

Table of Content

36 Long-Term Outcome of Dilated Cardiomyopathy Phenotypes: 25-year single centre experience

Mr Samuel Brown¹, Dr Minoth Kanagaratam², Dr Oscar Nolan³, Dr Adrian Crucean³, Mr John Stickley³, Ms Esther Poole³, Dr Ashish Chikermane³, Dr Milind Chaudhari³

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18 Introducing a bedside guide to improve paediatric ECG interpretation and clinician confidence

Dr Paolo Hollis¹, Dr Leonie Wong², Dr Prashanthi Katta³

¹Great Ormond Street Hospital For Children, ²Royal Brompton Hospital, ³East Surrey Hospital

33 Increasing incidence of Infectious Endocarditis in children with Congenital Heart Disease: A COVID 19 casualty or A need for improved awareness

Dr Neeraj Nirmal¹

¹University Hospitals of Leicester NHS Trust

52 Right to left or left to right?

Harriet Riggs¹, Dr Gopi Vemuri, Dr Giri Soda

¹Manchester University Foundation Trust

43 Impact of undiagnosed cardiac vascular anatomy on neonatal peripheral long line insertion leading to unneeded withdrawal.

Dr Jamie Shah¹, Dr Bikash Bhojnagarwala¹

¹Chelsea and Westminster Hospital

50 Paediatric COVID Experience in a District General Hospital

Dr Wan Yung Siu¹, Dr Ramona Onita¹

¹East Of England NHS

26 An Unusual Cause of Headache

Dr Rachel Hoi Khoon Tan¹, Dr Abdulla Tarmahomed¹, Dr Giri Soda²

¹Alder Hey Children's Hospital, ²Royal Manchester Children's Hospital

46 Appropriate Use Criteria for Paediatric Echocardiograms: Friend or Foe?

Dr Marisa McMillan¹, Dr J Higgins¹, Dr Emma Hulbert-Powell², Dr Indranil Misra³, Dr N Brennan⁴, Dr C Zaborowski⁵, Dr Tara Bharucha⁶, **Dr Lucinda Winckworth¹**

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49 Current trends in district general PEC clinic referrals

Dr M McMillan¹, Dr J Higgins¹, Dr E Hulbert-Powell², Dr I Misra³, Dr N Brennan⁴, Dr C Zaborowski⁵, Dr T Bharucha⁶, **Dr Lucinda Winckworth¹**

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Long-Term Outcome of Dilated Cardiomyopathy Phenotypes: 25-year single centre experience

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Introduction:

Dilated cardiomyopathy (DCM) is the commonest cardiomyopathy and the main indication for heart failure (HF) admission and heart transplantation (HTx) in children. Management protocols have evolved beyond HTx to include long-term inotropes and mechanical cardiac support (MCS), influencing long-term outcomes.

Methods:

A single-centre retrospective study of children (<16 years) with DCM from 1994 to 2019. Clinical presentation, management and outcomes were analysed for survival and their predictors.

Results:

