After obtaining permission from the university ethics committee and completing the consent form by the families of the children were assessed by Peabody development motor scale and pediatrician assessment and compared with independent samples t-test.

**Findings:** The mean ages in case and control group were 24.5 (±5.6). Fine motor skills of children Phenylketonuria (PKU) is weaker than healthy children (p<0.01) gross motor skills don’t show statistically significant differences between the two groups. In sub test , balance of gross motor skills and hand-eye coordination of fine motor scale is weaker than healthy children. (p<0.01)

**Conclusion:** The result showed that screening and following acurrent diet treatment have not completely reduced the motor delay and have had lower motor development than normal children. parents and professionals offer rehabilitation circumstances, occupational therapists have special attention to this issue

**Keywords:** Motor development, Phenylketonuria, children

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

Morteza Mearadji the Netherlands

Pediatric metabolic bone diseases A classification and an overview of clinical and radiological findings M. Mearadji International Foundation for Pediatric Imaging Aid The metabolic bone diseases affecting bone formation and mineralization can be etiologically classified in four categories. Endocrine bone diseases and skeletal dysplasia are excluded from this topic. A. Disorders with insufficient mineralization organic matrix: This group contains all different types of rickets associated with vitamin D abnormalities, such as nutritional deficiency and other similar conditions. Also hypophosphatasia as a rare inherited disease with deficiency of serum and bone alkalinephosphatase activity (Phosphopenic rickets) can be classified in this group. B. Abnormalities of bone matrix (osteoid) formation: Osteoid functioning as organic framework for mineral deposition. An example of such metabolic bone disorder is osteogenesis imperfecta, with signs and symptoms of osteopenia and bone fragility, due to defective matrix formation. Menkes disease (copper deficiency) and shortage of vitamin C, which are both associated with deficiency of matrix. C. Diseases associated with increased or decreased bone resorption: The major cause of increased bone resorption in children is secondary hyperparathyroidism, a common finding in chronic renal failure osteodystrophy. Instead, in osteopetrosis, the bone resorption is decreased. In this abnormality the failure of osteoblasts in bone resorption results in an increased bone mass. D. Pharmacologic and toxic changes of skeleton: The bone metabolism can be affected exogenic by some drugs, such as biphosphonate, which are usually used in treatment of osteoporosis and osteogenesis imperfecta. This is based on potent inhibitory effects of biphosphonate on bone resorption. Hyperostosis of the long bone as an adverse reaction of Prostaglandine 1 therapy which is used in pediatric cardiology. With lead poisoning, an increased density of growing metaphysis is characteristic for toxic effect on skeleton. Systemic oxalosis with deposition of calcium oxalate on tubular bone is an endogenic metabolic disease involving not only bone, but other organs as well. In this presentation clinical and radiological findings of a large number of metabolic bone disorders will be shown and discussed with special attention on etiology and differential diagnosis.

**Keywords:** Pediatric metabolic bone disorders

**A nationwide report on blood pressure of children and adolescents according to socioeconomic status: the CASPIAN-IV study**

Zahra Fallah1, Roya Kelishadi2, Mohammad Esmaeil Motlagh2, Gelayol Ardalan2, Ramin Heshmat3, Amir Kasaeian1,4, Hamid Asayesh5, Mostafa Qorbani5

1. Department of Pediatrics, Child Growth and Development Research Center, Research Institute for Primordial Prevention of Non-communicable Disease, Isfahan University of Medical Sciences, Isfahan, Iran; 2. Department of Pediatrics, Ahvaz University of Medical Sciences, Ahvaz, Iran; 3. Department of School Health, Bureau of Population, Family and School Health, Ministry of Health and Medical Education, Tehran, Iran; 4. Chronic Diseases Research Center, Endocrinology and Metabolism Population Sciences Institute, Tehran University of Medical Sciences, Tehran, Iran; 5. Department of Biostatistics & Epidemiology, School of Public Health, Tehran University of Medical Sciences, Tehran, Iran; 6. Non-communicable Diseases Research Center, Endocrinology and Metabolism Population Sciences Institute, Tehran University of Medical Sciences, Tehran, Iran; 7. Department of Medical Emergencies, Qom University of Medical Sciences, Qom, Iran; 8. Department Epidemiology, School of Public Health, Alborz University of Medical Sciences, Karaj, Iran

**Background:** Hypertension is a major leading factor for global burden of diseases. Blood pressure (BP) tracks from childhood to adulthood. So it is important to investigate its affecting factors. Aim: To compare the BP status in the Iranian pediatric population according to the socioeconomic status (SES) of their living area. Setting and design: A nationwide survey.

**Methods:** A representative sample of 14880 students, aged 6-18 years was selected by multistage random cluster sampling from 30 provinces in Iran. Anthropometric
indices and BP were measured. A validated questionnaire including the questions of the World Health Organization Global School-based student Health Survey (WHO-GSHS) was completed. Findings were compared across the four regions of the country, categorized based on their elevating SES: Southeast, North-northeast, West and Central.

**Findings:** Participants consisted of 13486 children and adolescents, i.e. a participation rate of 90.6%, consisting of 49.2% girls and 50.8% urban residents. The mean (SD) age of participants was 12.47 (3.36) years. The region with highest SES (Central) had the lowest rate of high blood pressure (HBP), i.e. 3.0% (95% confidence interval: CI)12.4-3.9), and the region with lowest SES (Southeast) had the highest rate, i.e. 7.4% (4-12.2). The mean (95% CI) values of systolic BP (SBP) for the four regions from lowest to highest SES were 100.5(99.6-101.3), 100.9(100.3-101.4), 101.7(101.3-102) and 101.7(101.2-102.1) mm Hg. The corresponding mean Diastolic BP (DBP) values were as follows: 65.4(64.6-66.1), 63.4(62.9-63.8), 65.6(65.3-65.8) and 64.4(64.0-64.7) mm Hg. **Conclusion:** We found significant differences in mean BP and the frequency of HBP according to the SES of the living area. Further studies are necessary to find the underlying factors resulting in such differences.

**Keywords:** Blood pressure, high blood pressure, child, adolescent, socioeconomic status

---

**No Association between Nodular Thyroid Disease with rs1256049 Polymorphism of Estrogen Receptor 2 Gene in Women From Markazi Province**

Maryam Yousefi, Ahmad Hamta, Abdorrahim Sadeghi, Afsoon Talaei
1. Department of Biology, Arak University; 2. Molecular and Medicine Research Center, Department of Biochemistry and Genetics, Arak University of Medical Sciences; 3. Endocrine and Metabolism Research Center, Arak University of Medical Sciences, Arak, Iran

**Background:** Thyroid nodules are common, so that 4-7% of adults have a palpable nodule and up to 50-70% of nodules are detected in high-resolution ultrasound. Thyroid nodules in women are 4 times greater than men and thyroid cancer in women compared to men is ratio 3:1, and is the sixth most common cancer in women. Epidemiological findings and experimental evidences of thyroid damages, suggesting that sex hormones, especially estrogen, may have effect on this gland and its neoplasm. The aim of this study was to investigate the association between rs1256049 polymorphism in the estrogen receptor 2 gene with thyroid nodule disease.

**Material:** In this case-control study, 146 Patients with nodular thyroid and 151 health individuals were referred in Amiralmomenin hospital Arak, Iran. They were recruited in study. Diagnosis is based on by ultrasoundography and were confirmed by an endocrinologist. Genomic DNA was extracted from EDTA treated whole blood. The genotypes were determined using tetra-amplification refractory mutation system-polymerase chain reaction (T-ARMS-PCR) and analyzed by statistical methods.

**Findings:** The frequency of CC, TC and TT in case group 136(93.2%), 10(6.8%) and 0(0%) and in the control group 139(92.1%), 12(7.9%) and 0(0%) was obtained respectively. No statistically significant association (P=0.72) between disease and rs1256049 polymorphism was observed.

**Conclusion:** Our findings showed no significant association between rs1256049 polymorphism and nodular thyroid disease.

**Keywords:** Estrogen receptor 2, Polymorphism, rs2987983, rs1256049, Thyroid nodule

---

**Comparison of intellectual and developmental status in children with hyperphenylalaninemia and PKU with normal population**

Parisa Aghasi1, Aria Setoodeh1,2, Azadeh Sayarifard3, Maryam Rashidian4, Fatemeh Sayarifaru5, Ali Rabbani1,2, Javad Mahmoudi Gharei1,2
1. Children’s Medical Center, Tehran University of Medical Sciences, Tehran, Iran; 2. Growth and Development Research Center, Tehran University of Medical Sciences, Tehran, Iran; 3. Preventive and Community Medicine, Tehran University of Medical Sciences, Tehran, Iran

**Background:** Hyperphenylalaninemia (HPA) and PKU is a metabolic error caused by deficiency of the phenylalanine hydroxylase enzyme, which results increasing level of phenylalanine, this increase is toxic to the growing brain. The purpose of this study was to compare the intellectual and developmental status in children with HPA and PKU with normal population in national screening program referred to Children’s Medical Center.

**Methods:** In this study, 41 PKU patients and 41 healthy children were studied. Wechsler test was used in order to assess the intellectual status of children 4 years and older and Ages and Stages Questionnaire (ASQ) was used to assess the developmental status of children 5 years and younger.

**Findings:** In intellectual test comparison, two groups showed significant difference in Wechsler’s performance intelligence score and some performance subscales (P<0.01). In comparison of developmental status, no significant difference was observed in two groups (P> 0.05).

**Conclusion:** Even with early diagnosis and treatment of PKU patients, these children show some deficiencies intellectually compared to normal children. This study emphasized on necessity for screening intellectual and developmental status of PKU patients so that effective medical or educational measures are taken in case of deficiencies.

**Keywords:** Hyperphenylalaninemia, PKU, intelligence, development

---

**Diabetic ketoacidosis follow to L-Asparginase therapy in acute lymphoblastic leukemia.**

Nadereh Tare, Majgun Faraji Goudarzi, Hamid-reza Sherkatolabbasieh Lorestan University of Medical Sciences, Khoramabad, Iran

**Background:** Diabetic ketoacidosis is rare complication of L-Asparginase therapy in acute lymphoblastic leukemia (ALL). Nearly 10% of all patients that receiving L-Asparginase may develop mild hyperglycemia that resolve after few days, but severe hyperglycemia and Diabetic ketoacidosis is very rare. We present the case of acute lymphoblastic leukemia and severe hyperglycemia and
Diabetic ketoacidosis following to L-Asparaginase therapy. Case presentation: 12 years-old female admitted to Madany children hospital for severe abdominal pain, nausea, vomiting, severe weakness and unexplained dehydration. She was known case of ALL 10 months ago that received L-Asparaginase for chemotherapy 7 days ago. On the time of hospitalization she had severe respiratory distress (RR =60), tachycardia (PR=180), blood pressure 90/60mm/Hg and temp=37.9°C. In physical examination severe dehydration, severe abdominal pain and drowsiness were presented. In Lab tests PH =6.74, Hco3=21.1 mmol/L, Pco2=15.4 mm/Hg, Hemoglobin 14.2g/dL with 80% neutrophil. blood glucose =319mg/dL, blood urea nitrogen 29 mg/dL, creatinine 0.5 mg/dL, sodium 131 mmol/L and potassium 4.9 mmol/L. Urin analysis showed ketonuria +2, glucosuria+2 with 1025 specific gravity. The patient was managed for Diabetic ketoacidosis. Fluid therapy and insulin infusion were done and wide spectrum antibiotic started. Finally she was better and discharged after 6 days with insulin injection.

**Conclusion:** hyperglycemia is known complication of L-Asparaginase but it usually resolves 2 weeks after last dose. Mechanism of hyperglycemia may be inhibition of insulin synthesis, secretion and impaired insulin receptors. We concluded that glucose monitoring regularly is necessary in patients receiving L-Asparaginase.

**Keywords:** L-Asparaginase, Diabetic ketoacidosis, acute lymphoblastic leukemia

**Association between rs2987983 polymorphisms in the estrogen receptor 2 gene with thyroid nodular disease**

Maeryam Yousefi1, Ahmad Hamh1, Abdorrahim Sadeghi2, Afanesh Taleae2

1. Department of Biology, Arak University, Arak, Iran; 2. Assist. prof. Molecular and Medical Research Center, Department of Biochemistry and Genetics, Arak University of Medical Sciences, Arak, Iran; 3. Endocrine and Metabolism Research Center, Arak University of Medical Sciences, Arak, Iran

**Background:** The thyroid is a butterfly-shaped gland that located in the lower front of the neck. Thyroid nodules are common, so that 4.7% of adults have a palpable nodule and up to 50-70% of nodules are detected in high-resolution ultrasound. Thyroid nodules in women are 4 times greater than men. Epidemiological findings and experimental evidences of thyroid damages, suggesting that sex hormones, especially estrogen, may have effect on this gland and its neoplasm. The aim of this study was to investigate the association between rs2987983 polymorphisms in the estrogen receptor 2 gene with thyroid nodular disease was determined.

**Methods:** In this case-control study, 152 Patients with nodular thyroid and 158 health individuals were referred in Amiralmomenin hospital Arak, Iran were recruited in study. Diagnosis is based on by ultrasonography and were confirmed by an endocrinologist. Genomic DNA was extracted from EDTA treated whole blood. The genotypes was determined using tetra-amplification refractory mutation system-polymerase chain reaction (T-ARMS-PCR) and analyzed by statistical methods.

**Findings:** The frequency of rs2987983 genotypes AA, AG and GG in case group 51(33.6%), 86 (56.6%) and 15(9.8%) and in the control group 67(42.4%), 67(42.4%) and 24(15.2%) was obtained respectively. Statistically significant association (0.039) between disease and rs2987983 polymorphisms was observed.

**Conclusion:** Regarding to results we could be suggested rs2987983 polymorphism as a diagnostic biomarker. Since this polymorphism is located in the promoter region of the gene, probably the effect of this polymorphism is the change in the level of gene expression. Regarding to association between this polymorphism with nodular thyroid disease reported for first time in the Iran, it is recommended to be done in other populations and a larger sample size.

**Keywords:** Thyroid nodule, rs2987983 Polymorphism, Estrogen receptor 2

**Congenital hypothyroidism: incidence, etiology, risk factors, and genes.**

Seteila Dalili1, Shahn Koolhmane2, Arvand Akbari2, Zivar Saheli2

1. Pediatrics Growth Disorders Research Center, 17 Shahriar Hospital, School of Medicine, Guilan University of Medical Sciences; 2. Department of Genetics, International Campus, University of Guilan; 3. Department of Genetics, Faculty of Sciences, University of Guilan, Rasht, Iran

**Background:** Congenital hypothyroidism (CH) occurs in approximately 1:2,000 to 1:4,000 newborns. The clinical manifestations are often subtle or not present at birth. This is likely due to placental passage of some maternal thyroid hormone. Delayed diagnosis and early treatment influence the outcome of CH. We aimed to assess congenital hypothyroidism: incidence, etiology, risk factors, and genes.

