

Cardiology

Oral Presentation

Report of a Child with CHARGE Syndrome and Idiopathic Diffused Splenic Calcification

Elahe Malakan-Rad^{1,2}, MD

1. Department of Pediatrics, Tehran University of Medical Sciences, Tehran, IR Iran
2. Children's Medical Center, Pediatrics Center of Excellence, Tehran, IR Iran

Case Presentation: The patient is a 4-year-old male child who was brought to the pediatric cardiology clinic because of dyspnea. He was the fourth and the last offspring of a nonconsanguineous parents with a low income status. In present history, mother complained of chronic constipation and inability to eat solid food. On physical examination, he looked ill with severe stunting according to WHO assessment of malnutrition. He had clubbing and a grayish hue on the lips that showed a combination of anemia and cyanosis. His face was asymmetric due to unilateral facial palsy with bilateral external ear deformity. He had bilateral congenital deafness with absent expressive language and a short frenulum. He could neither sit nor stand or walk. Eye examination revealed chorioretinal coloboma, ptosis, orbital fissure phimosis, microcornea and microphthalmia of the left eye and normal right eye. No evidence of chorioretinitis was seen. On palpation, the precordium was hyperactive. A grade 4/6 ejection systolic murmur was present at the left sternal border. First and second heart sounds were normal. There was a cutaneous lesion with vitiligo in the buttock surrounding the rectal area. Deep tendon reflexes were normal. Bilateral undescending testes were present.

Complete blood count showed severe hypochromic and microcytic anemia (Hb=3.5gr/dL). Immunologic studies showed normal Tcell and B cell immunity. No microorganism could be isolated from blood, urine, bronchoalveolar lavage fluid, gastric contents and bone marrow aspirates. Serologic study for Echinococcus was negative. There was no skull calcification. Bone marrow aspiration showed erythroid hyperplasia. Serum analysis for antibody to histoplasma capsulatum and culture of blood and bone marrow for histoplasma were negative. TORCH study and PPD were negative. The secretions of the stomach were tested for acid-fast bacilli and was negative. Chromosomal study revealed normal karyotype.

Study for microdeletion of 22q11 could not be done because of low income of family. Skin lesion was diagnosed as vitiligo by dermatologist.

Electrocardiogram showed right axis deviation and right ventricular hypertrophy. Echocardiography revealed tetralogy of Fallot with right aortic arch. Cardiomegaly, congested lung fields and diffused splenic calcification were apparent on chest roentgenogram. Barium meal imaging revealed gastroesophageal reflux. CT scan of ear showed hypoplasia of petrous bone and cochlear hypoplasia. Brain computer tomographic scan showed atrophic changes and accompanying ventricular dilation. Abdominal CT and ultrasonography did not show any cysts and serologic testing for antibody of Echinococcus was negative. After stabilization of the clinical status of the patient, he was scheduled for cardiac catheterization and cardiac surgery at the due appointment time, but he did not return to clinic.

Conclusion: Based on clinical examination, CHARGE syndrome was diagnosed for the patient. This is the first reported child with CHARGE syndrome and idiopathic diffused splenic calcification. Absence of immunodeficiency and diarrhea despite severe and chronic malnutrition also merits attention in this boy.

Key Words: CHARGE syndrome; Heart anomaly Cyanosis; Malnutrition

Poster Presentation

A Study on the Side Effects of Losartan in Pediatric Hypertensive Patients

Mirza Shahid Arshad¹, MD; Shamsi Allami¹, MD

Al-Shafa Hospital; Pakistan

Objective: There is quite some controversy over Losartan caused side effects in Pediatrics amongst medical practitioners. Thus, the aim of our study was to determine the side effects of Losartan amongst Pediatric Essential Hypertensive patients.

Methods and Subjects: We involved 15 Pediatric patients in this study, starting July 2008 to September 2008, at Al-Shafa Hospital, Pakistan, who were diagnosed as Essentially Hypertensive. Ten were males and five females. All were between 10-15 years of age. All patients were having a blood pressure between 140/95 to 150/105. We provided them with Losartan tablets 25mg (provided/donated by a pharmaceutical company in, Tehran, Iran), two times daily, for three weeks and observed them for the Losartan related side effects.

Findings: All patients showed marked clinical improvement/reduction in their hypertension and

hypertensive symptoms. Six patients presented with moderate dry cough side effects related to Losartan in the initial days of treatment which disappeared after 12-15 days of use. Five patients complained of muscle weakness and pain, which was too cleared in a few days. Three patients complained of sore throat which cleared by reduction of dosage.

Conclusion: We find that Losartan tablets administered for Pediatric Essential Hypertension, has a lower incidence of side effects as compared to ACE Inhibitors. We suggest its use in Pediatric Essential Hypertension, but further research is also required.

Key Words: Essential Hypertension; Losartan; Paediatrics; Side effects; Symptoms

Oral Presentation

Management of Supraventricular Tachycardia in Children

Bahram Mohebbi¹, MD; Azar Tol², MS.PH

1. Iran University of Medical Sciences, Tehran, IR Iran
2. Deputy of Health, Tehran University of Medical Sciences, Tehran, IR Iran

Introduction and Epidemiology: Supraventricular tachycardia (SVT) can be defined as an abnormally rapid heart rhythm originating above the ventricles, often (but not always) with a narrow QRS complex; it conventionally excludes atrial flutter and atrial fibrillation. The prevalence of SVT is not well defined, but is estimated to be between one in 250 and 25,000 children. The prevalence is much higher in critically ill children and adults with congenital or acquired heart disease treated in a pediatric cardiac intensive care unit.