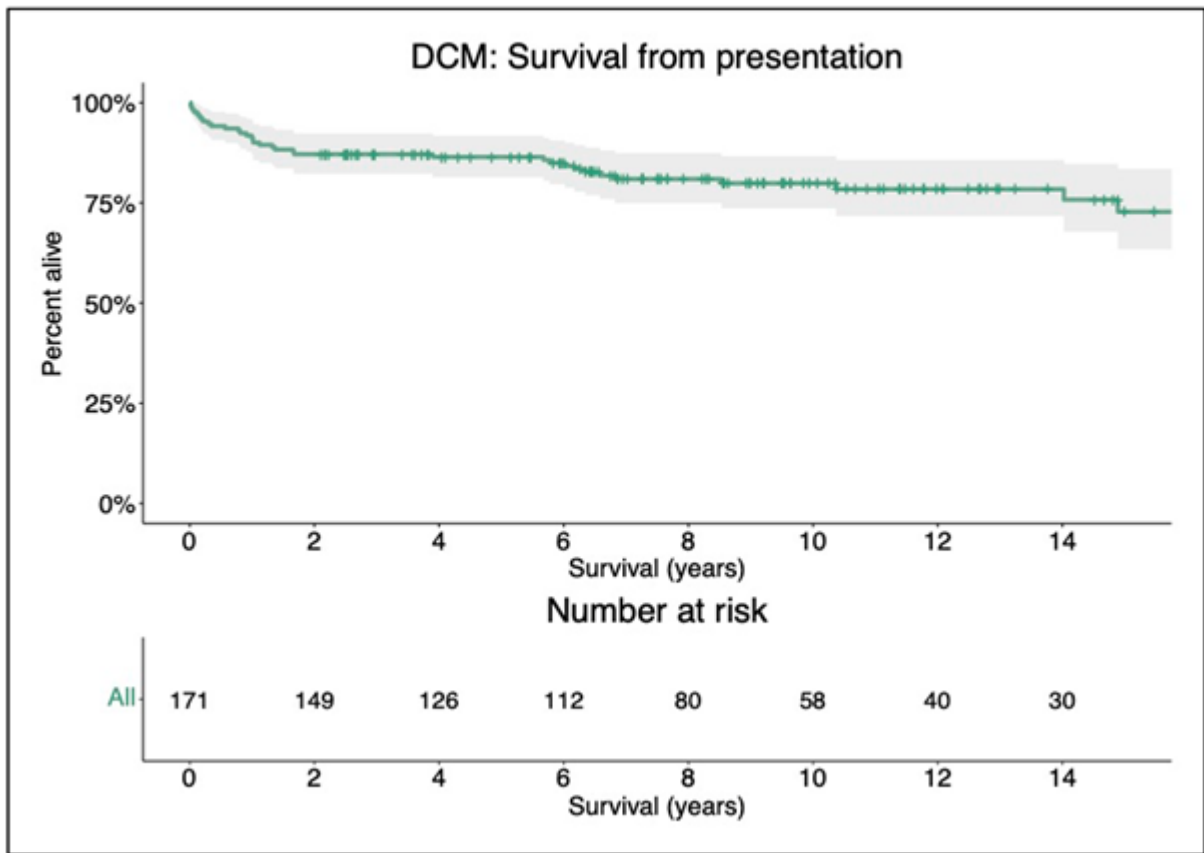
176 patients were identified. The median age at presentation was 1.6 years (Interquartile range, IQR 0.4-8.5) and follow-up 5.8 years (IQR 2.4-9.0). Aetiologies; idiopathic DCM (IDCM) (31%), myocarditis (19%), neuromuscular diseases (12%), familial (9%), Alström syndrome (6%) and metabolic (5%) amongst others. Almost three-quarters (72%) presented with acute HF and a fractional shortening of 14.0% (IQR 10.0-17.0%). Of these, 46% required intensive care, 33% intravenous inotropes and 20% MCS. Twenty children underwent HTx, mostly IDCM (n=13) and familial (n=4). There were 38 deaths, 20 with acute HF. Kaplan-Meier survival estimates were 91% (95% CI, 87.1-95.6%) and 80% (95% CI, 73.7-86.7%) at 1 and 10 years, respectively (Figure 1). Myocarditis had the best 10-year survival (91%; 95% CI, 74.6-94.9), whilst syndrome-associated-DCM, the lowest (67%; 95% CI 51.8-86.5). Children presenting aged 5 had the lowest relative mortality risk (Hazard ratio 0.52 (95% CI 0.20-1.39), p=0.03).

Conclusion:

In this 25-year single-centre study the mortality from DCM varied considerably according to age, severity and aetiology. Presenting at a very young or old age, syndromic association, and severity of cardiac dysfunction were indicators of poor survival.

Graph

Figure 1 - Kaplan-Meier survival curve (with confidence intervals, CIs) for all dilated cardiomyopathy (DCM) patients.



