



# **22<sup>nd</sup> International Congress on Pediatrics**

**&**

## **8<sup>th</sup> Congress on Pediatrics Nursing**

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## Cardiology

### Intrapericardial teratoma with pericardial effusion causing tamponade in a 2 day old neonate

*Radvar, M., MD; Salehi Ardabili, Sh., MD; Motahari & Seied-Al-Shohada Hospitals, Urmia University of Medical Sciences*

**Background:** Intrapericardial teratoma is a rare benign cardiac tumor which can be potentially fatal, because of pressure on the heart and great vessels. It is usually surgically removable. Macroscopically this tumor is a lone, large, polycystic mass, in contact with the base of the heart and accompanied by pericardial effusion.

**Case report:** Patient was a 2 day old, term, male neonate that admitted in NICU, because of respirator distress. The neonate had tachypnea (RR>55/min), tachycardia (HR>180/min) and weak pulses. Echocardiography showed moderate pericardial effusion and a large, cystic, space-occupying, intrapericardial, lesion on the anterior of the heart, which was pressing on the right atrium and right ventricle. Chest CT scan confirmed echocardiographic findings. The baby emergently referred for cardiac surgery and the tumor successfully removed. The postoperative course was uneventful and in outpatient follow up six months later, The infant shows normal development and has a normal echocardiogram. **Conclusion:** Intrapericardial teratoma with pericardial effusion can cause tamponade in early days of life, early cardiac surgery is life saving. These tumors are usually surgically resectable. In this case there was no recurrence of tumor 6 months later.

**Keywords:** Intrapericardial teratoma, Tamponade, Neonate

### Cardiovascular Effects of Methylphenidate in Children and Adolescents with Attention Deficit Hyperactivity Disorder

*Isa Tafreshi, R., MD; Jalili, B., MD; Iran University of Medical Sciences*

**Background.** Attention deficit hyperactivity disorder (ADHD) is a common neuropsychiatric syndrome in children and adolescents that impair daily living activities. Although benefits of methylphenidate (MPH) have been documented, increasing concerns have been raised about cardiovascular (CVS) effects of this drug. The aim of the present study was to assess the short-term CVS effects of MPH. Methods.

We studied 130 patients with ADHD (aged: 6-17 years, mean age: 7.9 ± 1.3 years, 68% male) who were entered into an open-label study of MPH up to 6 months. The subjects received each of three methylphenidate doses (0.3, 0.5 and 1 mg/kg) for a period of 2 weeks per medication dosage. ECG indices, BP and HR were recorded at 2, 8, and 24 weeks after treatment. Results. The patients were followed up over 6 months period. Mild increase in systolic BP was detected in 35% of the patient but the difference was not statistically significant. Two subjects met the criteria to be considered hypertensive (systolic BP values > 95th percentile for age and height). Palpitation and sinus tachycardia were observed in two (1.8%) patients early after the medication started. One subject had prolonged PR interval that no change was seen after initiation of medication. MPH does not associate with HR-corrected QT prolongation. Conclusions. MPH was generally well tolerated. No serious CVS adverse effect occurred. However, further investigations on different doses of MPH are required to replicate these findings in children with ADHD.

**Keywords:** Attention deficit hyperactivity disorder, cardiovascular effects

### Pediatric chest pain: a retrospective study

*Towhidifar, MH., Social Security Organization Yazd*

**Background:** Chest pain is a common presenting complaint in children. The etiology is benign in most cases, but it may lead to considerable anxiety in patients and their families. A thorough history and physical examination can determine positive findings which are useful guide to diagnosis. Laboratory testing is necessary only in a small number of patients. However, in the absence of characteristic findings some modalities such as chest x-ray (CXR) and electrocardiography (EKGs), could be helpful. This study was performed to find a particular diagnostic role of EKGs in children with chest pain who visited in emergency room. Methods: In this retrospective study, all children in age range of 4-12 years, with chest pain who were admitted to the emergency room in one year were included. A total of 73 (39 boys 24 girls) were evaluated. The most common causes of the pain were as followed; 30 (41%) due to respiratory problem particularly asthma, 13 (17.80%) due to musculoskeletal causes, 9 (12.3%) idiopathic and 8 (10.9%) related to gastrointestinal source. Cardiac problems were found in (3) 4.1%. For patients with ill-defined chest pain CXR, EKGs, echocardiograms or other test were performed for individual indications. ECGs were obtained in (27) 36.98%; except for tachycardia and arrhythmia that was seen in one patient, results of EKGs were normal in emergency room. More evaluation including CXR and echocardiogram, discovered cardiomyopathy in this patient who was treated for

asthma previously. Of note, one patient with simultaneous chest pain and abdominal pain, that her pain supposed to be for abdominal source and admitted to surgery, candidate for appendectomy, the day after diagnosed as rheumatic fever with sever cardiac involvement. For this patient EKGs was not performed first time.

**Conclusion:** By through inspection, determination of sources of chest pain often is straightforward, but it couldn't discover some serious problem in special condition. Considering that psychogenic and idiopathic pain are less common than previously believed, we suggest to obtain EKGs for every child with unexplained chest pain.

**Key Words:** chest pain, EKG, cardiac pain

### **Device closure of patent ductus arteriosus in Marfan patients: safety and effect on the aortic root diameter**

*Keyhan Sayadpour, AR., MD, Masood Sadiq, W. Weber, HS., Cutle, NG.; Children's Medical Center, Tehran University of Medical Sciences*

The evidences in the medical literature about safety of PDA device closure and its effects on aortic root diameter in Marfan patients are limited. We report our experience of PDA closure by coil or Amplatzer Duct Occluder in nine Marfan patients. Two patients had aneurismal type E ducts which were closed by a single coil respectively. The others had conical ducts for which even Amplatzer occluder, stiff, or double coils were used. No untoward consequence was found during a median follow up period of 18 months. Two patients had residual shunt. Seven patients had complete data on aortic root diameter before and after the procedures: that was decreased in 5 and increased in 2. We concluded that PDA device closure in Marfan patients is safe. Risk of residual shunt is higher even after closure of small ducts.

**Keywords:** Marfan Syndrome, Patent Ductus Arteriosus

### **Cardiac tumor in fetus with diabetic mother**

*Voshtani, SH. MD, Guilan university-Rasht-Iran*

**Introduction:** Neonate cardiac tumors are rare entities. Cardiac tumors are benign or malignant neoplasms arising primarily in the inner lining, muscle layer or the surrounding pericardium of the heart. Rhabdomyoma is the most common cardiac tumor during fetal life and childhood.

**Method:** Today the non-invasive diagnostic methods of these tumors are used the instrument such as echocardiography, CT and MRI. A 25 year old

pregnant woman with gestational diabetes mellitus was checked for diagnosis of congenital heart defect at 32nd weeks of pregnancy. The mean blood sugar of mother was 160 mg/100 ml at the time of pregnancy. The woman uses insulin for controlling the blood sugar in normal range. In her fetal echocardiography, a mass was observed in right ventricle which attached to intraventricular septum right ventricular apex by pedicle. Five days after cesarean section, the infant echocardiography revealed the tumor by size of 19mm length and 10mm width which was attached to right ventricle apex. Result: The infant was followed for twice every four months. In the second echocardiography after delivery the tumor size decreases to 11mm length and 6.2mm width, at the third echocardiography done at eight months after delivery the size also decreases more and reached to the size of 8.4mm length and 4.5 mm width.

**Conclusion:** It is known rhabdomyoma is the most common cardiac tumor during fetal life which can be regressed at several months after delivery, so surgical intervention is no longer indicated.

**Keywords:** cardiac tumor, neonate, diabetic

### **Evaluation of symptoms frequency and risk factors related with neonatal Patent Ductus Arteriosus(PDA) in neonates affected with PDA in Qaem Hospital Neonatal Intensive Care Unit(NICU) in 2007-2008**

*Saeidi, R. MD, Mahmoodi, E., Esmaeili M., Gholami Robatsangi, M.; Qaem Hospital*

**Background:** Ductus Arteriosus which is an essential route part of fetal circulatory system, tends to Patent Ductus Arteriosus(PDA) if remains patent after birth. Since this anomaly has become more apparent with the advance in medical equipments and because of the high prevalence of PDA which was (15%) in patients admitted at Rejaee Hospital in Tehran according to the research done there, and compared to the prevalence of (2-7%) in United States, we have decided to make a research to reveal the prevalence of the manifestations and risk factors of this disease.

**Methods:** This research is a cross-sectional descriptive study and done on 100 neonates admitted in Neonatal Intensive Care Unit(NICU) in Mashhad Qaem Hospital. Our study has focused on data include sex, diagnostic age, maternal diabetes and hypertension, mother age; and on patients signs like apnea, repetitive apnea, bounding pulse, heave, systolic and continuous murmur, respiratory distress and the risk factors: prematurity, birth weight, hypoxia, acidosis.

**Findings:** Through 100 neonates affected by PDA, 54% were males and 46% were females. The most common sign within this neonates was systolic murmur (at 89% of the cases), then respiratory distress (72%), continuous murmur (37%), bounding pulse (28%). With respect to the risk factors which have been detected, hypoxia (71%) and acidosis (70%) are

considered as first degree risk factors, where as prematurity(41%)and low birth weight (40%)as 2nd degree ones.68% of all neonates have had another anomaly than PDA.

**Conclusion:** Based on the high concurrence of cardiac and respiratory manifestations in PDA patients, strict cardiac and respiratory examination helps us to set the diagnosis of PDA on time.Since prematurity and low birth weight have been important risk factors in this disease,neonates having these 2risk factors should be evaluated to rule out PDA in them.

**Keywords:**Patent Ductus Arteriosus, systolic murmur, low birth weight, prematurity

### **Treatment of drug refractory ventricular tachycardia by omega-3 fatty acid supplementation**

*Abtahi, S., MD; Islamic Azad University, Mashhad Branch*

**objective:**conventional pharmacological and interventional approaches are often ineffective in refractory VT. We present a novel case report illustrating the potential use of omega-3 fatty acid supplementation in such patients. Both conventional anti-arrhythmic drugs and anti-tachycardia pacing often provide only partial control. Radiofrequency ablation, the mainstay of treatment is technically challenging in children, and associated with significant procedural risks. We present the potential use of omega-3 fatty acid supplementation in such patients.

**method:** A 5-year-old child presented with hypotensive sustained ventricular tachycardia sinus rhythm was restored by DC cardioversion. Echocardiography revealed Dilated cardiomyopathy. intravenous and then oral amiodarone were administered. the maintenance dose of oral amiodarone was increased to maximum dose, He continued to experience frequent episodes of monomorphic sustained ventricular tachycardia Mexilitine was added, but had little effect on the frequency of ongoing ventricular tachycardia. Radiofrequency ablation was not available for this age, then he was commenced on treatment with

omega-3 fatty acid supplementation (seven seas, pure cod liver oil 0.5 cc/kg/day)).

**Results:**At further review 4 weeks later, he felt well and, dramatically Prior to the initiation of omega-3 fatty acid supplementation, he had been experiencing 8-10 documented episodes of sustained ventricular tachycardia per day conclusion:He has remained well for the past 4 months, since the introduction of omega-3 fatty acid supplementation, and there have been no further episodes of sustained ventricular tachycardia, such that radiofrequency ablation has not been required to date.

**Keywords:**Ventricular Tachycardia, Omega-3, Treatment

### **Surgical outcome in coronary artery fistulas repair in children**

*Malekhamdi, MR, Hajar Hospital, Shahrkord University of Medical Sciences*

**Background:** Coronary artery fistula (CAF) is a rare congenital anomaly that can be complicated by endocarditis, myocardial infarction or coronary aneurysms. The purpose of this article is to review the clinical characteristics and surgical outcome in 10 patients with CAF in shahid Rajaei hospital. Patients and Methods: From 2000 to 2009, ten patients (age, 6 months to 15 years, mean age, 8.5 years) were diagnosed with CAF by echocardiography and cardiac catheterization. Six were female and 4 were male. All patients with isolated CAF (9) were asymptomatic. One patient with associated anomaly (MVP with severe MR) had dyspnea on exertion and palpitation. Results: five fistulas originated from the right coronary artery, three from the left and two from the left circumflex. Drainage was to the right ventricle (7) right atrium (2) and pulmonary artery (1). The ratio of pulmonary to systemic flows ranged between 1 to 1.6. All patients had surgical ligation. In the symptomatic patient in addition to ligation, mitral valve replacement was performed. There was no operative or late death. Follow up evaluation (ranged from 1 year to 6 years, mean 4.2 years) showed no evidence of recurrent or residual CAF.

**Conclusion:** Surgical management of CAF is a safe and effective treatment resulting in 100% survival and clousure rate. Key words: Coronary artery fistula, Surgical ligation, Outcome

**Keywords:**Coronary artery fistula, Surgical ligation, Outcome

## Endocrinology and Metabolic Disorders

### Diabetes insipidus in children

*Mostafavi, F., MD, Children's Hospital Medical Center, Tehran University of Medical Sciences*

In diabetes insipidus, the amount of water ingested and the quantity and concentration of urine produced needs to be carefully regulated if fluid volume and osmolality are to be maintained within the normal range. One of the principal mechanisms controlling urine output is vasopressin which is released from the posterior pituitary gland and enhances water reabsorption from the renal collecting duct.

In diabetes insipidus, the excessive production of dilute urine, and the causes of this clinical picture can be divided into three main groups: the first is primary polydipsia where the amount of fluid ingested is inappropriately large; the second group is cranial diabetes insipidus where the production of vasopressin is abnormally low; and, the third group is nephrogenic diabetes insipidus where the kidney response to vasopressin is impaired. The history and examination may suggest an underlying explanation for diabetes insipidus but a range of baseline and more extensive investigations may be required before a diagnosis can be reached. These investigations are not without risk, and the results need to be interpreted carefully because children do not always segregate neatly into a particular diagnostic category on the basis of one test alone. Children with cranial diabetes insipidus typically respond to arginine vasopressin or its manufactured analogue, desmopressin, with an increase in urine osmolality and an associated reduction in urine output. Such children usually require neuroimaging to look for evidence of evolving CNS pathology, such as an intracranial tumour. Vasopressin 'replacement' with desmopressin is the treatment of choice in patients with cranial diabetes insipidus although extreme caution is required when treating babies or small children because of the danger of fluid overload. Abnormal production of other pituitary hormones in children with CNS disease can also influence fluid balance. Nephrogenic diabetes insipidus can be due to abnormal electrolyte concentrations; therefore these should be measured as part of the initial assessment. In a small number of children the defect is a primary abnormality of the vasopressin receptor or one of the water channel proteins (aquaporins) involved in water transport. The treatment of these patients is difficult and typically involves therapy with a diuretic such as chlorothiazide, as well as indomethacin. These agents enhance urine osmolality by their effect on circulating volume and renal solute and water handling. The fluid intake of most young children with primary polydipsia can be safely reduced to a more appropriate level.

The mechanisms that control water balance in health and disease have been extensively studied in adults. The establishment of reliable biochemical assays to measure circulating vasopressin, an increased knowledge of intracellular events within the renal tubular cells, and the application of molecular biological techniques to define hormonal and receptor abnormalities has meant that it is possible to accurately characterize disorders of water balance. The physiology and pathophysiology of water balance is similar in childhood although there are age-related changes in water distribution, diet, renal function, hormonal production and cognition that need to be considered when investigating and managing the young patient.

**Keywords:** Diabetes insipidus, Children, Vasopressin

### Ophthalmic finding in children with Diabetes Mellitus type1 in hamadan, west province of Iran

*Razavi, Z., MD, Bazzazi, N., MD; HamedanUniversity of Medical Sciences*

**Background:** the aim of this study was to determine frequency of retinopathy and other ophthalmic problem among patients with type 1 diabetes mellitus visited in Pediatric endocrine Clinic in Hamedan west province of Iran in 2008.

**Methods:** Diabetic patients visited in Pediatric diabetes Clinic were included in our study. Variable data such as age, sex, disease duration, stage of puberty, blood pressure, dose of insulin intake/kg /day and HbA1c of patients were obtained according to patients' history and physical examination. Eye examination by biomicroscope and indirect ophthalmoscope to evaluate for any signs of DR through dilated pupils was done by an ophthalmologist, Hb A1c level and screening for microalbuminuria were requested for patients. Data analysis were assessed by independent t. test and Chi-square test.

**Result:** In this study 125 patients were evaluated. 67 patients were female. Mean age of patients was 11. 69 ±3. 89 years and their average duration of disease was 3. 36± 1. 77 years. 8 cases (6. 4%) had eye involvement. 5 of patients had retinopathy and 3 of those had cataract. we also found 3 case of optic atrophy due to wolfram syndrome. 25% of patients with DR had heigh blood pressure (P=0. 02). frequency of DR was higher in patients taking less amount of insulin corrected to their body weight(p=0. 008). There was also significant correlation between DR and HbA1C level (P<0. 024).

**Conclusion:** Our results revealed that frequency of ophthalmic problem was significant, There was also

significant correlation between HbA1C level, height, blood pressure, low insulin consumption, good glycemic control and treatment in patients with diabetes, and regular screening is highly recommended for early detection and timely treatment of diabetic retinopathy in order to prevent progression to blindness. Abbreviations: DR, Diabetic Retinopathy.

**Keywords:** Cataract, Children, diabetes type 1, retinopathy, wolfram

### **Leptin concentration before and after insulin therapy in children with new onset insulin dependent diabetes mellitus**

*Zaridoost, A., MD, Mostafavi, F., MD, Setodeh, A., MD; Children's Hospital Medical Center, Tehran University of Medical Sciences*

**Background:** Serum leptin concentration reflects the body fat mass. It is still controversial reports about the insulin effect on serum leptin concentration. The aim of this study was to examine the effect of insulin therapy on serum leptin in children with new onset type I diabetes.

**Methods:** This was Cross-Sectional study has been done in Children's Medical Center with 34 children who had new onset type I diabetes. Serum leptin levels were measured at presentation, before insulin therapy was initiated baseline, three to five days after treatment and at third months of follow up. The linear regression by SPSSv. 16 used for analysis.

**Results:** there was a meaningful difference between the baseline level of leptin and as the 3-5 days ones. ( $p=0.000$ ). The dosage of insulin was the most important factor affects the leptin levels after 3-5 days of treatment ( $p=0.006$ ). In 3th months of follow up, sex and insulin dosage were the variables that had effect on leptin level. ( $p=0.003$ ).

**Conclusion:** The results of our study showed that children with new-onset type I diabetes have low leptin levels before insulin therapy which increased in 3-5 days after treatment. We concluded that the acute insulin therapy alters the leptin secretion/ action.

**Keywords:** Leptin; Insulin; Type I Diabetes; Children

### **The association between puberty stages and age in sample healthy children in Iran**

*MirOliai, M., MD, Pahloosye, A, MD; Shaheed Sadoughi Medical Sciences*

**Background:** The Greulich-Pyle bone age atlas is prepared in the US for white children who were born between 1917-1942 and is being used to evaluate children's bone age. This study is performed to determine bone age according to G. P. atlas and assess its correlation with different puberty stages.

**Materials:** In this cross-sectional study, 600 healthy children between 5-18 years of age that presented to Ali-Asghar hospital during 2007-2008 were evaluated. Their bone ages were calculated according to G. P atlas; children were examined for puberty stages by physicians. puberty stages, heights, weights and other demographic datae were collected.

**Results:** 53. 5% of participants were boys. Mean bone age and chronological age were 11. 2 and 11. 6 years respectively. In age groups of 5-8, 8-11, 11-14 and 14-18 years, boys' bone ages were 1. 2, 1. 3, 0. 1, 0. 2 years respectively behind that of G. P atlas. These findings were 0. 5 and 0. 1 years behind and 0. 5 and 0. 37 years ahead respectively. There was a significant correlation between bone age and chronological age with puberty stages in both sexes,  $p<0.05$ , Height growth was not associated with puberty stage in boys, in contrast to that in girls,  $p>0.05$ .

**Conclusion:** Bone growth was not associated with the G. P atlas indices in most age groups and were behind the G. P standards. in our study

**Key Words:** Puberty, Bone age, Chronological age, Height growth

### **The evaluation of relationship between consumption of various food and the prevalence of Asthma, Allergic rhinitis, and Eczema symptoms in children and teenage of Yazd**

*Karimi, M., MD, Mirzaei, M., MD; Shahid Sadoughi University Of Medical Sciences, Yazd*

**Background:** Allergy is the condition that promoted some people to diseases such as Allergic Rhinitis, Asthma and Eczema. The prevalence of Asthma and other allergic disease is various in the world, and there seems to be potential increase in worldwide prevalence of allergic disease during 3 past decades. Many studies show the probability of relation between environmental factors such as diet and the increase in prevalence of allergic disease, but they don't have the same results. This current study which is the part of ISAAC (International Study of Asthma & Allergy in childhood) study reveals the evaluation of relationship between consumption of daily food and frequency of intake with the prevalence of allergic symptoms in Yazd.

**Methods:** This cross sectional study involved 3000 children in grade one of primary school and 3000 teenagers of elementary school of Yazd city. The data was collected by ISAAC questionnaire. The 13-14 teenagers and the parents of 6-7 years children filled the questionnaire. We used Epi6 and Spss for analyzing the data.

**Results:** A total population of 5969 children was evaluated in 2 aged groups. According to the findings, low consumption of vegetables (11. 7%vs 8. 4%) in 6-7 years children had meaningful relation with wheezing.

In this group of children allergic rhinitis had meaningful relation with high consumption of butter (17.9% vs. 14.2%), Corn puff and Chips (18.6% vs. 14.6%), Chocolate (19.4% vs. 14.6%) and Traditional Candy (21.9% vs. 15%). In 13-14 years teenage high consumption of corn puff and Chips had meaningful relation with 3 diseases of wheezing (22.9% vs. 19.2%), Allergic Rhinitis (47.4% vs. 42.5%) and Eczema (19.1% vs. 13.6%).

**Conclusion:** The consumption of some food such as Fast food, Pizza, hamburger, Sausages and Salami, Chocolate, Egg, Corn puffs, Butter and Chips in children have an effective role on increasing the symptoms of Asthma, Allergic rhinitis and Eczema. These findings are the same as other studies in the country.

**Keywords:** food, Allergic symptoms, children

### Townes Brocks syndrome with congenital adrenal hyperplasia

*Bozorgmehr, B., Kariminejad, A., Afroozan, F., Hadavi, V., Kariminejad MH.*

Townes-Brocks syndrome (TBS) characterized by imperforated anus, dysplastic ears (overfolded superior helices and preauricular tags) frequently associated with sensorineural and/or conductive hearing impairment, and thumb malformations (triphalangeal thumbs, duplication of the thumb, preaxial polydactyly and rarely hypoplasia of the thumb). Renal impairment, including end-stage renal disease (ESRD), may occur with or without structural abnormalities (mild malrotation, ectopia, horseshoe kidney, renal hypoplasia, polycystic kidneys, vesicoureteral reflux). Congenital heart disease and genitourinary malformations are the other problems. Mental retardation occurs in approximately 10% of cases. It is autosomal dominant disease with variability in the severity of expression. We are reporting a 8 year girl with dysplastic ears, deafness, dysplastic thumbs, small kidneys, history of repaired imperforated anus, and rectovaginal fistula. She is also diagnosed with congenital adrenal hyperplasia with congenital hyperplasia. We believe our patient is the first case of Townes-Brocks syndrome, with congenital adrenal hyperplasia.

**Keywords:** Townes Brocks Syndrome, ambiguous Genitalia, Congenital Adrenal hyperplasia,

### Defibrillation and cardioversion in children

*Mohebbi, B., MD, Iran University of Medical Sciences; Tol, A., PhD Student, Isfahan University of Medical Sciences*

**Introduction-** Defibrillation (DF) and cardioversion are methods of delivering electrical energy to the heart

through the chest wall in an attempt to restore the heart's normal rhythm. Defibrillation and cardioversion may be accomplished using a manual defibrillator which requires users to recognize the dysrhythmia and preselect the energy to be delivered. Alternatively, automated external defibrillators (AEDs) may be used. AEDs are computerized machines that automatically diagnose ventricular fibrillation (VF) and use voice prompts to instruct rescuers to defibrillate, if appropriate. In addition, based on preset values for heart rate and R-wave morphology, AEDs may advise defibrillation for ventricular tachycardia (VT).

**Definitions:** There is an important distinction between defibrillation and cardioversion: Defibrillation-Defibrillation is the asynchronous delivery of energy, ie, the shock is delivered randomly during the cardiac cycle. Cardioversion -Cardioversion is the delivery of energy that is synchronized to the QRS complex. Mechanism of action Cardioversion- Cardioversion terminates arrhythmia by the delivery of a synchronized shock that depolarizes the tissue involved in a reentrant circuit. By depolarizing all excitable tissue of the circuit and making the tissue refractory, the circuit is no longer able to propagate or sustain reentry. As a result, cardioversion terminates those arrhythmias resulting from a single reentrant circuit, such as supraventricular tachycardia, atrial flutter, or monomorphic ventricular tachycardia. Defibrillation-Despite its wide use, there is still controversy concerning the electrophysiologic mechanisms by which electrical current terminates atrial or ventricular fibrillation, arrhythmias which involve multiple microreentrant circuits. Fibrillation involves the entire atrial or ventricular myocardium and is considered to be a very recalcitrant unstable rhythm.

**Indications:** The application of electrical current is an essential component of advanced cardiopulmonary resuscitation (CPR) guidelines for the treatment of ventricular fibrillation (VF), ventricular tachycardia (VT), and unstable and/or drug resistant organized cardiac rhythms such as supraventricular tachycardia. Defibrillation is indicated in the treatment of the following: ventricular fibrillation and pulseless ventricular tachycardia. Cardioversion should be used in unstable patients with organized cardiac rhythms such as supraventricular tachycardia (SVT), atrial fibrillation, atrial flutter, or ventricular tachycardia with a palpable pulse.

**Keywords:** Defibrillation, Cardioversion, CPR

### Hypercalciuria in children with vesicoureteral reflux

*Nafisi-moghadam R, Shahid Sadughi Yazd Medical University, Yazd*

**Background:** Renal malformations including vesicoureteral reflux (VUR) are associated with urolithiasis. However, studies on urinary calcium excretion in

children with VUR have not been reported. This study was conducted to find out whether children with VUR have a higher prevalence of hypercalciuria and whether their family members are affected by hypercalciuria and/or urolithiasis. Methods: We studied the prevalence of hypercalciuria and urolithiasis in 46 children (12 males and 34 females) with VUR and in their parents. Results: Three out of 46 children had renal colic and nine out of 46 exhibited calyceal microlithiasis in the renal sonography. According to Stapleton's criteria, we found that 27 out of 46 children (58.6%) had hypercalciuria. These children were significantly shorter than children with normal calciuria and showed lower values of maximal urinary osmolality. We found no differences in urinary calcium excretion values related to the VUR grading, or to the presence or absence of renal scars, or to whether VUR was still unresolved or already resolved at the time of study. Seventeen out of 27 children with hypercalciuria (63%) had one or both parents affected by hypercalciuria, and there was a history of urolithiasis in six first-degree relatives and in four second-degree relatives (37%). Besides, 10 out of 19 children without hypercalciuria (52.6%) had one or both parents affected by hypercalciuria and there was a history of urolithiasis in three first-degree relatives and in three second-degree relatives (31.6%). Among the 27 children whose parents had hypercalciuria, four had both parents affected, 19 had only the mother affected and in four patients only the father was affected.

**Conclusion:** Our results showed that the prevalence of hypercalciuria was greater in paediatric patients with VUR than in the general population. Urolithiasis in patients with VUR had a metabolic origin. Hypercalciuria was inherited as an autosomal dominant trait although with a higher probability to be inherited from the mother.

**Keywords:** hypercalciuria; inheritance; vesicoureteral reflux

### **Mucopolysaccharidosis, report of more than fifteen years experience in Iran**

*Shafeghati, Y., MD; Karimi-Nejad. MH., MD; Almadani, N., MD; Afroozan, F., MD, Hadipour, F., Hadipour, Z.*

Because of the high rate of consanguineous marriages, metabolic disorders, both micromolecular and macromolecular types such as Mucopolysaccharidoses are very prevalent in our population, and now they constitute a very important health problem in this community. They are "Enzymopathies" and the result of homozygous mutations in many different genes which are responsible to encode specific enzymes inside the cells. Each enzyme should catalyze one of the metabolic reactions that is necessary to degrade intra-lysosomal glycosaminoglycans. Result of the mutation in a coding gene may cause severe enzyme deficiency, intra-lysosomal accumulation of

glycosaminoglycan macromolecules, and a specific phenotype. Typical sequence of symptoms may include progressive neurodevelopmental delay or regression, dysmorphic and coarse facies, corneal clouding, hearing loss, visceromegaly, skeletal dysplasia, and dysostosis multiplex congenita. We have studied 231 families with 300 cases in the past 20 years, that were referred for evaluation and diagnosis of a dysmorphic, neurodevelopmental regressive disorders with the MPS phenotype. The diagnosis in the index cases (after a detailed clinical, biochemical, radiological, imaging studies) confirmed by enzyme assay or molecular analysis. Prenatal diagnostic tests also carried out in 89 pregnancies and 27 affected fetuses were detected (about 33% of the pregnancies). I will present my experience and practical points regarding epidemiology, symptomatology of these very important and rare diseases. Also, I will offer a very simple diagnostic approach for diagnosis, treatment, follow up, and prevention by PND in the affected families.

**Keywords:** LSDs, MPSs, Enzymopathies, Iran experience

### **One year treatment with two brands of recombinant human growth hormone (rhGH) in children with growth hormone deficiency**

*Tabatabaei-Malazy, O., Mohajeri-Tehrani, MR., Heshmat, R., Taheri, E., Rabbani, A., Razzaghy-Azar, M., Adibi, H, Larijani. B.; Endocrinology & Metabolism Research Center, Tehran University of Medical Sciences; Growth & Development Center, Tehran University of Medical Sciences; Children's Medical Center, Tehran University of Medical Sciences; Endocrine research Center, Iran University of Medical Sciences, Tehran*

**Background:** The principle use of recombinant human growth hormone (rhGH) is in children with documented growth hormone deficiency (GHD) for induction of normal growth velocity (GV). In this randomized clinical trial (RCT) study, our main objective was to compare the efficacy and side effects of standard dose of rhGH (0.3 mg/kg week, Iranian brand, Samen Company) to FDA approved rhGH (NOVO Nordisk Company) in children with GHD. Methods: 22 pre-pubertal children with documented GHD were treated either with Samen or NOVO rhGH for 12 months. Upstanding height, height standard deviation score (HSDS), GV, serum level of Insulin like growth factor-1 (IGF-1) were measured before and 6 months after starting treatment. The bone age (BA) of them also was evaluated before and post 12 months. During the study, side effects of rhGH therapy were assessed. Findings: We evaluated 22 subjects, 12 in Samen and 10 in NOVO groups. In each group, 50% of the participants were male. The mean age in both groups was similar (12 yr). The mean difference of

height, HSDS, IGF-1 and BA in NOVO group before and 6 months after study were 8.76 cm, 0.52, 190 ng/ml and 2.85 yr, respectively, while these results in Samen group were 9.13 cm, 0.57, 222 ng/ml, 1.7 yr, respectively. Although, these differences were statistically significant within each group, there weren't statistically significant difference between groups. The mean  $\pm$  SD of GV in Samen and NOVO groups were 9.4  $\pm$  1.75 and 8.8  $\pm$  1.15 cm, respectively, which wasn't significantly different between groups. The serum level of IGF-1 was similar within groups. We didn't find any side effects such as edema, headache, and pseudotumor cerebri within groups.

**Conclusion:** Considering the above results, it seems the efficacy of Iranian brand of rhGH (Samen) is similar to NOVO brand after 12 months treatment in children with GHD. We suggest a post marketing surveillance with a large sample size in order to confirm our findings.

**Keywords:** growth hormone deficiency, recombinant human growth hormone, insulin like growth factor-1.

### **Incidence of tetrahydrobiopterin deficiency in neonatal hyperphenylalaninemia in Fars province, south Iran**

*Ordooei, M., MD, Shaheed Sadoughi University of Medical Science; Karamifar, H., MD; Karamizadeh, Z., MD; Amirhakimi, GH., MD; Shiraz University of Medical Science*

**Background:** BH4 is the cofactor for phenylalanine hydroxylases. infant with BH4 deficiency are diagnosed as having pku, but they deteriorate neurologically despite adequate control of plasma phenylalanine. Oral administration of BH4 to patients with milder forms of hyperphenylalaninemia due to phenylalanine hydroxylase deficiency (no BH4 deficiency) may reduce plasma level of phenylalanine without the need to remain on a low phenylalanine diet. The objective of this study was to determine the incidence of BH4 deficiency and responsive to BH4 in patient hyper phenylalaninemia in Fars province, south of Iran. Method: In a period of one year from November 2007 to November 2008 blood samples were with drawn from all newborns born in Fars province for measurement of serum phenylalanine. The samples with a serum level of  $\geq$  2 mg/dl confirmed by using high-pressure liquid chromatography (HPLC) method. For BH4 deficiency was performed by the BH4 loading test and phenylalanine level check at 0, 4, 8, 24 hours after BH4. in order to determine which of the affected infant had BH4 deficiency and would respond to BH4 therapy.

**Results:** Nine out of 76966 new borns had a serum phenylalanine level  $\geq$  2 mg/dl of which 8 cases were confirmed by HPLC. after BH4 loading test, one patient had serum phenylalanine level below 2 mg/dl with falls into the category of BH4 deficiency and one

patient had a 32% decrease in serum phenylalanine level after 8h of BH4 administration who was considered BH4 responsive. after patient not show any significant change in serum phenylalanine level after BH4 loading test. **Conclusion:** In this study for BH4 loading test serum phenylalanine checked before BH4 and then 4, 8, 24 hours there after. responds to BH4 after 8 hours no much different with 24 hours.

**Keywords:**Hyperphenylalaninemia ; BH4 ; New born ; PKU

### **Comparison of hashimoto thyroiditis in diabetic patients (type1) with control group in Qods pediatric hospital in 1388**

*Safari, F., Asgari, A., Sadeghi, T., Ghazvin University of Medical Sciences*

**Background:** Type 1 diabetes is the most common metabolic disease worldwide. thyroid autoimmune diseases accompany with type 1 diabetes is the most common Endocrinopathy. Therefore, thyroid function tests and anti thyroid antibodies in patients suffering from Type 1 diabetes is essential to prevent the first level of thyroid autoimmune disorders. The purpose of this study is to compare the anti-thyroid antibodies in patients with Type 1 diabetes and healthy individuals together. Materials and Methods: In this descriptive-analytic study, 65 children with Type 1 diabetes and 65 healthy children who had referred to Children's Hospital in Qazvin, using simple sampling were selected and for them, the amount of Anti-TG and Anti-Tpo and TSH and T4 hormones were measured. The amount of antibodies in tow group compared together using Chi-square statistical analysis, t-independent, Kruskal Valis. findings: There was not significant difference between two group of samples( case and witness group) in case of sex and age but BMI percentile of two group was significantly different. Positive Anti-TG in patients gorup was 10.8% and for control group it was 1.5% which this difference is significant statistically (p=0.029). 16.9% of patients group and 3.1% of control group had positive Anti-Tpo that was significantly different between tow group(p=0.024). In patients gorup, percent of TSH had been increased. In the control group 4/6 percent of TSH had been increased that its difference was not significant statistically.

**Conclusion:** according to the results of research, it seems that the prevalence of Hashimoto thyroiditis in patients with type 1 diabetes is more than healthy people. Hence, thyroid function tests (TFT) and antithyroid antibodies (Anti-Tpo) in patients with Type 1 diabetes is necessary in order to early diagnosis of autoimmune thyroid disorders, prevention of complications of thyroid disorders and timely treatment.

**Keywords:** Type 1 diabetes, Hashimoto thyroiditis, anti thyroid antibodies, Anti-TG

### **The optimal dose of vitamin D in growing girls during academic years a randomized trial**

*Shakiba, M., Shahid Sadoghi Medical University, Yazd*

**Background:** Prevalence of Vitamin D deficiency is remarkable during childhood and adolescence throughout the world. Sufficient intake of vitamin D is contributed to numbers of health outcomes. The aim of this study is specifying the optimal dose of vitamin D in growing girls in a Muslim country during an academic year. Methods: this randomized clinical trial study is carried out in Yazd in center of Iran in 2007, 120 junior high school girls(aged12-15years) were randomly divided into 4 groups. 60 students in group I and II were treated as vitamin D deficiency with 300, 000IU vitamin D3 and then randomly received 50, 000U/monthly or 100, 000IU/three months vitamin D3;60 other students in group III and IV received 50, 000IU/three months and 100000/three months from the beginning of the academic year. Medication continued for the entire academic year, One month after the last dose, Serum 25(OH)D levels were measured. Results: The mean level of 25(OH) D was 29. 7±4. 60 ng/ml in group I and 30±5. 61ng/ml in group II. Mean serum levels of 25(OH) D were 15. 2±6 ng/ml and 23±6. 8 ng/ml for group III and IV respectively. Conclusions: Neither doses of about 800IU/day nor 1000IU/day are sufficient to maintain 25(OH)D in optimal level( $\geq 20$  ng/mL)for all, but after the treatment of deficiency, intakes of about 1000 IU/day or 2000IU/day of vitamin D maintains optimal level in all of the students.

**Key word:** vitamin D, girl, bolus therapy, 25OH vitamin D

### **Vitamin D requirement in pregnancy to maintain sufficient vitamin in newborn a randomized trial**

*Nafei, Z. , MD, Shakiba, M. , Shahid Sadoghi Medical University, Yazd*

Vitamin D requirement concept was changed in recent years, pregnancy and intra uterine life is a crucial period of life for development and we donot know exact amount of vitamin requirement in that. this study is a randomized trial to find doses of vitamin D in pregnancy to produce sufficient level in newborn. In 2009 at Yazd in center of iran on 52 healthy pregnant women this study was performed, they randomly recieved 50, 000 or 100, 000 IU vitamin D3/month from second trimester and 17 others treat first as deficiency with 200, 000 IU vitamin D3 then 50, 000IU/month. 25-OH vitamin D in cord blood was check by chemiluminescence immunoassay and level more than 20ng/ml accept as sufficient. Result:all

mothers had vitamin level below 30 ng/ml at the end of first timester, 24 % of monthers on 50, 000/month(about2000IU/day ) could not acheived sufficeint level in newborns. all in other two group had level more than 20ng. ml in their babies. we couldnot find any side effect during consumption. Coclusion: in area with high prevalence of vitamin D deficiency in mothers about 4000IU /day vitamin D3 from second trimester of pregnancy is needed to make level more than 20ng/ml of 25-OH vitD in newborn, consuming less than 2000IU /month is not sufficient in all.

**Key words:** vitamin D, pregnancy, 25 hydroxy vitamin D, vitamin supplementation

### **The Comparison of the height mean in high school boy students (18 years old) in Ahwaz city with NCHS chart in 2008**

*Vaziri Esfarjani, Sh., Jafariyan Shooshtari, A., Jondi-Shapour University of Medical Sciences, Ahvaz*

Growth is one of the important aims during childhood. Thus, growth monitoring is necessary for routine medical health services throughout the world. Since some of the most effective factors on growth, are envoiroment and human race, therefore growth charts for each area needs to be determine. The aim of this study is to define the growth charts in one of the regions in middle east, Iran, in 2008. In this study, height mean-as one of the growth indexes-in high school boy students (18 years old) in Ahvaz city was determined and compared with NCHS chart. This cross-sectional study was conducted on 1292 healthy students (male, 18 years old), living in Ahvaz. The sampling approach was multi stage and coincidenty. The data was determined by measuring the teenagers height in this study. Statistical analyzes was done by t test. The results showed that the mean of height was significantly less than the NCHS in this study ( $P<0.05$ ). According to these results shortening of the height in Iranian community may can consider as a normal fact. However, more studies should be done in future to assess effective height factors in Iranian pupollation in this regard.

**Keywords:**Height, NCHS'S chart, boys of high school

### **BMI, Waist Circumference and the Prediction of Metabolic Syndrome in Highschool Students**

*Mehrkash, M. , MD; Mohammadian S, MD; Gorgan University of Medical Science, Gorgan, Iran.*

*Qorbani M; PhD Student of Epidemiology, Tehran University of Medical Science; Shafa, N., MD;*

**Background:** The metabolic syndrome is a cluster of metabolic disorders that are associated with elevated

risk of cardiovascular diseases and diabetes type 2. This study investigated the prevalence of metabolic syndrome and its components among adolescents population in Gorgan - Iran.

**Methods:** A cross-sectional survey was conducted involving 450 adolescents 15 -18 years, selected through stratified random sampling from high schools in Gorgan (north of Iran). Anthropometric measurements, blood pressure, FBS and lipid profile were assessed. Metabolic syndrome was defined according to NHANES III criteria. Collected data analyzed by SPSS software.

**Results:** The Mean±SD age of the subjects was 16±0.8 years. Between two groups (girls and boys), age difference was not significant. Prevalence of metabolic syndrome rate was %3.2 (15 cases). Prevalence in boys and girls was %4.9 (11 cases) and %1.8 (4 cases), respectively. The prevalence of obesity was %19.4 (BMI ≥ 85 percentile), the ratio was %19.6 in boys and %18.7 in girls. The relationship between obesity and metabolic syndrome is significant (OR: 6/97), (CI: 20/16- 2/41). It means that the obesity increase odds of the metabolic syndrome incidence about 7 times. The relation between the waist circumference and metabolic syndrome is significant (OR: 21/64), (CI: 6/7- 69/8).

**Conclusion:** This study shows that obesity and overweight (BMI ≥ 85 percentile) increased metabolic syndrome incidence about 7 times, and abdominal obesity increased odds of metabolic syndrome incidence 21 times. Therefore, it is recommended to further study about the percentile cutoff point of obesity risk and further attention to the waist circumference and the abdominal obesity in new planning is necessary to prevent the weight gain.

**Keywords:** Metabolic Syndrome, BMI, Waist Circumference, Obesity

### Children and adolescents obesity

*Soheilipoor, F., MD; Iran University of Medical Sciences*

Children and adolescents obesity Children and adolescents obesity (BMI > 95%) is a big problem and chronic involvement in today's society. It can be screened in the best way and any intervention in the first years of this chronic involvement can be as an effective step in health promotion programs of this critical age group. If we as family physicians, nurses or any other member of the health system do not do something about it today, our future practices will be filled with a majority of obese adults with more time needing to be spent in disease management. The most important common reason of obesity is environmental in nature. Genetic and Endocrine causes also play a role, but this is rare. Risk factors are: race, parental obesity, high birth weight and low birth weight. There are some important co-morbidities with obesity that include: Diabetes, Hypertension, Dyslipidemia (three of the biggest risk factors for cardiovascular disease),

Sleep apnea, Gall bladder disease and Joint pain. In evaluation of overweight children and adolescents attention to dietary history, activity history, review of systems, Family history, psychosocial history and careful physical exam is necessary. They will determine laboratory testing and imaging that is required. But what do we do as an effective member of the health system? What is the answer of this important health problem? The role of medications is limited and therefore, Education is the most important step. Simply spending time with the family may be the most important thing we do. Education also includes the adults in the family, school teachers and society managers. Remember, it is usually the parents who provide the meals. Small steps are always the key and can be done in the best way.

**Keywords:** Obesity, Children, Adolescents

### Evaluation of TSH cut off In Iranian neonatal screening of hypothyroidism

*Hashemipour, SV., MD, Rabbani, A., MD, Sotudeh, A., MD, Paravaneh, N., MD; Growth and development Research Center, Department of Pediatrics, Tehran University of Medical Science*

Thyroid hormone is important for normal development of the central nervous system from fetal life until at least the first 2 yr after birth. Children with congenital hypothyroidism (CH), if left untreated, are at risk for impaired cognitive and motor development. The newborn screening program was developed to prevent mental retardation by early detection of congenital hypothyroidism (CH). Current methods of screening are the primary thyrotropin (thyroid stimulating hormone- TSH) screening alone and primary thyroxine (T4) with secondary TSH screening (T4/TSH). In Iran, TSH screening is employed almost universally to detect CH. The TSH concentration of greater than 5 mIU/ml was chosen in Iran as the cut-off value for diagnosis of CH in the screening program. Our experience shows a high rate of false positive results of neonatal hypothyroidism screening program in Iran. In this study, we followed 50 neonates with primary TSH greater than 5 mIU/ml (12 cases, 5-10 mIU/ml, 12 cases, 10-15 mIU/ml, 10 cases, 15-20 mIU/ml and 16 cases greater than 20 mIU/ml) for at least 13 months (13-38 months), to determine if they are really hypothyroid. During follow-up 12 infants were hypothyroid; 11 cases of them had primary TSH greater than 20 mIU/ml and one case had primary TSH equal to 19.90 mIU/ml. None of 12 cases who had primary TSH between 5 to 10 mIU/ml were hypothyroid. (ppv=0%) None of 12 cases who had primary TSH between 10 to 15 mIU/ml were hypothyroid. (ppv=0%) one of 10 cases who had primary TSH between 15 to 20 mIU/ml were hypothyroid. (ppv=10%) 11 of 16 cases who had primary TSH greater than 20 mIU/ml were hypothyroid. (ppv=69%) 12 of 50 cases who had primary TSH greater than 5 mIU/ml were

hypothyroid. (ppv=24%) 12 of 38 cases who had primary TSH grater than 10 miu/ml were hypothyroid. (ppv=32%) 12 of 26 cases who had primary TSH grater than 15 miu/ml were hypothyroid. (ppv=46%) Based on our findings, we conclude that, cut off in iran is low and choose a greater cut off is necessary.

**Keywords:** neonatal hypothyroidism, screening, TSH cutoff

### Neonatal diabetes

*Abbasi, F., MD, Amoli, MM., Asayahpoor, FA., Sajjadian, N., MD., Taii, N., MD; Metabolic and Endocrine Research Center*

Patients with neonatal diabetes usually characterized by hyperglycaemia within the first SIX months of age. and requiring insulin treatment For control of symptoms. it can either be transient (TNDM) or permanent (PNDM). In majority of cases, the cause is unknown. Heterozygous activating mutations in KCNJ11, the gene encoding the Kir6. 2 subunit of the ATP-sensitive potassium channel causes 30 to 58 percent of cases of neonatal diabetes. Method: We sequenced the KCNJ11 gene in a patient with permanent neonatal diabetes. his neonatal diabetes was characterized by marked hyperglycemia and was treated with insulin. the patient never had any developmental delay, muscle weakness; epilepsy or dysmorphic features. he was mutation carrier therefore was invited for switching from insulin to sulfonylureal. result: he was successfully transferred to SU, dramatically improved his blood glucose and HBA1C in next follow ups. Conclusion: This case illuminates that, the molecular genetic testing of patients with permanent neonatal diabetes may lead to change of the treatment of this disease with oral hypoglycemic agents like sulfonylureas.

**Key words:** Diabetes, Neonatal, Mutation

### Newborn screening by tandem mass is one of the nation's most successful public health programs

*Farshidi, S., MPH, MD, Gafari, SR., Farokhi, B., Brogerdihope MA., Gene Center*

Newborn screening programs test babies for disorders that are often not apparent at birth. Such disorders may be inherited, infectious, or caused by a medical problem of the mother. If these disorders are not detected and treated soon after birth, they may cause mental retardation, severe illness, or premature death. More than 4million newborns are screened annually in the United States, and thousands of infants are rescued from disability and death. Newborn screening begins within 24 to 48 hours of a child's birth when a few drops of blood are obtained from a heel stick. The

blood spots are sent to a laboratory that is a part of the state or territorial public health department. The spots are analyzed by several different laboratory methods to test for biochemical and genetic markers that reveal hidden congenital (present at birth) disorders. If such markers are found, the newborn screening follow-up program notifies the parents and physicians so that the baby can receive immediate attention. Follow-up programs arrange for diagnostic tests to confirm the newborn screening results. Follow-up programs also refer the child to a treatment center to provide access to the essential medical services needed to minimize the effects of the underlying disorder. Common considerations in determining whether to screen for disorders: 1. A disease that can be missed clinically at birth 2. A high enough frequency in the population 3. A delay in diagnosis will induce irreversible damages to the baby 4. A simple and reasonably reliable test exists 5. treatment or intervention that makes a Result of Expanded NBS by MS/MS in difference if the disease is detected early Iran 5000 neonates screened for 23 IEM during 2007-2009 – 11 newborn with confirmed metabolic disorders. • 8 required treatment – Overall prevalence of metabolic disorders was 2. 2/1000 – 16 false positive (0. 32%) – Overall specificity (98. 68%) and sensitivity (100%) – 7/11 were judged to have benefited from screening & treatment.

**Keywords:** Newborn screening, Tandem mass

### 24 Hydroxylase Polymorphism as a Possible Explanation for the Higher Level of 1, 25 (OH)<sub>2</sub> Vitamin D in African American Ethnicity

*Salehpour Sh., Assistant professor of pediatric endocrinology, Shahid Beheshti University of Medical Sciences; Ardeshirpour, L. , Yale University, Wong BYL., Molecular Genetics, Toronto University; Cole DEC., Clinical Biochemistry and Genetics, Toronto University; Carpenter TO., Yale University*

**Context:** States of vitamin D insufficiency are important determinants of rickets, as well as osteoporosis and other common complex disorders like diabetes, cancer and infectious diseases. Although serum concentrations of the vitamin D metabolites are primarily driven by vitamin D supply (by diet or cutaneous synthesis), there is emerging evidence to suggest that single nucleotide variants (SNVs) are important genetic determinants. **Objective:** The aim of this study was to determine whether a functional SNV in the 24-hydroxylase gene promoter (c. -686A>G in CYP24A1) shows significant association with blood levels of vitamin D metabolites Methods: Genomic DNA from 776 inner-city New Haven children aged 6 mon to 3 yr with different ancestries (African American, Caucasian, and Hispanic) was genotyped for the c. -686A>G SNV. Serum 25-hydroxy vitamin D and 1, 25-dihydroxy vitamin D [1, 25(OH)<sub>2</sub>D] were

measured by RIA. Ancestry was assessed using a validated panel of 108 Ancestry Informative Markers (AIMs) and data from well-characterized African, Native American, and European population samples. Main outcome measures: The main outcome measures were significance of associations between the c. -686A>G CYP24A1 SNV and vitamin D metabolites, with modeling to adjust for age, season, vitamin D intake and other co-variables. Secondly we examined the strength of these associations in relation to SNV frequency in the three major ancestral groups. Results: Subjects with the variant allele of CYP24A1 (and decreased 24-hydroxylase activity) had a significantly higher mean 1, 25(OH)2D ( $p < 0.001$ ), but all variants were found in African-Americans who, as a group, had higher mean 1, 25(OH)2D ( $p < 0.0001$ ). Since the effect was not significant when the association was AIMs-adjusted for ancestry, we cannot exclude confounding by stratification. Conclusion: Further studies of the CYP24A1 SNV are warranted, but the 24-hydroxylase polymorphism may be considered as one possible contributor to the increased 1, 25(OH)2D that is widely observed in African Americans.

**Keywords:** 24 Hydroxylase gene polymorphism, 1, 25(OH)2 Vitamin D, African American Ethnicity

### Evaluation of serum liver enzymes levels in patients with turner syndrome

*Rohani, F., MD; Karimi, M., MD; Endocrine Research Center*

**Background:** Turner's syndrome is the most common sex-chromosomal abnormality in female which may be associated with various disease conditions: both normal and elevated levels of liver enzymes has been reported.

**Objectives:** The aim of this study was to assess serum levels of liver enzymes in patients with turner syndrome.

**Methods:** In this descriptive-Analytic study, we measured liver enzymes levels; aspartate (AST), alanin amino transferase (ALT) and alkaline phosphatase (Alk. ph) in 27 patients with turner syndrome in Institute of Endocrinology and Metabolism of Iran.

**Results:** Mean age of patients was 14.34±8.6 years. Mean age of diagnosis was 9.23±6.3 years. Most of patients (70.4%) had 45XO karyotype. Short stature and edema and webbed neck were the most prevalent symptoms (44.4% and 26%, respectively). Associated disease were: Heart (48.14%), kidney (14.82%), Thyroid diseases (11.11%) and diabetes mellitus (3.7%). Serum levels of liver enzymes in all patients were normal. There wasn't significant difference in level of liver enzymes in estrogen receiving patients (n=9) and other patients.

**Conclusion:** Despite of recent reports about liver enzymes abnormalities in turner's syndrome, serum

levels of liver enzymes in all patients with turner syndrome in our study were normal.

**Keywords:** Turner's syndrome, Aspartate transferase (AST) Alanin amino transferase (ALT) Alkaline phosphatase

### Autoimmune thyroid disease in children and adolescents with type 1 diabetes mellitus: a survey in the northwest of Iran

*Shiva, S., MD, Ghalehgalab Behbahan, A., MD, Tabriz University of Medical Sciences*

**Background:** The prevalence of autoimmune thyroid disease in diabetic children considerably varies in different countries depending on the age, sex, and ethnic origin of the subjects. The situation in Iranian population especially children is still a much debated subject. This study has been designed to investigate this problem in the Northwest of Iran.

**Methods:** From February 2006 to November 2007, serum levels of anti-thyroid peroxidase and anti-thyroglobulin antibodies and thyrotropin hormone were measured with ELISA method in 176 diabetic children (M/F: 78/98) at a mean age of 8.3±3.7 and mean diabetes duration of 1.6±2.5 years, who were referred to pediatric-endocrinology clinic of Tabriz University of Medical Sciences (Tabriz-IRAN).

**Results:** Autoimmune thyroid disease was found in 12% of patients (8.6% female and 3.4% male). Significant levels were found for anti-thyroid peroxidase (10.2%), anti-thyroglobulin (8%), and both antibodies (6.3%) in all patients.

**Conclusion:** We concluded that autoimmune thyroid disease in Iranian children and adolescents with type 1 diabetes has a medium prevalence rate compared with those of other countries. The disease is more common in female and older diabetic patients.

**Keywords:** Anti-thyroglobulin; Anti-thyroid peroxidase; Autoimmune thyroid disease

### Serum Zinc level in children with retarded growth and normal control children in Ahvaz, Iran

*Monajemzadeh, SM., Riahi, K., Rahimi, M., Assar, S., Sharifan, M.*

**Background:** Zinc is an essential micronutrient for human metabolism. It catalyzes more than 100 enzymes. Also, it contributes to protein structure, and regulates gene expression. Therefore, it has an important role for growth and also for development and highly relevant functions such as immunity, tissue repair, protection against oxidative damage, neuropsychological function and hormone action. Data from many studies of infant childhood growth have

shown strong associations. between zinc deficiency and growth retardation. This study was done to evaluate plasma zinc level in children with growth retardation and healthy control group aged 2 to 15 years, referred to Endocrine clinic of Ahvaz Golestan Hospital in 2009-2010.

**Method:** In a cross-sectional study, 76 children with growth retardation (case) and 76 well developed children (control) were evaluated. Both case and control group were the same according to age, sex and geographical region. 3ml venous sample was taken from each child and plasma zinc level was measured by atomic absorption spectrophotometry.

**Results:** Mean plasma zinc level in children with growth retardation was 78. 89 +- 19. 36 and in control group was 85. 27+- 18. 46 µg/dl. There was a significant statistical difference between the two groups. (p value=0.03).

**Conclusions:** According to the results, mean serum zinc level in children with growth retardation was lower than well-developed children. So we found a relationship between growth retardation and zinc deficiency in this population. Improving zinc intake should be a component of interventions to promote growth in children living in settings where zinc intake is inadequate.

