Utility of echocardiography and electrocardiography for the evaluation of cardiac status in Duchenne muscular dystrophy: validation with cardiac magnetic resonance imaging

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Aims and Background Dilated cardiomyopathy and related problems are inevitable consequences of Duchenne muscular dystrophy (DMD). Cardiac magnetic resonance (CMR) imaging is a gold standard to evaluate ventricular function and myocardial fibrosis in DMD, as many of the patients have poor acoustic window for echocardiography. The aim of this study was to investigate whether echocardiography or electrocardiography can determine cardiac functional status or the degree of myocardial fibrosis in comparison to CMR. Methods Medical recordings of 20 patients with DMD (aged 17.57 ± 5.22 years) were retrospectively analyzed. We evaluated CMR data and compared with echocardiographic and electrocardiographic data conducted within 1 month of the CMR examination. Results Left ventricular ejection fraction on CMR correlated significantly with z-value of peak early diastolic tissue velocity at medial mitral annulus (e'(z); r = 0.566, P = 0.044) and showed a negative correlation with z-value of mitral inflow peak E-to-septal tissue velocities (E/e'(z); r = -0.660, p = 0.014) on echocardiography. There was significant negative correlation between left ventricular ejection fraction on CMR and tissue Doppler imaging–derived left ventricular performance index by echocardiography (r = -0.523, p = 0.046). Indexed left ventricular end-diastolic dimension on CMR showed a tendency of negative correlation with left ventricular ejection fraction on echocardiography, but the statistical significance was low (r = -0.500, p = 0.057). There was positive correlation between amount of myocardial fibrosis (%) assessed by late gadolinium enhancement (LGE) on CMR and frequency of fragmented QRS complexes on electrocardiography (r = 0.614, p = 0.014). The LGE area on CMR showed no significant correlation with other functional indices on both echocardiography and CMR. Conclusion There is an association between severity of myocardial dysfunction defined by left ventricular ejection fraction on CMR and e', E/e', and tissue Doppler imaging–derived left ventricular performance index on echocardiography. We found that fragmented QRS is related to the amount of myocardial fibrosis defined by LGE CMR. Further investigation should be conducted to determine if echocardiography and electrocardiography data can reflect cardiac findings by CMR in DMD patients.