Notes: 5 patients did not have a recorded date of presentation.

Introducing a bedside guide to improve paediatric ECG interpretation and clinician confidence

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Objective: Does a bedside guide improve Paediatric ECG interpretation and clinician confidence?

Introduction: Paediatric ECG interpretation is often found to be challenging by paediatric trainees, particularly in the context of the changing physiology with age of paediatric patients. We felt that a brief but clear bedside guide would be beneficial in improving both the quality of interpretation, and clinician's confidence.

Study design: We created a brief bedside paediatric ECG interpretation guide limited to an A4 sheet. We invited junior doctors working in general paediatrics department to take part. Participants were asked to review an ECG and complete a questionnaire. They were then given a copy of the ECG interpretation guide and asked to review the same ECG and complete the same questionnaire again.

Results: 19 participants completed the survey, including GP trainees and general paediatrics trainees at various stages of training. Without the guide, 66% of questions asked about the ECG were answered correctly, and this increased to 82% when the participants were supplied with the guide. 90% of trainees correctly identified Right ventricular hypertrophy after using the guide showing an increase of 26%. 82% of respondents said that the guide made a difference to how they interpreted the ECG and 72% felt that their confidence in their interpretation was improved with access to the guide.

Conclusion: A brief ECG interpretation guide in general paediatrics improved the quality of interpretation as well as clinician's confidence in interpretation of Paediatric ECG.

Increasing incidence of Infectious Endocarditis in children with Congenital Heart Disease: A COVID 19 casualty or A need for improved awareness

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Infective Endocarditis (IE) is a major concern in patients with congenital heart disease (CHD). Adequate awareness and dental hygiene carry an important role in its prevention. An increase in the incidence of these cases has been reported during the COVID 19 pandemic. This could be due to the constraints in routine dental surveillance during the pandemic. Our centre also witnessed an increase in such cases requiring an intervention during this period.

AIM:

We sought to identify and understand parental knowledge between CHD and IE, and measures taken for maintenance of good dental hygiene.

METHODS:

We conducted a face to face survey amongst our local cohort of CHD patients and their families during outpatient clinics and ward admissions.

RESULTS:

We surveyed 48 patients and their families. 33 patients had undergone a cardiac surgery and 15 patients had undergone an interventional catheter procedure.

Preliminary analysis showed the following, only 50% (n=24) were under routine dental surveillance. Of the remaining population, 25% (n=12) were not enrolled in a dental practice. Parents reported constraints in access to dental care during the pandemic. Only 79% (n=39) were meeting daily dental hygiene needs, as advised. Amongst the parent population, only 56% (n=27) understood the association between poor dental hygiene and IE.

CONCLUSION:

Our survey highlights the importance of awareness in primary prevention of IE in patients with CHD. Given the increasing shift towards remote consultations, the importance of patient awareness should not be overlooked

Right to left or left to right?

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PH was admitted to the Neonatal Unit at 12 hours of age with low oxygen saturations of 85% picked up on NIPE. Examination was otherwise unremarkable and there was no significant history or risk factors for sepsis. She was commenced on high flow oxygen initially requiring 40% FiO₂ to maintain saturations > 92%. Local Echocardiogram was unremarkable, apart from a right to left shunt at the atrial septum across the PFO and a bulging septum towards the left. However PDA was left to right and there was only trivial TR. A diagnosis of Persistent Pulmonary Hypertension was made and following discussion with tertiary cardiology she was managed locally with high flow oxygen. Oxygen requirements escalated to 80-100% over the next 2 days. She was intubated and ventilated, commenced on Nitric Oxide and transferred to the tertiary neonatal unit. She remained difficult to oxygenate and was commenced on high frequency oscillation ventilation, however this did not improve oxygenation. Nitric Oxide was continued in addition to Prostin due to ongoing difficulties with oxygenation. An echocardiogram was repeated which showed no signs of PPHN and a structurally normal heart. Following cardiology review and further detailed echocardiography a diagnosis was made of large Eustachian Valve causing obstruction to the Tricuspid Valve inflow. The Eustachian valve formed a loopy dainty structure in the right atrium directing flow from the IVC into the left atrium, hence resulting in a right to left shunt. Oxygen saturations stabilised at 85-90%.