**Keywords:** Zinc, Growth retardation

### Major inborn errors of metabolism presenting in the first 12 years of life in 79100 consecutive births in Ghazvin Province

*Movafagh, A., Department of Medical Genetics, Shahid Beheshti University of Medical Sciences; Saffari F., temeh' Mohamadzadeh, Ghesmat-Assistant PMD, Metabolic Disease Research Center, Qazvin University of Medical Science.*

**Background:** The clinical features of an inborn errors of metabolism result from deficient enzyme activity. Expression of metabolic diseases are congenital and inherited in an autosomal recessive pattern. This article aimed to report the occurrence of metabolic diseases in geographic of Ghazvin province.

**Material:** A Cross-sectional study was perform between 2000- 2010 on 79100 children from the age infancy to age 12 years. Clinical manifestation, update laboratory findings and all essential information have been approached to confirm precise diagnosis of metabolic diseases. The sorted inborn errors of metabolic patients, checklist and data were analyzed by SPSS (v. 16 inc. Chicago IL).

**Results:** A total of 531 metabolic disorders were recorded and the prevalence of metabolic disorders was higher in male (372 cases) than female (159 cases). The more frequent malformation associated inborn errors of metabolic among our patients were noticed for G6PD

(4. 0%) and the lowest rate recorded for Galactosemia (0. 37%).

**Conclusion:** Detection and management of total inborn errors of metabolic can be life save and spanning for children. However, screening program, prenatal and molecular gene therapy, consanguineous marriage counseling could be considered as the important factors for the inborn errors of metabolic prevention.

**Key words:** inborn error of metabolism prevalence, population Ghazvin

### Cockayne Syndrome, Reporting a cases from Iran Confirmed by DNA-repair and direct sequencing analysis

*Hadipour, F., Hadipour, Z., Noruzinia, M., Behjati, F., Shafeghati Y., 1. Sarem Cell Research Center; Medical Genetics Department; 2. Genetics Research Center, University of Welfare Science and Rehabilitation, 3. Department of Medical Genetics, School of Medical Sciences, Tarbiat Modares University*

Cockayne syndrome is a very rare genetic disorder with a recessive autosomal inheritance characterized by dwarfism, microcephaly, mental retardation, deafness, photosensitive dermatitis and a peculiar form of retinal pigmentation. We report here an Iranian family with oneaffected child who is suffering from Cockayne syndrome. Cardinal features were: failure to thrive, short stature, premature aging, microcephaly, dysarthric speech, photosensitivity, sunken eyes, and dental caries. There was no blindness or deafness, and the fundus examination showed tapetoretinal degeneration. Direct sequencing of all coding sequences of CSA and CSB genes, showed a novel mutation (c. 2382+57G>T in intron 10 of CSB) that was not reported before. This variation might perturb splicing in CSB. However to prove the pathogenicity of this mutation mRNA analysis on fibroblast is planned to be investigated.

**Keywords:** Cockayne syndrome, premature aging, photosensitivity, microcephaly, CSA and CSB genes

### The Treatment of ambiguous genitalia and presentation of 84 cases

*Delshad, S., MD; Razzaki Azar, M., MD, Iran University of Medical Sciences, Ali Asghar children Hospital*

**Background:** Many factors such as chromosomes, gonads, hormones and enzymes influence the development and differentiation of embryo's genitalia. A defect in any one of these factors can result in child genital ambiguity. The ambiguous genitalia is a contradiction between the patient's gonads and external genitalia and it is difficult to identify the neonate's true gender according to his or her external genitalia.

**Material:** The patients records reviewed retrospectively and they were evaluated periodically. In the recent 15 years, 84 patients with ambiguous genitalia had admitted to ali asghar children and tehran children hospitals. There were 65 female pseudohermaphrodits due to CAH with (46 xx) karyotype, 13 male pseudohermaphrodits with (46xy) karyotype, 4 cases with mixed gonadals dysgenesis and two patients with true hermaphroditism. The majority of these cases diagnosed and managed earlier at infancy. After preliminary diagnosis in order to determine the gonads sonography performed.

**Results:** The most common complication which is seen of vaginoplasty, is vaginal orifice stenosis. in 21 cases with one stage group (36 cases) versus in 4 patients from 2 stages proup (21 cases) which underwent revision was performed. In 6 patientes of first group while the after treated by dilatations. The difference between two method was statistically significant (P-

value: 0. 047). in intestinal vaginoplasty prosedures one vaginal orifice stenosis, was observed which treated by dilatations. The most common complication for hypospadias repair in male pseudohermaphroditism group were urethral fistula that occurred in one of the cases.

**Conclusion:** In ambigious genitalia external genitalia has to correct in the first months of birth for decreasing parents anexity. In female pseudohermaphroditism, 2 stage operations have little complications such as vaginal stricture. In female pseudohermaphroditism diagnosed in the first months of birth, the choice management is making her as a female. In male pseudohermaphroditism testicular feminization type, the choice management is gonadectomy, vaginoplasty (with an intestinal segment) and changing the sex to female.

**Keywords:** Ambiguous genitalia, Pseudohermaphrodits, Congenital adrenal hyperplasia

## Gastroenterology & Nutrition

### Comparison of effectiveness between Beclomethason Dipropionate and evaluation of therapeutic effect of vitamin E on persistent diarrhea in less than 5 year-old children in Urmia

Gheibi, Sh., MD, Fakoor, Z., MD, Karamyyar, M., MD, Aabedini, AA., MD, Urmia University of Medical Sciences

**Background:** Persistent diarrhea usually commences acutely but lasts for more than two weeks. It consists 3-20% of all diarrhea cases in children younger than 5 years old but responsible for more than 50% of diarrhea related deaths. Antioxidant agents such as Zinc reduce the diarrhea-related complications especially in developing countries, so this study aimed to investigate the effect of vitamin E on the improvement of Persistent diarrhea.

**Material:** In this double-blinded clinical trial children within 3 months to 5 years old who were admitted due to persistent diarrhea in Shaheed Motahhari Hospital of Urmia enrolled to study. Patients were randomly allocated into two groups: in first group, all subjects received zinc supplement, but in the second group, the subjects received vitamin E in association with zinc supplement. Age, Sex, improvement, hospitalization period, weight gain, stool firmness in two study groups were analyzed using SPSS software ver16.

**Results:** There were no significant difference between two groups regarding age, sex ( $P=0.343$ ), weight ( $P=0.377$ ), height ( $P=0.723$ ), head circumference ( $P=0.971$ ), nutritional status ( $P=0.159$ ), and the duration of diarrhea ( $P=0.464$ ). Mean hospitalization period in the intervention group was  $3.39\pm 0.78$  days, and  $3.92\pm 1.94$  days in the control group ( $P=0.054$ ). Regarding the improvement of appetite in the follow-up period, there was a significant difference among two groups ( $P=0.026$ ). Chi-square analysis revealed no significant difference in complete or relative improvement of diarrhea or response to treatment between two groups ( $P=0.374$ ).

**Conclusion:** Although the improvement process was not significantly different among two groups in our study, the difference in the improvement of appetite during follow-up period was supporting the hypothesis which the administration of vitamin E in children with persistent diarrhea could be useful in restoring the health status and wellbeing of children. Moreover, considering weekly significant difference in the hospitalization period of patients in two groups, further similar studies with larger sample size is

recommended to investigate the role of vitamin E in decreasing the diarrhea related costs.

**Keywords:** Persistent Diarrhea, Vitamin E, Double-blinded Clinical Trial, Children

### Efficacy of ursodeoxy cholic acid in management of hepatitis due to anticonvulsive drugs in children

Asgarshirazi, M., MD; Keyhanidoost Z., MD; Department of Pediatrics, Tehran University of Medical Sciences

**Background:** The liver plays a central role in drugs biotransformation that have two aspects: activation & detoxification. Sometimes the liver will be damaged in this process. Anticonvulsants are an important group of drugs that have been associated with a broad range of adverse effects, specially in liver. Patterns of liver test abnormalities in drug hepatotoxicity can be hepatitis or hepatocellular ( $ALT/ALP \geq 5$ )- cholestatic ( $ALT/ALP \leq 2$ ) or mixed ( $ALT/ALP > 2$  to  $< 5$ ). Ursodeoxycholic acid (UDCA) is a naturally occurring dihydroxy bile acid that has been effective in treatment of cholestatic disorders. It acts through replacement of intracellular hydrophobic bile acids, has a direct hepatoprotective effects & improves mitochondrial OXPHOS & stabilizes the mitochondrial membrane.

**Methods:** from 2009 April to 2010 August, 22 infants & children whom have been on anticonvulsive therapy with Sodium valproate- Primidone- Kepra (Levetiracetam)- & phenobarbital & have shown transaminases rise (2 times or higher than normal upper limits), after exclusion of other causes for hepatitis (viral & autoimmune) & abdominal sonography have been treated with UDCA. All of them had been undergone metabolic diseases screening before anticonvulsant therapy. Before treatment with UDCA, minimum doses of anticonvulsant drugs that have been effective in control of convulsion were tried & in case of transaminases rises after 2 wks, treatment with UDCA has begun.

**Results:** In these 22 patients 16 boys and 6 girls with mean age of 22 mos, with aminotransferases rise during anticonvulsant therapy we started UDCA in doses of 10-20 mg/kg/day & follow up them. 10 patients had received Sodium valproate, 6 patients Primidone, 2 patients Kepra & 4 patients Phenobarbital. The range of pretreatment ALT was 164-423 U/L & AST 95-216 U/L. By starting of UDCA, both of them declined to normal or near normal range after mean 21 days.

**Conclusion:** Our findings showed that although UDCA is used in cholestatic disorders but can be also useful in

management of hepatocellular damage arising from drug hepatotoxicity.

**Keywords:** Drug hepatotoxicity, Anticonvulsants, UDCA

### Hepatobiliary diseases in Cystic Fibrosis

*Kavehmanesh, Z., MD; Baqiyatallah Medical Sciences University*

Liver involvement in Cystic Fibrosis is much less common than both respiratory and pancreatic problems which can be present as much as 80-90%. Liver involvement at postmortem studies is up to 70%. Liver disease can be asymptomatic or progressive. Clinical liver disease affects about 30-40% of CF patients which are increased by age from childhood to adulthood. Increase life expectancy due to better control of extrahepatic problems brings liver disease as a challenging issue. A broad clinical spectrum of hepatobiliary problems are associated with CF, the problems may be mild steatosis due to poor nutrition or multilobular cirrhosis and portal hypertension. Other hepatobiliary complications of CF are neonatal cholestasis (1-2%), biliary obstruction, microgallbladder, cholelithiasis (12%), atretic cystic duct, distended gallbladder, common bile duct stenosis, sclerosing cholangitis, etc. The factors responsible for the progression of liver disease in CF are unknown, but may be influenced by genetic modifiers. Liver disease are shown to be increased in patients having  $\Delta F508$  mutation. Early diagnosis and treatment of CF hepatobiliary problems may decrease their morbidity and mortality. Clinical evaluation of the patients include routine liver physical examination, liver function tests, total protein, serum albumin, PT, PTT and also ultrasonographic imaging of hepatobiliary systems. Treatment relies on correction of nutritional status, supplementation of fat soluble vitamins, essential fatty acids, carnitine, choline and minerals such as zinc. Treatment of end stage liver disease complications such as ascites, pruritis and portal hypertension also may be necessary. Early treatment with ursodeoxycholic acid may be helpful in chronic liver disease of CF patients.

**Keywords:** Hepatobiliary, liver, cystic fibrosis

### Role of synbiotic in the treatment of childhood constipation: a double-blind randomized placebo controlled trial

*Khodadad, A., MD, Sabbaghian, M., MD; Children's Medical Center, Tehran University of Medical Science*

**Background:** Constipation is a common problem in children. There is some clinical evidence for the role

of probiotics in the treatment of constipated children. This is the first study about the therapeutic effect of synbiotic (combination of probiotic and prebiotic) in treatment of childhood constipation.

**Methods:** A double-blind randomized placebo controlled study (RCTs) was performed. 102 children aged 4–12 years with functional constipation according to Rome III criteria were assessed for 4 weeks. They randomized in 3 groups: Group A, received 1.5 ml/kg/day oral liquid paraffin plus placebo, group B, 1 sachet of synbiotic per day plus placebo and group C, 1.5 ml/kg/day oral liquid paraffin plus 1 sachet of synbiotic per day. Frequency of bowel movements (BMs), stool consistency, number of fecal incontinence episodes, abdominal pain, painful defecation per week, success treatment and side effects determined in each group before and after treatment.

**Results:** The frequency of BMs per week increased in all groups ( $P < 0.001$ ), but it differ between groups and was higher in group C ( $P < 0.05$ ). Stool consistency increased and number of fecal incontinence episodes, abdominal pain and painful defecation per week decreased in all groups similarly and was not statistically difference between them. No side effects were reported in group B and the main side effect in group A and C was seepage of oil ( $p < 0.001$ ). Treatment success was similar and there was not significant different between groups ( $p = 0.559$ ).

**Conclusion:** This study shows that synbiotics have got positive effects on symptoms of childhood constipation without any side effects.

**Keywords:** Constipation, Synbiotic, Probiotic, Prebiotic, Paraffin oil

### Cholestatic form of hepatitis A; atypical presentation of hepatitis A

*Modaresi, V., MD, Social Security Organization, Shohada Hospital, Yazd*

**Background:** Prolonged cholestasis is an unusual feature of Hepatitis A. Common manifestations of cholestatic viral hepatitis A are pruritus, fever, diarrhea, and weight loss. Serum bilirubin levels are often more than 10 mg/dl and the clinical course lasts at least 12 weeks. Cholestasis will spontaneously resolve, although corticosteroids will hasten the resolution but may predispose the patient to develop a relapse of the hepatitis. Autoimmune hepatitis is a chronic hepatitis of unknown etiology characterized by hyperglobulinemia, the presence of certain circulating autoantibodies, and inflammatory changes on liver histology. However one of several "triggers" for autoimmune hepatitis, particularly are viral hepatitis, which may incite the development of autoimmunity in predisposed individuals. Case series presentation: In this case series, we report the cases of three patients presenting with a clinical syndrome typical of cholestatic hepatitis who had previously acute hepatitis A. Our patients showed usual features of cholestatic hepatitis including; pruritus, mild fever and diarrhea, with serum total bilirubin levels greater than 10 mg/dL and direct

bilirubin around 6-8 mg/dl, The finding that was attractive to us, was high globulin level which is supposed to be related to autoimmune hepatitis, assessment of serologic markers for it were negative. Followed our patients revealed a clinical course lasting about 12 weeks with resolution of globulin level. All patients recovered completely without sequel.

**Conclusion:** Actually presentation of autoimmune hepatitis may be similar to what is seen in cholestatic prolonged jaundice after Hepatitis A. Thus management often is including evaluation of both autoimmune hepatitis and obstructive conditions such as sclerosing cholangitis. In addition it is advised to assessment of Wilson disease. Since prolonged cholestasis has not been associated with a specific serologic marker, this study demonstrates that high globulin which is seen in some unusual cases of Hepatitis A could be an expected variant and there is no need to do invasive procedures such as liver biopsy that is involved in the evaluation of suspected obstructive jaundice or autoimmune hepatitis.

**Key Words:** cholestatic jaundice, hepatitis A, Hyper globulinemia

### Evaluation of different clinical manifestations in gastroesophageal reflux in infants referred to Madany hospital clinics, Khorram Abad

*Taee, N., MD, Faraji Godarzi, M., MD; Tarhani, F., MD; Abbasi, F., MD; Lorestan University of Medical Sciences*

**Background:** Inactive back of milk or food from stomach to esophagus is gastroesophageal reflux or GERD. Prevalence of GERD is frequent and usually are mild and asymptomatic in infants but in severe from weight loss, weight gain retarded based on age and other factors clinical manifestations are different. Goals of study were evaluation of different clinical manifestations in infants that GERD established with Ultrasonography.

**Materials:** This is retrospective study in 50 infants with GERD and different clinical manifestations. For all patients Ultrasonography done and GERD reported. Tools of study were answer sheets and then data analyzed. Patients treated with Ranitidine – Omeprazole- Metoprolol.

**Results:** In 50 infants with GERD, clinical manifestations were refused feeding in 95%, vomiting after feeding 80%, recurrent restlessness in 86%, feeding only at night in 74%, refuse feeding in day 64%, regurgitation of liquid and milk in 42%, Thumb sucking in 74%, up right position desire in 92%, sandfire position in 62%, miss diagnosed with infant colic and treatment for colic in 95%, all infants were 20 days to 6 months age.

**Conclusion:** refused feeding, vomiting after feeding, restlessness with feeding, Thumb sucking, sandfire position were the most signs and symptoms in GERD.

**Keywords:** Khorram Abad, GERD, Gastroesophageal reflux, Clinical manifestation

### Analgesic effect of expressed breast milk in neonates during venepuncture

*Tarhani, F., MD, Taee, N., BSc., Faraji, M., Dalvand, Sh., Tarrahi, M.J., Ahmadi, M., BSc; Lorestan University of Medical Sciences, Khorramabad*

**Background:** Newborn infants routinely undergo invasive procedures, even after uncomplicated birth. Evidence shows that neonates do feel pain and may even have increased sensitivity to pain and to its long term effects compared with older infants. Non pharmacologic interventions are valuable for pain relief during minor procedures in neonates. The aim of this study to assess the effectiveness of expressed breast milk (EBM) in reducing pain due to venepuncture and comparison with formula and 50% dextrose solution.

**Materials:** A clinical trial study was conducted in 30 term neonates. Each infant received 3 treatment with EBM, formula and 50% dextrose solution during consecutive Venous blood sampling and their responses were measured by behavioral acute pain rating scale for neonates (DAN Score). This study was done in form of double blind and observers were unaware of what was being given to the baby.

**Results:** This study showed that pain score significantly is lower in neonates who received EBM ( $P < 0.001$ ). There was no statistical relationship between age, sex and weight with pain score.

**Conclusion:** Expressed breast milk effectively reduces pain of venepuncture in term neonates and it could be natural, noninvasive and readily available analgesic when mother can not be present to breastfeeding.

**Keywords:** Analgesia, Expressed breast milk, neonate

### Breastfeeding and Helicobacter pylori Infection in Children with Digestive Symptoms

*Monajemzadeh, M., Farahmand, F., Vakilian, F., Mahjoub, F., Alam, M., Kashef, N.; Children's Medical Center, Tehran University of Medical Science*

**Background:** This study aims to evaluate the role of breastfeeding in the acquisition of H. pylori infection in Iran and to compare the histopathologic changes occurring in children feeding on breast milk with those in infants feeding on formula.

**Materials:** In a case-control study parents of children with and without H. pylori infection who had undergone endoscopic survey and gastric biopsy in the Children's

Medical Center, Tehran, were asked about their feeding practices during the first 6 months after birth, the duration of breastfeeding period, the symptoms, and the duration of symptoms and concomitant diseases.

**Results:** A total of 154 children were included in this study. From this sample, 77 children formed the case group and 77 children formed the control group. A significant difference was found between *H. pylori* infection and feeding with formula ( $P=0.045$ ). In case group, a significant difference was found between breastfeeding and age of the infected child ( $P=0.034$ ), shorter duration of symptoms ( $P=0.016$ ), and finally degree of *H. pylori* colonization ( $P=0.021$ ).

**Conclusion:** It appears that breastfeeding in the first 6 months after birth can decrease the degree of *H. pylori* colonization, postpone infection until older age, shorten the duration of symptoms, and be concomitant with milder gastritis.

**Keywords:** Breastfeeding, Gastritis, Helicobacter pylori, Formula

### An infant with hepatomegaly due to carnitin deficiency

*Famouri, F., MD, Department of Pediatrics, Shahrekord University of Medical Science*

**Background:** Carnitin is an aminoacid that transport fatty acids from cytosol into mitochondria for oxidation and produce energy. So, carnitin deficiency causes muscle weakness, accumulation of unmetabolized fat in liver and cardiomyopathy. One type of carnitin deficiency is carnitin-acylcarnitin translocase deficiency. It is an autosomal recessive disorder. The carnitin-actyl carnitin carrier is a protein of inner mitochondrial membrane which catalyses mole to mole exchange of internal carnitin and external acyl carnitin. The most common presenting features are: lethargy, poor feeding, arrhythmia, hypoketotic hypoglycemia and hyperammonemia. Hepatomegaly and hypertrophic cardiomyopathy may develop secondary. Treatment is avoiding hypoglycemia by frequent feeding, low fat high carbohydrate diet and supplement of carnitin for prevention of complications.

**Case report:** The 8 month old infant was referred to pediatric ward with hepatomegaly. She was well nourished (Wt: 8 kg, Lt: 68 cm, Hc: 44cm). The patient had mild sitting at 8 months old. Sonography showed increased echogenicity of liver probably due to fatty infiltration. She had no splenomegaly. Liver function test showed only moderate elevation of transaminases since 2 months old age. Laboratory data was- VBG: PH = 7.29, pCO<sub>2</sub>= 24 mmHg, PO<sub>2</sub>=45 mmHg, HCO<sub>3</sub>=12, serum Na=134 mEq/l, K=3.8 mEq/l, uric acid= 2.9 mg/dl, random blood sugar=65 mg/dl; Urine analysis: SG= 1015, no ketone, no sugar and protein; CXR: normal;

Ammonia= 88 mg/dl (NL: <90), lactate: 25 mg/dl (NL: <20). CPK and LDH were normal. MCHAD, LCHA and VLCHAD were normal but acylcarnitin was deficient. Liver biopsy showed severe hepatic steatosis without glycogenosis. Other metabolic screenings were normal. With diagnosis of carnitin deficiency, carnitin solution was started with dose of 100 mg/kg/day. After 3 months, she had good muscle tone, hepatomegaly subsided and liver enzymes became normal.

**Conclusion:** There are many diseases with fatty liver and elevation of transaminases. One of them is carnitin deficiency which can involve the other organs. Early diagnosis and treatment can prevent progression of disease and its manifestations. Key words: Hepatomegaly; Carnitin; Acyl carnitin

**Keywords:** Hepatomegaly; Carnitin; Acyl carnitin

### Effect of zinc supplement on treatment of pulmonary infection in children with CF

*Nemat Khorasani, E., MD, Mansouri, F., Kermanshah University of Medical sciences, Iran*

**Background:** Zinc has significant anti-oxidant and anti-inflammatory activity. Zn deficiency can occur in subsets of patients with cystic fibrosis (CF) especially those with malabsorption. Although Zn has significantly reduced infections in various disorders, But its efficacy has not been thoroughly investigated in CF.

**Methods:** We performed a double blind placebo controlled study to investigate the effect of daily 5mg/Kg (Max 30mg) elemental Zn for one year on rate of respiratory tract infection in 20 children with CF (ages 7-18). Plasma Zn, pulmonary function tests (PFT), Rate of respiratory infections and use of antibiotics measured at baseline and end of the study.

**Results:** Rate of respiratory infections and use of antibiotics was lower in Zn treated compared to placebo ( $P<0.05$ ).

**Conclusion:** Oral Zn supplementation (5mg/Kg) reduce the rate of respiratory infections and antibiotic usage in children with CF.

**Keyword:** Cystic fibrosis, Zinc, Respiratory Infection, antibiotic

### Dietary Factors And Dental Caries

*Ghandehari Motlagh, M., DDS, Department of Pediatric Dentistry, Tehran university of medical sciences*

**Background:** Sugars (sucrose, fructose, glucose, and others) are one of the major etiologic factors in dental caries (Newbrun, 1967), but in fact animal studies have shown other sugars, notably glucose and fructose, to be as cariogenic as sucrose. This poses potential difficulties in making dietary recommendations because many fruits and vegetables containing sugars further studies into the

relationship between diet and dental caries have Refined the knowledge given by the vipeholm and hopewood house data, but in general the Findings of those earlier studies have been confirmed. Much effort in recent years has gone into Assessing the relative cariogenicity of foodstuffs. The multiplicity of food factors requires that Relative cariogenicity tests be interpreted with caution. These factors include cariogenicity tests be Interpreted with caution. Detergent quality texture, the effect of mixing foods, the sequence of ingestion, the frequency Of ingestion, and the pH of the food itself.

**Keywords:** dental Caries, ingestion, Dietary factors

### Effect of Interferential electrical stimulation on constipation in children with myelomeningocele

*Sharifi-Rad, L., Physical Therapy, Children's Medical Center, Tehran University of Medical Sciences*

**Background:** To evaluate safety and efficacy of transcutaneous Interferential (IF) electrical stimulation on constipation symptoms in children with myelomeningocele (MMC) induced refractory neurogenic bowel dysfunction.

**Methods:** Twenty MMC children (11 girls, 9 boys), mean age 6.7±2.9, with moderate to severe retractable constipation were enrolled in this study. They underwent Anorectal manometry before and after IF therapy with attention to the rectoanal inhibitory reflex (ml) and sphincteric pressure (mmHg). Parents were instructed to complete a bowel habit diary by providing data about laxative intake, number of defecation per week, form of stool, episodes of pain during defecation. Fifteen courses of abdomen area of interferential electrical stimulation for 20 minutes 3 times per week were performed with low frequency current, duration of 250 µs and repeated time of 6.6 seconds. All children were followed for the next 6 months.

**Results:** Of Anorectal manometry parameters after IF implication, sphincteric pressure and rectoanal inhibitory reflex significantly improved pre-treatment measures ( $p < 0.000$ ). In 80% patients constipation symptoms were decreased immediately after IF therapy and persisted for 6 months ( $p < 0.000$ ). Frequency of defecation reduced from 2.5±1.1 per week before treatment to 5.3±2.3 per week after treatment.

**Conclusion:** This study demonstrated that noninvasive IF therapy is effective to improve constipation symptoms and Anorectal manometry parameters of MMC children with neurogenic bowel dysfunction.

**Keywords:** constipation, children, electrical stimulation

### Gastroesophageal Reflux Disease in Children Presenting with Chronic Cough

*Faghihinia, J., Modaresi, MR., MD, Isfahan University of Medical Sciences*

**Background:** Coexistence of Gastroesophageal Reflux (GER) with a variety of respiratory symptoms has been reported. Among children, cough is a very common symptom of respiratory disease and reason for parents to seek medical attention. We investigated the prevalence of GER in children with chronic cough using ultrasonography as the diagnostic method.

**Methods:** This cross-sectional study was conducted in Pediatric Clinics of Isfahan University of Medical Sciences in 2008. Children with 3 months to 6 years of age who referred with chronic cough (cough for more than 3 weeks) of unknown etiology were evaluated for GER by two radiologists using ultrasonography.

**Results:** During the study period, 163 children were investigated. The mean age was 2.7±1.2 years and 92 (54.8%) were male. GER was diagnosed in 94 (56%) of the children. There was not significant difference between GER positive and GER negative children in sex but the prevalence of GER was significantly higher in those less than one year old (61.8%) and those with wheezing (60.3%);  $P < 0.05$ .

**Conclusion:** There is a considerable prevalence of GER in children referring with chronic cough and those with wheezing are at a greater risk. Ultrasonography is a useful and non-invasive method for diagnosis of GER in this population.

**Keywords:** Chronic cough, gastroesophageal reflux, respiratory symptoms, wheezing

### Reliability, Safety and Effectiveness of Bravo Capsule, A Catheter Free pH monitoring System for Evaluation of Gastroesophageal Reflux Disease in Children

*Karjoo, M., MD, Beg, M., MD, Kesselring, Sh., MS; Division of Pediatric Gastroenterology and Nutrition Upstate Medical University, Syracuse, NY*

**Background:** Gastroesophageal reflux disease is a common problem seen in pediatrics and adolescents. The Bravo capsule pH monitoring system is used for detection and to quantify the severity of reflux disease. This test is also valuable for evaluation of patients with extra-gastrointestinal manifestations of GERD such as asthma, chronic cough, hoarseness, aspiration pneumonia, and pre/post fundoplication surgery. Our aim was to evaluate the safety and reliability of this procedure in children.

**Methods:** From January 2002 to June 2010, 166 patients, 67 males and 99 females' ages 6-18 years underwent upper endoscopy with biopsies and placement of Bravo capsule. The endoscopy procedure was done in the operating room, under general anesthesia, using GIF 160

scope. Readings were done after 48 hours. In two patients, the Bravo and conventional pH probe were placed simultaneously and the recordings were compared.

**Results:** 1). in 2/166, both males, the pH dropped to below 2 after half hour. This indicated that the capsule fell down from the esophagus to the stomach. One of the patients deleted all data while playing with the recorder at home. 2). In 2 patients where Bravo and conventional pH monitoring were done simultaneously, the recording of the pH probe showed a slightly higher, more alkaline pH recording. This may indicate a possible decreased sensitivity of Bravo capsule compared to the conventional pH probe. 3). In 157/166, of patients tolerated the Bravo capsule placement with no complaints. Nine patients had a mild sensation of sub-sternal pain for 2 hours or less. In 120/166 had abnormal results, with fraction time Ph. below 4 ranging 5.6- 19.7%, except in one case with severe reflux disease ranging 51.6% in day one and 45.4% in day 2. In 46/166 with normal pH., recording with fraction time pH below 4 ranging 1 to 3.2%.

**Conclusions:** Bravo capsule pH monitoring of the esophagus is a reliable, natural, safe, well- tolerated procedure, free of significant complication, which is preferred by young' adolescents. With Bravo capsule, adolescents can go to school and continue their normal physical activities without interruption.

**Keywords:** GERD, esophageal pH, asthma, fundoplication, heartburn

### A study of the factors related to hospital malnutrition with a grading system

*Sayyari, AA., MD, Imanzadeh, F., MD, Javaherizadeh, H., MD, Moradi, M., MD, Ghoroubi, J., MD; Peyvaste, M., MD, Shamshiri, AR, Ph.D*

1. *Mofid Children's Hospital, Shahid Beheshti University*

2. *Imam Khomeini Hospital, Joundi-Shapour University of Medical Sciences, Ahwaz, Iran*

**Background:** About 0.5% of children that admitted in medical and surgical ward are prone to nutritional insufficiencies. The aim of this study was to create a simple system to identify a child who is prone to malnutrition during hospital stay.

**Methods:** Patients whom admitted to hospital aged  $\geq 1$  mo and length of hospital stay more than 48hrs were included in this study. Every patients with renal diseases, hepatic disease, heart failure, with special diet regimen ( diabetic, ketogenic) were excluded from this study. Parental agreement must be signed for each cases. Nutritional risk factor was evaluated in 322 children. Anthropometric, intake of calorie, type of illness, ability to swallowing, and conflicting factor with nutrition was assessed during 48hr after admission. Diseases were classified into

mild (1), moderate (2), and severe (3). Weight reduction was evaluated by regression analysis. Disease severity were classified into 3 grade (grade 1: mild, grade 2: moderate, grade 3: severe). Patients were divided in 2 groups: group I: without weight reduction or  $<2\%$ , group II: weight reduction  $\geq 2\%$ . Grade I disease include admissions for diagnostic purposes, minor surgery, gastroenteritis, urinary tract infection. Grade 2 disease includes common surgical procedure, non fatal infection, non progressive chronic disease, and mild chemotherapy. Diseases with severe grade include major surgery, severe infection, acute illness on the chronic disease, and high dose chemotherapy. Age, sex, disease severity, respiratory difficulty, calorie intake, diarrhea, vomiting, dysphagia, length of hospital stay, type of disease ( surgical vs medical) were assessed as the predictor of weight reduction. With regression analysis predicted probability was calculated from the following formula.  $F = 1.779 \times (\text{calorie intake} \leq 50\%) + 2.089 \times \text{grade II disease} + 5.3 \times \text{grade III disease} - 2.608$ . If each of factor is not present, its value equal =0. Nutritional risk factor has a range from 0 to 4. Risk degree 1 means moderate and risk degree  $\geq 2$  means high risk for malnutrition.

**Results:** Group I include 156 patients (48.44%) and group II include 110 patients (51.56%). From all cases, 226 (70.2%) patients was admitted in non surgical ward and 96 cases ( 29.8%) were admitted in surgical ward. One hundred and fifty patients (46.6%) had caloric intake less than 50% and 172 cases (53.4%) had caloric intake  $\geq 50\%$ . From our cases depends on acute malnutrition: 64 (19.9%) cases had mild malnutrition, 15 (4.7%) cases had moderate, and 8 patients (2.5%) had severe malnutrition and 235 cases (73%) had not evidence of malnutrition. Caloric intake in group I was  $>50\%$  in 124 cases and  $\leq 50\%$  in 32 cases and in group II were  $>50\%$  in 48 cases and  $\leq 50\%$  in 118 cases ( $p < 0.05$ ). Malnutrition analysis showed that calorie intake equal or less than 50%, disease grade 2 and 3, and length of hospital stay were related to weight reduction  $\geq 2\%$  ( $P < 0.05$ ).

**Conclusion:** Calorie intake, disease severity, and length of hospital stay are the most important factor for predicting hospital related malnutrition. This method is a easy method for predicting possibility of malnutrition in hospital and may prevent hospital malnutrition.

**Keywords:** malnutrition, Nutrition, risk factor

### Study of Helicobacter pylori infection in diabetic cases

*Imanzadeh, F., MD, Mofid Children's Hospital, Shahid Beheshti University of Medical Sciences*

**Background:** Helicobacter pylori is one of the most important pathogen in the gastrointestinal tract. There is some report about helicobacter pylori and diabetes mellitus. The aim of this study was to evaluate helicobacter pylori infection in cases with diabetes mellitus (Type I).

**Methods:** this case control study was carried out in Mofid Children's Hospital and Loghman Hospital during 2008-2009. Diabetes in each patients was confirmed by an experienced pediatrician with subspecialty in pediatric endocrinology. Each diabetic cases who had not history of antibiotic therapy in 2 months ago were included in this study. Urea breath test was done with ISOMAX 2000 made by Canada. 49 cases ( M:28, F: 21) underwent urea breath test to detect *H. pylori* infection. 108 controls, who without history of antibiotic therapy and diabetes were selected. Urea breath test was performed for all controls children. Chi-square test was used for analysis with SPSS ver 11.5.

**Results:** Forty- nine cases with mean age=8.1±2.6 (Max=13,Min=3.5) were studied. From cases group, 14 cases had positive UBT test . From 108 control persons, 30 (27.8%) (M=16 (27.6%), F=14 (28%)) had UBT positive results. From 49 cases, 14 cases (28.6%) had positive UBT. In case group, 28.6% in male and female group had positive results (P value>0.05). In our cases, 10 cases had abdominal pain, 6 cases had anemia, 8 cases had vomiting.

**Conclusion:** We did not find significant differences between two groups. Another study with more sample is recommended.

**Keywords:** Helicobacter pylori, diabetes, Infection

### **Vomiting in GI Tract Diseases: Clinical findings and Imaging procedures: When, why, how?**

*Mearadji, M., MD, International Foundation for Pediatric Imaging Aid, Netherland*

Disorders of GI-tract are the most common cause for vomiting in pediatric age group. Generally vomiting occurs in conjunction with other symptoms which vary depending on the underlying disease, disorders or condition such as abdominal pain or distention, diarrhea or constipation, complicated incidentally with dehydration. The type and content of vomities (emesis) are valuable clinical information such as hematomases or bile stained emesis and projectile or less severe vomiting. From differential diagnosis point of view the pediatric patients with a GI-tract disorder should be classified in 3 age groups as neonates, infants and older children. In the neonatal period vomiting is frequently related to congenital abnormalities of GI-tract, while in infancy and childhood acquired disorder are the most usual findings. An accurate clinical evaluation leads to an indication for the imaging of GI tract. The conditions not related to GI with vomiting, as CNS, pulmonary and renal diseases, etc as well as short term vomiting by gastro enteritis should be clinically excluded and differentiated from GI disorders. The diagnostic approach by one or more modalities depends on the clinical findings. Nowadays the available modalities

used in imaging of GI are abdominal plain films, ultrasound, examinations with contrast media, CT, MRI and nuclear scanning. Obviously endoscopic examination has priority in numerous acute and chronic diseases of upper and lower GI-tract. An abdominal plain film is usually the first step by an acute abdomen. Otherwise the use of an adequate modality depends on additional clinical symptoms of GI, age and condition of patient. Within scope of this presentation the clinical signs and symptoms of frequent observed disorders, congenital of acquired in different age group will be shown. Answering the question when and why an imaging procedure is indicated will be discussed. The use of an appropriate modality to achieve an optimal result (how) should be demonstrated.

**Keywords:** vomiting, GI tract, pediatric

### **Infantile colic due to cow's milk protein allergy**

*Razmi, V., MD, Social Security Organization, Kerman*

**Background:** Despite of benign nature of infantile colic, it can be a source of anxiety in family and disturb mother's reactions. Recent evidence suggests that presentation of infantile colic in breast fed infants may be the consequence of delayed gastric emptying and dysmotility due to sensitization to trace amounts of maternally ingested antigens passing into breast milk. This study was conducted to investigate the effectiveness of elimination of milk products from mother's diet whose infants suffered from severe colic.

**Methods:** Twenty three breast fed infants with age range of 2-4 months, who had crying more than 3 hours a day and every pathologic etiology were ruled out in their initial assessment, were taken part in this study. All parents' subjects received necessary education about home management of colic and reassured. Then using simple method of random sampling, participants divided into two groups, (group I including 10 and group II including 13 infants). Mothers in group I advised to continue to previous method of care and feeding, and in group II they advised to not to take any cow's milk product for following 4 weeks. All subjects of both groups visited regularly every week and for each case, time of crying recorded in addition to growth monitoring and control for other problems.

**Results:** 2 out of 10 infants in group I (20%) and 8 out of 13 (61.53%) in group II showed improvement in well being and also decreased crying time every day. Mothers in group II were complained about relative decrement in milk production, although their infants gained weight properly.

**Conclusion:** Improvement in infantile colic was significantly obvious in group whose mothers eliminated cow's milk products from their diet, suggests a trying of this option for infants who suffered from prolonged crying.

**Keywords:** Food Allergy, Enteropathy, Infantile colic

### **A randomized clinical trial with probiotics in acute viral diarrhea in Iranian children**

*Allahverdi, B., MD, Ghorbani Vaghi, A., Foroughi, S., Miri, M., Delfan, B., Mozafari Kermani, R., MD*

**Background:** Gut microbial composition has major impacts on health . Villous absorptive capacity, normal barrier mechanism and local immune defense all act in optimal level when intestinal microbiota are intact . Viral diarrhea can commonly disturb enteric microbial milieu . Replacement of normal flora can potentially reduce disease severity .

**Materials:** 100 children between 12 and 60 months with acute viral diarrhea enrolled a double blind clinical trial from April to September 2008 in Lorestan, Iran. 50 children were given one sachet lyophilized probiotic extract (10 (9)CFU) daily for 7 days and 50 ones took placebo (in similar sachet form) for same duration . Dietary and hygienic orders were

similar. Each child was closely observed regarding complications and disease severity. After 4 weeks patients were weighed. Data were analysed by SPSS-13 and paired t-test was used .

**Results:** There was no significant difference regarding sex ( $p=0.754$ ) and mean age ( $p=0.983$ ) between two groups. Mean duration of diarrhea was  $4.5\pm 1.03$  days in probiotic group vs.  $6.8\pm 2.6$  days in placebo one ( $p=0.000$ ). Daily times of watery stool in case and control groups were  $4.36\pm 0.95$  and  $5.08\pm 0.89$  respectively ( $p=0.000$ ). Nobody in probiotic group suffered by persistent diarrhea or any significant complication . Weight decrement after probiotic challenge was significantly less than placebo ( $p=0.000$ ) .

**Conclusion:** Industrial and natural probiotic containing meals have documented promising effects in viral diarrhea. They can be considered as part of management protocol in childhood diarrhea .

**Keywords:** Probiotic, Children, Viral diarrhea, Lorestan, Iran

## Hematology & Oncology Abstracts

### Pre- and post-hematopoietic stem cell transplantation (HSCT) evaluation of children.

*Kato, Sh., MD, PhD, Department of Cell Transplantation & Pediatrics Tokai University School of Medicine, Isehara, Japan*

Allogeneic HSCT is nowadays largely accepted as a standard treatment for children with many hereditary and/or hematological disorders, of both malignant and non-malignant nature. More than 40 years have elapsed since the first successful HSCT in a patient with SCID, and many children were cured from their original disease and have become long term survivors. The following issues will be presented and discussed in this lecture. 1- Patient selection: Indication of HSCT has been changed from time to time and from disease to disease. Current guideline for a variety of childhood diseases (hematologic malignancies, immunodeficiencies, bone marrow failure syndromes and metabolic disorders) of the Japan Society for Hematopoietic Cell Transplantation (JSHCT) will be presented. 2- Donor selection: HLA-identical sibling was the only type of donor routinely accepted for the first two decades, but matched unrelated voluntary adult donors, unrelated cord blood (UCB) units and partially matched family members have become available as alternative donors in the last two decades. Special ethical consideration should be paid when minors are selected as donors for HSCT. The ethical and technical guidelines for childhood donation of hematopoietic stem cells of the Japanese Society of Pediatric Hematology will be presented. 3- Quality of life in long-term survivors: any children have become long-term survivors, and they are at risk of developing many kinds of organ dysfunction, including endocrinological dysfunction, growth impairment, infertility, cardiac dysfunction, respiratory dysfunction, secondary malignancy and so on. The details of our experiences will be presented.

### Cord blood banking and cord blood transplantation in children in Japan

*Kato, Sh., MD, PhD; Department of Cell Transplantation & Pediatrics; Tokai University School of Medicine, Isehara, Japan*

Cord blood transplantation (CBT) has become a therapeutic option in hematological malignancies and genetic diseases not only in children but also in adults without a histocompatible sibling donor or a

matched unrelated donor. More than 550,000 cord blood units have been stored, and more than 18,000 unrelated CBTs have been performed through cord blood banks worldwide. CBT has several advantages, including rapid availability without donor coordination, decreased viral transmission, reduced acute and chronic graft-versus-host disease (GVHD) and less HLA matching. CBT, however, has several limitations, including delayed neutrophil and platelet engraftment, prolonged immune recovery and increase in infectious complications. We report the current status of CBT and CB banking in Japan. The first CBT from a sibling donor was performed in 1994 in Japan. Several local cord blood banks were born between 1995 and 1996, and the first unrelated CBT was done in 1997. The Japanese Government decided to establish and financially support a network of those banks in 1999. Since then more than 43,000 cord blood units have been stored for transplantation, and more than 6,600 UCBTs have been reported to JCBBN. The number of UCBT has been increasing steadily, and becoming almost equal to that of unrelated bone marrow transplantation in the last five years. In the first five years (1997-2001), majority of UCBTs were done in children. However, early promising results of UCBT in adults encouraged adult patients and physicians, and the number of UCBTs in adults have been increased very rapidly in the following years.

### Relationship between MCV / MCH and severity of beta globin gene mutations in beta-thalassemia carriers

*Ehsani, MA., MD, Salamati, P., MD, Jamshidi, R., PhD, Holakouie Naieni, K., PhD, Derakhshandeh-Peykar, P., PhD,*

1. Dept. of Pediatrics, Tehran University of Medical Science
2. Dept. of Statistics, Iran University of Medical Science
3. Dept. of Epidemiology, School of Public Health and Institute of Public Health Tehran University of Medical Science
4. Dept. of Medical Genetics, School of Medicine, Tehran University of Medical Science,

**Background:** Thalassemia as a heterogeneous disease is one of the most common single gene disorder with a worldwide distribution. The aim of this study was finding a relationship between blood indexes and severity of beta globins gene mutations in beta-thalassemia carriers.

**Methods:** In this cross-sectional study, we were determined 30 beta globin gene mutations in 1206 unrelated beta thalassemia carriers. Furthermore their blood indexes, including CBC and electrophoresis were

also prepared. Then, by using SPSS software and t-test, the relationship between genetic findings and the results of their blood parameters were analyzed.

**Results:** In this study, the relationship between the severity of beta globin gene (b<sup>+</sup>/b<sup>o</sup>), in beta thalassemia carriers, and their average blood indexes, were evaluated. The results indicated that b<sup>+</sup> thalassemia in comparison with b<sup>o</sup> thalassemia had a lower mean MCV and MCH value. That means with less time and expense it could be possible to find a statistically significant relationship between a specific ranges of blood indexes and type of mutations in beta thalassemia carriers.

**Conclusion:** The results confirmed a significant correlation with blood indexes and certain type of mutations in beta thalassemia carriers.

**Keywords:** Thalassemia, Globin mutation, Blood beta

### Prevalence of Histopathologic Solid tumor in Khozestan province

*Abdollahi, A., MD, Rashidi, I., MD, Akyash, N., MD, Bozar, M., Moaven Jola, E., Tehran University of Medical Sciences*

**Background:** Malignancy is the most common disease causing death among children between 1 & 16 yr of age. In this study, with the evaluation of solid tumor histopathologic prevalence, thoroughly and separately, and also their generic and age-wise prevalence in Golestan and Imam Khomeini university hospital, So that, by using the results, we can perform more studies for preventing and treating the patients.

**Materials:** During a cross-sectional study By using the pathology departments archives of Golestan and Imam Khomeini hospital, the name of children whose their pathologic specimen referred to above-mentioned department, and their pathologic diagnosis was extracted. Then, these tumor types, age and sex of the patient were noted to evaluate the types and distribution in different age groups, and findings analyzed, reported descriptively.

**Results:** The most findings in solid tumors(27.9%) are related to the neural(12.5%) and lymphatic tumors(12.5%). We have germ cell tumors that include 4.16% of lesions. And finally, 70.83% of cases are related to other tumors. 72.9% of patients were male, 27.08% were female.

**Conclusion:** The prevalence of solid malignant tumors at this age group was less than other studies. The prevalence of central nervous system tumors was the most and after that was Lymphoma like other studies. The prevalence of Lymphoma was higher than other studies that causes of it should be researched.

**Keywords:** Malignant tumors, Children, Prevalence, Histopathology

### Aleration of Proapoptotic genes BAK, BAX and BIK in ALL patients.

*Isaian, A., Ph.D, Izadyar, M., MD, Kochakzadeh L., MD, Ramyar, A., MD, Alavi, S., MD, Nourbakhsh, S., MD, Sanati, MH., Ph.D; Tehran Medical Sciences University, Children's Medical Center*

**Background:** This study was designed to study the role of proapoptotic BAK, BAX, and NBK/BIK genes in a group of patients with ALL elucidate the possible role of these genes in progression of malignancies in this diseases.

**Methods:** Fifty Iranian patients with ALL and hundred Iranian healthy controls were investigated in this study. The entire coding regions of the BAK gene (exons 2–6), NBK/BIK gene (exons 2–5), and BAX gene (exons 1–7) were amplified using polymerase chain reaction (PCR). The PCR products were separated by 2% agarose gel electrophoresis, and all positive samples were verified by direct sequencing of PCR products using the same primers used for PCR amplification, Big Dye chemistry, and Avent 3100 Genetic Analyzer following the manufacturer's instructions (Applied Biosystems).

**Results:** In ALL groups had the BAK exon 2 c342C>T alteration with frequency of (34%) and P<0.0000001 which was significantly associated with increased risk of developing leukemia and lymphoma. Higher frequency of another nucleotide substitution in the noncoding region of exon 7 in BAX gene (6855G>A) was also identified with in 96% and with P<0.1343 OR; 3.273 95% CI (0.63-17.07). There were alteration in BAX exon 1 (g.146C>T) in 72% ALL patients with P value <0.0001; OR; 6 which was significant.

**Discussion:** Several alterations in the proapoptotic genes BAK, NBK/BIK, and BAX were found in our study, which could elucidate involvement of the mitochondrial pathway mediated apoptosis in accelerating and developing of cancers. There were cases where chemotherapeutic drug treatment has resulted to chemoresistance due to mutations in BAX and BAK genes. Understanding mitochondrial apoptosis process will go a long way in developing new rationally designed drugs which could be used to pharmacologically manipulate this process to induce apoptosis in treating cancers and autoimmune disorders.

**Keywords:** ALL; Acute lymphoblastic leukemia, BAX; BCL-2 associated x protein,

### An In Vitro Investigation Of Anticancer Effect Of Biopigment On Leukemia

*Ahmadi Fakhri F., Khanafari A., Baseri M., Yaghoobi R., Shahghasempour Gh., Ph.D Student, Microbiology Department,*

1. Islamic Azad University, Sciences and Researches Branch, Fars- Iran.
2. Microbiology Department, Islamic Azad University, North Tehran Branch, Tehran,

3. *Islamic Azad University, Kazeroon Branch*
4. *Islamic Azad University, Jahrom Branch*
5. *Tissue Engineering and Nanomedicine Research Center, Shahid Beheshti University of Medical Sciences*

**Background:** The leukemias of childhood are the hematopoietic system cancers. These are the primary cause of cancer related mortality of children in the world. Biopigment isolated from bacteria attracts interest as a consequence of its anticancer activity. The objective of present research was to investigate the anticancer effect of violacein as a biopigment extracted from *Janthinobacterium lividum* against Jurkat E6.1 cells to introduce as a new generation of pharmaceuticals.

**Methods:** Violacein was extracted from *Janthinobacterium lividum* DSM1522T with ethanol, and purified by filtration and high performance liquid chromatography. It was loaded on dendrimers (G=2) and capped with ascorbic acid and evaluated by infrared spectroscopy (FTIR). The cytotoxicity assays of free violacein and dendrimer-ascorbic acid-violacein as a complex delivery system were evaluated against Jurkat E6.1 cells by tetrazolium reduction assay. In vitro release kinetics of violacein and ascorbic acid loaded in the final powder was carried out.

**Findings:** The results of the inhibitory concentration for 50% of the cells (IC50) values for free violacein and dendrimer-ascorbic acid-violacein as a complex delivery system, were 7 and 4  $\mu$ M, respectively. Total loaded of violacein rates for dendrimer-violacein and dendrimer - ascorbic acid - violacein were determined to be 28% and content of ascorbic acid release in dendrimer-ascorbic acid-violacein were determined as in 30 hours.

**Conclusion:** This complex delivery system was 5.0 $\pm$ almost 85 observed to be almost 2 more efficient compared with free violacein. Biopigments can lead to pharmaceutical delivery at specific sites and reduce side effects.

**Keywords:** Leukemia, Anticancer, Violacein, Dendrimer

### **Immunogenicity of trivalent influenza vaccine in children with acute lymphoblastic leukemia during maintenance therapy**

*Shahgholi, E., MD, MPH, Ehsani, MA., MD, Salamati, P., MD, MPH, Maysamie, A., MD, MPH, Sotoudeh, K., MD, Mokhtariazad, T., PhD, Mousavinia, F., MD, Khademi, J., MD. Tehran University of Medical Sciences*

**Purpose** The aim of this study was to assess the immune response of children with acute lymphoblastic leukemia (ALL) to influenza vaccine

and to compare it with healthy controls. Procedure Thirty-two children aged 1–18 years with ALL on maintenance therapy and 30 healthy sibling controls were enrolled in the study. All children were vaccinated with trivalent inactivated influenza vaccine. Hemagglutinin-inhibition (HI) antibody titers were determined in sera of both patient and control groups just before and 4 weeks after vaccination. The ability of each group to mount a protective ( $\geq 40$ ) and/or fourfold titer was measured. Results The protective response for virus subunits among patients and healthy controls were 43.4% versus 88% for H1N1 ( $P = 0.04$ ), 63.3% versus 80% for H3N2 antigens ( $P = 0.06$ ), and 26% versus 73% for B antigen ( $P = 0.001$ ). Responses for H1N1 and B subunits were significantly lower in patients than controls. In the patient group, the significant response to each virus was demonstrated in the analysis of pre- and post-vaccination geometric mean titer (GMT) ( $P = 0.001$ ). The percentage of patients and controls with fourfold increase in HI titers were 56.2% versus 80% for H1N1 ( $P = 0.04$ ), 40.6% versus 53.3% for H3N2 ( $P = 0.31$ ), and 59.4% versus 83.3% for B ( $P = 0.038$ ). Immune responses for H1N1 and B subunits were significantly lower in patients than Controls Influenza vaccine is tolerated well in ALL patients with acceptable but limited immune response compared to healthy controls. These findings support the recommendation for annual influenza vaccination in children with ALL

**Keywords:** ALL;infection in immunocompromised hosts;immunology;pediatric hematology/ oncology; vaccines

### **Evaluation of Rituximab Treatment for Children with Chronic Immune Thrombocytopenic Purpura in Iran**

*Mirbehbahani, NB., Rashidbaghan, R., Payab, Z, Hemathology and Oncology Research center- Shahid Beheshti University*

**Background:** Immune thrombocytopenic purpura (ITP) is an autoimmune disease that is resulted from increasing autoantibodies against self-antigens and T-cell mediated cytotoxicity. This occurrence causes platelet destruction. In this study, we evaluated the rituximab treatment in children with chronic immune thrombocytopenic purpura (ITP). Rituximab is an anti-cd20 chimeric monoclonal antibody which has been used for treating different autoimmune hematologic diseases like ITP and has had beneficial role in their treatment.

**Method:** This investigation included 4 children (2 boys and 2 girls) with immune thrombocytopenic purpura, their ages arranged from 30 mth to 66 mth and the mean age was 47.25  $\pm$  14.10 mth.

Rituximab was given with a dosage of 109/L $\times$ 375mg/m2 weekly for 4 weeks. Platelet count in initial was less than 20 and responses were assessed in follow-up.

**Results:** The follow-up period after rituximab treatment ranged between 6 to 11 mth. Findings: the incidence of fever urticaria, pruritus, throat tightness and serum sickness was zero. On this treatment we didn't have any rising in platelet count.

**Conclusion:** this condition may be associated with the age of our patients whom were young at the time of commenced using rituximab. Rituximab may be an effective and safe treatment for adult with steroid-resistant ITP. We believe that more cases and studies are required to elucidate the reasons for different case series reported in literature.

**Keywords:** ITP, rituximab, autoimmune, hematologic diseases

### Approach to a Patient with Congenital Neutropenia

*Rezaei, N., Molecular Immunology Research Center; and Department of Immunology, School of Medicine, Tehran University of Medical Sciences, and Research Group for Immunodeficiencies, Pediatrics Center of Excellence, Children's Medical Center, Tehran University of Medical Sciences*

Neutropenia (or granulocytopenia) is a reduction in the absolute neutrophil count to less than 1,500/mm<sup>3</sup>. Impaired production, peripheral destruction, and abnormal distribution of neutrophils may lead to low numbers of circulating granulocytes. Congenital neutropenia is the most commonly encountered phagocytic defects, which could be associated with oculocutaneous hypopigmentation, pancreatic insufficiency, combined immunodeficiency, metabolic disease, and bone marrow aplasia/infiltration. Although the patients with severe congenital neutropenia and cyclic neutropenia usually have normal appearance, a group of disorders including Chediak-Higashi syndrome, Griscelli syndrome type 2, Hermansky-Pudlak syndrome type 2, and p14 deficiency are associated with oculocutaneous hypopigmentation in addition to neutropenia. Exocrine pancreatic insufficiency could be seen in Shwachman-Diamond syndrome and Pearson Marrow syndrome, whereas there are some evidences of combined immunodeficiency in CD40L deficiency, WHIM syndrome, cartilage hair hypoplasia and reticular dysgenesis. In addition to the above-mentioned disorders, congenital neutropenia can be seen in some metabolic diseases (such as glycogen storage disease Ib and Barth syndrome) and bone marrow aplastic diseases. Timely referral to a hematologist and/or clinical immunologist is the key point in early diagnosis of the patients, whilst appropriate approach to definite diagnosis and proper management can prevent further complications in this group of disorders.

