Mechanical dispersion by speckle tracking echocardiography in fabry disease

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Topic(s):
Tissue Doppler, Speckle Tracking and Strain Imaging

Citation:
Background: Fabry disease is a rare X-linked storage disorder caused by a deficiency of the lysosomal enzyme a-galactosidase A and generally causes multi-organ dysfunction. Heart disease is the main cause of death, due to severe left ventricular (LV) systolic dysfunction and sudden death. In several heart diseases, the LV systolic dysfunction and ventricular arrhythmias are associated with mechanical dispersion (MD). The presence of MD in patients with FD has not been studied yet. In this cross-sectional study, we investigated the prevalence of MD in patients with FD.

Methods: Complete echocardiographic and speckle tracking echocardiographic (STE) data were collected. MD is an index of inter-segmental discoordination of contraction which has been used to quantify LV dyssynchrony and was defined as the standard deviation (SD) of time to peak negative strain in 17 left ventricular segments. Patients were divided into two groups according to whether or not they had left ventricular hypertrophy (LVH). MD was defined as an SD > 49 msec.

Results: We studied 108 patients with FD, 24 patients (22%) were excluded due to inadequate imaging quality or presence of comorbidities, so the final study population consisted of 84 patients (mean age 33.3±14.6 years, 60.7% women). LVH in FD appears at older ages than in patients without LVH (48±12.5 y/o vs 27.8±11.1 y/o, p < 0.0001). Patients with FD without LVH (Group I) showed normal global longitudinal peak strain (GLPS) (21.2±2.5%) and no MD (32.7±8.8 msec). In Group II (n=23) patients with FD with LVH, 17 (73.9%) had MD > 49 msec prolonged mechanical dispersion (73.3±20.7 msec) and reduced GLPS (13.6±4.0%). MD was more pronounced in Fabry patients with LVH than in patients without LVH (63.4±24.7 msec vs. 32.7±8.8 msec, p < 0.0001). GLPS was lower in Fabry patients with LVH than in patients without LVH (15.3±4.7% vs 21.2±2.5%, p < 0.0001).

Conclusions: To our knowledge, this is the first study to demonstrate the prevalence of mechanical dispersion in patients with FD. Mechanical dispersion was seen in 73.9% of patients with FD with LVH. This dyssynchrony should be taken into account in patients who develop heart failure or life-threatening ventricular tachyarrhythmias.
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FD without LVH (normal MD)

FD with LVH (prolonged MD)