Impact of undiagnosed cardiac vascular anatomy on neonatal peripheral long line insertion leading to unneeded withdrawal.

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Background:

Peripheral long lines require large bore vessel placement due to known increased risks of perforation and extravasation in line tips in superficial or fine vessels. Multiple asymptomatic variants in cardiac and vascular anatomy exist within the neonatal population that are often undiagnosed. We reviewed 2 cases (Figure 1) where the lines were almost or completely removed due to undiagnosed anatomical variants.

Case 1:

A peripheral long line was inserted via scalp vein. The clinician was worried to see it taking a very abnormal course via the left side of heart and entering the cardio chamber. However, the neonate had Bilateral Superior Vena Cava's (SVC's) with left SVC draining into coronary sinus, explaining x-ray findings. Once the diagnosis was established the line was withdrawn so that the tip was in the left SVC suitable for use.

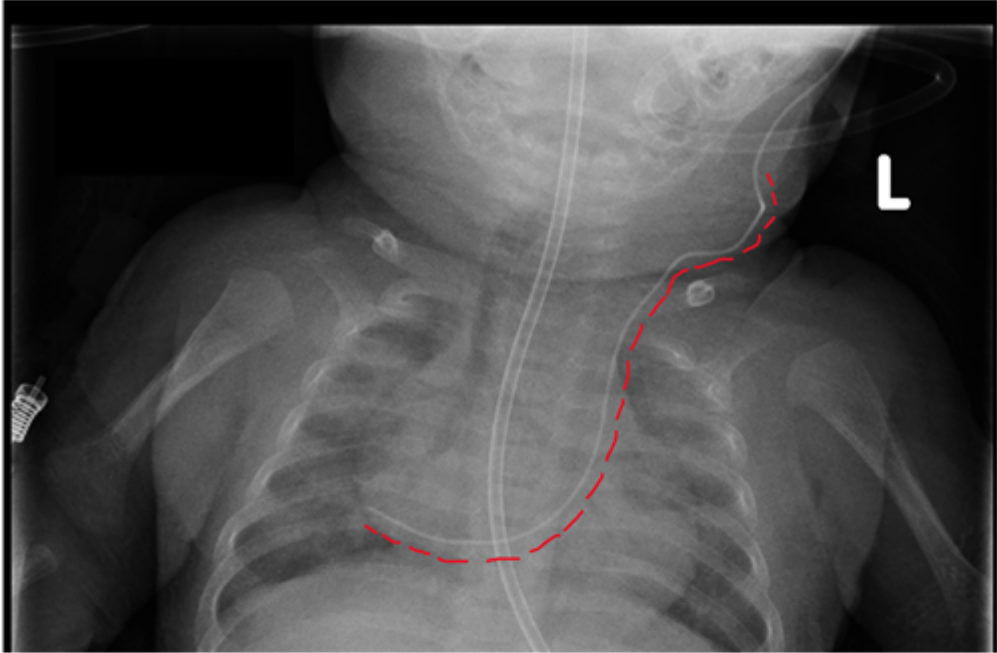
Case 2:

The first long line in a premature infant (inserted via right saphenous vein), was found to be crossing the midline and therefore removed. The second line, (inserted in the left saphenous vein), was found on the left side until the diaphragm and was also removed. Subsequent echocardiogram showed an interrupted infrahepatic Inferior Vena Cava (IVC) with azygos continuation and left atrial isomerism. Retrospectively both long lines were removed unnecessarily.

