Echocardiography

A case of congenital myopathy with severely hypertrophied ventricles

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Background: Congenital myopathies comprise a heterogeneous group of muscle disorders that usually present in the early life initially as hypotonia and weakness at birth and later as motor delay, muscle and respiratory weakness. They are caused by genetic defects in contractile proteins with characteristic histological abnormalities and show no evidence of regeneration and degeneration in contrast to muscle dystrophies which are caused by defects in the anchoring/supporting membrane proteins. Primary cardiac involvement is rare though in addition to arrhythmias both DCM and hypertrophic cardiomyopathy have been reported. Rod like structures may be seen in cardiac myocytes and conduction tissue in addition to striated muscles in nemaline myopathy, whereas centronuclear myopathies show evidence of dilated cardiomyopathy.

Case: A 1-year-old child with recurrent episodes of LRTI and generalized hypotonia presented with severe respiratory distress. The child was having tachycardia with gallop and Echo revealed severely hypertrophied ventricles with almost complete obstruction of cavity. Such severe hypertrophy of ventricles is rarely seen in hypertrophic cardiomyopathy associated with congenital myopathies.

Use of endothelial function, strain, strain rate and tissue velocity imaging for early detection of cardiovascular involvement in patients with beta thalassemia

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Background and objective: Increased iron stores have been linked to the risk of cardiovascular disease in thalassemia patients. Cardiac failure due to due to iron overload remains the most common cause of death in these patients. Structural alterations leading to functional alterations of vascular endothelial cells have been described. Tissue velocity and strain imaging might prove useful in early detection of the regional myocardial dysfunction in these patients even when the conventional echo parameters as the dimensions, fractional shortening (FS) and the ejection fraction (EF) remain within the normal range, so as to improve the prognosis with earlier aggressive chelation therapy. Simultaneously the alteration of the endothelial function was also assessed in vivo.

Materials and methods: The study included two age matched groups: 30 thalassemia patients (23 boys, 7 girls: 12.4 ± 5.2 years) and 20 controls (13 boys, 7 female: 12.5 ± 5.2 years) enrolled between June 2014 and June 2015. Conventional echo-Doppler measures of LV dimensions and function were obtained. TVI measures including systolic and diastolic myocardial velocities (Sm, Em, Am and Em/Am) of the basal lateral and septal LV segments. Systolic strain (SI) and strain rate (SRI) values were measured in the basal and mid segments of the LV, RV and septum. These values were compared between the two groups. The brachial artery endothelial function was assessed by vascular response to reactive hyperemia (flow-mediated dilation [FMD]) and sublingual glyceryl trinitrate (nitrate mediated dilatation [NMD]) to check the endothelial dependent and independent responses respectively. The carotid intima-media thickness (CIMT) was also compared to see structural alterations in addition to the endothelial function.

Results: The conventional echo and the Doppler measures like diastolic and the systolic dimensions, FS, EF etc. were comparable between the two groups (p = NS) except LV mass index (169.45 ± 61.145 vs 104.66 ± 24.4, p = 0.009) which was higher in the thalassemia patients. The Sm had a trend towards lower values (8.7 ± 1.1 vs 10.85 ± 1.13, p = 0.248) and Am had a trend towards higher values (12.85 ± 1.84 vs7.18 ± 1.84, p = 0.903) in the patients. The Em (10.12 ± 1.16 vs 17.9 ± 2.11, p = 0.002) & Em/Am (0.811 ± 0.192 vs 2.06 ± 0.62, p < 0.001) were significantly lower in the patients. Basal lateral LV SI (−19.5 ± 3.98 vs 24.19 ± 1.81, p = 0.003), SRI (−0.82 ± 0.53 vs 0.867 ± 0.38, p = 0.38) mid LV SI (−19.07 ± 3.98 vs −25.56 ± 2.62, p = 0.042) & SRI (−0.671 ± 0.23 vs 1.07 ± 0.417, p = 0.029) were significantly lower for the patients though the similar values for the RV, even though less than the control group, did not reach statistical significance. Although the CIMT (0.366 ± 0.134 vs 0.357 ± 0.112, p = 0.304) was comparable, the FMD was impaired (7.57 ± 3.16 vs 18.08 ± 1.9, p = 0.018) inspite of the preserved NMD (16.6 ± 2.1 vs 17.4 ± 3.2, p = 0.56) implying endothelial dysfunction.

Conclusions: Endothelial dysfunction, and LV hypertrophy occur in patients with thalassemia, which may result in reduction of mechanical efficiency of the heart. These patients have regional systolic and diastolic dysfunction. TVI and the newer modality of SI and SRI are promising tools for quantitative assessment of regional myocardial function detecting myocardial involvement earlier than the conventional echo parameters.

Assessing change of right ventricular longitudinal strain in mitral stenosis patients undergoing percutaneous transvenous mitral commissurotomy

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