

Myocardial strain using speckle tracking echocardiography in pediatric Duchenne Muscular Dystrophy patients

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Introduction

Duchenne muscular dystrophy (DMD) is an inherited X-linked disorder, with an incidence of 1:3500 male births. Cardiomyopathy is the leading cause of death. Cardiac MRI (CMR) with late gadolinium enhancement (LGE) is an important tool to analyze myocardial fibrosis, but early changes could be detected analyzing myocardial strain by echocardiography.

The aim of this study is to analyze the myocardial strain pattern in our pediatric DMD population and to correlate these parameters with global LV function and CMR data.

Methods

We analyzed 17 patients diagnosed with DMD. We obtained epidemiological, clinical, CMR and echocardiography data (LVEF) including speckle tracking offline analysis with QLAB® , Philips.

Results.

We collected data from 17 pediatric DMD patients. Mean age 13,3y (median 14, IQR 3). All of them received treatment with oral corticoids. Except in the younger case (5yo), the other patients were not still capable of autonomous deambulation.

Concerning global LV function analysis 7 patients showed LV systolic dysfunction (LVEF <55%).

Three of these patients showed LGE in the CMR (inferolateral regions). All 7 patients showed regional impairment in the circumferential strain. Longitudinal strain was altered only in 2 patients, with LVEF less than 40%.

The other 10 patients showed normal LVEF values. We detected LGE in the CMR in 3 patients (same inferolateral regions and anteroseptal regions). In 6 of these 10 patients, we detected an abnormal behaviour of myocardial deformation in the speckle tracking analysis: abnormal deformation in basal and mid inferolateral regions (two of these 6 patients, only mid inferolateral region abnormalities). Two of the 3 patients with LGE showed impairment of these regional abnormalities.

Conclusion

Circumferential strain is altered in DMD patients, including in those with normal LVEF values. The majority of patients with preserved LVEF and LGE in the CMR showed abnormalities in the regional circumferential strain, specially inferolateral regions. Circumferential strain by speckle tracking echocardiography in pediatric DMD patients could be a marker of myocardial fibrosis.