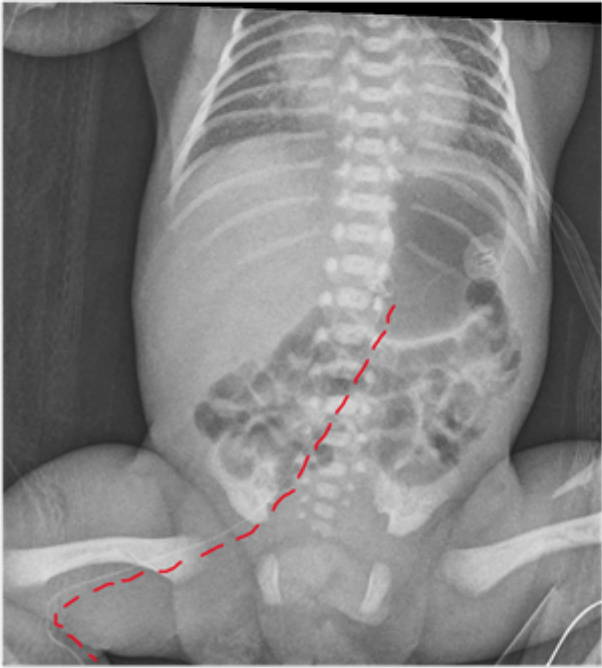
Conclusion:

These cases highlight the importance of being aware of anatomical variants to avoid unneeded complete withdrawal of long lines especially in such difficult cases (where all veins have been exhausted). A timely diagnosis by Echocardiography should be prompted whenever in doubt.

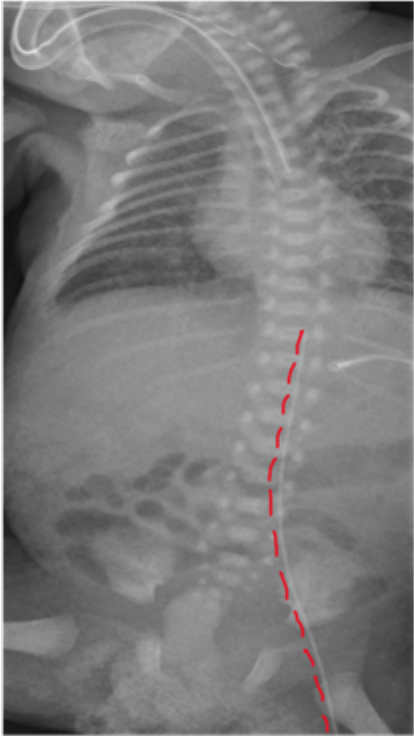
Image



Case 1



Case 2 - 1st Long line



Case 2 – 2nd Line

Paediatric COVID Experience in a District General Hospital

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Objectives

SARS-COV2 infection caused 2 major period of lockdowns and school closure in England. Our study aims to undertake a review of (1) the number of COVID swab positive paediatric patients (under 16 years) attending children's assessment unit (CAU) with possible COVID symptoms as per RCPCH guideline in the 2 periods of national lockdown (2) cardiac involvement of COVID swab positive patients, or patients positive for COVID and presented as PIMS syndrome (3) patients with long COVID syndrome

Design

Retrospective study from 23/3/2020-1/6/2020 and 30/10/2020-8/3/2021 at the time of lockdown

Settings & Patients

Paediatric patients (under 16 years) presenting to CAU with one or more COVID symptoms as per RCPCH guideline

Results

Between 23/3/2020-1/6/2020, 90 patients were symptomatic with one or more COVID symptoms and 1 patient was COVID swab positive (1%). Between 30/10/2020-8/3/2021, 964 were symptomatic with one or more COVID symptoms and 22 patients were COVID swab positive (2.3%). 5 swab positive patients had cardiac review: 2 of them due to new heart murmur, 1 of previous heart murmur and 2 existing cardiac patients. 2 patients were diagnosed with PIMS syndrome: one presented with cardiogenic shock and one presented as myocarditis with globally depressed myocardial function and reduced ejection fraction on echocardiograms. 1 patient was referred to tertiary centre for long COVID syndrome after encountering COVID infection.