Acute management: Acute management of the child who presents in SVT can be a challenge because the exact mechanism of the tachycardia often is unknown. The treatment strategy depends upon the patient's presentation and clinical status (hemodynamically stable or unstable). The approach consists of initiating therapy while continuing to assess the patient's condition. Hemodynamic assessment - The most important initial clinical determination to make in children presenting with a tachyarrhythmia is whether signs and symptoms related to the rapid heart rate are present. These include hypotension, heart failure, and signs of shock, pallor, or decreased level of consciousness. Signs in infants may include irritability, tachypnea, and poor feeding. Hemodynamically unstable- Unstable patients with hemodynamic compromise, such as unconsciousness or shock with severe heart failure, require immediate termination of the tachyarrhythmia. Cardioversion- Direct current cardioversion at 0.5 to 2.0 J/kg should be

performed. A narrow complex tachycardia should be converted in synchronous mode in which a shock is not delivered during the vulnerable repolarization period. Vagal maneuvers - In children who have mild or no symptoms, vagal maneuvers should be attempted while supplies and personnel are assembled to proceed to medical therapy, if needed. These maneuvers should be performed while the ECG is continuously monitored. The electrocardiographic pattern seen during termination of the tachycardia can help determine its mechanism. Antiarrhythmic drugs - If the vagal maneuver does not convert SVT that is hemodynamically stable to normal rhythm, an intravenous catheter should be placed for the administration of antiarrhythmic drugs. The most commonly used pharmacologic agent for acute management of SVT is adenosine; procainamide and amiodarone are sometimes given for tachycardia that is refractory to adenosine. Chronic Therapy- After the acute episode is terminated, a 12 lead ECG should be performed to look for evidence of WPW syndrome. In addition, an echocardiogram should be obtained to assess for structural heart disease, since SVT can be associated with congenital disorders (21 percent of patients referred for catheter ablation in one series. Recommendations- The treatment strategy for SVT depends upon the patient's presentation and clinical status.

Acute treatment: An infant or child who presents in the nursery or to the emergency department in SVT should have a 15 lead ECG performed while a hemodynamic assessment is made. Chronic therapy- After the acute episode is terminated, a 12 lead ECG and echocardiogram should be performed to look for evidence of WPW syndrome and structural heart disease.

Key Words: Supraventricular tachycardia; Children; Management

Poster Presentation

International Statistical Classification of Congenital Malformations of the Circulatory System

Mohtaram Nematollahi, PhD

Health Information Management, Shiraz University of Medical Sciences, IR Iran

The International Statistical Classification of Diseases and Related Health Problems (most commonly known by the abbreviation ICD) provides codes to classify diseases and a wide variety of signs, symptoms, abnormal findings, complaints, social circumstances and external causes of injury or disease. Every health condition can be assigned to a unique category and given a code, up to six characters long. Such categories can include a set of similar diseases. ICD-10 contains

the following codes for Congenital malformations of the circulatory system: Q20 Congenital malformations of cardiac chambers and connections, Q21 Congenital malformations of cardiac septa, Q22 Congenital malformations of pulmonary and tricuspid valves, Q23 Congenital malformations of aortic and mitral valves, Q24 Other congenital malformations of heart, Q25 Congenital malformations of great arteries, Q26 Congenital malformations of great veins, Q27 Congenital malformations of peripheral vascular system, Q28 Other Congenital malformations of circulatory system.

Key Words: Classification of Diseases, ICD-10, Circulatory system

Oral Presentation

Interventional Therapies for Congenital Heart Diseases in Children

Keyhan Sayadpour Zanjani, MD

1. Department of Pediatrics, Tehran University of Medical Sciences, Tehran, IR Iran
2. Division of Cardiology, Children's Medical Center, Pediatric Center of Excellence, Tehran, IR Iran

Traditional treatment for congenital heart diseases (CHD) is surgery. However, transcatheter therapies are now replacing many operations. Balloon

valvoplasties (aortic, pulmonary, mitral) were the first interventional procedures practiced. Patent ductus arteriosus (PDA) was the first CHD treated by device implantation. There are several types of coils (Cook, and pfm) and umbrella devices (Amplatzer duct occluder I and II) which make closure of almost all PDAs feasible. Atrial septal defect (ASD) can also be closed by device. Large ASDs in small children are difficult, but small to medium defects and those in adults are easily treatable without surgery. The list of devices available for ASD closure is even larger. Ventricular septal defects are also amenable to device closure. The only concern is heart block in case of perimembranous defects. Muscular defects and defects in patients over 5 years of age are recommended for Amplatzer device closure. New devices (pfm VSD coil, transcatheter patch) are available with low risk of heart block.

Coarctation can now be treated without surgery for whole life in infants as small as 7 kg of weight. Stents can relieve the stenosis and can be redilated safely for several times (Palmaz-Genesis XD, Covered and bare CP).

Pulmonary branch stenosis, patent foramen ovale, aortopulmonary collaterals, aortopulmonary window, and some more diseases can also be treated by intracardiac devices. In this lecture, CHD amenable to transcatheter therapy will be reviewed briefly and examples of patients treated in Children's Medical Center will be reported.

Key Words: Congenital heart diseases; Patent ductus arteriosus; Atrial septal defect; Coarctation