Serum BNP level in screening of hemodynamically ductus arteriosus in preterm neonates with echocardiography

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Background: B-type natriuretic peptide (BNP) is secreted by heart ventricles when there is a hemodynamically stressful state and or congestive heart failure. The aim of this study was evaluation of serum BNP level as a reliable predictor and screening tool for detection of hemodynamically significant patent ductus arteriosus in preterm neonates.

Methods: Sixty preterm neonates with gestational age ≤34 weeks and birth weight ≤2500 grams, who admitted in NICU for respiratory distress and prematurity, enrolled in this study. After clinical assessment by a neonatalogist, the neonates underwent echocardiography and within 24 hours after echocardiography, blood sample for BNP was taken.

Findings: Sixty neonates with 30.98 ± 1.909 (Mean ± SD) weeks of gestational age and 1682.50 ± 350.480 (Mean ± SD) grams of weight were enrolled. BNP levels in neonates with hsPDA (n=13) were significantly higher than those with nonhsPDA (n=17) and those of the control group (n=30). (1667.061 ± 820.977 pg/ml vs. 667.1176 ± 666.369 pg/ml and 309.113 ± 170.957 pg/ml respectively, p<0.005).

At a cutoff point value of 450 pg/ml serum BNP level had a sensitivity of %92.31, specificity of %86.67, negative predictive value of %98.5, positive likelihood ratio of 6.92 and negative likelihood ratio of 0.089 for detection of hsPDA.

Conclusion: Elevation of Serum level of BNP detects hsPDA in premature neonates at cutoff point value of 450 pg/ml. BNP level is a reliable predictor of hsPDA and good screening tool for hsPDA detection.

Keywords: Brain natriuretic peptide, congenital heart disease, Patent ductous arteriosus, neonate

Left ventricular function in pediatric patients after renal transplant

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Background: Despite good outcomes of renal transplantation (RT), cardiovascular disease is the major cause of death in adults. These abnormalities become more important because of improved life span of children after successful RT. Few data are available regarding the effect of RT on left ventricular(LV) function or LV mass regression in children. The aim of this study was to investigate the effect of RT on cardiac structure and function assessed by echocardiography.

Methods: Twenty patients(mean age: 12±3.1 years, 45% males) with RT for a median time of 2.1 years(range 0.9-3.5) were compared to 30 age-matched healthy controls. Standard echocardiography was performed and myocardial performance index(MPI), as noninvasive index for assessing global LV function, was calculated.

Findings: The mean value of MPI of the patients was significantly different from those of the control subjects(0.47±0.09 vs. 0.37 ± 0.02, p<0.001). The LV mass index(LVMI) in patients was positively associated with duration of dialysis( r=0.53, P=0.04) and systolic blood pressure index(r=0.61, p<0.05). LVH was found in 3(15%) patients. Tissue Doppler values were different, showing an increased E/E' ratio (10.2±1.6 vs.7.8± 1.2, p<0.05); lower Em and Sm velocities(10.8± 2.8 vs. 13± 2.1 (p<0.001); 6.8±1.9 vs. 7.2 ±1 (p<0.001), respectively) indicating diastolic dysfunction in patients. However, no correlation was found with LVMI.

Conclusion: Subtle abnormalities in combined systolic and diastolic LV function are present in renal transplanted patients. These findings were confirmed the importance of early detection and initiation of interventions to modify the natural progression of cardiac dysfunction in children with chronic kidney disease and also in renal transplant recipients.

Keywords: kidney transplantation, cardiac function, children

The effect of surfactant on cardiac function in 1000-2500g infants with respiratory distress syndrome

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Background: Respiratory distress syndrome is one of the common causes of mortality and morbidity in preterm infants. Surfactant is widely used in the treatment of respiratory distress syndrome. It seems that surfactant has some effects on cardiac function; therefore, in the present study this effect has been investigated.

Methods: This semi-experimental study was performed on 35 preterm infants with respiratory distress syndrome. First, systolic and diastolic function of left ventricle was determined by echocardiography. Then, 4ml/kg surfactant(Survanta) was injected with INSURE method. After the injection of surfactant, echocardiography with the same method was repeated.

Findings: Surfactant administration significantly increased mean Ejection Fraction(EF) that shows systolic function of left ventricle(p<0.001). Moreover, Mitral valve E/A ratio improved significantly(p<0.001). Furthermore, arterial oxygen saturation and decreasing in heart rate showed significant improvement (p<0.001). The other important criterion was mean Tie index after surfactant administration that showed simultaneous improvement of systolic and diastolic functions of left ventricle(p<0.001).