**Methods:** In this study investigators reported the results obtained from their previous investigations on congenital hypothyroidism. Good-enough tests was applied to evaluate the outcomes. Also, for gene study, Fifty patients with congenital hypothyroidism and fifty healthy controls were genotyped for FOXE1 283 G˃A and PDE8B rs4704397 using the polymerase chain reaction- restriction fragment length polymorphism(PCR-RFLP) and allele specific PCR (AS-PCR).

**Findings:** In our research performed in Shiraz, congenital hypothyroidism noted in 1:1465 children with a female to male ratio of 1.19:1. Also, prolonged jaundice (73%), large anterior fontanel (56%) and wide posterior fontanel (55%) were the most common clinical findings in these patients. Dyshormonogenesis (57%) was the most common etiology of permanent CH and thyroid receptor blocking anti body (TRBAb) was found in 6.8% of patients.Low Birth Weight (LBW), postdate delivery. Normal Vaginal Delivery and macrosomia were more prevalent in patients with CH. Also, iodine was assessed and based on results, Guilan province can be classified as a none-IDD endemic area. Permanent and transient hypothyroidism were noted in 43.2% and 56, 8 %, respectively. In patients with permanent CH, %68.2 and %31.2 had dyshormonogenesis and thyroid dysgenesis, respectively. Demographic characteristics and intelligence quotient noted no statistical difference in patients. results showed no significant role of FOXE1 283 G˃A and PDE8B rs4704397 on congenital hypothyroidism.

**Conclusion:** according to results, it seems that considering the mentioned risk factors and online diagnosis can be helpful. Also, larger population-based studies are needed to clarify the role of FOXE1 and PDE8B polymorphism in congenital hypothyroidism.

**Keywords:** congenital hypothyroidism, incidence, Gene polymorphism, FOXE1, PDE8B
Abetalipoproteinemia case reports and literature review

Shahab Noorian, MD; Alborz Medical Science, Bahonar Hospital, Karaj, Iran

Abetalipoproteinemia is a very rare (<1:1000,000) autosomal recessive metabolic disorder that prevents from normal absorption of dietary fats. Mutations in the MTP gene encoding the MTP (Microsomal triglyceride transfer protein) cause the disease. This protein has an important role in the transfer of lipids on to apo B and its absence compromises the transport of absorbed fats into the lymphatic system and the general circulation. Disease is usually revealed during early childhood by steatorrhea and failure to thrive. In later childhood ataxia and retinitis pigmentosa will be appeared. Here a 19-month-old patient with Abetalipoproteinemia is presented. He was pale, and had a bulging abdomen. His weight and height were below the 5th percentile, The child’s appearance and severe failure to thrive pushed the physicians to look for possible many metabolic abnormalities, so many additional diagnostic and laboratory testing were performed for him at the time of the visit, but nobody could to noticed his problem. when his mother reported that he had foul-smelling stool from the first month of life, Microscopic examination of the stool revealed steatorrhea and confirmed this fact. In this regard extremely low plasma lipids levels, absence of beta-band in lipoprotein electrophoresis and hematologic manifestations of ABL include acanthocytosis were detected.

Conclusion: If your patient have any symptoms of ABL, for example abnormal growth patterns and fatty and frothy stools, you can think to abetalipoproteinemia. Remember ABL is treatable, but treatment delays can have lasting effects.

Keywords: Abetalipoproteinemia; steatorrhea; failure to thrive; APO B

Mutation analysis of androgen receptor gene: multiple uses for a single test


Iran University of Medical Science, Tehran, Iran

Background: Androgen receptor gene mutations are one of the leading causes of disorders of sex development (DSD) exhibited by sexual ambiguity or sex reversal.

Methods: In this study, 2 families with patients whom diagnosed clinically as androgen insensitivity syndrome (AIS) were physically and genetically examined. This evaluation carried out by cytogentic and molecular analysis including karyotype and sequencing of SRY and AR genes.

Results: In family 1, two brothers and their mother were hemizygous and heterozygous respectively for c.2522G>A (rs6152) mutation in two siblings who were reared as girls. The SRY gene was intact in all of the study’s participants. Our findings in family 1 could be a further proof for the pathogenicity of the c.2522G>A variant.

Conclusion: Given the importance of AR mutations in development of problems such as sex assignment in AIS patients, definitive diagnosis and phenotype-genotype correlation could be achieved by molecular genetic tests that in turn could have promising impacts in clinical management and also in prenatal diagnosis of prospect offspring. In this regard, phenotype-genotype correlation could be helpful and achieved by molecular genetic tests. This could influence the clinical management of the patients as well as prenatal diagnosis for the prospective offspring.

Keywords: Mutation, androgen receptor, disorders of sex development

G-genes predisposes type 1 diabetes

Shahin Kooshmanae, Arvand Akbari, Zivar Salehi

Rasht, Iran

Background: Oxidative damage can lead cells to apoptosis which is believed to be the main cause of pancreatic β-cell death and eventually ending up to the development of type 1 diabetes (T1D). Glutathione S-transferase enzymes (GSTs) play a crucial role in counteracting reactive oxygen species (ROS). In this study, we tried to evaluate the association of three well-known polymorphisms of GSTM1, GSTT1 and GSTP1 in the pathogenesis of T1D which are whole gene deletions in GSTM1 and GSTT1 and a single nucleotide polymorphism (SNP) in GSTP1 known as Ile105Val.

Methods: Samples were collected from 159 patients diagnosed with T1D and 210 control subjects. Genotyping for GSTM1 and GSTT1 was performed by Multiplex PCR and by PCR-RFLP for GSTP1.

Findings: The GSTM1 and GSTT1 null genotypes were present at frequencies of 54 % and 59.1 % in T1D cases, whereas in controls the frequencies were 41.4 % and 43.3 %, respectively. Double-null genotype was found to be elevated among T1D patients (Odds Ratio [OR], 2.04; 95 % Confidence Interval [CI], 1.08-3.85; P = 0.027). No effect of any genotype for GSTP1 on T1D susceptibility was detected. Individuals with both the double-null and GSTP1 Ile/Val genotype combined appeared to be at increased risk of T1D (OR, 4.95; 95 % CI, 2.11-11.6; P = 0.0002).

Conclusion: This is the first study conducted on Iranian children with T1D. The absence of GSTM1 and/or GSTT1 may be important risk factor for T1D. Furthermore, presence of Val allele for GSTP1 could strengthen this risk.

Keywords: GSTM1, GSTT1, GSTP1, deletion, Ile105Val, polymorphism, type 1 diabetes, oxidative stress

Prevalence of growth disorders in a nationally representative sample of Iranian adolescents according to socioeconomic status: the CASPIAN III Study

Maryam Bahreynian,1 Mohammad Esmail Molaighi,2 Mostafa Qorbani3,4 Ramin Heshmat,6 Gelayol Ardalan,7 Roya Kelishadi7

1 Pediatrics Department, Child Growth and Development Research Center, Research Institute for Primary Prevention of Non-communicable Disease, Isfahan University of Medical Sciences, Isfahan, Iran; 2 Department of Pediatrics, Ahvaz Jundishapur University of Medical Sciences, Ahvaz, Iran; 3 Department of Public Health, Alborz University of Medical Sciences, Karaj, Iran; 4 Endocrinology and Metabolism Research Center, Endocrinology and Metabolism Research Institute, Tehran University of Medical Sciences, Tehran, Iran; 5 Chronic Diseases Research Center, Iran J Pediatr; Vol 25 (Suppl 1), Oct 2015

Keywords: Growth disorders, socioeconomic status, adolescents, prevalence
The Effect of Education to Mothers and Girls on Knowledge and Practice toward Puberty Hygiene in Tabriz, Iran: a randomized controlled trial

Atousa Afsari1, Massomeh Abbasnezhadeh2, Samira Fatahi3
1. Tabriz University of Medical Sciences, Tabriz, Iran; 2. Nursing Department, Kermanshah University of Medical Sciences, Kermanshah, Iran

Background: Puberty and changes resulting from it are one of the most important events of everybody’s life. Mothers played a central role in raising the level of their knowledge on menstruation and abdominal obesity was defined as waist to height ratio> 0.5. Sub-national classification of the country was based on geography and social class of each region. Analysis of variance and Chi-square test were used to compare the prevalence of growth disorders between regions.

Findings: The mean and standard deviation for BMI was 19.42 (4.09) kg/m², showing a significant trend with lowest amount in Southeast (with lowest SES) and highest in the Central part (with highest SES) of Iran (p<0.001). Our findings showed that the abnormal parameters, such as the prevalence of obesity, combined overweight and obesity and abdominal adiposity, increased on a significant trend with rising socio-economic status (P<0.001, except for girls height, P=0.003). The opposite direction was documented for the prevalence of underweight and stunting, with the highest frequencies in Southeast (lowest SES) and the lowest in Central part (highest SES).

Conclusion: The present study showed significant national variation in the prevalence of weight disorders in Iranian adolescents. These findings underscore the necessity of implementing evidence-based health promotion programs and preventive strategies at national level.

Keywords: Adolescents, obesity, socioeconomic status, stunting, underweight, weight disorders

The Effect of Education to Mothers and Girls on Knowledge and Practice toward Puberty Health: a randomized controlled trial

Atousa Afsari1, Massomeh Abbasnezhadeh2, Samira Fatahi3
1. Tabriz University of Medical Sciences, Tabriz, Iran; 2. Nursing Department, Kermanshah University of Medical Sciences, Kermanshah, Iran

Background: Aattitude of a girl toward her menstruation and puberty has a considerable impact on her role of motherhood, social adjustment, and future marital life. This study was conducted in 2014 aiming at comparing the effect of education to mothers and girls on the attitude of adolescent girls of Tabriz city-Iran toward puberty health.

Methods: This randomized control clinical trial was conducted on 364 adolescent girls who experienced menstruation. Twelve schools were selected randomly among 107 girl secondary schools. One-third of the students of each school were selected randomly using a table of random numbers and their socio-demographic and attitude questionnaires were filled out by them. The schools were randomly allocated to the groups of mother’s education, girl’s education, and no-intervention. The attitude questionnaire was filled out by the participants again two months after intervention. The general linear model, in which the baseline values were controlled, was employed to compare the scores of the three groups after the intervention.

Findings: No significant differences were observed among the three groups in terms of the attitude score before intervention (p>0.05). Attitude score improvement after intervention in the girl’s education group was significantly higher than the one of both mother’s education (adjusted difference: 1.8 (confidence interval 95%: 0.4 to 1.3) and no-intervention groups (adjusted mean difference: 1.3; [95% CI: 0.0 to 2.6]) by controlling the attitude score before intervention.

Conclusion: Based on the research findings, it is more effective to educate girls directly about puberty health to improve adolescent girls’ attitudes than education to mothers and transferring knowledge from them to their daughters as a substitute for direct education of the girls in order to raise the level of their knowledge on puberty hygiene.

Keywords: Adolescent Girls, Knowledge, practice, Puberty hygiene

Endocrinology and Metabolism Population Sciences Institute, Tehran University of Medical Sciences, Tehran, Iran

Background: This study was performed to assess the growth disorders among a nationally-representative sample of adolescents according to the socioeconomic status (SES) of their living area.

Methods: This nationwide cross-sectional survey conducted among a representative multi-stage cluster sample of 5624 adolescents aged 10-18 years in Iran. The World Health Organization reference values were used to define growth disorders. Abdominal obesity was defined as waist to height ratio> 0.5. Sub-national classification of the country was based on geography and social class of each region. Analysis of variance and Chi-square test were used to compare the prevalence of growth disorders between regions.

Findings: The mean and standard deviation for BMI was 19.42 (4.09) kg/m², showing a significant trend with lowest amount in Southeast (with lowest SES) and highest in the Central part (with highest SES) of Iran (P<0.001). Our findings showed that the abnormal parameters, such as the prevalence of obesity, combined overweight and obesity and abdominal adiposity, increased on a significant trend with rising socio-economic status (P<0.001, except for girls height, P=0.003). The opposite direction was documented for the prevalence of underweight and stunting, with the highest frequencies in Southeast (lowest SES) and the lowest in Central part (highest SES).

Conclusion: The present study showed significant national variation in the prevalence of weight disorders in Iranian adolescents. These findings underscore the necessity of implementing evidence-based health promotion programs and preventive strategies at national level.

Keywords: Adolescents, obesity, socioeconomic status, stunting, underweight, weight disorders
Keywords: Adolescent, Education, Attitude, Puberty

Polycystic ovary syndrome in adolescents
Fatemeh Saffari MD
Children Growth Research Center, and Metabolic Diseases Research Center, Qazvin University of Medical Sciences, Qazvin, Iran

Polycystic ovary syndrome (PCOS) is the most common cause of infertility in women, frequently becomes manifest during adolescence, and is primarily characterized by ovulatory dysfunction and hyperandrogenism. The syndrome is heterogeneous clinically and biochemically. The diagnosis of PCOS has lifelong implications with increased risk for metabolic syndrome, type 2 diabetes mellitus, and possibly cardiovascular disease and endometrial carcinoma. PCOS should be considered in any adolescent girl with a chief complaint of hirsutism, treatment-resistant acne, menstrual irregularity, or obesity. There are several proposed diagnostic criteria for polycystic ovary syndrome (PCOS). Few studies focus on treatment of PCOS in adolescents, so management is based primarily on studies in adults. Hormonal treatment with estrogen-progestin contraceptives is ordinarily the first-line approach for management of PCOS, in combination with weight management for patients with obesity. Diet and exercise are first-line treatments for obese adolescents with PCOS.

Keywords: polycystic syndrome; adolescent; hyperandrogenism

Polycystic ovary syndrome in adolescents
Ehya Zaridoost, Rasht, Iran

Polycystic ovary syndrome is a heterogeneous syndrome of unexplained chronic hyperandrogenism and oligo-anovulation and the most common cause of chronic anovulation associated with hyperandrogenic state. The incidence of it is about 5-10% in reproductive-age women. Diagnosis is made by excluding other hyperandrogenic disorders (e.g., nonclassic and classic adrenal hyperplasia, androgen secreting tumors, hyperprolactinemia) in women with chronic anovulation and androgen excess. Major morbidities are related to reproductive and cardiovascular systems. The most important reproductive morbidities consisting infertility, irregular uterine bleeding, increased pregnancy loss and higher risk of endometrial cancer than non PCO people. Cardiovascular risks are linked to insulin resistance and common occurrence of obesity, although it also occurs in non-obese women with PCOS. PCOS is considered to be a heterogeneous disorder with multifactorial causes and it appears to account for 70% of the variance in pathogenesis. Both heritable and non-heritable factors contribute to arise it. So a positive family history of chronic anovulation and androgen excess increases the risk for occurring PCOs. There are many evidences that risk factors for PCOS can be recognized in childhood. Congenital virilizing disorders; above average or low birth weight for gestational age; premature adrenarche, particularly exaggerated adrenarche; atypical sexual precocity; or intractable obesity with acanthosis nigricans, metabolic syndrome, and pseudo-Cushing syndrome or pseudo-acromegaly in early childhood have been identified as independent prepubertal risk factors for the development of PCOS. During adolescence, PCOS may masquerade as physiological adolescent anovulation. Asymptomatic adolescents with a polycystic ovary occasionally (8%) have subclinical PCOS but often (42%) have a subclinical PCOS type of ovarian dysfunction, the prognosis for which is unclear. Identifying children at risk for PCOS offers the prospect of eventually preventing some of long-term complications associated with this syndrome once our understanding of the basis of the disorder improves.