Conclusion

All covid swab positive children did not have any cardiac complications. Those children presented with PIMS syndrome had some cardiac involvement at presentation but returned to normal upon follow-up.

An Unusual Cause of Headache

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A 15 year old girl presented with a prolonged history of headache lasting a few years. She was reviewed by the GP and found to be hypertensive with a blood pressure measuring 160/110mmHg in both arms.

She was referred to the paediatric cardiology clinic for further assessment. On examination, she was found to have a soft systolic murmur in the aortic region. A radio-femoral delay was noted with slightly weak femoral pulses.

Her echocardiogram demonstrated a well-developed ascending and transverse aorta; there was discontinuity of the aorta between the second and third branches. Collateral flow was seen around the descending aorta. The intracardiac anatomy and function were within normal limit.

A contrast CT aortic angiogram confirmed a left sided aortic arch with a bovine type branching pattern and a long segment (4.8 cm) Type A interrupted aortic arch.

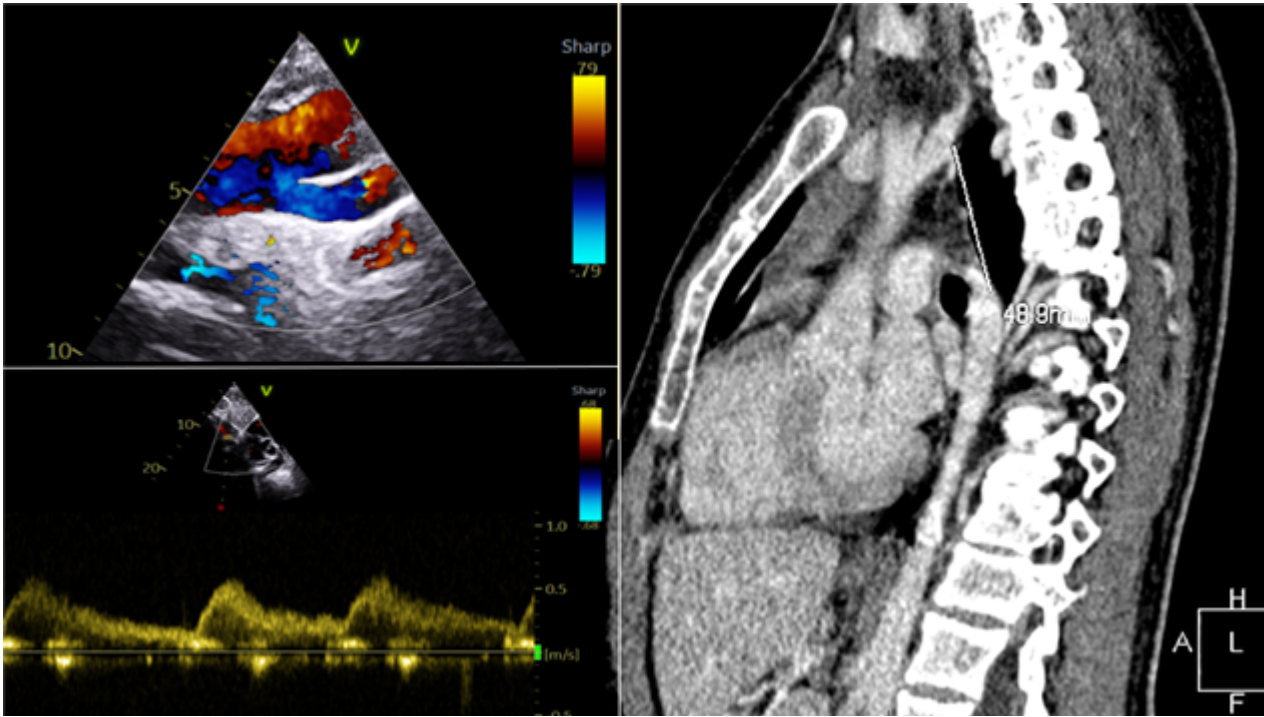
Discussion

Most paediatric headaches are benign with leading causes including tension-type, migraine and viral illness. Cardiovascular defects as a cause for headache remain uncommon but significant when present. A comprehensive history of the headache should be followed by a thorough systemic examination.