Conclusion: The present study showed that surfactant administration in preterm infants with respiratory distress syndrome can improve systolic and diastolic functions of left ventricle.

Keywords: prematurity, ejection fraction, surfactant, Tie index
Arterial Tortuosity Syndrome: a case report

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**Background:** Arterial tortuosity syndrome (ATS; OMIM 208050) is a rare autosomal recessive condition characterized by dysmorphic features, elongation, tortuosity, and aneurysm of the large and middle sized arteries.

**Case Presentation:** We report on a 3-month-old girl who presented with diaphragmatic hernia and respiratory distress. Based on angiogram, a diagnosis of ATS was made and subsequently confirmed by a homozygous one base-pair deletion at position g.318 of SLCA10. We stress similarities (facial appearance, inguinal herniae, ...) between ATS and autosomal recessive cutis laxa, both being connective tissue disorders disorganizing the elastin network.

**Keywords:** Arterial tortuosity syndrome, SLC2A 10

Prevalence, prognosis and management of diaphragmatic paralysis after congenital heart surgery in children

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**Background:** Diaphragmatic paralysis (DP) due to phrenic nerve injury is one of the complications of congenital heart surgery and may cause life-threatening respiratory distress and infections and related complications in infants and young children. We designed a study to determine the prevalence of diaphragmatic paralysis, clinical features and treatment outcomes in children who underwent congenital heart surgery in our center.

**Methods:** All patients who underwent congenital heart surgery in our center included in study. Only the patients died at surgery room or early after surgery were excluded from study. If 48 hours after congenital heart surgery weaning the patient from mechanical ventilation was not possible or there was respiratory distress after separation and need for reintubation, a chest x-ray was requested. If hemidiaphragm was elevated on chest x-ray, fluoroscopy was performed to confirm phrenic nerve paralysis.

**Findings:** Prevalence of DP was 5.5%. There was not any statistical difference between patients with DP and without DP in terms of age, gender and weight. But cyanotic congenital heart diseases, history of previous cardiothoracic surgery, mean duration of hospitalization and mortality rate were significantly higher in patients with DP. In 13 patients (52%), diaphragmatic plication was performed. The highest number of plication was performed following the arterial switch operation, VSD closure, and correction of tetralogy of Fallot. Plicated patients had lower age and weight than the group that received conservative treatments. Although considerable difference was observed between the average length of hospital stay in the plicated patients and the patients who received conservative treatment, this difference was not statistically significant.

**Conclusion:** In recent years, the most common cause of diaphragm paralysis in children is related to phrenic nerve injury during surgery. In different studies, the prevalence of DP after congenital cardiac surgery ranges from 0.3 to 12.8%. The prevalence of DP in this study is comparable with other series. In this study, phrenic nerve palsy was more common after arterial switch operations, Fontan and B-T shunt. Maybe low body weight was a risk factor in arterial switch operations that generally performed in neonates. The patients who undergo Fontan and B-T shunt operations usually have history of previous cardiac surgeries, which increase the chance of DP due to adhesions. As a conclusion, postoperative diaphragmatic paralysis after cardiac surgery is a serious complication and the first step in its diagnosis is to suspect.

**Keywords:** phrenic nerve, diaphragmatic paralysis, diaphragmatic plecation, infant, mortality

Assessment of myocardial systolic function by tissue doppler echocardiography in full term infants

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**Background:** Tissue doppler echocardiography (TDI) is a new technique echocardiography that assesses both systolic and diastolic ventricular function. Information about TDI is very low and normal values of TDI in neonates has not yet published.

**Methods:** In this study 66 full term healthy infants were assessed by tissue doppler echocardiography for ventricular function. Velocities of E’, A’, S waves were assessed at level of the mitral and tricuspid valves and interventricular septum.

**Findings:** Among 66 neonates, velicity was generally higher among boys than girls (*p*<0.01). Peak mitral and tricuspid flow velocities during diastole (E’) were 6.21+-2.03, and 6.87+-2.83 cm/s and A’ wave velocities: 7.2+-2.04 and 8.23+-3.20 cm/s and S wave velocities: 5.95+-1.34 and 6.85+-1.44 cm/s.

**Conclusion:** Tissue doppler echocardiography is a useful tool for assessing ventricular systolic function in neonates. In this study normal range of waves velocities of ventricular systolic function were assessed that can be used in diagnosis of neonatal heart diseases.

