

**Conclusion** The most prevalent abnormal geometry type was eccentric hypertrophy that was present in 9,6% of controls but in 41,3% of obese children ( $p < 0.05$ ).

PO-0040 WITHDRAWN

PO-0041 **IS ECHOCARDIOGRAPHY AN ESSENTIAL TOOL IN MECHANICALLY VENTILATED CRITICALLY ILL CHILDREN: WHERE DO WE STAND IN TRAINING OF PAEDIATRIC INTENSIVISTS TO PERFORM BEDSIDE ECHOCARDIOGRAPHY?**

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**Objective** To determine the indication and need for echocardiographic assessment according to clinical severity of patients and therapeutic interventions performed after echocardiographic evaluation in critically ill children.

**Design** Prospective observational study.

**Setting** 14-bed, tertiary care Paediatric Intensive Care Unit (PICU).

**Methods** The PICU is a multidisciplinary referral centre. A total of 140 children; 75 mechanically ventilated (MV) and 65 spontaneously breathing (SB) who were admitted to our unit consecutively from March to August 2013 were included into the study. The indication for echocardiography and therapeutic interventions performed after cardiology consultation were recorded. For the estimation of severity of disease the paediatric risk of mortality score (PRISM) III was determined. The correlation between Prism III score and the need for echocardiographic evaluations were analysed.

**Results** Ages of all patients were between 45 days to 18 years. The male-female ratio was 1.33. Echocardiography was indicated in 88% ( $n = 66$ ) of MV and 46.2% ( $n = 30$ ) of SB patients. In 35.4% ( $n = 34$ ) of patients who underwent echocardiographic evaluation no definitive change occurred in treatment strategy while in 64.6% ( $n = 62$ ) of patients decisive/supplemental information was obtained. Echocardiographic evaluation was necessary in MV children ( $p < 0.001$ ) when compared with SB group and there was a positive correlation between Prism III score and the need for echocardiographic evaluation.

**Conclusion** Echocardiographic evaluation is essential in mechanically ventilated children and the need for echocardiographic assessment increases according to clinical severity of patients. Basic training for intensivists in this technique is challenging and needs to be developed and supported in critically ill children.

PO-0042 **PREVALENCE AND OUTCOME OF DIASTOLIC DYSFUNCTION IN CHILDREN WITH FLUID REFRACTORY SEPTIC SHOCK – A PROSPECTIVE OBSERVATIONAL STUDY**

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**Background and aims** Severe sepsis is often characterised by disturbances in cardiac function, popularly known as sepsis induced

myocardial dysfunction (SIMD). SIMD may be systolic (SD), diastolic (DD) or global. Diastolic dysfunction has been infrequently reported in only a few studies on adult patients till date. Detecting DD early in the course has important therapeutic and prognostic implications in the management of septic shock. Our aim was therefore, to determine the prevalence and outcome of DD in children with septic shock and to generate evidence for possible early clinical predictors of DD in such patients.

**Methods** Consecutive children  $\leq 17$  years of age with fluid refractory septic shock and not on mechanical ventilation admitted to our ICU from June 2011 to August 2012 were included. DD was defined as an abnormal 'E', *mitral annulus velocity* on tissue Doppler imaging at admission.

**Results** A total of 56 children were included. The prevalence of DD was 41.1% (95% CI: 27.8 to 54.4) and the mortality rate was 43% in those with DD. On univariable analysis of possible early predictors of DD, we observed that children with DD tended to have higher mean CVP (13 vs. 6;  $p < 0.0001$ ) and greater positivity for cTnT (70% vs. 36%,  $p = 0.01$ ) as compared to others. On multivariable analysis, only an increased CVP remained significant (adjOR: 1.6; 95% CI: 1.12, 2.14,  $p = 0.008$ ).

**Conclusion** Diastolic dysfunction is common in children with fluid refractory septic shock and immediate outcome may be poorer in such patients. Increased CVP after initial fluid resuscitation may be an early indicator of DD and warrant urgent bedside echocardiography.

PO-0043 **CLASSIC INFANTILE-ONSET POMPE DISEASE: A STUDY OF 8 CASES**

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**Introduction** Classic infantile pompe disease is an autosomal recessive inherited disorder caused by a deficiency of alpha 1–4 glucosidase, resulting in accumulation of glycogen in skeletal muscle and heart. It's characterised by hypotonia, cardiomegaly and hypertrophic cardiomyopathy, feeding difficulties and respiratory distress, all patients died in the first year of life.

**Patients and methods** A retrospective study of cases of classic infantile pompe disease collected in the Department of Paediatrics, Hospital Hedi Chaker, Sfax over a period of 7 years (2008–2013).

**Results** During the study period we collected 8 cases of the disease. The average age at diagnosis was 3 months and 6 days.

All patients were eutrophic. Hypotonia was noted in all cases, Respiratory distress in seven cases, cyanosis in 4 cases and a systolic murmur in 5 cases. Hepatomegaly was constant, macroglossia was noted in one patient. Chest radiography showed cardiomegaly in all cases. Electrocardiogram, showed electrical signs of left ventricular hypertrophy, wide QRS complex, a shortening of the PR interval was seen in 4 cases. Echocardiography confirmed diagnosis of hypertrophic cardiomyopathy. The dosage of acid maltase activity practiced in 5 patients, showed no enzyme activity. The genetic study of 4 patients showed a mutation in the homozygous state of the GAA gene. Treatment was purely symptomatic involving propranolol, oxygen and antibiotics. All patients died.

**Conclusion** The fatal evolution of the classic infantile form of Pompe disease is being changed by the advent of enzyme

replacement therapy which 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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**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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**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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**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 ( $3.2 \pm 4.4$  to  $1.1 \pm 2.8$ ) and slowing of the heart rate (from  $101 \pm 34$  to  $94.7 \pm 30$ /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 ( $r = -0.2$ ,  $p = 0.2$ ).