**Keywords:** Neutropenia; phagocytic defects; oculocutaneous hypopigmentation; pancreatic insufficiency

### Allu-immunization in thalassemia major, and management

*Izadyar, M., MD, Children Medical Center, Tehran University of Medical Sciences*

Packed red cell transfusion is still the 1<sup>st</sup> line management in thalassaemia major. Regular red blood cell transfusion prevents of progressive, harmful anemia side effect such as cardiomyopathy and death, and also severe facial deformities. On the other hand transfusion has different side effects such as transmission of blood born infections, increased body iron load, and allu-immunization. In the allu...phenomenon, antibodies appear against transfused red blood cells and with each transfusion, the antibodies titers increase. So the patient with low hemoglobin level and no increment with transfusion. In this situation management is temporary and sometime unattainable: safe blood from a full compatible donor which never acceptable, immunosuppressive such as: Cyclosporine, azathioprine, IV IG that cost expensive. In my study I tried corticosteroids for 5 patients with unpreventable, progressive hemolysis. The patient's age were 3-25 years old, 1 girl, and the others were boys with history of transfusion on alternate day. Corticosteroids usage in long time, even months without transfusion, stabilized the hemoglobin level > 8 gm/dl. These 5 cases are on hydroxyurea for years, no transfusion, any changes in bone density, but with nearly normal life performances.

### Effect of miR-210 in hypoxia

*Mohammadi, Sh., Alizadeh, Sh., Mossahebi-Mohammadi, M., Degbakhsh, E.*

MicroRNAs (miRNAs) are small non-coding poly nucleotide which are encoded in genome of eukaryotes. This conserved molecules consist of 19-23 nucleotide which could regulate gene expression through specific mechanisms in Ratio of 1:1. miRNAs are capable of incorporation to different types of genes. According to high amount of miRs it is expected that these molecules have different roles in regulation of gene expression. miRs are responsible for regulation of 30 percent of genes. They also have roles in Development, proliferation, apoptosis, adipose metabolisms and cellular differentiation. miRNAs act via join to 3' UTR region of mRNA. miR210 encoded in intervening sequences (introns) of non-coding gene on chromosome 11. Hypoxia occurs in pathophysiologic conditions such as overgrowth of tissues, acute and chronic ischemia, tumor progression and mountain climbing. In response to hypoxia and regeneration of damaged cells HIF (Hypoxia induce factor) expression, which is a sensitive factor to hypoxia, is induced. HIF is a member of Helix-loop-helix family of transcription factors which causes activation of HIF through attachment to HREs and result in linkage of activated HIF to target genes. miR210 is One of the target genes of HIF. In hypoxic conditions

activated HIF attached to miR210 promoter which is the reason of enhancement in expression of miR210. Following Increase in levels of miR210 a change occurs in cell responses to hypoxia. One of these changed responses in this conditions is overcoming to arrest in cell cycle. This effect is due to suppression of transcription factor, MNT. MNT is a inhibitory receptor for transcription that links to MAX and is the antagonist of C-myc. Other effects of miR210 includes induction of angiogenesis in endothelial cells and increase in immigration and survival of these cells in hypoxia.

**Keywords:** HIF, miR-210, Hypoxia

### **Frequency of diabetes and its association with serum ferritin in thalassemia patients tehran 2009**

*Habibian, N., Azad University*

this study was conducted to determine the frequency of diabetes and its association with serum ferritin in thalassemia patients in Tehran 2009.

**Methods:** in this cross sectional descriptive analytical study 80 patients with thalassemia in Tehran in 2009 were recruited

**Results:** 15 patients 18.8% had diabetes. the mean serum ferritin level in diabetic patients was 3812.6 and in non diabetics was 1933.86 with a statistically significant difference.

**Conclusion:** according to obtained results, it may be concluded that cause of high frequency of diabetes among thalassemia patients is irregular or inadequate receiving of desferal or low quality of it.

**Keywords:** thalassemia -diabetes -ferritin

## Clinical Immunology, Allergy and Immunodeficiencies

### Fluticasone propionate in treatment of children with moderate asthma

*Ahmadi Afshar, A., MD, Moghimi Hadji, M., Zanjan University of Medical Science*

**Background:** Asthma is a common chronic disease. Beclomethason dipropionate (BDP) and Fluticasone propionate (FP) are two inhaled corticosteroids that frequently be used in treatment of patients with asthma. In this study, the effectiveness of BDP and FP in management of asthmatic children was investigated

**Methods:** In this trial, 50 children with moderate persistent asthma were randomly selected to receive either BDP 200 µg or FP 250µg for three months. Pulmonary function tests were measured in both groups at the beginning of study and monthly after treatment. Daily and night symptoms and consistency of drugs were also measured.

**Results:** There was significantly better FEV1 in patients receiving FP compared to the BDP group ( $P < 0.01$ ). There was also statistically significant difference in patients receiving FP in comparison with BDP group in increment of FVC, FEV1/FVC, FEF25-75 ( $P < 0.005$ ). Night symptoms were significantly improved in the FP group from the first month ( $P = 0.001$ ), whilst improvement of daily symptoms in this group compared to the BDP group was found from the second month ( $P = 0.001$ ).

**Conclusion:** Although symptoms and pulmonary function tests results were improved in both groups receiving either FB or BDP, this study suggested that FP was more effective than BDP in controlling moderate asthma in children.

### Prevalence of anxiety disorders among allergic asthma and rhinitis patients in Qazvin

*Kiani Amin, M., Fallahzadeh, M., Esfahani, A.; 1. Pediatric Immunology and Allergy Department, 2. Psychiatry department-Qazvin University of Medical Sciences*

**Background:** Asthma and Allergic Rhinitis are heterogenous disorders with different ongoing problems. The most important psychologic presentations which has demonstrated are anxiety and depression. Consequents of these psychosomatic problems recently has been shown, causes economic as well as social and medical disadvantages. This study was performed to assess the abundance of comorbid depression and anxiety in Allergic respiratory adult patients.

**Methods:** In this study 98 patients with respiratory allergy enrolled in three separate groups as Allergic rhinitis asthma and co-existence of asthma and allergic rhinitis (AR). From a total of 98, 61 out of 98 (61.3%) had allergic rhinitis, 26 out of 98 (26.1%) were asthmatic and 11 patients (11.2%) identified as both asthma and allergic rhinitis. Prevalence and severity of anxiety disorders in these subgroups identified by Hamilton questionnaire.

**Results:** Our results showed a general prevalence of 27.5% among these allergic patients. Prevalence of allergy revealed 26.2% in AR, 30% in asthma and 27.3% in simultaneous AR and asthma. It seems that asthmatic patients are more prone to psychosomatic disorders but severity was detected prominently in patients with AR.

**Conclusion:** Anxiety occurs significantly among allergic disorders and however it is not considered as an important factor, can cause problem in medical treatment or sometimes poor control.

**Keywords:** Allergic rhinitis, Asthma, Anxiety disorders

### Wheezing in children and infants: The response to l-epinephrine or aminophylline

*Modaresi, M., Khanlarpour, A., Phaghihinia, J.; Pulmonary Pediatrics Department, Al-zahra Hospital, Isfahan University of Medical Sciences*

**Background:** Respiratory distress is a leading cause of hospitalization in children. We compared the effect of intravenous (IV) Aminophylline and nebulized L-epinephrin on children with respiratory distress.

**Methods:** We included 100 infants and children 2 to 60 months old with a first episode of respiratory distress and wheezing admitted in the emergency department. The study was a double-blind, randomized trial comparing a three days of IV Aminophylline with nebulized L-epinephrin. The outcome was assessed for three days by the Respiratory Distress Assessment Instrument (RDAI).

**Results:** Although Aminophylline had significant improvement in wheezing and the total RDAI score on the first day of admission ( $p < 0.05$ ) but the improvement were not significantly different on the other days between both groups ( $p > 0.05$ ).

**Conclusion:** Because of narrow windows for therapeutics and toxic level of Aminophylline we recommend the Nebulized the nebulized L-epinephrine for improving the respiratory distress in hospitalized infants and children with first episode of respiratory distress

**Keywords:** Respiratory distress, Aminophylline, L-epinephrin

### **Exfoliative dermatitis : a rare presentation of allergic drug reaction in a patient with cystic fibrosis**

*Mansouri, M., MD, Chavouszade, M, MD, Khanbabae, Gh, MD; Mofid Hospital, Shaheed Behshiti Universit of Medical Sciences*

**Background:** Although Cutaneous adverse drug reactions are a frequent problem in clinical medicine. Exfoliative dermatitis is a rare presentation of a dermatological drug reaction and There are few reports in literatures regarding that. The incidence of allergic drug reaction in children is lower than adult and in early infancy it may still be extremely rare. The

**Case Report:** our patient was a boy aged 4 months admitted due to skin drug eruptions. The skin lesions included the erythematous plaques in malar area and throughout the body with the extensive exfoliation. He had no fever. He has received cotrimoxazole, forazolidon and ceftriaxone due to a chronic diarrhea since one months ago. The skin rash was compatible for drug allergy and resolved after receiving IVIG. Eventually the diagnosis of cystic fibrosis was also confirmed for him.

**Conclusion:** Nonimmediate allergic drug reactions comprise several diseases manifested as exanthematous, pustular and bullous eruptions with different evolution profiles and severity, among which, The most frequent are usually benign and comprise maculo-papular exanthema (MPE) and fixed drug eruptions (FDEs). 'Acute generalized exanthematous pustulosis' type IV delayed hypersensitivity reactions have been revisited to incorporate drug-induced Exanthema. There are few reports concerning exfoliative dermatitis in the elderly people, but its occurrence in early month of life is extremely rare. Risk factor for developing drug allergy are poorly understood but since sicker patients are often treated with multiple medications, increasing the risk for adverse drug interactions. Compromised renal and hepatic functions also contribute to drug allergy in chronically ill person. All these factors might have some roles in our patient's clinical presentations who had Cystic fibrosis as a background disease.

**Keywords:** Drug allergy, skin drug eruption, Nonimmediate allergic drug reaction

### **Allergic bronchopulmonary aspergillosis in differential diagnosis of hyper eosinophilic syndrome: a case report**

*Tabatabaei, A. MD, Hoshmand, S, Chavoshzadeh Z., MD, Eshghi P. MD, Ramyar, A., MD;  
1. Mofid Hospital. Shaheed Beheshti Medical University, 2. Children Medical Center, Tehran University of Medical Sciences*

**Background:** Allergic pulmonary aspergillosis (ABPA) can be complicated asthmatic or CF patients and must be considered in approach to patients with hyper eosinophilic syndrome (HES). A patient with HES was reported that had past history of hepatoblastoma and with lab workup, ABPA was detected.

**Case Report:** A 7 yrs boy was referred to our hospital with some short attack of dyspnea especially exertional. He did not mention any past history of asthmatic attack or recurrent cough of chronic lung disease. But he had past history of hepatoblastoma that was treated by chemotherapy 2 years ago. In physical examination he had fine crackles in two lungs. he didn't have splenomegaly or hepatomegaly. Diffuse infiltrative pulmonary lesion was detected. *By chest X ray. In lab data:* He had significant eosinophilia (WBC 30000, eso 80%) In several time. Work up for any parasitic infection was negative. With regard to past history of chemotherapy, second malignancy was suggested, But BMA & B was normal. And for R/out myeloproliferative HES, FIP1L1/ PDGFRA mutation was negative. But workup showed IgE > 6000 Iu/cc. Specific IgE to aspergillosis was highly positive in CAP study and prick test. Specific IgG to aspergillosis was positive too. Detailed history taking was discovered that patient had some night cough and dyspnea when he went to north and prick test to mite was highly positive. Patient was treated with prednisolone and itraconazole and after two months WBC was returned to normal (WBC -10000, Eso- 6%) and he had not have any cough and dyspnea.

**Conclusion:** ABPA must be considered in any patient with HES and full approach for previous Asthma or CF is necessary.

### **Diagnosis and treatment of a primary immunodeficiency disease**

*Aghamohammadi, A., MD; Department of Pediatrics, Tehran University of Medical Sciences*

Primary Immunodeficiencies (PIDs) are genetic diseases resulting in an increased rate of infection, allergy, autoimmune disorders, and cancer in affected individual. The main forms of PID are combined immunodeficiency, antibody deficiencies, and defects of innate immunity which are characterized by different susceptibilities to pathogens. Making a correct diagnosis of a primary immunodeficiency disease is crucial for the selection of proper therapy. Medical history (age at onset, severity of infections, type and location of infection) also past medical history, family history and physical examination provide important insights into the possible underlying mechanisms of immunodeficiency and differential diagnosis of PID. Initially, diagnosis of PID should be established by screening the immune system and by ruling out anatomic abnormalities and secondary immunodeficiencies. Clinical immunologic laboratory

tests are very important to validate the suspicion of PID. Lymphopenia and marked reduction of T-lymphocyte counts in particular, is a hallmark of severe combined immunodeficiency (SCID). Assessment of T-cell receptor excision circles (TRECs) Levels by means of PCR has been proposed for newborn screening of SCID. Levels of TRECs in circulating lymphocytes are particularly high in newborns and infants. No TRECs are detected in infants with SCID. Measurement of total immunoglobulins, assessment of specific antibodies to both protein and polysaccharide antigens and enumeration of B lymphocytes (CD19 or CD20) are used in screening of antibody deficiency. Diagnosis of different type of phagocytes disorders are made by absolute neutrophil count (ANC), evaluation of dihydrorhodamine-123 (DHR-123) oxidation, (by means of flow cytometric) and evaluation of CD18 expression on the surface of leukocytes. Measurement of hemolytic activity of the classical (CH50) and alternative (AP50) pathways of complement, as well as of C3 and C4 levels, might guide in the diagnosis of complement deficiencies. During last decades, significant advances have been made in the treatment of PIDs based on the use of immunoglobulin replacement therapy, hematopoietic cell transplantation from unrelated donors and cord blood, and gene therapy. Infants with SCID require prompt intervention with use of cotrimoxazole to prevent *Pneumocystis* infection, prophylactic use of antifungal drugs, immunoglobulin replacement therapy, and aggressive treatment of any infectious episodes. HLA-matched sibling donors remain the ideal option, however, advances in living donor unrelated HSCT and banked umbilical cord blood grafts provide hope for all children with severe PID. Treatment of antibody deficiency is based on immunoglobulin replacement therapy. This can be performed by use of intravenous immunoglobulin [IVIG] and subcutaneous immunoglobulin [SCIG]). Both are effective in reducing the incidence of infections. The usual dose of IVIG is 400 mg/kg per 21 days, but higher doses might be needed in patients with bronchiectasis or enteroviral meningoencephalitis. This article highlights on treatment of more common and severe forms of PID.

### **Hematopoietic cell transplantation in the treatment of primary immunodeficiency disorders**

*Friedrich, W., MD; Department of Pediatrics, University of Ulm, Germany*

A major proportion of genetic immunodeficiencies in children has a dismal prognosis unless a functional immune system can be established. Transplantation of hematopoietic precursor cells offers this possibility, and during the last 20 years the practical use of this curative approach has been markedly advanced. While initially restricted to exceptional patients with available

histocompatible family donors, its applicability now is much broader since histocompatible unrelated donors can frequently be identified. Furthermore, the use of HLA-nonidentical family donors such as parents has become an alternative, because severe GvHD as a main complication has become preventable using purified precursor cells purged of GvHD inducing donor T-cells. In this presentation, current experience of transplant strategies in various Primary Immunodeficiencies will be reviewed, as based on studies performed by centers of the European Society of Immunodeficiencies and by our own group, and main risks and complications will be discussed. Also current controversies including optimal timing of the treatment and the rationale of intensive pretransplant conditioning will be addressed. While straight forward as a therapeutical principle, hematopoietic cell transplantation still remains a sophisticated treatment strategy.

### **Primary immune deficiencies presenting in adults**

*Mansouri, D, MD; Division of Infectious Diseases & Clinical Immunology, National Research Institute of Tuberculosis & Lung Diseases, Shahid Beheshti University of Medical Sciences*

Primary immune deficiency diseases (PIDs) are genetically determined defects of immune system function resulting in increased susceptibility to infection. Although relatively rare, PIDs often raise diagnostic and treatment dilemmas and impose a significant burden on the health care system. As a group, the PIDs have provided invaluable insight into the fundamental workings of the human immune system, and more than 100 PIDs are now recognized. A commonly held misconception is that PIDs occur solely in the pediatric age group and hence, only pediatricians are involved in their management. It has become increasingly clear that many patients with PIDs survive into adulthood and fall under the care of physicians responsible for the management of adult patients. A range of factors result in internists being faced with the responsibility of managing PIDs. Firstly, some PIDs tend to present after adolescence, including a range of antibody disorders such as common variable immune deficiency (CVID), selective IgA deficiency, and IgG subclass deficiency. Secondly, some patients with PIDs have a mild phenotype delaying presentation into adulthood. Examples of PIDs presenting late with atypical or attenuated phenotypes include adenosine deaminase (ADA) deficiency, Wiskott-Aldrich syndrome, and chronic granulomatous disease (CGD). Thirdly, significant advances in treatment of PIDs involving utilization of carefully selected antimicrobials, immunoglobulin replacement therapy, and curative modalities such as bone marrow transplantation and gene therapy, now

ensure that many patients with PIDs live into adulthood and graduate from care provided by pediatricians. Finally, and of great concern is that some patients with PIDs present in childhood with recurrent infections, autoimmune disorders, and unusual malignancies but the underlying PID passes unrecognized for many years. Although the importance of recognizing PIDs in adult patients is often the focus of review articles, primary data on this subject is sparse and is generally limited to case reports. In this study we report 78 cases of previously unrecognized PIDs diagnosed in adults in Iran during a 11-year period. This series provides unique data regarding PIDs presenting in adulthood and serves as a timely reminder that physicians, in Iran and elsewhere, must consider the diagnosis of PIDs in their adult patients. Moreover, the often-significant delay between first manifestation of immune deficiency and diagnosis of PID in many of these cases offers a window into understanding the natural history of these rare conditions.

### Clinical phenotypes of primary immunodeficiencies

*Rezaei, N., MD, Molecular Immunology Research Center; and Department of Immunology, School of Medicine, Tehran University of Medical Sciences, Tehran, Iran.. Research Group for Immunodeficiencies, Pediatrics Center of Excellence, Children's Medical Center, Tehran University of Medical Sciences*

Primary immunodeficiency diseases (PID) are a heterogeneous group of inherited disorders with defects in one or more components of the immune system. The patients with PID suffer from a wide range of clinical manifestations, including recurrent infections, autoimmune diseases, and cancers. Recurrent infections, or infection with an opportunistic organism, are the most commonly recognized associations with PID. Patients with predominantly antibody deficiencies and combined immunodeficiencies are often associated with recurrent respiratory tract infections and gastrointestinal manifestations, whilst patients with phagocytes defects can also suffer from recurrent cutaneous and deep abscesses. Meningococcal infections could be associated with complement deficiencies. Autoimmunity is surprisingly common in patients with PID. Idiopathic thrombocytopenic purpura, autoimmune hemolytic anemia, systemic lupus erythematosus, juvenile arthritis, sclerosing cholangitis, and vasculitis are some autoimmune diseases that can be seen in PID. An increased susceptibility to malignancies is also common in some PID. PID are not often apparent by casual inspection. Certain patients may have dysmorphic features and there can be unique physical findings in some cases. Although clinical phenotypes of patients with PID can suggest a clinical diagnosis, definite diagnosis of

patients often rests on the identification of critical diagnostic laboratory features.

### Updates on Autoimmune lymphoproliferative syndrome (ALPS) and a case report

*Cheraghi, T., Jafroodi M, Almasi Aghdam, M., Achaea pour, M.; Department of Pediatrics, 17<sup>th</sup> shahrivar Hospital, Guilan University of Medical Sciences, Iranian Blood Transfusion Organization*

Autoimmune lymphoproliferative syndrome is a non-malignant lymphoproliferation that is characterized with lymphadenopathy, Hepatosplenomegaly, autoimmunity, and malignancy. Lymphadenopathy is chronic and lasts more than 6 months, Hepatosplenomegaly is associated with or without hypersplenism, autoimmunity is mostly directed against intravascular antigens including platelets, RBCs, and neutrophils and producing single lineage or multilineage cytopenias. These patients are primarily at risk to malignancy primarily Hodgkin's disease and non-Hodgkin lymphoma. The basic defect is thought to be dysfunctional apoptosis due to mutation in FAS (CD95), as a death receptor, its ligand, (FAS-Ligand), caspase10, caspase 8, and unknown genes. Abnormal persistence of unapoptotic cells lead to elevated TCRalpha/beta+CD3+CD4-CD8- double-negative T cells that is the most consistent finding in this syndrome. However although the defect in FAS-mediated apoptosis has been described in most patients, this is not the consistent finding and some patients with criteria compatible with ALPS have no defect in apoptosis. There are many diseases that may mimic the ALPS manifestations including lymphoma, infectious mononucleosis, Common variable immunodeficiency disease (CVID, Several non-X-linked forms of hyper IgM syndrome, X-linked lymphoproliferative syndrome (XLP), Wiskott-Aldrich syndrome (WAS), Evans syndrome, and Kikuchi-Fujimoto disease *Natural history and prognosis of ALPS is guarded, and there remains a lot to be elucidated.* Understanding the phenomenon occurring in ALPS may shed light on tolerance and mechanisms of autoimmune diseases. Here we report a case with chronic lymphadenopathy, and Hepatosplenomegaly. CBC ESR, CRP, malignancy survey including bone marrow aspiration and lymph node excisional biopsy and IHC staining has not shown any evidence of malignant cells. Immune screening study was ok. EBV-IgM was normal. HIV test was negative. Flowcytometry showed elevated TCR  $\hat{I}\pm\hat{I}^2+$  (CD3+, CD4 $\hat{\wedge}$ /CD8 $\hat{\wedge}$ ) double negative T cells.

**Key words:** Autoimmune lymphoproliferative syndrome, double-negative T cells, FAS

### Oral and dental health in iranian patients with primary antibody deficiency disorder

*Meighani, Gh., Aghamohammadi, A., Javanbakht, H., Abolhassani, H., Jafari, SM., Rezaei, N.; Growth & Development Research Center, Children's Medical Center, Tehran University of Medical Sciences*

Primary antibody deficiency (PAD) is a disorder of immune system associated with decreased values of secretory and protective immunoglobulins. Because of the important role of immunoglobulins in the protection of oral cavity, PAD patients were more susceptible to caries or oral manifestations. The objective of the present study was to investigate the oral and dental manifestations of PAD patients whom referred to Children Medical Center comparing with their matched healthy controls regarding age and gender. Materials and Methods: In this cross-sectional study, 33 PAD patients and 66 healthy matched controls (2 controls for each patient) were examined with standard techniques regarding oral and dental manifestations together with the number of decayed, missed and filled teeth (DMFT). Results: Twenty one patients were diagnosis as CVID, 8 patients had XLA and 4 patients had HIGM. Aphthous was most frequent disorder in PAD patients (38.7% of cases) which significantly differ with controls manifest with aphthous (16.7%; P value=0.033). Moreover cases had significant higher presentation of oral and dental manifestation indeed herpes sores, candidiasis tonsillitis, gingivitis, calculus, enamel hypoplasia and other ulcerations. The mean dmft scores were  $6.15 \pm 3.6$  and  $1.93 \pm 0.4$  in PAD patients and controls respectively (p value< 0.001). The mean DMFT values were not significantly different between various types of PAD, however CVID patients had higher means of DMFT. Conclusion: The study showed significant differences of oral and dental manifestation between PAD and control patients. therefore, CVID involvement increases the risk of oral lesions' incidence and caries development compared because of lower immunoglobulin levels and higher rate of oral and dental infections.

**Keywords:** Common variable immunodeficiency, X linked Agamaglobulinemia, Hyper IgM, Oral and dental manifestations, Immunoglobulin

### Primary Immunodeficiency diseases associated with increased susceptibility to viral infections and malignancies

*Hedayat, M., Rezaei N., Research Group for Immunodeficiencies, Pediatrics Center of Excellence, Children's Medical Center, Tehran University of Medical Sciences; Molecular Immunology Research Center; and Department of Immunology, School of Medicine, Tehran University of Medical Sciences*

Primary immunodeficiencies (PIDs) are commonly characterized by increased susceptibility to infections, often associated with autoimmune disorders and malignancies. The emergence of malignancies in a heterogeneous group of PIDs, namely X-linked lymphoproliferative disease, IL-2-inducible T-cell kinase deficiency, epidermodysplasia verruciformis, WHIM syndrome, autosomal recessive hyper-IgE syndrome, and autosomal recessive signal transducers and activators of transcription 1 deficiency, results at least partly from the interplay between the underlying genetic defect, immune dysregulation, and increased susceptibility to viral infection. However, the interaction between heritable and environmental factors associated with enhanced tumor susceptibility has not yet been fully elucidated in a variety of human PIDs, including purine nucleoside phosphorylase deficiency, immunoglobulin class switch recombination deficiencies due to defects in CD40-mediated signaling, idiopathic CD4<sup>+</sup> lymphocytopenia, selective IgA deficiency, X-linked agammaglobulinemia, Wiskott-Aldrich syndrome, and cartilage hair hypoplasia, all of which are characterized by marked vulnerability to viral infections. Hence, to find the missing links in the development of malignant tumors, it is of utmost importance to gain in-depth insights into the fundamental molecular nature of the diseases.

**Key words:** Primary immunodeficiency disease, viral infection, malignancy.

### X-Linked lymphoproliferative syndrome: a genetic condition typified by the triad of infection, immunodeficiency and lymphoma

*Mahmoudi, E., Rezaei, N., Aghamohammadi, A.; Growth and Development Research Center, Children's Medical Center, Tehran University of Medical Sciences*

X-linked lymphoproliferative disease (XLP) is a rare primary immunodeficiency disorder characterized by dysgammaglobulinemia, increased susceptibility to EBV infection and lymphoma. While the majority of cases are caused by germline mutations in the SH2D1A gene, which encodes the adaptor molecule SAP, a subset of patients carries mutations in the XIAP gene, which encodes the X-linked inhibitor of apoptosis protein. Studies of immune cells from SAP- or XIAP-deficient patients reveal impairments in numerous functional processes, including NK and CD8<sup>+</sup> T cell cytotoxicity, CD4<sup>+</sup> T cell cytokine production, CD4<sup>+</sup> T cell help to B cells and NKT cell development. Therefore, the phenotypes observed in XLP likely result from the disruption of numerous cellular processes. By dissecting the roles of SAP and XIAP in normal individuals and XLP patients, we will enhance our understanding of immune cell biology and

demonstrate how genetic defects in these molecules predispose patients to infection as well as malignancy.

**KeyWords:** Immunodeficiency, Infection, Malignancy, X-linked, Lymphoproliferative syndrome

### **A Case report of probable Gamma Interferon Deficiency in a 6 month baby girl admitted to Imam Khomeini Hospital of Arak**

*Alimohammadi, D., Tavallae, SE., Social Security Organization*

Primary immunodeficiency (PIs) contains a wide range of syndromes and disabilities along side variety of clinical symptoms and laboratory findings. allergies, auto immunity and acquired immunodeficiency are the most important troubles. Out break of recurrent bacterial infections of inferior respiratory tract which appears mostly among children, is the most common clinical feature. The central nervous system complications due to repetitious infection is also remarkable. Infants and children however have a normal growth, but are potentially in danger because of recurrent infection. Certainly, defense system of human body has a multi structure of different physical and cellular elements to decline these side effects. This report presents the clinical profile of a 6 month female baby, showing rare spread BCG related infection. clinical and laboratory studies (both in Arak and Tehran) discuss a possibility of Gamma Interferon Deficiency.

**Keywords:** Immunodeficiency, Defense system, BCGitis

### **Hyper IgM syndrome: a case report**

*Pourmand, N., Rahaei, M., Godarzi, V., Pourmand, Sh., Atarod, L.; Valiasr Hospital, Tehran University of Medical Sciences*

Hyper-IgM Syndrome (HIM) is a rare genetic (primary) immunodeficiency disorder that is typically inherited as an X-linked recessive genetic trait. patients with XHIGM have markedly reduced levels of IgG, IgA, and IgE but have normal or elevated levels of IgM. Patients with this form may have severe neutropenia and often present with recurring upper and lower respiratory infections within the first year of life. Treatment is IV immune globulin 400 mg/kg once/mo. For the X-linked form, granulocyte colony-stimulating factor is also given as needed for neutropenia. In this report we present a 13 year-old boy of CD40 ligand deficiency who suffered from chronic, severe neutropenia, osteomyelitis. Administration of IVIG was

started for him when the diagnosis was made at the age of 1.5 years and he was on the regular IVIG therapy after that until now for a period of 7 years. Treatment with IVIG regularly in patient who suffer from Hyper-IgM syndrome and neutropenia in CD40 ligand deficiency seems have an excellent clinical response and better quality of life.

**Keywords:**Hyper-IgM Syndrome, severe neutropenia, IVIG, osteomyelitis

### **Comparison of pulmonary diseases in common variable immunodeficiency and X-linked agammaglobulinaemia**

*Aghamohammadi, A., Allahverdi, A., Abolhassani, H., Moazzami, K., Alizadeh, H., Gharagozlou, M., Kalantari, N., Sajedi, V., Shafiei, A., Parvaneh, N., Mohammadpour, M., Karimi, N., Salehi-Sadaghiani, M., Rezaei, N.; Department Of Pediatrics, Children's Medical Center Tehran University of Medical Sciences*

Pulmonary disease is the most common complication in patients with common variable immunodeficiency (CVID) or X-linked agammaglobulinaemia (XLA). Pulmonary disease may progress despite immunoglobulin replacement therapy. In this study pulmonary complications were compared in patients with CVID or XLA.

Pulmonary complications were evaluated in 115 patients (76 with CVID and 39 with XLA) by reviewing hospital records of chest infections, pulmonary function tests and high-resolution CT scans. Thirty-two patients with XLA (82%) presented with 59 episodes of pneumonia before diagnosis, whereas 15 patients (38.4%) experienced pneumonia after immunoglobulin replacement therapy (1.67 vs 0.45 episodes per patient per year). Among the CVID patients, 196 episodes of pneumonia were documented in 59 patients (77.6%) before diagnosis, while 36 patients (47.3%) experienced pneumonia after therapy (1.11 vs 0.58 episodes of pneumonia per patient per year). Forty-seven (41%) patients (38 with CVID and 9 with XLA) developed chronic lung disease. The CVID patients developed more complications, including bronchiectasis and lymphoid interstitial pneumonitis, than the XLA patients.

Patients with CVID had a greater likelihood of developing lung disease, possibly due to delayed diagnosis and immune dysregulation, as compared with XLA patients. Early diagnosis of patients with primary antibody deficiencies and adequate i.v. immunoglobulin replacement therapy substantially reduces the number of pulmonary infections. However, CVID patients are prone to progression of lung disease despite optimal immunoglobulin therapy because of the nature of the disease. This important issue should be addressed in further studies.

**Keywords:** common variable immunodeficiency; lung complication; pneumonia; pulmonary function test; X-linked agammaglobulinemia

### Comparison of students' and their parents' responses to an asthma questionnaire

*Modaresi, MR., Golshan, M., Khanlarpour, A., Iranpour, R.; Pulmonary Pediatrics Department, Al-zahra Hospital, Isfahan University of Medical Sciences*

Asthma is one of the most common chronic diseases during childhood. Unfortunately little definite epidemiologic data is available concerning Iranian children.

To determine the degree of coordination to responses to an asthma symptom questionnaire completed by students and their parents in Isfahan junior high schools.

A questionnaire based on the international study of asthma and allergy in childhood (ISSAC) was given to 2484 students and their parents during 1998-1999. The answers were compared and analyzed by the McNemar X<sup>2</sup>-test.

According to the findings by wheeze, cough after exercise, and history of asthma based in student and parental questionnaires were 37.8%, 48.6%, 13.8%, 66.2%, 6.1% and 27.6%, 22.8%, 13.5%, 16.2%, 20.8%, 8.2%, 13.8%, 66.2%, agreement between parents and students' answers werenot related to sex, parental educational level and history of smoking of parents. Conclusion : Evaluation of both students' and their parents' answers is recommended to determine exact prevalence rate of asthma.

**Keywords:** Asthma, Epidemiology, Student

### Health Policy Making for Common Variable Immunodeficiency: The Burden of the Disease

*Abolhassani, H., Aghamohammadi, A., Abolhassani, F., Eftekhar, H., Rezaei, N.; Growth and Development Research Center; Department of Pediatrics, Pediatrics Center of Excellence, Children's Medical Center, Tehran University of Medical Sciences; National Institute of Health Research, Tehran University of Medical Sciences*

Common Variable Immunodeficiency (CVID) is a primary immunodeficiency disease, characterized by chronic and recurrent infections and increased susceptibility to autoimmune disorders and malignancies. The objective of this study was to estimate the burden of CVID in Iran in the period 1985-2008 based on incidence, mortality, and disability adjusted life years (DALY).

In this study, the methods developed by the World Health Organization for National Burden of Disease (NBD) studies were applied to estimate disease and

injury incidence for the calculation of Years of Life Lost due to premature mortality (YLL), Years Lived with Disability (YLD), and DALYs.

The average age-adjusted CVID epidemiological rates in this period were 1/200,000 in incidence and 1/91000 in prevalence. The burden of CVID (DALY) was 25.21/100000 (17.86/100000 for YLL and 7.35/100000 for YLD). The DALY showed significant increased tendency in the patients with age range of 5 to 14 years as well as in whom with polyclonal lymphocytic infiltration phenotypes ( $p < 0.001$ ).

Based on measurement of DALY of disease in CVID patients, reduction of immature death in polyclonal lymphocytic infiltration phenotype group and reduction in episodes of infections in patients with infectious only phenotype and appropriate management with regular intravenous immunoglobulin is the most important way to decrease the burden of CVID.

**Keywords:** Common Variable Immunodeficiency, Disability adjusted years, Years of life lost, Years lived with disability

### Sunshine Hypothesis

*Cheraghi, T., MD; Guilan University of Medical Sciences*

Sunshine Hypothesis Why asthma and allergic diseases are so progressively increasing worldwide? Many efforts has been made to explain this pattern. The 1st attempt was the introduction of "hygiene hypothesis". according to this theory; as exposure to infectious agents decreases as a result of clean lifestyle, antibiotic use, vaccination, pasteurization of milk and other foods, smaller family size, exposure of immune system to infections decreases; this deviates the immune system away from balanced TH1-TH2 to dominant TH2 and asthma and atopy that are TH2 – dependent diseases are increasing. Since the introduction of this hypothesis many questions have raised that cannot be explained ; such as asthma in inner city crowded families or obese patients, It also cannot explain why simultaneous TH1-dependent autoimmune diseases are increasing. Therefore we need an alternative hypothesis that could explain the increasing prevalence of both TH1 and TH2 – dependent Diseases. Litonjua and Weiss proposed the "sunshine hypothesis ". They hypothesize that "as populations grow more prosperous and more westernized, more time is spent indoors and there is less exposure to sunlight, leading to vitamin D deficiency, subsequently resulting in more asthma and allergy" Countries like England, Australia, and newzealand that are far from equator have the highest rate of asthma in the world. Studies have been performed at the cellular and molecular Level and show that vitamin D has receptor on many cells including T cells, activated B cells, dendritic cells, regulatory T cells, and respiratory epithelial cells. It has been shown that vitamin D induces the regulatory T cells; these cells have regulatory effects on both TH1 and TH2

cells and thereby prevent them from unabated activation. Thus vitamin D functions as a hormone. This effect explains why in vitamin D deficiency states both TH1 and TH2 dependent diseases has been so prevalent in recent decades. Vitamin D deficiency may explain at least a portion of asthma epidemic. It may play a role in fetal lung maturation, prevention of wheezing in early life, and reversal of steroid resistant in asthma.

**Keywords:** Vitamin D deficiency, asthma

### **Hyper-immunoglobulin M Syndrome Immunoglobulin Profile In Three Patients With Ataxia-Telangiectasia**

*Aghamohammadi, A., MD, Nasiri Kalmarzi, R., MD, Rezaei, N., MD; Pediatrics Center of Excellence, Children's Medical Center, Tehran University of Medical Sciences*

Ataxia-Telangiectasia (AT) and hyper-immunoglobulin M (HIGM) syndrome are both primary immunodeficiency diseases caused by different genetic defects. While the majority of patients with AT have decreased immunoglobulin levels, a small proportion of AT patients have increased serum immunoglobulin (Ig)M concentrations during the course of disease, a high level of IgM at onset is rare. This suggests that ATM mutation not only results in DNA repair defect but also immunoglobulin class switch defect.

**Case report:** we report 3 girls with recurrent sinopulmonary and otic infections from early infancy. The median age was 6(3-10) years. They were frequently admitted to hospital due to lower respiratory infections. All of the patients had hepatosplenomegaly and neurologic disturbance (gait defect) and oculocutaneous lesions (telangiectasia). Median levels of immunoglobulins were respectively: 8 for IgA, 0.7 for IgE and 504 for IgM. Median level of  $\alpha$ FP was 137(75-211). All patients

Were evaluated for Immunoglobulin class switching.....

AT could be classified as type 4 HIGM syndrome in which class switching defects are postulated to be in the DNA repair machinery. AT should be taken into account in the follow-up of autosomal recessive HIGM.

**Keywords:** Ataxia-Telangiectasia, IgM, Hyper-IgM syndrome

### **Hyper-IgM syndrome (HIGM)**

*Kashef, S., MD, Department of Immunology and Allergy, Shiraz University of Medical Sciences*

Hyper-IgM syndrome (HIGM), also called "immune deficiency with normal or elevated IgM", is a term denoting deficiency of IgG, IgA, and IgE, with normal

or elevated serum concentrations of IgM. Several gene defects have been grouped together under this eponym. They can be classified as defects of signalling through CD40 causing both a humoral immunodeficiency and a susceptibility to opportunistic infections or intrinsic defects in B cells of the mechanism of immunoglobulin class switch recombination (CSR) resulting in a pure humoral immunodeficiency. The first described, and most common defect, was X-linked hyper IgM syndrome due to deficiency of CD40 ligand (CD40L, also called CD154). This disorder and an autosomal recessive form due to deficiency of CD40 have associated abnormalities of cellular immunity. Another disorder, termed NEMO deficiency, is also sometimes classified as part of the HIGM syndromes.

Two additional rare forms of hyper-IgM syndrome, with autosomal recessive inheritance and normal T cell function, have been defined. These are caused by mutations in nucleic acid modifying enzymes called activation induced cytidine deaminase (AICDA or AID) and uracil nucleoside glycosylase (UNG). These enzymes are required for the process of immunoglobulin class-switching.

Recurrent and severe sinopulmonary infections, gastrointestinal infections (including Giardiasis), bacterial meningitis, viral encephalitis, and severe hepatitis B infections have also been observed. Lymphoid hyperplasia is seen in about two-thirds of patients. This may involve peripheral nodes, as well as mesenteric nodes, tonsils, liver and spleen. A variety of autoimmune pathologies have also been observed in these patients including diabetes mellitus, autoimmune hepatitis, rheumatoid arthritis, inflammatory bowel disease, and uveitis. Lymphoid malignancy is another complication in this syndrome.

Patients with AICDA and UNG deficiencies generally exhibit excellent responses to gamma globulin replacement therapy with reduction in bacterial infections. CD40 signalling defects may require corrective therapy with bone marrow transplantation.

### **Allergic rhinitis**

*Gharagozlou, M., MD, Allergy & Immunology Dept. Children's Medical Center, Tehran University of Medical Sciences*

Allergic rhinitis is a symptomatic disorder of the nose induced after allergen exposure by an IgE-mediated inflammation of the membranes lining the nose. It is a global health problem that causes major illness and disability worldwide. Over 600 million patients from all countries, all ethnic groups and all ages suffer from allergic rhinitis. It affects social life, sleep, school and work and its economic impact is substantial. Risk factors for allergic rhinitis are well identified. Indoor and outdoor allergens as well as occupational agents cause rhinitis and other allergic diseases. The role of indoor and outdoor pollution is probably very important, but has yet to be fully understood both for

the occurrence of the disease and its manifestations. In 1999, during the allergic rhinitis and its impact on asthma (ARIA) WHO workshop, the expert panel proposed a new classification for allergic rhinitis which was subdivided into 'intermittent' or 'persistent' disease. This classification is now validated. The diagnosis of allergic rhinitis is often quite easy, but in some cases it may cause problems and many patients are still under-diagnosed, often because they do not perceive the symptoms of rhinitis as a disease impairing their social life, school and work. The management of allergic rhinitis is well established and the ARIA expert panel based its recommendations on evidence using an extensive review of the literature available up to December 1999. The statements of evidence for the development of these guidelines followed WHO rules and were based on those of Shekelle et al. A large number of papers have been published since 2000 and are extensively reviewed in the 2008 update using the same evidence-based system. Recommendations for the management of allergic rhinitis are similar in both the ARIA workshop report and the 2008 update. In the future, the GRADE approach will be used, but it not yet available. Another important aspect of the ARIA guidelines was to consider co-morbidities. Both allergic rhinitis and asthma are systemic inflammatory conditions and often co-exist in the same patients. In the 2008 update, these links have been confirmed. The ARIA document is not intended to be a standard of care document for individual countries. It is provided as a basis for physicians, health care professionals and organizations involved in the treatment of allergic rhinitis and asthma in various countries to facilitate the development of relevant local standard-of-care documents for patients.

#### **Allergic rhinitis recommendations:**

1-Allergic rhinitis is a major chronic respiratory disease due to its: -prevalence -impact on quality-of-life-impact on work/school performance and productivity - economic burden -links with asthma 2-In addition allergic rhinitis is associated with co-morbidities such as conjunctivitis. 3-Allergic rhinitis should be considered as a risk factor for asthma along with other known risk factors. 4-a new subdivision of allergic rhinitis has been proposed: -intermittent(IAR) -persistent (PER) 5-the severity of allergic rhinitis has been classified as 'mild' moderate/ severe' depending on the severity of symptoms and quality-of-life outcomes. 6-Depending on the subdivision and severity of allergic rhinitis, a stepwise therapeutic approach has been proposed. 7-the treatment of allergic rhinitis combines: -Pharmacotherapy -Immunotherapy -Education 8-Patients with persistent allergic rhinitis should be evaluated for asthma by means of a medical history, chest examination, and, if possible and when necessary, the assessment of airflow obstruction before and after bronchodilator.

9-Patients with asthma should be appropriately evaluated (history and physical examination) for rhinitis. 10-Ideally, a combined strategy should be used

to treat the upper and lower airway diseases to optimize efficacy and safety.

### **Antibody Responses to Vaccination with Pneumococcal Polysaccharide Vaccine in Splenectomized Patients with Idiopathic Thrombocytopenic Purpura or Trauma**

**Mahmoodi, M., PhD, Ghaderi, H., Aghamohammadi, A., MD, Rezaei, N., MD, Isaean, A., PhD, Zargar, M., MD, Khaji, A., 1. Cancer Research Center, Tehran University of Medical Sciences; 2. Research Group for Immunodeficiencies, Pediatrics Center of Excellence, Children's Medical Center, Tehran University of Medical Sciences; 3. Growth and Development Research Center, Tehran University of Medical Sciences; 4. Molecular Immunology Research Center; and Department of Immunology, School of Medicine, Tehran University of Medical Sciences; 5. Sina Trauma & Surgery Research Center**

**Background:** An increased risk of invasive infections with encapsulated bacteria such as *Streptococcus pneumoniae* has been described among splenectomized patients. Pneumococcal vaccination has been recommended in these patients. In this study, the serum antibody response of splenectomized patients with idiopathic thrombocytopenic purpura (ITP) or trauma who were immunized with 23-valent pneumococcal polysaccharide vaccine (Pneumovax 23) were evaluated. We also addressed the question of whether an underlying hematological disease may affect the immune response.

**Methods:** Twenty patients with ITP and fifteen cases of trauma along with 40 healthy individuals as controls were vaccinated with PPV23. The serum antibody response (IgG and IgG2) to the Pneumovax antigens was determined by enzyme-linked immunosorbent assay (ELISA) prior to vaccination and 4 weeks post-vaccination. Specific antibodies against whole pneumococcal antigens were measured using Enzyme-Linked Immunosorbent Assay (ELISA) technique.

**Results:** Although there was not any significant difference in pre-vaccination anti pneumococcal IgG levels between these three groups, the mean of post-vaccination IgG titer to the pneumococcal antigens in ITP group was significantly lower than those in trauma group ( $P=0.02$ ) or in healthy control group ( $P=0.001$ ). No significant differences in antibody titer increase were found between trauma group and healthy control group ( $P>0.05$ ). Response to immunization was poor in 9 of 20 ITP patients (45%). There was direct association between IgG and IgG2 after vaccination in the studied groups ( $P<0.001$ ,  $R=0.984$ )

**Conclusion:** Patients suffered from ITP who have undergone splenectomy responded poorly to pneumococcal antigens and remained at significant risk for pneumococcal infection. These findings suggest that the immunization policy with pneumococcal

polysaccharides should be evaluated in splenectomized patients and cases with ITP required to be offered other prophylactic methods.

**Keywords:** Splenectomized patients, Pneumococcal polysaccharide vaccine, Idiopathic thrombocytopenic purpura.

### **Atopic dermatitis in children and adults, a general consideration**

*Zandieh, F., Isfahan University of Medical Sciences*

Atopic dermatitis (AD) is a pruritic disease of unknown origin that usually starts in early infancy (an adult-onset variant is recognized); it is characterized by pruritus, eczematous lesions, xerosis (dry skin), and lichenification (thickening of the skin and an increase in skin markings). Atopic dermatitis may be associated with other atopic (immunoglobulin E [IgE]) diseases (eg, asthma, allergic rhinitis, urticaria, acute allergic reactions to foods). Atopic dermatitis has enormous morbidity, and the incidence and prevalence appear to be increasing. Other conditions with different etiologies and prognoses are often grouped under the umbrella of a diagnosis of atopic dermatitis. Good evidence

indicates that genetic factors are important in the development of atopic dermatitis, but the pathophysiology is still poorly understood. Two main hypotheses have been proposed regarding the development of the inflammatory lesions. The first suggests an immune dysfunction resulting in IgE sensitization and a secondary epithelial-barrier disturbance. The second proposes a defect in epithelial cells leading to the defective barrier problem, with the immunological aspects being epiphenomena. The prevalence rate for atopic dermatitis is 10-12% in children and 0.9% in adults. Incessant itch and work loss in adult life is a great financial burden. A number of studies have reported that the financial burden to families and government is similar to that of asthma, arthritis, and diabetes mellitus. In children, the disease causes enormous psychological burden to families and loss of school days. Mortality due to atopic dermatitis is unusual. Treatment is based on several therapeutic modalities: - Moisturization - Topical steroids - Immunomodulator. - others: UV-A, UV-B, pre or probiotics, nonconventional therapies

With increase age most patients improve; this can occur at any age. While the frequency of atopic dermatitis is as high as 20% in childhood it is 0.9% in adults. One third of patients develop allergic rhinitis. One third of patients develop asthma.

## Infectious Diseases & Vaccination Abstracts

### Blood cell count characteristics of H1N1-positive pediatrics patients in Zahedan-Iran

*Soleimani, Gh., MD, Akbarpour, M., MD, Soleimani A., MD, Parsa, M., MD, Research Center for Children and Adolescents Health, Zahedan University of medical sciences, Ali ebne abitaleb Hospital, Zahedan Iran*

**Background** In influenza A (H1N1) infection, that spread globally since the spring of 2009, relative lymphopenia without leukopenia was observed in some studies.

**Methods:** This study is a prospective observational study of children requiring hospitalization for confirmed or suspected positive H1N1 infection between September and October 2009 to describe blood cell count characteristics of H1N1-positive pediatrics patients in Zahedan-Iran with 2009 influenza A (H1N1) infection. For all patients the throat swab set up for PCR testing to confirm or exclude the diagnosis of H1N1 Influenza A. Age, sex, laboratory data, outcome were documented.

**Results:** Thirteen patients was H1N1 positive (61.5% male versus 38.5% female). Mean age of patients was  $70.31 \pm 53.39$  months. The median white blood cell count (WBC) was 7092.31 cells/mm<sup>3</sup> (range: 2600-17900 cells/mm<sup>3</sup>; normal: 4000–10000 cells/mm<sup>3</sup>). The mean lymphocyte count was 37.28% (5-58%). Five patients had WBC less than 4000 (38.8%). Only 2 patients had leukocytosis due to superinfection that required intensive care support and both of them expired. The mean platelet count was 185920 cells/mm<sup>3</sup> (range: 70000-329000 cells/mm<sup>3</sup>; normal: 150000-450000 cells/mm<sup>3</sup>). Six patients had platelet less than 150000 (46.2%). All patients were followed 3 weeks after discharge and their cell counts were normal.

**Conclusion:** During the evaluation period of our study, 2009 H1N1 influenza caused leukopenia and thrombocytopenia.

**Key words:** H1N1, leukopenia, thrombocytopenia

### Comparison of high dose versus usual dose of rectal acetaminophen in the treatment of febrile children

*Hashemian, H., MD, Momtazbakhsh, M., MD, Department of Pediatrics, Guilan University of Medical Sciences*

**Background:** The antipyretic effectiveness of high dose versus usual dose of rectal acetaminophen is not

well established. This study is designed to compare the antipyretic effectiveness of high dose (30 mg/kg) rectal acetaminophen with the standard rectal dose (15mg/kg) in febrile children.

**Materials:** A randomized, controlled trial was performed in 80 patients aged between 6 months and 6 years with rectal fever > 39° Celsius who referred to 17 Shahrivar Hospital of Rasht- IR Iran. Children were excluded if they had taken any antipyretic drug or antibiotics within the previous 8 hours; required antibiotic treatment within the first 3 hours of receiving acetaminophen; had hepatic, renal, or neurological diseases; had a history of hypersensitivity to acetaminophen; had febrile seizures; or had diarrhea during the medical consultation. The patients were randomly assigned in one of two groups: First group (N= 40 cases) received 15 mg/kg and the second group (N= 40 cases) received 30 mg/kg acetaminophen rectally. The body temperature were assessed at the end of the first and third hour and compared between two groups. The data were analyzed by SPSS-14 using Independent Sample T Test, Paired Sample T Test, and Chi-Square Test.

**Results:** One hour after use of rectal acetaminophen, the mean temperature reduction in groups 1 and 2, were  $0.97 \pm 0.59$  °C and  $1.03 \pm 0.62$  °C, respectively (P= 0.663). There was significant difference in mean temperature reduction between the groups 1 and 2, three hour after use of rectal acetaminophen ( $1.22 \pm 0.72$  °C vs.  $1.57 \pm 0.65$  °C, respectively; P= 0.028).

**Conclusion:** Rectal acetaminophen with dose of 30 mg/kg is probably more effective in reducing fever.

**Keywords:** Acetaminophen, Children, Dose, Fever, Rectal

### Incidence of E.coli causing pediatric UTI and its antibiotic resistance in one major children hospital in Tehran.

*Fahimzad, SA, MD, Mofid Hospital*

**Background:** Urinary tract infection (UTI) is one of the most important and common disease in children, that needs immediate diagnosis and appropriate treatment. The aim of this study was to determine incidence of E. coli that cause UTI in pediatrics and its antibiotic susceptibility.

**Methods:** Medical records of 222 children 0-14 years old which admitted to mofid children Hospital with UTI diagnosis in 1387 were studied. Their urine cultures were positive with single bacterial species.

**Results:** E.coli was the most common pathogen (70.3 %), followed by klebsiella (26%), Enterococcus (8.1%), Proteus (2.3%), Enterobacter (2.3%) and some

other bacteria. In this study, Ecoli was resistant to cephalixin (72/8%), piperacillin (70%), cotrimoxazole (69/56%), cefotaxim (63%), cefixime (55/8%), nalidixic acid (45%), ceftazidime (39/7%), norfloxacin (31/6%), kanamycin (26/2%), ceftizoxim (19/3%) gentamycin (14%), ciprofloxacin (12/9%), amikacin (4/9%) and nitrofurantoin (3/5%). Conclusion: In order to high resistance of Ecoli to 3rd generation cephalosporins, especially ceftriaxon, it's better to be cautious to use it for empiric therapy.

**Conclusion:** In this study minimum resistance of E.coli was in nitrofurantoin followed by amikacin, ciprofloxacin and gentamycin. So, for empiric therapy we can add amikacin or gentamycin to 3rd generation cephalosporines for high risk patients like infants.

**Key words:** UTI, children, E coli, antibiotic resistance

### **Correlation of otitis media and Congenital Nasolacrimal Duct Obstruction in children aged 1 month to 6 years old in 22th Bahman Hospital**

*Sobhani, F., Al-Sheikh, A., Sobhani, R., A-Saeidi, S., Najafi, L.; Gonabad University*

**Background:** To determine the otolaryngological disorders associated with congenital nasolacrimal duct obstruction (CNLDO) and their correlation with otitis media.

**Methods:** This case-control study included 40 children with and 40 without CNLDO which aged 1 month to 6 years old in 22th Bahman hospital. Case selection was done through ophthalmologic clinics and for control group through general population or pediatric clinics. Both groups were followed up with routine otorhinolaryngological examination with tympanometry.

**Results:** Incidence of otitis media in case and control groups was 20% and 15% respectively. As the main result no significant difference in incidence of otitis media was mentioned between case and control groups. According to this study there were no significant differences in ophthalmic, external ear, tympanic membrane, mouth and throat examinations between two groups.

**Conclusion:** No statistically significant difference was achieved for otitis media in children with CNLDO and control group.

**Keywords:** congenital nasolacrimal duct obstruction, otitis media, effusion.

### **Recurrent meningitis as a complication of skull base meningocele**

*Sedighi, I., MD, Borzouei, B., MD, Sadrosadat, ST., MD, Ghodsi, M., MD; Department of Pediatrics, Hamedan University of Medical Sciences/ Besat*

*Hospital; Department of Neurosurgery, Tehran University of Medical Sciences*

**Background:** Malformations of the skull base are rare anomalies, but can cause severe complications such as meningitis. They are often detected not until after a history of recurrent meningitis.

**Case report:** A 6-year-old boy was admitted in our hospital due to change in mental status, fever, and vomiting. His past medical history revealed two episodes of bacterial meningitis in 12 and 22 months of age. His parents were cousins and had lost their 3 sons before this boy was born. One of them had died due to respiratory distress 15 days after birth, another one had been born dead, and the last one had died due to meningitis in 3 years of age. He received Vancomycin and Ceftriaxone upon diagnosis of recurrent meningitis. Due to seizure, intravenous Phenytoin was started. Complementary diagnostic evaluations included computed tomography (CT) and magnetic resonance imaging without any abnormal finding. Streptococcus pneumonia was isolated from CSF culture. Serum immunoglobulins, complement C3, C4, and CH50 levels, NBT slide test, and flow cytometry (CD3, CD4, CD8, CD19, and CD16+56) were normal. After ruling out primary immunodeficiency, we repeated CT and this time a skull base bony defect behind sphenoidal sinus with a meningocele was noted. Endonasal surgery confirmed this bony defect and closure was accomplished. No further meningitis has been observed.

**Conclusion:** Congenital skull base defects may be difficult to detect. All children with recurrent meningitis should be investigated for these defects. Early recognition of skull base defects is important to avoid delay of definitive surgical management.

**Keywords:** recurrent meningitis, skull base defects, meningocele

### **Fever on monday evening every week: an atypical PFAPA syndrome case report**

*Borzouei, B., MD, Sedighi, I., MD; Department of Pediatrics, Hamedan University of Medical Sciences/ Besat Hospital*

**Background:** PFAPA (Periodic Fever, Aphthous stomatitis, Pharyngitis and Adenitis) syndrome was first described in 1987 by Marshall et al. Its etiology is unknown. PFAPA is characterized by episodes of high fever lasting 3-6 days, then resolve spontaneously, recurring every 2-8 weeks, and accompanying with cervical adenitis, pharyngitis and aphthous stomatitis. Between the episodes of fever there are no symptoms.