**Keywords:** full term infant, ventricular systolic function, tissue doppler echocardiography

First report of Persistent fifth aortic arch and severe aortic coarctation in association with Goldenhar Syndrome

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**Background:** Goldenhar syndrome or oculo-auriculo-vertebral dysplasia is present in 1/3500 to 1/25000 live births. The most recognizable feature of this syndrome is hemifacial microsomia. It may be associated with several other craniofacial abnormalities and congenital heart diseases (CHD). Ventricular septal defect and tetralogy of Fallot have been reported as the most common CHD in these patients. However, to date, there is no report of persistent fifth aortic arch and severe coarctation in these patients. This is the first report of a 7-year-old boy with Goldenhar syndrome and persistent fifth aortic arch associated with aortic arch tortuosity and severe coarctation in the persistent fifth arch.
Case presentation: The patient was a 7-year-old boy, born to a consanguineous parent as a twin pregnancy, after 7 years of infertility. On general appearance, hemifacial microsomia, bilateral dermolioma of the eyes, deformity of both ears associated with skin tags were apparent. On cardiac examination, pulses were normal in the upper extremities, but were very weak in the lower extremities. A grade 3/6 ejection systolic murmur was heard at upper left sternal border with wide radiation to the back. Renal bruit was heard on the back. On CXR, the left upper border of the cardiac silhouette was almost absent due to the aortic arch abnormality. Aortic angiography showed common origin of all four brachiocephalic branches from the main aorta, and a torotous, hypoplastic persistent fifth aortic arch with severe coarctation. The patient was referred to pediatric cardiac surgeon for aortic repairment.

Conclusion: This is the first report of persistent fifth aortic arch and severe coarctation in a 7-year-old male patient with Goldenhar syndrome. Persistent fifth aortic arch is a rare abnormality of aortic arch with an unknown incidence. Persistent fifth aortic arch may be asymptomatic or may present itself as coarctation.

Keywords: Goldenhar syndrome, persistent fifth aortic arch, coarctation

Williams Syndrome
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Case Presentation: Proband is a 4-year-old girl from Marand city who was referred to our genetic center due to mental retardation, and developmental delay. She was the first only child of consanguineous parents (3rd familial marriage relationship). Her karyotype was normal. He had mental retardation, growth developmental delay, prominent lips with open mouth, normal calcium of serum, echo finding (pulmonary stenosis, ASD), blond and curly hair, blue eyes, periorbital fullness of subcutaneous tissues. According to clinical findings and normal karyotype, Williams Syndrome was considered. FISH study was done by professor Karymjinajad, and Williams Syndrome diagnosis was confirmed.

Conclusion: Although most individuals with this disorder represent sporadic cases, parent to child transmission has been documented. Studies using Fluorescent In Situ Hybridization indicate that both inherited and sporadic cases of Williams Syndrome are caused by a deletion at 7q11.23, a region that includes approximately 17 genes. Homozygosity for the elastin gene is responsible for supravalvular aortic stenosis as well as other vascular stenosis and LIM-KINASE 1 Homozygosity is a contributing factor to impaired visuospatial construction cognition in this disorder. Genetic counseling and prenatal diagnosis (C.V.S or Amniocentesis) advise for Future pregnancies.

Keywords: Williams syndrome, FISH technique, genetic counseling

A case with ARDS due to immune deficiency and CMV pneumonia
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A 6 months old boy without previous problem was admitted in pediatric ward due to fever and tachypnea. Reticular pattern was found in the CXR. During two days, respiratory distress and tachypnea increased so he was admitted in PICU. Progressive hypoxia and respiratory distress was prominent. In CXR, diffuse alveolar infiltration was seen. Non invasive positive pressure ventilation was started and hypoxia was diminished for 8 hours, but after that due to decrementing of PO2, tracheal intubation was done and mechanical ventilation was started. Hypoxia diminished by a high PEEP(15cm H2O) and low tidal volume(6 ml/kg) and high Fio2(95%). Because of positive CMV PCR (7000 CMV DNA Copies/mL) in respiratory discharges, Gancyclovir was started. Due to low IgG, Ig M and IgA levels, intravenous immunoglobulin was used. After 3 days, PEEP decreased slowly and after 8 days of admission, the patient was extubated. Feeding via NG tube was started. He again became febrile and in his blood culture and urine culture Candida nonalibicans was found so amphotericin started. The day after, he had tachypnea and metabolic acidosis, so amphotericin was changed to fluconazole. After 3 weeks due to negative CMV PCR, gancyclovir was discontinued. The patient discharged from PICU after 30 days of admission.

