replacement therapy wich significantly increased the survival of these patients.

PO-0044 A REVIEW OF THE CLINICAL PRACTICE OF PAEDIATRICIANS WITH EXPERTISE IN CARDIOLOGY (PECS) IN THE UK

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10.1136/archdischild-2014-307384.719

Background Recently there have been significant developments in PEC training and service aims including the recognition of Paediatrician with Expertise in Cardiology Special Interest Group (PECSIG) by the RCPCH and British Congenital Cardiac Association (BCCA), the creation of an agreed SPIN-module training curriculum for PECs and the development of PEC-specific service standards by the BCCA.
Aim To find out which clinical services are provided by PECs and to determine the extent of variability in their clinical practice.
Methods An internet-based questionnaire was sent out via PEC-SIG and NICHe (Neonatologists with Interest in Cardiology and Haemodynamics) contact databases and an NHS directory. Non-responders were followed up via telephone.
Results The response rate was 72% (129 of 179 hospitals). PECs carried out echocardiography in all hospitals in which they were employed (69%). Support for this service was provided by echo-technicians in 36% of hospitals and by neonatologists in 27%.

PEC-led outpatient clinics were held at least fortnightly in 66% of hospitals. The mode duration of appointment for a new patient was 30 min (range <20 min to >45 min) while for the follow-up appointment the mode was 20 min (range <20 min to >45 min).

Telemedicine facilities were established in only 45% centres, where sharing echocardiogram images via PACS was used most commonly.
Conclusion There remains significant variation in PEC services especially regarding the frequency of clinics, the duration of appointments and telemedicine utilisation. It was reassuring to see a relatively high number of hospitals offering PEC-led echocardiography and other PEC service provision.

Abstract PO-0044 Table 1 PEC service provision

Type of service	Percentage of centres providing service
Paediatric echocardiography	87%
12-lead ECG	98%
24-hour Holter ECG	91%
24-hour ambulatory BP monitoring	74%
Long term external cardiac monitoring (e.g. cardiac memo)	56%
Exercise testing	47%
Other services*	4%

** a handful offered TILT test and fetal echocardiography facilities*

PO-0045 A REVIEW OF THE AVAILABILITY OF PEC (PAEDIATRICIAN WITH EXPERTISE IN CARDIOLOGY) SERVICES IN THE UK

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10.1136/archdischild-2014-307384.720

Background A national survey completed in 2008 reported that PEC services were available in only 35% of non-specialist paediatric cardiology hospitals in the UK. In 2012, the NHS *Specialised Services Commission* recommended that there should be an increased level of PEC services in all hospitals as part of their 'Safe and Sustainable' review.
Aim To determine the availability of PEC services in the UK.
Methods An internet-based questionnaire was sent out via PEC-SIG and NICHe (Neonatologists with Interest in Cardiology and Haemodynamics) contact databases and an NHS directory. Non-responders were followed up via telephone.
Results The response rate was 72% (129 of 179 hospitals). Of these hospitals, 69% had established PEC services with at least 1 PEC employed and 19% had two or more PECs. In all centres echocardiograms were performed by either PEC consultants or specialist paediatricians, with support from echo-technicians in 36%.

Local PEC-led outpatient clinics ran at least fortnightly in two-thirds (66%) of hospitals, whilst 63% of hospitals held out-reach clinics with a paediatric cardiologist from a specialist centre at least monthly. However, 12 of the hospitals without any PEC services reported that they never held out-reach clinics either.
Conclusion There has been a substantial increase in PEC availability in non-specialist paediatric cardiology hospitals (69% as compared to 35% in 2008) but still 31% had no established PEC services. In most but not all hospitals without PEC services, support is offered by tertiary-centres for paediatric cardiology through out-reach clinics.

PO-0046 CLINICAL AND ECHOCARDIOGRAPHIC EFFECTS OF HYPOCALCEMIA SECONDARY TO SEVERE VITAMIN D DEFICIENCY (VDD) AND EFFECT OF TREATMENT

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10.1136/archdischild-2014-307384.721

Introduction Hypocalcemia (HC), without an underlying myocardial disease, is a relatively uncommon but reversible cause of congestive heart failure.
Objective We studied the cardiac functions (heart rate, blood pressure, ECG and Echocardiographic parameters (Fractional shortening (FR), left ventricular end diastolic diameter (LVEDD) of 14 children who presented with hypocalcaemia due to VDD before and 2-4 weeks after treatment with an IM dose of vitamin D 3 (VD) (10,000 IU/kg).
Results Correction of HC and VDD was associated with marked improvement of the LVEDDSDS (3.2 ± 4.4 to 1.1 ± 2.8) and slowing of the heart rate (from 101 ± 34 to 94.7 ± 30/min). The FS and QTc did not change. The LVEDDSDS was negatively correlated with serum calcium level (r =-0.46, p = 0.03) and PTH concentrations (r = 0.44, p = 0.032) but not with 25OHD level (r= -0.2, p = 0.2).