**Case report:** A 6-year-old boy was admitted in our hospital due to periodic fever. His problem began from 101 days ago at the age of 68 months. Episodes of fever were lasting 1 day, and recurring on Monday evening every week. Accompanying signs and symptoms

included pharyngitis, cervical adenitis, malaise, chills, nausea, and bone pain in lower limbs. In physical examination he had red throat and enlarged cervical lymph nodes. Complete blood count revealed 15600 WBCs/mm<sup>3</sup> (Neutrophils=75%), ESR=40mm/h. Because of positive throat culture he received antibiotic. After treatment throat culture was been negative but the patient was suffering from febrile episodes. In bone marrow aspiration myeloid hyperplasia was seen with no evidence for malignancy. After cyclic neutropenia rule out, on supposing PFAPA he received corticosteroid then febrile episode resolved dramatically. He had good growth and development with no any other remarkable health problem during 30 months follow up. The patient required only a single oral dose (1 mg/kg) of prednisilone for each episode. The febrile episodes recurred less frequently at intervals of 2-3 months.

**Conclusion:** If the febrile episodes remain after treatment of bacterial pharyngitis, keep in mind the PFAPA syndrome.

**Keywords:** PFAPA, periodic Fever, pharyngitis

### Sentinel Hospital-Based Surveillance of Rotavirus Diarrhea in Iran

*Keshkar, A., Eesteghamati, A., Gouya, M., Najafi, L., Zali, MR., Sanaei, M., Yaghini, F., El Mohamady, H., Patel, M., Klana, JD., Teleb, N.,*

1. *Department of Preventive Medicine, School of Medicine, Golestan University of Medical Sciences*

2. *Center of Disease Control and Prevention, Ministry of Health and Medical Education*

3. *Research Center of Gastroenterology and Liver Disease, Shahid Beheshti University of Medical Sciences, Tehran,*

4. *Naval Medical Research Unit 3*

5. *Centers for Disease Control and Prevention, Atlanta, Georgia*

6. *Regional Office for the Eastern Mediterranean, World Health Organization, Cairo, Egypt*

**Background:** Rotavirus is the most common causes of severe, acute diarrhea during childhood and is an important cause of morbidity and mortality in developing countries. We established active hospital-based surveillance of childhood diarrhea to assess the scope of severe rotavirus disease in Iran.

**Methods:** From May 2006 through April 2007, prospective surveillance of rotavirus diarrhea among children aged <math>15</math> years was conducted in 5 sentinel hospitals in Iran. Stool samples were tested for rotavirus using a commercially available enzyme immunoassay, and rotavirus-positive samples were genotyped using reverse-transcriptase polymerase chain reaction.

**Results:** Of 2198 children admitted to the hospital for acute gastroenteritis, 1298 (59.1%) had stool samples test positive for rotavirus by enzyme immunoassay. Of the rotavirus episodes, 85% occurred during the first 2

years of life, with the peak prevalence of severe rotavirus disease occurring from September through January. Among the 110 rotavirus-positive samples that were genotyped, G4P[8] was the most commonly detected rotavirus genotype (30.9% of strains). Other commonly detected genotypes included P[8] with G nontypeable (21.8%), G4 with P nontypeable (13.6%), G1[P8] (10.9%), and G2[P4] (5.5%).

**Conclusions:** Rotavirus is the most common cause of severe diarrhea in Iran, which indicates that safe and effective rotavirus vaccination in Iran is a public health priority.

**Keywords:** Sentinel surveillance, Genotyping, Rotavirus, Iran

### Infectious Disease Prevention Program : How Can Children Educate Their Family?

*Sedighi, I., Noori, Sh., Sadrossadat, T., Nemati, R.; Hamedan university of medical sciences*

**Background:** Knowledge Transfer has advanced as one of the priorities in most of the research centers in the world. There are many researchers, who try to find a way for transferring their knowledge to their society. The purpose of this study is to propose an innovative method of knowledge transfer.

**Methods:** After changing seven health topics (about infectious disease prevention) into childish poetries, we selected five kindergartens randomly and taught these poetries to the children. Teaching process was held after a pretest, containing 25 questions that examined 103 of their parents about above-mentioned topics, however those parents who was working in health care system or their education was related to health fields were excluded from study. The same post-test was given after 4 months of teaching process.

**Resultss:** The mean of correct answers to the pretest was 59.8% comparable with 81.6% for post-test (P: 0.00). There was neither significant difference between genders and correct answers nor between knowledge degrees and correct answers. Assuming one's correct answers to the questions as his/her Knowledge Mark, the mean of this variable increased 5.32 by this method.

**Conclusion:** This cost-effective and joyful method had successful results. Children can have an active role in enhancing the whole family's health situation. They sing these poetries repeatedly at home and no one gets tired of their teaching! Learning within family without any obligation makes our innovative scheme a suitable solution for paving the knowledge transferring way. In other words, we tried to overcome the barriers between researchers and society.

**Keywords:** Infectious disease, Knowledge Transfer, poetries, kindergarten

## **A Pregnant patient with a previous pregnancy complicated by group B streptococcal disease in the infant.**

*Khotaei, Gh., MD; Children's Medical Center, Tehran University of Medical Sciences*

The group B streptococcus (GBS, streptococcus agalactiae) is recognized as an important cause of invasive disease in neonates and pregnant women. Among neonates, premature infants are at the greatest risk of adverse outcomes from GBS infection. These premature infants account for 25% of cases of GBS disease among neonates. The disease manifests itself as an early-onset form (<7 days after birth), a late-onset form (7 days to 3 months after birth) and a very late form (>3 months after birth). Disease among infants usually presents as bacteremia, pneumonia and meningitis.

The organism colonizes the gastrointestinal tract of humans, with the genitourinary tract being the most common site for secondary spread. Colonization rates vary widely among different ethnic groups, geographic areas and age groups. These rates generally indicate that 10-30% of pregnant women have vaginal or rectal colonization with GBS. Data from the USA have suggested that prior to the implementation of recommendations for the Prevention of early-onset GBS using maternal intrapartum antimicrobial prophylaxis (IAP) the incidence of GBS neonatal disease was 1-4 cases per 1000 live births. Among these cases, early-onset disease occurred in approximately 1 infant per 100-200 colonized woman and was responsible for 75% of cases among infants. Since the widespread use of IAP the incidence of early-onset GBS disease has decreased significantly (by approximately 80%), resulting in an incidence rate of less than 1 case per 1000 live births.

The incidence of early-onset disease is higher in babies born to women less than 20 years of age and in those who are of black race in the USA. Intrapartum risk factors include premature onset of labor (<37 weeks gestation), prolonged rupture of membranes (>18 hours) and intrapartum fever (>100.4 °F/>38° C). Additional risk factors include heavy vaginal colonization with GBS, previous delivery of an infant who had GBS disease and the presence of low maternal levels of anti-GBS capsular antibody. Women who have GBS bacteriuria are at an increased risk of delivering an infected baby with early-onset disease. This is related in part to the fact that women who have GBS bacteriuria are usually heavily colonized with GBS. Bacteriuria caused by GBS is associated with an increased risk of preterm labor.

Any woman who has lost an infant as a result of GBS disease needs the usual understanding and support given to any woman who has lost an infant during the neonatal period. Intrapartum prophylaxis is recommended regardless of screening cultures because of the previous delivery of a baby who had GBS disease. Routine vaginal-rectal screening for GBS is

not necessary in this setting. However, it would be appropriate to obtain urine cultures at different antenatal visits to determine whether GBS bacteriuria is present, given that these women are at an increased risk of having GBS-affected infant. Penicillin G (5 megaunits intravenously initially followed by 2.5 megaunits intravenously q4h) should be given intrapartum until delivery. Ampicillin (2g intravenously initially and then 1g intravenously q4h until delivery) is an acceptable alternative. Penicillin G is preferred because it has a narrow spectrum and is thus potentially less likely to select out resistant bacteria. The use of IAP in women who are allergic to penicillin takes into account the risk of anaphylaxis and the background prevalence of GBS resistance to erythromycin and clindamycin. Given the relatively high rates of GBS resistance to these two latter agents, women who are allergic to penicillin but who are at low risk of anaphylaxis may receive cefazolin 2g initially then 1g q8h. If the isolate has been shown to be susceptible to clindamycin this agent may be used. Vancomycin should be reserved for women who are at high-risk of anaphylaxis in the setting where susceptibility testing of the GBS isolate has not been performed. Group B streptococci are associated with various complications during pregnancy. These include septic abortion, urinary tract infections, chorioamnionitis, wound infection and endometritis. Although IAP may have a beneficial effect on endometritis, an assessment is necessary in the immediate postpartum period in order to guide further antibiotic therapy directed at the mother. The newborn infant of a mother who has received intrapartum prophylaxis requires a special management approach. The approach outlined in Figure PP25.2 may be used as a guide to the management of the neonate.

## **Crimean-Congo hemorrhagic fever in children**

*Khalili, M., MD, Alavi-Naini, R., MD; Research Center for Infectious Diseases and Tropical Medicine Zahedan University of Medical Sciences  
Research Center for Children and Adolescents Health, Zahedan University of Medical Sciences*

**Background:** Crimean-Congo hemorrhagic fever (CCHF) is a tick-borne disease caused by the arbovirus which is a member of the Nairovirus genus. After a short incubation period, CCHF is characterized by a sudden onset of high fever, chills, headache, myalgia and in severe cases, hemorrhagic manifestations. In this study, the epidemiological, clinical and laboratory findings were described in children with CCHF. Our purpose was to emphasize the importance of this disease as a public health problem in Iran, particularly in the southeastern Iran.

**Methods:** In this study the patients under 18 years old with the diagnosis of CCHF were retrospectively

evaluated in means of sex, nationality, living environment (rural versus urban), past history of tick bite, contact history with livestock and fresh meat, the date of tick bite, contact with cattle or suspected persons, the date of the first clinical symptoms and outcome.

**Results:** In 27 patients with Congo hemorrhagic fever, 17 (63%) were men and 10 (37%) were women. The male/female ratio was 1.7. The mean age was 11.6±2.5 years (range 6-15 years). The mean incubation period was 5±3.3 days (range 1-14 days). The most common observed symptoms were fever (88.9%), bleeding (81.5%) and myalgia (66.7%). The most common site of bleeding was oral-nasal mucosa (66%) and then gastrointestinal tract (15%). Twenty-one patients (77.8%) had contact with livestock and 13 (48.1%) children had livestock in the living area. History of tick bite and contact with fresh meat were seen in 3 (11.1%) and 11 (40.7%) of the patients respectively. Laboratory data revealed thrombocytopenia and Leukopenia in 100% and 74% of the cases. Platelets less than 20000/μL were seen in 27.3% of the patients on admission. Nearly all the patients except two children were treated with ribavirin. Both these two children not treated with ribavirin died. Fatality rate was 22.2% in all the patients.

**Conclusion:** children were infected in several ways but the most common routes of infections in our patients were contact with livestock and fresh meat, and then tick bite. Sometimes the route of transmission is not quite clear-cut but the epidemiological factor of living in an endemic area is one of the important criteria to suspect to this life threatening disease. According to our study the most common clinical manifestations were fever, bleeding and myalgia. Consequently, Crimean-congo hemorrhagic fever is an endemic infection in the South-Eastern Iran and has high mortality rates among children so awareness of physicians is important for early diagnosis and treatment.

**Keywords:** Crimean-congo hemorrhagic fever, Children

### Laboratory Features of Crimean-Congo Hemorrhagic Fever in Children

*Alavi-Naini, R., MD, Khalili, M., MD;  
Research Center for Infectious Diseases and Tropical Medicine Zahedan University of Medical Sciences;  
Research Center for Children and Adolescents Health, Zahedan University of Medical Sciences*

**Background:** Crimean-Congo hemorrhagic fever (CCHF) is an acute viral hemorrhagic fever that emerges and re-emerges in many countries. The objective of this study is to determine the laboratory features of children having CCHF.

**Methods:** We conducted a retrospective study to determine the laboratory data of patients less than 18 years old in a period of six years. White blood cell

counts, hemoglobin, platelets and liver function tests were evaluated.

**Results:** Laboratory data revealed thrombocytopenia and Leukopenia in 100% and 74% of the cases, respectively. Platelets less than 20000/μL were seen in 28% of the patients on admission and in 78% during hospitalization. Hemoglobin less than 10 mg/dl were observed in 56% of the patients. Transaminases more than threefold of normal level were observed in 71% of the cases with the predominance of SGOT. BUN and creatinine were within normal limit.

**Conclusion:** The results of the study showed that thrombocytopenia, leukopenia and anemia are the most common laboratory features of children with CCHF. Crimean-Congo hemorrhagic fever should be kept in mind in any children with acute onset of fever with thrombocytopenia, anemia and leucopenia in an endemic region for CCHF.

**Keywords:** Crimean-Congo hemorrhagic fever, children, Laboratory data

### Acute respiratory viral infection: a clinical and radiological overview lecture

*Mearadji, M., MD; International Foundation for Pediatric Imaging Aid, Neaterland*

Acute respiratory viral infection (ARVI) A clinical and radiological overview M. Mearadji International Foundation for Pediatric Imaging Aid. Acute inspiratory infections (ARVI) are prevalent worldwide and directly responsible for morbidity and mortality mainly in children under 5 years of age. The most frequent agents by ARVI are respiratory viruses, including human respiratory syncytial virus, adenovirus, different types of influenza viruses, parainfluenza, rhinovirus and bocaviruses to be discovered. Generally similar clinical signs by different viral infection as fever, cough, wheezing and tachypnea will be found. A more severe clinical course is to be expected by different risk factors in all viral infection such as lower weight, prematurity, congenital heart diseases, chronic lung diseases as well as immunodeficiency. Depending on severity and condition of the affected children, the majority of patients with ARVI require hospitalization, intubation and mechanical ventilation because of respiratory failure. In some severe cases of respiratory syncytial virus infection and influenza, extracorporeal membrane oxygenation (in specialized centres available in West-Europe and USA) is absolutely indicated. Hypersecretion following tracheobronchitis is the initial cause leading to ARVI by most type of viral respiratory infection. It will also affect the lower respiratory tract following mucosal obstruction complicated with super infection resulting in severe dyspnea. The radiological features of ARVI are similar in all type of viral infection, depending on severity with hyperinflation, bronchial wall thickening and atelectases with or without

consolidation following bacterial superinfection. This presentation is based on a retrospective study of 79 patients suffered from ARVI including 41 children with respiratory syncytial virus infection. The remaining cases were affected from other agents as adenovirus, different type of influenza viruses parainfluenza, metapneumovirus, Bocavirus, Ebstein Barr virus affecting the lung, cytomegaly-pneumonia and varicella, nearly all patients were admitted to ICU and mostly intubated. A few cases underwent extra corporal membrane oxigation. The serial chestfilms or a performed CT by infants and children with different acute respiratory viral infection were reviewed. The radiological signs as hyperinflation, bronchial wall thickening and atelectases were the usual findings by nearly all type of viral infection. A bacterial superinfection was a frequent finding in a consolidated lung. However atelectatic area with partial superinfection were radiologically not recognized at all. In such cases the deterioration of patient condition, a positive bacterial culture by sampling and a elevation of CRP were all evident signs to suspect a bacterial superinfection leading to antibiotic treatment.

**Conclusion:** An acute respiratory viral infection in young children is a life threatening event. Serial chestfilm is needed to suspect the diagnosis and to follow the pulmonal condition. A bacterial super infections is a frequent complication by all type of viral infection. Atelectasis occurs more frequently in association with bacterial infection.

**Keywords:** ARVI

### **What is the most common causes of chronic cough in children attending to tabriz children's hospital**

*Jabbari-Mogaddam, Y., MD, ENT Department of Children Hospital; Sadeghi Shabestari, M., MD, Allergy Department of Children Hospital*

**Background:** Foreign body aspiration, chronic rhinosinosis, asthma were the most common causes of pediatric chronic cough, which is a daily cough lasting for > 4 weeks. cough is one of the most common complaints of childhood and has various causes and the causes of chronic cough are rarely studied in children of our region, and also the etiologies of chronic cough in children are different in comparison with adult. The aim of this study is evaluation of different etiologies of chronic cough in children.

**Methods:** All children referring with the chief complaint of resistant chronic cough more than 4 weeks to ENT and allergy clinic of Tabriz children hospital during a period of 6 month were evaluated. After accurate observation about type of cough, onset time and exact physical examination, the most common signs and symptoms in this period were recorded and necessary evaluations on probable diagnosis were made to reach a definite diagnosis.

**Results:** During 6 month 108 children with the average age of  $6.33 \pm 2.79$  evaluated (76 boys(70.4%) and 32 girls(29.6%) were studied. Productive coughs and post nasal discharge were the most common clinical findings. Chronic Rhino sinusitis (55%) along with allergic disorders(44%), asthma (31%) and foreign body aspiration (14%) were the common causes of chronic cough. Diffuse pulmonary hoarseness was the common finding in pulmonary examinations. The most common radiographic findings were maxillary sinusitis and pulmonary hyperinflation. Pollen and mites are the most allergens in patients with allergic states. With recognition of underlying cause, recovery was observed in one to three months.

**Conclusion:** Considering the common causes of chronic cough can be helpful in prompt diagnosis of this type of patients and their adequate treatment. Accurate history and physical examinations can be useful in presenting correct diagnosis. By diagnosing the certain cause of cough, recovery could be seen in most of patients in one month.

**Key words:** Chronic cough; children; etiology

### **Sepsis in pediatric burn patients**

*Sobouti, B., MD, Iran University of Medical Sciences; Fallah, Sh., MD, Shaheed Beheshti University of Medical Sciences*

Pediatric burns are an important cause of childhood morbidity, often resulting in carring and appearance change along with loss of function. Sepsis is common in the pediatric burn patient and can markedly increase mortality. Anticipation, prompt diagnosis of infection and effective therapy can result in successful outcomes for many of these children. Serious burns render children susceptible to a variety of infectious complications. Local and systemic factors contribute to this susceptibility. Local factors include the open wound, an incompetent gut barrier, exposure of the globe/ bones/ cartilages/ joints, central venous/ arterial catheters, endotracheal tubes, bladder catheters and transnasal tubes all increase the risk of injured children to potential infection. Systemic factors are also important contributors to the increased susceptibility to infection. There is a well-documented global decrease in humoral and cellular immune function associated with burns. Neutropenia is common, neutrophil function is depressed and T-cell transcription is altered. Data suggest that excessive transfusion of blood products may exacerbate global immunosuppression. Most infections in burned children are bacterial. The most common organisms are *S. aureus* (MRSA) and *p. aeruginosa*. Other organisms include CONS, VRE, GAS, *E.coli*, klebsiella, acinetobacter, serratia, proteus and enteric gram negatives. Burns patients have reduced cellular immune function and viral infections (VZV, HSV, CMV) are more common in burned children than is generally appreciated. Candida,

aspergillus, mucor and fusarium are the dominant fungal pathogens in this setting. Clinical diagnosis of sepsis is made by meeting at least three of the following criteria: -burn wound infection ( $> 10^5$  organisms/gr tissue) with histologic or clinical evidence of invasion. -Thrombocytopenia ( $< 50,000$ ) or falling rapidly -Leukocytosis or leukopenia ( $>20000$  or  $< 3000$ ) -Unexplained hypoxia, acidosis or hyper / hypoglycemia -Prolonged paralytic ileus -Hyper/hypothermia ( $> 39$  c or  $< 36.5$  c) -Positive blood culture- Documented catheter or pulmonary infection - Altered mental status -Progressive renal failure or pulmonary dysfunction. Local evidence of invasive wound infection includes: -Black or brown patches of wound discoloration -Rapid escher separation - Conversion of wounds to full – thickness -Spreading peri-wound erythema-Punctuate hemorrhagic sub-eschar lesions -Violaceous or black lesions in unburned tissue (ecthyma gangrenosum)

Common infectious complications in child burns: burn impetigo-burn – related surgical wound infection -burn wound cellulites-invasive burn – wound infection - otolaryngologic infections-ophthalmic infections-pulmonary infections-endovascular infections-intra-abdominal infections-musculoskeletal infections-urinary tract infections central nervous system infections Quick and effective closure of deep burns is the cornerstone of infection prevention. Other methods include systemic antibiotics, topical antimicrobial agents, infection control practices, IVIG, IFN- $\gamma$ , G-CSF and probiotics. Infection remains the largest single cause of morbidity and mortality in acutely burned children. Anticipation of the common related infection facilitates early and more effective treatment. If infection can be controlled and wounds closed, most seriously burned children will have very

### Epidemiological survey of measles in the cities covered by Medical University of Gonabad year 2006-2009

*Ramezani Awal Riabi, H.; Gonabad University of Medical Sciences*

**Background:** A viral disease, measles is an acute contagious attraction rate in developing countries is 5-1% and 30-10% in some countries is estimated. Improve immunization coverage (over 95%) reduced cases of measles and rubella, and reducing child mortality has to accompany. Our country due to high immunization coverage to reach 95% capacity is removed to reach the stage that all the efforts of health authorities to require this study to evaluate the risk of measles and centers have been set.

**Methods:** We conducted this study to the necessary information from the measles elimination program review forms, including linear lists measles and measles epidemiology review individual form from

health centers - public and private health city health centers were sent were extracted.

**Results:** Results from surveys conducted over the past four years a total of 10 cases of health centers - public and private treatment has been reported that four males and four females. 8 months of age  $\pm$  12 years old, eight were rural and one urban. 30% have a history of vaccination dose and 40% had history of measles immunity in turn and the rest of this age were not making. Index sampling (three samples of throat, urine and serum) from suspected cases %99.9 respectively of the type of center reporting health houses, %30, %60 of health centers - public health and 10% were from public hospital.

**Conclusion:** The results of a laboratory sample to the laboratory treatment of measles virus in the samples declared negative seems to immunization coverage above 95% of health authorities in reporting suspected cases of measles to prevent epidemic diseases effectively

**Keywords:** Measles, Epidemiology, Gonabad, Iran

### Epidemiology of pertussis in Iran; findings of national surveillance system

*Zahraei, SM., MD, Hodaiei, P., MD, Doosti, F., Babaei, A.; Center for Disease Control, Ministry of Health and Medical Education*

**Background:** Outbreaks of pertussis were first described in the 16<sup>th</sup> century. Before availability of pertussis vaccine in the 1940, pertussis was one of the most common childhood diseases and a major cause of mortality in the world. Pertussis remains a major health problem among children in developing countries with 295000 deaths resulting from the disease in 2002. Since 1984, due to well development of EPI program and high coverage of pertussis vaccine, incidence rate of pertussis has decreased in Iran.

**Methods:** This article is based on the findings of the National Surveillance System for pertussis which was established in 2006. Based on the national guidelines for pertussis surveillance, all health facilities immediately report suspected cases of the disease to the district health center, completed case investigation forms will be send with nasopharyngeal specimens to the designed reference laboratory. Specimens will check for pertussis by culture and RT-PCR. In this descriptive study, information which produced by the national surveillance system in 2007-2009, will be discussed.

**Results:** Total reported suspected cases of pertussis were 1965 in the study period, out of each 1394 specimens has been sent to the reference lab. Confirmed cases by culture were 2, 5 and 10 in 2007 to 2009, respectively. At the meantime, 27, 32 and 82 cases confirmed by RT-PCR in 2007 to 2009, respectively. Distribution of suspected cases in different age groups were as follows: 21% in  $>2$  month,

37% in 2-12 months, 26% in 1-4 years and 16% in 5 and more years old. There was a seasonal pattern with high prevalence in summer and spring. The disease was mainly in children who lived in urban areas (73%) in comparison to rural areas (27%).

**Conclusion:** Pertussis surveillance system has been improved step by step since its establishment in 2007. However, it could be strength by establishment of a network of laboratories in different part of the country under supervision of the reference laboratory. In addition, low number of reported cases in adults would be due to modified clinical presentation of the disease in this age group.

**Keywords:** Epidemiology, Pertussis, Iran

### **Epidemiologic study of recent measles outbreaks in East and South–East of Iran in 2010**

*Soltanshahi, R., MD, Sabouri, A., Zahraei, SM., MD, Center for Disease Control, Ministry of Health and Medical Education*

**Background:** Measles is still one of the most infectious killers of children in the world. Two regions of WHO (World Health Organization) nearly have achieved in Measles Elimination (America and European regions). WHO has candidate 22 countries in EMRO Region for Measles elimination to 2010. At present this aim hasn't been implemented in all 22 countries that Iran is one of those Immunizations of 33 millions of people in age groups 5 to 25 years in 2003, was a successful project and one main step for Measles elimination in IR Iran. The number of lab confirmed Measles dramatically was decreased in 2004 and 2005 (less than 10 cases annually). 10 outbreaks have been occurred in some districts of 4 provinces (Khorasan, Sistan Baluchestan, Hormozgan and Kerman) in east and south–east of Iran Since beginning of 2010 (by August). This article studies these outbreaks epidemiologically.

**Methods:** This study is an epidemiologic investigation (cross sectional). All of lab confirmed measles cases have investigated by sex, age, nationality and vaccination history in 10 districts in 2010 (first 6 months).

**Results:** From 521 measles suspected cases in areas with outbreaks, 199 cases were confirmed by laboratory. 100 cases (51%) were male and 99 cases (49%) were female. In review of nationality 86% were Iranian, 11% were afghan and 3% were from Pakistan. In review of age groups 20%, 40%, 23%, 9% and 8% of cases were respectively in age groups under 1, 1-4, 5-9, 10-14 and 15 and more years old, respectively. In the case of living area, 61% and 39% were from rural and urban areas. About history of vaccination 20% (38 persons) were under 1 year (haven't reach age vaccination), 42% (79 persons) had been vaccinated and 37% (68 persons) had not been vaccinated.

**Conclusion:** Although measles vaccination coverage is determined above 95%, 68 (37%) cases had not been vaccinated. So it is proposed one survey be designed that could investigate both vaccine coverage and vaccine efficacy.

**Key Words:** Measles, Elimination, Outbreak, Iran

### **Polio outbreak in Tajikistan: a real experience of Polio importation**

*Mousavi Firouzabadi, ST., MD, Zahraei, SM., MD, Abdol Yaghini, F., Zamani, Gh., Center for Disease Control, Ministry of Health and Medical Education*

**Back ground:** The last case of laboratory confirmed Polio was reported in 1991 and the last case of clinically confirmed Polio was reported in 1997. Following an increase in number of Acute Flaccid paralysis (AFP) cases, additional investigations revealed a Polio outbreak in Tajikistan. At this time 70% of global Polio cases in 2010 are from Tajikistan.

**Methods:** This article is about an outbreak investigation reflected in World Health Organization (WHO) reports and reviews potential risk for importation of Wild Polio virus for other Polio-free countries.

**Results:** The last laboratory confirmed Polio case in Tajikistan was reported in 1991. Tajikistan as a member state of European region of World Health Organization was certified as Polio-free in 2002. Since then formal national reports indicated satisfactorily national coverage of routine immunization with OPV3 and also AFP surveillance system performance indicators. following detection of an AFP cluster outbreak (with date of onset of the first case on 26 December 2009), primary laboratory investigations confirmed 32 Polio cases. Further investigations revealed more AFP cases throughout Tajikistan which ensued by more laboratory confirmed Polio cases. While primary epidemiological investigations suggested that Afghanistan should be the origin of an importation, final virologic sequencing of wild viruses showed an Indian virus from Uttarpradesh as cause of importation.

There are 456 laboratory confirmed Polio cases in 2010 from Tajikistan (70% of global Polio count). 12 Polio cases have been reported from Russian Federation which 5 of them are believed to be local transmission. No other Polio-free countries with Tajikistan have reported Polio cases.

**Conclusion:** Experience of Polio outbreak in Tajikistan indicates that Polio-free status should be considered vulnerable if special attention is not paid to strict monitoring of routine immunization coverage or AFP surveillance indicators. Performing Supplementary immunization activities should also be considered in High risk areas and countries.

**Keywords:** Tajikistan, Poliomyelitis, Outbreak, Importation

## Miscellaneous Abstracts

### Specialized pediatric care in Afghanistan, an example of the third type of humanitarian medicine

*Leis, A., French Medical Institute for Children, Kabul Afghanistan*

Afghanistan has suffered from three decades of conflicts and war resulting in a serious affection of infrastructures and services, like the health system. Numerous health care providers left the country during the war, the access to education and professional activities for females was restricted by the policies during Taliban regime. Today Afghanistan's health system is suffering from lack of resources; the indicators of the quality of the health system confirm the observation. Afghans are obliged for an important number of diseases to leave the country to get adequate care abroad. The government of Afghanistan with the help of the international community has undertaken considerable efforts in rebuilding the health system. An example for the establishing of a center of specialized pediatric care is the French Medical Institute for Children in Kabul, a private public partnership between the governments of Afghanistan and France, the Aga-Khan-Development Network and the French NGO "La Chaîne de l'Espoir" inaugurated in 2006. In the first four years of the existence of the institute numerous treatments have been installed that never had been performed before in Afghanistan, like pediatric cardiac surgery, pediatric intensive care, pediatric spine surgery or pediatric laparoscopic surgery. Capacity building of local staff has led to an increasing autonomy even in advanced techniques. In 2010 an Afghan cardiac surgery team performed the first independent pediatric open heart surgery.

The French Medical Institute of Children in Kabul is an example of a successful concept of transfer of knowledge and competences in specialized pediatric care even in an environment with limited resources

### Congenital disorders and its relation with familial marriage in the dead infants of Kalale Township in 1388

*Fadaei, E., MD, Charabin, A., Mallah, A.*

**Background:** The congenital disorders are some of important causes of infant's death. Other than creating emotional problems for parents, the birth of an infant with congenital disorders needs to spend high costs, too. The familial marriage in Iran is one of the most important causes of congenital diseases.

**Methods:** The descriptive study and the discussed society of all infants of township who have died for different reasons in the current year. A death discussion form is filled for all cases of infant's death after the death report in the Township, whose information is extracted from the health profile of households, interviewing with the households and referring to hospital. The tools of collecting data designed in the shape of a form. After collecting data, the questions were encoded, the data entered to spss software, and studied by the discretionary and analytic statistics.

**Results:** The findings represent that 41 cases of infant death occurred during the year of 88 (urban and rural). The most cause of infant death is immaturity (41.46%). The congenital disorder was reported in the second rank of infant's death reason at the township (32.02%) which the cardiac system disorder is at its top (50%). 16 cases of infant death were reported because of disorder. 13 cases (81.25%) of death occurred for congenital disorder on the first week. The results showed that the disordered infants, (10 cases), parents (62.5%) have had a familial relation. There was a meaningful infection between disorders led to death and the parents, familial relation with the certitude ration of 95%.

**Conclusion:** The results showed that the familial marriage is higher in the disordered infants showed that the familial marriage is higher in the disordered infants meaning fully which is like the results acquired from the others, research. So its necessary to teach the households and young's based on the dangerous familial marriage.

**Keywords:** infant death- congenital disorder

### Severe pleural effusion due to hydrocarbon poisoning in patients admitted to Madany hospital, Khorram Abad

*Faraji Godarzi, M., MD, Taei, N., MD, Tarhani, F., MD, Bajelan, H., MD, Lorestan University of Medical Sciences, Madany hospital*

**Background:** Hydrocarbon poisoning is one of causes of patient's admission to pediatric emergency centers. Oil and other Materials are ingested by children instead of Water, Syrup or juice. Hydrocarbon poisoning usually are without complication but occasionally Complicated with pleural effusion and Pneumothorax and pneumatocele Formation and leads to prolonged hospitalization and may be leads to child death (1%). Pleural effusion is one of the infrequent complications of Hydrocarbon poisoning that may need to administration of broad spectrum antibiotics and chest tube insertion. Goal of study were evaluation of

Hydrocarbon poisoning patients with severe complications.

**Methods:** This is retrospective study in 15 patients that admitted to Madany hospital for Hydrocarbon poisoning from winter to summer in 1389. Tools of study were Files patients and varieties were age, sex. Living place (Rural or urban), clinical signs and symptoms and terminal events. Then data analyzed and performed.

**Results:** In 15 patients admitted for Hydrocarbon poisoning, (86/6 %) were boys, (60%) were rural, and age range were from 15 months to 7 years. Clinical manifestations were (40%) chemical pneumonia, cough in (33/3%), vomiting in (50%), Fever in (20%) Sleepy condition in (13%), chest pain and tenderness in (13%) and the most time poisoning were in winter. In two patients high Fever, unresponsiveness to broad spectrum antibiotic were seen. In Laboratories findings high level of ESR upon to 90 and severe Leukocytosis and shift to left were reported. Physical examinations reveal severe chest pain and tenderness.

**Conclusion:** Hydrocarbon poisoning is one of causes of children admission in hospital that occasionally complicated and leads to morbidity and mortality. Then parents learning for children care and unused milk, juice or syrup bottle for taking oil and on time referred patients to hospital were recommended.

**Keywords:** Hydrocarbon poisoning- pleural effusion- Pneumothorax

### The impact of molecular medicine on clinical pediatrics

*Ranjbar, A, Bonn, Germany*

Molecular medicine is a wide and exciting field of basic research, changes medical sciences from phenomenological to causal, increasing our understanding of molecular errors and pathologic processes in different diseases. The recognition of fundamental molecular errors led to increase the precision and predictability of diagnostic procedures and targeting therapeutic approaches, especially in pediatric illnesses. In present article we discuss about various aspects of the growing impact of molecular medicine on clinical pediatrics by presenting pediatric cases with particular regards to cellular and molecular interactions and immunologic disorders.

### Improvement of Vision in Children

*Omid Salehpour –Negah Center*

In spite of general belief, using the eyes doesn't cause weakness of vision meaning worsening of refractive error. In children with amblyopia who are wearing glasses and doing patching or both, it would be better to use the eyes to improve the vision. In children with low vision, it is the best to use the eyes more. Moving too close to the items in these children is not harmful

and by magnification of the object could help the vision to improve. Using the eyes in children with visual impairment is a part of vision Physiotherapy. In conclusion, prescription of glasses, patching the eye and others are useful when the eyes are used more.

**Keywords:** Amblyopia, Low vision

### Accident prevalence Leads to hospitalization in under 8 years old in ashayer hospital in 1388

*Jaferian, M., MD, Taei, N., MD, Yarahmadi, A., MD; Naghavi, Z., Jazayeri, E.; Lorestan University of Medical Sciences*

**Background:** After advancing hygienic and vaccination program, reducing death from pneumonia and gastroenteritis, accident are the most common causes of children mortality and morbidity. Goal of study were evaluation of accident mortality and morbidity in children under 8 years in ashayer hospital in 1388

**Methods:** This is retrospective study in all patients admitted to ashayer hospital for accident in 1388 patients that referred to ashayer hospital for accident. 208 file patients were found, varieties were age, sex, duration, and then data performed and analyzed with statistical methods.

**Results:** In 208 patients that referred to ashayer hospital for accident 64.4% were boys and they were 9% under 2 years old, 25.8 % were between 2-4 years old, 31.1 % were between 4-6 years old, 34.1 % between 6-8 years old. total duration were from 1 day to 57 days. 79.3% were under 7 days. 13% were between 7-14 days, 7.7% were upper then 14 days. 72.2% were urban. Unfortunately 15.86% died.

**Conclusion:** In this study the most mortality and morbidity were boys, and 6 years old and 7 days were most duration then we propose that parents must be careful for take care of their children and traffic training

**Keywords:** Accident, Khorramabad, Child

### Evaluation of causes of mortality in 1-59 months in Lorestan province in 1387

*Setayesh, M., MD, Naghavi, Z., Jaferian, M., MD, Taei, N., MD, Yarahmadi, A., MD; Lorestan University of Medical Sciences*

**Background:** Accidents and events after pneumonia and diarrheal diseases are the most causes of mortality in childhood period. Childhood mortality related to many causes for example: age, sex, occupants parents knowledge. Annually, fire, car accidents, falling, drowning, are the most events that leads to death in childhood period. Goals of study were evaluation of

childhood mortality in 1-59 months in Lorestan province in 1387.

**Methods:** This is a retrospective study in child hood mortality in Lorestan province. Varieties were sex, age, causes of mortality. In formations accepted from hospitals or health centers. The data performed and analyzed with statistical methods. Goals of study were evaluation of childhood mortality in 1-5q months in Lorestan province in 1387.

**Results:** 125 children died in Lorestan province in 1387. 42.2% died in hospital and 57.6% died in out of hospital in events sites. 64% were rural and 36% were urban. 53% were boys and 47% were girls. 27% died with congenital anomalies, 28% with accidents and events, 12% died after respiratory and diarrheal disease.

**Conclusion:** Respiratory disease and diarrhea, events and accidents, congenital anomalies are the most causes of mortality in Lorestan province. Parents' knowledge and broadcasting information, training classes for parents are the best way to prevention accidents and events. Genetic counseling and prenatal diagnosis are the most way to prevent congenital anomalies

**Keywords:** Events, Accident, Mortality, Khorramabad

### Does Pre-Adenotonsillectomy Echocardiographic Findings Change Postoperatively in Children with Severe Adenotonsillar Hypertrophy

*Jabbari-Moghaddama, Y., MD, ENT Department of Tabriz Children Hospital; Ghaffari Babilb, Sh., MD, Cardiology Department of Tabriz Children Hospital, Abavisania, Kh.*

**Background:** Adenotonsillar hypertrophy (ATH) is the most common cause of upper airway obstruction and obstructive sleep apnea symptoms in children. The aim of this study is to compare pre-adentonsillectomy echocardiographic findings with postoperative findings.

**Methods:** From August 2007 to November 2008, fifty-five children with adnotonsillar hypertrophy and obstructive-sleep symptoms (aged 4 to14 years old of which 35 were males and 20 females) were randomly selected. Preoperatively echocardiography was performed for all patients by the same pediatric cardiologist. One month and six months after operation, patients with positive findings were followed up and again echocardiography was performed separately.

**Results:** All the patients' parents complained about severe open mouth snoring, agitated sleep and hyperpnoea. No complaints of apnea were reported. Tonsillar grades of all cases were type III or IV. The preoperative mean pulmonary arterial pressure levels of the 4 (7.3%) children were higher than normal range (25mmhg) and it decreased significantly after operation ( $P<0.000$  Npar test) (Preoperative MPAP =  $32 \pm 3$  mmHg, and six months postoperative follow up, MPAP= $11 \pm 5$  mmHg). The preoperative tricuspid

regurgitation pressure level of 7 children was higher than normal range and it decreased significantly after operation ( $p<0.000$  preoperative TR= $34 \pm 8$  mmHg postoperative TR= $19 \pm 6$ mmhg). AC/ET in these 7 patients were lower than 0.4.

**Conclusion:** This study showed that chronic symptomatic obstructive adenotonsillar hypertrophy cause higher tricuspid regurgitation pressure and mean pulmonary arterial pressure, and relief by adenotonsillectomy.

**Keywords:** Adenotonsil, Echocardiography, Children

### Sport Dentistry And Mouthguards In Children

*Ghadimi, S., DDs, Ghandehari Motlagh, M., DDs, Ahmadi, R.; Department of Pediatric Dentistry, Tehran University of Medical Sciences*

Many children and adolescents attract to participate in athletic activities. Due to lack of standard medical and dental forms most of physicians and dentists who treat children do not have any information about involvement of these children in sports. Participation in sports has several benefits to growing children and adolescents, dentists and physicians goal is maximizing the benefits and reducing injuries. According to The International Academy for Sports Dentistry; "Sports Dentistry involves the prevention and treatment of orofacial athletic injuries and related oral diseases". Dental injuries are common among orol/facial injuries during participation in sports which most of these dental injuries are preventable. Victims of these injuries face lifetime dental costs. The important appliance for protecting teeth and surrendering structures is intraoral mouthguard. This article reviews the guidelines and presents some preventive mouthguards which can significantly reduce dental injuries.

**Keywords:** Sport dentistry, Mouthguard, Children

### Is parental consanguinity a risk factor for cleft lip and cleft palate in their neonates?

*Akhavan- Karbasi, S., MD, Golestan, M., MD; Shaheed Sadoughi Medical Sciences University, Yazd*

**Background:** Today, frequency of consanguineous marriages declined in the developed countries but consanguinity is a recognized common practice among marriages in Iran and traditional cities. The purpose of this study was to evaluate and compare frequency of cleft lip (CL) and cleft palate (CP) in consanguineous and unconsanguineous marriages.

**Methods:** In a descriptive, cross-sectional study, all live births in all maternity hospitals evaluated between March to December 2008 in Yazd from veiw of cleft lip and cleft palate.

**Results:** 4800 live delivered neonate evaluated. Eleven live births had CL and/or CP (2.29 per 1000 live births). Positive family history of CL and/or CP was seen in 45.4% of patients. Thirty percent of patients had both CL and CP and 8% had only cleft lip. Cleft lip and/or palate was more prevalent in consanguineous marriages (63.4% versus 35.6%,  $p$ -value = 0.001). The incidence rate of cleft lip and cleft palate was not statistically different based on maternal age, birth order, mother's educational level, maternal occupation.

**Conclusion:** In this study, consanguineous marriages was a risk factor for cleft lip and cleft palate in their neonates. Therefore, genetic counselling and recurrence risk assessment of congenital anomalies should be done in consanguineous marriages especially in families with positive family history.

**Keywords:** Cleft lip, Cleft palate, Consanguineous marriage

### Scorpion sting in children in Tehran-Iran: a review of 28 cases.

*Farnaghi, F., MD, Bayat, M., MSc, Jazayeri-far, F., MSc; Nurse. Shahid-Beheshti University of Medical Sciences.*

**Background:** Scorpion sting is a rare but medically important emergency condition in children with significant mortality and morbidity.

**Methods:** It was a retrospective study of all children admitted after Scorpion sting in Loghman Hakim Hospital (the only referral pediatrics toxicology center in Tehran) from April 2005 to August 2009. General characteristics of the children, species of the scorpions, anatomic site of the sting, clinical and laboratory findings, treatment approaches, complications, and outcomes were recorded.

**Results:** A total of 28 scorpion sting envenomation children (mean age 5.1 ± 2.2 years; from 9 mo to 12.5 yr) were investigated. The male to female ratio was 1.1. Most of stings (84%) occurred in summer months & 23 patients (82%) were admitted from rural areas. Ten stings (36%) were inflicted by *Androctonus crassicauda* (black fat tail scorpion), 64% by *Mesobuthus eupeus* (yellow scorpion) and there were no cases of *Hemiscorpius lepturus*. The lower extremities were the most common site of stings (62%). 64% of our patients were stung between 6pm to 12 mn. Two patients were completely asymptomatic and all of the others became symptomatic during the first 6 hours after their stings. Local manifestations such as pain were the most common clinical manifestations. Only 8 patients showed systemic manifestations. CNS, autonomic (miosis or salivation), somatic skeletal neuromuscular dysfunction and cardiovascular manifestations were the most frequently seen clinical findings. Four patients (14.2%) had signs of serious envenomation and required admission to the PICU. Twenty patients

received 1 vial and 4 patients received 2 or 3 vials of antivenom without any complication. Two cases with severe carpopedal spasm and opisthotonus position who were unresponsive to initial treatment, successfully treated with midazolam drip without any significant complication. There were no cases of pulmonary edema, hypertension or DIC. Fortunately there was no mortality or morbidity.

**Keywords:** scorpion, sting, children.

### Breastfeeding in infants conceived by assisted reproductive techniques in Royan Institute

*Nedaeifard, L., Mozafari- Kermani, R.; Child Health and Development Research Department of Iran Medical Sciences Branch of Academic Center for Education, Culture and Research*

*Asadpour-Tehrani, M., Nateghi, MR., Shahzadeh Fazeli, A., Gourabi, H., Department of Genetics, Royan Institute for Reproductive Biomedicine*

**Background:** Breast feeding is a major determinant of infants' health and survival. The aim of this study is to determine the pattern of breastfeeding and to assess its determinants in assisted reproductive techniques (ART) infants.

**Method:** In a descriptive cross sectional study, 312 infants were selected by the non-incident consecutive method from ART infants who refer by Royan institute to our center in the period of study. Infants were born between September 2007-September 2009 in Tehran. Infant nutrition, mother age, number of gestation, ART techniques, birth growth indexes (weight, height and head circumference), beginning time and kind of nutrition after birth and growth pattern until 6 months old were considered. Data were analyzed by SPSS-version 10 by using Chi-square test.

**Results:** 158 (50.6%) of infants were boys and 154 (49.4%) were girls. 110 (35.2%) were breastfed, 46 (14.8%) were formula feed and 156 (50%) were both breast and formula feed. 65 (60%) were exclusive breast feeder (EBF) till 6 months. 253 (81.1%) infants were fed first day after birth. Rate of feeding in the first day after birth and EBF of neonate with low birth weight were less than normal birth weight ones ( $p=0.001$ ). Approximately 92% of infants who had normal increase of weight and height till 6 months, began feeding in first day of life ( $p=0.016$  and  $0.05$  respectively). Mothers age, multi fetal pregnancy and ART technique did not affect beginning time and kind of infant feeding, and EBF.

**Conclusion:** According to our study the most important factor which affect ART infants nutrition pattern is their birth weight. Low birth weight infants were fed with formula more than normal birth weight infants and as a result they were less EBF. Beginning time of feeding affect their weight and length increase till 6 months.

**Keywords:** Assisted Reproductive Techniques, Infants, Breast feeding, Exclusive Breast feeding, Growth

### **Methadone is the most common childhood acute poisoning in Yazd**

*Golestan, M., MD, Owliaee, H, MD, Amiri, N., Harand-Aavar, M., Sallary, R., MD, Shaheed Sadoughi University Medical Sciences, Yazd*

**Background:** Poisoning is a significant public health issue for children. Poisonings, like other types of injuries, are predictable, and preventable events. Epidemiologic surveillance in each area is necessary for prevention and treatment planning program. The purpose of this study was to clarify the characteristics of acute poisoned children admitted at hospital in yazd city.

**Methods:** In this retrospective study, all children who were admitted due to acute poisoning in affiliated hospitals of the Shaheed Sadoughi University of Medical Sciences in Yazd from October 2006 to March 2009 were included. Data were collected from medical records. Results : Among 116 poisoned children from birth to 15 years old who were admitted in hospital, 49% were under 5 years. The most common age group were the 1-5 year s old ( $P < 0.01$ ) Opiate was found to be the most common cause of poisoning in children (36% ) and 60% of them was Methadon, the significant signs were impaired consciousness, respiratory depression and miosis. In 90% of children with methadone poisoning, one of their parents were on methadone maintenance treatment.

**Conclusion:** Methadone was the most common cause of childhood acute poisoning in Yazd and in almost all one of their parents were on methadone maintenanc treatment (MMT). MMT is a widely used pharmacological intervention for people recovering from heroin addiction and risks to children for unintentional methadone exposure are real and can result in serious health complications

**Keywords:** Acute poisoning, children, unintentional, methadone

### **Evaluation of medical errors in hospitalized children in khorramabad Madani hospital in first half of 2008**

*Mohsenzadeh, A., MD, Lorestan University of Medical Sciences, Khoram Abad*

**Background:** Hospitalized children are suffered from medical errors may cause serious injuries. The aim of this study to evaluate medical errors in hospitalized children in khorramabad Madani hospital in first half of 2008.

**Methods:** This study was a cross sectional that was performed for all medical errors in hospitalized children in khorramabad Madani hospital 21/3/2008 to 21/9/2008. The sampling method was simple. Studied variables including: age, sex, weight, kinds of errors, education of parents, job of parents. Data collected by questionnaire and analyzed by spss software.

**Results:** In this study out of 2250 records, 151 (6/3%) had medical errors. 53% were girls and 47% were boys that there is a significant relation between sex and medical errors. 46/4% were related to age group lower than 2 years. Most of errors were occurred in weight group of 6kg. Types of medical errors including drug ordering 46/3% (involved incorrect dosing (37%), frequency 28%, rout 19% and others 16%), transcribing 10%, administering 32/4%, dispensing 11/3%. Most errors related to liquid therapy 76/2% and intravenous rout 85/4%. Most errors were occurred during night 47% and during weekend 56/6%. Conclusions Medication errors are common in pediatric inpatient settings, in this study was 6/3% and further efforts are needed to reduce them.

**Keywords:** medical errors, hospitalized children

### **Barriers to Physical Activity in A Population-Based Sample of Children and Adolescents in Isfahan, Iran**

*Kelishadi, R., Ghatrehsamani, Sh., Hosseini, M., Mirmoghtadaee, P., Manouri, S., Poursafa, P.; Isfahan Cardiovascular Research Center*

**Background:** To explore the barriers to physical activity in a representative sample of Iranian children and adolescents

**Methods:** The study was conducted in 2007 in urban and rural areas of Isfahan district in Iran. In the qualitative part, we used the grounded theory approach, including semi-structured focus group discussions and in-depth interviews. The quantitative part comprised 600 randomly selected students.

**Results:** The qualitative study included 34 school students (16 girls), 20 parents (11 mothers) and 11 school staff. All students disclosed that studying was a priority, they pointed to lack of safe and easy-access place for physical activity and unsupportive family as the main barriers, lack of self-confidence and low self-worth were the two other concepts developed in this context. Parents pointed to lack of safe and easy-access place for activity followed by the priority of studying. The concepts derived from interviews with school staff were unhealthy modeling of parents, priority of studying, and inadequate public knowledge about how to integrate physical activity in routine daily life. The quantitative survey comprised 600 students including 286 (47.8%) girls. Parents' education level had inverse association with children's physical activity level. Significant inverse associations of self-efficacy and physical activity levels were documented.

**Conclusion:** Increasing the public knowledge about adopting physical activity habits in daily routine, and informing the families and students about the benefits of physical activity for improving learning, as well as providing safe places, as using the school facilities in non-school hours should be considered in planning effective preventive strategies and interventions

**Keywords:** Physical activity, Pediatric, Barriers, Prevention, Iran

### **A ten years Experience of Posterior myectomy in patients with primary short segment aganglionosis**

*Nahvi, H., MD, Department of Pediatric Surgery, Bahrami Children Hospital, Tehran University of Medical Sciences*

The term of short segment is not clearly defined. Short-segment aganglionosis is not rare. Posterior myectomy has proved to be effective treating infants and children with a primary short segment Hirschsprung Disease without prior colostomy and also in patients with secondary aganglionosis (incomplete correction of the aganglionosis colon after a prior definitive pull through. in this study 30 patients with primary Short-segment aganglionosis who underwent Posterior rectal myectomy during a ten-year period was reviewed. Patients with secondary aganglionosis excluded from this study. Selected patients with primary short-segment aganglionosis, proved by muscle biopsy, responded well to a single transanal operation. The complications were minimal. the average postoperative follow-up were 15 months (range, 1 month to 5 years). the results were good in 90% and poor in 10%. Of patients . 4 patients was required an pull-through after the myectomy, there was not any technical difficulties.

**Conclusion:** Within the limits of this study , posterior rectal myectomy has a definite place in the treatment of primary short-segment Hirschsprung disease.

**Key words:** Constipation - posterior myectomy - short segment Hirschsprung's disease

### **Polyorchidism**

*Kajbafzade, AM., MD, Taqizade, F., MD, Sabetkish, N., MD; Pediatrics Urology Research Center, Tehran University of Medical Sciences*

**Background:** Polyorchidism is defined as the presence of two or more testes and is a rare congenital abnormality. The majority of reported cases are asymptomatic patients about polyorchidism is frequently associated with additional urological pathologies such as undescended testis, inguinal hernia, testicular torsion, hydrocoele, malignancy and infertility. Differential diagnosis includes spermatocoele, hydrocoele, epididymal cysts or aberrant epididymis. Ultrasonographic examination of the scrotum and inguinal region confirmed the findings. It is found predominantly on the left side. Classification systems for polyorchidism have developed including those proposed by Leung, Hancock and Wolf based on anatomical arrangements of the testes and their outflow paths.

**Case Report:** We checked all urologic dossier in Children's medical center, and found a very top case of Polyorchidism case. Patient was a 9 month diagnosed with left undescended testis. During operation we seen two testis, one was atrophied which resected. In microscopy studies section show fibrovascular connective tissue with smooth muscle bundles and remnants of epididymis and vas deferens.

**Conclusion:** Although rare, polyorchidism is an anomaly which should be considered when assessing any scrotal mass. Imaging with ultrasound should help clarify the presence of polyorchidism. Current management would be for surgical exploration and fixation of the masses after counselling of the patient. If there is any doubt then biopsy may also be undertaken.

**Keywords:** Polyorchidism, Testis abnormalities, Undescended testis

## Neonatology Abstracts

### Neonatal jaundice in the 21st century; still a concern

*Kadivar, M., MD, Department of Pediatrics, Children's Medical Center, Tehran University of Medical Sciences*

Jaundice is still the most common and certainly vexing problems encountered in the neonatal period. From long times ago, there are assortments of attempts to detect early hyperbilirubinemia and prevent its adverse effects on newborn infants.

Postnatal hyperbilirubinemia is universal and manifests as newborn jaundice in near 60% of full-term newborns and more than 80% of premature infants in the first week of life. Unmonitored and untreated hyperbilirubinemia may progress to excessive levels that can be associated with evident bilirubin neurotoxicity. In the spite of several years of research, many aspects of neonatal jaundice remain a mystery to the health-care workers. The elevated serum bilirubin concentration presents as much of therapeutic as a diagnostic problem to the health care personnel because of its potential to damage central nervous system. Acute bilirubin encephalopathy remains a clinical emergency and its delayed management represents an easily preventable neonatal brain injury. There are many questions regarding kernicterus as the risk and timing of bilirubin-related neurotoxicity and safe bilirubin level among newborns. Most frequent clinical and contributory risk factors for extreme hyperbilirubinemia and kernicterus are late prematurity, undiagnosed hemolytic disease, genetic abnormalities deficiency, and concurrent complications of dehydration, sepsis, or acidosis, hypoalbuminemia or poor feeding. Kernicterus, a preventable form of neonatal bilirubin induced brain injury, is presently prevalent worldwide and is a matter of public and societal concern. It may present as classic or subtle. Subtle kernicterus or bilirubin-induced neurologic dysfunction (BIND) refers to individuals with subtle neurodevelopmental disabilities without classical findings of kernicterus. Acute bilirubin encephalopathy or chronic kernicterus results in neonatal death or multisystem disabilities, including irreversible isolated auditory neuropathy (hearing loss); extrapyramidal movement disorders; neuromotor disorders; and visual disabilities. Kernicterus can be further classified as auditory predominant or motor predominant and characterized based on the severity of clinical sequelae. Treatment of jaundice includes encouraging frequent breastfeeding, phototherapy, medication, and exchange transfusion. But, phototherapy is still commonly used to treat severe neonatal hyperbilirubinemia that can be used as traditional methods using overhead or fiberoptic pads. On the other hand, the harms of

detection and early treatment of hyperbilirubinemia could include potential unknown side effects of phototherapy and possible adverse events related to exchange transfusion.

However, there is no known screening test that will identify infants at risk of developing chronic bilirubin encephalopathy. Screening tests for hyperbilirubinemia consist of clinical risk assessment, measuring bilirubin level directly either in plasma or by transcutaneous methods. Although, the universal predischarge screening combined with assessment of gestational age is most predictive of subsequent severe hyperbilirubinemia at early days after birth, but it seems to be associated with an increase in phototherapy and hospitalization rates. The strategy of early intervention should be implemented efficiently because of a narrow safety margin in neonatal period especially among newborn infants over 72 hours and during the first week after birth such that timely. There is adequate evidence that screening using hour-specific bilirubin measurement and clinical risk factors can identify infants who are likely to develop severe neonatal hyperbilirubinemia.

**Conclusion:** All newborns should be systematically followed up for risk of developing severe hyperbilirubinemia prior to discharge from hospital. It seems that universal predischarge bilirubin screening reduces the chance of developing very high bilirubin level among newborn infants. In addition to bilirubin levels, severe neonatal hyperbilirubinemia adjusted for postnatal age in hours, gestational age, and hemolysis should be considered.