Keywords: CMV pneumonia, ARDS, CMV PCR, high PEEP

Congenital chylothorax: presentation a case
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Background: When lymphatic fluid accumulates within the pleural cavity, a rare condition in newborns occurs that called congenital chylothorax. Here is a presentation of a birth traumatic case (a newborn male with 3100g birth weight) with bilateral (right prominent) pleural effusion successfully treated by Ocreotide.

Case presentation: This newborn male, weighing 3100 g, was delivered at 38 weeks of gestation by a 33-year-old mother through normal vaginal delivery. There was acrocyanosis and subcostal and intercostal retraction and he used acceleratory muscles for breathing that showed respiratory distress. So the infant was ventilated with ambo bag (O2 sat was about 75-80%) and admitted to the neonatal intensive care unit (NICU) of Baqiyatallah Hospital, Tehran, Iran. An early chest x-ray showed bilateral (right more than left) pleural effusion with left-sided heart shift. Insertion of Intercostal tubes (ICD) was performed and about 60cc of milky pleural fluid was drained. Biochemistry analysis of pleural fluid showed following characteristics: glucose: 35 mg/dl, protein: 1.1 gr/dl, cholesterol: 50 mg/dl and triglycerides: 300 mg/dl. Counts of RBC and WBC were 10000 and 2500 per Cubic mm respectively. After this, congenital chylothorax was diagnosed and enteral feedings with a medium-chain triglyceride(MCT) was started. Then instead of reinsertion of ICD, we decided to start Octreotide infusion. Finally chest x-ray and ultrasound did not show any pleural effusion accumulation in the both sides, and the patient was discharged. Follow up CT showed no pleural effusion.

Conclusion: In the postnatal period, supportive methods like ventilatory care, immediate drainage of effusion, total parenteral nutrition(TPN) and Ocreotide must be considered as first line treatments; but when they could not help, finally surgical method can be useful.

Keywords: congenital chylothorax, newborn, pleural effusion
A rare case of organophosphate poisoning by accidental injection of toxin

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Background: Organophosphate poisoning is common in our country. It can cause accidental poisoning or can be used for committing suicide. The exposure might be dermal, gastrointestinal and/or inhalational. Poisoning in children usually happens due to the negligence of parents. The case which is introduced here suffered from organophosphate poisoning due to accidental injection of poison into the buttocks area.

Case Presentation: The patient was a 5-year old boy that the organophosphate poison was injected to his buttocks accidentally. To kill the insects, his mother filled a syringe with the poison, sprayed on them and left the container and syringe at the corner of the room. The kid accidentally sat on the container and the content of the syringe was injected to his buttock muscle. He was reported with loss of consciousness, miosis, respiratory distress, and severe tearing. He was then transferred from Malayer District to a health care center located in Hamadan. After being treated by atropine and pralidoxime, he got healed in a few days. The abscess in the site of injection healed later.

Conclusion: Organophosphate poisoning could take place by inhalational, gastrointestinal and/or dermal exposure. Poisoning by injection is very rare. Moreover, this specific case emphesises on training the parents about how to look after their children. We also considered the possibility of a child abuse case which the investigations did not confirmed.

Keywords: Organophosphate poisoning, injection, child

Accidental poisonings of methadone syrup in children admitted at the emergency department of Children's Hospital of Khorramabad

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Background: Methadone is a synthetic narcotic analgesic prescribed for the relief of moderate-to-severe pain. It is also used in the detoxification treatment of opiate dependence, and for maintenance in heroin and narcotic addiction. If methadone be consumed by children accidentally, shortly, the toxic symptoms emerge, including: nausea, vomiting, chest pain and other symptoms of poisoning. Accidental methadone poisoning in children is often fatal.

Methods: We retrospectively studied the records of all children admitted by accidental methadone poisoning at the emergency department of Children's Hospital of Khorramabad from 2009 till 2012. Variables studied included age, sex, symptoms, treatment, and final status of the patient during the hospitalization period. The data collected were analyzed by Spss.

Results: Of the 90 patients who had an overdose of methadone syrup, 53% was male and 47% female. Of age groups, 47% under one year of age, 41% between one and five years, 12% over five years. The cause of toxicity, in majority of cases, methadone syrup stored in containers such as mineral water, beer bottles and containers of medicine (acetaminophen, diphenhydramine) and was mistakenly given to the child. Lethargy in 100%, Pin point pupils in 63%, 12% loss of consciousness, respiratory disorder 15%, 67% weak pulse, 76% had vomiting 100% were urban. 53% stay for a day 29%/4% in two days, and 17%/6%/three days. 12% of patients admitted to ICU. 30% were discharged with physician orders.

