Results In the last 10 years, 11 babies had a documented diagnosis of Ebstein's anomaly. Ten of these were antenatally diagnosed. Four were born prematurely (<37 weeks). Eight babies required resuscitation at birth and nine babies required intubation within the first 24 hours of life. Many of the babies had other significant co-morbidities including: hydrops fetalis, seizure activity, ventriculomegaly, trisomy 21 and persistent pulmonary hypertension (PPHN). Two babies also underwent therapeutic hypothermia. All of the babies born prematurely died within the neonatal period. Of the seven term babies with Ebstein's anomaly, three were discharged home, two were transferred to the Regional Paediatric Cardiology unit and two died on the neonatal unit. The two babies that were transferred the Regional Paediatric Cardiology unit subsequently died. The three surviving infants continue to be followed up. One of these patients is now seven years old and has not required any surgical intervention.

Conclusion In our experience, Ebstein's anomaly was uniformly fatal in babies born prematurely. Furthermore, in term babies the survival was 43%. The overall survival of Ebstein's anomaly presenting in the neonatal period in our cohort was 27%. Although this is an outcome of a small cohort of neonates with a rare congenital heart disease, it is important that parents are counselled appropriately antenatally so that they are adequately prepared for the potentially poor outlook of babies born with Ebstein's anomaly in the neonatal period.

G23(P) CHILD WITH HYPERTROPHIC CARDIOMYOPATHY DUE TO PRIMARY CARNITINE DEFICIENCY (PCD)

Y Kodeeswaran, S Rajendran. *Paediatrics, Luton and Dunstable Hospital, Luton, UK*

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Introduction Primary carnitine deficiency (PCD) is a rare autosomal recessive disorder of fatty acid oxidation, in which the transport of carnitine into cells is impaired. Carnitine deficiency blocks the oxidation of long-chain fatty acids in the mitochondria that leads to heart and hepatic disease, myopathy, nonketotic hypoglycaemia, and neurological complications. **Case Report** 7 month old child presented with 3 day history of vomiting.

He was born at term to non-consanguineous parents. He had returned to the UK recently, after a 4 month stay in India. Whilst in India he had had 2 hospital admissions with vomiting; one of which required admission to PICU with hypoglycaemia and metabolic acidosis, diagnosed as gastroenteritis and the next was for UTI.

O/E: Baby was drowsy, with evidence of subcostal recession and hepatomegaly.

Started on antibiotics for possible sepsis, given saline bolus, and started on maintenance fluids. He later developed hypoglycaemia, metabolic acidosis, and fluctuating consciousness. After a discussion with the metabolic team, he was intubated and transferred to PICU.

Investigations Ammonia levels increased from 164 to 340 micromol/L in 6 hours, high ALT and deranged clotting. Free carnitine <1 umol/l.

Initial ECHO showed mild left ventricular hypertrophy. He had cardiopulmonary arrest on day 3 of PICU admission and consequently needed ECMO for 4 days. He was noted to have hypertrophic cardiomyopathy with left ventricular dysfunction. He required multiple inotropes and nitric oxide

and also developed intracranial bleeding and subsequent seizures, requiring Levetiracetam. Other complications included AKI (required CVVH), liver failure, MRSA Bacteremia and E-Coli septicemia, which were treated.

Significant improvement was noted following levocarnitine replacement- he made a good recovery and was discharged home.

Discussion Primary carnitine deficiency (PCD) usually presents with cardiomyopathy and muscle weakness and is one of the treatable causes of cardiomyopathy.

Treatment of this disorder with L-carnitine is highly effective in correcting cardiomyopathy and muscle weakness.

Child may be asymptomatic until they develop signs of cardiomyopathy and congestive heart failure. Cardiac function does not respond to inotropes or diuretics. If no carnitine is supplemented, progressive heart failure eventually leads to death.

G23(P) A 10 YEAR EXPERIENCE OF NEONATES WITH TRUNCUS ARTERIOSUS

M Jagga, EL Stockley, T Desai, SV Rasiah. *Birmingham Women's and Children's NHS Foundation Trust, Birmingham, UK*

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Background Congenital heart defect affects around 10 in a 1000 live born babies. Truncus arteriosus is a rare conotruncal defect in which a common arterial trunk supplies systemic, pulmonary and coronary circulation. The incidence of truncus arteriosus is quoted between 1–2% of all congenital heart malformations.

Aim To evaluate the initial neonatal management and outcomes of newborns with truncus arteriosus.

Method We conducted a retrospective review of all newborns with truncus arteriosus admitted to our neonatal unit between 01/04/2009 and 31/03/2019. Data were collected from Badger and Cardiology databases to evaluate the initial neonatal management and long-term outcomes of this cohort of patients.

Results Sixteen newborns with truncus arteriosus were admitted to our neonatal unit in the last 10 years. Their mean gestational age was 36 weeks with a mean birth weight of 2407 grams. Cardiac defects were detected prenatally in 13 patients (82%). Three babies were diagnosed postnatally. One following failed pulse oximetry screening requiring non-invasive respiratory support and 2 diagnosed following on-going respiratory support on echocardiography. Respiratory support was initiated within the first 24 hours of life in 8 patients; these included all patients with a postnatal diagnosis of truncus arteriosus. Four patients required mechanical ventilation. Whilst in the neonatal intensive care unit, 3 babies developed heart failure and were treated with diuretics and 2 developed necrotising enterocolitis which was managed conservatively. Median length of stay before transfer to our local surgical cardiology centre was 3 days. Fourteen patients (87%) underwent common arterial trunk repair. 11 (78%) babies survived to date. Two patients died in the initial post-operative period and one patient at 14 months of age. The remaining two patients did not receive any surgical intervention and subsequently died (4 and 11 days old).