### Pulse Oximetry in the First day of life in Newborns Delivered in Kashan Shabihkhani Maternity Hospital

*Mosayebi, Z., MD, Tehran University of Medical Sciences, Movahhedian, AH. MD, Kashan University of Medical Sciences*

**Background:** Failure in timely diagnosis of asymptomatic congenital heart diseases in newborns contribute to significant morbidity and mortality in this age group. To evaluate whether pulse oximetry can be used as a screening test to detect congenital heart disease in otherwise healthy newborns before nursery discharge.

**Methods:** In this cross-sectional study during a 6 month period we performed hand and foot pulse oximetry in 1506 term and healthy newborns in Shabihkhani Maternity Hospital. Babies with functional oxygen saturation below 95% considered abnormal and a second saturation measurement was

performed 2 hours later. If the repeated measurement was again below 95% they were referred for performing an echocardiogram.

**Results:** A total of 1506 newborns were screened. A postductal functional saturation of less than 95% was found in 100 cases, in whom the second measurement was performed. In 29 babies that a second measurement was also less than 95%, echocardiography was performed. Of whom 6 neonates with congenital heart disease (prevalence 4 in 1000 live birth) were detected. The types of congenital heart disease in these asymptomatic newborns were of critical type and some with ductal dependent lesions.

**Conclusions:** According to the significance of diagnosis of congenital heart disease, pulse oximetry screening along with the clinical examination could help in the early detection of critical asymptomatic congenital heart diseases. Performing leg pulse oximetry seems to be adequate for screening, because in all the circumstances it was lower than arm saturation.

**Keyword:** Oxygen saturation, Congenital heart disease, Pulse oximetry, Newborn

## Late Preterm Outcome

*Razi, N., MD, Tehran University of Medical Sciences*

In 2005 NIH, NICHD recommended that preterm infants born between 34<sup>0/7</sup> wks and 36<sup>6/7</sup> wks to be called "Late Preterm" instead of "near term". Some recent studies show increased risk of mortality and morbidity in this group compared with infants born at term. Maternal risk factors for late preterm delivery include chorioamnionitis preeclampsia, diabetes, thrombophilia, PPROM, Primigravida, multiple gestation, teenage pregnancy. 75% of preterm births are late preterm. NICU admission for 34 wks, 50% for 35 wks, 15 % and for 36 wks 8%. Late preterm infants are at risk for hypothermia. Respiratory distress in 29% of late preterm vs 4% in term, higher incidence of TTN, RDS, PPHN and respiratory failure than term infant, 10% of late preterm have significant apnea. SIDS rate is twice as term infant. Feeding problem is a common problem in late preterm. Late preterm is one of the major risk factor for severe hyperbilirubinemia and kernicterus. Hypoglycemia is more frequent than term infants. Hospital stay is longer in late preterm. Risk ratio (RR) for death comparing to term infants in first day 11.4. For < 7 day 7, for < 28 day 5.6 and < 1 yr 3.5, Neurologic abnormalities. Learning difficulties, poor scholastic achievement and behavioral problems are more common in late preterm. The rate of CP is 3 times of term newborn. It has been recommended, no discharge before the age of 48hrs and 24hrs successful feeding or if weight loss is >7%. Formal evaluation of breastfeeding and follow up visit in 24-48hrs are necessary.

In conclusion it has been found that most of prematurity related adverse neonatal outcomes are

significantly more common in the late preterm group than at full-term. Thus efforts to reduce the rate of late preterm should address both spontaneous deliveries, in the form of tocolysis and corticosteroids for fetal lung maturation, as well as re-evaluation of current obstetric practices regarding timing of indicated delivery in cases of PPROM and other obstetric complications. In addition, better understanding of short and long term morbidity associated with late preterm is important for appropriate counseling regarding expected outcome in this group of preterm infants.

## Prebiotics and the neonate

*Mohagheghi, P., MD, Iran University of Medical Sciences*

Prebiotic oligosaccharides are a hugely significant component of breast milk. Consequently, acquisition of gut flora in breastfed babies is significantly higher than formula fed infants. Prebiotics are the oligosaccharides that survive in acidic conditions, evade digestion in the small intestine, must be selectively fermented in the colon and stimulate one or a limited number of beneficial bacteria. Development of gut flora: prebiotics induce a bifidogenic effect and promote a healthy intestinal micro flora. Prebiotics promote healthy digestion and favourable stool characteristics of infants and protecting against Gastroenteritis. Also prebiotics repress pathogenic microbial growth. Human milk oligosaccharides (HMOS) are the most likely component responsible for the bifidogenic properties of human milk. New data from clinical studies show that a specific mixture of prebiotics (GOS/FOS 9:1) in infant formulas has similar functional properties to HMOS on the Faecal Flora and stool PH in Term Newborns, stool consistency, healthy digestion (reducing incidence of regurgitation and crying in connection with feeding), and Protecting against Gastroenteritis (reducing number of episodes of gastroenteritis significantly).

**Keywords:** Human milk oligosaccharides (HMOS), Prebiotics, Galacto oligosaccharides (GOS), Fructo oligosaccharides (FOS), Bifidogenic effect

## Anterior Fontanelle size Tehran Iranian infants

*Shajari, H., MD, Jeddian, A., MD, Rashidiranjbar, N., Ashrafy, MR. MD; Shariati Hospital School of Medicine; Children's medical Center, Tehran University of Medical Science*

**Background:** The aim of this study was to determine the mean size of anterior fontanelle, its distribution in gender, and the relationship between anterior fontanelle size and infant's gender, weight, height, head circumference, and type of delivery.

**Method:** 400 Iranian infants who were term and healthy participated in this study. They were examined for anterior fontanelle size, weight, height and head circumference within 24 hours of birth. The type of delivery type of each was recorded as well.

**Results:** The mean size of anterior fontanelle was  $25.34 \pm 13.27$  mm. The mean size of anterior fontanelle was  $26.70 \pm 13.19$  mm in boys, and  $23.67 \pm 13.20$  mm in girls which showed a significant difference between both genders ( $P = 0.023$ ). A significant negative correlation was found between anterior fontanelle size and infant's weight or height ( $P < 0.05$ ). No significant correlation was found between the anterior fontanelle size and infant's head circumference. The anterior fontanelle size and type of delivery, showed no correlation ( $P > 0.05$ ).

**Conclusion:** The mean size of anterior fontanelle in term Iranian infants is  $25.34 \pm 13.27$  mm. This finding could be used as an indicator for neurodevelopmental evaluation, and a clue for early diagnosis of some disorders such as hypothyroidism and skeletal disorders in Iranian newborns.

**Keywords:** Anterior Fontanelle, Infant, Clinical examination, neurodevelopmental evaluation.

### Apgar score and arterial blood gas in the first hour of birth in neonates

*Kaveh, M., MD, Davari-Tanha, F., MD, Farahani, M., Valiasr Reproductive Health Reserch Center*

**Background:** Although, the high efficacy of Apgar score to find the neonates who have respiratory distress, a low number of Apgar score couldn't determine asphyxia definitely, and other factors could reduce it, too.

**Methods:** This case control study was carried out in Mirza Koochak Khan hospital. Arterial blood gas in the first hour and Apgar score in minute 5 were evaluated in subjects (half of them had an Apgar score below 7 and the rest were 7 or over). Furthermore, the kind of delivery, gravidity, gender and gestational age were investigated.

**Results:** All of neonates were divided into two groups based on Apgar score (below 7 or over). Each group consisting of 100 newborns. We found a significant association between Apgar score and ABG in the first hour of life (pH and BE being the most important factors of ABG). In this study a positive effect was shown in male gender, the Cesarean delivery and gestational age over 37w on the association between Apgar score and ABG.

**Conclusion:** According to an association between Apgar score and ABG, ABG is recommended as a routine procedure in neonates with a low Apgar.

**Keywords:** Apgar score, Arterial blood gas, ABG, Asphexia, Base excess

### Determination risk factors of macrosomia neonatal birth, with regard to Gestational diabete

*Mohammad-Beigi, A., Mohammad-Salehi, N., PhD, Arak University of Medical Sciences, Arak*

**Background:** Macrosomia defined as infant birth weight over 4-4.5 kilogram or 90 or higher percentile of weight for age. Prevalence of macrosomia is equal to 10% in all pregnancies in united stated. This study conducted to determinates the neonatal macrosomia predictors in Shiraz

**Methods:** This case control study carry out on 420 infants that borne in Shiraz hospitals. Data analyzed with univariate method by t-test and fisher exact test and logistic regression multivariate models after gathering by questionnaire.

**Results:** In univariate method mother age, diastolic blood pressure and BMI showed a significant relationship with macrosomic neonatal birth. Also GD, macrosomia and cesarean history, preeclampsia, hydramnios, glocosuria in second and third trimester, diabetes disease and cesarean delivery related to macrosomic birth. But the most important factors in regression model were GD history (OR= 20.06), macrosomic birth history (OR= 10.52) and preeclampsia in pregnancy period (OR= 4.86), respectively.

**Conclusion:** With regard to this study results, mothers with macrosomic birth and GD history and preeclapmsia are at risk for delivering macrosomic neonatal. So in order to preventive of macrosomic side effects, adoption of weight taking in pregnancy period advised.

**Keywords:** Macrosomia, Neonatal, Risk factors, Gestational diabetes (GD), Shiraz

### Evaluation the causes of exchange transfusion in hospitalized icteric newborns from 2007 to 2009

*Eghbalian, F, MD; Hamadan University of Medical Sciences*

**Background:** Exchange transfusion is effective in sever jaundice when phototherapy may not be helpful. Present study was done in order to assess the causes leave to exchange transfusion in hospitalized icteric newborns at Besat Hospital of Hamedan city from 2007 to 2009.

**Methods:** In a descriptive cross-sectional study, a questionnaire containing sex, order of birth, age of neonate at first referral of parents for assessing neonatal jaundice, age of neonate at time of exchange transfusion, history of neonatal jaundice in family, blood group and Rh of mother, blood group and

Rh of neonates, G6PD, Coombs, Hb, blood culture and bilirubin level was fulfilled from patients dossiers for 100 neonates who had jaundice and admitted in Beasat hospital of Hamedan city and undergone exchange transfusion.

**Findings:** Mean age of studied neonates at time of first referral of parents for assessing neonatal jaundice was 3.01 days and SD of 1.47 days. At time of first referral of parents; 38% of neonates were three days old. Mean age of studied neonates at time of exchange transfusion was 8.36 days with SD of 6.69 days. 76% of neonates were male. History of neonatal jaundice in family was present in 17% of cases. ABO incompatibility was present in 34% of studied neonates and Rh incompatibility was present in 21% of studied neonates. Concomitant ABO and RH incompatibility was not present in any studied neonates. G6PD deficiency was seen in 4% of patients. Positive blood culture was reported in 15% of patients.

**Conclusion:** In present study, ABO incompatibility has been the most common cause of exchange transfusion and Rh incompatibility is located in next order. Also because in 18% of cases, parents had referred for neonatal jaundice on fifth day of birth and afterward, it seems that necessary instructions could be effective for parents of neonates with jaundice specially those at high risk of kernicterus, in order to decrease both delayed treatment and need to exchange transfusion.

**Keywords:** Exchange transfusion, Causes, Newborn

### Hypocalcemic effect of phototherapy in icteric newborns

*Alizadeh-Taheri, P., MD, Eivazzadeh-Khoei, B., Salamati, P., MD; Bahrami Hospital, Tehran University of Medical Sciences*

**Background:** Hypocalcemic Effect of Phototherapy in Icteric Newborns Abstract Back ground: Hyperbilirubinemia is a common problem in newborns. Phototherapy is the most common treatment of indirect hyperbilirubinemia in icteric newborns. Although phototherapy is a safe method, complications may occur. A significant complication of phototherapy is hypocalcemia that may cause serious problems in newborns. Finding just few studies of hypocalcemic effect of phototherapy in newborns, this study has been held up to evaluate this complication. Objective: To investigate the frequency of hypocalcemia in icteric newborns after phototherapy.

**Methods:** This study was performed on 147 icteric newborns treated with phototherapy. These newborns were completely normal in physical examination. Plasma bilirubin and calcium levels were measured before and after 48hrs of phototherapy. The frequency of hypocalcemia, the age of admission, the time of jaundice appearance, the birth weight and level of serum bilirubin were recorded.

**Results:** eighty –three newborns (56%) had decline in serum calcium level and ten newborns (7%) developed hypocalcemia. None of hypocalcemic neonates was symptomatic clinically. All hypocalcemic patients responded to oral calcium therapy.

**Conclusion:** This study shows that phototherapy in icteric newborns lowers serum calcium level. Phototherapy- induced hypocalcemia is a transient phenomena and is asymptomatic in most patients..

**Keywords:** Hypocalcemia, Jaundice, Neonate, Phototherapy

### Assessment of febrile neonates with and without low risks for serious bacterial infection

*Zarkesh, M., MD, Hashemian, H., Momtazbakhsh, M., Rostami, T., Gilan University of medical Sciences*

**Background:** It is propounded that febrile neonates with low risk criteria (LRC) can be carefully observed without parenteral antimicrobial therapy; but yet, reliability of LRC to exclude serious bacterial infection (SBI) is uncertain.

**Methods:** Epidemiological and clinical data and final diagnosis of all febrile term neonates  $\leq 28$  days of age, with no history of admission, reception of antibiotics or chronic diseases, seen in the emergency room and admitted in neonatal ward of 17 Shahrivar children's hospital of Rasht- Iran from January 2004 to January 2009 were reviewed. All of them were underwent full sepsis workup. The prevalence of SBI in total population and LRC positive and negative neonates were calculated

**Results:** A total of 202 previously healthy febrile neonates were evaluated. SBI was shown in 38 (18.8%), and was significantly more common in neonates  $> 7$  days of age. The most common type of SBI was urinary tract infection (UTI). 62 (31%) cases of 202 neonates had LRC, and only one of them (1.6%) had SBI (UTI with E.Coli). SBI was significantly more common in neonates without LRC (26.6% versus 1.6%,  $p= 0.0001$ ). The negative predictive value (NPV) of the LRC to exclude SBI was 98.4% (95% CI: 96.7% to 100%).

**Conclusion:** These findings suggest that LRC may be relied upon to exclude SBI in febrile neonates. Then, we propose that all febrile neonates should be admitted, ill or LRC negative neonates should undergo a full sepsis work up and be administered systemic antibiotics. LRC positive neonates should be under close observation, if their clinical status deteriorates, their cultures show bacterial growth, and/or they have persistent fever  $>48$  hours, undergo a full sepsis work up and be administered systemic antibiotics.

### Study of cord blood insulin in relation to maternal blood glucose in 75 gram oral glucose tolerance test

*Hematyar, M., MD, Mahboubi, M., Fadaki, FK, Mozafari, R., Islamic Azad University, Tehran Medical Branch*

**Background:** The insulin level of fetus indicate the effects of maternal diabetes on fetus and it's health in future. There is not exact glucose threshold in maternal glucose tolerance test (GTT) for fetal hyperinsulinism and it's correlation with fetal morbidity and mortality. The aim of this study is to determine the correlation between insulin levels in cord blood with maternal blood glucose levels in 75 gram oral GTT.

**Methods:** This is a cohort study on 200 pregnant women in Tehran Javaheri hospital in 2009. The 75 gram OGTT was performed at 24-28 weeks of gestation and then 2 cc of cord blood for measuring insulin was gotten at delivery time. The data analysed by SPSS ver 13 and correlation, regression and independent t test. Results: 5.5% of neonates have hyperinsulinism. The correlation between maternal fasting blood sugar and cord blood insulin was significant. The correlation between infant birth weight and cord blood insulin and also maternal first hour glucose level in 75 gram OGTT was significant. Although the level of cord blood insulin was higher in mothers with impaired GTT but the correlation was not significant.

**Conclusion:** The concentration of insulin in fetus indicate the levels of glucose that transfer from mother to fetus intrauterine. In this study the most correlation between hyperinsulinemia with maternal fasting blood sugar was observed. It seems maternal FBS has the most sensitivity for detection of hyperinsulinemia in fetus

**Keywords:** insulin, gestational diabetes, 75 gr oral glucose tolerance test, macrosomia

### Surfactant replacement therapy at NICU in Amirkola children Hospital, Iran; Experience for a decade

*Mousa Ahmadpour-kacho, MD; Yadollah Zahedpasha, MD; Masoomeh shirvani, MD; Tahereh Jahangir, BS - Babol University of Medical sciences*

**Background:** This study was carried out to determine the profile, underling diseases, complication and the out come of the neonates who received surfactant replacement therapy at NICU in Amirkola children hospital, a referral hospital in the north of Iran.

**Methods:** In this descriptive study we analyzed the age and the time of surfactant replacement, the rate of mortality, underlying diseases and some of the

complications such as: Pneumothorax, nosocomial infection, pulmonary hemorrhage and PDA in 133 neonates who received surfactant between 1999- 2007 at NICU in Amirkola children hospital. The type of surfactant which used since 1999 to 2006 was Survanta with the dose of 4 ml/kg. In 2007 because of difficult availability of Survanta some times Curosurf with the dose of 1.25 ml/kg was used. The indication for surfactant replacement therapy was respiratory failure and requirement to mechanical ventilation.

**Results:** The mean gestational age of neonates was  $32.39 \pm 3.8$  weeks and mean weight of them was  $1950 \pm 875$  grams. The mean age and time of receiving surfactant was 7.3 hours after birth. The mean length of ventilating was  $4.05 \pm 3.93$  days. The more prevalent underlying diseases were: RDS (83.56%), sepsis or pneumonia (8.45%), MAS (6.10%) and CDH (1.87%) The prevalence of complications were pneumothorax (42.48%), secondary sepsis (6.05%), PDA (3.67%) and pulmonary hemorrhage (3.42%). The mortality rate decreased from 48.87% for the first 8 years of the study to 24.88% for the last two years.

**Conclusion:** Survival rate of the surfactant treated neonates increased during the last two years of the study.

**Keywords:** neonate, surfactant, NICU, RDS

### Maternal serum zinc and copper and infant birth weight

*Mirzarahimi, M., MD, Saadati, H., Msc, Enteshari, A., MD, Mazani, M., PhD, Shahizadeh S, Isazadeh-far, Kh. MD; Ardabil University of Medical Sciences*

**Background:** Trace element deficiencies have been documented to play an important role in determination of the fetal outcome. It has been reported that the pregnant women in developing countries consume diets with a lower density of minerals and vitamins. Zinc and copper are essential trace elements and their deficiency can lead to a variety disorders. We evaluated whether is any association between infant birth weight and maternal serum zinc and copper levels.

**Methods:** This case-control study was carried out at the Obstetrics and Gynecology Department of the Alavi Hospital in Ardabil between August 2008- August 2009. Fifty six women who had delivered low- birth-weight infants (<2500gr) were taken as the case group, and from the mothers who had delivered normal birth weight ( $\geq 2500$ gr) infants 56 were selected at random as the control group. Venous blood sampl were obtained from the mothers. Serum zinc and copper levels were determined by the Atomic Absorption Spectrophotometer method.

**Results:** Mean of birth weight in infants, maternal age, body mass index in mothers and socioeconomic or demographic factors did not differ between cases and control groups. Maternal zinc concentration ( $\mu\text{g/dl}$ ) did not differ between Cases and Controls;  $55.84 \pm 14.40$

µg/dl vs.  $52.16 \pm 8.84$  µg/dl respectively. The mean serum copper level of case group was ( $231/75 \pm 38/12$  µg/dl) significantly higher ( $p < 0/05$ ) than control group ( $204/42 \pm 31/30$  µg/dl). There was a negative correlation between maternal serum copper level and birth weight.

**Conclusion:** Maternal zinc concentration has no effect on neonatal birth weight infants. There was a negative correlation between birth weight and maternal copper concentration. Maternal serum zinc and copper concentration declined as gestation progressed

**Key word:** birth weight; gestational age; copper; zinc

### Neonatal mortality in kuhdasht hospital, in 1388

*Yarahmadi, AA, MD, Taei, N., MD, Jaferian, M., MD, Naghavi, Z., Lorestan University of Medical Sciences*

**Background:** Neonatal mortality is one of the most score in developed country. The most causes of neonatal mortality were prematurity, PROM, IUGR, Abnormalities and others. Goal of study were Evaluation of neonatal mortality in kuhdasht hospital, in 1388.

**Methods:** This is retrospective study in neonatal mortality in kuhdasht hospital, in 1388. Variables were sex, neonatal weight, gestational age, congenital abnormality, causes of labor, causes of mortality, causes of, causes of CS, tools of study were file patients and then data performed and collected and analyzed with statistical methods.

**Results:** In 37 neonatal died in kuhdasht hospital, gestational age of neonates was between 22-40 weeks with 30 week average. Mean maternal age were 27.2 years. NVD were 46% and CS were 54%, Causes of death 45% were premature labor, 13% congenital abnormality, 22% other causes. 41% were girls and 59% were boys. Neonatal weight 1000-2500 gram was 60%, below 1000 gram 12% and upper 2500 were 28%.

**Conclusion:** Prematurity and congenital abnormality were the most predisposing factor for neonatal death in kuhdasht Hospital. then prenatal care, infectious disease prevention, routine prenatal examination were the most useful way to prevent neonatal death.

**Keywords:** Kuhdasht, Neonatal death, NMR

### A survey on laboratory tests in neonatal sepsis and their application in treatment hospitalized patients in the neonatal ward, Ekbatan Hospital, Hamedan, 2004-2005

*Teymouri, R., Ph.D., Pediatric Neurorehabilitation Research Center, University of Social Welfare &*

*Rehabilitation Sciences, Teymouri, F., MD, Kurdistan University of Medical Sciences, Bijar Health Network*

**Background:** Neonatal sepsis is referred to as any type of bacterial infection with positive blood culture during the first month of life. It is one of the main causes of admission at neonatal wards and neonatal intensive care unit (NICU) and it still remains as one of the main reasons of mortality and morbidity, despite advent of new effective antiseptic. The aim of this study is to evaluate laboratory tests in neonatal sepsis and their application in the treatment of hospitalized patients in neonatal ward and NICU.

**Methods:** This descriptive cross-sectional study comprised 88 neonates admitted at neonatal ward or NICU, Ekbatan Hospital with neonatal sepsis from 2004 to 2005. Basic workup included CBC diff, Plt, ESR, CRP, blood culture (BC), urine analysis, urine culture and lumbar puncture. The results of mentioned laboratory tests entered the questionnaires. Gestational age, neonatal age, sex, birth weight, and treatment duration were extracted from the medical records. Data were statistically analyzed by SPSS 13.

**Results:** 56.8% out of 88 neonates were boys and 43.2% were girls. 18% neonates were preterm and 82% were term. Early onset sepsis was found in 39.8% and late onset sepsis was found in 60.2% Blood, CSF (cerebro spinal fluid), and urine culture were positive in 9 (10.2%), 2 (2.3%), and 8 (9.1%) patients respectively. The most common first manifestation was poor feeding which was seen in 41% neonates. From 9 patients with positive blood culture, 11.1% had leukocytosis. Leucopenia, neutrophilia, neutropenia, thrombocytosis and thrombocytopenia were found in 33.3%, 11.1%, 22.2%, and 44.5% respectively. CRP was positive in 77.8% patients. ESR was not increased in any cases of positive blood cultures. The mortality rate was 10.2% generally.

**Conclusion:** Neonatal sepsis is a disease with a wide spectrum of non specific symptoms and signs, which is confirmed by positive BC. The results of this study showed that there is no specific sensitive (100%) laboratory test for the diagnosis of neonatal sepsis and only positive CRP, abnormal PLT and WBC counts may be helpful in the diagnosis of definitive neonatal sepsis cases.

**Keywords:** Neonatal sepsis, laboratory tests, neonatal ward

### Inanition Fever in Neonates Referred to Tabriz Children's Hospital

*Ghalehgholab Behbahan, A., Farhangmehr, T., Tabriz University of Medical Sciences*

**Background:** Fever due to dehydration in the first days of life is called "inanition fever" and is associated with electrolyte disturbances including: hypernatremia and indirect hyperbilirubinemia that may result in serious

and permanent sequelae. Exclusive breastfeeding as a cause of low fluid intake, over-clothing and over-heating may lead to inattention fever in a newborn infant during the first few days after birth. This study evaluates clinical and laboratory findings of neonates hospitalized with fever and dehydration to uncover their associated risk factors.

**Methods:** In a cross-sectional and analytical-descriptive study on 300 febrile neonates who were consecutively referred to Tabriz Children's Hospital, from April 2007 to September 2009 (after exclusion of sepsis and surgical problems). Needed data on admission, including: body temperature, weights at birth and admission, type of delivery and feeding, beside serum sodium, bilirubin, glucose and calcium levels were collected through file review. Chi-square statistics and Independent samples t-test were used.

**Results:** We studied 300 febrile neonates (66% males and 34% females) at the mean age of 6.16 (1 to 28) days; hypernatremia was found in 27 (9%) of them. Vaginal±5.39 delivery (P=0.009) and being term (P=0.001) were significantly more frequent in hypernatremic cases. The birth weight (P=0.031), percentage of weight-loss after birth (P=0.003), body temperature (P=0.007) and serum levels of direct (P=0.037), indirect (P=0.029) and total (P=0.031) bilirubin all were significantly higher in hypernatremic than non-hypernatremic cases; but reversely, respiratory rate (P=0.03) and calcium serum level (P=0.033) were significantly higher in non-hypernatremic neonates. Moreover, we found 9 (3%) dehydrated neonates. The frequency of hypernatremia (P<0.001), the age of newborns at admission (P=0.012), their body temperature (P<0.001) and heart rate (P=0.04) were significantly higher in dehydrated than non-dehydrated cases; but other studied variables made no significant difference between dehydrated and non-dehydrated neonates. Type of feeding (including breastfeeding) and serum level of glucose showed no significant correlation with dehydration or hypernatremia in our cases.

**Conclusion:** All febrile neonates who have tachycardia, jaundice, or weight loss of more than 10% of birth weight, should be evaluated for hypernatremia.

**Keywords:** Inattention, Fever, Dehydration, Hypernatremia, Neonates

### The Relationship between Maternal Environmental Tobacco Smoke Exposure during Pregnancy with Anthropometric Parameters of the Newborn

*Sadat, Z., Bazarganipour, F., Sehat, Z., Barati, M., Kashan University of Medical Sciences*

**Background:** Maternal environmental tobacco smoke exposure (ETS) or passive smoking would be a risk factor for maternal and new born morbidity. This study was conducted to determine this relationship with in

pregnant woman referring to Shabihkhani Maternity Hospital of kashan, Iran, during 2006-2007.

**Methods:** The study was carried out on 150 Nonsmoking pregnant women who were exposed to ETS greater than or equal to 5 cigarettes per day smoked by others in the mother's presence in all of pregnancy period as exposed group and 150 Nonsmoking pregnant women who were not exposed to ETS as the non-exposed group. Birth weight, height and head circumference were compared in the two groups and analyzed by t- and X<sup>2</sup> tests.

**Findings:** The mean birth weights in the exposed group and non exposed group were and 3137.5±437.8 and 3270±499.5 grams respectively, the t- test showed this difference to be significant (p=0.015). The mean birth height and head circumference in the two groups were not significantly different.

**Conclusion:** Nonsmoking pregnant women who were exposed to ETS gave 133 g reduction in birth weight compared to unexposed women. Further studies are needed to determine if ETS decreases birth height and birth head circumference

**Keywords:** Birth Weight; Body Height; Birth Head Circumference; Tobacco Smoke Pollution; Pregnancy

### Analysis of some maternal risk factors in preterm delivery

*Bayat-Mokhtari, M., MD, Tabrizian., F., MD, Dolatabadi, E., MD; Khalesian, B., Islamic Azad University of Mashhad*

**Background:** The aim of this study is to determine some of the maternal risk factors that lead to preterm delivery and with recognition of these risk factors, it is possible to prevent the preterm labor; which is the most common cause of neonatal morbidity and mortality.

**Methods:** This study is descriptive-analytical research. Our data had been collected from registered information in files of mothers and preterm neonates, that had been born in 22 bahman and 17 shahrivar hospitals, mashhad at 2007-2009. The case group consist of 360 preterm delivery (before 37 weeks of gestation). Control group consist of 360 term delivery (after 37 weeks of gestation). In this study, some of maternal risk factors leading to preterm labor such as maternal age, previous abortion, numbers of previous pregnancies, preeclampsia, diabetes, vaginitis, UTI, premature rupture of membrane, were extracted from registrations records. Two groups were compared and data analysis performed with SPSS software and chi-square test.

**Results:** In this study, the prevalence of preterm labor was 6.1%. Maternal risk factors such as diabetes, UTI, maternal age, number of previous pregnancies, gender of fetus, had not significant p-value, but we found significant p-value for risk factors such as vaginitis, PROM, preeclampsia, previous abortion, number of fetuses.

**Conclusion:** With prevention of maternal genitourinary infections, premature rupture of membrane and early diagnosis and treatment of preeclampsia and prevention of repeated abortions and iatrogenic multiple gestation with inappropriate induction of ovulation, we can reduce the incidence of preterm labor and then morbidity and mortality of preterm neonates.

**Keywords:** Preterm labor, Maternal risk factors, Prevention

### **Transcutaneous feeding: The effect of massage with MCToil on weight gaining in preterm newborns**

*Saeidi R., MD, Patovi S, Kianifar H., MD, Gholami, R., Robatsangi M, Ghorbani Z; Mashhad University of Medical Sciences*

**Background:** Transcutaneous feeding is suggested for Very Low Birth Weight infants. MCToil provides a source of medium chain triglycerids and highly efficient cellular food. Objectives: This study evaluates the effect of massage with coconut oil on weight gaining in preterm newborns.

**Methods:** This Clinical trial study was conducted from Aug 2007 to Mar 2008 in NICU of Ghaem Hospital in Mashhad medical university. 121 newborns were randomly assigned to three groups: In group A: Massage with MCT oil was given by a trained person, under controlled conditions of temperature during 5 minutes, 4 times a day for 7 days. In group B: newborns received only massage during 5 minutes, 4 times a day for 7 days and in group C: there was no intervention. Weight of newborns was measured daily during study in three groups by a nurse who was blinded to study. Data were analyzed by spss 14. Analysis Of Variance, Kruskal-wallis and Paired t-test were performed to evaluate the effect of massage with coconut oil on weight gaining.

**Results:** There was a significant difference between three groups in weight gaining after intervention ( $p=0.001$ ). A and C ( $p=0.005$ ) is significant but there was not a significant difference between groups B and C ( $p=NS$ ).

**Conclusions:** Massage with MCT oil has a positive effect on weight gaining in preterm newborns.

**Keywords:** Growth, MCT oil, preterm newborn

### **The study of infants icterus hospitalized in Kalaleh Hospital at the first three month of 1389**

*Salahi, R., MD, Malekzadeh, M, MD, Fadaei, E., MD, Cherabin, A., MD*

**Background:** the icterus is the most common cause of repeated hospitalization of infants after permission

from the maternity which its premature recognition according to the creating causes can lead to decrease the icterus complication. The major causes of icterus billirubin are incompatibility in RH, ABO system, secondary groups of blood, decreased enzyme of G6PD, infection, immature infant, hypothyroid, lack of evolution in hepatic system, physiological icterus, ... This study accomplished in order to discuss the relationship between the amount of blood billirubin finiculus with the mothers blood group and icterus causes.

**Methods:** The type of study was analytic and carried out on infants whom hospitalized in Hazrat-e-Rasool Hospital of Kalaleh at first three month of 1389 because of icterus. Two cc siterated and colded blood were gotten to determinethe blood groups and the amount of billirubin in finiculus and mothers blood group pulled out from their file and was analyzed. The CBC test carried out by the hematology Sysmex set, the G6PD enzyme by the way of florescent and the amount of billirubin through of billirubinmetry. **Findings:** Among 74 infants hospitalized for icterus, 9 infants were put aside from the study because of deficiency in the file of mothers pregnancy period. 43.07% were in O blood group, 26.15% in B blood group, 20% in A blood group, and 10.76% in AB blood group. 6.15% have blood incompatibility, 9.23 lack of G6PD enzyme, 84.23% related to the other causes of icterus (lack of evolution in hepatic system-immaturity and so on), the most amount of billirubin were observed in the mothers with the O blood group.

**Conclusion:** This study showed that the billirubin of finiculus in the infants born from the mothers with the O blood group was at a higher level. So the infants with the O blood group mothers must be looked after and followed more carefully after permission from the maternity for affection to the icterus.

**Keywords:** the blood of finiculus-billirubin-blood group-infant

### **The macrosomic frequency and its reasons on the born infants by caesarean operation in Hazrat-e-Rasool Hospital at Kalale Township**

*Salahi, R., MD, Malekzadeh, M, MD, Cherabin, A., MD, Fadaei, E., MD*

**Background:** Macrosomy is known to infants weighing at least 4000g or more. The amount of microcosmic break-out in the natural pregnancies reported about 8 to 12, it was in Tehran 3.8% and in Finland 3.4%. The break-out and epidemiology is different in various areas and the frequent studies have used of caesarean freely for ending pregnancy in the macrosomy. The current research carried out in order to study the fetal macrosomic break-out in the caesarean patients so that we can take an influential step to decrease the side effects and promote the mothers and infants health.

**Methods:** The type of study was sectional-descriptive and analyzed the mothers who resorted to the operation room of Hazrat-e-Rasool Hospital from Farvardin 88 to the end of year where the mothers with infants weighing 4000g and more were studied. Then the data analyzed using spss software and descriptive statistics.

**Results:** Among the whole caesarean infants the macrosomic break-out was 9% in a year. The 21-30 years old mothers were of 86.6% macrosomic infants and the first and second pregnancy number was 73.3%. The lack of progress and dense meconial are of common causes at caesarean operation. The pregnancy age was 78.98%, 38 weeks and higher, the weight of macrosomic infants with the average of 4150g, 66.6% the male sexes and the first minute apgare was less than 8.

**Conclusion:** The macrosomy considered among the side effective diseases and has the probability of bad effects on the mother and fetus. Regarding to the research result it is suggested in women under 30 years old and mostly in the common first and second pregnancies, in the sex of boy and the first minute low apgare which needed to revive immediately after the operation and these considered among the risky-full infants: the mothers training during pregnancy period, and providing certain cares for the child birth in this group respecting to bad effects on the infant at the birth time need to special attention.

**Keywords:** macrosomy-caesarean-Kalale

### Is Ceftizoxime An Appropriate Surrogate for Amikacin in Neonatal Sepsis Treatment

*Eslamieh, H., MD, Alizadeh-Taheri, P., MD, Salamati, P., MD*

**Background:** Neonatal sepsis, a life-threatening condition, presents with non-specific clinical manifestations and needs immediate empirical antimicrobial therapy. Choosing an appropriate antibiotic regimen covering the most probable pathogens is an important issue. In this study we compared the effectiveness of ceftizoxime and amikacin in the treatment of neonatal sepsis both in combination with ampicillin.

**Methods:** In a randomized clinical trial, all term neonates with suspected sepsis referred to Bahrami hospital during March 2008 to March 2010 were evaluated. Patients were randomly recruited into two groups; one group receiving ampicillin and amikacin and the other ampicillin and ceftizoxime. Blood, urine and csf cultures, leukocyte count and C-reactive protein level were performed for all neonates.

**Results:** A total of 135 neonates were evaluated, 65 in amikacin group and 70 in ceftizoxime group. 60 neonates (85.7%) in ceftizoxime group and 54 neonates (83.1%) in amikacin group responded to the treatment (P-value = 0.673 and  $\chi^2 = 0.178$ ). Only 24 (18%) blood

samples had a report of positive blood culture. The most frequent pathogen was coagulase negative staphylococcus with the frequency of 58.32% of all positive blood samples. Conclusion: Ceftizoxime in combination with ampicillin is an appropriate antimicrobial regimen for surrogating the combination of ampicillin and amikacin to prevent bacterial resistance against them.

**Keywords:** antibiotics, neonatal sepsis, coagulase negative staphylococcus, Neonatal Intensive Care Unit (NICU)

### Neonatal jaundice in neonates hospitalized for indirect hyperbilirubinemia in Besat and Fatemeh hospital, Hamedan

*Basiri, B., Sabzehei, MK., Hamedan University of medical Sciences*

**Background:** Jaundice is a common problem in neonatal period that can be associated with potential side effects. Study about predisposing factors of neonatal hyperbilirubinemia would be valuable to identify strategies for risk prevention and better management.

**Method:** This is a prospective descriptive – cross sectional study was performed from November 2007 to August 2008 in Besat and Fatemeh hospitals in Hamedan. All infants from 1-28 days of life who admitted for indirect hyperbilirubinemia were included. Infants with incomplete data were excluded. Data were described using spss.

**Results:** The subject group included 143 (61.4) male and 90 (38.6) female. The prevalence of prematurity, ABO incompatibility, Rh- incompatibility, breast-fed infants, G6PD- deficiency, cephalhematoma, echymosis, hypothyroidism, sepsis, polycythemia, previous sibling with icter and previous admission was: 60.1% (n=140), 2.1% (n=5), 0%, 60.1 (n=140), 5.2% (n=12), 11.2% (n=26), 2.5% (n=6), 9.01% (n=21), 6% (n=14), 0%, 11.6% (n=27) and 9.9% (n=23). The mean of total and conjugated bilirubin, age of icter onset, age at admission and maternal age was: 18.63±4.78, 0.51±0.41mg/dl, 3.43±1.78 ds, 5.1 ±3.06 ds and 27.56±5.09 ys. Most of neonates were fed in their first 2 hours of life (62.7%, n=146) and had meconium passing in the first 24 hours (70.4%, n=146). There was first trimester hemorrhage, diabetes mellitus, hypertension, PROM, NVD and labour complications in: 12.4% (n=29), 0.9% (n=2), 4.3% (n=10), 10.7% (n=25), 54.1% (n=126) and 6% (n=14). Significant weight loss was not seen in 85% (n=198) and 89.7% (n=209) of neonates were not visited for icter before admission. Most of neonates (61%, n=142) were discharged in the first 48 hours and 73.4% (n=171) had TSB in high risk zone. Phototherapy and blood exchange were done in 88% (n=205) and 0.9% (n=2) as the first therapy.

**Conclusion:** We recommend more attention to neonates who are breast – fed, have previous sibling with icter, have cephalhematoma and echymosis and with A+ blood group, because it is possible that have higher chance for developing significant indirect hyperbilirubinemia. Besides neonates of mothers with DM, HTN, PROM, first trimester hemorrhage, O blood group and who have NVD or complications of labour are probably at higher risk of hyperbilirubinemia. Performing the G6PD and thyroid test and sepsis evaluation are valuable in determining the cause of icter in neonates. Early discharge (48 hours) and neglecting follow up are associated with rehospitalization of neonates with TSBs mostly in high risk zone.

**Keywords:** Neonatal jaundice – neonatal risk factors – maternal risk factors

### **Anthropometric measurements at birth as predictor of low birth weight**

*Sajjadian, N., MD, Department of Neonatology, Shariati Hospital, Tehran University Medical Science*

**Object:** Low birth weight neonates are responsible for two – third of neonatal mortality in developing Countries where direct weighting at birth is not feasible especially when birth happens at home. So easier measurement of other anthropometric measurements may define the high risk neonate.

**Methods:** A cross sectional study was carried out in SHARIATI Hospital in Tehran, between September and February 2008-2009. All Consecutive full-term. Single ton, live born babies were included and anthropometric measurements carried out within six hours after birth-by authors.

**Results:** 500 newborn studied. 52.2% were male and 47.8% were female. The mean birth weight was 3195.4±399.9 gram. And 3.8% of newborns were LBW. Anthropometric value had differences between gender ( $p<0.05$ ). The maximum Correlation were between chest circumference followed by mid arm circumference ( $r=0.70$ ) and birth weight. The minimum Correlation was between mid arm to head circumference ratio ( $r=0.4$ ) and birth weight. The optimal cut off-points for chest circumference to identify LBW newborns were  $\leq 31.25$ cm and  $\leq 10.25$ cm respectively.

**Conclusion:** anthropometric values are simple, practicable, quick and reliable indicator for predicting LBW newborns in the community and can be easily measured by paramedical workers in developing nation.

**Keywords:** Low birth weight, Anthropometric measurements, Neonatal mortality

### **Stroke in a child with type IV sensory autonomic neuropathy: a coincidence or**

### **complication?**

*Barzegar, M, Shoaran, M., Pediatric Health-Research Center, Tabriz University of Medical Sciences*

Sensory autonomic neuropathy Type IV (Congenital Insensitivity to Pain and Anhidrosis) is a rare syndrome inherited as an autosomal recessive trait. This type of hereditary neuropathy is characterized with episodes of hyperpyrexia, anhidrosis, insensitivity to pain and developmental delay. These patients often have self-amputated fingers and some kinds of self-mutility behaviors. A six-year -old male child was admitted with left hemiplegia. He had unexplained recurrent fevers constantly especially during summer months, There was a global delay in the developmental milestones, after his teeth started to grow in, he began to chew his fingers and bit off the tip of his tongue. The patient responded well to touch sensation but pain and temperature sensations were absent. In recent admission Routine hematologic and chemistry measurements were normal. Negative or normal results were noted in uric acid, serum glucose, cholestol, triglycerides, ceatine kinase, liver, renal, thyroid and parathyroid function tests and lumbar puncture. Nerve Conduction Study findings were normal but somatosensory reflex (SSR) were unobtainable compatible with a sensory-autonomic neuropathy. Intradermal injection of 0.05 mL 1:1000 histamine solution gave rise to the expected wheal without axon flare. Iodine-starch test revealed no sweating. The parents did not give consent to sural nerve biopsy, genetic analysis was not available. Brain CT scan showed a large hypodense lesion in left temporo-parietal lobe suggestive of severe ischemia. By our knowledge our 6-year- old patient is the first case, of the association of HSAN IV with stroke.

### **The risk factors of hearing loss among Iranian deaf children**

*Amirsalari, S., MD, Ajallouian M., Baqyatallah University of Medical Sciences*

**Background:** The main aim of the present study was to demonstrate the risk factors of profound bilateral severe sensorineural hearing loss among some Iranian children candidate for cochlear implantation.

**Methods:** Our study was designed to collect information about profound hearing impaired cases referred from all over the country through the ENT services to Baqiyatallah Cochlear Implantation Center. A total of 310 children with diagnosis of profound hearing impairments, aged from 6 months to 4 years were admitted to this study.

**Results:** Risk factors were obvious in 218 (70.3%) patients with profound hearing loss but was Unknown

in 92 (29.7%) . 103 (33%) children had one or more close deaf relatives in their family, so they were considered as hereditary deafness cases, followed by prematurity 32 (10.3%), syndromic cases 25 (8%) , TORCH infections 12 (3.9%), severe hyperbilirubinemia 9 (3%), eruptive infections 9 (3%) , meningitis 8 (2.6%), asphyxia 6 (2%) and oto-toxic drugs 6 (2%).

**Conclusion** The present study was an attempt to identify risk factors of severe to profound hearing impairment in Iranian population. We concluded that the most common risk factor is having a positive family history of deafness.

**Keywords:** Risk factors of hearing loss, Deaf, children

### Question and answer in newborn gastroesophageal Reflux

*Soltanzadeh, MH., MD, Shahid Beheshti University of Medical Sciences*

GER is seen in up to 50 % of infants with recurrent emesis. In this session we discuss about GER in neonate. It is clinically evident in 50 % of infants in 1st 3 months of age-Only 5-10 % of children have reflux at the age of 1 yr of life There is gradual resolution of vomiting by the age of 1-2 yrs of life-If regurgitation has not resolved by 24 months of age-Further evaluation is recommended.

the red flags of GER disease are Bilious vomiting and Hematemesis. These findings should suggest an alternative diagnosis to GERD.

### Comparing micronutrients concentration before and after blood exchange transfusion in newborns by neutron activation analysis method

*Khatami, SF., MD, Parvaresh, P., Shakerian-Ardekani, Th., Gharib, M., Mashhad University of Medical Sciences; Department of Physics Payame Noor University; Iran Atomic Energy Organization*

The existence of various elements in human body, especially 17 trace elements and their effects on body metabolism is very important. Investigating the amount and variation of these elements is usually the main focus of many researchers. Also, the blood exchange transfusion can make some changes on trace elements in icteric newborns. There are two reasons for paying attention to the case in this study. One is the abundance of blood exchange transfusion and easy access to newborn's blood in Iran and the second is the importance of NAA method in analyzing trace elements. The present study, as a sort of pioneer study, has been done in the National Research Laboratory of Payame Noor University (PNU) by the cooperation of Iran Atomic Energy Organization (IAEO) and Tehran University of Medical Sciences (TUMS) during the year 2007. Three elements such as iron, selenium and zinc, has been measured by the above mentioned method and the results will be reported. The outcomes indicate the existence of the three elements on blood both before and after blood exchange transfusion. Also there are some indications of some significant changes in the three elements after transfusion.

**Keywords:** micronutrients, blood exchange transfusion, neutron activation analysis method, newborns

## Nephrology

### Randomised trial comparing short and long intravenous antibiotics in children with acute pyelonephritis: DMSA scan evaluation at 6- month follow-up

Ataei, N., Safaian, B., Madani, A., Esfahani, ST., Ataei, F.; Department of Pediatrics, Children's Medical Center, Tehran University of Medical Sciences

**Background:** Urinary tract infection (UTI) is a common infection in infants and children. It may lead to irreversible changes in renal parenchyma and long term complications. In pediatrics patients with febrile UTI the administration of intravenous antibiotics is often recommended. However, there is no general agreement regarding the duration of intravenous treatment, ranging from a few days to more than two weeks. A potential advantage of prolonged parenteral treatment might be that it would decrease the frequency of renal scar formation. The aim of this prospective study was to compare the prevalence of renal scarring following initial treatment with antibiotics administered intravenously for 14 or 3 days.

**Methods:** In a prospective study (November 2005 to March 2007) 73 children [ 67 girls (91.8%) and 6 boys (8.2%)] aged 3 months to 12 years with positive urine culture and acute renal lesions on initial DMSA scan were randomly assigned to receive intravenous ceftriaxone (75 mg/kg in two divided doses) for 14 days or 3 days, followed by oral cefixime (4 mg/kg/bid) to complete a 14 days course. After six months, scintigraphy was repeated in order to diagnosis renal scars. All children studied with DMSA scan and ultrasonography within 3 days of admission. Investigations were completed by voiding cystourethrography to detect vesicoureteric reflux when urine culture became negative.

**Results:** Renal scarring developed in 19.2% of the 37 children in the 14 days intravenous group and 24% of the 36 children in the 3 days group. Of 29 kidneys with reflux, 28 (96%) were found to have abnormal renal scan. Among 115 kidneys with non-refluxing ureters 99 (86%) revealed parenchymal changes on renal cortical scintigraphy ( $P > 0.05$ ). Children between 1 to 5 years had more renal scarring than infants and group older than 5 years [38 (52%), 18 (24.6%) and 17 (23.3%) , respectively]. After adjustment for age, sex, delay of treatment (>7 days) degree of inflammation, presence of vesicoureteral reflux there was no significant difference between the two treatment groups on renal scarring ( $P=0.3$ ). Also there was no significant difference in creatinine clearance (CCr) at the first and six months later between two treatment groups. In final analysis rate of remission on DMSA scan was significantly high in both groups ( $P=0.001$ ).

**Conclusion:** In children with acute pyelonephritis initial intravenous treatment for 14 days. compared with 3 days, dose not significantly reduce the development of renal scar formation process. Additionally, we found that an antibiotic course of 3 days is safe when powerful antibiotic such as ceftriaxone is used

**Keywords:** Acute pyelonephritis, Children, DMSA scan, Vesicoureteral reflux, Scar

### A case report of bladder exstrophy in 4 years old girl complicated with bilateral vesicoureteral reflux in a 4 years follow up

Mohseni, N., Kajbafzadeh A., Mortazavi, H.; Children's Medical Center, Tehran University of Medical Sciences

A case report of bladder exstrophy in 4 years old girl complicated with bilateral vesicoureteral reflux in a 4 years follow up Background: The bladder exstrophy is a rare cloacal membrane defect with failure of fusion of the lower abdominal wall, the symphysis pubis, urinary tract and the external genitalia to different degrees such as exteriorization of the pelvic viscera on the abdominal surface, inferiorly displaced umbilicus and abnormal exterior genitalia. Complete bladder exstrophy and epispadias with concomitant separation of the pubic bones is the most common in clinical cases. bladder exstrophy is rare, occurring once in 25 000–50 000 births, with a 2 : 1 male to female ratio. There is some evidence that genetic factors may play a role to this pathology and other cloacal malformations. Case presentation: The Case was a 4 years old girl, born after a normal term pregnancy with a normal birth weight. She was the child of a healthy mother. On physical examination all the characteristic features of bladder exstrophy were present. No other pathological findings was found. Neck of bladder was open in cystoscopy. The patient underwent the excision of exstrophic bladder through a successful operative process. 4 years after the surgery the patient referred to children's medical center with the chief complaint of incontinency. A kidney, ureter, and bladder (KUB) x-ray showed evidences of exstrophic bladder and grade 2 of bilateral vesicourethral reflux. ureteric orifices was abnormal (stadium or the golf hole). There was no evidence of trabeculation, stenosis and/or diverticulum and the bladder was in normal size. The length of the duct was about 10 cm. After the assessments through cystoscopic procedure the diagnosis changed in to the presence of short urethra and bilateral Vesicoureteral reflux (VUR). The patient was discharged with good general condition and further surgery was

recommended in order to treat Vesicoureteral reflux.  
**Conclusion:** Nowadays, death from exstrophy alone is uncommon and improvement of surgical techniques have made it possible to cure children with this abnormality.

**Keywords:** Bladder exstrophy, Vesicoureteral reflux, VUR

### Prevalence and symptoms of Idiopathic Hypercalciuria in primary school children of Rasht

*Safaei-asl, A., MD, Guilan University of Medical Science*

**Background:** Hypercalciuria is defined by urinary calcium excretion more than normal rate for age. Hypercalciuria causes many of urinary symptoms like hematuria, frequency, dysuria, UTI and et. It will be silent in children. For determine the prevalence and clinical symptoms of Hypercalciuria in primary school children in Rasht, we did a descriptive and cross – sectional study. The aim of this study was to determine the prevalence of idiopathic hypercalciuria (IH) in school children in Rasht.

**Methods:** This study was descriptive and crosses – sectional we had 30 primary school. We evaluated 340 primary school children (age 6-11, mean 9.1 years) in two steps: first (Screening test), we measured urine calcium to urine creatinine ratio (UCa/UCr) and in the second step (Definitive test), for those children who had UCa/UCr ratio more than 0.21 mg/mg we measured 24 hours urine calcium excretion. Children with secondary forms of hypercalciuria were excluded from the study. In the end of study we take a good history and did physical examination, kidney sonography and necessary laboratory exam to determine the cause of hypercalciuria

**Results:** Of 340 children, 180 were males and 160 females. the mean age of patients were 9.3 years old. In the first screening, 47 (13.8%) children (26 males, 21 females) had an abnormal UCa/UCr ratio. But in the end only 19 had the criteria of IH, i.e. the prevalence of IH was Prevalence of Idiopathic Hypercalciuria in primary school children of Rasht 5.6%. The prevalence in females and males was 3.3% and 2.3%, respectively. Of these children 3 had hematuria (including 2 cases of gross hematuria), 5 children gave a history of recurrent abdominal pain, 3 children suffered from dysuria and 12 persons had a history of personal or familial (first or second degree) urolithiasis.

**Conclusion:** Prevalence of IH in our children was 5.6% and its manifestations were: hematuria, dysuria, recurrent abdominal pain, incontinence, urgency, urinary tract infections and urolithiasis

**Keywords:** Renal stone; Hypercalciuria; Hematuria; Abdominal pain; Calcium

### Non-Calculus Signs and Symptoms of Hyperoxaluria and Hyperuricosuria in Children: A Single Experience

*Beiraghdar, F., Panahi, Y., Madani, A., Jahani, Y.; Nephrology & Urology Research Center, Baqiyatallah University of Medical Sciences; Research Center of Chemical Injury, Baqiyatallah University of Medical Sciences; Department of Pediatrics, Tehran University of Medical Sciences*

**Background:** Non-calculus presentations of hyperoxaluria (HX) and Hyperuricosuria (HU) are not common. The aim of this study was to investigate the relationship of symptomatic non-calculus idiopathic HX, HU and both of them with dysuria, failure to thrive (FTT), recurrent urinary tract infection (UTI), dysmorphic red blood cells (RBCs) and abdominal pain in children.

**Methods:** A cross sectional study was done on 58 children who were aged less than 14 years with history of persistent microscopic or macroscopic hematuria with HX and/or HU, regardless of having renal calculi, between October 2007 and October 2008. The patients were divided into three groups according to the type of crystalluria (I, 10 HX; II, 20 HU; and III, 28 HX+HU).

**Results:** The common presenting symptoms were abdominal pain (63%) and dysuria (45%). FTT frequently occurred in female (68%). No significant relation was seen between the groups in terms of gender, macroscopic hematuria and recurrent UTI. We found that dysuria, positive family history, FTT, abdominal pain and dysmorphic RBCs in patients with HX were higher when compared to HU group. Moreover, logistic regression analysis showed the higher odds ratio of FTT, abdominal pain and dysmorphic RBCs in patients with HX+HU group when compared to patients with HU.

**Conclusion:** Although our study showed that non-calculus symptoms and signs of crystalluria such as dysmorphic RBCs, FTT, abdominal pain and dysuria are frequently seen in children with HX, however, further studies are needed.

**Keywords:** Hyperoxaluria, Hyperuricosuria, Non-Calculus Signs and Symptoms

### Relationship between Pathologic and Laboratory Data of Children Suffering from Hemolytic Uremic Syndrome (HUS): A Center study

*Abdollahi, A., Mehrazama, M., Hooman, N., Otukesh, H.; Iran University of Medical Sciences*

**Background:** Hemolytic uremic syndrome (HUS) is the most prevalent cause of children renal insufficiency which in many cases (90%) occurs following diarrhea.

Hemolytic microangiopathic anemia, thrombocytopenia, and renal insufficiency are main symptoms of hemolytic uremic syndrome. This study aims to consider the relationship between pathologic data of nephro-biopsy and laboratory data of children suffering from the disease.

**Methods:** This study has been carried out in retrospective, cross-sectional and descriptive procedures. For this purpose, 28 patients with an average age of 6 years suffering from uremic hemolytic syndrome referred to Ali Asghar Hospital over the last 10 years. Light microscopic data of glomeruli, arterioles, arteries, interstitial tissue, medullary vessels and tubules were evaluated. Laboratory data including hematology, biochemistry, and urinary tests were extracted from patients' files. Data were analyzed using SPSS software.

**Results:** The most prevalent damages in glomeruli were decreased capillary lumen and thickening of its wall and in arterioles were mild decrease of lumen and in artery thickening of intima and mild infiltration of inflammatory cells and mild edema in interstitial and hyperemia in vaso recta and the most prevalent pathology in tubules was the existence of cast. Significant relationship was found out between time of recovery of hematological disorders and medullary vessels congestion and reduplication of arterial inner elastic lamina and also improvement of biochemistry changes with glomerulus necrosis and leucocytes assembly in vaso recta. Arteriolar rate with creatinine serum level at discharge time was related and tubular rate with platelet count at discharging time was also related.

**Conclusion:** Biopsy is an important tool for prognosis and determination of disease intensity. There was valuable statistical relationship between some laboratory data at the time of referral and pathological data which even could influence intensity or prognosis of disease.