Keywords: polycystic syndrome; adolescent; hyperandrogenism
Determination of the Nutritional Status of Under Two Years Old Children of Khorramabad City in 2014-2015

Dr. Mahnaz Mardani1, Nafiseh Soleimani2, Maryam Eslami2, Pouria Reapour2
1. Faculty of Health and Nutrition, Lorestan University of Medical Sciences, Khorramabad, Iran; 2. Faculty of Health and Nutrition, Lorestan University of Medical Sciences, Khorramabad, Iran; 3. Faculty of Foreign Languages, Khorazmi University, Tehran, Iran

Background: Children are considered to be every society’s future, hence their healthy growth guarantees the society’s health and progress. According to World Health Organization’s (WHO) released statistics in 2013 the prevalence of malnutrition in the world is 15%. This prevalence is as up to 25% in undeveloped and developing countries. Considering the rapid rate of physical growth resulting in the enhanced nutritional needs of this age group, children are considered to be the most endangered age group from malnutrition. The aim of this research is to determine the prevalence of malnutrition among children up to two years old referring to the health clinics in Khorram-Abad city.

Materials and Methods: In this descriptive-analytic study the files of families available at the health clinics in Khorram-Abad city were analyzed. Using clustering method 70 children up to the age of two were studied. The needed information was acquired through a self-made questionnaire which included the demographic information of the mother and the child, the mother’s BMI before pregnancy, and the child’s health index gathered from the family files of the families. Waterloo classification was used for the identification of severe malnutrition (weight for height and age), chronic malnutrition (height for age) and the WHO classification was used for the identification of the contemporary malnutrition (weight for height index includes: low weight under 5 percentile, normal between 5-85 percentile, overweight between 85 – 95, and obese equal to or more than the 95 percentile. Then the relation of these amounts to the mother’s BMI before pregnancy was analyzed. The acquired information was analyzed through the SPSS software, version 21. In the descriptive part, the central and diversity indexes and for the analysis of the relationship between the variables the Fisher test was used.

Findings: In this survey according to the index for height to age 58.2% of the children were normal in height to age ration, and 41.8% were short in height. Using the fisher test, the mother’s BMI and the child’s height to age ratio were correlated and had a meaningful relationship. Also considering the index for weight for age, 61.7% of the children were considered normal, 16.4% were overweight, 16.4% were underweight, and 5.2% were obese (p<0.05). According to the exact fisher test there was a meaningful relationship between the mother’s BMI and the child’s weight to age ratio (p<0.01). Furthermore according to the index for weight for height, 64.7% of the children were normal, 3.5% were overweight, 10.6% were underweight, and 21.2% were obese. There was no meaningful relationship found between the mother’s BMI during pregnancy and the index for children’s weight for height index.

Conclusion: According to the present study’s results the children under study were suffering from a high prevalence of short height (chronic malnutrition) also the prevalence of low weight is higher than the society’s mean. According to these findings it is necessary to convey the appropriate nutritional teachings, especially to the feeding of mother’s breast milk, and appropriate and on time supplemental nutrition. Furthermore, considering the mother’s BMI during pregnancy, the index for weight for age, and the index for height for age of the child attention to the mother’s nutrition during pregnancy is also critical.

Keywords: Body Mass Index (BMI), Children under 2, Malnutrition.

Relationship Between Breast Feeding and Obesity in Children with Low Birth Weight

Farzad Shidfar1,2, Mitra Zarrati1, Maryam Moradof 2, Farinaz Naziri nejad1, Hossein Keyvani1, Mohsen Rezaei Hemami1
1Department of Nutrition, School of health, Iran University of Medical Sciences; 2Department of Exercise Physiology, Islamic Azad University, Tehran, Iran.

Background: breastfeeding appears to play a role in determining obesity and abdominal obesity during childhood especially in children with low birth weight. The purpose of this study was to investigate the association between breast feeding, abdominal obesity and obesity in Iranian children with low birth weight (LBW).

Methods: 1184 students (625 girls and 559 boys), aged between 10 – 13 years old, selected from 112 elementary schools in Iran. Measured parameters were family’s economic situation, level of parent’s education, history of obesity in First-degree relatives, breast feeding, food pattern and birth weight by questionnaire. Also height, weight, waist circumference and blood pressure were measured.

Findings: 13.68% (n=160) of students had low birth weight, 59.91% (n=701) were normal and 26.1% (n=309) had weight more than 4000 grams at birth. 26.41% of participants had abdominal obesity. 5.32% of students were obese and 22.04% were overweight, girls were more obese than boys (p<0.05), history of obesity in first-degree relatives, severe weight gain in pregnancy and birth weight had significant relationship with prevalence of obesity and abdominal obesity in childhood (p<0.05). The prevalence of abdominal obesity in students with low birth weight was significantly correlated with breast feeding (p<0.05); But this correlation was not significantly due to obesity in our participants (p>0.05). Duration of breast feeding had significant relationship with obesity and abdominal obesity in children with low birth weight (p<0.05).

Conclusions: As the prevalence of obesity in children is increasing, more research is urgently needed to clarify whether breast feeding might even have negative consequences for risk of chronic disease in childhood, especially in children with low birth weight. Breast feeding and its long-term consequences were important factors for prevention of metabolic syndrome criteria in childhood and older ages.
Key words: Low birth weight, Abdominal obesity, Obesity, Breast feeding, Iran

Mother's employment and level of mother's education in reduction of height and weight in the infants
Mohsenzadeh N, Mohsenzadeh A, Ahmadipour SH, Lorestan University of Medical Sciences, Koramabad, Iran

Background: Children's growth is one of the most important sources of information for the diagnosis of delayed growth and malnutrition in children. Therefore, continuous measurement of height and weight can detect the deviation in the growth pattern. The purpose of this study was to evaluate the effect of mother's employment and level of mother's education in reduction of height and weight in the infants under two years and referred to khorramabad health centers in first six months of age.

Methods: This is a cross-sectional study. The population included in the study were all the infants under 2 years old referred to khorramabad health centers in 1392. All information collected by questionnaire and was analyzed by SPSS software.

Findings: 150 infants were studied, 24.8% had reduction in the weight curve and 7% had a reduction in the height curve. There is a significant relationship between reduction of weight and height with employment of mother so that in housewife mothers 22.7% and in employed mothers 64.3% have children’s growth curve reduction (p=0.0001). There is a significant relationship between mother's education and growth curve reduction; uneducated mothers were 56% and college-educated mothers were 18%.

Conclusion: There are meaningful relationship between mother's employment and mother's education level with reduction of height and weight graphs.

Key words: height reduction, weight reduction, health centers.

Abdominal pain in children
Bahar Allahverdi, MD, Pediatric GI Research Center, Children's Medical Center Hospital, Pediatric Center of Excellence
Bahareh Yaghmaei, MD, Pediatric Intensive Care Fellowship, Children's Medical Center Hospital, Pediatric Center of Excellence, Tehran, Iran

Abdominal pain is one of the most common complaints in childhood and one that frequently requires urgent evaluation in the office or emergency department. The cause is typically a self-limited minor condition, such as constipation, gastroenteritis, or viral syndrome. The challenge for the clinician is to identify those patients with abdominal pain who have either serious, potentially life-threatening conditions, such as appendicitis or bowel obstruction (as can occur from volvulus, intussusception, or adhesions), or infections that require specific treatment (streptococcal pharyngitis or pneumonia). The likely diagnosis is often suggested by the child's age and clinical features (associated symptoms, past medical history, and physical examination).

Neurologic Basis of Abdominal Pain
Pain perception is incompletely understood, but depends upon the type of stimulus and the interpretation of receptor signals in the central nervous system (CNS). Different types of stimuli may act simultaneously to influence the perception of pain. As an example, the gastric mucosa typically is insensitive to pressure or chemical stimuli, however, these stimuli may cause pain if the gastric mucosa is inflamed. The processing of visceral pain signals by the CNS and psychologic factors also influence the perception of pain. The threshold for perceiving pain from visceral stimuli may vary among individuals. Pain originating in the viscera is perceived to originate at a distant site (ie, referred pain). Pain receptors in the abdomen respond to mechanical and chemical stimuli. Stretch is the principal mechanical stimulus involved in visceral pain; other stimuli include distention, contraction, traction, compression, and torsion. Pain receptors in the gastrointestinal tract are located on serosal surfaces, within the mesentery, and within the walls of hollow viscera (between the muscularis mucosa and submucosa). Mucosal receptors respond primarily to chemical stimuli (eg, substance P, bradykinin, serotonin, histamine, prostaglandins), which are released in response to inflammation or ischemia. Other visceral pain receptors respond to chemical or mechanical stimuli. Referred pain usually is located in the cutaneous dermatomes sharing the same spinal cord level as the visceral inputs. As an example, nociceptive stimuli from the gallbladder enter the spinal cord at T5 to T10. Thus, pain from an inflamed gallbladder may be perceived in the scapula. Precise localization of the pain to the right upper quadrant in patients with acute cholecystitis usually occurs once the overlying parietal peritoneum (which is somatically innervated) becomes inflamed.

Chronic abdominal pain is common in children and adolescents. Complaints of chronic abdominal pain occur in 10 to 19 percent of children. The prevalence is increased in children age four to six years and early adolescents. According to one study, as many as 17 percent of middle- and high-school students reported weekly episodes of abdominal pain. Among the patients who reported abdominal pain, approximately 21 percent had discomfort that was sufficiently severe to affect activity. The initial evaluation of the child or adolescent with chronic abdominal pain includes history, physical examination, and stool testing for occult blood to determine whether the child has any “alarm findings”, which help to distinguish organic from functional abdominal pain and direct the need for additional evaluation. Information other than alarm findings obtained in the initial evaluation helps to distinguish among organic etiologies and provides insight into biopsychosocial factors that may trigger or reinforce pain (independent of etiology) and are helpful formulating a management plan.

Comprehensive initial history and examination helps to reassure the patient and family that the clinician is taking their complaints seriously. At the time of presentation, the parents and child may be frustrated and anxious; they may have tried over-the-counter or dietary interventions without improvement and may be increasingly concerned that the child has a serious disorder. The parents should be asked what they think is causing the pain (or what they are worried is causing the pain) so that their concerns can be directly addressed. It is important to establish a therapeutic alliance early in the course of evaluation and treatment. It may be helpful to introduce the concept of functional gastrointestinal disorders(FGID’s) as a possible cause at the initial evaluation, even if the pain has not been present long enough to qualify as “chronic”. It also may be helpful to educate the patient/family about the biopsychosocial model.
of pain, and possible role of stress. Providing examples of how stress can directly cause abdominal pain or other symptoms (churning of the stomach or headache before a test) may help them to understand this relationship.

Minilaparoscopic cholecystectomy: first reported patients series in Iran

Seyed Mojtaba Moussavi-Khoshdeh
Allaghar Children’s Hospital, Iran University of Medical Sciences, Tehran, Iran

Background: Laparoscopic surgery has been the standard method for cholecystectomy since many years. Minilaparoscopy is a new subdivision of laparoscopy that can be done by fine instruments and through smaller incisions. The use of smaller instruments during minilaparoscopic cholecystectomy has been proposed to reduce postoperative pain and improve cosmesis. Surgical technique Under general anesthesia with OTT(orotracheal tube) in supine position a 5 mm incision performed in umbilicus for insertion of endoscope port by open technique and CO2 insufflation performed at appropriate pressure. Then a 3mm port was introduced in epigastric region and a grasper introduced trough a 2mm stab in RUQ without port. Cholecystectomy was done after clipping of the duct and bladder was extracted trough umbilical port. In this way cholecystectomy can be done by 3 incisions ,sum of them equal to 10 mm.

Method: 56 minilaparoscopic cholecystectomis were done in children since 2010 at four medical centers. We studied their recordings retrospectively. Findings: The age range of patients was 19 mounts to 18 years. Mean operative time was 96 min. Conversion of epigastric 3 mm port to 5 mm port was done in 10 patients. No major complication was seen in fallow up. Conclusion: Minilaparoscopic cholecystectomy can be safely performed using 5-mm umbilical, 3-mm epigastric and 2-mm subcostal incisions. The use of minilaparoscopic techniques resulted in decreased early postoperative incisional pain, avoided late incisional discomfort and produced superior cosmetic results. Although improved instrument durability and better optics are needed for widespread use of miniport techniques, this approach can be routinely offered to many properly selected patients undergoing elective cholecystectomy.

Keywords: cholecystectomy, Minilaparoscopy, Children

Evaluation of serum Adenosine deaminase in Cystic Fibrosis patients in an Iranian referral Hospital

Parisa Tajdini 1 , Fatemeh Farahmand 1, Gholamhossein Falahi 2, Sedigheh Shams 3, Shima Mahmoudi 4
1- Department of Pediatrics Gastroenterology, Children’s Medical Center, Pediatrics Center of Excellence, Tehran University of Medical Sciences; 2- Pediatric Infectious Disease Research Center, Tehran University of Medical Science, Tehran, Iran

Background: Adenosine, a signaling nucleoside, is controlled in part by the enzyme adenosine deaminase (ADA). There are rare reports on the role of adenosine levels and ADA in cystic fibrosis (CF) patients. The aim of this study was to assess serum ADA in CF patients in order to find whether the severity of lung disease in CF is related to significant changes of ADA or not.

Method: Venous blood samples (1cc) of each individual of CF (3y-15y) and 49 healthy children (3y-15y) referred to children medical center Hospital was taken and their serum ADA was measured. Classification of respiratory and gastrointestinal disease severity in CF patients as well as their Body Mass Index (BMI) was performed. The results were compared with values obtained from healthy children matched for age and gender.

Findings: This study included 49 children of both genders (20 females and 29 males) with CF (mean age: 6.36±2.22 years). The mean serum ADA in CF patients group and control group was 9.38±2.72 and 16.04± 1.27, respectively (p=0.001). The mean serum ADA in CF patients with normal BMI was higher than patients with low BMI (p=0.002).

Conclusion: In this study the lower serum level of ADA was seen in CF patients compared to control group. The clinical symptoms in CF patients’ especially respiratory symptoms might be associated with reduction of serum ADA and rising serum adenosine; therefore, further studies on the uses of ADA enzyme therapy in CF patients are highly recommended.

Keywords: Adenosine deaminase, Cystic Fibrosis, children

Association between Outcome and Demographic Status in Azeri Turkish Population with Cystic Fibrosis

Mandana Rafiey 1, Morteza Jabarpoor-Bonyadi 2, Morteza Ghojazadeh 3, Leila Vahedi 4
1. Tabriz Children's Hospital, Liver & Gastrointestinal Diseases Research Center, Tabriz, University of Medical Sciences, Tabriz, Iran; 2. Faculty of Natural Sciences, Center of Excellence for Biodiversity, University of Tabriz, Tabriz, Iran

Background: Outcomes in cystic fibrosis are improving rapidly. The demographic parameters are as impressive factors in outcome which can be evaluated and modified. The aim of this study was to investigate the association between outcome and demographic status of patients with cystic fibrosis from Azeri Turkish population in Iran.

