Aim Prolonged QT syndrome is now recognised as a major cause of sudden unexpected cardiac death, especially in children and adolescents.

Methods Selected from Paediatric Cardiology clinic list

Results A 3 1/2 year old fit and healthy boy, was found to be unresponsive by his parents at home. He was given Cardio-pulmonary resuscitation (CPR) in Emergency dept as noted to have Ventricular Fibrillation (VF) rhythm and was given 2 shocks. His total down time (from the onset of CPR to return of detectable pulse) was approximately 26 mins. CT head were normal. In Cardiac ICU he was cardioverted twice using 60J energy and started on bolus dose of Amiodarone followed by maintenance dose. He was intubated total of 3 days and was passively cooled for total of 72 hrs.

His ECG shown long QT syndrome and was started on Nadolol, titrated the dose based on serial 24hr tapes. His MRI head and Cardiac MRI was normal. His Cardiomyopathy screening blood tests were normal. His bloods were taken for Long QT syndrome genetic testing and was sent home with a consideration for inserting Intra cardiac defibrillator (ICD) when results available.

ECG performed on both parents and his sister were normal. He was sent home with automated external defibrillator for parents to carry around. Parents were advised to attach 12 lead ECG probes to his body at night and oxygen saturation monitor. Parents and nursery staff received basic life support (BLS) training.

His genetics blood tests came back positive for Long QT syndrome 3. Whole family was referred for genetics review and was listed for consideration of insertion of epicardial pacemaker.

Conclusions Long QT syndrome can be asymptomatic and be an incidental finding. However it can present as sudden syncope and malignant ventricular arrhythmia or sudden death in previously healthy individuals. All first degree family members of affected patients should be screened with ECG to determine whether others are affected.

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G76 TRANSFER TO REGIONAL CARDIAC CENTRE: ARE WE GETTING IT RIGHT?

doi:10.1136/archdischild-2013-304107.088

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Background Antenatal detection of cardiac malformation is currently 35%.¹ Although this has been steadily improving, still majority of cardiac malformations present unexpectedly in the postnatal period. Over recent years, Neonatologists have acquired echocardiographic skills to make diagnosis locally, thus reducing unnecessary transfer of patients to cardiac centre for diagnostic evaluation alone.

Aims and Methods: We audited all acute inpatient referrals to our regional cardiac centre at Liverpool over 2.5 years period from Mar-2008 – Oct-2010. Neonatal Badger dataset was used to identify all cardiac transfers to Liverpool. Patient notes were reviewed to look at the antenatal diagnosis, clinical presentation, local echocardiographic diagnosis, subsequent diagnosis concordance at cardiac centre and their outcomes.

Results There were 15 transfers to regional cardiac centre in the audit period. Echocardiography was performed in 14/15 patients (93%) locally. Of these 14 patients, 2 patients were referred semi-electively for PDA ligation and both were confirmed by cardiologist to be suitable for ligation. Of the remaining 13 patients, local echocardiographic findings were concordant with cardiologist's findings in 12 patients (92%). One patient suspected to have congenital

cyanotic heart disease turned out to have structurally normal heart and was treated as PPHN (discordance rate 8%).

Antenatal diagnosis was made in 5/13 (38%) which is in keeping with national statistics.¹

Excluding patients with PDA, 10/12 (83%) babies had abnormal saturations (<95%) and 2/12 (17%) had normal saturations (> 95%). Both the babies with normal saturations had aortic arch abnormalities.

3 babies had inoperable cardiac conditions and 1 baby had underlying severe chromosomal anomaly and multiple congenital malformations leading to palliative care plans.

Conclusions Our attempt at initial cardiac diagnosis was 93% compared to 41% and correct cardiac diagnosis concordance from our unit was 92% compared to 64% in the study published from Embrace transport service.²

Our concordance rates are comparable to similar work published from our local regional Neonatal unit.³

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G77 A CASE OF DILATED CARDIOMYOPATHY DUE TO NUTRITIONAL VITAMIN D DEFICIENCY RICKETS

doi:10.1136/archdischild-2013-304107.089

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Introduction Dilated Cardiomyopathy is an important cause of heart failure in children. It often has an idiopathic aetiology, but may also be caused by infection and can be inherited. Hypocalcaemia is a rare but important reversible cause of dilated cardiomyopathy. Vitamin D deficiency is the main cause of hypocalcaemia in almost all reported cases.

Case Report: An 8 month-old, Afro-Caribbean boy, presented with acute heart failure and respiratory distress. He was born in the UK and exclusively breastfed until 6 months of age. Cardiovascular examination revealed a gallop rhythm and a grade 2/6 pan systolic murmur at the apex, with a 2cm liver edge palpable. There was clinical evidence of rickets.

Investigations included a chest X-ray, which showed cardiomegaly and plethoric lung fields. ECG showed increased left ventricular voltages and a prolonged QTc interval. Venous blood gas revealed a metabolic acidosis and a low ionised calcium level (0.6mmol/l). Echocardiogram revealed a poorly functioning and dilated left ventricle with severe mitral regurgitation. Hypocalcaemia secondary to severe vitamin D deficiency was diagnosed by screening blood tests. In addition to supportive treatment for cardiac failure (furosemide and captopril) he was treated with alphacalcidol and Calcium-Sandoz.

Echocardiogram 2 months later showed a near normal left ventricular function with a well contracting ventricle. His biochemical profile normalised within 4 weeks of treatment.

Discussion Nutritional vitamin D deficiency leading to dilated cardiomyopathy has been increasingly reported, and indicates it as an important aetiology to consider in heart failure. A recent retrospective review showed that such infants can present in extremis, requiring inotropic and ECMO support. However, compared to other cardiomyopathies, there is a relatively good prognosis, with a mean recovery of left ventricular shortening reported at 12.4 months.¹

Rickets itself has seen a resurgence, especially in developed countries² This is a preventable condition, but obstacles^{3,4} have been reported, despite NICE⁵ and DoH⁶ guidelines for antenatal care, post-natal supplementation and clinical management.

Conclusion This case represents a serious consequence for undiagnosed vitamin D deficiency. In view of its preventable nature,