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PO-0041 WITHDRAWN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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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 echocardiographic evaluation in critically ill children.

Design Prospective observational study.

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

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.

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

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 evaluations were analysed.

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-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-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, macrognosia 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. However, the need to establish early diagnosis and treatment when enzyme replacement therapy is not available continues to be a challenge.