Conclusion:

1. It is important to measure blood pressure, palpate peripheral pulses and assess for radio-femoral (or brachio-femoral) delay when examining a child presenting with headache and/or hypertension.
2. Cardiac causes of hypertension – such as coarctation of aorta and interrupted aortic arch remain an important differential diagnosis for the child presenting with headache and/or hypertension and must be explored if an alternative diagnosis is not found.

Image



Appropriate Use Criteria for Paediatric Echocardiograms: Friend or Foe?

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Objectives and background:

Appropriate Use Criteria (AUC) published in 2014 guide the requesting of paediatric echocardiograms based on specific history points or examination findings, with each criteria rated “appropriate”(A), “may be appropriate”(M) or “rarely appropriate”(R). Generated in the USA, they have not been assessed for the UK paediatric population. Our aim was to use AUC to review level 3 paediatric clinic referrals across the Wessex/Thames Valley region.

Methods:

All first cardiac referrals from five district general hospitals over a 3 month period (01/12/19-01/03/20) were included. Prospective data collection was used to assign an AUC to each patient and secondary outcomes recorded.

Results:

128 patients were seen, with 34/113 AUC represented. Half of referrals were rated “R” with only 33% (n=42) having an “A” rating. Referral appropriateness was not significantly affected by age (P 0.86) or referral source (P 0.56). The yield of significant pathology from “A” referrals was higher, trending towards significance (P 0.056) but with a specificity and sensitivity of 0.72 and 0.75 respectively and positive predictive value of only 0.14.

Conclusion:

AUC were easy to apply and interpret, appearing applicable to our population. However sensitivity and specificity for predicting pathology were relatively poor. Although only referring “A” rated patients would have meant a 65% referral reduction, 25% of patients with echocardiogram findings necessitating tertiary referral would have been missed. Before dismissing the potential usefulness of AUC though, larger scale evaluation is needed due to small study numbers after the COVID pandemic forced an early termination to data collection.

Current trends in district general PEC clinic referrals

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Objectives and background:

National recommendations are that district general hospitals have a paediatrician with expertise in cardiology (PEC). Despite an increase in PEC-led clinics, the annual number of referrals are increasing and current services risk becoming overwhelmed. Our aim was to review new PEC-led clinic referrals in our region.

Methods:

Data was collected from PEC-led clinics in five DGHs in Wessex/Thames Valley between 01/12/19–01/03/20. Inclusion criteria were first referral and under 16 years old. Recorded information included age, referral source, referral reason, risk factors, examination, echocardiogram findings and onward plan.

Results:

There were 128 eligible patients, the majority aged under 5 (62%). The commonest referral reasons were: asymptomatic ejection systolic murmur (42%), cardiac-related family history (23%), pathological murmur (8%) and genetic disorders with cardiac risk (6%). Only 17% had positive findings on echocardiogram, with less than half deemed significant (36%, 8/22). Abnormal scan yield was significantly higher in children under 1 year ($P < 0.01$). Overall a quarter required ongoing follow-up, mostly in PEC clinics (80%, 28/35).

Conclusions:

PEC clinic activity is increasing but the abnormal echocardiogram yield is low. All children over 1 year with significant pathology had pre-alerting symptomatology or signs. With the long post-COVID-lockdown waits, this supports a general paediatric review of children over 12 months with an innocent-sounding ejection systolic murmur and no risk factors or symptoms, giving adequate safety netting rather than automatic referral to PEC clinic. Echocardiogram appointments could then be reserved for children with risk factors, pathological examination or those aged under 1 year.