Cardiology, Respirology and Emergency Medicine Abstracts

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

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Background: Isolated non compaction left ventricle (NCLV) is a rare genetic disorder that involved in enhanced large ventricular trabeculation due to cardiac heart failure, arrhythmia and mural thrombosis. Echocardiography and cardiac MRI is the best diagnostic imaging for it.
Case Presentation: We reported 2 cases (single originated family): a 9 year-old girl and a 16 year old boy reported (family members) that after exercise and excitement were involved sudden cardiac Death. Both patients had a history of seizures and syncope (in the female case) that treated by anticonvulsant drugs. Following the occurrence of two patients' sudden cardiac death in their family, other family members examined by echocardiography and then to be diagnosed as isolated NCLV in their family.
Findings: Seizure and syncope could be as the initial manifestations of NCLV, which is the embryonic origin of the myocardium. Syncope and seizure could be warning signs (risk factor) for these patients who are at risk of sudden death if they have exposed. Echocardiography which was done in patients with seizure attack, or syncope in the absence of underlying neurological disease, and especially in familial case of NCLV may be beneficial. Arrhythmias, SCD and thromboembolic events compared with cardiac heart failure were happen rarely, but the prognosis of medical treatment by anti-convulsion drugs extremely is poor in these patients after diagnosis was made.
Keywords: Cardiomyopathy, Left Ventricle Non Compaction, Failure, Death

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

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Background: Today progression in cardiac surgery and anesthetic technique causes a good chance for small children who are undergone surgery due to coarctation of aorta (COA). On the other hand with increasing diagnosed cases of COA and concomitant intera cardiac lesion, then surgical options must be noted. Based of this fact, we tried to decrease complication and cost by shortening of duration of hospitalization. In this study we compared hospitalization time for different manners including L.thoracotomy and Midsternotomy in coarctation only or coexisted with Ventricular Septal Defect (VSD) or another intracardiac anomaly. When coarctation coexists with a VSD in a neonate or infant with heart failure or VSD of membranous, outlet, or malalignment type one-stage repair of the coarctation (by end-to end anastomosis) and the VSD through a mid-sternotomy is the procedure of choice for such situations. Two stages coarctation repair alone may be performed, with later VSD closure if it remains large or the infant has failure to thrive. Coarctation repair with concomitant banding of the pulmonary trunk can be performed, with later removal of the band and VSD closure.
Methods: Our study was a retrospective- descriptive study. Duration of study was from 2009 to 2012 at Children's Medical Center. Number of patients were 22 (L. Thoracotomy=8 cases, sternotomy=14 cases). Results were analyzed by using statistical software spss, 15.5 (assurance=95%). T-test descriptive (freq), Pearson (correlation age & Wt by admission time).
Findings: The minimum and maximum age of patients was 6-574 days. The minimum and maximum weights were 2.5-8.5 kg. Average stay in groups L. Thoracotomy and sternotomy were 33.62 and 32.42 days, respectively. According to the Pearson correlation ($P<0.041$) the relation between patient age or weight and duration of hospitalization was inversed and whatever the age and weight of patients increased, discharge time was decreased.
Conclusion: We advise prospective study and more cases for getting better results and comparison complications of coarctation surgery between Lat sternotomy and Thoracotomy.
Keywords: Hospitalization, Coarctation of Aorta Repair, Lat Thoracotomy, Mid Sternotomy

The bronchopulmonary dysplasia in infants with retinopathy of prematurity

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Background: For the last decade the bronchopulmonary dysplasia (BPD) became a growing problem in pediatrics and has taken a second place on frequency among chronic respiratory diseases after childhood asthma. In infants of early age the BPD is determineed at 15-38% of cases, with the birth weight less than 1500g who need mechanical ventilation because of respiratory distress syndrome of newborn. With the development of nursing and respiratory therapy technologies in preterm infants the increase of BPD frequency has occurred with the reducing of mortality among children with birth weight less than 1000g and gestational age less than 30 weeks. The toxic effect of high concentrations of inspired oxygen and hyperoxia causes the damage not only for the lung tissue, but also non-infectious nonimmune inflammation with subsequent proliferation of endothelial and retinal neovascularization- retinopathy of prematurity (ROP). The objectives of the study were determining the frequency of bronchopulmonary dysplasia in infants with retinopathy of prematurity.
Methods: 263 preterm infants with retinopathy of prematurity were studied by chest radiography, and binocular indirect ophthalmoscopy.
Findings: Out of 263 children with ROP, 71.5% (188) were preterm infants with BPD, gestational age 27- 32

