Abstracts

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Objectives There is not yet a general consensus about the diagnosis of left ventricular noncompaction. The echocardiographer may miss the areas with maximal noncompaction leading to a misdiagnosis. Accordingly, we suggested a new method to measure the percentage of myocardial hypertrabeculation/noncompaction using two-dimensional echocardiography.

Patients In this study, this new method was examined on 3 non-compaction, 26 dilated cardiomyopathy, and 25 normal subjects.

Results The mean percentages of myocardial hypertrabeculation/noncompaction were 3.59 ± 2.27 in control group, 8.86 ± 5.52 in dilated cardiomyopathic patients, and 38.1 ± 30.8 in noncompaction patients. A value of 16% could distinguish left ventricular noncompaction from dilated cardiomyopathy with 92% specificity and 100% sensitivity and from normal subjects with 100% specificity and sensitivity. This percentage had a statistically significant association with noncompacted to compacted myocardial thickness ratio ($P < 0.001$).

Key Conclusions This method showed good correlations with the other echocardiographic and magnetic resonance criteria. However, it is not dependent on finding the area of maximal involvement.

508 REGIONAL CARDIAC NETWORKS: CAN THEY WORK?

doi:10.1136/archdischild-2012-302724.0508

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Background Only eleven paediatric intensive care units (PICUs) in UK have cardiac surgical services on-site. Small proportion of patients admitted to non-cardiac surgical PICUs require transport to surgical centres. North West and North Wales Paediatric Transport service (NWTs) provides PIC transport service, including ability to conference call specialists. Alder Hey Children's hospital (AHCH) is the cardiac surgical centre and Royal Manchester Children's hospital (RMCH) only has cardiology on site.

Methods Retrospective review suspected cardiac cases transferred in 12 months:- assessing if time critical surgical patients underwent single transfer to surgical centre- interventions by referring hospital or NWTs.

Results Total 29 patients of suspected cardiac diagnosis.

Abstract 508 Graph 1

Abstract 508 Table 1 Interventions

INTERVENTIONS	REFERRING HOSPITAL	NWTS
Intubation	22	4
Central Venous Access	6	7
Dinoprostone (PGE2)	20	2
Inotropes	7	10
Inhaled Nitric oxide	0	3

Seven patients had time critical cardiac surgical lesions. One patient required a second immediate transfer to surgical centre. Retrospectively, this could have been predicted from clinical picture. 68% patients had dinoprostone started by referring team following advice from cardiologist or NWTs. Inotropes initiated on advice or by NWTs.

Conclusions Regional cardiac network can work effectively with improved communication particularly in acute scenario. Potential surgical cases may be predicted from clinical picture, especially if not resolving with full medical treatment.

509 PREVALENCE OF SIDEROPENIC ANEMIA IN CHILDREN WITH CONGENITAL HEART DISEASE LESS THAN ONE YEAR IN COMPARISON WITH HEALTH CHILDREN

doi:10.1136/archdischild-2012-302724.0509

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Introduction Syderopenic anemia is prevalent in all children with congenital heart disease, especially in children with signs of heart failure.

Objectives The purpose of this study is to determine the prevalence of sideropenic anemia in children with CHD and compares these results with data of health children less than one year in Kosova population.

Methodology Between 2000 and 2010 4236 children were evaluated retrospectively clinically and by echocardiography for congestive heart failure (CHF) caused by CHD. CHD was diagnosed in 832 patients (19.6%) while 78 patients (1.84%). Also from the study have been exclude children with complex CHD caused central cyanosis. In all patients were analyzed heart failure symptoms, hemoglobin levels, age of diagnosis, duration of hospitalization per year and death during medical attendance.

Results Anemia was present in 78% of children with CHD, and in 85% children with CHF. Hospitalization days per year in anemic patients had a significantly higher than in non-anemic patients (mean 32.5 ± 39.6 days per year versus 12.3 ± 14.3 days per year ($p < 0.05$). There was not found significant relation onset of heart failure symptoms and the risk of developing anemia between anemic and non-anemic patients. The evaluation showed also needed for the two times longer period of the treatment of patients with CHD with antianemic medication in compare with healthy anemic children.

Conclusion Anemia is prevalent in pediatric patients with CHD especially patients with heart failure. Study showed no higher mortality in anemic patients.

510 A NOVEL MYH7 VARIANT IDENTIFIED IN A CHILD WITH RESTRICTIVE CARDIOMYOPATHY

doi:10.1136/archdischild-2012-302724.0510

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Restrictive cardiomyopathy (RCM) is very rare in children and usually associated with a poor prognosis. Identification and thorough

diagnostic work-up are essential. We report the case of a 3 year old previously well girl, who was referred to A&E with shortness of breath. On admission, she was found to be in acute heart failure.

Echocardiography showed a restrictive cardiomyopathy and a very large pericardial effusion which was subsequently drained. A full cardiomyopathy screen was performed but all her metabolic indices were normal. Genetic investigations identified a novel MYH7 variant. The MYH7 gene is usually associated with hypertrophic cardiomyopathy. This patient is the second in the world in whom RCM has resulted from a variant invariant in MYH7, the first with a phenotypic effect and provides further evidence that this gene is linked with different cardiac phenotypes.