Methods: Data on 331 patients, from March 2001 to September 2014 were gathered, and the association of outcome with demographic variables was studied using logistics regression, Chi-square, and Independent-Samples T test by SPSS.18. Odds ratio calculated with confidence intervals of 95% and P < 0.05 were considered significant.

Findings: There were 85 (25.7%) dead patients and 246 (74.3 %) alive patients at the time of the study were collected. Two hundred and two (82.2%) of the 246 CF patients who were alive were under nine years, and 77 (90.6 %) of 85 dead CF-patients had died under 4 years. The risk of mortality was 50% less in urban patients than rural patients. The ratio of mortality for positive family history was two times or 97% greater than negative family history. The proportion of mortality was approximately two times or 94% higher for positive consanguineous marriage than negative consanguineous marriage.

Conclusions: The results demonstrated that the mortality rate is higher in CF-patients with positive family history, positive consanguineous marriage, and residency in villages. Therefore, demographic factors play an important role on outcome of cystic fibrosis. Unfortunately, these parameters, which can be managed easily and with less cost, have been overlooked.
Keywords: Cystic Fibrosis; Outcome; Demographic Status, Azeri Turkish population; Iran

Determination of the Nutritional Status of Under Two Years Old Children of Khorram-Abad City in 2014-2015 and its Relationship With Mothers’ Body Mass Index During Pregnancy

Mahnaz Mardani1, Nafiseh Soleimani1, Maryam Eslami2, Pouria Reapour1
1. Faculty of Health and Nutrition, Lorestan University of Medical Sciences, Khorramabad, Iran; 2. Faculty of Foreign Languages, Kharazmi University, Tehran, Iran

Background: Children are considered to be every society’s future, hence their healthy growth guarantees the society’s health and progress. According to World Health Organization’s (WHO) released statistics in 2013 the prevalence of malnutrition in the world is 15%. This prevalence is as up to 25% in undeveloped and developing countries. Considering the rapid rate of physical growth during childhood resulting in the enhanced nutritional needs of this age group, children are considered to be the most endangered age group from malnutrition. The aim of this research is to determine the prevalence of malnutrition among children up to two years old referring to the health clinics in Khorramabad city.

Methods: In this descriptive-analytic study the files of families available at the health clinics in Khorramabad city were analyzed. Using stratified sampling 170 children up to the age of two were studied. The needed information was acquired through a self-made questionnaire which included the demographic information of the mother and the child, the mother’s BMI before pregnancy, and the child’s health index gathered from the family files of the families. Waterloo classification was used for the identification of severe malnutrition (weight for height age), chronic malnutrition (height for age) and the WHO classification was used for the identification of the contemporary malnutrition (weight for height index includes: low weight under 5 percentile, normal weight between 5-85, over weight between 85-95, and obese equal to or more than the 95 percentile. Then the relation of these amounts to the mother’s BMI before pregnancy was analyzed. The acquired information was analyzed through the SPSS software, version 21. In the descriptive part, the central and diversity indexes and for the analysis of the relationship between the variables the Fisher test was used.

Findings: In this survey according to the index for height to age 58.2% of the children were normal in height to age ratio, and 41.8% were short in height. Using the fisher test, the mother’s BMI and the child’s height to age ratio were correlated and had a meaningful relationship. Also considering the index for weight for age, 61.7% of the children were considered normal, 16.4% were underweight, 16.4% were overweight, and 5.2% were obese (p<0.03).

Conclusion: According to the present study’s results the children under study were suffering from a high prevalence of short height (chronic malnutrition) also the prevalence of low weight is higher than the society’s mean. According to these findings it is necessary to convey the appropriate nutritional teachings, especially to the feeding of mother’s breast milk, and appropriate and on time supplemental nutrition. Furthermore, considering the mother’s BMI during pregnancy, the index for weight for age, and the index for height for age of the child attention to the mother’s nutrition during pregnancy is also critical.

Keywords: Body Mass Index (BMI), Children under the age 2, Malnutrition.

Simultaneous Presentation of Wilson’s Disease and Autoimmune Hepatitis; A Case Report

Farid Imancadeh, MD; Naghi Dura, MD; Amir Hossein Hosseini, MD; Peiman Nasri, MD; Ali-Akbar Sasyari, MD; Department of Pediatric Gastroenterology, Mofid Children Hospital, Shahid Beheshti University of Medical Sciences, Tehran, Iran

Background: Coexistence of Wilson’s disease and autoimmune hepatitis has been rarely reported in English literature. In this group of patients, there exist features of both diseases and laboratory and histopathological studies may be misleading. Medical treatment for any of these entities, per se, may result in poor response. Therefore, by considering the acute hepatitis resembling Wilson’s disease and autoimmune hepatitis, simultaneous therapy with immunosuppressive and penicillamine may have a superior benefit. Case Presentation: We present the case of a 10-year-old boy with nausea, vomiting, yellowish discoloration of skin and sclera, abdominal pain and tea-color urine. Physical examination showed mild hepatomegaly and right upper quadrant tenderness. Laboratory and histochemical studies and atomic absorption test were done and the results were highly suggestive of both Wilson’s disease and autoimmune hepatitis, in him.

Conclusion: This case study highlights, although rare, the coexistence of Wilson’s disease and autoimmune hepatitis and the need to maintain a high level of awareness of this problem. Therefore, it is reasonable to consider this type of hepatitis in rare patients, with dominant features of both diseases at the same time.

Keywords: Hepatitis, Hepatolenticular Degeneration; Autoimmune

Long term outcome of children with biliary atresia after Kasaei surgery in children medical center

Mojtaba gorji Growth and Development Research Center, Children Medical Hospital, Tehran University- of Medical Sciences, Tehran, Iran

Background: Long- term outcome of children with biliary atresia after Kasai surgery in children medical center

Abstract: Aim: The aims of the present study were to evaluate the long-term prognosis of children with biliary atresia after the Kasai surgery and to analyze the present status of survivors retaining their own livers. Background: Biliary atresia is the most common cause of pathologic
infantile icter that result from obstructions of extra hepatic bile ducts due to inflammation and fibrosis. It is a progressive disorder and gradually results in cirrhosis, portal hypertension and hepatic failure. With hepatopancreatoenterostomy (Kasai surgery) within first 8 weeks of infantile life by production of biliary drainage we can slow the speed of progression of disease to cirrhosis, hepatic failure and its complications.

**Method:** This is a descriptive retrospective cohort study in which we evaluate patients with biliary atresia who admitted in children medical center hospital in Iran and had Kasai surgery during 10 years. (From 1998-2008)

**Findings:** 2-5 years survival rates of patients were 14.3% , 5-10 years survival rates of patients were 8.1% and more than 10 years survival rates of patients were 2.1%. The survival rates varied significantly depending on the age at Kasai operation. The incidence of cholangitis was 59.1% and gastrointestinal bleeding were 38.7% in post surgical state. 4 patients had liver transplantation (8.1%) of them 2 patients survived (50%). From 10 patients who perform kasai before 8 weeks of life 6 patients survived (60%) and 4 patients were died. From 23 patients who performed kasai between 8-12 weeks of life 4 patients survived (17.3%) and 19 patients (82.6%) were died and. From 16 patients who perform kasai after 12 weeks of life 2 patients survived (12.5%) and 14 patients ( 87.5%) died. Conclusion: according to our findings, this study resembles to previous studies. Kasai surgery is an effective procedure in treatment of patients with biliary atresia and the earlier it performed the better outcome and survival of patients. The long-term survival rate can be improved if Kasai operation is performed earlier.

**Keywords:** biliary atresia. outcome. kasaei surgery

**Comparison of lead levels in patients with chronic abdominal pain with the control group**

Mohabbi A; pirhadi N; vafaei N;Ashati S; Khodadad Ahmad*Growth and Development Research Center, Children Medical Hospital, Tehran University- of Medical Sciences, Tehran, Iran- (*corresponder)

Chronic abdominal pain (CAP), which happens at least once a week for 2 months, is a kind of abdominal pain and one of the most common symptoms especially in children. Organic causes been identified by physical or laboratory routine examinations. Nonetheless there are several cases with indeterminate reason abdominal pain, that is not justifiable by physiological or biochemical disorders. We assumed lead as a cause for this kind of (AP). Lead absorbed by intestine and causes to be poisoning. Lead poisoning is an environmental disease and causes gastrointestinal disorders. this study included 48 patients at the age of 3 to 16. we analyzed the amount of serum lead levels with atomic absorption method. Results compared with the control group at the same age. In this study, the average lead level in patients with abdominal pain than the control group.

**Keywords:** chronic abdominal pain (CAP-pb)

**Impact of nutrition support team on nutritional status and outcome of postoperative congenital gastrointestinal anomalies patients**

Parisa Zarei-shargh,MSc; Emad Yazbashian,MSc; Atieh Mehdiizadeh-Hakkaei,MD,PhD; KhadiemiGholamreza, MD ; Abdolreza Norouzy, MD,PhD;Bahareh Imani, MD 6

**Background:** The aim of current study was to evaluate post-operative nutritional status in pediatric patients who undergo operation due to congenital gastrointestinal anomalies in pediatric intensive care unit (PICU) and investigate the role of nutrition support team (NST) on outcome.

**Method:** A retrospective clinical study, was carried out at two PICUs in Dr Sheikh pediatric hospital, Mashhad, Iran; one being supported by nutrition support team and the other not. A total of 120 patients were included through a non-random simple sampling. Different variables such as age,sex, prematurity, type of anomaly, birth weight, use of dopamine, weight gain in PICU, length of PICU stay, post-operative enteral nutrition initiation , duration of mechanical ventilation, mortality rate, maximum of blood sugar, calorie delivered to calorie requirement ratio and distribution of energy from enteral or parenteral roots were compared between patients of two PICUs.

**Result:** Median weight gain and the amount of calorie delivered during PICU stay in subjects of NST-supported PICU was significantly more than non NST-supported PICU. There was no significant difference in length of PICU stay, enteral nutrition initiation after operation, ventilation days and percent of mortality between two groups. Percentage of enteral feeding also increased about 2.8% which was not significant.

**Conclusion:** Nutrition support team (NST) could increase post-operative weight gain and calorie delivery in patients as well as providing an increase in enteral feeding rather than parenteral.

**Keywords:** Nutritional support; Digestive System Abnormalities; Nutritional status; Intensive Care Units, Pediatric; Outcome Assessment; Postoperative period

**Primary sclerosing cholangitis, a case report**

Vajiheh Modaresisaryazdi, Iranian Social Security Organization

**Background**

Primary sclerosing cholangitis (PSC) is a chronic progressive disorder of unknown etiology that is characterized by inflammation, fibrosis, and stricture of medium and large size intrahepatic and extrahepatic biliary ducts. The great majority of affected patients (~90%) have underlying ulcerative colitis. Fatigue and pruritus are common features at presentation. Here we report a 16 year old girl suffering from pruritus for more than 4 years who was treated for allergic disease. Case report: This is a 16 year old girl with itching started from 4 years ago. Several medications including antihistaminic drugs were used in order to relieve itching, but she didn’t improve. She also experienced some diet modification for possible food allergy as etiology of pruritus with no response. Except for itching ,she had no other problems. After two years she evaluated for systemic disease and disturbance of liver enzymes was detected. Alt=125.Ast=98.Alk=989 and GGT=132, Bilirubin Total =2.Direct=1.2 .Pt and INR were in normal range. Abdominal ultra sound showed...
The prevalence of malnutrition in the 0-2 year’s children of the Khorramabad city and its association with the mother’s pre pregnancy BMI

Mahnaz Mardani, Nafise Solaymani, Maryam Esami, Soodabe Zare, Pouria Rezapour
Lorestan University of Medical Sciences, Khorramabad, Iran

Background: According to the World Health Organization’s data the prevalence of malnutrition in 2013, was15% although, this prevalence has been 25% for undeveloped and developing countries. Due to the extremely rapid growth of children, they are would be at risk of malnutrition more than other aged groups, so, the aim of this study was to determine the prevalence of malnutrition in 0-2 years of age children that has been referred to health centers in the Khorramabad city.

Methods: In this cross-sectional study by using the two-stage cluster sampling 170 children from 0 to 2 years of old were selected and their information was collected through a customized questionnaire that included demographic information, child health indicators, and maternal pre-pregnancy BMI. The acute malnutrition (weight for age) and chronic malnutrition (height for age) were identified by the Waterloo classification and also past time malnutrition was identified by the WHO classification (weight for height index including: underweight = lower than the 5th percentile, normal weight = between the 5th and 85th percentile, overweight = between the 85 – 95th , and obese≥ 95th percentile). Then the relationship of the children’s status and mother’s pre-pregnancy BMI values were assessed. Data were analyzed using SPSS version 21.

Findings: In this study, based on height-for-age index, 41.8% of children were short stature and between mother’s pre pregnancy BMI and children’s height for age there was a significant relationship (p<0.03). Based on weight for age index, 16.4% of children were underweight, 16.4% were overweight and 5.2% of them were obese, and between mother’s pre pregnancy BMI and BMI was a significant correlation (p<0.01). In addition, based on weight for height index, 64.7% of the children were normal, 3.5% were underweight, 10.6% were overweight, and 21.2% of them were obese. Between mother’s pre pregnancy BMI and children’s weight for age index was no significant relationship.

Conclusion: Based on the results of the present study, children are suffering of chronic malnutrition (short stature) as well as the prevalence of underweight in this group of children is also higher than the average prevalence of community. Accordingly, we recommend that, attendance to exclusive breastfeeding and also supplementary nutrition can improve children nutrition and their health indices.

Keywords: Malnutrition, children, overweight, underweight

Eosinophilic esophagitis

Mehri Najafi, MD
Tehran Medical School, Children Medical Center Hospital, Tehran University of Medical Sciences, Tehran, Iran.

Eosinophilic esophagitis is a chronic immune / antigen mediated inflammatory disease of the esophagus. The disease is characterized by histological evidence of dense esophageal tissue eosinophilia. The disease is more common in males and in patients with atopic disease.

Symptoms: Clinical manifestations are variable depending on age and the disease phenotypes. Feeding difficulties are the most common symptoms in infants and toddlers, vomiting and pain in children, and dysphagia and food impaction in adolescents. Patients may or may not be atopic. Total IgE and specific IgE to food antigen are not reliable. The most important allergens are milk, nuts, wheat, fish, soya and eggs.

Diagnosis: Endoscopy and histology are the main diagnostic ways.

Treatment: Dietary treatment: that is most effective in atopic patients. Three elimination diets have been developed for patients : amino acid-based formula (AAF), targeted elimination diet, and empiric elimination diet.

Drug therapy: Corticosteroids (oral, topical) are highly effective.