weeks and birth weights from 720g to 2150g. The most significant factor affecting to the development of ROP, was a long oxygen dependence, which was typical for children with BPD. The duration of oxygen therapy ranged from 1 to 3 months of life, to an average of 30-38 weeks corrected age. Founded that of children with BPD of 1-2 stages, 48.4% (91) diagnosed with ROP with minimum vascular activity - ROP I-II, at zone 2, which ended with spontaneous regression. In infants with BPD 3-4 degrees the most severe forms of ROP - ROP II-III was revealed in 1-2 zone in 12.5% of cases, "+" disease in 4.8% and backagressive form of ROP in 2.7%. 3-4 degree BPD was clinically manifested with respiratory failure, obstructive syndrome, episodes of desaturation and oxygendependence. Threshold ROP was needed the conduction of laser photocoagulation of the retina.
Conclusion: Thus, BPD has unfavorable effects on the immature retina of a premature baby. Without affecting the incidence of ROP, the BPD is a risk factor for its progression. Recommended to maintain the oxygen saturation SaO2 within 88-92%, not to avoid the sharp fluctuations in the oxygen level to prevent episodes of hyperoxia and hypoxia. The gradual elimination of oxygen is important for the prevention of proliferative ROP.
Keywords: Bronchopulmonary Dysplasia, Retinopathy System, Infants

The effects of one course of endurance training on functional and structural indices of the heart in children

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Background: The strenuous and perseverant exercises have profound effects on the functional and structural status of the heart in children. The goal of this study was to determine the effects of the above mentioned exercices on some cardiac indices such as stroke volume(SV), cardiac output(CO), left ventricle ejection fraction(LVEF), left ventricle end-diastolic diameter(LVEDd), left ventricle end-diastolic volume(LVEDV), left ventricle mass(LV mass), and left ventricle posterior wall diameter(LVPWd).
Methods: The study was done on 20 children(10 patients as the case and 10 ones as the control groups), having the average age, height and weight of 10.8+/- 6yr, 149 cm and 36+/- 8 Kg, respectively. The case group had a perseverant running three times a week for 12 weeks, reaching 50-70% of their maximal heart rate. In the both groups, all the above mentioned indices were measured before the study, by echocardiography. Again, all the parameters were measured 12 weeks after the perseverant running.
Conclusion: The results of the study showed no significant difference between the case and control (without exercises) groups who had increased their SV index by 5.3 and 1.37%, respectively. The CO index with an increase by 7.29 and a decrease by 6.9 in the case and control groups, respectively, also showed no significant difference. Also LVEF displayed no significant difference in none of the groups with(the case) and without(the control) exercises. LVPWd showed an increase by 2.14 & 2.17 in the case and control groups, respectively, which indicated no significant difference. Neither LVEDd nor LV mass didn't show any significant difference in the control group, with a decrease by 2.26 and 0.31, respectively. Meanwhile, the case group demonstrated a significant difference in the latter indices, with an increase by 6.78(P value<0.023) & 12.64(0.003), respectively. Finally LVEDV showed a significant difference with an increase by 24.7(P value<0.002), in the case and no significant difference with an increase by 2.29 in the control groups, respectively.
Keywords: Children, Aerobic Exercises, Structural Changes, Functional Changes, Cardiovascular System

Acquired pericardial cyst with degenerative changes rare entityin Iranian Child: a case report

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Background: Pericardial cysts constitute two forms congenital and acquired. Second form is rare with unusual clinical presentations. Also keep minding, the second form had rare complications, such as spontaneous rupture and hemoptysis. Our article is based on the record of a boy with acquired pericardial cyst with degenerative changes and review of literatures.
Case presentation: A 9 year old boy presented with intermittent, non-exertional, retrosternal chest pain since13 months ago. The pain radiated to the left arm. He denied constitutional symptoms, hemoptysis, chronic cough and sputum production. He also underwent surgery for lung hydatid cyst 1.5 years previously. On physical examination, the cardiopulmonary system was unremarkable. Chest X-ray revealed a rounded calcified cystic mass along the left cardiac border. A intrapericardial cyst was seen at thoracotomy. Histologic examination of excised lesion shown cystic wall including dense fibrous connectivetissues with calcification, focal ossification, aggregation of amorphous material containing cholesterol crystal and chronic inflammatory cells infiltration. Mesothelial cells lining was not found. He had uneventful postoperative period and was symptoms free when seen six months later in the clinic.
Conclusion: Acquired pericardial cyst has exceptional features in location of the cyst, macroscopic and microscopic pathology, and clinical presentations. Some of patients have past medical history of tuberculosis, hydatid cystand other infectious diseases. Therefore, more investigations including serology for hydatid disease, search for specific granuloma, acid fast bacilli and … are highly mandatory.
Keywords: Acquired Pericardial Cyst, Children

Guidelines for sinusbradycardia in newborns

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

bradycardia include sinus bradycardia, abnormalities of sinus node function and abnormalities of atrioventricular conduction. In the instances where sinus bradycardia is pathologic, it usually results from non-cardiac disease. Sinus node dysfunction is rare early in life but can arise from surgical interventions, congenital heart disease, or endovascular manipulations. Abnormalities of atrioventricular conduction have a similar etiology but are more common than sinus node disease. Atrioventricular nodal disease can also result from maternal collagen vascular disease, even in the absence of symptoms in the mother. In these cases, epidemiological issues such as heart block in subsequent pregnancies and the maternal risk of developing symptomatic collagen vascular disease become important. The approach to treatment and long-term prognosis for bradycardia in the neonate is highly dependent on the underlying etiology and on the presence of concurrent factors such as structural heart disease.
Keywords: Sinusbradycardia, Neonatal