511 ATYPICAL WHEEZING CASES DURING INFANCY DUE TO GASTRIC VOLVULUS

doi:10.1136/archdischild-2012-302724.0511

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Background Gastric volvulus is rarely seen abnormality during childhood. Nonbilious vomiting, abdominal distension and dyspepsia are the most common presenting symptoms. Yet, it may cause reactive airway disease symptoms such as wheezing attacks, because of gastroesophageal reflux, and chronic cough in infants. Surgery after early diagnosis in gastric volvulus resolves symptoms completely and its prognosis is excellent.

Aim Here, two infants having chronic cough and recurrent wheezing attacks, unresponsive to bronchodilators, are presented.

Patients/Methods Physical examination of both cases revealed about 6-month-old male patients having recurrent wheezing, tachypnea, dyspnea and subcostal retractions. Widespread ronchi and sometimes rales were heard on both lungs of the patients. Rest of the examination was normal. In their laboratory evaluations, acute phase reactants, renal and liver function tests were within normal. Immunoglobulin G, its subgroups and other immunoglobulins were found to be normal. Sweat tests were normal. High resolution computerized tomography (HRCT) demonstrated minimal mosaic pattern on both lung parenchyma. Bronchoscopy was normal in the first infant. Cranial, abdominal ultrasonography and fundus (eye) examinations showed normal findings in both patients. Cardiac examination and echocardiography were normal. Oesophago-gastro-duodenography showed gastric volvulus, organo-axial and mesentero-axial, in both infants; respectively.

Results After the surgery for gastric volvulus and gastroesophageal reflux in the first case, the symptoms entirely improved. But, the symptoms of second case resolved without surgery.

Conclusion Our cases are being reported to emphasize the necessity of thinking gastric volvulus in the differential diagnosis of atypical wheezing infant, even which it is very nadir cause.

512 NEONATAL TESTES TORSION: CASE REPORT

doi:10.1136/archdischild-2012-302724.0512

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Introduction Torsion of the testes results from twisting of the spermatic cord, which leads to a compromised testicular blood supply and subsequent testicular infarction. The consequent ischemic damage affects long-term testicular morphology and sperm formation.

Case We describe a case of 2 day-old boy who presented with bilateral visible swelling in the scrotum. Physical examination findings

showed enlarged, hard, nontender right scrotal mass, and left hydrocele. Doppler ultrasonography demonstrated torsion of right testes and bilateral hydrocele. Exploration was performed using a median raphe incision in the scrotum. Right testes was delivered, detorsed, and placed in warm, moist sponges but it appeared nonviable and necrotic. Right orchiectomy was done. Left testes was normal, it was fixed to the scrotum. Postoperative recovery was uneventful.

Conclusion Perinatal testicular torsion is a rare condition. During newborn examination it must kept in mind if a scrotal mass or swelling occurs.

513 HEERFORDT'S SYNDROME IN AN ADOLESCENT BOY

doi:10.1136/archdischild-2012-302724.0513

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Introduction Heerfordt's Syndrome is characterized by bilateral uveitis, facial paralysis, fever and parotitis which is a rarely seen condition in sarcoidosis with neurological manifestations.

Case Report A fifteen year old boy admitted to our pediatric emergency service with fever, vomiting and swellings on his cheeks. Bilateral conjunctivitis, bilateral preauricular swellings, maculopapular rash on his anterior and posterior body areas and uncertain neck stiffness were detected on his physical examination. The rest of the examination was considered as in normal range. His medical history revealed an upper respiratory tract infection one week ago.

Lomber puncture was performed because of patient's neck stiffness. No cell and culture growth were detected on puncture sample. During his observation, left peripheral facial paralysis was developed and bilateral uveitis was detected while his high fever was still persisting.

Whole body gallium 67 scan was performed. Focal accumulations of gallium 67 in both lacrimal glands, parotid and submandibular glands(panda sign) were seen. Because of existence of uveitis, parotitis, left peripheral facial paralysis and fever, the patient was diagnosed as Heerfordt's Syndrome. Steroid (Prednisolone) treatment was started. The regressions of facial paralysis and parotid gland size were noticed after two weeks of the steroid therapy.

Result This case report was chosen to take attention to a rare cause of parotitis.

514 THE AUTOIMMUNE LYMPHOPROLIFERATIVE SYNDROME: A CASE REPORT

doi:10.1136/archdischild-2012-302724.0514

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Background The Autoimmune Lymphoproliferative Syndrome (ALPS) is an impairment of lymphocyte apoptosis expressed by generalized non-malignant lymphoproliferation, lymphadenopathy and/or splenomegaly. Majority of patients with ALPS harbor heterozygous germline mutations in the gene for the TNF receptor-family member Fas (CD 95, Apo-1) which are inherited in an autosomal dominant fashion. Somatic Fas mutations are the second most common genetic etiology of ALPS.

Case report We describe a two year old boy who was admitted with hepatosplenomegaly, generalized lymphadenopathy and anemia. Histopathological and immunohistochemical analysis of lymph nodes suggested a lymphoproliferative disorder in large granular lymphocytes. The lymphocyte phenotyping performed in the patient showed an increased population of T cells $\alpha\beta$ double