Keywords: eosinophilic esophagitis, milk allergy, vomiting, abdominal pain

Effect of plasmapheresis on treatment of acute pancreatitis in infant with familial chylomicronemia

Enayatollah Nemati Khorasani, MD; Fariba Mansouri, MD
Faculty of Baghiyatallah University Medical of Sciences, Tehran, Iran

Background: Familial chylomicronemia is a rare genetic disorder.(autosomal recessive)with incidence 1/1000,000 that there is difficulty in clearance serum content of triglyceride & cholesterol due to deficiency of Apo-B-lipoprotein lipase or lipoprotein lipase(LPL) or cofactor Apo-C II. Difference between them is decrease level of serum TG after prescription FFP in Apo-CII and without response in LPL deficiency.

Case Presentation: We introduce a female baby 41 days old that referred by hyperlipidemia (TG=25000 mg/dl and cholesterol= 1500 mg/dl) and acute pancreatitis attack and signs and symptoms denoting to familial chylomicronemia who treated by plasmapheresis. After plasmapheresis decreased the TG of plasma: 400mg/dl and cholesterol: 232mg/dl and improved the symptoms and sings of her pancreatitis.
Conclusion: According to this finding, It was denoted that plasmaphersis is a useful method for treatment of acute pancreatitis caused by chylomicronemia.

Keywords: Plasmapheresis, Acute pancreatitis, Familial Chylomicronemia

Effect of plasmapheresis on treatment of acute pancreatitis in infant with familial chylomicronemia

1-Dr. Enayatollah Nemat Khorasani-MD. Ped Gastrologist, Associated Prof.(Faculty of Buhegyatlallah university medical of sciences). 2-Dr. Fariba Mansouri-MD. Internist. pulmonologist. Associated Prof(Faculty of Tehran university of medical sciences.)

Familial chylomicronemia is a rare genetic disorder (autosomal recessive) with incidence 1/1000,000 that there is difficulty in clearance serum content of triglyceride & cholesterol due to deficiency of Apo-B-lipoprotein lipase or lipoprotein lipase(LPL) or cofactor Apo-C II. Different between them is decrease level of serum TG after prescription FFP in Apo-CII and without response in LPL deficiency. Clinical futures are: Hepatosplenomegaly, Pancreatitis attack, Xantem eruptions on limbs&butects, Paleness of retina(libemia reticolaris). We introduce a female baby 41 days old that referred by hyperlipidemia (TG=25000mg/dl & cholesterol= 1500mg/dl) and acute pancreatitis attack and signs & symptoms denoting to familial chylomicronemia who treated by plasmapheresis.

Keywords: Plasmapheresis-Acute pancreatitis-Familial Chylomicronemia

Effects of mother’s age and BMI on failure of exclusive breastfeeding

Reza Saedi1, Saeed Soltani asl heri2
1. Neonatal Research Center, Imam Reza Hospital, Faculty of Medicine, Mashhad University of Medical Sciences, Mashhad, 2. Azad University of Medical Sciences, Mashhad, Iran.

Background: Exclusive breastfeeding for the first 6 months of life has been recommended by numerous health and professional medical organizations, but unfortunately the prevalence of exclusive breastfeeding remains far too low in many areas of the developing world. In some studies observed suboptimal coverage of exclusive breastfeeding, with less than 40% of infants younger than six months of age estimated to be exclusively breastfed in 2010. The purpose of this research was to assess the relationship between age, height, weight and BMI of mother in exclusive breastfeeding.

Methods: This is a cross-sectional study among 150 infants who discharged from NICU and normal nursery of Imam Reza and Ghaem hospitals in Mashhad University, Iran. Method of data collection was filling the questionnaire based on information in thefile, physical examination and interviewing with mother. To determine the possible relationships between variables within the sample, we used chi-square and t test. We analyzed the data by SPSS11.5.

Findings: There was no relationship between age of mother and success in exclusive breastfeeding. (P>0.99). Also there is no significant relationship between height and weight of mother with succeeding in exclusive breastfeeding (p=0.55, p=0.85). Results showed that there was no relationship between mothers BMI and succeeding in EBF.

Discussion and Conclusion: based on our research there was no significant relationship between height, weight, age and maternal BMI with failure or succession in EBF.

Keywords: exclusive breastfeeding, Body mass index, Neolnetes.

Measurement properties of an outcome measure for evaluation of pediatric functional constipation

Hirbod Nasiri Bonaki, Aidin Ahedi
Tehran University of Medical Sciences, Tehran, Iran

Background: Pediatric functional constipation is common among young children. Clinical evaluation of functional constipation plays an important role in management of pediatric patients. Reliable, valid and comprehensive measures are critical for evaluation of constipation severity. Our aim was to develop an outcome measure and evaluate its measurement properties.

Methods: A new measure was developed using the items of functional constipation section of the Rome III diagnostic criteria. After translation process, 40 patients with a diagnosis of chronic functional constipation were assessed using the new measure twice with a one week interval to evaluate the test-retest reliability of the measure. Validity of the measure was assessed using correlation with VAS score of overall severity of constipation. Patients were reassessed after two months of standard medical treatment, for evaluation of responsiveness of the measure.

Findings: Of 40 participants, 22 (55%) were male and mean age ± SD of the patients was 6.2 ± 1.2. The new measure showed good test-retest reliability with 90% agreement of total scores and ICC value of 0.87. Construct validity of the measure was supported by high correlation (r= 0.89) of the total score with VAS score of overall severity. Change in scores of the new measure and VAS score were also highly correlated (r= 0.8), indicating the responsiveness of the new measure.

Discussion and Conclusion: Proposed measure showed good reliability, validity and responsiveness in studied population. Further evaluation of its validity is recommended by evaluation of its correlation with other measures of constipation.

Keywords: Pediatric functional constipation, Outcome measure, Reliability, Validity, Responsiveness

Assessing vitamin D status in children residents of Tehran; are supplements really necessary?

Mohammad Hossein Khosravi1,2, Mohammad Torkaman2, Hassan Abolghasemi2
1. Students’ Research Committee, Baqiyatallah University of Medical Sciences, 2. Department of Pediatrics, Baqiyatallah University of Medical Sciences, Tehran, IR Iran

Background: Vitamin D is one of the most necessary lipid-soluble vitamins for the body; helping the growth and development of bones. Vitamin D deficiency in children
has several adverse effects. The most important preventing factor is the determination of vitamin D deficiency and prescribing vitamin D-containing supplements in the initial phase.

**Methods:** Three hundred children between 1-10 years of age referred to our pediatrics clinic enrolled to participate in this study. Parents were asked to answer a questionnaire including questions about demographic information, nutrition and supplements. Then the level of vitamin D, calcium and phosphorus were measured in their blood.

**Findings:** Finally 286 children, 140 male and 146 female, with a mean age of 4.46±2.82 underwent analysis. Of them 218 (76.22%) cases had vitamin D deficiency with a mean age of 5.09±2.82 and 76 cases (23.78%) had normal vitamin D levels with a mean age of 2.58±1.88 (p=0.001). Vitamin D level had a mean of 29.71±14.42 ng/ml in 88 (30.8%) cases up to 2 years of age and 17.11±14.02 ng/ml in 198 (69.2%) cases more than 2 years old (p=0.0001).

**Conclusion:** We concluded that children more than 2 years of age have lower vitamin D levels in comparison with those less than 2 years old. According to the significant higher level of vitamin D in supplement-consuming children, prescribing vitamin D–containing supplements in children more than 2 years of age is highly recommended.

**Keywords:** Vitamin D; Children; Sunlight exposure; Supplements.

---

**Geographical distribution of weight disorders in Iranian children and adolescents**

Silvia Hosepian, Roya Kelishadi, Masoumeh Rashidi, Mostafa Qorbani

**Background:** Weight disorders are considered as an important health concern as well as good indicator of nutritional status in a community. The aim of this study was to determine the geographical distribution of weight disorders in Iranian children and adolescents according to the geographical and climatic conditions.

**Methods:** In this cross-sectional study, students aged 6-18 years, living in urban and rural areas of 30 provinces in Iran, were studied. The prevalence of underweight and overweight, were determined in different geographical regions of Iran using the geographic information system (GIS) and ArcGIS software.

**Findings:** The prevalence rate of underweight and overweight in most regions ranged between 5-15%. The higher rate of underweight was seen in very hot and hot regions including Hormozgan, Sistan, Kerman, Zanjan and Ilam. The prevalence of overweight and obesity was higher in regions with cold and rainy weather including the Northern provinces, East Arzabayejan, Tehran, Ghom, and Boushehr with a prevalence rate of 15-20%. The lowest rate was documented in Southeastern provinces, Kerman and Hormozgan with a prevalence rate of 0-5%.

**Conclusion:** The findings of our study indicated that the distribution of weight disorders had large variations across different provinces. The higher rate of overweight/obesity in Tehran is due to the lifestyle of its population (less physical activity or unhealthy diet). Due to weather in Northern provinces, the outdoor physical activity is unavailable in most of the times of the year for children which could explain the higher rate of obesity in this region. It is supposed that the main factors that could explain the higher rate of underweight in very hot regions are under nutrition and low intake of essential micro and macronutrients. However, the role of other environmental and ethnic factors in this field should be evaluated also.

**Keywords:** Obesity, Overweight, Underweight, Children, Adolescents

---

**The Effect of Zinc Supplementation on Linear Growth and Growth Factors in Primary Schoolchildren in the Suburbs Mashhad, Iran**

Rahim Vakitili, Maha Yarzun Bakhtil, Mohammad Vahedian, Mahmoud Mahmoudi, Masoumeh Saeidi, Saba Vakili

1. Department of Pediatrics, Mashhad University of Medical Sciences; 2. Department of Public Health, Mashhad University of Medical Sciences; Immunology Research Center, Mashhad University of Medical Sciences; 4. Students Research Committee, Faculty of Medicine, Mashhad University of Medical Sciences, Mashhad, Iran

**Background:** Zinc is an essential trace element required for the functional activity of several enzyme systems. Several studies have been carried out to assess the effect of zinc supplementation on children’s growth, but controversy exists as to the effect of zinc on growth and growth hormone – insulin-like growth factor-I (GH-IGF-I) system. The aim of this study was to evaluate the effect of zinc supplementation on linear growth and serum level of IGF-I, Calcium(Ca), Phosphorus (P), Alkaline Phosphatase (ALP) in elementary school children living in a low socioeconomic suburbs of Mashhad, Iran.

**Methods:** The study was a randomized double-blind, placebo-controlled efficacy trial. Subjects were 200 volunteer primary school children. Both case and control groups comprised of 100 individuals each with 50 males and 50 females. Intervention supplementation was zinc sulfate tablets (10 mg elemental) and placebo tablets for both groups, administrated for a period of six months. The height, weight, height for age and weight for age Z-scores and Body Mass Index (BMI) were measured at 0, 2, 4, and 6 months. After six months the level of IGF-I, Ca, P and ALP were measured using blood samples taken from 50 volunteer children, 33 from the case and 17 from the control group. The statistical analysis was performed by SPSS version 11.5.

**Findings:** There was a significant increase in linear growth and weight amongst both male and female of the case group compared to the control after six months of receiving zinc (p<0.05). However, there was no significant difference in the serum level for the measured parameters between the two groups.

**Conclusion:** This study provides evidence of positive effect of zinc supplementation on the growth of school children living in a low socioeconomic suburbs of Mashhad city in Iran.

**Keywords:** Alkaline phosphatase, Calcium, IGF-I, Linear Growth, Phosphorus, Zinc
Effect of peppermint essence on satisfaction of patient and medical team with pediatrics’ endoscopic examination

Mohammad-Ali Kiani1, Ali Ghasemi1, Elham Poursoltani1, Bibi Leila Hoseini2, Hamid Ahanchian1, Masumeh Saeidi3
1. Department of Pediatrics, Faculty of Medicine, Mashhad University of Medical Sciences, Mashhad; 2. Midwifery Department, Nursing and midwifery school, Sabzevar University of Medical Sciences, Sabzevar; 3. Students Research Committee, Faculty of Medicine, Mashhad University of Medical Sciences, Mashhad, Iran

Background: Endoscopic examination of gastrointestinal (GI) system is a diagnostic and therapeutic instrument in children. Endoscopy usually encounters some difficulties because of intensive and spastic response of GI muscles during endoscopic examination. So this study aimed to assess the effect of peppermint essence on satisfaction of patient and therapeutic team during endoscopic examination.

Methods: This clinical trial study was conducted on 120 children less than 14 years who affected to pyloric spasm under endoscopy. Patients were randomly divided into two groups. Control group (n=60) received placebo and case group (n=60) received peppermint essence. Data were analyzed by descriptive-analytic (Mann-Whitney test, T-test, correlation) statistics and using SPSS 11.5.

Findings: More than half of case group (73.3%) were satisfied with endoscopy process, while 51.6% of control group were dissatisfied. Mean of endoscopy duration time was 9.30±0.35 min in peppermint group and 10.14±0.34 min in control group; which it had a significant difference in two groups (P<0.05). Mean duration time of pylorus spasm relaxation was less than 60s in case group, while it took time more than 60s in 60% of control group (P<0.05).

Conclusion: Findings showed that peppermint administration to children during endoscopy caused to improve satisfaction of endoscopy team. It also caused to reduce duration time of endoscopy and pyloric spasm.

Keywords: Children, Endoscopy, Peppermint, Satisfaction

Junk Food Consumption and Effects on Growth Status among Children Aged 6-24 Months in Mashhad, Northeastern Iran

Rahim Vakili1, Mohammad Ali Kiani1, *Masumeh Saeidi2, Bibi Leila Hoseini3, Gholamreza Khademi1
1. Department of Pediatrics, Faculty of Medicine, Mashhad University of Medical Sciences, Mashhad, Iran.
2. Students Research Committee, Faculty of Medicine, Mashhad University of Medical Sciences, Mashhad, Iran.
3. Midwifery Department, Sabzevar University of Medical Sciences, Sabzevar, Iran.

Background: Junk food, due to the lack of vitamins, minerals and trace amounts of energy and protein, there is the risk that the child’s stomach filled and by reducing her/his appetite, reduce the chance of nutritious foods. So it is necessary to determine the relationship between using of junk food with growth rate in children.

Methods: This cross-sectional descriptive-analytic study was conducted on 300 mothers and their babies, who were referring to 10 selected Mashhad health-care centers for monitoring their 6-24 months children. Participants were selected by cluster and simple random sampling and valid and reliable questionnaire was used to collect data. Data were analyzed by descriptive-analytic statistics and using SPSS version 16.

Findings: In growth chart, 86.7 percent of children showed appropriate growth, 10.3 percent had delayed growth and 3 percent had horizontal growth curve. In 11.3 percent of families, the junk food has been used for children regularly, 44.7 percent did not believe in these snacks and 44 percent of mothers sometimes used this junk food for their children. Results showed the statistical correlation between junk food consumption and growth status of children was significant, so children whom haven’t had junk food, have grown more favorable than the other kids (P<0.05).

Conclusion: Use snacks interfere with the child's growth. Junk food consumption among the study population was high relatively. Mothers need to be aware of the effects of junk food to children’s development.