Noninvasive evaluation of myocardial systolic dysfunction in the early stage of Kawasaki disease: a speckle-tracking echocardiography study

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Background: Evaluation of myocardial function by speckle-tracking echocardiography is a new method for the early diagnosis of systolic dysfunction. We aimed to determine myocardial speckle-tracking echocardiography indices in Kawasaki disease (KD) patients and compare them with the same indices in control subjects.
Methods: Thirty-two patients (65.5% male) with KD and 19 control subjects with normal echocardiography participated in this study. After their demographic characteristics and clinical findings were recorded, all the participants underwent transthoracic echocardiography. Strain (S), Strain rate (SR), time to peak strain (TPS), and Strain rate (TPSR), Longitudinal velocity and view point velocity images in the two-, three-, and four-chamber views were semi-automatically obtained via speckle-tracking echocardiography.
Findings: Among the patients, 24(75%) were younger than 4 years old. Mean global S and SR was significantly reduced in the KD patients compared to controls (17.03±1.28 vs. 20.22±2.14% and 1.66±0.16 vs. 1.97±0.25 1/s, respectively), while there were no significant differences regarding mean TPS, TPSR, Longitudinal velocity and view point velocity. Using repeated measure of analysis of variances, we observed that S and SR decreased from base to apical level in both groups. The change in the pattern of age adjusted mean S and SR across levels was significantly different between the groups (P<0.001 for both parameters).
Conclusion: We showed changes in S and SR in KD patients versus control subjects in the acute phase of KD. However, we suggest that further studies be undertaken to compare S and SR in the acute phase and thereafter in KD patients.
Keywords: Kawasaki Disease, Left Ventricular Function, Myocardial Velocity, Strain Rate Imaging

Assessment of neonatal sepsis on myocardial function by tissue Doppler imaging

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Background: Neonatal period is the most critical stage in the life of newborn babies and neonatal sepsis is one of the most common causes for mortality at this age. Cardiovascular complications, myocyte damage and modification of cardiac blood flow induced by inflammatory mediators are among the consequences of neonatal sepsis on newborns. Nevertheless only a limited number of echocardiographic studies have been performed to investigate the pathologic effects of neonatal sepsis on heart. Tissue doppler echocardiography is a useful method for the assessment of regional heart systolic and diastolic functions. In this study, we aimed to determine the myocardial performance during neonatal sepsis by tissue echocardiography in term and preterm newborns.
Methods: This study was a descriptive research and the information was collected through questionnaires and sampling on 61 preterm and term neonates. Tissue echocardiography (TDE) and doppler echocardiography were performed. TDE was done at the level of mitral and tricuspid valve annulus and intraventrivular septum level. The results were then statistically analyzed and evaluated.
Findings: In this study, 30 neonates were term and the rest were preterm with the age of 1 to 9 days.We have not observed significant difference between the tissue echocardiographic indices of mitral and tricuspid valves and interventricular septum between healthy and septic newborns. Our results, however, revealed significant difference in A&E waves at the level of tricuspid and mitral valves between sepsis and none sepsis group(p<0.001).
Conclusion: Doppler echocardiography is a useful tool to evaluate the myocardial function during neonatal sepsis. It is also possible to investigate the myocardial modifications during sepsis by Tissue Doppler Imaging (TDI).
Keywords: Sepsis, Neonate, Myocardial Function, Tissue, Doppler Echocardiography

Primary lung tumors in children

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Background: Primary lung tumors are rare in children; usually, single case reports appear in literature. The collective review cases are also limited, and there is much diversity in reported cases.
Methods: To review the literature on reports of primary lung tumors in children and adolescents, and to present personal experiences of cases with pediatric primary tumors.
Findings: Among the reported pediatric primary lung tumors, some are similar to adult lung pathology, while others are quite different and unique to the pediatric lung. The tumors are mostly endobronchial and the commonly reported cases are carcinoid and mucoepidermoid carcinoma. The pseudoneoplastic tumors, which are considered the most common benign primary lung tumors in children, are controversial in their nature.

Conclusion: Primary lung tumors in children are rare and histopathologically diverse. They mostly present as endobronchial and there is usually a delay in their diagnosis. For children with persistent respiratory symptoms and chronic wheezing who are not responding to conventional treatment, the possibility of an endobronchial lesion should also be considered in their differential diagnosis. The author's personal cases highlight the issue.
Keywords: Children, Lung Tumor, Endobronchial Tumor

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

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

Cardiovascular complications in a burned child; a case report

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

Congenital chylothorax treated with octreotide

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

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

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Background: Acute exogenous lipoid pneumonia is a consequence of exposure to significant quantities of mineral oils. Accidental poisoning is the most common cause of the acute form in children. Chest x-ray findings include consolidation, nodular lesions, reticular and alveolar-interstitial patterns. Lower lobes or right middle

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