Abstract PO-0046 Table 1 Comparison between cardiac and biochemical parameters before versus after vitamin D therapy

	Before	After
Systolic BP mmHg	105 ± 15	104 ± 15
Diastolic BP mmHg	61.3 ± 10.3	62.4 ± 9
HR (beat/min)	101 ± 34	94.7 ± 30*
LVEDD	7.1 ± 11.5	3.8 ± 0.8*
LVEDDSDS	2.23 ± 4.4	1.1 ± 2.8*
FS	0.32 ± 0.08	0.34 ± 0.08
25 OHD ng/ml	6.7 ± 5	2.6.5 ± 17*
calcium nmol/l	1.9 ± 0.45	2.27 ± 0.15*
PTH	152 ± 151	43 ± 34*
QTc msec	375 ± 39	386 ± 36

*$p < 0.05$ after versus before vitamin D therapy.

Discussion Improvement of the recorded changes in the heart rate and LVEDD after VD therapy implements an important role of VD through its effect on the concentration of the extracellular calcium ion that could modify the strength of the myocardial contraction through excitation-contraction coupling.

Conclusion These data strongly indicate that the maintenance of an optimal vitamin D status may be a promising approach for the prevention and/or therapy of myocardial diseases and in countries with high prevalence of VDD, vitamin D supplementation can prevent this risk of cardiac dysfunction.

PO-0047 WITHDRAWN

PO-0048 WITHDRAWN

PO-0049 CORONARY ARTERY ANOMALIES IN PATIENTS UNDERGOING ARTERIAL SWITCH OPERATION

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10.1136/archdischild-2014-307384.722

Background and aims The arterial switch operation (ASO) is the surgical standard of care for repair of transposition of the great arteries (D-TGA). Until recently, anatomical variations of the coronary arteries, especially the intramural course of a single coronary artery, were considered contraindications for ASO. Transfer of the coronary arteries may be a surgical challenge in these cases increasing the risk of (sub-)acute coronary artery occlusions.

Methods We report our management of two exemplary cases of D-TGA with coronary artery anomalies:

(1) Single coronary ostium of RCA, Cx and LAD originating from aortic sinus II and an intramural course of the proximal LAD.

(2) Side-by-side position of the great arteries, RCA and LAD originating from sinus I and Cx from sinus II.

Results Both neonates successfully underwent ASO with transfer of the coronary arteries.

During the post-operative period, patient 1 was diagnosed with a subacute anteroseptal ischemia and was then managed conservatively. Follow-up echocardiogram at 12 months demonstrated satisfactory left and good right ventricular function.

The postsurgical course of patient 2 was uneventful with good biventricular function at follow-up.

Conclusions Anatomical variations of the coronary arteries require adaptions of the surgical technique of coronary artery transfer. Nowadays, ASO is even possible in patients with D-TGA and complex coronary anomalies. The long-term management, however, has to be evaluated, e.g. regarding the need for coronary artery re-surgery.

PO-0050 ABSENT PULMONARY VALVE IN A PATIENT WITH ALAGILLE SYNDROME

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10.1136/archdischild-2014-307384.723

Background and aims Absent pulmonary valve (APV) is a rare congenital defect of the right ventricular outflow tract (RVOT). The genetics of APV are unknown. However, mutations in the NOTCH-signalling pathway have been associated with RVOT obstruction. Mutations in the *JAG-1* gene cause a broad spectrum of symptoms, ranging from an isolated heart defect to the complete clinical features of Alagille syndrome.

We present the case of a 14 month-old girl with APV and a family history of Alagille syndrome.

Methods Pulmonary stenosis and a large ventricular septal defect (VSD) had been diagnosed prenatally. Postnatal echocardiogram revealed an APV, pulmonary stenosis, a large sub-aortal VSD, and right ventricular hypertrophy.

Genetic analysis of the *JAG-1* gene showed a frame-shift-mutation in exon 12 of the *JAG-1* gene that had not been described before.

The patient underwent corrective heart surgery at 9 months of age. The VSD and the native pulmonary artery orifice were closed surgically. A valved xenograft conduit (Contegra®, 14 mm) was implanted between the RV and the pulmonary artery.

Results The last follow-up echocardiogram at 12 months of age demonstrated a sufficient pulmonary valve, closed VSD, resolving right ventricular hypertrophy and good biventricular function.

Conclusions Genetic mutations affecting the NOTCH-signalling pathway can be involved in the pathogenesis of APV. Therefore it is essential to characterise patients with NOTCH-signalling pathway defects by their clinical features and by the underlying mutations in order to develop future therapeutic approaches of APV.

Endocrinology/Diabetes/Metabolism

PO-0051 ZINC AND COPPER DISORDERS IN CHILDREN WITH DIABETES TYPE 1

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10.1136/archdischild-2014-307384.724