Key Words: Children, Growth status, Junk food, Mashhad
Silibinin enhance anti proliferative effect of vitamin D3 on acute myeloid leukemia cells

Vahid Lesan, Department of Biology, Faculty of Food Industry and Agriculture, Standard Research Institute (SRI), Karaj, Iran

Background: The non-toxic polyphenolic component of the extract of milk thistle in known as Silibinin.. In vitro anti-cancer against different cancers, it has been proved that Silibinin is effective. A hematologic cancer which occurs because of blocked differentiation of hematopoietic stem and/or progenitor cells is known as Acute Myeloid Leukemia (AML) that is as a result of different genetic and epigenetic errors and is identified by the uncontrolled proliferation of myeloid blasts. AML accounts for 15-20% of childhood leukemias.

Methods: Cell viability and apoptosis were evaluated trough MTT assay and flowcytometry respectively. By using a commercially kit the activity of caspase 3, 8 and 9 was determined. The differentiation of HL-60 cell to monocyte was estimated by the NBT reduction assay and morphologic researches. All the experiments were accomplished in triplicate and the data are displayed as means±SD. Statistical significances of difference throughout this study were calculated by using a Student’s t-test and one-way variance analysis. \( P \) values <0.05 were considered significant.

Findings: In this study it is indicated that silibinin potent inhibitory effect on vitamin D3 and on proliferation of AML cells is dose dependent. The fact that silibinin increases the differentiation of HL-60 cells into monocytes which is induced by vitamin D3 is pointed out by Cytofluorometric analysis and morphologic studies.

Conclusion: In this study it is proved that silibinin and vitamin D3 separately induces apoptosis and inhibit the growth of leukemic cell lines. In addition, we see synergistic effect between silibinin and vitamin D3. This synergism seems to be occur via activation of caspase 3, 8 and 9 expression, showing the extrinsic and intrinsic apoptotic pathways involvement.

Keywords: Silibinin, Leukemia, vitamin D3, caspase 3

Silibinin inhibit proliferation of Acute myeloid leukemia cells and induce differentiation into monocyte

Vahid Lesan, Department of Biology, Faculty of Food Industry and Agriculture, Standard Research Institute (SRI), Karaj, Iran

The main involved component of silymarin, a standardized extract of the milk thistle seeds is called Silibinin. In vitro anti-cancer impacts on human prostate adenocarcinoma cells, both small and nonsmall human lung carcinoma cells, human eocctercival carcinoma cells, estrogen-dependent and -independent human breast carcinoma cells, and also human colon cancer cells, silibinin has been indicated. Acute myeloid leukemia (AML) accounts for 15-20% of childhood leukemias. Though remission is achieved by following treatment with front-line chemotherapy, almost half of the patients are faced with disease relapse associated with chemoresistance. Consequently, therapies that could maintain the remission phase in pediatric AML are urgently needed. Methods: Cell viability and apoptosis were assessed trough MTT assay and flowcytometry respectively. The activity of PKC was calculated by using a commercially kit and the protein levels of PKC isoforms were determined by western blotting. The differentiation of HL-60 cell to monocyte was estimated by the NBT reduction assay and morphologic studies. Findings: In this study it demonstrated that silibinin inhibit proliferation and induces apoptosis in AML cells in dose dependent manner. The issue that silibinin caused differentiation of HL-60 cells predominantly into monocytes is proved by cytofluorometric analysis and morphologic surveyes. Silibinin improves protein levels of both PKCa and PKCb in HL-60 cells and protein kinase C (PKC) activity. Conclusion: PKC and extracellular signal-regulated kinase (ERK) restrains significantly restrained HL-60 cell differentiation caused by silibinin, showing that PKC and ERK might be involved in silibinin-induced HL-60 cell differentiation. Finally silibinin could be a potent anti cancer agent for targeting acute myeloid leukemias.

Keywords: Silibinin, Leukemia, Apoptosis, PKC, ERK

Evaluation of children with idiopathic thrombocytopenic purpura

Shokoufeh Ahmadipour1, Azam Mohsenzadeh1, Saeedeh Ahmadipour2
1. Pediatrics Department, Lorestan University of Medical Sciences, Khorraramabad, Iran; 2. Pharmaceutics Student, Isfahan University of Medical Sciences, Isfahan, Iran

Background: Idiopathic thrombocytopenic purpura (ITP) is one of the most common causes of thrombocytopenia in healthy children with acute onset. This disease is an autoimmune disorder due to the function of auto antibodies against platelets. Platelets destruct by macrophages FC receptors especially in the spleen and thrombocytopenia occurs. ITP is estimated about 1 in 20,000 in children. The peak age is 1-4 year, males and females are equally affected. ITP is in 2 forms; acute and chronic. Usually thrombocytopenia seen 1-4 weeks after the acute viral infections, and almost cures on their own within 6 months. Approximately 20% of patients who present with acute ITP have persistent thrombocytopenia for \( \geq \) 12 month and said to have chronic ITP.

The aim of this study was to evaluate clinical and laboratory characteristics of children admitted to hospital with a diagnosis of ITP in Khorraramabad pediatric hospital.

Methods: This is a cross-sectional study. All hospitalized children who diagnosed with ITP during 1391-1394 were studied. Necessary tests such as CBC, platelet count, hemoglobin, peripheral blood smear, bone marrow aspiration was requested for patients. Variables such as; age, sex, clinical symptoms, platelet count, anemia, viral infection, treatment, hospitalization time was studied. All collected data were analyzed with SPSS software.

Findings: 56 patients were hospitalized. 85.7%, male and 14.3% were female. 30.3% were between 1-5 yr, 64.3% 5-10yr and 5.3% above 10yr. The most common symptom was purpura and petechiae rapidly throughout the
body. 70% of patients had epistaxis. 50% of patients had platelet counts less than 10,000. 5% of patients had anemia. In the most cases there was history of viral infection 2 to 3 weeks before. 3 patients were positive for H. pylori antibodies. In 30% of patients bone marrow aspiration was done. For 65% of patients IVIG was used. In 85.5% of patients, duration of hospitalization was 2-5 days. 95% fully recovered within 1 month and 15% were suffering from chronic disease.

**Keywords**: children, purpura, thrombocytopenia.

### Evaluation of skeletal changes in children with Acute Lymphoblastic Leukemia

Farzad Kompani, Nazila Rezaei
Children Medical Hospital, Tehran University of Medical Sciences, Tehran, Iran

Evaluation of skeletal changes in children with Acute Lymphoblastic Leukemia Farzad Kompani1, Nazila Rezaei2 1-Department of Hematology and Oncology, Children’s Hospital Medical Center, Tehran University of Medical Sciences, Tehran, Iran 2-Non-communicable Diseases Research Center, Endocrinology & Metabolism Population Sciences Institute, Tehran University of Medical Sciences, Tehran, Iran

**Introduction**: Childhood leukemia is the most common neoplasm in children. Children with acute lymphoblastic leukemia frequently show skeletal changes thus we decided to conduct this study to evaluate skeletal changes in children with acute leukemia.

**Method and materials**: All patients with acute lymphoblastic leukemia that were admitted to our hospital were evaluated. All of them have a chest X-ray that was reported by a radiologist for skeletal changes in chest bone and Humerus.

**Results**: We examined 50 children with acute lymphoblastic leukemia with reference for skeletal pain as well as the nature and the degree of skeletal changes. Metaphisial bands were found in 15% and periosteal reactions in 5% of patients'. There were no skeletal changes in 80% of patients. There was no relations between skeletal changes and sex, age but related with immunologic cell type.**

**Conclusions**: Duration of disease apart from the immunological cell type seems to be an important factor in the incidence of initial skeletal roentgenographic finding.

**Keywords**: Leukemia, skeletal changes, Chest X- Rays
**Etiologies of first attack of acute urticaria in children admitted in Pediatric**

A. Mohsenzadeh, S.H. Ahmadiipour, K. Shahkarami
Emergency Department of Children's Hospital of Khorramabad, Lorestan University of Medical Sciences

**Background:** Acute urticaria (AU) is a common condition in children and affect 20% of individuals at some point in their lives. The most common etiologies of a first attack of acute urticaria in children are foods, medications, insect bites, infections, contact allergy and transfusion reactions.

**Methods:** We retrospectively studied the records of all children admitted by first-attack of acute urticaria at the emergency department of Children's Hospital of Khorramabad from 1992- 1993. Studied variables included age, sex, symptoms, etiologies, treatment, and length of hospital stay. The collected data analyzed by SPSS software.

**Findings:** Of the 50 patients who admitted by first-attack of acute urticaria, 54% was male and 46% female. Age group distribution was: 47% under one year of age, 41% between two and five years and 12% over five years. The most common etiology of first-attack acute urticaria in children was infection 32.4%, foods 30.5%, idiopathic causes 11.2%, medications 12.5%, transfusion reactions 7%, inhalants 1.5%, insect bites 3.5%, and contact materials 0.4%. Ibuprofen and antibiotics were the most common etiology. Clinical presentations were; only skin lesions 48%, respiratory tract symptoms 38.4%, gastrointestinal symptoms 12%. Allergic rhinitis 24.4% was the most prevalent allergic disease, followed by asthma 10% and atopic dermatitis 4.6%. The mean duration of symptoms was 5.4 ±2.05 days. The most common medical treatments were oral antihistamines 62.6%, intravenous steroids 35.4%, intravenous antihistamines 34.6% and oral steroids 18.9%.

**Conclusion:** The most common etiology of first-attack acute urticaria in children was infection, foods and medications.

**Keywords:** acute urticaria, children, emergency department

**Is Meconium aspiration producing asthma in Children?**

F. Naghdi, MD, S.H. Gholyan, MD; Department of Pediatrics Azad University of Medical science

**Background:** Asthma is a chronic inflammatory disease of respiratory tract due to increase of response to stimulators. These are allergens, pollutions, exercise in cold and stress. In many countries, Asthma is most etiology of non attendance students. It produces high morbidity and mortality. In our country, incidence of Asthma is 10%-15% in children and adolescence. Prenatal factors are considered effective for producing Asthma. For example kind of delivery, birth weight, birth age, asphyxia and meconium aspiration.

**Methods:** this is a case- control study. We assessed 622 child between 2-12 years that refer to allergy clinic of Imam Khomeini hospital. We choose patients with Available method. We analyzed the information with SPSS software.

**Findings:** We assessed 622 Asthmatic child between 2-12 years. The results revealed that 76.1% of those with meconium aspiration and 46.8% of those without meconium aspiration had asthma ,showing significant difference(P=0.0001),54.2% were boys and 42.9% were girls. In Asthmatic children there were high incidence of going to kinder garden and contact with pets.

**Conclusion:** In children between 2-12 years. Asthma can relate with meconium aspiration.

**Key word:** Aspiration meconium, Asthma, children.

**Multidimensional relation between allergic disease and early life infections.**

Seid Mohammad Fathi, Department of Allergy and Clinical Immunology, Qazvin University of Medical Sciences, Qazvin, Iran

Allergy and infection are related events and the immune system plays a critical role in both of their pathogenesis. They have multidimensional relationship and interactions. In the second half of 20th century along with universal successes toward control of infectious disease, significant increase in incidence and prevalence of allergic disease was observed. Recent studies show that parasitic and helminthic infections during childhood and presence of specific intestinal bacterial flora like Bifidobacteri in infancy can be protective factors against allergic disease at older ages, accordingly use of probiotics agents as an allergic disease preventive strategy in infants is widely accepted. On the other hand, incidence of bacterial, viral and fungal infections in patients with asthma, allergic rhinitis and atopic dermatitis are significantly higher than normal population. This can demonstrate defects in defense mechanism and immune system of this patients and can cause exacerbation of their symptoms. Infections with micro organisms according to type of micro organism and age of patient can cause significant effect on clinical presentation of atopic infants and children. These interactions can be as an inducer, exacerbator or inversely as a protective and reducer factor.

**Keywords:** Allergy, Infection, probiotics

**The effect of viola odorata flower syrup on the cough of children with asthma: A double-blind, randomized controlled trial**

Mohammad Javad Qasemzadeh1, MD; Hosein Shariﬁ2, MD, PhD; Mohammad Hamedanian1, MD; Mohammad Gharehbagh1, MD; Mojtaba Heydar1, MD; PhD; Mehdi Sardari2, MD; PhD; Meisam Akhlaghoud1, MD; Mohammad Baghey Minaz3, PhD
1. Department of Medicine, Qom Branch, Islamic Azad University, Qom, Iran. 2. Persian Medicine & Pharmacy Research Center, School of Traditional Medicine, Tehran University of Medical Sciences, Tehran, Iran; 3. Research Center for Traditional Medicine and History of Medicine, Shiraz University of Medical Sciences, Shiraz, Iran.

The collected data analyzed by SPSS software.
Background: This study aimed to investigate the effect of violet syrup on cough alleviation in children with intermittent asthma.

Methods: In a parallel, double-blind, randomized controlled trial, 182 children aged 2 to 12 years with intermittent asthma were randomly assigned 1:1 to receive violet syrup or placebo along with the common standard treatments in both groups (short-acting b-agonist). Both groups were evaluated in terms of the duration until cough suppression was achieved.

Findings: No significant difference was observed in basic characteristics. The duration lasting to yield more than 50% cough reduction and 100% cough suppression was significantly less in the violet syrup group compared to placebo (P<0.001, P<0.001, respectively). There was no significant difference in therapeutic effects between boys and girls. There was a significant inverse correlation between the age of children and rate of cough alleviation and suppression by violet syrup.

Conclusion: This study showed that the adjuvant use of violet syrup with short-acting b-agonist can enhance the cough suppression in children with intermittent asthma.

Keywords: Viola odorata, cough, children, asthma, herbal medicine, traditional persian medicine

Relation between asthma and mode of delivery in the asthmatic children

Iraj Mohammadzadeh, Amirkola Hospital, Babol, Iran

Background: Asthma is the most frequent chronic respiratory disease and number of studies shows that the relevance of caesarean with asthma is denied. Regarding high numbers of caesarean in this area, this study was performed with the propose of “determining the relation between asthma and mode of delivery in the asthmatic children” referring to the Amirkola Children’s hospital compared with the control group.

Methods: This analytic study was with case-control groups and performed on 562 children in two groups. 283 ones were attending the case group while the control group included 279 children (between 3-14 years old) referring to the Amirkola Children’s hospital. The collected information in the questionnaire contained: age, gender, birth weight, age of mother at delivery, and environmental factors. After completing the questionnaire, data entered computer and analyzed.

Findings: Distribution of sex between case group (38.2% female, 61.8% male) and control (42.7% female, 57.3% male) did not show any meaningful difference with mode of delivery. The average age in case and control group was respectively 6.2±2.8 and 6.7±3.8 years. Birth weight and weight of cases, exposure to cigarette smoke, gestational age and affecting to severe infection did not show any meaningful difference between two groups. (p>0.05) but contact to animals and maternal age at the time of delivery in both groups was meaningful. (p<0.05) Distribution of delivery mode in case-group (39.1% NVD and 60.9% caesarean) and control (42.2% NVD and 57.8% caesarean) was not meaningful.