Conclusions: In this study, most cases of poisoning in children who take methadone accidentally. Key words: methadone, poisoning, emergency department

Mortality risk prediction by application of PRISM scoring system in pediatric intensive care unit

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The Pediatric Risk of Mortality (PRISM) score is one of the scores used in the pediatric intensive care units (PICU) for predication of the mortality risk by many pediatricians. Herein, we intended to evaluate the efficacy of PRISM score in the prediction of mortality rate in a PICU.

Methods: In this cohort study, 221 children admitted during an 18 month period to PICU were enrolled. PRISM score and mortality risk were calculated. Follow up was noted as death or discharge. Results were analyzed by Kaplan-Meier curve, ROC curve, Log Rank (Mantel-Cox), Logistic regression model using SPSS version 15.

Findings: Totally, 57% were males. Forty seven patients died during the study period. The PRISM score was 0-10 in 71% patients, 11-20 in 20.4% and 21-30 in 8.6%. Increase in PRISM score showed an increase of mortality from 10.2% (in 0-10 score patients) to 73.8% (21-30 score patients). The survival time significantly decreased as PRISM score increased (P=0.0001). A 7.2 fold mortality risk was present in patients with score 21-30 compared with score 0-10. ROC curve analysis for mortality according to PRISM score showed an under curve area of 80.3%. Conclusion: PRISM score is a good predictor for evaluation of mortality risk in PICU.

Keywords: PRISM score, mortality, intensive care unit, children

Transport of critically ill patients

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Moving a critical patient is a dangerous medical intervention; there are risks and benefits. The benefits still revolve around providing specialized treatments and diagnostics not available at every facility. Recent literature has shown that time until definitive treatment is an important consideration. Critically ill patients are at increased risk of morbidity and mortality during transport. Risk can be minimized and outcomes improved with careful planning, the use of appropriately qualified personnel and selection and availability of appropriate equipment. Furthermore, the accompanying personnel and equipment are selected by training to provide for any
زمان بستهٔ کودکان تحت عمل جراحی با استفاده از سیستم‌های نهایی‌محاسبه‌ای برای ارزیابی ناهنجاری‌های قلبی از انجام‌پذیری و باعث توصیه می‌شود.

روش: این تحقیق توصیفی شامل داده‌های از 8 نمونه از بیماران می‌باشد که در بیمارستان مرکزی علمی تهران تحت عمل جراحی جنگل کودکان با تعرض به گیری و مهار دیابتی از آنها به دست آمد.

نتایج: در این بیماران، نتایج مربوط به مقدار میزان دما در عضله کبدی، ساعتی، و ویتامین اکسید وکسید نشان می‌داد که در بیمارانی که در دووزن‌های مختلفی بوده‌اند، نتایج کمتری داشته‌اند.

SPSS 15 سنتی در سطح 95% اطمینان نشان داده بود که در بیمارانی که در دو وزن‌های مختلفی بوده‌اند، نتایج کمتری داشته‌اند.

کلمات کلیدی: بیماری‌های قلبی، جراحی قلبی، کودکان، نوزادان.
ترمیم کورکاتاسپسون اوتورت در نوزادان و شیرخواران زیر ۲ ماه

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مرکز طبی کودکان

زمینه: شایع‌ترین آنومالی مادرزادی قلبی که در هفته دوم بعد از تولد برای می‌کند کورکاتاسپسون اوتورت می‌باشد. این بیماران در صورت وجود علامت‌های ابتدایی، تحت دنیا جراحی قرار گیرند. در بیماران بدون علامت‌هایی می‌توان عمل جراحی را به بعد از سه ماه‌های موقت نمود. در کشور ما بیشتر نتایج ضعیفی از این مکاتب عمل جراحی است. این بیماران به بعد از سه ماه‌های موقتی می‌شود.

روش: این مطالعه بر روی بیماران انجام شد. از سال ۱۳۸۷ تا ۱۳۹۲، ۲۲ بیمار زیر ۲ ماه مبتلا به کورکاتاسپسون اوتورت و ۲۸ بیمار زیر ۲ ماه بدون نشانه‌های لازم از کورکاتاسپسون، به بیمارستان تهران آمده بودند. هر دو گروه به صورت تصادفی در بیمارستان هم‌زمان درمان گردیدند.

نتیجه‌گیری: جراحی اصلاح کورکاتاسپسون در این گروه سنتی رخ خطر ناپذیر نبود. قابل معافیت قابل قبول‌ترین است. بنابراین می‌توان به نتایج درمان کورکاتاسپسون اوتورت ارجاع داد. در صورت وجود علامت‌های قلبی نیاز به عمل جراحی ندارید.