**Keywords:** Hemolytic Uremic Syndrome, Histopathology, Laboratory

### Subcutaneous terbutaline use in CKD to reduce potassium concentrations

*Shjari, A.; Shaheed Sadoughi University of Medical Sciences, Yazd*

**Background:** Acute hyperkalaemia is a common, potentially life-threatening problem for patients receiving maintenance haemodialysis. Patients with chronic kidney disease (CKD) can tolerate only a small potassium load, such as that from dietary ingestion, because of the inability of the kidney to excrete potassium. Maintenance of potassium homeostasis is therefore dependent on extrarenal mechanisms and intermittent haemodialysis. Extrarenal potassium balance is governed by mechanisms that include  $\beta$ -adrenergic-receptor ( $\beta$ AR) mediated stimulation,

insulin, aldosterone, acid-base balance and plasma osmolality, but some or all of these mechanisms may be impaired in patients with CKD.  $\beta$ AR agonists reproducibly decrease plasma potassium concentrations in healthy volunteers, suggesting that they might be useful for treating symptomatic hyperkalaemia in patients with CKD.  $\beta$ 2AR agonists can be administered by a variety of routes, including nebulization, metered-dose inhalation and intravenous infusion. Each route of administration has its pros and cons. The aim of the study was to assess the effectiveness of weight-based subcutaneous terbutaline dosing in reducing the plasma potassium levels of patients with CKD who require haemodialysis.

**STUDY DESIGN:** Clinicaltrial.  
**ENDPOINTS:** Changes over time in plasma potassium concentration and heart rate in response to terbutaline; and percentage change in responses from baseline.

**Method:** Fourteen patients with CKD (aged 10-15 years) on long-term haemodialysis received terbutaline 7  $\mu$ g/kg subcutaneously. Heart rates and the potassium concentration in blood samples were measured serially for 7 hours. The effects of terbutaline on heart rate and potassium responses were determined for each patient.

**Results:** Terbutaline significantly reduced plasma potassium concentrations and significantly increased heart rates during the study ( $p < 0.001$ , repeated-measures analysis of variance). The mean reduction in peak potassium concentration ( $-1.31 \pm 0.5$  mEq/L) and the mean increase of peak heart rate ( $25.8 \pm 10.5$  beats/min) relative to baseline were statistically significant ( $p < 0.001$ , baseline versus peak for both responses). No significant adverse effects were observed.

**Conclusion:** Subcutaneously administered terbutaline is an effective acute treatment for hyperkalaemia in patients with CKD undergoing long-term haemodialysis. In patients without active ischaemic heart disease, use of subcutaneous terbutaline renders intravenous access unnecessary and should be considered as an alternative to nebulized or inhaled  $\beta$ -agonists. Close cardiovascular monitoring is necessary to minimize treatment-related toxicity, as with any  $\beta$ -adrenergic receptor agonist.

**keywords:** hyperkalaemia-ckd-terbutaline

### Undescended testis among six-year-old boys in I.R.Iran - 2009

*Mahram, M., Qazvin University of Medical Sciences, Motlaq, ME., Jondishapour University of Medical Sciences, Ziaedini, H., Ardalan, G.; Ministry Of Education and Training; Taherian, F., Qazvin Health Center; Taheri, M.*

**Background:** Screening can be considered as the best way for early diagnosis of many diseases to prevent subsequent complications and imposed socio-economic costs to the communities. Some diseases such as

undescended testis have a golden time to be diagnosed and any delay may lead to irrecoverable results including infertility, carcinogenicity and some other health complications. This study performed to assess the prevalence of undescended testis in all six-year old boys over the country (I.R.Iran) in 2009.

**Method:** This study was based on a national screening program for screening six-year-old children before entering primary schools, in all the country. As a rule, all children before registration in primary schools must refer to specified assessment centers to be screened for probable diseases or disabilities by trained General Practitioners. Suspicious cases referred to related specialists for more assessment. All findings were registered.

**Results:** Out of all 955,388 children at the said age group, 484,891 were boys among which undescended testes were detected in 2261 cases (0.47%).

**Conclusion:** Regarding the golden time for treatment of undescended testis and probable serious complications of the disease, reaching this large number of involved children to age six without any diagnosis or treatment, as an alarm emphasizes on more attention of health decision makers to design earlier screening programs and physicians, especially pediatricians to examine testes in all referred boys, before expiration of golden time.

**Keywords:** Undescended Testis, Screening, Children

### The evaluation roles of hypercalciuria and hypocitraturia in children with urolithiasis in the north west of Iran

*Azarfar, A., Mortazavi, F., Rahimi, M., Mosavi, E., Maleki, M., Tabriz University of Medical Sciences*

**Background:** now the prevalence of renal stone are increasing because of the change in diet (uses high level protein) and life style. Renal stone is not a disease but it is the side effect of the other disease. in the most of the studies hypercalciuria is the most frequent metabolic Disorders in children with renal stone In this study we evaluated the metabolic factors that they may cause renal stones in children

**Method:** it is a cross sectional study. in this study we studied 56 children with renal stones in one years from north west of Iran. The mean ages of them were 2.4 years old. (Boys46%--54% girls).we wanted urine analysis about metabolic elements. we finded that only 3 of them had hypercalciuria(0.05) but 47 of them had hypocitraturia(83%) and 1 child had hyperexcretion of uric acid and non of them had oxaloria

**Result:** this study showed that in our patient are not statistical significancy about sex and age. the evaluation of metabolic elements in children with renal stone in this region of Iran showed that hypocitraturia had the more important role in producing stones than hypercalciuria

**Conclusion:** the evaluation of the level of urine citrate in patient with renal stones in this region of country is necessary. but other studies had need.

**Keywords:**hypercalciuria-hypocitraturia-renal stone

### Kidney involvement has been reported in different bacterial, viral, fungal and parasitic infections.

*Nickavar, A., Iran University of Medical Sciences*

Kidney involvement has been reported in different bacterial, viral, fungal and parasitic infections. According to better control of infectious disease, renal involvement has been reduced recently, occurs more in primary severe infections, late diagnosis of native infection with progressive manifestations and inappropriate treatment. Immunologic pathway (immune complex deposition), toxemia (endotoxemia and other toxins), ischemic damage, direct invasion, antimicrobial nephrotoxicity and combination of these are the pathophysiologic mechanisms in kidney involvement. Renal vasculature, glomeruli, tubulointerstitium and collecting system may be involved. Different pathologic features has been reported from focal or diffuse proliferative Glomerulonephritis, tubulo interstitial disorder (interstitial edema, mononuclear infiltration and tubular degeneration), MPGN, MGN, glomerular sclerosis, crescentic GN and hyalinosis. Complement, immunoglobulin, antigenic deposition and organism detection in renal biopsy is possible in some of the infections. Some of them have also characteristic pathologic features. Clinical manifestations range from urinary tract infection, renal tubular acidosis, isolated hematuria or proteinuria, hematuria plus proteinura, sterile pyuria, urinary spreading, acute nephritis, nephrotic syndrome, acute renal failure and renal transplant dysfunction. Characteristic radiologic manifestation such as renal cyst, necrosis, calcification, abcess formation (direct invasion) and obstructive uropathy has been reported in some infections. Improvement of primary infection could improve renal manifestations. Adjunctive treatments is necessary for better control. An interesting case of hydatid cyst renal involvement with a rare clinical manifestation is discussed in this lecture.

**Keywords:**kidney, infection, pathogenesis, pathology, symptoms

### Persian manna as a prebiotic

*Shakiba, M., MD, Nafeie, Z., MD, Lotfi, MH.; Shaheed Sadoughi University of Medical Sciences, Yazd*

**Background:** "Taranjabin" or Persian manna has been used for centuries as a remedy to improve infantile health in Iranian traditional medicine, it supposed by some to have been the "manna" of holy books Koran and scripture. Peribiotic are new healthy foods that improve microbial flora of GI system to have beneficial effects on host's by increasing number of bifidobacter and lactobacilli.

**Methods:** This before and after study conducted on 20 formula fed infant aged between 2-12 months in Yazd. Stool culture was taken before and after Taranjabin consumption (30 gram/ two weeks) and counted for lactobacilli, Ecoli and bifidobacter.

**Results:** Data showed significant increase in number of lactobacilli, bifidobacter & decrease in E coli count ( $P < 0.05$ ) mean while there was correlation between duration of usage and its bifidogenic effects of the manna ( $P < 0.05$ ). According to Mother's opinion their child sleep improved and less sweating took place during study, meanwhile stool was softer than previous. Four Infant developed diarrhea at the beginning of study which exclude from study they all improved without medical intervention no other complication was reported.

**Conclusion:** Taranjabin" has peribiotic effect and its effect is depending on duration of consumption, it may improve child wellbeing.

**key word :** Peribiotic, Taranjabin, Persian manna, Bifidogenic

### New Renal Function Tests in Neonates

*Otukesh, H., Hoseini Gohardehi, R., Khalesi, N., Rahimzadeh, N., Mohaghegh, P.; Iran University of Medical Sciences*

Renal function tests are common procedures used to evaluate renal function. Plasma creatinine and urea are relatively insensitive. Limitations of serum creatinine as an assessment of GFR in neonates have been attributed to effects of maternal load, tubulae secretion of creatinine, passive back diffusion of creatinine through tubules particularly in preterm infants. The serum creatinine concentration immediately after birth reflects the maternal creatinine concentration, neonatal muscle mass and GFR at the time of delivery. There is thus a practical need for an easy alternative to plasma creatinine, which would be more specific-sensitive and reliable form the analytical and clinical viewpoint. These tests include cystatin C, beta trace protein, NGAL,... Cystatin C is a nonglycosylated protein produced in all nucleated cells with a constant rate. Cystatin C is freely filtrated by glomeruli and is catabolized by proximal tubules without any tubular secretion. In neonates the cys C concentrations are not influenced by gender, weight, height gestational age and serum billirubin level. Moreover the cystatin C doesn't cross through the placenta and neonatal cystatin C concentrations does not correlate with maternal

cystatin C level. In contrast to creatinine, the cystatin C protein is not reabsorbed by immature kidney. Some authors believe that cystatin C is a better marker of GFR than creatinine in neonates and pediatric age group because it mirrors the maturation of renal function more closely. In this study we measured serum cystatin C and creatinine in 50 neonates. We used several equations for estimation of GFR by serum cystatin C and compared the GFR calculated by these equations with creatinine based GFR.

**Keywords:** neonate, renal function test, cystatin C, creatinine

### The renal stone in children

*Rahimzadeh, N., Otukesh, H., Golmohamadi, G., Iran University of Medical Sciences*

Iran is considered as a country with high prevalence of nephrolithiasis among children, hence we decided to study the epidemiology and etiology of this common disease in Iran. In this cross sectional study, 177 children with nephrolithiasis from 1382 till 1388 were evaluated. The mean age was 6.91 (S.D: 6.15) years. Male to female ratio was 1.88/1. Abdominal pain (51.9%) and hematuria (37.5%) were the main clinical presentation. FTT (5.1%) and UTI (36.4%) were other clinical manifestations. Of the 177 patients 46% had a family history of stones. RTA was seen in 6.5% of cases. Hypercalciuria (6.5%) and hyperoxaluria (5.1%) were the main metabolic disorders among our patients. 63% of patients had opaque stones. Nephrocalcinosis was observed in 5% of our patients. Stage horn stone was in 12.7%. 50% of patients treated only medically. Chronic renal failure and end stage renal disease was seen 3% and 1.5% of patients respectively.

**Keywords:** stone, children, epidemiology

### The Incidence and Outcome of Focal Segmental Glomerulosclerosis in Iranian Children

*Hoseini, R., Otukesh, H., Fershtehnejad, SM., Tahoori, A.; Shahid Beheshti University of Medical Sciences*

Focal segmental glomerular sclerosis (FSGS) is a cause of nephrotic syndrome characterized by proteinuria with and without renal insufficiency in children. The incidence of FSGS among children diagnosed with nephrotic syndrome has been increased in recent years in some studies. In contrast, some other studies did not show any increase in the incidence of FSGS. Since ethnicity seems to play an important role in the incidence of FSGS, multiple studies in different races should be done to determine disease frequency in varying demographics. To date, there has been no study of the outcome in children with FSGS and also its

frequency over several decades in Iran. In this study, we determined the frequency of FSGS in children who underwent renal biopsy over several decades in a major pediatric nephrology center in Tehran, Iran. Other aims of this study were to analyze clinical characteristics, course, resistance to steroid and other immunosuppressive medications and outcome of Iranian children with primary FSGS referred to Ali Asghar Children Hospital for the first time in Iran. Between 1982 and 2009, 716 renal biopsies were performed in Aliasghar children hospital. Eighty four children diagnosed as FSGS at Ali Asghar Children Hospital. In three periods of time we evaluated the incidence of FSGS in patients who underwent renal biopsy. Between 1982 and 1990 the incidence of FSGS was 10.1, between 1991 and 2000, this incidence was 9.2%. After 2000 the incidence of FSGS increased significantly and reached 20.5% (Pv=0.001). In our study, fifty eight patients with FSGS were followed for the mean of 5.7 yr (range: 3 months-20 years). Among these 58 cases, initial steroid resistance was seen in 47 (81.3%) and late resistance in 5 patients (8.6%). In contrast, two patients completely recovered and responded to steroid without any recurrence. Other four patients responded to steroid but suffered the recurrence. The clinical status at last visit in patients with initial and/or late steroid resistance showed that 14 patients (26.9%) gained complete recovery, while 32 patients (61.5%) were resistant to immunosuppressive drugs other than steroid. In 14 patients who recovered, nine patients responded completely to CsA. Four patients had recurrence on CsA therapy and responded completely to the combination of CsA and MMF. Additionally, one patient received MMF only and recovered. Twenty patients (20/58: 34.4%) progressed to ESRD in a mean time of 4.9 yr (range: 3 mo-12 yr) and the mean survival time of 11.45 [Standard Error (SEM) = 1.34] yr. The kidney survival rates were 90.4%, 69% and 47% at 1, 5 and 10 years of follow-up, respectively.

**Keywords:**FSGS, outcome, incidence, children

### Genetic in congenital nephritic syndrome

*Akhavan Sepahi, M., Qom University of Medical Sciences*

Congenital nephritic syndrome (CNS) is defined as proteinuria manifesting in the first 3 months of life. NS appearing later during the first year (4-12 months) is defined infantile, and NS manifesting thereafter is called childhood NS. Primary CNS is typically caused by mutations in genes encoding for components of the glomerular filtration barrier. The classical form is the Finnish type of CNS (CNF), which is caused by mutations in the nephrin gene (NPHS1) leading to massive proteinuria, hypoproteinemia and edema in the newborn period. Other known genes causing CNS are podocin gene (NPHS2), Wilms' tumor factor 1 gene (WT1), laminin  $\beta$ 2- gene (LAMB2), and PLC $\epsilon$ 1 gene (PLC $\epsilon$ 1). Glomerular Filtration Barrier: This barrier

is located in the glomerular capillary wall and comprises three layers: fenestrated endothelium, glomerular basement membrane (GBM), and epithelial cell (podocyte) layer with distal foot processes and interposed slit diaphragms (SDs). The GBM is a protein network formed by type IV collagen, laminin, nidogen and negatively charged proteoglycans. The role of GBM in glomerular permselectivity has been debated, but it is now known that a primary defect in a GBM component results in heavy proteinuria. Podocytes and SD are, however, even more important in preventing proteinuria than the GBM. The glomerular basement membrane also depicts the five important molecules in the pathogenesis of CNS. WT1 is a transcription factor important for podocyte functions. PLC 1 is cytoplasmic enzyme involved in cell signaling. Nephrin is a major component of SD and podocin is an adapter and scaffold protein for the SD components. laminin is an important component of GBM. CNF originally denoted to a severe form congenital nephritic syndrome typically seen in the Finnish newborns. The disease (also called as NPHS1) is highly enriched in Finland, the incidence being 1:8,200 live births. However, patients are reported all over the world among various ethnic groups.

**Genetics:** CNF is inherited as an autosomal-recessive trait. The NPHS1 gene is located to chromosome 19q13.1 and exon sequencing analyses has revealed two important mutations in over 90% of the Finnish patients (Fin-major and Fin-minor). Both mutations result in a stop codon and a truncated nephrin protein not expressed in SD. Several reports on NPHS1 mutations in non-Finnish patients have been published. The patients come from Europe, North America, North Africa, Middle East and Asia. Most non-Finns have individual mutations including deletions, insertions, nonsense, missense, and splicing mutations spanning over the whole gene. Missense mutations are all located within the extracellular part, and clustering to. Congenital Nephrotic Syndrome 25603 exons coding for the Ig-like motifs two, four and seven have been reported. The Fin-major and Fin-minor mutations are rare outside Finland, but enrichment of other mutations has been reported also in non-Finns. In Mennonites, 1481delC mutation is common and leads to a truncated protein of 547 residues. On the other hand, a homozygous nonsense mutation R1160X in exon 27 has been found in all Maltese cases. Importantly, six of the 16 cases with this mutation had an atypically mild disease. The same mutation has been reported in six French patients and two of them had a mild disease.

**Podocin Gene Mutations Genetics:** Mutations in the NPHS2 gene, encoding for a podocyte protein podocin, are a common cause of a steroid resistant NS (SRNS) in children and adults, accounting for up to 28% of sporadic and over 40% of familial cases of SRNS manifesting at various ages. The podocin gene mutations, however, are also an important cause of CNS. In a recent report, recessive NPHS2 mutations accounted for half of the CNS cases in 80 European families, while NPHS1 mutations were responsible for only one third of the cases.

### Joubert syndrome present as unilateral dysplastic kidney, hypotonia and respiratory problem :A case report

Malaki, M.; Tabriz University of Medical Sciences

**Abstract:** A girl eight months old who had history of asphyxia and respiratory distress immediately after birth was hospitalized in 4th month of age due to kidney infection and revealed that she is affected to a unilateral multicystic dysplastic kidney. she comes in recent admission at 8th months to emergency room because of fever, hyperpnea and apnea. In appearance she is a hypotonic girl with broad forehead, hypertelorism, depressed nasal bridge and bitemporal area, rapid vertical and horizontal nystagmus and open mouth with salivation. In spite of normal physical growth she has delayed developmental milestones. Blood gas O<sub>2</sub> saturation drops after Phenobarbital prescription; her urinary and blood tests are normal but cranial MRI has vermian agenesis and molar tooth sign. These physical and Para clinic findings suggest Joubert syndrome.

**Key Words:** Multicystic Kidney, Hyperpnea, Vermian Agenesis

### Validation of Persian version of PedsQLTM end stage renal disease module version 3 in children under 18years

Gheissari, A., Farajzadegan, Z., Salehi, F., Heidary, SM., Mazroei, A., Masaeli, A., Varni, JW., Fallah, Z., Zandieh, F.

**Background:** to evaluate the quality of life in Iranian children with ESRD, we validate the Farsi version of PedsQLTM End Stage Renal Disease Module version 3 in children with ESRD in Isfahan (2008-2010).

**Method:** To evaluate the reliability, validity and feasibility of the Farsi version of PedsQL TM ESRD module version 3 in children with ESRD ages 2-18 we followed the translation methodology proposed by developer (Forward-backward translation). A sample of 25 children with ESRD ages 2-18 (chosen by census method from patients come to Alzahra hospital in Esfahan 2009-2010) and their parents completed the questionnaires. The data after collection analyzed by SPSS 18.0 soft ware.

**Results:** In the Farsi version of PedsQL TM End Stage Renal Disease Module version 3 Cronbach Alpha s internal consistency values was 0.82 for children self reports and 0.88 for proxy reports. Constructive, criterion face and content validity were good. Missing items were less than 5%. The Farsi version of PedsQL TM End Stage Renal Disease Module version 3 was feasible.

**Conclusion:** Results show The Farsi version of PedsQL TM End Stage Renal Disease Module version

3 is suitable for pediatric health researches children with ESRD and their parents.

**Keywords:** quality of life, ESRD, children

### Renal calculi due to cystinuria in children

Fakhrossadat Mortazavi, MD Tabriz University of Medical Science, Department of pediatric nephrology

**Background:** Cystinuria is a rare autosomal recessive disorder of proximal tubular amino acid transport, characterized by excessive urinary excretion of cystine and dibasic amino acids lysine, arginine, and ornithine. Its overall prevalence is 1/7000-1/15000, with an estimated gene frequency of 1/100. Cystinuria results in considerable morbidity as recurrent stone formation. Its incidence in pediatric urolithiasis has been reported from 1% to 12.9% in different studies. First line therapy consists of high oral fluid intake and a low salt diet with urinary alkalization to a PH of 7. Sulfhydryl agents such as tiopronin, D-penicillamin can be added when stone formation is not well controlled. The aim of this study was to investigate the role of cystinuria in pediatric urolithiasis in our institution.

**Method:** Medical records of 25 children with cystine calculi who referred to Children's Hospital of Tabriz from 1999 to 2010 were studied retrospectively. Cystinuria was confirmed by measurement of cystine in urine, urine nitroprusside test or stone analysis.

**Results:** From 356 children with urolithiasis 25 children had Cystinuria (7%). Mean age of patients at the time of diagnosis of cystinuria was 13.7±7 months (4-36 months). Male to female ratio was 14/11. Eighteen patients (72%) had bilateral and 28% had unilateral kidney stones. Mean stone diameter was 11.4±5.9 mm (3-23 mm). Consanguinity was found in 16 (64%) patients. Four patients (16%) presented with acute renal failure due to bilateral urinary obstruction. Staghorn stone and pyonephrosis detected each in one patient. Twelve patients (48%) experienced at least one episode of urinary tract infection. All patients received conservative management with hydration and urine alkalization with potassium citrate. Captopril was added to treatment in 6 (24%) patients. ESWL and surgical interventions were performed in 7 (28%) and 14 (56%) patients respectively. DMSA scan revealed unilateral scarred kidney in 5 patients (20%). All patients had normal serum creatinine level at the end of follow up period.

**Conclusion:** The incidence and clinical course of cystinuria in this study was similar to literature. Even with good medical management long-term outcome may be poor due to insufficient efficiency and low compliance. All children with urolithiasis need complete metabolic evaluation for cystinuria. Key words: Urolithiasis, Cystinuria, Children

**Keywords:** Urolithiasis, Cystinuria, Children

## Neurology & Psychiatry

### Stroke in a child with type IV sensory autonomic neuropathy: a coincidence or complication?

*Barzegar, M., Shoaran, M.; Pediatric Health Research Center, Tabriz University of Medical Sciences*

Sensory autonomic neuropathy Type IV (Congenital Insensitivity to Pain and Anhidrosis) is a rare syndrome inherited as an autosomal recessive trait. This type of hereditary neuropathy is characterized with episodes of hyperpyrexia, anhidrosis, insensitivity to pain and developmental delay. These patients often have self-amputated fingers and some kinds of self-mutility behaviors. A six-year-old male child was admitted with left hemiplegia. He had unexplained recurrent fevers constantly especially during summer months. There was a global delay in the developmental milestones, after his teeth started to grow in, he began to chew his fingers and bit off the tip of his tongue. The patient responded well to touch sensation but pain and temperature sensations were absent. In recent admission Routine hematologic and chemistry measurements were normal. Negative or normal results were noted in uric acid, serum glucose, cholesterol, triglycerides, creatine kinase, liver, renal, thyroid and parathyroid function tests and lumbar puncture. Nerve Conduction Study findings were normal but somatosensory reflex (SSR) were unobtainable compatible with a sensory-autonomic neuropathy. Intradermal injection of 0.05 mL 1:1000 histamine solution gave rise to the expected wheal without axon flare. Iodine-starch test revealed no sweating. The parents did not give consent to sural nerve biopsy, genetic analysis was not available. Brain CT scan showed a large hypodense lesion in left temporoparietal lobe suggestive of severe ischemia. By our knowledge our 6-year-old patient is the first case, of the association of HSAN IV with stroke.

### Office management of Epilepsy

*Ghofrani, M., Shaheed Beheshti Medical University-Pediatric, Neurology Research Center*

**Office Management of Epilepsy:** If a clinician is supposed to manage a child with epilepsy, needs to have an understanding of different aspects of this subject. The following discussion aims to achieve this purpose. The most conservative statistics reveal that

febrile seizure affects 3% of children. Consequently, great numbers of children suffer from both febrile and afebrile seizures. Dangers and Difficulties of epilepsy 1- status epilepticus (convulsive) 2- Fear of another ictus 3-Long-term AED usage 4- Teratogenic side-effects of AED 5-Stigmatization 6- School absenteeism 7- Social limitation

**Definition:** Seizure consists of sudden and abnormal neuronal discharge manifested by changes of behavior, cognition, level of consciousness, and motor and sensory activities. Seizure may be epileptic or nonepileptic.

During convulsive seizure, glucose consumption increases and cannot be replaced. Subsequently, if lasts long, leads to brain damage. Most epileptic syndromes do not follow Mendelian laws. The exception is 3 CPS SW EEG discharges of absence seizure, which is transmitted as autosomal dominant. Recurrence of epilepsy in the family increases the risk of epilepsy in other members. In addition, the family with epilepsy is more prone to develop seizure after trauma, or any encephalopathic process.

**Principles of treatment:** 1- Choose single most effective AED. 2-Dose to be increased gradually till either seizure ceases or side-effect appears. 3- If 1<sup>st</sup> AED fails, DC it and give 2<sup>nd</sup> AED. 4-Poly therapy is allowed if several single AEDs fails. 5- Maintain equilibrium between epilepsy and AED side effects. 6-Measure AED level if: 7- Seizure persists with adequate dosage 8-New AED is added (in view of drug interaction) 9- Evidence of AED toxicity appears. When interviewing patient and/or family we should tell them: 1-Epilepsy is not necessarily a whole-life disease. 2. Usually is not a sign of brain tumor 3. Usually seizure ceases with antiepileptic drugs. Child's family may have many questions. We should volunteer their questioning and explain the matter in simple words with friendly attitude. We should discuss about: 1. Our goals of treatment; 2. How much achievable are our goals; 3. AED side effects; 4. Conceivable difficulties which we may encounter in the course of therapy; and 5. Duration of treatment. Although majority of epileptic patients enjoy good intellect, 1/5 to 1/3 suffer from some kind of CNS lesion which cause their seizure. 20% of mentally retarded children are affected with seizure disorder and one third of them are suffered from cerebral palsy. Also many children with epilepsy have learning disorder even enjoy adequate intellect. Incidence of behavior disorder is higher in epileptics which its origin is obscure. Poorer school performance is noted in epileptic children compared to healthy ones which may stem from mental defects, epilepsy per se, AED and social factors. Drug therapy is the major form of therapy for a vast majority of children with seizure disorders. Currently, the following drugs are used for the treatment: 1. Carbamazepine 2. Phenytoin 3. Phenobarbital 4. Primidone 5. Valproic acid & SVF 6. Ethosuximide 7. Clonazepam 8. Diazepam 9.

Lorazepam 10. Midazolam 11. Acetazolamide 12. Trimethadion 13. Lamotrigine 14. Gabapentin 15. Felbamate 16. Topiramate 17. Vigabatrin 18.

ACTH& corticosteroid ; ACTH and corticosteroid are used in cases of infantile spasm and refractory seizure, specially if behavior disorder complicates the problem. New AEDs like Lamotrigine and Vigabatrin may be successful to cease intractable epilepsies such as Lennox-Gastaut, atonic, myoclonic and atypical absence.

**Institution of Treatment:** Institution of an antiepileptic treatment is a serious decision that require a firm diagnosis of epilepsy.

Drug therapy should not be initiated for children with seizure of uncertain origin, for those with a single seizure (unless it is of a type observed only in chronic epilepsy) or for those with EEG abnormalities without clear-cut clinical manifestations. Children who have had two or more seizures are candidates for drug treatment. Some investigators, however, tend to withhold treatment for patients with infrequent seizures that are of limited expression.

**Choice of Drug:** The choice of drug depends, in the first place, on the epileptic syndrome, or at least on the type of seizure experienced by the patient. Any choice is of necessity arbitrary: in part because neither the efficacy nor the unwanted effects of any drug is entirely predictable.

### **Development of Infants Conceived Assisted Reproductive Techniques in Royan Institute**

*Mozafari-Kermani, R., Nedaeifard, L., Nateghi, MR., Shahzadeh-Fazeli, A., Gorabi, H.,*

*1. Child Health Research Department of Iran Medical Sciences Branch of Academic Center for Education, Culture and Research; 2. Genetic of Royan Institute for Reproductive Biomedicine; 3. Associate professor of genetic of Royan Institute for Reproductive Biomedicine*

**Background:** Any study has not been done in Iran about development of infants of Assisted Reproductive Techniques (ART) even in comparison with infants of normal conception (NC). This study is a comparative study for developmental assessment of these infants.

**Methods:** In one case- control study 400 ART ' infants have been compared with 420 NC ' infants. Their developments have been assessed two times with test of Denver II until 9 months old. All of them were citizen of Tehran and assessed in one center. They have been adjusted for duration of pregnancy and delivery result. Chi- Square test and Wilcoxon Singned Ranks test were used for analysis of data.

**Results:** From 400 infants of ART, 31/3% was preterm and 42/8% were multi fetal pregnancy, which were higher than NC. There was not significant difference in development of infants of ART and NC in different methods of ART in two times assessment (without

concerning premature and multi fetal pregnancy) ( $P>0/005$ ). There was not significant difference between development of term and single fetal pregnancy of ART and NC in first and second time of assessment of development (p values were 0/7 and 0/4 respectively).

**Conclusion:** We concluded that there is not any difference in development of term and single fetal pregnancy in infants of ART and NC until 9 months old.

**Key words:** Assisted Reproductive Techniques, Development, Infants

### **Evaluating Preschool Children's IQ with Severe and Profound Hearing Loss After Auditory Rehabilitation**

*Mahmoudi-Rad, M., University of Social Welfare And Rehabilitation Sciences Tehran*

**Background:** Speech and language is the most common way of communication and a fundamental of learning. Hearing loss is one of the sensory impairments that prevent speech & language and other aspects of psychological development in children. The present study evaluated IQ among preschool children with severe and profound hearing loss after auditory rehabilitation.

**Methods:** To evaluate IQ among preschool children with severe and profound hearing loss after auditory rehabilitation, Goodenough Draw-a-Person and Picture Completion Tests were used. Subjects were nine preschool children with severe and profound hearing loss who had been received early auditory rehabilitation and had been followed regularly. Tests were performed on the available subjects by a child psychologist when they were 5-year old and repeated a year later. Findings: The average scores of the subjects in each two years was the same as normal children and increasing scores was significantly difference after a year. The Goodenough Draw-a-Person and Picture Completion tests' P-values were 0.008 and 0.007 respectively.

**Conclusions:** This study indicated the important role of early and continuous auditory rehabilitation on IQ among preschool children with severe and profound hearing loss.

**Keywords:** Hearing loss, Auditory rehabilitation, Intelligent quotient

### **Evaluation of developmental status of hypoglycemic neonates at the age of two years**

*Fallah, R., MD; Shaheed Sadoughi University of Medical Sciences University, Yazd*

**Background:** Hypoglycemia in neonates can cause seizures, brain injury and long-term neurological

impairments. Developmental surveillance should be performed in all children at every well-child visit and if there is developmental concerns by the parent. Parental reports screening tests are cost-effective, easy to complete and time saving. Among them, Ages and Stages Questionnaires (ASQ) is currently the most widely used and assess in five different domains: communication, gross motor, fine motor, problem solving and personal social skills by questions on what the child can or cannot do. The answer of “yes: 10 point” indicates that the child does this item, “sometimes: 5 point” indicates an occasional or emerging response and “not yet: 0 point” indicates that child does not yet do the behavior. The score on any domain below the cut-off point is considered abnormal and referral for further evaluation. The purpose of this study was to evaluate developmental status of neonates with hypoglycemia and seizure via Persian version of 24-month ASQ.

**Methods:** In a follow up study, developmental status of all hypoglycemic neonates admitted to Shaheed Sadoughi Hospital of Yazd, central city of Iran between 2006 and 2008, evaluated at the age of two.

**Results:** Twenty six boys and 15 girls with glucose level of  $30.1 \pm 2.3$  mg/dl were evaluated. Developmental delay in fine motor and problem solving domains were not seen in any child. In gross motor, communication and personal social skills, 9, 6 and 2 children had developmental delay, respectively. Birth weight less than 1000 grams and those presented after the first week of life were more likely to have gross motor and communication delay.

**Conclusion:** Prompt and timely diagnosis and control of neonatal hypoglycemic are necessary for preventing developmental delay in these children and evaluation and monitoring of developmental status of them should be emphasized for early and timely diagnosis, management and also rehabilitation.

**Keywords:** Hypoglycemia, Developmental delay, Ages and Stages Questionnaires (ASQ)

## Minor Head injury in children

*Fallah, R., MD, Shaheed Sadoughi Medical Sciences University, Yazd*

Head trauma (HT) occurs commonly in childhood however most of them is mild. Clinical features of predictors of intracranial injury after minor HT include: prolonged loss of consciousness, amnesia, persistent vomiting, seizure, severe headache, GCS <15, signs of a depressed or basilar skull fracture, bulging fontanel, focal neurologic examination, nonfrontal scalp hematoma for children <2 years of age and severe mechanism of injury [ fall >0.9 m (3 feet), motor vehicle collision with patient ejection, death of another passenger, or rollover, penetrating injury and inflicted injury ], preexisting conditions that place child at risk for intracranial injury (ventricular shunt, arteriovenous

malformation, bleeding disorder) Neuroimaging should be done in all high risk children include focal neurologic findings, depressed or basilar skull fracture, depressed or altered mental status (irritability, agitation, lethargy, repetitive questioning, or slow response to verbal questioning) , seizure, definite loss of consciousness 2 years old children with-(especially more than a few seconds), and also in bulge fontanel, persistent vomiting, suspicion of child abuse, underlying condition predisposing to intracranial injury, large nonfrontal scalp hematomas 3 months with nontrivial trauma, <12 months), infants <(especially in significant or prolonged behavioral change, vomiting that is delayed by several hours after injury or occurs multiple times, clinical deterioration, loss of consciousness less than a few seconds now resolved, injury caused by high force (fall more than three to four feet, patient ejection, death of a passenger, rollover, high impact head injury), skull fracture more than 24 hours old (nonacute). Children should be admitted in hospital if: • Brain injury or depressed or basilar skull fracture • Persistent, significant alteration in mental status despite normal head CT • Unremitting vomiting • Caretakers who are unreliable or unable to return for care • Suspected inflicted injury • Extracranial injury requiring admission Neurosurgical consultation should be done in brain injury detected by CT, depressed, basilar, or widely diastatic skull fracture and deteriorating clinical condition.

**Keywords:** head injury, children, infant

## Sumatriptan compared to Acetaminophen in the treatment of Paediatric Migraine

*Arshad, MS., Allami, SH., Azaad Medical university, Tehran*

**Objective:** To determine the safer and more effective between Sumatriptan and Acetaminophen in Paediatric Migraine headaches.

**Methods:** A total of 25 patients, ten female and fifteen male aged between 9-12 years were treated with Sumatriptan 25mg tabs s.o.s and Acetaminophen tabs s.o.s for their Migraine headaches who were reporting to Al-Shafa Hospital, Gujrat, Pakistan over a period of four months, during my administration of that hospital in the paediatrics outdoor department. The patients were followed up either on subsequent visits or by family telephone. The medicines were provided by Tehran Darou/Chemie pharmaceutical company Tehran, Iran for this study.

**Results:** Sumatriptan had lesser side effects and was more effective in relieving migraine headaches in 13 of these patients whereas 2 patients reported Acetaminophen to be better.

**Conclusion:** Sumatriptan is superior to Acetaminophen for treating Paediatric Migraine headaches.

**Keywords:** Migraine, Sumatriptan, Acetaminophen, Headache, Paediatric

### **The impact of epilepsy and its treatment on the quality of life in children with unprovoked epilepsy visiting Children's Hospital Medical Center, Tehran.**

*Mohammadi, M., Hussein, I.; Children's medical center, Tehran University of Medical Sciences*

**Background:** To study the impact of epilepsy and its treatment on the quality of life in children with unprovoked epilepsy visiting Children's Hospital Medical Center, Tehran.

**Method:** 60 children with known epilepsy were selected from the out patient department of Children Medical Center of Tehran University of Medical Sciences, Tehran, between Farwardin 1383 and Mehr 1383 (March 2004 and September 2004 AD). All subjects and their parents were asked about the quality of life assessment in a questionnaire prepared according to Child self-report scale & Parent proxy response scales which were modified according to the local culture and awareness of disease, in an interview setting.

**Result:** The sample of 60 children was predominantly male with 36 boys (60%) and 24 girls (40%) with a mean age of 10.33 years (S.D. =1.38). Out of these, 40 were from big cities (66.7%), 15 from small cities (25%) and 5 from villages (8.3%). The types of seizure distribution were Generalized Tonic Clonic Seizure (56.7%), Absence (23.3%) and others. 48 patients (80%) were being treated with one antiepileptic drug (AED), and 10 (16.7%) were being treated with two or more AEDs. Analysis of the QOLQ-20 revealed the overall QOL score expressed by patients themselves as: mean score as 27 (SD=4.66) out of total score of 40, the minimum and the maximum score as 15 and 35 respectively. The groups of QOL as graded in analysis plan were scored by both patients and parents. 18 subjects (30%), who were patients themselves, were categorized in group A (Satisfactory) of QOL scoring ( $\geq 30$  point), 28 (46.7%) were in group B (Relatively Satisfactory) (25-29), 10 (16.7%) were in group C (Dissatisfactory) (20-24) and only 4 (6.7%) were listed in group D (Poor) ( $< 20$  score). On the other hand, 28 subjects (46.7%) were categorized by parents in group A of QOL scoring ( $\geq 30$  point), 17 (28.3%) were in group B (25-29), 7 (11.7%) were in group C (20-24) and only 8 (13.3%) were listed in group D ( $< 20$  scores).

**Conclusion:** In accordance with the previous studies from the developed countries, we also found reduced QOL in children with epilepsy. However, our results show an upper trend, reflecting a better QOL in patients. We have shown that four sub-scales which include life status, worries, school performance and relationship with others, are obviously affected and around one-third of the subjects had satisfactory QOL based on child measure scale and less than half of the subjects had satisfactory QOL based on parent measure scale. In addition, the concerns that were not

significantly affected in this study included going away to picnic or similar places, comfort at school, good teacher behaviors, number of friends and well being that is explainable by previous studies. These results can guide the caring physicians with regards to the relevant concerns of patients and parents in providing better treatment and further improvement of QOL in pediatric epilepsy patients.

**Keywords:** Quality of Life, Measures, Epilepsy, Children, Parents

### **Treatments used by parents of children with autism in Tehran, Iran: Trends in biomedical approach**

*Memari, AH., MD, Kordi, R., MD, Mirfazeli, F., MS, Sport Medicine Research Center, Tehran University of Medical Sciences*

**Background:** During the last decades, the prevalence of Autism Spectrum Disorder (ASD) has been increased significantly. Although there is no definite cure for the autism, numerous treatments regarding biomedical therapies, relative services and alternative medicine have been proposed to ameliorate the developmental and behavioral problems related to ASD. New findings about biological association of autism may direct practitioners and families to use medications as a primary treatment.

**Methods:** A cross-sectional survey in autistic children between ages of 7 to 15 was employed through the autism schools to set a data base for the students. Parents provided general information about the children; through the study they were asked to answer questions about any treatments used currently and or previously.

**Results:** One hundred and fifteen children with ASD consisted of 93 boys ( $9.63 \pm 2.04$  years old) and 23 girls ( $9.82 \pm 1.69$  years old) where assigned to the study. A total of 87 various treatment consisted of 74 medications were used by families; Speech therapy was the most commonly reported intervention followed by occupation therapy and Risperidone as a medication. In biomedical treatments, Risperidone was mostly used by the parents followed by Sodium Valproate and Ritalin. Additionally, 97% of children have already used at least one drug in regard to autism syndrome and 80% of parents were currently using at least one drug to three per child.

**Conclusion:** Parents of children with ASD were using too large number of medications, many of each were not supported by empirical evidences; future research should focus on lifelong consequences of this approach in autistic pediatric population.

**Keywords:** Autism, Medication, Treatment and Parent

## Evaluating Preschool Children's IQ with Severe and Profound Hearing Loss After Auditory Rehabilitation

*Mahmoudi-Rad, M., University of Social Welfare and Rehabilitation Sciences*

**Background:** Speech and language is the most common way of communication and a fundamental of learning. Hearing loss is one of the sensory impairments that prevent speech & language and other aspects of psychological development in children. The present study evaluated IQ among preschool children with severe and profound hearing loss after auditory rehabilitation.

**Methods:** To evaluate IQ among preschool children with severe and profound hearing loss after auditory rehabilitation, Goodenough Draw-a-Person and Picture Completion Tests were used. Subjects were nine preschool children with severe and profound hearing loss who had been received early auditory rehabilitation and had been followed regularly. Tests were performed on the available subjects by a child psychologist when they were 5-year old and repeated a year later. Findings: The average scores of the subjects in each two years was the same as normal children and increasing scores was significantly difference after a year. The Goodenough Draw-a-Person and Picture Completion tests' P-values were 0.008 and 0.007 respectively.

**Conclusions:** This study indicated the important role of early and continuous auditory rehabilitation on IQ among preschool children with severe and profound hearing loss.

**Keywords:** Hearing loss, Auditory rehabilitation, Intelligent quotient

## Febrile seizure and anemia: is there any correlation

*Malek, A., MD, Afzali, N., MD, Islamic Azad University, Mashhad Branch*

**Background:** Febrile seizure (FS) is the most common neurologic emergency disorder in childhood. It occurs about 2-5% in 6months to 6 years old children. The common age of FS is similar to the age of anemia specially iron deficiency anemia. Because the anemia results to decrease in tissue oxygenation especially brain, this study was designed to evaluate relationship between anemia and febrile seizure.

**Methods:** The study was performed on 160 hospitalized children between 6 months to 6 years during one year. They were divided to two groups: with FS (Case group) and without FS (control group). Each group contains 80 persons. Hematocrit (Hct) of all patients was measured and compared with normal

standard table. Anemia was determined as Hct less than 2 standard deviation of mean values for age (with 95% confidence interval). Statistical analysis was done with version 15 SPSS and P value less than 0.05 was significant.

**Results:** Prevalence of anemia was similar in case and control groups (31.3% in each group) and there was no significant correlation between anemia and FS in this study; In case group 64 persons had simple FS and 16 persons had complex FS. The prevalence of anemia was higher in simple FS cases than complex FS. In case group anemia was detected in 37.5% of simple comparing to 6.3% of complex which was significantly different (P value: 0.016). Prevalence of anemia was not different in cases with positive history of FS comparing to cases with first attack of disease. Anemia was detected in FS boys in 28.6% and in FS girls in 34.2%; which was not significantly different (P value: 0.585)

**Conclusion:** Anemia was not significantly more common in children with febrile seizure than similar age persons. In simple FS cases, anemia was more detected than complex FS. Because of high incidence of simple FS, treatment and prevention of anemia can decrease the start and relapsing seizure attacks. The type of anemia in FS patients should be determined. Relating to high incidence of iron deficiency anemia in this age group, serum Ferritin level and Red Blood Cell indices should be considered in the next studies.

**Keywords:** Anemia, Children, Febrile Seizure, Hematocrit

## Subclinical epileptiform discharges and its effect on cognitive impairment

*Karimzadeh, P., MD, Pediatric Neurology Research Center, Shahid Beheshti University of Medical Sciences, Mofid Children Hospital*

Subclinical EEG discharges (SEDs) in children with psycho cognitive impairment are not uncommon. Subclinical discharges can occur without significant clinical manifestations. Formal testing during EEG recording can demonstrate transitory cognitive impairment (TCI). This term means an episode of epileptiform discharges happens without any clinical seizure. Short epileptiform paroxysmal discharges of few seconds in children without any clinical epilepsy are not noticed by clinical observation of children. Additionally some studies have indicated such mild epileptiform discharges can accumulate over time, when these paroxysmal persist over years and results in effects on cognitive function. About one-half of children with subclinical epileptiform discharges demonstrate transient cognitive impairment (TCI) during these discharges. Children with predominantly left-sided discharges have problem in reading and those with right-sided discharges in visual spatial task. Some studies revealed left-sided spiking often produces

problems in verbal tasks but right-sided discharges show impairment in nonverbal skills. According to some reports suppression of these discharges by antiepileptic drugs (AEDs) has remarkable improvement in psychocognitive function and development and development of children. Early detection of subclinical epileptiform discharges and treatment may prevent its effect on psychocognitive and learning development of children. Suppression of the EEG discharges with "Valproic Acid" "Lamotrigine" "Carbamazepine" and "Prednisolone" has been reported to improve cognitive performance. Author has noticeable results in suppression of epileptiform discharges with Na-Valproate Methylprednisolone and Lamotrigine. These antiepileptic drugs were effective in improving developmental-behavioral conditions of children with cognitive impairment and subclinical discharges. In these studies we treated children with psychocognitive impairment who had normal results for neuroimaging, genetic and neurometabolic investigations.

**Key words:** Subclinical EEG discharges, Transitory Cognitive Impairment, Antiepileptic drugs

### **When to exempt children from DPT immunization: A neurological point of view**

*Tonekaboni, SH., Pediatric Neurology Research Center, Mofid Children Hospital, Shahid Beheshti University of Medical Sciences*

Serious adverse reactions following immunization is extremely rare, but occasionally can result in permanent sequelae or life-threatening illness. The occurrence of an adverse reaction after immunization does not prove that the vaccine is responsible for the signs and symptoms. Vaccines are administered to infants and children during the stage of their lives when certain diseases most commonly become clinically apparent (e.g. seizure disorders). Because chance temporal companionship of an adverse reaction to the timing of prescription of a specific vaccine commonly occurs, a true causal association requires that the event occur at a significantly higher rate in vaccine recipients than in unimmunized groups of similar age and residence. Exempting children from DPT vaccination, especially pertussis immunization, should be limited to the narrow recommended contraindications: These children will be in increased risk for pertussis and also they have negative role in outbreaks of pertussis. A vaccine should not be administered when a contraindication is present. In contrast, a precaution is a condition in an individual that might increase the risk of a serious adverse reaction. However, immunization might be indicated in the presence of a precaution, because the benefit of protection from the vaccine outweighs the risk of an adverse reaction. Usually, precautions are temporary conditions and a vaccine can be administered later. A generic contraindication for all

vaccines is anaphylaxis to a prior dose. Convulsions and family history of convulsion are not a contraindication to DPT immunization. Encephalopathy within 7 day of a DPT vaccination not attributable to an identifiable cause, is a contraindication for further pertussis vaccine injection. But, fever within 48h. after a dose of DPT, collapse or shock like state within 48h., seizures within 3 day after a dose and persistent, inconsolable crying lasting for minimum 3 hours within 48h. of a dose are not a contraindication but a precaution: we should consider carefully the benefits and risks of this vaccine under these circumstances. Children with progressive neurologic conditions should not be vaccinated with DPT until the condition stabilizes. Chronic progressive neurologic conditions that are stable do not constitute a reason to delay DPT, this is in contrast to unstable or evolving neurologic conditions (e.g. cerebrovascular events and acute encephalopathic conditions). Whether and when to administer DPT to children with proven or suspected underlying neurologic disorder should be decided individually. Generally infants and children with stable neurologic conditions including well controlled seizures, may be vaccinated.

**Keywords:** DPT, Vaccination, Contraindication

### **Mental health services for hospitalized children and their parents as a part of consultation-liaison child psychiatry program in Children Medical Center**

*Mahmoudi-Gharaei, J., Delavari, M., Dehghani, F., Sadeghi-Nejad, H., Gohari, Z., Karami, E., Abniki, E., Naji, T., Talebzadeh, H.*

*1. Psychiatry and Psychology research Center, Roozbeh Hospital, Tehran University of Medical Sciences; 2. Consultation-Liaison Psychiatry Clinic, Children Medical Center, Tehran University of Medical Sciences; 3. Children Medical Center, Tehran University of Medical Sciences*

**Background:** To report the characteristics of patients seen and mental health services in Consultation-Liaison Child Psychiatry Unit of Children Medical Center.

**Method:** All the children who were admitted in emergency, rheumatology, hematology, neurology and hemo-dialysis wards, and the children who referred to outpatient CL child psychiatry clinic in Children Medical Center were included. All the children were screened for psychiatric disorders and adjustment problems. Mental health interventions at the primary level included individual and group interventions for children and parents and group interventions for hospital wards nurses. In secondary and tertiary level, psychoeducational and psychological counseling were performed by 5 trained psychologists under supervision of a child and Adolescent psychiatrist. The entire psychologists were participating in weekly supervision

sessions. The children who need special psychiatric interventions were consulted with a child psychiatrist. Qualitative statistical method was used for data analysis and report.

**Results and Conclusion:** The study is currently ongoing and the data will be presented at the meeting.

**Keywords:** Mental Health, Children, Medical illness, Liaison-consultation, Psychiatry

### **Ionized Calcium and Ionized Magnesium in Children with Seizure**

*Shams, S., PhD, Bavarian, B., MD, Ashrafi, MR., MD, Hadadi, M., MD, Haghi-Ashtiani, MT., MD, Jafari, F., Pediatrics Center of Excellence, Children's Medical Center, Tehran University of Medical Sciences*

**Background:** There are compelling physiological reasons that calcium and magnesium active forms are ionized types. The aim of this study was to compare ionized calcium (iCa) and ionized magnesium (iMg) levels in children complaining of seizure attack within 24 hours after seizure with matched control ones. We also established the reference interval for iCa and iMg.

**Methods:** 60 patients with a seizure attack who presented to Emergency Department (ED) of Children Medical Center in Tehran from April 2008 to June 2009 and 68 children as the control group included in this prospective study. Serum total and ionized Ca (iCa), total and ionized Mg (iMg) were measured in patients and controls. The concentrations of serum total Ca and serum total Mg were measured by commercial kit on Hitachi 717 autoanalyzer and serum ionized Ca and ionized Mg were measured with an ion-selective electrodes (Nova and Stat Profile critical care Xpress Analyzer). The results were compared between groups using general linear model considering age as the covariance.

**Finding:** 36 and 32 of children were male in case and control group respectively. Mean age of patients was 4.1 years while it was 5.4 years in controls which was significantly higher in controls ( $P$ -value=0.041). Total serum Ca ( $8.96 \pm 0.96$  vs.  $9.38 \pm 0.81$  mg/dl), iCa ( $4.23 \pm 0.97$  vs.  $4.77 \pm 0.37$  mg/dl) and Mg ( $2.14 \pm 0.25$  vs.  $2.28 \pm 0.17$  mg/dl) were significantly lower in the seizure group (all  $P$ -values < 0.002). Ionized Mg was lower in the seizure group ( $1.17 \pm 0.27$  vs.  $1.20 \pm 0.10$  mg/dl). However, this difference was not significant ( $P$ -value=0.397). iCa /iMg ratio was also significantly lower in seizure group ( $3.70 \pm 0.98$  vs.  $3.98 \pm 0.26$ ,  $P$ -value=0.03).

**Conclusion:** We found significantly lower serum ionized calcium levels while serum iMg level was not significantly different from normal children. These findings suggest that although the correction of iCa deficiency may result in better seizure control for some pediatric patients, the role of hypomagnesemia is

overestimated in children. Also these findings are not affected by age of patients or their seizure type.

**Keywords:** seizure-children-ionized calcium- ionized magnesium

### **A survey on the prevalence of depression among high school boy students in Hamedan, year 2008: looking for influential factors on its incidence**

*Batebi, A., MD, Pourreza, A., PhD, Rezaee Mosamam, H., MspH*

**Backgrounds:** Depression is known as a very widespread mental illness among human populations. In this respect, it is compared, most of the time, with cold. According to studies conducted in mental hospitals, about 75% of patients hospitalized due to psychiatric reasons, suffered by depression. Because of higher rates of depression among children and adolescents, which in fact are the architectures of the future of any community, studying the prevalence of depression among them and recognizing main influential factors on it seems to be very important.

**Method:** 300 students from grade 1 to grade 4 of Hamedan's high schools randomly selected as the sample of the study. A questionnaire with two separate parts was employed for data collection. The first part of the questionnaire included demographic characteristics of the respondents and the second part was Beck Depression Inventory (BDI). Collected data analyzed through SPSS and findings demonstrated by tables and figures.

**Results:** Findings revealed that 40% of the students demonstrated nothing relevant to depression. But 32.3, 10.7, 11.7, and 4/7 percent of them suffered from, mild, moderate, severe, and very severe forms of depression respectively. Further analysis showed that depression among students has developed direct significant relationship with factors such as grade, previous year rank, birth order, physical activities, past unpleasant events happened for family, and Background of mental illness ( $p < 0/05$ ). A non parametric Kruskal-Wallis test conducted for assessing the relation between depression and grade.

**Conclusion:** The present study revealed a high rate of depression (60%) among the student. Different factors have been recognized as responsible for this illness. At the same time such a prevalence rate may indicate an almost forgotten or devalued part of the health system function. For the purpose of being efficient, and saving resources, it is suggested that students at the time of registration for new year of schooling, should be screened or consulted for this very common illness.

**Keywords:** Depression, students, Beck Depression Inventory (BDI), influential factors

## Selenium & Glutathione peroxidase in Epilepsy

Ashrafi, MR., MD, Shams, S., PhD, Children's Medical Center. Tehran University of Medical Sciences

**Background:** It is not a long time since oxidative stress and its resultant free radicals are known to be the both cause and the consequence for epileptic seizures. In order to cease the increasing damage to the brain neurons following epileptic seizures, natural anti-oxidative systems are playing the main role. One of the most important of these detoxifying systems is composed of the trace element Selenium and its dependent detoxifying enzyme, Glutathione peroxidase.

**Method:** In a case-control study, 53 epileptic patients (29 females and 24 males) with a mean age of 5.5 years, were studied during a 15 months period. The control group was consisted of 57 healthy children with no history of any neurologic disease and a mean age of 5.6 years. From all children of either group, blood specimens were drawn and Serum Selenium level and RBC Glutathione peroxidase activity were evaluated and finally the results were compared and statistically analyzed.

**Findings:** Statistically significant differences in the means for Serum Selenium level ( $p$  value  $<0.05$ ) and also RBC Glutathione peroxidase activity ( $p$  value  $<0.001$ ) between the two groups were observed. On the other hand, after analyzing the study results, trying to introduce a value for GPx activity that could be accepted as a reliable indicator for Serum Selenium deficiency in patients, was not successful. **Conclusion:** Findings of this study strongly support the proposed crucial role for the trace element Selenium and its dependent enzyme, Glutathione peroxidase, deficiency in epilepsy pathogenesis first as a predisposing condition and second as a consequence of the disease that has Longley been a controversy.

**Keywords:** Selenium, glutathione peroxidase, seizure, epilepsy, oxidative stress, free radicals

## Mothers Expressed Emotion (EE) toward children with epilepsy

Khodabakhshi-Koolae, A., MD. Family Counseling Department University of Social welfare and Rehabilitation

**Background:** The expressed emotion (EE) concept (Leff and Vaun, 1985), measures of attitudes and behavioral patterns of relative toward patients (Widemann et al, 2002). Expressed Emotion including; Hostility, criticism and emotional over Involvement. Propose of this study is to identify factors with maternal expressed emotion (EE) toward their child with epilepsy.

**Methods:** 40 mothers who had a child with epilepsy and 40 mothers who had not a child with epilepsy (control healthy group) selected in present study. A total 80 mothers were answered Family Questionnaire (FQ) Persian version (Khodabakhshi Koolae, 2008). Epilepsy children were selected from welfare centers in Tehran. The data of epilepsy children collected from medical records. The mean of age of children was 11 years old.