Conclusion Truncus arteriosus is a rare conotruncal defect with the majority being detected antenatally in our cohort. They

are likely to require respiratory support in the neonatal period. Despite the complex nature of their congenital heart disease, their surgical outcomes are good in our cohort of babies. The following information can be used when counseling parents antenatally and managing their expectations.

G24(P) ANXIETY AND CHEST PAIN: A CHICKEN AND EGG SITUATION?

L Doherty. Paediatric department, Torbay Hospital, Torbay and South Devon NHS FT, Torquay, UK

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Introduction Chest pain is a common paediatric presentation and can cause anxiety in patients, parents and healthcare professionals. Media coverage can heighten anxiety, but underlying cardiac pathology is rare in children. It is important to rule out serious underlying pathology and reassure appropriately.

Case History A 16 year old boy with Type 1 Diabetes Mellitus was referred by his diabetes nurse non-specifically unwell with hyperglycaemia and pallor. He had a two day history of anxiety associated with improving dyspnoea and chest pain, and was taking his GCSEs. He was fully immunised but had been given a recent telephone diagnosis of measles, and is needle phobic.

On presentation, he was tachycardic with otherwise normal observations, and felt to be anxious. He became febrile and tachypnoeic with capillary blood gases showed a worsening non-ketotic metabolic acidosis, and a CRP of 80. He eventually agreed to a cannula under midazolam sedation 10 hours post attendance.

He developed worsening central chest pain and dyspnoea which prevented him lying flat. An ECG and CXR were requested, however there was no ECG machine on the ward. Furthermore, nursing staff are not trained to record ECGs. On examination, he was pale, sweaty with a soft systolic murmur. His venous bloods showed a haemoglobin of 70, MCV 49 and platelets of 673.

The ECG showed inferior ST elevation and inferolateral T wave inversion. A troponin was 1595 and an echocardiogram showed an akinetic inferior wall, dilated right heart, bright pericardium and no pericardial fluid. He was transferred to a tertiary adult cardiac centre, but due to the challenges with his diabetes and needle phobia, was moved to PICU under the paediatric cardiologists. A diagnosis of viral myocarditis was made causing left coronary inflammation, possibly exacerbated by microvascular disease secondary to diabetes, and anaemia.

Conclusion Always rule out serious pathology with a background of chest pain first, and be aware that anxiety may be a cause but also an effect of chest pain. Be aware of becoming task focused, and the risk of not seeing the bigger picture.

G25(P) DRUG INDUCED SHORT QT INTERVAL WITH USE OF ANTI-EPILEPTIC MEDICATIONS

S Choopa, R Sesham, P Nair. Paediatric Department, Bedford Hospital NHS Trust, Bedford, UK

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Introduction Although drug induced long QT intervals is known to have proarrhythmic effect, the same is not known about drug induced short QT intervals. Drug induced short QT syndrome is a rare occurrence and have mainly been reported with the use of anti epileptic medications. Proarrhythmic effects of drug induced QT shortening has only been shown with digitalis. The clinical relevance of drug induced QT shortening is currently unclear.

Case Report We report a case of drug induced QT shortening in a 15 year old boy being treated for Juvenile Myoclonic epilepsy. He was initially being treated with Sodium Valproate with later addition of levetiracetam. He started having complaints of intermittent palpitations and occasional dizziness. He was otherwise well with no family history of sudden deaths etc. His examination and echocardiography were normal. His ECG showed sinus rhythm with a QT interval of 340 milliseconds. His previous ECG 6 months ago had shown a QT interval of 330 milliseconds. An ECG done 3 years ago at diagnosis had shown a normal QT interval of 400 ms. He had subsequent multiple ambulatory monitoring and exercise stress tests which were also normal. He is under ongoing follow up with the cardiac team and his anti-epileptic medications have been continued.

Discussion Drug induced short QT interval is a rare phenomenon and have been mainly seen with use of some anti-epileptics, digitalis & promethazine. Although there have been case reports of QT prolongation with use of levetiracetam, we could not find any reports of QT shortening. There were few reports of QT shortening when sodium valproate was used as a co-medication. In this case we are not sure if it is the valproate on its own or the addition of levetiracetam which caused the QT shortening. However based on no evidence of proarrhythmic effect with drug induced QT shortening, we agreed to continue his anti epileptic medications while closely monitoring him. More evidence base and research is needed to throw light on management of patients with drug induced short QT-intervals.

G26(P) SCREENING FOR CONGENITAL HEART DISEASE AMONG PRIMARY SCHOOL CHILDREN IN KHARTOUM STATE

¹E Babikir, ²B Elbasheer, ³R Berair. ¹MBBS, MD Paediatrics SMSB, Fedail Hospital, Khartoum, Sudan; ²MBBS, MD Paediatrics U of K, PCIC Malaysia, El-Mawadah Hospital, Khartoum, Sudan; ³MBBS, MD paediatrics CES, El-Akademi Hospital, Khartoum, Sudan

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Background Congenital heart defects are the most common type of birth defects and remain the leading cause of death from congenital malformations.

Aims To determine the prevalence of latent congenital heart disease (CHD) among Governmental primary school children in Khartoum state.

Methods This is Descriptive cross sectional school based study which was conducted in the three localities in Khartoum state (the capital of Sudan) which are Sharq Alneel, Khartoum and Omdurman locality using a multistage clustered sampling technique recruited 2400 pupils aged between 6 and 14. Team of doctors worked the history and clinical examination to be related to Echocardiography (Echo) findings which had been done by consultant