REVIEW ARTICLES

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Cardiomyopathy secondary to Duchenne muscular dystrophy in children

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Abstract

Background: Cardiomyopathy (CM) associated with Duchenne muscular dystrophy (DMD) is a commonly recognized appearance of this neuromuscular disease, significantly increased morbidity and mortality, as well as the necessity for cardiological management. CM in DMD is defined by left ventricular (LV) systolic dysfunction and both atrial and ventricular dysrhythmias and is associated with higher mortality than other cases of pediatric dilated CMs. Notwithstanding the high rate of cardiac involvement, patients are usually asymptomatic despite significant LV dysfunction, because of likely poor mobility that masks the usual heart failure (HF) symptoms. Also, imagistic predictors are provided to be very helpful in defining early LV dysfunction, especially electrocardiogram and cardiac imaging (transthoracic echocardiography, speckle-tracking, cardiac magnetic resonance) are used to detect the onset and progression of dilated cardiomyopathy (DCM) in DMD.
Conclusions: As most DMD patients are asymptomatic for a long time of their life, so identifying predictors of HF is crucial to support these patients. Ventricular dysfunction based on the ejection fraction (EF) measurement helps to choose therapy. In the case of early DCM (LVEF≥50%) the great purpose is to prevent ventricular dysfunction incipience with first-line HF therapy with Angiotensin-converting-enzyme inhibitors (ACE-I) or angiotensin receptor blockers (ARBs). Current guidelines recommend the use of conventional HF medication in case of disease progression and DCM with Mid-Range Reduction of LV EF (40-49%). The therapeutic approach for patients with DCM and severe ventricular dysfunction (<40%) has been studied less profoundly and contemporary guidelines recommend all drugs used for HF treatment.
Key words: Duchenne muscular dystrophy, cardiomyopathy, heart failure, neuromuscular.

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