**Results:** The results indicated family questionnaire (FQ) was dramatically increased in comparison to healthy controls. Mothers with epilepsy children reported high level of EE than healthy group. Also, 58% of mothers reports very high level EE. It is noteworthy to noted, the type of EE in mothers with epileptic children were with Hostility and emotional over Involvement.

**Conclusion:** According to the results mothers with epileptic Children experienced high level of EE toward epilepsy children. High EE in mothers with epilepsy may lead to children to relapse. Furthermore, mothers with high emotional over Involvement limited their children. It seems to be; new family education and family therapy are strongly suggested.

**Keywords:** Mothers, Expressed Emotion (EE), and children with epilepsy

## Static and Dynamic Balance Function in Congenital Severe to Profound Hearing-Impaired Children

Jafari, Z., Ph.D, Iran University of Medical Sciences Faculty of Rehabilitation Sciences; Malayeri, S., Rezazadeh, N., Ph.D, Candidates of Audiology University of Social Welfare & Rehabilitation, Audiology Department; Hajiheydari, F., MS., University of Social Welfare & Rehabilitation

**Background:** Past behavioral evaluations have revealed some balance disorders in hearing impaired children. This study was done to evaluate static and dynamic balance skills in congenital severe to profound hearing impaired children in comparison with normal age-matched children.

**Methods:** This analytical case-control study was conducted on 30 congenital severe to profound hearing impaired children (14 girls, 16 boys) and 40 normal children (20 girls, 20 boys) from 6 to 10 years old in Tehran city. Bruininks-Oseretsky Test of Motor Proficiency 2 [BOT-2], balance subset was used for evaluation of balance skills. The balance subset has 9 parts and determines effect of decrease or omission of somatosensory and/or visual inputs on balance function.

**Results:** Hearing-impaired children showed 16.7% to 100% fail results in 7 parts of the balance subset that were 2.5% to 57.5% in just 3 parts of balance subset in normal children. Mann-Whitney test showed significant

differences between two groups ( $P < 0.0001$ ). There was significant difference between two groups in time of two static balance skills of "Standing on 1 leg on a line" and "Standing on 1 leg on a balance beam" with eyes closed with t-independent test ( $P < 0.0001$ ). Sex had no effect on results of the hearing-impaired children. In normal children, Mann-Whitney test revealed effect of sex on two static balance skills ( $p \leq 0.029$ ). In hearing-impaired children, kruskal-Wallis test showed effect of age on 2 static and 1 dynamic balance skills ( $p \leq 0.031$ ), but age had no effect on results of the normal children.

**Conclusion:** It seems that development of static balance skills are longer than dynamic ones. Because severe to profound hearing-impaired children showed more weakness than normal children in both static and dynamic balance abilities, use of functional tests of balance proficiency like as balance subset of BOT-2 that has been designed to decrease redundancy of effective sensory inputs in balance function including somatosensory and visual data can help to identify balance disorders in this children. Key words: hearing loss, static balance, dynamic balance, BOT-2 test

**Keywords:** hearing loss, static balance, dynamic balance, BOT-2 test

### Acute cerebellar ataxia as manifestation of pediatric stroke

Aminzadeh, V., MD, Khaleghjoo, A., MD, Khaligi, N.; Guilan University of Medical Science

**Background:** The term ataxia denotes disturbances in the fine control of posture and movement. The cerebellum and its major input systems provide this control. The initial and most prominent feature is usually an abnormal gait (wide based, lurching, and staggering). The two most common causes of acute ataxia among children are drug ingestion and acute postinfectious cerebellitis. Migraine, brainstem encephalitis, and an underlying neuroblastoma are the next considerations. (1) Vascular disorders is a very rare cause of acute cerebellar ataxia. (1) The aim of paper is presentation of rare cause of ataxia in children (stroke) for pediatrician till through investigation of ataxia regarding to history & physical examination is performed.

**Case Report:** A 9 years old boy present acutely with unsteady gait after wake up sleep. Patient had occipital headache & vomiting since previous day. There is no history of febrile illness & any drug ingestion before this. Past medical & developmental history is unremarkable. His uncle had sudden death in adolescent period. On physical exam had normal vital sign, no neck rigidity, no papilledema. Only positive findings in neurologic exam was hypoactive deep tendon reflexes in lower extremities, slurred speech, wide base gait, dysmetria, veering to right

side & intension tremor. After initial brain CT & role out of mass lesion lumbar puncture is performed that had normal finding. Regarding to history & neurologic examination brain MRI & MRA is performed that show large right hemisphere infarction. Through stroke investigation show not any predisposing factor only vertebral artery anomaly. Acute cerebellar ataxia due to large right sided cerebellar hemispheric infarction in brain MRI & vertebral artery anomaly show with brain MRA.

**Conclusion:** In evaluation for acute cerebellar ataxia in children through history & physical examination is important. At first most common causes in pediatric must be role out (drug ingestion & post infectious cerebellitis). Then regarding to history & physical examination through evaluation is indicated. Also because Multiple risk factors are often present in individual patients with pediatric stroke, complex etiologic investigations are often necessary. The cause of stroke in children is established in  $\approx 75\%$  of cases.

**Key words:** Ataxia, Cerebellum, Stroke

### Vestibular evoked myogenic potential (VEMP) test in evaluating children with benign paroxysmal vertigo

Nilforoush, MH., M.Sc, Sepehrnejad, M., M.Sc, Rahbar, N., M.Sc; Audiology Department, Rehabilitation faculty, Medical University of Isfahan

**Background:** Benign paroxysmal vertigo is defined as recurrent (at least 5) attacks of severe vertigo resolving spontaneously after minutes to hours. Since diagnosis of BPV was made by the initial assessing conflicting results may be probably due to selection bias. Therefore, objective parameter, e.g. vestibular function test may help to evaluate BPV children. Recently, vestibular evoked myogenic potential (VEMP) test has been validated to reflect the sacculo-collic reflex.

**Methods:** in this study, tried to present a review article from twenty original articles during 2000 to 2010 on this subject.

**Results:** VEMP test showed 50% abnormality in BPV children, including absent and delayed responses. VEMP results reveal higher abnormality in BPV children.

**Conclusions:** Vestibular evoked myogenic potential (VEMP) is a non-invasive, fast and reliable test in balance system evaluation. Thus, VEMP tests may serve as supplementary diagnostic tool in evaluating children with BPV. Key word: Benign paroxysmal vertigo, vestibular evoked myogenic potential, Children  
**Keywords:** Benign paroxysmal vertigo, vestibular evoked myogenic potential, Children

## Body mass index in Children with Autism Spectrum Disorders in Tehran, Iran

*Memari, AH., MD, Kordi, R., MD, Sport Medicine Research Center, Tehran University of Medical Sciences*

**Background:** In children and adolescents, the prevalence of obesity and overweight has tripled in the last two decades and many studies to investigate the mechanisms and solutions for this threat of child healthy life are underway for several pediatric subpopulations. In children with autism spectrum disorder (ASD) this is an important undertaking, since not only the obesity mechanisms may not be the same as for typically developing children but also it can be followed by more complex consequences for the autistic counterparts. We aimed to estimate the prevalence of obesity in children with ASD attending autism special schools.

**Methods:** A Representative sample from the autism special schools students (7-15 years old) consisted of eighty nine boys and twenty three girls diagnosed with autistic spectrum disorder have recruited; the height, weight and BMI of participants was investigated and analyzed to find the body composition of children with ASD. Further, their parents were asked to complete the checklists of general information.

**Results:** Overall, 22 (19.6%) of the children were obese, 18 (16.1%) overweight, 63 (56.3%) healthy weight, and 9 (8%) underweight; Rate of obesity in girls was 30.8% against 16.9% in boys. Further analysis showed that a significant correlation existed between age and BMI ( $r=0.26$ ,  $Pvalue<0.01$ ).

**Conclusions:** Based on present findings, children with ASD have a prevalence of obesity at least as high as children overall; and further research is warranted to follow up the consequences of the obesity in the ASD population. Moreover, Particular attention should be given to the obesity in autistic girls rather than boys.

**Keywords:** autism, BMI & obesity

## Hi's Healer Neurogenetics of Epilepsy

*Taqizade, F., Department of Genomic Psychiatry, Ruzbeh Psychiatric Hospital, faculty of Medicine, Teheran University of Medical Sciences*

**Background:** A very important challenges in epilepsy is the hereditary of it. As 400 years ago (FD) Hippocrates discovered to hereditability of epilepsy and Tissot on seventeenth century captioned the genetics factors role in epileptic patient susceptibility. Because the epilepsy is a complex disease therefore the genetics study of that isn't easily than single gene disorders. There are very wide disparities about be genetically origin or vulnerability gene(s) in this disorder.

According to last researches is the lower brain provocation threshold in some persons for expression epilepsy. Meantime are special heritable syndromes that there are convulsions high Incidence. (e.g. Sphingolipidosis, Mucopolysaccharidosis, Lipid and Amino acid and glucose metabolism disorders.

**Method:** this study made a premetha-analysis and mentioned genetics aspects of all infantile seizure disorders. The keyword was Genetics, Gene, Epilepsy, Convulsion and Hereditary; and Database was Pubmed. Our limited results by "Human studies". So arranged the results by types of clinical manifestations of epilepsies.

**Results:** all types of genetically epilepsies which identified those genes include Idiopathic epilepsies (generalized and focal), Symptomatic epilepsies (lenox syndrome and Tuberos sclerosis), Progressive epilepsies (Unverricht-Lundborg and Lafora's Disease), Epilepsies with complicated inheritance (Benign rolandic epilepsy), Epilepsies with simple inheritance or single gene (ADNFLE, FTLE). This is clarify most types of epilepsies have Atosomal Dominance (AD) inheritance and about epilepsies with complex inheritance demanded most studies.

**Conclusion:** for net diagnosis and effective treatment of epileptic patients must be extensive genetics investigations and design genetherapy based on individual genomics characters.

**Keywords:** Epilepsy – Gene – Inheritance – Chromosomal locus

## Assessment and management of common psychiatric problems in pediatricians' clinics

*Mahmoudi-Gharaei, J., MD, Tehran University of Medical Sciences*

Many psychiatric disorders such as Attention deficit/Hyperactivity disorder, anxiety, sleep and feeding disorders are detectable and manageable in pediatricians' clinics. Attention deficit/Hyperactivity disorder has been viewed as a disorder of elementary school-age children. Considering morbidity of this disorder, early detection and management of ADHD became a priority in clinical practice. Anxiety is among the most common and disabling psychiatric problems of childhood. Effective psychopharmacological interventions have been developed and could potentially be helpful for anxiety disorders in children and adolescents. Recent studies demonstrated that approximately 30% of young children have some kind of sleep problem, ranging from mild, time-limited difficulties with bed time to chronic, serious sleep disorders. Many of children with sleep problems may experience impaired daytime functioning. Eating disorders are also common in children but not much attention has been given to them. For example in a study the authors found that at 4 month of year 36% of

the infants were reported to have feeding problems. Considering the high prevalence of these psychiatric problems in children and their morbidities, early detection and appropriate therapeutic interventions are essential.

## Stroke in Children

*Taghavinejad, MT., MD, Behzad moghaddam, MH., Milad Hospital*

**Background:** Stroke is an acute neurologic injury occurring in CNS specially brain due to ischemia or hemorrhage. 80% of case are ischemic and 20% are hemorrhagic. Ischemic stroke may be due to ischemic thrombosis, embolism and local or systemic hypoperfusion. Hemorrhagic stroke is due to intracerebral or subarachnoid hemorrhages. Stroke is not as common in children as in old adults, but it may occur in neonates, infants children and young adults as well. Annual incidence rates of arterial ischemic stroke (A.I.S) in infants and young children is from 0.6 to 7.9 /100000 children per year. It occurs more among boys (m/f:1.5/1) The risk factors and etiologies of AIS in children and young adults are: 1)Congenital and aquired heart problems. 2)Vasculopathies 3)Hematologic problems (malignancies, coagulopathies) 4)Metabolic disorders 5)Drug ingestions 6)Infections.

**Case report:** Younger children with stroke mostly present with seizure and altered mental status but older children usually present with hemiparesis or other focal neurologic signs such as aphasia, visual disturbance and cerebellar signs although seizures, headache and lethargy are not uncommon. During the last year we had 7 cases of stroke. 3 of them had hematologic etiologies, one of them was due to congenital vascular agenesis. cardiac problem(TOF) and the complication of its treatment was seen in another case. In one of the cases no etiologic cause could be detected. The last case occurred during H1N1 pandemic following an encephalopathy secondary to this infection.

## Early Detection of Newborn Hearing Loss: Suggestion and new Protocol

*Abdi, S.*

Hearing loss is one of the most common congenital anomalies. Universal newborn hearing screening (UNHS): early detection of infants with sensorineural hearing loss well-before three months of age. The program offers all parents the opportunity to have their baby's hearing tested shortly after birth. The current screening programs in many countries consisting of: an initial screening with Transient evoked otoacoustic emissions (TEOAE) following the

second stage protocol with Automated Auditory Brainstem-response (AABR) in someone who had the "Fail" result form the first stage. Babies who need a follow-up from screening will have an audiology assessment carried out by three months of age. In our country consisting of: an initial screening with Transient evoked otoacoustic emissions (TEOAE) following the second stage protocol with the same test, Transient evoked otoacoustic emissions (TEOAE) in someone who had the "Fail" result form the first stage. The objective of our study was to recommend a new protocol for early detection of hearing loss in infants.

**Keywords:**Hearing loss, Screening, TOAE

## Is Prescreening Developmental Questionnaire-II (PDQ-II) a Valid & reliable tool for two step developmental screening of 0-6 years old children in Tehran city?

*Shahshahani, S., MD, Sajedi, F., MD, Vameghi, R., Azari, N., MD, Kazemnejad, A., PhD, Tonekaboni, SH., MD,*

*1. Pediatric Neurorehabilitation Research Center-University of Social Welfare & Rehabilitation Sciences(USWR); 2. Clinical Sciences Department-Pediatric Neurorehabilitation Research Center – USWR; 3. University of Tarbiat Modares; 5. Shahid Beheshti university of Medical sciences*

**Background:** This research was designed to identify the validity and reliability of Prescreening Developmental Questionnaire 2 (PDQ- II) in Iranian children, and also determining its agreement coefficient with Denver Developmental Screening Test- II (DDST-II).

**Methods:** At first a precise translation of the test was done by the research team and then the Persian version was back translated by three English language experts, who were unfamiliar with the PDQ. The back translated version was compared with the original version after which corrections were made and the Persian version finalized. Next the content validity of the final Persian version was verified by three pediatricians familiar with child development and also by reviewing relevant books and journals. Then, test was performed on 237 children ranging from 0 to 6 years old, in four health care clinics, in north, south, east and west regions of Tehran city. In order to determine the agreement coefficient, these children were also evaluated by DDST II at the same time. Available sampling was used until the desired sample number was achieved. Interrater methods (between mother and examiner) and Cronbach's  $\alpha$  were used in order to determine reliability of the test. The Kappa agreement coefficient between PDQ and DDST II was estimated. The data was analyzed by SPSS software.

**Results** All of the questions in PDQ had content validity, and there was no need to change them. The

total Cronbach's  $\alpha$  coefficient of 0-9 month, 9-24 month, 2-4 year and 4-6 year questionnaires were 0/951, 0/926, 0/950 and 0/876 respectively. The Kappa measure of agreement for interrater tests was 0/89. The estimated agreement coefficient between PDQ and DDST II was 0.383. Based on two different categorizing possibilities for questionable scores that is "Delayed" or "Normal", sensitivity and specificity of PDQ will be 35.7 – 63% and 75.8- 92.2 % respectively. Developmental disorders were observed in 38% of children (18% delayed and 20% questionable) who were examined by PDQ, and in 35% of children who were examined by DDST II.

**Conclusion** This research showed that PDQ has a good content validity and reliability and moderate sensitivity and specificity but by considering its relatively weak agreement coefficient with DDST-II, using it along with DDST-II for a two-stage developmental screening process, remains doubtful.

**Key words:** development, developmental delay, screening, DDST II, PDQ

### **Investigative of the prevalence of physical problems on the basis of the different ways of carrying bag at the deaf and mental retarded students in Hamedan city**

*Mortazavi, Sa., Mortaza, Se, Azad University of Medical Sciences, Hamedan Branch*

**Background:** The schools an educational and social center deal with a lot of mental, physical, social, growth and personality of people at youth and child hood. Carrying the luggage in the wrong ways can created problems for students that can not be compensated. This problem for exceptional students is of the most importance

**Method:** The study was carried out in a descriptive – analysis manner by a questionnaire referring to the exceptional elementary school the subject s under the study consisted of 80 deaf and mental retorted students who were selected randomly in order to instigate the type of the trouble of carrying bay and the kind of the bag used.

**Results:** There is a meaningful relationship between the kind of the bag used by the deaf and mental retarded student and having trouble when carrying the bag. ( $p<0.05$ ) The students who used back packs were reported to have lees problems while carrying bag than those used the normal bags. ( $p<0.05$ )

**Conclusion:** According to the most of the experts, backpack, if used correctly, is the best device for carrying educational instruments. Regarding the conditions of such students, this is recommended that all the parents, teachers, principals and exceptional education staff to pay more attention to this problem.

**Keywords:** physical problems, kind of bags. mental retarded students, deaf students

### **Anencephaly in Northern Iran (1998-2005)**

*Najafi, L., MD, Golalipour, MJ, PhD, Keshkar, AA., MD, Ph.D, Gorgan Congenital Malformations Research Center Gorgan University of Medical Sciences; Gorgan Congenital Malformations Research Center, Department of Social Medicine*

**Background:** Anencephaly is a neural tube defect that is incompatible with life. The aim of this prospective study was to determine the prevalence of anencephaly in North of Iran.

**Methods:** Since 1998 through 2005, 49534 births in Dezyani hospital in Gorgan, North of Iran, were screened for (NTDs). Clinical and demographic data of diagnosed cases were recorded in a pre-designed questionnaire for analysis. These included sex, ethnicity, parental consanguinity and residential area.

**Results:** The overall prevalence of NTDs and anencephaly were 28 and 12 per 10000 births respectively. The prevalence of anencephaly was 11 and 12 per 10000 births in males and females respectively. According to the parental ethnicity, the prevalence of anencephaly was 12, 16 and 7 per 10000 in Fars, Turkman and Sistani races, respectively. The rate of anencephaly was 13.1/10000 in newborns with mothers aged 15-19 years. Thirty six percent of the parents had consanguinity. Also 63% of the parents resided in rural areas and 37% in urban areas. The highest rate was in the year 1999 (23/10000) but the least was in 2003(2/10000). The most prevalent season for occurrence of anencephaly was winter (16/10000).

**Conclusion:** The present study indicated that the prevalence of anencephaly among Iranian newborns in North of Iran was higher than European population. More studies should be performed to ascertain if improved folate supplementation has contributed to the drop in overall incidence. The incidence in Turkman population is significantly higher than other ethnicity groups and this phenomenon should be further studied.

**Keywords:**Anencephaly, Prevalence, Neural tube defects, birth defect, Iran

### **Case study of a child's math learning disorder**

*Abniki, E., MA, Delavari, M, MA; Children's medical center, Tehran University of Medical Sciences*

**Background:** The main characteristic of disorder in mathematics is that the ability in mathematics regarding to age, calendar year, measured intelligence and education commensurate with his age, significantly lower than is expected. The aim of this study is a case study of children with disorders in mathematics and its related ability to rehabilitate. Case report: the desired child is 9-year-old girl is in second grade who studying elementary. The child complained of impaired subjects, especially mathematics lessons and behavior problems

by school teacher was referred. Tools: to assess child intelligence, an intelligence test for children was used. The questionnaire related to diagnostic capabilities up to three years old child, diagnostic questionnaire mathematical abilities ages three to six years, the content of the questionnaire by first grade math book, math book content questionnaire for primary school children were used to measure ability. Results: Using an intelligence test became clear that the intelligence test child is 92 and is normal. The questionnaire listed the amount of child disability-related concepts of mathematics were identified. Then regarding to specific disability, the rehabilitation program to enhance the capabilities of a variety of children mathematical concepts related to planned and implemented. The program for 5 months and a week was carried out. After completing the program, reporting results to evaluate teachers and mathematics tests showed that these rehabilitation programs to treat child has math disorder. Further results will be mentioned in the main article.

### Reliability, validity and feasibility of persian version of PedsQL™ generic core Scales

*Heidary, M., Gheissari, A., Farajzadegan, Z., Salehi, F., Masaeli, A., Mazroi, A. ; Isfahan University of Medical Sciences*

**Background:** To evaluate the reliability, validity and feasibility of the Farsi version of the Pediatric Quality of Life inventory (PedsQL™ 4.0) Generic Core Scales

in Iranian healthy students ages 7-15 and chronically ill children ages 2-18.

**Methods:** We followed the translation methodology proposed by developer. Sample of 160 healthy students (chosen by random cluster method between 4 regions of Esfahan education office) and 60 chronically ill children (chosen consequently from patients come to Alzahra hospital specialty clinics) and their parents completed the Farsi version of PedsQL™4.0 Generic Core Scales.

**Results:** The Farsi version of PedsQL™4.0 Generic Core Scales discriminated between healthy and chronically ill children (healthy students mean score was 12.3 better than chronically ill children,  $p < 0.001$ ). Cronbach Alpha's internal consistency values exceeded 0.7 for children self reports and proxy reports of children 5-7 years old and 13-18 years old. Reliability of proxy reports for 2-4 years old was much lower than 0.7. Although proxy reports for chronically ill children 8-12 years old was more than 0.7 these reports for healthy children with same age group was slightly lower than 0.7. Constructive, criterion face and content validity was good. The Farsi version of PedsQL™4.0 Generic Core Scales was feasible easy to complete.

**Conclusion:** Results show the Farsi version of PedsQL™4.0 Generic Core Scales is suitable for pediatric health researches children over 8 and parents of chronically ill children over 5. It is necessary to alternate scoring for 2-4 years old questioner and to find a way to increase reliability healthy children ages 8-12 especially according to Iranian culture.

**Keywords:** PedsQL; Pediatric; Quality of life

## Rheumatology

### Refractory Juvenile Dermatomyositis and Infliximab “Case Report”

*Aghighi, Y., MD, Fallah, F., MD, Raeeskarami, SR., MD; Tehran University of Medical Sciences*

**Background:** JDM is a multisystem, autoimmune inflammatory disease that affects children younger than 18 years and characterized by acute & clinically inflammation of the skeletal muscle and skin rash. The disease manifest as skin findings and muscle weakness. Traditional treatment for JDM includes high dose corticosteroid with MTX. For refractory or severe disease, the use of cyclosporine, IVIG and pulse therapy with intravenous methylprednisolone are recommended. Second line agents include azathioprin, mycophenolate mofetil and hydroxychloroquine used for skin disease. More recently biologic agents such as anti-TNF  $\alpha$  (infliximab) and rituximan (Anti CD20- Anti body) has shown major clinical benefit in patients with refractory JDM. Infliximab is a drug that blocks a key protein of all inflammatory processes called tumor necrosis factor (TNF  $\alpha$ ). TNF is a signaling protein that increase the activity of cells involved in inflammation. Infliximab stops TNF from binding to cells, there by reducing inflammation & should only be used in patients who do not respond to other treatments. Patients receiving infliximab should continue to receive MTX or a similar type of drug.

**Case Report:** An 8 year old girl with refractory JDM who was treated by infliximab is reported. Onset of clinical manifestations started were at 6. Initial symptoms were dermatological followed by muscle involvement which results symmetrical, proximal muscle weakness. Gradually increased fatigue on walking and loss of ability to perform activities of daily living. She had an inadequate response to conventional therapy, including IVIG, MTX, oral Prednisolone and high dose intermittent intravenous methylprednisolone. She was admitted in PICU due to respiratory distress secondary to intercostals weakness. Because of unresponsibility to conventional treatments, administration of infliximab was begun. Infliximab was prescribed at a dosage of 3mg/kg/per dose. She is receiving the infliximab on weeks 0, 2,6,14 and then fallow at 8 weekly interval. After administering of four courses of drug obvious clinical improvement in muscle and normalization of and enzyme were recorded. She tolerated well and remained in remission.

**Conclusion:** Infliximab has beneficial effects in children with refractory JDM, especially in those with severe muscular manifestations.

**Keywords:** refractory JDM, Infliximab.

### Linear Scleroderma with Facial Involvement (en coup de sabre) “Case Report”

*Raeeskarami, SR., MD, Saeidi, M., MD, Aghighi, Y., MD; Tehran University of Medical Sciences*

**Background:** Localized fibrosing disorder include a spectrum of rare conditions that frequently begin in childhood and characterized by circumscribed fibrotic areas involving different levels of dermis, subcutis and sometimes underlying soft tissue and bone. The causes of localized fibrosing disorder are unknown. Localized fibrosing disorder is subdivided into subgroups: linear scleroderma, lesion en coup de sabre, progressive hemifacial atrophy (Parry-Romberg syndrome). Lesions en coup de sabre is a linear scleroderma which involve the face and scalp. The lesions starts with contraction. The lesion in localized sclerosing condition is characterized by infiltration of lymphocytes, mast cells, plasma cells and eosinophiles with excess collagen deposition extending into the dermis, the subcutaneous fat and in some forms deeper structures. Treatment including; systemic, topical corticosteroids. Modifying agents are necessary to control a severe inflammatory process. Plasma pheresis, psoralen plus ultraviolet light, tacrolimus can be useful in localized scleroderma. Often patients with en coup de sabre systemic require surgical reconstruction of the face and scalp.

**Case Report:** A 5 year-old girl with localized scleroderma without internal organ involvement is reported. The skin involvement is remarkable in the unilateral, atrophic and contracture lesion on right side of the face which became progressive to atrophy on the same side. The sign began at age 5 by dyspigmentation patchy lesion. She was suffering from a polyarthropathy that began at the sametime. She referred to dermatologist just at the beginning of her sickness and used topical cream “Elider” and then referred to neurologist and tried physiotherapy, with usefulness, result. According to lab data, she had a positive “Anti ds DNA”, “Anti CCP” and a negative “ANA”, “SCL 70”, “Anti centromer” respectively. RF- C3- C4 CH50 were within normal limits. The chest x-Ray was normal. Barium swallow and echocardiography were also within normal limits.

**Keywords:** Linear Scleroderma, En coup de sabre

### Kawasaki disease (KD)

*Moradinejad, MH., Children Medical center, Tehran University of Medical Sciences*

**Background:** Kawasaki disease (KD) or (infantile poly arteritis nudosa) is an acute intense life-threatening vasculitis. Kawasaki disease was first described in

Japanese in 1967 by Tomisaku Kawasaki. The diagnosis of K.D. is made by clinical criteria. The child with K.D. must have a fever lasting 5 or more days, and four of the following criteria: 1) bilateral, nonexudative conjunctival injection; 2) red lips or fissured lips, injected pharynx, or "strawberry tongue"; 3) erythema of the palms or soles, edema of the hands or feet, 4) polymorphous exanthem; 5) acute nonsuppurative cervical lymphadenopathy. An additional category of atypical K.D. includes patients with only four criteria and the presence of coronary artery abnormalities on echocardiography. This disease is a common vasculitis after henoch-schonlein syndrome in Iran. The aim of this study was to describe the demographics and clinical features of KD in Iranian patients.

**Methods:** A review was conducted for all cases of KD treated at Pediatric rheumatology department in Children's Medical Center between January 2000 and July 2010. The diagnostic criteria for typical Kawasaki were based on the criteria of the Japan Kawasaki Disease Research Committee. Atypical or incomplete KD has been described in which patients not strictly meeting the diagnostic criteria but have coronary artery changes. Color doppler echocardiograms were done at the time of diagnosis, 14 to 21 days, 60 days, and 1 year after treatment. Dilatation of coronary arteries may be detected by echocardiography beginning 7 days after the first appearance of fever, with the coronary dilation usually peaking around 4 weeks after the onset of illness. In general, clinical and laboratory indices of greater inflammation are associated with a higher likelihood of aneurysm development.

**Results:** Two hundred seventy patients were identified. In our study two hundred twenty children (81%) fulfilled criteria for typical KD, and fifty (19%) cases with atypical Kawasaki. One hundred fifty cases were boys and 120 cases were girls, the disease is more common in males than in females, with a male-to-female ratio of (M to F Ratio was 1.25: 1). The age distribution pattern showed from 3 months to 5 years of age, the peak prevalence is in children aged 18-24 months. Echocardiographic abnormalities were found in 60(21%) cases, including 60% with typical and 40% with atypical Kawasaki. Male to female ratio was more in patients with cardiac complication (2.3:1). Coronary artery ectasia or aneurysms occur in 15% to 25% of children with K.D. who do not receive treatment with IVIG in the acute phase. Therapy for K.D. in the acute phase is aimed at reducing inflammation of the coronary artery wall and preventing coronary thrombosis; treatment with high-dose intravenous gamma globulin G and aspirin has become the standard of care and reduces the risk of development of coronary artery. Even when treated with high-dose (IVIG) regimens within the first 10 days of illness, however, approximately 5% of children with K.D. develop at least transient coronary artery dilation, and 1% develop giant aneurysms.

For those with persistent or recrudescing fever despite initial IVIG infusion, multiple courses of gamma globulin and treatment with corticosteroids may be indicated.

**Conclusion:** Kawasaki disease should be considered in any infants or child (especially less than 2 years old) with a prolonged febrile illness. Demographic features of our patients were similar to reports from other countries. The incidence of atypical Kawasaki in our study was about 19%.

**Keywords:** Kawasaki, Vasculitis, Aneurism, Coronary artery, Atypical Kawasaki

### B-cell Lineage Study in Patients with Juvenile Idiopathic Arthritis

*Rezaei, A., MD, Aghamohammadi, A., MD, Moradinejad, MH, MD, Parvaneh, N., MD, Rezaei, N., Seyedtabaei, R., Asgarian-Omran, H., MSc, Shahrestani, T., MSc, Amirzargar, AA., PhD; Tehran University of Medical Sciences*

**Background:** Juvenile idiopathic arthritis (JIA) is the most common rheumatic disease in children. The exact causes of disease are still poorly understood. It seems that B cells have several functions in JIA, including production of autoantibodies, antigen presentation, production of cytokines, and activation of T cells. Here, we aimed to evaluate B-cell lineage and its precursors in the bone marrow of patients with JIA.

**Methods:** Twenty consecutive patients with JIA were enrolled in this study. JIA is subdivided into three groups of Pauciarticular, Polyarticular, and Systemic JIA. Bone marrow mononuclear cells were separated. Then we analyzed the immunophenotype of the JIA patients by flow cytometry. After separation, the mononuclear cells were stained specific for B cell lineage [CD10, CD19 and CD20], T cell lineage [CD3] and non specific lineage [CD34, HLA-DR and TdT].

**Results:** Flow cytometric study of bone marrow showed that JIA patients had low level of CD10, CD19, and CD20. Polyarticular patients had lower level of CD10, CD19, and CD20 than pauciarticular JIA patients and systemic onset JIA patients had lower levels than both of them.

**Conclusion:** Decreasing of B cell precursor in bone marrow is one of mechanisms for pathogenesis of JIA and the more decreased B cell precursors in bone marrow are, the worst severity of the disease is. Significant differences in CD10 content of bone marrow were detected between the polyarticular and pauciarticular groups. So, it seems that polyarticular JIA patients had lower percentage of pre B cell stage.

**Keywords:** B-cell Lineage, Immunophenotyping, Juvenile idiopathic arthritis, Chronic Arthritis

## Young Researchers Abstracts

### Frequency of different types of first cousins marriages among parents of female patients affected to CLP and CP

*Davoodi, E., Joyain, N., Senemar, S., SHojaei, A.; Department of Genetics, Tehran University of Medical Sciences*

**Background:** Congenital cleft lip with or without cleft palate (CLP) and cleft palate alone (CP) are common craniofacial anomalies in different countries. The etiology of these disorders involves interaction of genes on different chromosomes including X and environmental factors. Consanguineous marriages can increase the incidence of congenital malformations. The special manner of transition of X chromosome suggests that different first cousins marriages can have different influences on incidence of multifactorial disorders with susceptibility genes on X chromosome. The aim of this study is investigation of frequency of different types of first cousins marriages among parents of female patients affected to CLP and CP. **Materials and Methods:** The data about patients were extracted from pedigrees of 9964 genetic counseling files belonged to referred people to genetic counseling center of Iranian Academic Center for Education, Culture & Research, Fars Province Branch. The frequency of different types of first cousins marriages were estimated among female patients' parents and compared to this frequency among 714 first cousins couples referred to this center for genetic counseling before marriage or pregnancy. **Results:** 215 and 81 patients affected to CLP and CP were identified. The frequency of consanguineous marriage was 50.3 and 66.2 respectively in CLP and CP patients. The percents of parallel matrilineal marriage, parallel patrilineal marriage, matrilineal cross-cousin marriage and patrilineal cross-cousin marriage among parents of female patients affected to CLP and CP were 21.4%, 21.4%, 46.4%, 10.7% and 16.7%, 22.2%, 27.8%, 33.3% respectively while these percents were 32%, 35%, 19% and 14% among first cousins couples referred to this center that had not significant differences based on X2 test.

**Conclusion:** The high frequency of consanguineous marriage among parents of these patients was expected but frequencies of parallel matrilineal and matrilineal cross-cousin marriages that were expected to have higher risk weren't higher than two other types of first cousins marriages that weren't according our prediction. It seems that complex etiology of these disorders and also small

sample size used in this study have made it hard to comparison of the influence of different types of first cousins marriages on incidence of these disorders.

**Keywords:** Consanguineous marriages, multifactorial disorders, susceptibility genes, first cousins marriages

### Relationship between cardiovascular fitness and serum leptin, ghrelin, cortisol, lipid profile and body fat in overweight and obese boys

*Fallahi AA., Ravasi, AA., Abasi, A., Department of Sport Physiology of Tehran University; PhD Student in field of Sport Physiology of Tehran University*

**Background:** Previously we conclude some inflammatory markers (IL-6, CRP, and WBCs) were inversely associated with cardiovascular fitness in children. In recent years 2009-2010, this have been reported leptin maybe a risk factor of cardiovascular disease and ghrelin have a protective effect (improved left ventricular contractility and cardiac output, reduction of arterial pressure and systemic vascular resistance, improves endothelial dysfunction) to cardiovascular system. But this hypothesis not survived in obese children and adolescence. Therefore, the purpose of this study was, survey the relationship between cardiovascular fitness and serum leptin, ghrelin, cortisol and body fat in overweight and obese boys.

**Methods:** Twenty three male overweight and obese boys with mean aged  $15.52 \pm 0.66$  years old were participated to study. Criteria of this study was BMI > 25, and there are not any of subjects with any diseases. All subjects were participated to a graded exercise testing breath to breath gas-analyzer by Bruce protocol for measurement of cardiovascular fitness or VO<sub>2</sub>max. Serum leptin, ghrelin, cortisol levels were determined at baseline, before of graded exercise testing. Body fat percentage was calculated by the sum of 8 measurements of skinfold thickness sites (biceps, triceps, subscapular, iliac crest, suprailiac, abdominal, front thigh and medial calf). Enzyme immunoassay (ELISA) method and BioVendor R&D and DRG kit were used for measurement of serum ghrelin and also leptin. Data were analyzed using pearson correlation and regression model.

**Results:** Analysis of data showed, mean VO<sub>2</sub>max for subjects was  $41.13 \pm 5.17$  ml/kg/min. Cardiovascular fitness negatively correlated ( $P < 0.05$ ) to leptin ( $r = -0.451$ ,  $p = 0.031$ ) and body fat ( $r = -0.500$ ,  $p = 0.015$ ), but

not with Ghrelin ( $r=0.137$ ,  $p=0.533$ ), cortisol ( $r=-0.034$ ,  $p=0.879$ ), triglyceride ( $r=-0.180$ ,  $p=0.412$ ), cholesterol ( $r=-0.287$ ,  $p=0.185$ ), HDL ( $r=0.214$ ,  $p=0.327$ ), LDL ( $r=-0.321$ ,  $p=0.135$ ). Regression by stepwise model showed, body fat is a most important negatively predictors of cardiovascular fitness.

**Conclusion:** In summary, this maybe conclude elevated levels of leptin and percentage of body fat were a new and important risk factor for cardiovascular disease in overweight and obese boys. Also body fat is a most important negatively predictors of cardiovascular fitness than levels of serum leptin, lipid profiles.

**Keywords:** Vo2max, Ghrelin, Obesity, Lipid profile, Leptin

### Promoting safety in children with disabilities

*Tol, A., PhD Student, Sharirirad, GR., PhD, Isfahan University of Medical Sciences; Deputy of Health, Tehran University of Medical Sciences*

Children with disabilities are at increased risk for accidental (unintentional) injuries, inflicted (intentional) injuries, and child neglect. Behavioral, physical, and cognitive characteristics of the child and environments that are not well adapted for individuals with disabilities contribute to the increased risk. Predisposing factors Behavioral factors- Children with behavior disorders (eg, increased activity, impulsivity) are more likely than those without behavior disorders to engage in hazardous behaviors that may result in unintentional injury. Intellectual factors - The prevalence of injury in children and adolescents with intellectual disabilities is 1.5 to 2 times that of children and adolescents without intellectual disabilities. Biologic factors - Biologic features of certain conditions may increase the risk or severity of injury. Environmental factors - Environmental factors may increase the risk of injury if the environment is not adapted to the needs of individuals with disabilities. Summary And Recommendations Children and adolescents with disabilities have an increased risk of injuries compared to their nondisabled peers. Primary care providers play a key role in promoting the safety of children with disabilities through anticipatory guidance, treatment, counseling, and referral. Factors that may contribute to the increased risk of injury include behavioral problems, cognitive impairment, medical conditions, environments not well adapted to the needs of disabled children, psychosocial stressors associated with caring for a child with disability, and the increased vulnerability of a child with disability. Prevention of unintentional injury in children with disabilities encompasses the injury prevention strategies that are recommended for all children. In addition, primary care providers should tailor injury prevention advice to their

patients' specific disabilities (eg, osteopenia, attention deficit hyperactivity disorder, visual impairment). Prevention of maltreatment entails provision of anticipatory guidance regarding the increased risk of maltreatment in children with disabilities, provision of support to the families of children with disabilities, recognition and fostering of family strengths, and provision of developmentally appropriate self-protection training for children with disabilities.

**Keywords:** Disabilities, Children

### The effects of swimming primary training on the leptin of the serum and related hormones in children and adolescents

*Yarahmadi, H., MS, Hamedinia, MR, PhD, Porjahed, MJ., MS, Sabzevar Tarbiat Moallem University*

**Background:** leptin is a protein with a helical structure similar to cytokines and a relative mass of 16 kDa, assists in the regulation of body weight and energy homeostasis. The purpose of present study was to examine the effect of swimming primary training on leptin of the serum and related hormones in obese children and adolescents.

**Method:** 60 obese subjects (children and adolescents) were randomly assigned to one of four obese groups: 1) swimming primary training groups (children), 2) control groups (children), 3) swimming primary training groups (adolescents), 4) control groups (adolescents). Our experimental subjects received swimming primary training for 8 weeks and 3 sessions per week and 60 minutes per session. Before and after training period, blood samples, anthropometric and body composition measurements were taken in fasting state from all subjects.

**Findings:** The swimming primary training prevent significant increase of serum leptin and insulin hormone in adolescents. Furthermore swimming primary training caused a significant decrease in body fat percent, body fat mass and body mass index, a significant increase of Vo2max in children and adolescents, a significant decrease in cortisol hormone and a significant increase in fat free mass in adolescents. Between levels of leptin hormone changes and body fat percent and fat mass after swimming primary training there was positive and significant correlation and between levels of leptin hormone change and fat free mass after swimming primary training there was a negative correlation. Levels of basal serum leptin were significantly higher in children than adolescents ( $p<0/05$ ).

**Conclusion:** 8 week swimming primary training improve serum leptin and insulin hormone some of the anthropometric and metabolic parameters.

**Keywords:** Swimming primary training, Leptin, Obesity, Children, Adolescents, Insulin

### Clinical and immunohistochemical factors predicting for primary focal segmental glomerulosclerosis (FSGS) in children

*Beigi, H., Mozafarpour, S., Samanianpour, S., Medical Student, Isfahan University of Medical Sciences; Gheissari, A., MD, Department of Pediatrics, Pediatric Nephrology Department, Al-Zahra Hospital, Isfahan University of Medical Sciences*

**Background:** Focal Segmental Glomerulosclerosis (FSGS) in children is a clinicopathological syndrome characterized by segmental sclerosis of some glomeruli. It is the most common cause of End Stage Renal Disease (ESRD). There is no study so far to determine immunohistochemical predictors of pediatric FSGS. In this study we tried to search for prognostic values of immunohistochemical (Vimentin, Desmin and  $\alpha$ -smooth muscle actin) and clinical factors in primary pediatric FSGS.

**Methods & materials:** In this historical cohort survey, we have studied the expression of cytoskeletal proteins (Vimentin, desmin,  $\alpha$ -SMA) in the paraffin embedded tissue sections from the kidneys of 31 children with primary FSGS from 2001 until 2009 in Isfahan, Iran with evaluating clinical features. To evaluate renal survival and outcomes we used Kaplan-Meier's curves and Cox's multivariate Regression. The end point was GFR less than 90 ml/min/1.73 m<sup>2</sup>. **Result:** Thirty-one patients were included in the study, 20 (64.5%) were male and 11 (35.5%) were female. At presentation, the mean age was 64.3 $\pm$ 49.35 months, proteinuria ranged from 0.1-4.3 g/day/1.73m<sup>2</sup>, the median estimated Glomerular Filtration Rate (eGFR) was 96.8 ml/min/1.73m<sup>2</sup> and 45.2% had high blood pressure at presentation. After a mean follow up of 46.3  $\pm$  56.5 months, 61.3% and 16.1% reached complete and partial remission respectively. Five-year renal survival was 41.7%. Among clinical and immunohistochemical parameters, only proteinuria adjusted by body surface area (BSA) was associated with renal survival (P=0.003) and none of other parameters were determinants of renal survival (P>0.05). **Conclusion:** In our study, we did not find any correlation between immunohistochemical parameters and renal survival in childhood primary FSGS and among clinical factors, just BSA proteinuria at presentation was renal survival independent predictor. Further studies with bigger samples and longer follow up period are needed to identify more correlations.

**Keywords:** Primary Focal Segmental Glomerulosclerosis, Immunohistochemical factors, Renal survival

### Yellow nail syndrome with CD4 deficiency: a case report

*Behforouz, A., Medical Student, Atarod, L., MD, Shaqafi, MR., Medical Student, Tehran University of Medical Sciences*

**Background:** The yellow nail syndrome is a rare disease with a classic triad of yellow nails, lymph edema and pleural effusion. Nail alterations include; yellow discoloration, onycholysis, onychodystrophy, thickened and over curved nails. Some respiratory manifestations like sinusitis, chronic bronchitis and bronchiectasis could be observed in this syndrome. Lymph edema mostly affects lower limbs and pleural effusion is rare and often exudative.

**Case report:** In this article, we report a 14 years old girl with yellow nail syndrome. She had nail alterations (like onychodystrophy, onycholysis and yellow over curved nails), painful edema of extremities specially in lower limbs, facial edema, bronchiectasis and pan sinusitis. She had dystrophic nails since birth. Lower limb edema started with fingers of left leg at first month and progressed to the knee. At 10 the right foot became edematous and now it is up to knee. The treatment of YNS depends on the signs. Vit E, zinc sulfate, oral antibiotics and hydrochlorothiazide were prescribed for our patient.

**Conclusion:** The diagnosis of YNS, depends on accurate examinations and paying enough attention to the clinical signs. Although most cases are around 40 years old, it is important to know that the syndrome could be seen in all ages.

**Key words:** yellow nail syndrome, lymph edema, bronchiectasis, plural effusion, CD4+ deficiency

### The incidence of nephrocalcinosis in very low birth weight neonates

*Azizi-asl, MR., MD, Gylan University of Medical Sciences; Ataei, N., MD, Amini, E., MD, Tehran University of Medical Sciences*

**Background:** Nephrocalcinosis (NC) refers to diffuse perception and growth of either calcium oxalate (CaOx) or calcium phosphate in the parenchyma of the kidney. The risk of nephrocalcinosis (NC) in preterm neonates is considerable, but conflicting numbers are given for actual incidence (10-65%). Ultrasonography (US) has been found to be a sensitive and reliable for detection of nephrocalcinosis. Furosemide induced hypercalciuria is said to be the main risk factor. This study designed to determine the incidence and possible contributory factors

toward renal nephrocalcinosis in our population of preterm neonates.

**Methods:** We assessed prospectively the incidence, causes and outcome of nephrocalcinosis in very low birth weight (less than 1500g) preterm neonates by serial renal ultrasound scans using a 7/5 MHz and 5 MHz probe and urine analysis. Infants born elsewhere and transferred after 24 hours of age were excluded.

**Results:** Two out of the 50 infants developed nephrocalcinosis giving an overall incidence 4% in the study group. The follow up showed persisting nephrocalcinosis in both preterm neonates. Urinary investigation showed no consistent findings in infant with nephrocalcinosis.

**Conclusion:** The incidence of nephrocalcinosis was lower in our population than is usually reported. Routine renal ultrasound scanning of very low birth weight preterm neonates is valuable in detecting nephrocalcinosis

**Keywords:** Nephrocalcinosis, Ultrasound scan, Very low birth weight, Preterm neonate

### **Case report: An Amphotericin B resistant thoracic vertebral osteomyelitis due to Aspergillus in a patient with chronic granulomatous disease (CGD) dramatically treated with antifungal agent; Vericonazole**

*Rahaei M, Atarod L, Goodarzi V, Pourmand N, Khaki S; Tehran University of Medical Sciences*

**Background:** Chronic granulomatous disease (CGD) is a rare inherited disorder of the nicotinamide adenin dinucleotide phosphate (NADPH) oxidase complex in which neutrophils and monocytes are incapable to destroy pathogens. This situation predisposes patients to bacterial and fungal infections. Some manifestations of this disease are several abscess, pneumonia, multiple infections and osteomyelitis. Fungal osteomyelitis due to Aspergillus is infrequent in CGD patients specially long bone osteomyelitis due to Aspergillus has been rarely described. Common anti fungal agents such as Amphotericin B, Antimicrobial agents and interferon-gamma are the principles of current therapy for chronic granulomatous disease (CGD) Case presentation: A 17 years old boy presented with hypodermic abscess. After clinical and paraclinical tests, the patient was known as a rare case of osteomyelitis of the ribs and vertebrae. The infection was resistant to Amphotericin so the patient was treated with newer generation of anti fungals, Vericonazole, INF- $\gamma$  and IVIg which resulted in an excellent clinical response with no further complication. Conclusion: In patient with CGD because of several disorders that involve various organs of the body during

life, serious and constant follow up are needed. Some of the disorders such as resistant osteomyelitis of the ribs and the vertebrae can be treated dramatically with newer antifungals so they should be in the special focus of attention.

**Keywords:** thoracic vertebral osteomyelitis, Aspergillus, chronic granulomatous disease (CGD), Vericonazole

### **Sublingual traumatic ulceration in a CP child**

*Kay, G., MD; Resident of Pediatric Dentistry of Tehran University of Medical Sciences*

Sublingual traumatic ulceration in a CP child Riga-Fede disease (RFD), first described in 1881 by Italian physician Antonio Riga and in 1890 by Fede who performed histological studies of the lesions, is a benign and uncommon mucosal disorder. and usually presents with ulceration of the oral mucosa and usually the ventral surface of the tongue. It is most commonly associated with natal or neonatal teeth in newborns. it may also occur in older infants after the eruption of primary lower incisors with repetitive tongue thrusting habits, or in children with familial dysautonomia (in sensitivity to pain), or, in children with cerebral palsy Dyskinesias of various body parts accompanies CP, including the tongue, which may predispose the infant to this lesion when the tongue is involved. Although more common in infants, RFD has been reported in older patients and, recently, in patients with acquired immunodeficiency syndrome. Individual lesions present as erythema surrounding a centrally removable, yellow, fibrinopurulent membrane. Often, the lesion develops a rolled white hyperkeratotic border immediately adjacent to the ulceration. As it often mimics many oral malignant and benign disorders, the differential diagnosis is important. Case report An 10-month-old infant boy suffering from cerebral palsy (CP) presented with a large exophytic lesion located on the anterior ventral part of the tongue.(figure1)the parents reported that the child had inadequate nutrient intake, and difficulty in suckling. Contact of the anterior part of the tongue with the newly erupted anterior mandibular teeth was suspected to be the cause. so, two central incisors was extracted. after 2 months the parents came back and complain about the return of the lesion according to the eruption of the lateral incisors (figure2). But this time we decided to grind round the incisal edges of the anterior mandibular teeth and covering with composite resin material. The patient did not return for follow-up. During a telephone interview with the mother, 6 year later, she reported that the ulceration of the tongue was almost completely healed. Conclusion Although these oral lesions are self-limiting and may heal spontaneously, delayed or false diagnosis

and ineffective therapy may result in mutilation, permanent deformity of the tongue, nutritional insufficiency, and growth retardation in the long term. Restorative dental treatment such as placement of a protective barrier over the sharp edge of the incisor teeth and smoothing of rough incisor edges, changing feeding habit or style (ceasing use of a feeding bottle), and topical corticosteroid application can be used as conservative treatment options. Extraction of the lower incisor teeth has been suggested as a more radical treatment choice.  
**Keywords:**Sublingual traumatic ulceration, Riga Feda Disease

### **A survey on experiences of parents of children with major thalassemia: Qualitative study**

*Mnamjou, Z., Khodayarian, M., Motevasselian, M., Bakhshi, F*

**Background:** Major Thalassemia is the most severe Congenital Hemolytic anemia which is known due to its high prevalence in Iran and should be considered in many aspects. Because of chronic and lifelong nature of this disease, can remains psychosocial and economic problems in parents and their conflicted child. Therefore, this research has being done with the aim of determining the experiences of children with Major Thalassemia in 1389. Method: This research has done as a qualitative research and semi-structured interview method which 25 parents of children with Thalassemia disease who had referred to Specific Disease Center, has been participated to this study. Target-based sampling has been done for choosing father or mother of the child. Open questions has been asked such as, the first reaction of parents toward informing from their child disease, the time of accepting it, ways of encountering and communication of parent's family and relatives toward the child their suggestions to parents, limitations which this disease make for sick child, etc. Then, analysis of content quality for determining classes and subclasses of parent's descriptions has been taken. Results: Results showed that the first reaction of parents were sense of distress, denial and disbelief, blame and guilt, but during the time factors such as doctors and nurses explanation about the treatment, starting of blood transfusion, observation of children's physical retardation and communication with other parents, helping parents to accept their child disease. They described their sensation toward their child by doing some works such as obtaining all of their possible needs, creation of silence and calmness in family circumference, expressing more love to their child, avoiding punishment because of inaccessibility to a certain treatment and blaming themselves. The most important limitations of thalassemia has been expressed

as reduction in appetite, loss of sexual ability, early fatigue and dyspnea, social isolation, developmental retardation. Other problems which these parents had been face with were being renounced with relatives and keeping their child disease secret from them, frequent changing in doctors, relative's recommendations for doing graft and necessary experiments before other pregnancy. Conclusion: Based on finding, there are suggestions for increasing positive experiences among parents of children with major thalassemia: 1. Making facilities for grafting(cost, information, education, etc) 2. Consult with children and their parents as routine therapeutic approach. 3. Strengthening/upgrading training provided by doctors and nurse. 4. Provide special educational-entertaining tours for children and their parents also can be effective.

**Keywords:**Thalassemia, Experiences, Parents, Child

### **Evaluation of blood utilization at Central Paediatric ward, Afzalipour hospital in Kerman**

*Nikpoor, AR. Saneie-Moghadam, E., DLS, Daneshvar, H., PhD, Raesi, F., Farahi, A.; Immunology Department, Kerman Medical University*

**Background:** The preparation, storage and distribution of blood and blood components are expensive. In addition, specially in pediatric wards, request of bloods more than needed, adversely affect blood quality and freshness of bloods transfusing to the infants and pediatrics. The aim of this study was to show the current pattern of packed red cell (P.C) requestion and transfusion, according to the blood cross match to transfusion ratio (C/T ratio), transfusion index (TI) and transfusion probability index (%T) in the Central Paediatric's ward, Afzalipour hospital in kerman.

**Material:** This is a cross sectional study on 120 patient's blood requests and reservation orders. we collected the data of all blood orders of central pediatrics ward from May to July 2010. C/T ratio, TI and %T index were analyzed using SPSS 17.

**Results:** During these months, 120 blood request or reservation order forms have been sent to blood bank. Fifty seven of them had blood reservation order and 63 [ 27 boy (%42) and 36 girl (%57)] patients, had orders for blood administration. the overall ratio of C/T, TI and %T index were 1.078, 1.015 and %92 respectively. these findings were compared with standard figures of C/T < 2.5, TI ≥ 0.5 and %T ≥ %30.

**Conclusion:** In this study, we show that despite the findings of other wards of hospitals, blood requests and utilization in pediatric ward of kerman is based on appropriate protocol and it will be help to follow the

policy of proper usage of blood and its components in health system.

**Keywords:** Blood Utilization, Pediatric ward, Kerman

### **The Effect of Therapeutic Touch in preterm Infants**

*Rahiminia, E., Neonatal intensive care nursing student, member of the research (Association of the student, Tabriz University of Medical Sciences)*

**Background:** Therapeutic touch (TT), a complementary therapy, has been shown to decrease stress, anxiety, and pain in adults and children, as well as improve mobility in patients with arthritis and fibromyalgia. However, less has been reported about the effectiveness of this therapy with infants, particularly preterm infants. The aim of this study is the effect of therapeutic touch in neonates, particularly preterm infants.

**Methods:** The present study has been collected with the review of literature and internet resources.

**Results:** According to studies, the preterm infants have physical problems such as respiratory distress syndrome, respiratory insufficiency and apnea of immaturity, cardiovascular complications including patent ductus, bradycardia and anemia of prematurity and behavioral problems including avoidance by closing eyes, grimacing or crying and difficulty sleeping and resting and the preterm infant's energy fields as being complete but disorganized, lacking coherence. TT is a subtle energy therapy and the use of the TT transfers the energy to the infants.

**Conclusion:** Results have been shown that the preterm infant's physical responses to TT: decrease heart rate and respirations and seizure activity and increase respiratory functioning & oxygenation & feeding & weight over time & increase time in deep sleep and relaxation, at least the TT treatment process for preterm infants facilitates preterm infant's physiological, behavioral, energy field development and well-being.

**Keyword:** Therapeutic touch, preterm infants, energy field

### **The Perspective of children about obese persons: Painting Findings**

*Sarrafi-Kheyraabad, S., Sanagoo, A., MD, Jouybari, LM., MD; Golestan University of Medical Sciences*

**Background:** Children's paintings have a kind of instant charisma. They are simple, attractive and full of life and identity. Children talk through their paintings. The words that they are not able to tell them directly due to a variety of reasons. What is important to them is projecting what they have learned. Obesity is one of the

health problems in many countries, including Iran. The perceptions and behaviors are formed since childhood that play an important role in the future of their life. The purpose of this study was exploring the perspective of preschool children about obese persons.

**Methods:** In this descriptive study (2009) sixty children 4 to 6 years were studied. To gather the data we asked the preschool children in kindergarten in Gorgan to draw an obese man or woman as their perception. Also they were asked to describe their painting by words. The drawing and its description were analyzed using "content analysis" method.

**Results:** The children drew an obese person with "big abdomen, inactive, lazy and ugly who can not play". The non-obese one drew as active, lovely, winner and able to play". The data indicate that the children assume low mobility as obesity complication and believed overeating and laziness as the reason. They suggested "exercise" against obesity.