Conclusion: The results of our study show that mode of delivery in asthmatic children and control group did not show any meaningful difference.
severities of allergic reactions were recorded after each dose. The serum IgE and SPT were measured at the beginning and at the end of the study.

**Findings:** 14 patients with male to female ratio of 5:2 and the median age of 4.75 (3.7 – 7) years were studied. 13 patients (92.9%) completely closed the buildup and maintenance phase successfully and became desensitized to Cow’s milk. During the build up and maintenance phase 24 (2.0%) and 11 (0.9%) episodes of allergic reactions occurred. The median serum IgE level against Cow’s milk proteins and casein decreased from 39.30 to 10.40 and 7.72 to 2.83 (mm/L), respectively. The median of the difference of the waist diameter with the control, decreased from 10 to 6 mm during the immunotherapy protocol.

**Conclusion:** Oral immunotherapy is effective to decrease the frequency and the severity of allergic reactions. **Keywords:** desensitization, food allergy, cow’s milk allergy

### Assessment of the prevalence of asthma symptoms diagnosed by ISAAC questionnaire in Sanandaj, Kurdistan

**Rasoul Nasiri Kalmazri, Majid Tajik, Kasra Rabeti, Daem Roshani, Seyran Nili, Ali Shekari**

**Besat Medical Research Center, Kurdistan University of Medical Sciences, Sanandaj, Iran**

**Background:** Asthma is one of the most important childhood diseases in developing countries. The main causes of asthma are still unknown. The prevalence, mortality, and economic burden of the disease have taken a rising trend since 1960, and this rise was more marked in children. The present study was aimed to assess the prevalence of asthma symptoms diagnosed by ISAAC questionnaire among children and adolescents living in Kurdistan province, in the western part of Iran.

**Methods:** This study was a cross-sectional study. To collect the required data 2000 questionnaires were distributed in elementary and junior high schools among people aged 6-7 years and 13-14 years; a total of 1940 questionnaires were completed, and the response rate was 97 percent. The utilized sampling method was a combination of cluster sampling (cities) and classification (sex, and educational grade). The students were randomly selected. The sample size was in proportion with the number of students in each area; the number of students in each category was determined by calculating the relevant ratios for sexes and educational grades. ISAAC questionnaire was used as the main data collection tool. After entering the data into SPSS 19 software, they were analyzed using logistic regression and Chi square test.

**Findings:** According to the results of this study, the prevalence of asthma was previously estimated to be 3.9% as diagnosed by physicians; it was estimated to be 3.5% and 4.5% in elementary and junior high schools, respectively, and no statistically significant difference was found. The prevalence of wheezing was 26.5% in Sanandaj in the past 12 months, and there was no statistically significant difference between the two grades. There was a large difference between different areas in terms of the prevalence rates, the difference was statistically significant (P < 0.035).

**Conclusion:** Based on the results of this study, there was a relatively high prevalence of wheezing among the students in Sanandaj the past 12 months. Moreover, in this study the previous diagnosis of asthma in children aged 6-7 and 13-14 years old was 3.4% and 4.3%, respectively, which was higher than that in other similar studies.

**Keywords:** Asthma, ISAAC questionnaire, Children

### Toxic epidermal necrosis and aplastic anemia

**Mansooreh Shariat, MD; Mohammad Gharagozloo, MD; Mohammad Hossein Eslamian, MD; Children Medical Center, Iran. Tehran, Tehran University of Medical Sciences, Iran**

Toxic Epidermal necrosis and aplastic anemia Objective: The objective of this case report is to present a patient with TEN that after receiving IVIG and relative recovery, developed a prolonged fever and aplastic anemia. We started pulse therapy with methyl prednisolone and the patient had a dramatic response. Background: SJS/TEN is a severe drug reaction with skin and mucosal membrane involvement and high mortality. There is keratinocyte apoptosis followed by skin detachment. In TEN skin involvement is more than 30%, and in SJS is less than 10% total body surface area involvement. Visceral involvement including glomerulonephritis, pulmonary and GI bleeding often reported in SJS/TEN. The most common cause of death is sepsis. The recommended treatment is IVIG. Case presentation: A three years old boy developed diffuse erythematous maculopapular eruption one week after Buphen, Amoxicillin and cephalaxin intake. The patient also had positive nikolsky sign and pharyngeal exudation. He was ill, toxic and irritable. IVIG 1 gm/kg was started for patient (4 doses). In hospital course the patient developed anemia, neutropenia, thrombocytopenia, respiratory difficulty and prolonged fever without response to conservative management, IV antibiotic, antifungal and G CSF. Chest X Ray and abdominal sonography was normal. Eventually pulses of methyl prednisolone (30 mg/ kg/ day for three days) was started for patient on 36th days of admission. Fever stopped after few hours, neutrophil count increased after three days and patient discharged with good condition 6 days later. Conclusion: Pulse therapy with corticosteroids can be effective in managing patients with TEN who develop aplastic anemia and prolonged fever after receiving IVIG.

**Keywords:** Toxic epidermal necrosis, Aplastic anemia

### Hereditary angioedema: misdiagnosis, mismanagement and report of seven cases from a family

**Javad Ghaffari, MD; Mohammalsadegh Rezaei, MD; Amir Bahari, MD;**

**Mazandaran University of Medical Sciences, Sari, Iran**

Hereditary angioedema is a rare disorder of complement system which is often seen with autosomal dominant hereditary. Clinical characteristics include non-pruritic and non-pitting mucocutaneous edema that could involve all parts of the body. This study reports seven cases of hereditary angioedema with classical manifestations accompanied by low function of C1INH (type 2). One death occurred due to laryngeal edema. This case study aimed at increasing the knowledge regarding hereditary angioedema, its early diagnosis and correct managements.

**Keywords:** Hereditary angioedema, complement, diagnosis, complication
Evaluation of T-bet and GATA-3 Gene expression in allergic asthma patients

Javad Ghaffari, Atieh Rafatmanesh, Siavash Abedi, Mojtaba Najafi-Saeid Abediankheili
Mazandran University of Medical Sciences, Sari, Iran

Background: Allergic asthma is a chronic inflammatory disease identified by high response to allergens and excessive air ways edema. T-bet and GATA-3 are two transcriptional factors that differentiate Th1 and Th2 from Thnaive. In this study, we examined the expression levels of these two factors in patients with allergic asthma in comparison with healthy controls.

Materials and Methods: In a case-control study, 26 patients with allergic asthma and 26 healthy subjects were studied who were matched for age and sex. Sampling was done and peripheral blood mononuclear cells were isolated and CDNA was synthesized after RNA extraction. Gene’s expressions were evaluated by Real-time Polymerase chain reaction.

Findings: The results showed that T-bet and GATA-3 expression levels significantly increased in patients in comparison with control group. In addition, GATA-3/T-bet ratio showed a significant increase in case group (P=0.005).

Conclusion: This study showed that the genes expression of T-bet and GATA-3 regulate the balance of Th1/Th2. Therefore, evaluation of T-bet and GATA-3 is believed to have an important role in treatment and clinical condition of asthma patients.

Keywords: Allergic asthma, T-bet, GATA-3

In vitro comparison of chromosomal radiosensitivity in common variable immunodeficiency and acute lymphocytic leukemia patients

Majid Mahmoodi, Hassan Abolhassani, Farideh Farzanfar, Nima Rezaei, Gholamreza Azizi, Reza Yazdani, Kouroos Divsalar, Rassoul Nasiri-Kalmar
Asghar Aghamohammadi
1. Cancer Institute, Tehran University of Medical Sciences, 2. Research Center for Immunodeficiencies, Children’s Medical Center, Tehran University of Medical Sciences; 3. Department of Immunology, School of Medicine, Tehran University of Medical Sciences, Tehran, Iran

Background: Common variable immunodeficiency (CVID) is one of the predominantly antibody deficiencies that cause significant morbidity and mortality. Despite in vitro assays to evaluate radiosensitivity have been frequently used in order to select the most appropriate future treatments, the frequency of chromatid breaks and rearrangements. Surprisingly first degree relatives of CVIDs had similar G2 aberration frequencies to CVID cases which showed significant difference with normal controls. Conclusions: Results indicate that most of the CVID patients and their first degree relatives are sensitive to ionizing radiation similar to the ALL cases. These patients should be protected from unnecessary diagnostic and therapeutic procedures using ionizing radiation

Keywords: Common variable immunodeficiency; chromosomal radiosensitivity; acute lymphoblastic leukemia; Ataxia telangiectasia

Scoring the recurrent infections in children with primary antibody deficiencies

Ahmad Bahrami, Iran University of Medical Sciences, Tehran, Iran

Background: Primary antibody deficiencies (PADDs) are heterogeneous group of disorders, characterized by hypogammaglobulinemia and increased susceptibility to recurrent infections. To evaluate the diagnostic delay in Iranian PADDs in association with their infections, we scored such manifestations to find an association between such delay and the scoring system.

Methods: Forty-eight patients with PADDs, who were referred to our center during a 25-year period, were enrolled in this study. Each episode of infection, before making the PADDs diagnosis, got a score of 5 or 10 based on the severity of infections.

Findings: The diagnosis was made with median delay of 34.5 months, when the patients had mean score of 94.48 ± 52.89. There was a significant direct association between this scoring system and delay diagnosis. The score of 50 was considered as the cutoff point in our patient group. In this score, the suspicions to PADDs in more than 90% of patients true positively lead to diagnosis of PADDs.

Conclusion: Although diagnosis delay significantly decreased over time, PADDs still continue to be diagnosed late. Based on the results of this study, the assessment of immune system should be performed in the patients with total score or about 25 score per year.

Keywords: primary antibody deficiencies, scoring system, recurrent infections

Neonatal effects of substance abuse during pregnancy

K. Shahkarami, MD, PhD Student of Addiction Studies; A. Mohsenzadeh, MD; S.H. Ahmadipour, MD
1. Neuroscience Department, School of Advanced Technologies in Medicine; 2. Lorestan University of Medical Sciences, Iran

Background: Nearly 90% of drug-abusing women are of childbearing age. Maternal drug addiction can cause problems for the fetus and the newborn. Perinatal death, prematurity, fetal growth retardation, neonatal abstinence syndrome (NAS), and a wide variety of other perinatal complications have been frequently observed in the offspring of addicted women. The aim of this study was to...
assess infant with prenatal drug exposure.

**Methods:** This study was a retrospective record review of data from 25 infant with addicted mothers compared with the control group who were referred to health centers of Khorramabad from 1388-1393. Studied variables included gestational age, preterm deliveries, birth weight, Congenital anomalies, and length of hospitalization. The data collected by questionnaire and analyzed by SPSS.

**Findings:** Preterm delivery (35%) was significantly more frequent compared with control group (6%). Gestational age at delivery was 34.1 ± 2.5 which is less than control group (37.4 ± 1.6). Children born to addicted mothers had lower birth weight (2,315 ± 69 vs 3,280 ± 540.5). The risk of various congenital anomalies was 6% in the group of children born to addicted mothers. Congenital cardiac defects were more common. Neonatal abstinence syndrome developed in 75%. Length of hospital stay was 18 ± 9 days.

**Conclusion:** Addicted pregnant women are high-risk group, according to perinatal outcome.

**Key words:** neonate, substance abuse, pregnancy

---

**Update on Pediatric Phagocyte Defects**

Nima Rezaei, MD, PhD, Research Center for Immunodeficiencies, Children's Medical Center, Tehran University of Medical Sciences, Tehran, Iran; and Department of Immunology, School of Medicine, Tehran University of Medical Sciences, Tehran, Iran

Phagocyte defects are a category of primary immunodeficiency diseases (PIDs), consist of defects of neutrophil differentiation, defects of motility, defects of respiratory burst, and Mendelian susceptibility to mycobacterial diseases. Severe congenital neutropenia is one of the most frequent phagocyte defects, which could be due to underlying genetic defects in one of the following genes: ELANE, GFI1, HAX1, G6PC3, VPS45. Cyclic neutropenia, glycogen storage disease type 1b, p14 deficiency, Barth syndrome, Cohen syndrome, and poikilodermata with neutropenia are other neutrophil function/differentiation defects. Leukocyte adhesion deficiency (types I-III), 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 (CGD) is the prototype of defects of respiratory burst, which could be due to underlying genetic defects in one of the following genes: CYBB, CYBA, NCF1, NCF2, NCF4. Finally, Mendelian susceptibility to mycobacterial diseases is another group of phagocyte defects, which predispose individuals to mycobacterium, and could be due to mutation in one of the following genes: IL-12RB1, IFNGR1, IFNGR2, IL12B, STAT1, CYBB, IRF8, ISG15. GATA2 deficiency (Mono MAC Syndrome), pulmonary alveolar proteinosis along with autosomal recessive form of IRF8 deficiency are other diseases that have been classified as phagocytes defects. Clinical description of patients could help us to have better understanding on nature of these diseases, while long-term follow-up of patients with known gene mutation(s) could give us more insights into the pathophysiology of diseases.

**Keywords:** Immunodeficiency, phagocyte defect, approach, neutropenia

---

**Association of Chediak-higashi syndrome and Hemophagocytic lymphohistiocytosis, a case report**

Seyed-Saeed Hosseininodeh1; Farzad Kompani2; Mojtaba Gori3

Children's Medical Center. Tehran University of Medical Sciences.

**Background:** Chediak-Higashi syndrome (CHS) is an infrequent, autosomal recessive disorder characterized by severe immunodeficiency, frequent bacterial infections, tendency to bleeding, variable albinism, and progressive neurologic dysfunction. Hemophagocytic lymphohistiocytosis (HLH) is a clinical syndrome, caused by severe hypercytokinemia due to a highly stimulated but ineffective immune response. Association of CHS and HLH is rare and few cases have been reported since now.

**Case presentation:** Here we report a case of HLH that diagnosed as a CHS by skin and hair biopsy and multiple admissions for pulmonary infections thereafter. We also describe two same associations of HLH and CHS.

**Conclusion:** Early diagnosis and treatment of HLH and CHS may help prevention of multiple infections and reduced number of hospitalizations.

**Keywords:** Chediak-higashi, HLH, Hemophagocytic lymphohistiocytosis
Detection of Cytomegalovirus (CMV) Antibodies or DNA Sequences from Ostensibly Healthy Iranian Mothers and their Neonates

Fatemeh Hoda Fallahi1, Bahman Abeli Kiasari2, Seyed Hamidreza Monavari3
1. Loghman Hospital, Shahid Beheshti University of Medical Sciences, Tehran, Iran; 2. Human Viral Vaccine Department, Razi Vaccine & Serum Research Institute, Hesarak, Karaj, Iran; 3. Department of Virology, Tehran University of Medical Sciences (TUMS), Tehran, Iran

Background: Cytomegalovirus (CMV) remains the most common cause of viral intrauterine infection. The objective of this research was to determine the prevalence of at-risk pregnancies for congenital cytomegalovirus transmission in a randomly selected pregnant women and their newborns.

Methods: Enzyme link immunosorbent assay (ELISA) and real-time polymerase chain reaction (PCR) were utilized to screen the sera of mothers (n=100) and consecutive umbilical cord blood samples from their newborn (n=100).

Findings: Of the 100 mother's sera analysed, 100 (100%) and 3 (3%) were positive for cytomegalovirus IgG and IgM antibodies respectively. Of the 100 cord serum specimens analysed, 99 (99%) and 2 (2%) were positive for cytomegalovirus IgG and IgM antibodies respectively.

Cytomegalovirus DNA was detected in 4 out of 100 (4%) cord blood samples of newborns. From four CMV DNA positive cases, case 1 had no IgM in cord serum, but had IgM in mother's sera. Cases 2 and 4 were positive for IgM in both mother's sera and cord serum. Case 3 had no detectable CMV IgM in sera and cord serum.

Conclusion: As many as 66% and 100% of CMV IgM-positive women in this study also had CMV IgM and CMV DNA in their delivery cord blood samples respectively suggesting an increased risk of congenital CMV infection in those pregnancies. A paired women sera/cord blood CMV IgM-negative was found to be positive for CMV DNA. The data may also suggest the utility of PCR in place of CMV IgM as a diagnostic method for congenital CMV infection.

Keywords: Cytomegalovirus, Health care worker, PCR

The common bacteria species isolated from urine culture, and their drug resistance

Temenk Samice-Rad1, Meht Kahriz2, Mahsa Zaare-Ardestani2, Maryam Rahmani3, Fatemeh Mohamadi1, Nader Khosro Shahi1
1. Qazvin Metabolic Research Center, Qazvin University of Medical Sciences, Qazvin, Iran; 2. Khorramabad, Tehran University of Medical Sciences, Tehran, Iran; 3. Tehran University of Medical Sciences, Tehran, Iran

Background: Urinary tract infection (UTI) is a main cause of neonatal fever. But the diagnosis of UTI can be missed because the symptoms are often non-specific and the sterile sampling may be difficult. In the neonate the clinical approach and radiologic investigations of UTI are quite different from that of older children and adults. Our objective is to determine the common bacteria species isolated from urine culture, and their drug resistance in the Kosar Hospital.

Methods: For this cross-sectional study data were evaluated from 21 babies with positive urine culture reports that admitted at the Kosar Hospital between March 2008 and December 2012. The data from these participants were obtained by a check list and review of laboratory report sheets of urine culture results and antibiograms. Both descriptive and statistical analysis methods were applied.

Findings: Eleven subjects were boys. Six of the 21 subjects had low birth weights (< 2,500 gm). Their mean gestational age was 34 ± 3/7 weeks. The most common complaint was fever (83%), followed by poor appetite, diarrhea, vomiting and abdominal distention. Pyuria was found in 17 patients. Escherichia coli was the most common pathogens, found in 14 cases, followed by Klebsiella pneumonia (2 cases), Enterococcus (3 cases) and Enterobacter cloacae (2 cases).

Conclusion: The clinical presentations of UTI in neonate patients are nonspecific and urinary tract obstruction manifestations often followed by repeated UTI. Physicians need to be alert and sensitize to these important clinical presentations. Also any UTI in neonate patients should be worked up for urinary obstruction or reflex. Further prophylactic approach, either medical or surgical, is performed to decrease the renal damage.

Keywords: Urine culture, Urinary tract infection

Prevalence of thrombocytopenia in patient who admitted to the intensive care unit and its relation to bacterial sepsis

Hamid-Reza Sherkatolabasieh1, Shiva Shafezadeh2, Azade Jafarstae, Azam Mohsenzadeh1, Mojgan Goodarzy, Naderie Taei, Aliakbar Razliarnsy
1. Department of Pediatrics, School of Medicine, Lorestan University of Medical Sciences, Khorramabad, Iran; 2. Internal Medicine Department, School of Medicine, Lorestan University of Medical Sciences, Khorramabad, Iran

Background: Thrombocytopenia is a common disorder in patients who admitted to the intensive care unit (ICU) that has been showed as a bad prognostic factor for mortality and has a various causes. The aim of this study was to determine the prevalence of thrombocytopenia and its relation to bacterial sepsis as one of causes of this disorder in patients who admitted to the ICU.

Methods: This study was a retrospective analysis of all patients who hospitalized in ICU of a teaching hospital affiliated to Kermanshah University of Medical Sciences from 2012 to 2014.

Findings: Of 387 patients, 231 patients (59.6%), at least one time, afflicted by thrombocytopenia. There wasn’t any significant difference between age and gender by thrombocytopenia incidence (P>0.05). Totally, 76 patients (19.6%) had positive blood culture that 58 cases (76.3%) occurred in thrombocytopenic patients.

Conclusion: The results of this study was determine that thrombocytopenia incidence is a good prognostic factor in morbidity and mortality of patients who hospitalized in...
The prevalence and antimicrobial pattern of gram-negative bacteria isolated from urine of children with urinary tract infection

Bahram Nikmanesh1, Mohammad Taghi Haghi Ashtiani1, Molood Barzan2, Narges Nodar Jafari1
1. Children’s Medical Center, Pediatrics Center of Excellence, Tehran university of medical sciences, Tehran, Iran; 2. Department of Lab Medical Sciences, School of Allied Medical Sciences, Tehran University of Medical Sciences, Tehran, Iran; 3. Department of Microbiology, Faculty of Advanced Sciences & Technology, Pharmaceutical Sciences Branch, Islamic Azad University, Tehran, Iran

Background: One of the most important childhood infections is urinary tract infection (UTI). In order to prevent serious complications of UTI in children such as hypertension and renal failure, definitively diagnose and prompt treatment are essential. Since bacteria of the family Enterobacteriaceae are known to be the most common causes of UTI, the present study aimed to determine the frequency and antimicrobial resistance patterns of them in children with UTI.

Methods: The present study was conducted on urine samples of children with UTI referred to Children’s Medical Center of Tehran during one year. The urine samples were cultured on selective media and the bacteria were identified by biochemical tests. Antibiotic resistance pattern of isolates were investigated by disk diffusion method.

Findings: Out of 1348 positive urines for Enterobacteriaceae bacteria, more cases of UTI were observed in outpatient (1050, 77.89%) than in hospitalized patients (298, 22.11%). Female patients were 911 (67.58%) and male patients were 437 (32.42%) and 68% of patients were aged less than 2 years. E.coli was the most common bacteria isolated among family Enterobacteriaceae, with prevalence of 76%. Results of antimicrobial susceptibility testing showed that clinical isolates had the most sensitivity to Amikacin and Piperacillin-Tazobactam, respectively (93%), and (98%) and resistance to Cephalothin (80%).

Conclusion: Considering the prevalence of urinary tract infections, especially in children under 2 years and also in girls, the knowledge of local resistance pattern and well-timed eligible treatment are imperative. Accordingly, Amikacin and Piperacillin-Tazobactam are recommended for empirical treatment in children with UTI.

Keywords: gram-negative bacteria, UTI, Piperacillin-Tazobactam,

Status of vaccination coverage in children 24 months to 6 years in urban areas and high risk areas covered by Tehran University of Medical Sciences

Malahat Khalili, Ali Nikfarjam, Ahmad-Reza Shamsihi, Zahra Hassanpour
Gilan University of Medical Sciences, Rasht, Iran

Background: Children immunization program is the most successful and most cost-effective public health interventions and medical programs. The study was designed and conducted in order to assess vaccination coverage in areas covered by Tehran University of Medical Sciences.

Methods: This study was a cross-sectional study of children aged 24 months to 6 years living in urban areas and high-risk and marginal areas is covered by Tehran University of Medical Sciences. 630 children in 90 clusters in urban areas and cluster sampling in areas with high risk were done as well as Probability Proportional to Size Sampling and sample size of 768 children were determined. Questionnaires were completed by interview with mothers or guardian of a child and evaluation of vaccination cards. The collected data were analyzed using descriptive statistics and Chi-square analysis was used for statistical analysis.

Findings: 750 children (97.7%, 95% CI: 96.32-98.60%) in high risk and marginal areas and 616 children (97.8%, 95% CI: 96.3-98.78%) in urban areas had complete immunization histories. Ignorance of immunization program (50%) was addressed as the main reason for incomplete immunization.

Conclusion: Universal mass vaccination coverage of children and equal coverage in marginal and urban areas indicate the success of the authorities in the implementation of this program. Organizing training courses for mothers about immunization program, providing the required facilities and services, monitoring and supervision of vaccination at health care centers, home health care seem to be essential in maintaining and improving the existing status.

Keywords: vaccination, immunization, converge, children

The comparison of methicillin resistant staphylococcus aureas (MRSA) nasal colonization in children of health care workers (HCWs) and non HCWs attending day care centers in Hamedan, 2014

Iraj Sedighi, Mehrnaz Olfat.
Hamedan university of medical Sciences, Hamedan, Iran

Background: Staphylococcus aureus is a gram-positive cocci responsible for significant human infections and its main colonization site in humans is anterior nares. Methicillin resistant Staphylococcus aureus (MRSA) colonization occurs more frequently among health care workers (HCW) and they can transmit MRSA to other peoples and provoke community acquired MRSA (CA-MRSA) which is incresingly seen among children without risk factors. In this survey, we compare the prevalence, antibiotic susceptibility pattern and risk factors associated with nasal carriage of Staphylococcus aureus and MRSA between healthy 1 to 6 years old children of HCWs and non HCWs attending day care centers. Method: This cross sectional study was conducted on 67 one to 6 years old healthy children of HCWs (case group) and 67 of non HCWs (control group) attending day care centers in Hamedan, Iran in 2014. Demographic features and other nessecary informations collected via questionnarie. Nasal swabs were obtained and trasfered to our microbiology laboratory and cultured for staphylococcus aureus. Antibiotic susceptibility and erythromycin induced clindamycin resistant test (D-test) was done for positive specimens.
Findings: 12 (17.9%) and 22 (32.8%) of children were colonized with Staphylococcus aureus in case and control group respectively (p.value=0.052). only one child in case group was colonized with MRSA, which was resistant to vancomycin and clindamycin too. Any child of control group was colonized with MRSA. Factors associated with nasal colonization of Staphylococcus aureus were older age in both case group (p=0.023) and control group (p=0.017) and lack of breast feeding history in control group (p= 0.037).

Conclusion: we conclude that parent jobs has no effect on nasal colonization of staphylococcus aureus and MRSA. In addition, MRSA prevalence among children of our area is uncommon.

Keywords: nasal carriage, methicillin resistance staphylococcus aureus, parent jobs, healthy children

A 14 month-old baby with secretory, brown to black lesion on his nostril with edema around that

Parisa Rahmani Ahranjani1, Azadeh Ghare Zadeh Shirzadi2, Hamid Eshaghi1
Children’s Medical Center, Tehran University of Medical Sciences, Tehran, Iran.

A 14 month-old boy was brought to the emergency clinic of Children Medical Center (CMC) presented with large area of edema around that, extending to his eye and neck on the same side. This lesion appeared firstly as a small erythematous papule 3 days before bringing to hospital which ulcerated from the center and changed to red-brown ulcer with large edema. He also had 3 reactionary cervical lymph nodes on that area, low grade fever and a honey-colored scab around the lesion. He didn’t have poor-feeding or lethargy in recent days. He received Cephalexin with the impression of impetigo and Hydroxyzine for one day. As he was living in rural area, he had a history of contact with animals like sheep, cows and manure. On the dorsum of his father’s hand we found a small well defined black painless crusted ulcer since 2 weeks ago without any treatment. He was a normal baby and except the lesion mentioned before had unremarkable physical examination. He was vaccinated as routine of Iran’s vaccination program. The CBC and CRP were normal but ESR was as high as 17 mm/h. The smear and culture from secretion revealed bacillus anthracis, so Penicillin as the antibiotic of choice for Anthrax started for the patient. The patient became better dramatically by even the first doses of antibiotic. The antibiotic course continued for 10 days.

Comparison of urinary antigen test and PCR for detection of Legionella pneumophila in hospital-acquired pneumonia patients

Ali-Akbar Rahbarimanesh, Hosein Shokri, Samileh Noorbakhsh, Mohammad-Reza Shokri, Abolfazel Farid
Bahrami Children Hospital, Tehran University of Medical Sciences, Tehran, Iran

Legionella pneumophila is the causative agent of more than 95% cases of severe legionella pneumonia. Nosocomial pneumonias in different hospital wards impose an important health threat. The main objective of this study is to detect legionella with two methods of polymerase chain reaction (PCR) and urine antigenic test in patients suffering from nosocomial pneumonia admitted to pediatric intensive care unit (PICU) of children hospitals.

Methods: This study was conducted in PICU wards of Rasool Akram and Bahrami children hospitals, Tehran, Iran during 2013 - 2014. In patients diagnosed with hospital-acquired pneumonia, intratracheal secretion samples for PCR and urine sample for urinary antigen test were taken. Simultaneously, PCR and urinary antigen test were conducted using commercial kits. The results of urinary antigen test and PCR were analyzed by SPSS v.19 for statistical comparison.

Findings: With 96 patients being studied, the average age of patients was 2.77 years with two age peaks of under 1 year and 7-8 year. More than half of the patients were under 1 year old. The most common underlying diseases were seizure, ALL, Down syndrome and metabolic syndromes. The positivity rate of legionella urinary antigen test was 16.7% and positivity rate of PCR test was 19.8%. No significant association was found between the results obtained by both assays with age, gender or underlying diseases.

Conclusion: PCR method is merely more efficient in legionella detection than urinary antigen test. The difference between these two methods was not significant.

Keywords: Legionella pneumonia, Urinary antigen test, hospital-acquired pneumonia, pediatric intensive care unit

Prevalence of Human T-lymphotropic Virus in patients with Multiple Transfusions

J. Ghaffari (MD)1, J. Yazdanicharati (PhD)2, M.R. Haghshenas (PhD)3, Mazandaran university of medical sciences,sari,Iran

Background: Human T-lymphotropic virus type I (HTLV-1) is a member of the retrovirus family, which often leads to the asymptomatic infection of individuals. Transmission of this virus is possible through breast-feeding, sexual contact, transfusion of contaminated blood products and use of contaminated needles. This study aimed to review the prevalence of HTLV-1 infection in high-risk patients, particularly those with thalassemia, hemophilia and hemodialysis in Iran and other countries.

Methods: In order to find related articles, we searched in different sites including Google, Yahoo, PubMed, Irandoc, Iranmedex, Magiran and SID using keywords such as HTLV-1, high-risk, transfusion, thalassemia, hemodialysis, Iran and world.

Findings: In total, 45 articles were found in relation to the subject of the study. Based on the specified criteria of the study in the Iranian population, 17 papers conducted in relation to the prevalence of HTLV-1 infection in patients with thalassemia major and hemophilia were collected, 14 cases of which were about patients with thalassemia alone or those with a combination of hemodialysis and hemophilia. As for the studies conducted in other countries, 12 articles were found, 7 cases of which were about thalassemia patients; therefore, they were selected and studied.

Conclusion: According to the results of this study, the prevalence of HTLV-1 infection in high-risk patients with thalassemia, hemophilia and hemodialysis in endemic areas was higher than non-endemic areas.

Keywords: Thalassemia, Hemodialysis, Hemophilia, Human retrovirus type I