**Conclusion:** According to the literature "playing" for children and "working" for the adults are very vital. The children emphasized on obesity as an inhibitor for playing. The children's perception is rooted from personal experiences, family and society's perspective. The studied children had a negative view on an obese person, then we can use this opportunity to train them to make a healthier future.

**Key words:** Preschool Children, Obesity, Paintings, Drawing

### **Hypospadiac urethral duplication in an 8 years old boy with an ex diagnosis of bilateral vesicoureteral reflux, a Case report**

*Khaki, S., Mortazavi Sh., Kajbafzadeh, AM., Pediatrics Urology Research Center, Tehran University of Medical Sciences*

**Background:** Urethral duplication is a rare congenital anomaly of the urinary system which is characterized by two urethral canals and presents in three anatomic variants of epispadiac, hypospadiac and Y-type. It may present with a double stream or with urinary tract infection (UTI).  
**Case presentation:** We report a case of hypospadiac urethral duplication in an 8 years old boy referring to children's medical center with a chief complaint of recurrent UTI and bilateral vesicoureteral reflux. After conducting voiding cystourethrogram (VCUG) in this case, bilateral vesicoureteral reflux (VUR) as the third grade for the right and the second grade of the left part was detected. Then the patient underwent cystoscopy and endoscopic treatment in which the right and the left ureters' orifices were observed as golfhole and horsehole respectively in the normal anatomical area with moderate bladder trabeculation. Dorsal ductal hydrodilatation was

then performed. Followed by the diagnosis of hypospadias in the physical examination including two dorsal and ventral urethral canals, the repair of hypospadiac urethral duplication was done through urethroplasty. As the ventral urethra is almost always the functioning channel and the dorsal urethra is hypoplastic, atretic, or abnormal at hypospadias repair, due to hypoplastic dorsal urethra in this case, the dorsal urethra was ablated and the ventral urethra was anastomosed to sagittal plane of the urethral area. At postoperative radiological evaluation, no other associated anomaly was revealed. The patient had no postoperative complications and then was discharged. At further assessments, urethroscopy and cystoscopy were performed and found to be normal. Conclusion: In any child presents with a the duplication of the urethra along with hypospadias thorough investigation is necessary and further surgical and medical evaluations are needed to establish a normal stream and prevent the occurrence of the recurrent UTIs.

**Keywords:** Hypospadiac, Urethral duplication, Bilateral Vesicoureteral reflux, Urethroplasty

### **Assessment of growth indexes and its correlation with type of rheumatoid arthritis in patients with chronic juvenile arthritis in pediatric rheumatology clinic of Imam Khomeini hospital**

*Khadamy, J., Mousavi-Nia, SF., Medical Student Aghighi, Y., MD, Moayeri, H., MD, Raeeskarami, SR, Vali-e-Asr (as) Pediatric Raees Karami SR., MD, Vali-e-Asr Hospital, Tehran University of Medical Sciences*

**Background:** Chronic inflammatory arthritis is the most prevalent pediatric rheumatoid disease, an inflammatory process which other than causing to fixed deformities also to developmental problems, including growth inhibition.

**Methods:** In the study growth indexes of patients suffering from chronic juvenile rheumatoid arthritis was measured. And correlation between age at onset, age at diagnosis date, patient gender, systemic corticosteroid administration, previous history hospitalization, duration of active arthritis, type of arthritis, patient functional class, patient appetite, positive family history, last hemoglobin, ESR, RF, CRP and growth indexes (BMI, weight, length) were assessed. Materials and Methods- In this cross sectional study, 102 known cases of juvenile rheumatoid arthritis with simple sampling method were selected. Height, weight and BMI of patients were compared with age and sex matched normal values and SDS was calculated. Other than was applied for evaluation of weight, height and BMI growth situation with other variables. SDS of weight, height and BMI were calculated.

**Results:** Mean ( $\pm$  SEM) of weight SDS was  $-0.68(\pm 0.160)$ , Mean ( $\pm$  SEM) of height SDS was  $-0.957(\pm 0.172)$  and Mean ( $\pm$  SEM) of BMI SDS was  $-1.93(\pm 0.094)$ . Weight SDS was correlated with hemoglobin, previous history of hospital admission, JRA type and function class and height SDS was correlated with hemoglobin, previous history of hospital admission, kind of therapy and function class and BMI SDS was correlated with JRA type.

**Conclusion:** According this study, growth disturbance is associated with more severe disease and inflammation.

**Keywords:** Growth indexes, Chronic juvenile arthritis

### **Isolation of alkaligenesis from the cerebrospinal fluid of an 18-month-old male child with meningitis and arthritis**

*Abdollahi, M., Daneshju, Kh., MD, Izadi, P., Sasanpoor, P. Department of Pediatrics, Imam Khomeini Hospital, Tehran University of Medical Science; Sport Medicine Research Center, Tehran University of Medical Sciences*

Achromobacter xylosoxidans is a rare cause of bacteremia. Literature review illustrates that A. xylosoxidans bacteremia mostly occurs in predisposed and immunosuppressed patients particularly in those with concurrent neoplastic diseases. Wide range of clinical manifestations including primary bacteremia, pneumonia, catheter-associated bacteremia, meningitis, endocarditis, cholecystitis, peritonitis and pyelonephritis has been reported for patients infected by Achromobacter xylosoxidans. A case of arthritis and meningitis due to Achromobacter in a previously healthy patient is reported herein. Strains of Achromobacter xylosoxidans are commonly insensitive to routine anti-microbial therapy. Tazocin was the preferred antimicrobial regimen in treatment of our patient. To our knowledge, this is the first documented case of A. xylosoxidans with presentation of arthritis.

**Keywords:** Alkaligenesis, Arthritis, Achromobacter, Immunodeficiency

### **The evaluation of ventricular septal defect presentations in children admitted at Mashad Ghaem hospital**

*Hashemian, M., Mahmudi, E., Sabzevar University of Medical Sciences*

**Background:** Congenital heart disease affects 1/100 births. Ventricular septal defect (VSD), the most common

congenital heart disease, has variable presentations as american text; but not obvious in our country. Hence we investigated these presentations in this article.

**Method:** We assessed patients with VSD, who were admitted at Ghaem hospital (Mashad, Iran) from March 1991. Findings: 130 patients recruited into the study. The most common chief complaint was exertional dyspnea (96.9%) and failure to thrive (30%). the most common sign was systolic murmur(65%) and thrill (65%). Seventy percent of patients had right ventricular hypertrophy in Electrocardiogram. The most common abnormality in XR was increased cardio- thoracic ratio, and increased pulmonary vascular marking. The most common associated anomalies were Pulmonary stenosis(PS), Atrial septal defect(ASD), Tricuspid regurgitation (TR) and Aortic insufficiency(AI).

**Conclusion:** In children with exertional dyspnea, failure to thrive, systolic murmur and thrill, we should think of VSD.

**Keywords:** Congenital heart disease, Ventricular septal defect, dyspnea

### **Ambulatory blood pressure monitoring: a great tool for early detection of hypertension in reflux nephropathy**

*Saedi, M., MD, Hooman, N., MD, Tavassoli, A., MD, Eisa- Tafreshi, R., MD; Iran University of Medical Sciences, Tehran*

**Background:** Reflux nephropathy is a common problem in childhood and is the most probable cause of secondary hypertension (HTN) in this age group. The aim of this study was to determine the prevalence of hypertension in Iranian children with renal scars after recovery from urinary tract infection (UTI) by ambulatory blood pressure monitoring (ABPM) as well as casual clinic blood pressure (BP) measurements. We also evaluated end organ damages in terms of microalbuminuria, left ventricular hypertrophy (LVH) and sleep disturbances. Although similar studies are performed in other countries, such sort of investigations has not been reported in Iranian patients so far.

**Methods:** Seventeen patients aged 5-15 years with at least one year history of reflux nephropathy and normotensive according to casual blood pressure measurements were included in this study. According to the severity of renal scar, the subjects were divided in three groups and underwent 24 hours ABPM. Data of microalbuminuria, glomerular filtration rate, echocardiography and sleep score were also compared in these groups.

**Results:** Although based on casual BP measurements all patients were normotensive, ABPM data revealed a prevalence of 23% of HTN in the examined subjects.

Mean ABPM systolic and diastolic load were greater in the group with the most severe renal scar compared to the mild and moderate groups. Forty seven percent of the patients did not show the normal nocturnal drop in BP (non-dipper). ABPM data indicated that these patients had higher systolic BP load compared to the other subjects. Accordingly, the mean casual systolic and diastolic BP was higher in these non-dipper subjects. Eighteen percent of the patients had poor sleep quality index which was associated with higher ABPM systolic load and mean casual systolic and diastolic BP. None of the examined subjects had microalbuminuria or LVH.

**Conclusion:** It could be concluded that compared to LVH and microalbuminuria, HTN is an early complication of reflux nephropathy. In addition, our data indicate that ABPM could be suggested as a sensitive method for screening of HTN in these patients.

**Keywords:** Reflux Nephropathy, Ambulatory Blood Pressure Monitoring, Hypertension

### **Cutaneous granulomas in common variable immunodeficiency: case report and review of literature**

*Abolhassani, H., Rezaei, N., Kalantari, N., Tamizifar, B., Cheraghi, T., Parvaneh N., Yeganeh, M., Moazzami, K., Ebrahimi-Daryani, N., Anaraki, MR., Aghamohammadi, A.; Children's Medical Center, Tehran University of Medical Sciences*

Common variable immunodeficiency (CVID) is a heterogeneous disease characterized by recurrent infections, autoimmunity, malignancies, and granulomatous inflammation. Granulomatous lesion is one of the important manifestations of CVID, which continues to be unknown to many clinicians. While noncaseating granulomatous lesions can be detected in lungs, liver, spleen or conjunctiva of CVID patients, there are only few reported cases with skin granuloma. This report presents a 27-year-old female with multiple persistent cutaneous granulomatous lesions on both hands. The patient had been well until age of 20 years, when she developed these skin lesions and frequent upper respiratory infections and bacterial pneumonia. Also, she experienced recurrent diarrhea (more than 10 episodes). Laboratory evaluation showed decreased serum levels of all immunoglobulin isotypes and low specific antibody responses. The diagnosis of CVID was based on clinical and laboratory findings. Intravenous immunoglobulin therapy at a dosage of 400-500 mg/kg monthly was introduced and improved skin lesions. In conclusion, taking history of recurrent infections and measuring immunoglobulin levels can be suggested in patients with granulomatous lesions instead of other expensive tests

**Keywords:** Common variable immunodeficiency, Granulomatous

### Dental treatment of children under General anesthesia

*Baghalian, A., Afshar, H., DDS, Tehran university of Medical Science, Dental School*

**Background:** Some children can not tolerate dental treatment under local anesthesia, barriers to treatment may be dental fear or behavior management problems or some sort of medial problems of patient. The indications for dental treatment under general anesthesia have been described by various authors as extensive decay, behavioral management problems, a medically compromised patient, a handicapped patient, and a combination of these. General anesthesia can be considered an option for dental treatment of most of the children who aren't able to communicate because of little age or mental retardation.

**Method:** A literature search using the pubmed database was conducted. The following keywords were used in the search: general anesthesia, pediatric dentistry. The original literature search encompassed articles from 1990 till present. Also we have treated a lot of cases under general anesthesia at pediatric medical center with follow up visit and meaningful treatment success.

**Results:** The use of general anesthesia for dental care in children is sometimes necessary to provide safe, efficient and effective care. Depending on the patient this will be done in an ambulatory care setting or inpatient hospital setting. It should be only one component of the dentist's overall treatment regimens. Oral hygiene and preventive care must be implemented at the onset of treatment with parents or guardians and patients to eliminate the cause of dental problem. The safety of the patient as well as the need to diagnose must justify the use of general anesthesia.

**Conclusion:** Most of the literature that pertains to use of general anesthesia for dental treatment agrees on its appropriateness for facilitation of treatment for fearful and/or behaviorally resistant individuals. There is, however, very little in the way of solid evidence to support other decisions, such as exactly what constitutes appropriate procedures to be performed or optimal frequency of treatment under general anesthesia. At the current time with the lack of meaningful evidence in this area, it remains for each practitioner to be knowledgeable about each individual patient's cognitive functioning, behavioral capabilities, medical status and dental treatment needs.

**Keywords:** Pediatric dentistry, General anesthesia

### Behavior abnormality following intravenous immunoglobulin treatment in patients with primary antibody deficiencies

*Hyrbod-Mobarakeh, A., Saroukhani, S., Aghamohammadi, A., Mahmoudi-Gharaei, J., Abolhassani, H., Cheraghi, T., Imanzadeh, A., Moazzami, K., Parvaneh, N., Rezaei, N.; Children's Medical Center, Tehran University of Medical Sciences*

**Background:** Treatment with intravenous immunoglobulin (IVIG) is considered a safe therapy for patients with primary antibody deficiencies (PADs), whilst adverse effects have been frequently reported. Meantime behavioral disorders reactions have not been reported yet. In this study, we describe for the first time a group of patients with PADs, who were under IVIG therapy and experienced some behavioral disorders.

**Methods:** Five patients, including two hyper IgM syndromes, one X-linked agammaglobulinemia, one common variable immunodeficiency, and one hypo IgM disease, were surveyed. Analysis of Conner's Parents Rating Scales-Revised Short (CPRS-R:S) and child behavior checklist (CBCL) was performed for the patients, suspected to hyperactivity.

**Results:** Analysis of CPRS-R:S showed an evidence of mild hyperactivity before IVIG administration in four patients, whereas another patient had evidence of severe hyperactivity. After IVIG administration, hyperactivity scores of three patients were changed from mild hyperactive behavior to markedly hyperactive behavior or attention deficit hyperactivity disorder range of hyperactivity. In the CBCL scores, there were abnormal externalization scores for three patients; while two remaining patients had abnormal internalization scores. **CONCLUSIONS:** Although predisposition to behavioral disorders can be due to a genetic background, further investigations are necessary to test the hypotheses about responsibility of either IVIG or underlying disease in progression of behavioral abnormalities

**Keywords:** Adverse drug reaction, Behavior abnormality, Intravenous immunoglobulin, Primary antibody deficiencies, Hyperactivity

### Eventration of the diaphragm: a case report

*Abbasi, M., Jouybari, LM., Golestan University of Medical Science, Students Research Committee*

**Background:** Diaphragm of eventration is a defect of whole or a part of diaphragm muscle and replacing it with fibroplastic tissue. The incidence of eventration is uncertain, although according to a study it occurred in 1 in 1400 patients based on chest radiography. Diaphragmatic

eventration can be congenital or acquired; isolated or association with other abnormalities. It is also more common in male than female newborn. The aim of this article is reporting a newborn with congenital diaphragmatic Eventration.

**Case report:** The study conducted in a central teaching hospital in Gorgan, North of Iran in 2010. The case is dysmorphic male premature newborn which born at 33 gestational age. He was the first child of the family and the mother received prenatal care during her pregnancy period. Because of Oligohydramnios, preterm labor and fetal distress cesarean section was done at 32 weeks of pregnancy. There was no abnormality history among the family. Due to sever respiratory distress the patient admitted immediately in NICU and underwent associated ventilation. Thoracotomy done after three day however no improvement observed. He expired after 30 days.

**Conclusion:** Considering that the parents rejected genetic analysis we as health care givers are responsible to inform people about prevention of genetic abnormalities and importance of genetic consoling before decision for having a child.

**Keywords:** Eventration of the diaphragm, case report, Iran

### Protective role of bilirubin as an antioxidant from free radical related illnesses among neonates

*Poorsattar-Bejeh-Mir, A., Poorsattar-Bejeh-Mir, K., MD, Fereshtehnejad, SM., MD, MPH, Mohagheghi, P., MD; Center for Gifted & Talented Students; Medical Education and Development center (EDC), Babol University of Medical sciences; Amir Mazandarani General Hospital, Sari; Iran University of Medical Sciences and Health Services*

**Background:** There are disparate data regarding whether protective or toxic charectersitic of bilirubin considering free radical related illness among neonates.

**Methods:** Seventy one infants with gestational age (GA) of <32 wk and/or birth weight (BW) of <1500 g, who survived beyond 4 weeks and completed physical examinations were enrolled in this study. Regarding the presence or absence of advanced retinopathy of prematurity (ROP), grade III intraventricular hemorrhage (IVH), grade III necrotizing enterocolitis (NEC), respiratory distress syndrome (RDS), bronchopulmonary dysplasia (BPD), sepsis or severe fungal infection (SFI) the infants were divided into two groups. And the mean of total serum bilirubin (TSB) of the first 14 days of life were measured and compared between these two groups.

**Results:** A significant lower TSB were found in severe form of ROP (P <0.001), grade III NEC (P=0.008), grade III IVH (P=0.021), SFI (P=0.003) and sepsis (P=0.007) in comparison to mild or disease free status. Moreover, the

cut-off point of 5.1 mg/dl for the mean of TSB had the sensitivity of 88.1% and specificity of 84.6% to detect severe grades of ROP. Also the cut-off point of 3.25 mg/dl had 97.2% sensitivity and 100% specificity in order to distinguish SFI.

**Conclusion:** It is concluded that bilirubin may play an antioxidant role in vivo as in vitro; and protect preterm infant against these free radical related disorders. Our findings suggest that not only the upper limits of serum bilirubin, but also the lower limits must be taking into account in order to both preventing from neurotoxic effects and free radical based illnesses, respectively.

**Keywords:** Bilirubin, Free Radical, Preterm, Antioxidant, Retinopathy of prematurity Sepsis, IVH

### Gaps in mothers' knowledge and understanding on childhood immunization in health centers in Iran

*Khaheshi, I., Sedaghat, M., Habibi, A., Salehi-Sadaghiani, M., Torkashvand, A.; Department of Community Medicine, Tehran University of Medical Sciences, Tehran*

**Background:** Immunization during childhood is one of the major factors in improving children survival especially in families suffering from poverty. Improvement in maternal education and social interaction also improves the child vaccination. **Methods:** This study was carried out in 2 villages and 2 suburban health centers in the district of Tehran. A questionnaire was designed in order to assess the knowledge of mothers on vaccination.

**Results:** The literacy level of mother had significant correlation with knowing the name of vaccination (p value = 0.04) and the aim of vaccination (p value = 0.005).

**Conclusions:** This study showed the importance of education in improving health status of the society.

**Keywords:** Childhood immunization. Knowledge. Iran

### Use of complementary and alternative medicine for children with epilepsy in three hospitals in Tehran (2009-2010)

*Djafari-Naeini, S., Pediatric Neurology Research Center, Mofid Pediatric Hospital*

**Background:** Although the use of complementary and alternative medicine (CAM) have been studied globally, in our country, there are a few studies which have worked on it. The aim of this study was to determine the

prevalence, pattern of use, parental sources of information and adverse effects of CAM in children with epilepsy in Mofid and Emam Hossein and Children medical center from 2009 to 2010.

**Methods:** 92 parents or relatives of epileptic children who presented to pediatric neurology clinics or were admitted at ward, were randomly interviewed. The information obtained by the questionnaire was some demographic data from patients and parents, the type of CAM, if any, used by the patients and the sources and effects of the CAM used. Also they were asked if they had consulted the physician about using CAM.

**Results:** A total of 38 CAM were used by the patients from 92 people, either alone or in combination with other CAM. The most frequently used CAM was writing the prayers and the next was herbal medicine and supplements. CAM was introduced to them by relatives mainly. Only 20.5 percents of parents who had used CAM before, had discussed with their physicians about it. The main result was having no effects on seizure control. The mean number for using these methods was  $0.67 \pm 0.81$  for each responder.

**Conclusion:** This study showed that use of CAM is common in our country and it can make an effect in patient's treatment. So it is necessary for physicians to get enough information about CAM use in their patients.

**Keywords:** Epilepsy, Complementary medicine, Alternative medicine

### The effect of a period aquatic exercise therapy on muscle strength and joint's range of motion in hemophilia patients

*Dehghadani, M., Kargarfard, M., MSc; Faculty of Physical Education and Sport Sciences, University of Isfahan*

**Background:** Hemophilia is a X-linked genetically transmitted disorder, which is caused by a deficiency in a circulating blood clotting factor. The most common type is classic hemophilia that occurs in one in 10,000 male newborns. The Purpose of this study was to Evaluate The effect of a period aquatic exercise therapy on muscle strength and joints range of motion in hemophilia patients.

**Method:** The Present study is a Semi-Experimental and pre test-post test with control group Investigation. the hemophilia patients who referred to Sayedo- Shohada hospital, Were statistical society of this study. Twenty moderate to sever hemophilia child and teen selected accessible and targeted, then placed randomly in experimental (10 persons) and control (10 persons) groups. Aquatic exercise therapy group's subjects, began activity in water for 8 weeks, 3 sessions per week of 45 to 60 minutes, while the control group, was only under

follow-up, and during this period did not experience any effective physical activity. The patients' muscle strength and joint's range of motion were evaluated, through standard laboratory tools, using a isokinetic dynamometer (Biodex, Systems III) and a standard goniometer, In the beginning and end of period. Finally, data obtained, were analyzed using analysis of covariance (ANCOVA) in  $p \leq 0.05$ .

**Results:** in this study the strength of the muscles around knee joint for perform extension and flexion movements, significantly were increased in the experimental group while the control group remarkable change observed. Range of motion also was improved than before the course in the experimental group in all joints, while the control group staid without significant change.

**Conclusion:** the results showed that aquatic exercise therapy can be a useful method, to improve joints' strength and range of motion in hemophilia patients and follow it improve their daily functioning and quality of life.

**Keywords:** hemophilia, aquatic exercise therapy, muscle strength, joint range of motion.

### Hearing loss and ear defects in newborns conceived by ART

*Ahmadi, SA., Shahzadehfazeli, SA. Gourabi, H., Nateghi, MR., Ashrafi, M., Jarollahi, F., Afsharipur, S., Kouhpayehzadeh, J.,*

*1. Child Health and Development Research Center, Iran Medical Science Branch of ACECR, Tehran; 2. Royan Institute for Reproductive Biomedicine, ACECR, Tehran; 3. Iranian Biological Recourse Center, Tehran; 4. Faculty of Medicine, and Faculty of Rehabilitation, Iran Medical Science, Tehran,*

**Background:** The assisted reproductive techniques (ART) are used more frequently throughout world. The present research was conducted to determine the effects of these techniques on hearing defect and ear abnormalities.

**Methods:** In a descriptive, cross-sectional and non-randomized study, the status of hearing and ear abnormalities was assessed in 300 newborns conceived by ART in Royan Institute, Tehran, for sixteen months. The data were collected from parents, otoscopic examination and otoacoustic emissions (OAEs) test of newborns. The external ear was assessed by otoscopic examination, then OAE test, an objective test that does not need to collaboration of infant, was performed by audiologist. In this test, the OAE wave was registered after a click (stimulus) in 5-20 millisecond intervals with 82 dB SPL altitude. The data were analyzed by statistical tests.

**Results:** 300 cases were examined by otoscopy. This examination showed that, 2 cases (0.66%) had bilateral malformation in auricle, 2 cases (0.66%) had unilateral perforation of tympanic membrane, 5 cases (1.66%) had unilateral retraction of tympanic membrane, 8 cases (2.66%) had bilateral retraction of tympanic membrane, 1 case (0.33%) had unilateral tympanic membrane inflammation, 1 (0.33%) case had bilateral tympanic membrane inflammation and 1 case (0.33%) had obstruction of external ear canal by wax. 289 cases of 300 newborns were tested by OAEs test. 3 cases (1.03%) did not have bilateral registered wave and had bilateral hearing loss.

**Conclusion:** This study shows that the hearing and ear screening in newborns conceived by Assisted Reproductive Techniques is contemplative and emphasizes the profitability of continual check up in these newborn infants.

**Keywords:** Newborn, Hearing, Assisted Reproductive Techniques, Ear

### Sublingual traumatic ulceration in a CP child

*Kay, G., Pediatric Dentistry of Tehran University*

Sublingual traumatic ulceration in a CP child Riga-Fede disease (RFD), first described in 1881 by Italian physician Antonio Riga and in 1890 by Fede who performed histological studies of the lesions, is a benign and uncommon mucosal disorder. and usually presents with ulceration of the oral mucosa and usually the ventral surface of the tongue. It is most commonly associated with natal or neonatal teeth in newborns. it may also occur in older infants after the eruption of primary lower incisors with repetitive tongue thrusting habits, or in children with familial dysautonomia (in sensitivity to pain), or, in children with cerebral palsy Dyskinesias of various body parts accompanies CP, including the tongue, which may predispose the infant to this lesion when the tongue is involved. Although more common in infants, RFD has been reported in older patients and, recently, in patients with acquired immunodeficiency syndrome. Individual lesions present as erythema surrounding a centrally removable, yellow, fibrinopurulent membrane. Often, the lesion develops a rolled white hyperkeratotic border immediately adjacent to the ulceration. As it often mimics many oral malignant and benign disorders, the differential diagnosis is important. Case report An 10-month-old infant boy suffering from cerebral palsy (CP) presented with a large exophytic lesion located on the anterior ventral part of the tongue.(figure1)the parents reported that the child had inadequate nutrient intake, and difficulty in suckling. Contact of the anterior part of the tongue with the newly erupted anterior mandibular teeth was suspected to be the

cause. so, two central incisors was extracted. after 2 months the parents came back and complain about the return of the lesion according to the eruption of the lateral incisors (figure2). But this time we decided to grind round the incisal edges of the anterior mandibular teeth and covering with composite resin material. The patient did not return for follow-up. During a telephone interview with the mother, 6 year later, she reported that the ulceration of the tongue was almost completely healed.

**Conclusion:** Although these oral lesions are self-limiting and may heal spontaneously, delayed or false diagnosis and ineffective therapy may result in mutilation, permanent deformity of the tongue, nutritional insufficiency, and growth retardation in the long term. Restorative dental treatment such as placement of a protective barrier over the sharp edge of the incisor teeth and smoothing of rough incisor edges, changing feeding habit or style (ceasing use of a feeding bottle), and topical corticosteroid application can be used as conservative treatment options. Extraction of the lower incisor teeth has been suggested as a more radical treatment choice.

**Keywords:** sublingual traumatic ulceration, Riga-Fede Disease

### Obesity determinants and effects on children's health

*Jabbari, H., Fazelipoor, S., Medical Student, Azad University of Medical Sciences, Tehran*

Obesity is a multi-factorial syndrome involving genetic, environmental and behavioral alterations. In this study, daily energy and macronutrient intake, physical activity, parental education, season of birth, frequency for eating fast-foods and fizzy drinks, time spent watching TV and videos or working with computers turned out to be risk factors. Some factors in the infancy can lead to a deleterious effect on childhood Obesity. The results indicate that the mean birth weight is not significantly different between the obese children and normal children while frequency and duration of breast feeding is significantly lower in obese children than the normal children. Externalizing behaviors in early childhood are associated with children's weight status early in childhood and throughout the elementary school years. Also, there is a positive association between a range of family stressors (lack of cognitive stimulation, emotional support and financial strain) and child overweight or obesity. Moreover, regular physical activity is an important factor in reducing the prevalence of overweight and obesity. The rural population presents higher prevalence for malnutrition while in the urban area there is higher prevalence of risk for overweight in children. Also in the same situation the prevalence of obesity is more in black or another race in comparison with white

children. Obese children present changes in the autonomic nervous system characterized by decreases in parasympathetic activity and overall variability. Most obese girls are born in winter or autumn while non-obese girls are born mostly in spring and summer. Overweight and obesity are marginally higher in the pubertal age groups of 13 to 15 years, perhaps because of increased adipose tissue and overall body weight in children during puberty. Also, one of the major reasons for childhood obesity is watching television or using computers. TV viewing and male gender have direct association with eating fast food, sugar foods and pastry pattern so obesity while maternal education and longer sleeping have positive association with eating fruit and vegetables in children

**Key Words:** Obesity, Overweight, Children

### Comparing quality of life in children with hemophilia receive prophylactic treatment and without it in Tehran city

*Khanali, L., MS, Abdallah-gorgi, F, MS, Eshghi, P., MD, Abedsaedi, J. PhD, Farahani, H., PhD*

**Background:** Assessment of quantitative parameters is easier than qualitative ones. But it should not be forgotten that it is important to know quality of life (QOL) as a qualitative parameter in medicine to evaluate and if necessary to improve the patients care. Severe hemophilia A and B can lead to recurrent bleeding in joints, deteriorate joints functions and significantly affect the QOL. Aims. The aim of this study is to determine and compare the quality of life in children (4-7 y) with severe hemophilia A and B in two groups (prophylaxy and on- demand treatment) in the hemophilia centers of Tehran.

**Methods:** 30 registered patients with severe hemophilia A or B in every group (prophylaxy and on- demand) were asked to fill out the standard Haemo-Qol questionnaire. This questionnaire was translated to Persian language by authors, pediatric hematologist and an epidemiologist (all together). According to patients self-report and on the base of scoring list of standard Haemo-Qol questionnaire, and descriptive and Inferential statistics the QOL was determine and compare for each groups.

**Results:** In group prophylaxy, according to self-reports they had lower QOL in family subscale [mean 58/9 (SD: 14/9)]. In group on- demand according to self-reports they had lower QOL in physical subscale [mean 65 (SD:16/2)] and family subscale [mean 66 (SD:12)]. On the whole, patients with prophylaxy had a better QOL [mean 40/9 (SD: 9/8)] in comparison with patients in on-demand group [mean 58/3 (SD:10/9)].

**Conclusions:** According to results it seems that most of our patients in prophylaxy group experience a better QOL. Further efforts are necessary to considering

therapies that reduce bleeding events, like prophylaxis treatment, warrant more attention. 1-Shahid Beheshti University Of Medical Sciences, Tehran, Iran 2-Islamic Azad University Of Medical Tehran, Tehran, Iran 3-Pediatric Mofid Hospital, Tehran, Iran

**Keywords:** Hemophilia, Quality of life, Prophylaxy

### The Papillon-Lefevre syndrome -case report

*Kamali-sabeti, A., Bagheri, M., Kay, G., Tehran university of Medical Science, Dental School*

The Papillon-Lefevre syndrome (PLS) is a rare genodermatosis of autosomal recessive inheritance manifesting as palmer plantar hyperkeratosis with periodontitis. It was first described by two French physicians, Papillon and Lefevre, in 1924. It has a prevalence of 1-4 cases per million persons and both males and females are equally affected with no racial predominance. The disorder is characterized by diffuse palmoplantar keratoderma and premature loss of both deciduous and permanent teeth. The palmoplantar keratoderma typically has its onset between the ages one and four years. The sharply demarcated erythematous keratotic plaques may occur focally, but usually involve the entire surface of palms and soles resulting in foul-smelling odor. Well-demarcated psoriasiform plaques occur on elbows and knees. This may worsen in winter and be associated with painful fissures. The second major feature of PLS is severe periodontitis, which starts at the age of three or four years. The development and eruption of deciduous teeth proceeds normally, but their eruption is associated with gingival inflammation and subsequent rapid destruction of the periodontium. The resulting periodontitis characteristically is unresponsive to traditional periodontal treatment modalities and the primary dentition is usually exfoliated prematurely by the age four years. After exfoliation, the inflammation subsides and gingiva appears healthy. However, with eruption of the permanent dentition the process of gingivitis and periodontitis is usually repeated and there is subsequent premature exfoliation of the permanent teeth, although the third molars are sometimes spared. The case is a 7-year-old girl who presented with deciduous tooth loss. The parents and other family members were not affected. Pregnancy and delivery were normal. The mother had noticed skin lesions on the palms and soles of the child when she was 5 months old.

**Keywords:** Papillon-Lefevre syndrome, progressive periodontal disease

### Molecular epidemiology of human respiratory syncytial virus in Iranian children less than 5 years in 2007

*Faghihloo, E., Rezaei, F., Salimi, V., Naseri, M., Mamishi, S., Mahmoodi, M., Mokhtari-Azad, T., Tehran university of Medical sciences*

**Background:** Human respiratory syncytial virus (HRSV) is the most important viral agent of acute lower respiratory tract disease in infants and young children worldwide. This virus is responsible for 50% bronchiolitis and 25% pneumonia in infants. There are limited data of molecular epidemiology of HRSV from developing countries. This is the report on the molecular epidemiology of human respiratory syncytial virus in Iran.

**Methods:** In this study, RT-PCR for second hypervariable region of the HRSV G glycoprotein was performed on 72 throat swabs collected from children less than 5 years of age with acute respiratory symptoms in 1386.

**Results:** Of the 72 throat swabs collected from children with acute respiratory symptoms, 14 (19.44%) were positive for HRSV. Phylogenetic analysis revealed that all HRSV-positive samples clustered in three genotypes of subgroup A: 12 strains (85/71%) in genotype GA2, 1 strain (7/1%) in genotype GA1, and 1 strain (7/1%) in genotype GA5. In this study we couldn't identify any genotype of subgroup B.

**Conclusion:** Our results revealed that multiple genotypes of subgroup A were cocirculated during 1386 in children less than 5 years of age in Iran. Also this study revealed that genotype GA2 was predominant genotype in isolates were obtained from several cities (Tehran, Isfahan, Karaj, Qazvin, Bandar Abbas, Shahreza), so we speculate that this genotype may be predominant during 1386 in Iran. This study supported that RT-PCR for second variable region of G protein is an effective method for further studies of HRSV genotype designation in Iran.

**Keywords:** Human respiratory syncytial virus, Molecular epidemiology, Children, Iran.

### Genetic diversity in the g protein gene of human respiratory syncytial virus among Iranian children with acute respiratory symptoms, 2009

*Faghihloo, E., Salimi, V., Rezaei, F., Naseri, M., Mamishi, S., Mahmoodi, M., Mokhtari-Azad, T.; Tehran university of Medical sciences*

**Background:** Acute respiratory infection (ARI) is the major cause of morbidity and mortality in children

worldwide. Human respiratory syncytial virus (HRSV) is main viral agent of ARI in infants and young children in terms of effect and prevalence. The aim of this study was to investigate HRSV genotypes during one season in Iran.

**Methods:** In this cross-sectional study, a total number of 107 throat swabs collected from children less than 5 years of age with acute respiratory infection from October to December 2009. The respiratory samples were obtained from several provinces; Tehran, Isfahan, Hamadan, Zanjan, Kordestan, Lorestan, Azarbayjan garbi and were tested for G protein gene of HRSV by RT-PCR.

**Results:** Of the 107 respiratory samples, 24 (22/42%) were positive for HRSV, that 16 (66/6%) belonged to subgroup A and 8 (33/4%) to subgroup B. Phylogenetic analysis revealed that subgroup A strains fell on two genotype GA1 and GA2, whereas subgroup B strains clustered in genotype BA.

**Conclusion:** This study revealed that multiple genotypes of HRSV were cocirculated during the season 2009 in Iran. Also subgroup A strains were more prevalent than subgroup B strains, and genotype GA1 was predominant during this the season.

**Keywords:** Human respiratory syncytial virus, Genotype

### Infectious outcomes in the injured children hospitalized in Tehran and Qom University Hospitals after the bam earthquake in Iran in 2003

*Motamedi, A., MD, Ziaee, V., MD, Raeeskarami R., MD; Tehran university of Medical Sciences*

**Background:** Many earthquakes occur in our country every year, leaving great number of people killed or injured. For instance an earthquake with intensity of 6.3 devastated the city of Bam and adjacent villages, leaving more than 26,000 people killed and 30,000 injured. Many children lost one or both of their parents. Many of children suffered from disabilities and many kinds of infectious disease may occur during natural disasters. These children witnessed the most stressful event throughout their life while they were not prepared for such a devastating natural event.

**Methods:** We considered the consequences of infectious complications, their different kinds and also the mortality and morbidity in a group of children suffered from Bam earthquake who were admitted in Tehran and Ghom medical university hospitals in 2003. We reviewed the hospital documents of all patients suffered from Bam earthquake with inclusion criteria of being 18 or under 18 years old, retrospectively. Data analyzed with SPSS version 17.5 using descriptive analysis for describing the demographic results. We used t-test, chi-square and Odds ratio to analyze the data.

**Results:** 21(20.4%) patients had some kind of infectious complications. The different kinds of infections are as follows: 13(12.6%) wound infection, 4(3.9%) urinary infection, 2(1.9%) pneumonia, 2(1.9%) septicemia, 2(1.9%) cellulites, 2(1.9%) necrotizing faciitis, 1(1%) osteomyelitis, and 1(1%) peritonitis. According to our findings, there were infectious complication in 17 patients from 55 patients with lower limb trauma and this relation was statistically significant. There was a significant correlation between the number of days after hospitalization and the frequency of infection exist and a significant relation between surgical procedures especially orthopedic procedures and the frequency of infectious complications. 84 patients (81.5%) had complete relief at the end, 18 patients (17.4%) had some kind of morbidity, and one 4 year old girl (0.9%) died due to sepsis.

**Conclusion:** It seems that the infections prevalence and incidence of these complications can be effective to conduct the health funding and planning for natural disasters and management of crisis.

**Keywords:** Infectious outcomes, Children, Earthquake

### Compare the mental health of mothers of children with exceptional mental health of mothers of children normal

*Ahmadi, A., Shafiei-Amiri, M., Shabani, S., Zahmatkesh, Z.; Islamic Azad University & Young Researchers Club, Qhaemshahr Branch, Qhaemshahr; Islamic Azad University & Young Researchers Club, Roudehen Branch, Roudehen; Allameh Tabatabai University; Islamic Azad University, Tabriz*

**Background:** One of the most natural Hhayy pledges that human needs can be satisfied families. Family child care task Vtrbyt it, healthy communication with members and help, children's independence, even if mentally retarded child, is blind or deaf. The process of child birth is a joy for parents, although this process with derivatives and frequency is associated with discomfort. In spite of these problems must be said that the hope of being healthy and normal child usually feels confidence in them to create and accept their child, but as soon as the knowledge of parents of disabled child, all hope will be a disappointment converter problems begins.this study compared mental health of mothers of children Bamadran exceptional children is normal. **METHODS:** This study's method is comparative. The sample of this research consisted of 100 mothers of exceptional children and mothers of elementary school students in ordinary city of Babol were randomly selected. Tool used by a 90 item scale, SCL-90-R is. To analyze data statistically independent T method through SPSS software was used. Findings: The results of the findings showed that in

general mental health among mothers and mothers of normal children in exceptional children have significant difference. Also in terms of mental health symptoms, ie depression, anxiety, mental Parish, aggression and phobia have significant difference. But in terms of somatization, obsessive - compulsive, sensitivity in relations and ideas Paranoid significant difference between groups has not been observed.

**Conclusion:** This study showed that mothers of exceptional children than mothers of normal children have lower mental health.The first person who directly communicate with the child to mother. Madrvqty faced with a handicapped child can be (especially if mental retardation is) due to constant maintenance and the need to provide special conditions and development of these children face stress behavior such as format, language difficulties, Qshqqr and lack of self-care skills, undermine normal function of the mother is.

**Keywords:** Mental health, Mothers Exceptional Children, Normal children

### Epidemiological survey of measles in the cities covered Gonabad University of Medical Sciences Year 2006-9

*Ramazani-Awal-Riabi, H., Gonabad University of Medical Sciences*

**Background:** A viral disease, measles is an acute contagious attraction rate in developing countries is 5-1% and 30-10% in some countries is estimated. Improve immunization coverage (over 95%) reduced cases of measles and rubella, and reducing child mortality has to accompany. Our country due to high immunization coverage to reach 95% capacity is removed to reach the stage that all the efforts of health authorities to require this study to evaluate the risk of measles and centers have been set.

**Methods:** We conducted this study to the necessary information from the measles elimination program review forms, including linear lists measles and measles epidemiology review individual form from health centers - public and private health city health centers were sent were extracted.

**Results:** Results from surveys conducted over the past four years a total of 10 cases of health centers - public and private treatment has been reported that four males and four females. 8 months of age  $\pm$  12 years old, nine were rural and one urban. 30% have a history of vaccination dose and 40% had history of measles immunity in turn and the rest of this age were not making. Index sampling (three samples of throat, urine and serum) from suspected cases %99.9 respectively of the type of center reporting health houses, %30, %60 of

health centers - public health and 10% were from public hospital.

**Conclusion:** The results of laboratory sent 10 samples to the laboratory in terms of measles virus in the samples declared negative seems to be over 95% immunization coverage and health authorities on alert timely reporting suspected cases of measles in the prevention of epidemic diseases be effective

**Keywords:** Measles, Epidemiology, Gonabad, Iran

### Leisure and sports services availability for children with Autism spectrum disorder

*Ghanooni, P., MS, Shahid Beheshti University of Medical Sciences, Memari, AH., MD, Kordi, R., MD, Sports Medicine Research Center, Tehran University of Medical Sciences*

**Background:** Individuals with Autism spectrum disorder (ASD) may be experience significant life span disabilities whom need early and specific services at different levels of their needs and life span continuity of services and opportunities for inclusion in the society. Physical activity and Sports as a main aspect of quality of life and socialization process for children depend more on the availability of adapted and proper facilities than on the severity of individual impairment. This study investigated to what extent autistic children access the community based opportunities and facilities for sports and leisure activities.

**Methods:** A total of 101 families of autistic children (6-15 years old) surveyed about familiarity and accessibility of special communities and or clubs of leisure and sports activity which could provide qualified and adapted services for ASD children. Further, they determined to what extent their children involve in regular physical activity.

**Results:** Only 10.9% of families were member of a special community and 7.3% of children have done regular physical activity. 87.9% of families didn't know anything about adapted sports and activities for children with ASD. They reported the factors like time (16%), cost (28%), injury risk (1%) etc, may be contribute to not participation of children in regular leisure and sports activities. **Conclusions:** Findings showed limited opportunities in leisure and physical activities for children with ASD. Providing adapted and qualified facilities besides Educational programs in terms of regular leisure and sports activity promotion in ASD population will be warranted.

**Keywords:** Autism, Sports, Physical activity, Facility

### Autism and associated comorbidities in boys and girls students of Tehran, Iran

*Mirfazeli, F., MS, Memari, AH., MD, Kordi, R., MD, Sports Medicine Research Center, Tehran University of Medical Sciences*

**Background:** Autism is a neurodevelopmental disorder with almost unknown etiologies. The strong association between autism and genetic factors has been long established but not clearly. We aimed to Survey the comorbid conditions of autism as physical and psychiatric pathologies.

**Methods:** The sample consists of students (N = 104) with autism spectrum disorder (ASD) diagnosis. For all subjects, their parents were asked to complete the checklist of general and biomedical information in terms of medical and psychiatric comorbidities.

**Results:** 30.8% (N = 32) of subjects had at least one medical condition, and 27.5% (N = 28) had at least one additional psychiatric condition. Data analysis for sex difference showed that Boys had 28.9% medical and 21.3% psychiatric comorbid conditions and girls had 38.1% and 55% respectively.

**Conclusions:** In the present study, children with ASD exhibited high rates of medical as well as psychiatric problems. Based on these findings, further investigation of the effects of these problems on behavioral and cognitive abilities among students' boys and girls with ASD is warranted.

**Keywords:** autism spectrum disorder, comorbidity, medical, psychiatric problem

### The effect of kangaroo care on crying response to pain in preterm neonates

*Rahiminia, E., Neonatal Intensive Care Nursing Student, Member of the Research Association of the Student, Tabriz University of Medical Sciences*

**Background:** Crying is a common response to pain in infants and is considered to be the most sensitive measure of pain. Crying has many adverse physiologic effects, such as elevated heart rate and blood pressure. Blood sampling from infants by lancing and squeezing the heel is one of the most frequently occurring acutely painful procedures. Premature infants have even more heel sticks. Kangaroo care (kc) may reduce the audible and inaudible crying response. "Kangaroo Care," skin contact between mother and infant, reduces pain and may reduce crying in response to pain. The purpose of this study was to test Kangaroo Care's effect on the preterm infant's audible and inaudible crying response to heel stick.

**Methods:** The present study have been collected with the review of literature and internet resources

**Results:** In a study Fifteen preterm infants given 20 minutes of KC immediately before a venipuncture in incubator had lower beta-endorphin levels than when lying in an incubator for 20minutes before the venipuncture. In another study, 3 hours of KC followed by a heel stick in KC produced lower peak heart rate and shorter duration of crying in response to heel stick pain than incubator care. In another study the effect of 30 minutes of KC before and KC during a heel stick on audible and inaudible crying responses to pain. The preterm infants would have less crying time (audible cry + inaudible cry) with a heel stick done in KC than with a heel stick done in the incubator

**Conclusion:** Kangaroo Care could have an analgesic effect in several ways. First, KC's continuous tactile stimulation may serve as a pain inhibitory system by activating endogenous pain-modulating systems. As little as 20 minutes of KC significantly alters cortisol and beta-endorphin release. Second, KC's analgesic effects may be mediated through oxytocin. During skin-to-skin contact, oxytocin secretion increases in both infants and mothers. Third, KC may indirectly reduce neonatal pain by decreasing the total amount of noxious stimuli to which neonates are exposed. A final mechanism may be the maternal presence, which includes the presence of maternal odor. Maternal odor provides a degree of analgesia

**Keywords:** Kangaroo care, Crying response, Pain, Preterm neonates

### **Finding of brain CT scan in traumatic children that referred to Ayatollah Taleghani Hospital of Kermanshah 2010**

*Salehi-Zahabi, S., Moradi, F., Karimi, H., Asgari, Sh., Godarzi, M., Department of Radiology and Nuclear Medicine, School of Paramedical, Kermanshah University of medical science, Kermanshah*

**Background:** Trauma is the one of most common reason for death in children. In patient with trauma, the skull is the most common involved part. The importance of computed tomography (CT scan) in diagnosis of brain trauma is well established. CT scan is actually a choice method in evaluating patients with cranial trauma. Considering the importance of CT scan in the diagnosis of brain lesions, goal of this study is a survey in the results of brain CT scan in traumatic children that referred to Ayatollah Taleghani Hospital of Kermanshah 2010.

**Methods:** In this cross-sectional and descriptive study brain CT scan results of 496 children with cranial trauma that referred to CT scan department of Taleghani Hospital

of Kermanshah in April to August 2010 was evaluated. For data collection pre-prepared tables containing demographic data, cerebral results, lesion and location of fractures was used. Finally the data were analyzed using descriptive statistics.

**Results:** The results show that 68.54 % of 496 patients were male and had the most patient rate. In this study we evaluated computed tomograms of traumatic patients, 424 (85.48%) cases of 496 patients had normal brain CT scan and 72 (14.52%) cases had positive finding including: 2 cases(2.77%) of brain contusion, 9 cases(12.5%) of epidural hematoma, 6 cases(8.33%) of subdural hematoma, 2 cases(2.77%) of subarachnoid hemorrhage, 2 cases(2.77%) of intracranial hemorrhage, and 51 case (70.86%) of skull fracture.

**Conclusion:** Due to the high frequency of normal CT scan in this study and disadvantages of no indication CT scan especially in children, it is necessary for clinical physicians to pay attention and do accurate efforts because there is some concerns that the CT scan of brain is going to be a routine application.

**Keywords:** Brain CT scan, Children, Trauma, Kermanshah

### **The experience of children with leukemia from Intrathecal procedure in IT room; a qualitative study**

*Hedayat, K., Medical Student, Beheshti Medical University, Hedayat Z., Medical Student, Iran Medical University, Alhani, F., Tarbiat Modares University Ansari, Sh., Gholipour, F., Iran University of Medical Sciences*

**Background:** Acute lymphoblastic leukemia (ALL) is the most common malignant cancer in pediatric. Many hours of their life times spend in hospital under treatment protocol like IT. This quantitative research was done by means The experience of children with leukemia from Intrathecal procedure and their experiences in IT room.

**Methods:** The design chosen for this research project is Grounded theory a qualitative study. The type of sampling method was purposive sampling. Data gathering was done by In-depth interview with fifteen children under treatment of IT and also their parent and another key participation such as Oncologists, supervisor, staff and residents in charge of performing IT. In addition researcher field note, observation of their behaviors during the IT process. Data analysis: Constant comparison analysis continues to data saturation, Member checking and expert review was done too.

**Result:** Data analysis by Glaser method showed the main categorize are: 1- Fear and anxiety of child from hospital's environment, treatment's procedures like IT 2- Inappropriate relationship with child 3- Lake of child

cooperation The core variable of the codes is fear that must reduce with child centered communication.

**Conclusion:** This study shows that psychological problems and the fear which they suffered are added to their pain and weakness and cause to lower quality of life. Therefore healthcare staffs should do their best to reduce these kinds of problems and facility the therapeutics process.

**Keywords:** children, leukemia, IT, Fear& anxiety  
Qualitative study

### **Rated stressors influencing mothers with infants hospitalized in neonatal intensive care medical centers in Yazd 1388-89**

*Javadi, SS., Dehghan, A., Salimi, T.; Shaheed Sadooghi University of Medical Sciences, Yazd*

**Background:** Premature delivery with hospitalization and separation from family is associated. Neonatal intensive care environment can be stressful for parents, noisy, is strange and frightening. Climate that moment of death or life in it is current. In such an space to create an effective attachment with the baby and parents to be vulnerable. According to the study insufficient studies in this regard was carried out in our country.

**Methods:** Descriptive study with 39 mothers with infants hospitalized in two neonatal intensive care medical centers in yazd questionnaire (parent stressor scale, PSS) Funk 1998 Miles and has been done. Data using statistical software spss 16 change were statistically analyzed.

**Result:** The mean age of 25/76 years of maternal 30/8 percent of natural parity 61/5 percent had cesarean. 53/8 percent female infanticide and 46/2 percent were male. Of environmental stressors "are other children under mechanical ventilation in 77 percent "And stressors appearance and behavior of infant and therapy-specific: "to see needles reverberation of embedded body child 100 percent "And stressors associated parents and offered the role of parent: "Fear of touch or hug a child 46/2 percent" in order of most stressors mothers were.

**Conclusion:** The non avoid premature infants being hospitalized, preparing parents, especially mothers for the first exposure, and neonatal intensive care units to communicate with the appropriate infant, special environment, and provide appropriate parental role, as the basis of family-centered services should focus more Be.

**Keywords:** Preterm infant, stressor, neonatal intensive care unit

### **A survey on Posttraumatic Stress disorder among mothers of very low birthweight**

### **infants after discharging from the Neonatal Intensive Care Unit**

*Bakhshi, F., Namjou, Z., Purmovahed, Z., Behmardi, M.*

**Background:** Very low birthweight (VLBW) they require prolonged critical care hospitalization in a neonatal intensive care unit (NICU). Given these circumstances, investigators have speculated that mothers of VLBW infants may be at risk for posttraumatic stress disorder (PTSD). This study improves on existing studies by assessing mothers' current PTSD symptoms, by using a reliable valid indicator to measure how ill the infant was during the NICU hospitalization, and by utilizing a measure of PTSD symptoms designed to assess symptoms related to childbirth.

**Methods:** The study design was descriptive and correlational. In this study 17 mothers were assessed when their infants were more than 2 month old (2-6 month). Maternal perinatal posttraumatic stress symptoms were assessed with the Perinatal PTSD Questionnaire (PPQ).

**Results:** Eighty-three percent (n=14) of mothers scored in the clinical range on a measure of PTSD. How ill the infant was during the NICU hospitalization was related to mothers' PTSD symptoms. Mothers were so dependent to their children and they had lost their desire for doing other works. Many mothers said they are depressed and they could not forget memories of their child hospitalization time.

**Conclusion:** This study shows that most of mothers suffer from PTSD and this influences mother's interaction with their child and reinforces negative feelings in them. If clinicians were able to identify mothers at risk for developing PTSD, intervention might be provided to prevent PTSD, as well as problems for children. Hens, we suggest researchers to do more studies for finding correct ways of screening and treating stricken mothers.

**Keywords:** Mother, Posttraumatic Stress disorder (PTSD), Neonatal Intensive Care Unit (NICU)

### **The effects of two weight loss protocol on resting plasma concentration of IL-6 in overweight and obese health sedentary female of college students**

*Delphan, M., Rashidlamir, A., Delphan, F., Izadpanah, N., Rahbarizadeh, F., Physical Education and Sport Sciences of Ferdowsi University of Mashhad*

**Background:** Interleukin-6 is excreting by adiposities, which increased by overweight and obesity. Overweight and obesity causes a large number of health problems.

Obesity-related inflammatory marker is a promising predictor for diseases. However, the effect of exercise training, and other therapy that can reduce weight, on IL-6 is still unclear. We examined the effects of two weight loss protocol on resting plasma concentration of IL-6 in the overweight and obese health sedentary female of college students.

**Methods:** A total of 30 subjects (age = 21.9±2.4, weight=75.6±5.57, Height=161±4.79, BF% =29.53±3.18, BMI = 29.04±2.22 kg/m<sup>2</sup>) randomly divided into three experimental and one control groups. Experimental groupI: Experienced twelve-day of researchers proposed diet (D), experimental groupII: either Experienced twelve-day and researchers proposed diet along with twelve-day running training (intensity was 60 to 70 %HRmax for 50 minutes) every other day mid consumption 250Cc water beverage during per aerobic exercise period (AEDW), experimental groupIII: Experienced twelve-day and researchers proposed diet along with twelve-day running training (intensity was 60 to 70 %HRmax for 50 minutes) every other day mid consumption 250Cc 12% glucose soluble during per aerobic exercise period (AEDG) and control group (C) remained sedentary with normal diet in this period.

**Results:** After doing the programs, weight in experimental groups was reduced (P<0.0001). Resting plasma concentration of IL-6 was significantly decreased, for both AEDW (p<0.001) and AEDG (p<0.01) groups.

**Conclusions:** AEDW and AEDG, lowered IL-6 plasma concentration at rest. AED is an optimal pace for decreasing of rest plasma IL-6. Furthermore, according to finding of present study, AEDW is better than AEDG for weight loss.

**Keywords:** Obesity, Interleukin-6, Aerobic exercise, inflammatory marker, overweight

### Reliability, validity and feasibility of persian version of PedsQL TM generic core Scales

*Heidary, M., Gheissari, A., Farajzadegan, Z., Salehi, F., Masaali, A., Mazroi, A. ; Isfahan University of Medical Sciences*

**Background:** To evaluate the reliability, validity and feasibility of the Farsi version of the Pediatric Quality of

Life inventory (PedsQLTM 4.0) Generic Core Scales in Iranian healthy students ages 7-15 and chronically ill children ages 2-18.

**Methods:** We followed the translation methodology proposed by developer. Sample of 160 healthy students (chosen by random cluster method between 4 regions of Esfahan education office) and 60 chronically ill children (chosen consequently from patients come to Alzahra hospital specialty clinics) and their parents completed the Farsi version of PedsQLTM4.0 Generic Core Scales.

**Results:** The Farsi version of PedsQLTM4.0 Generic Core Scales discriminated between healthy and chronically ill children (healthy students mean score was 12.3 better than chronically ill children, p<0.001). Cronbach Alpha s internal consistency values exceeded 0.7 for children self reports and proxy reports of children 5-7 years old and 13-18 years old. Reliability of proxy reports for 2-4 years old was much lower than 0.7. Although proxy reports for chronically ill children 8-12 years old was more than 0.7 these reports for healthy children with same age group was slightly lower than 0.7. Constructive, criterion face and content validity was good. The Farsi version of PedsQLTM4.0 Generic Core Scales was feasible easy to complete.

**Conclusion:** Results show the Farsi version of PedsQLTM4.0 Generic Core Scales is suitable for pediatric health researches children over 8 and parents of chronically ill children over 5. It is necessary to alternate scoring for 2-4 years old questioner and to find a way to increase reliability healthy children ages 8-12 especially according to Iranian culture.

**Keywords:** PedsQL; Pediatric; Quality of life