Electromechanical discoordination is present in patients with Duchenne Muscular Dystrophy independent of tissue fibrosis.

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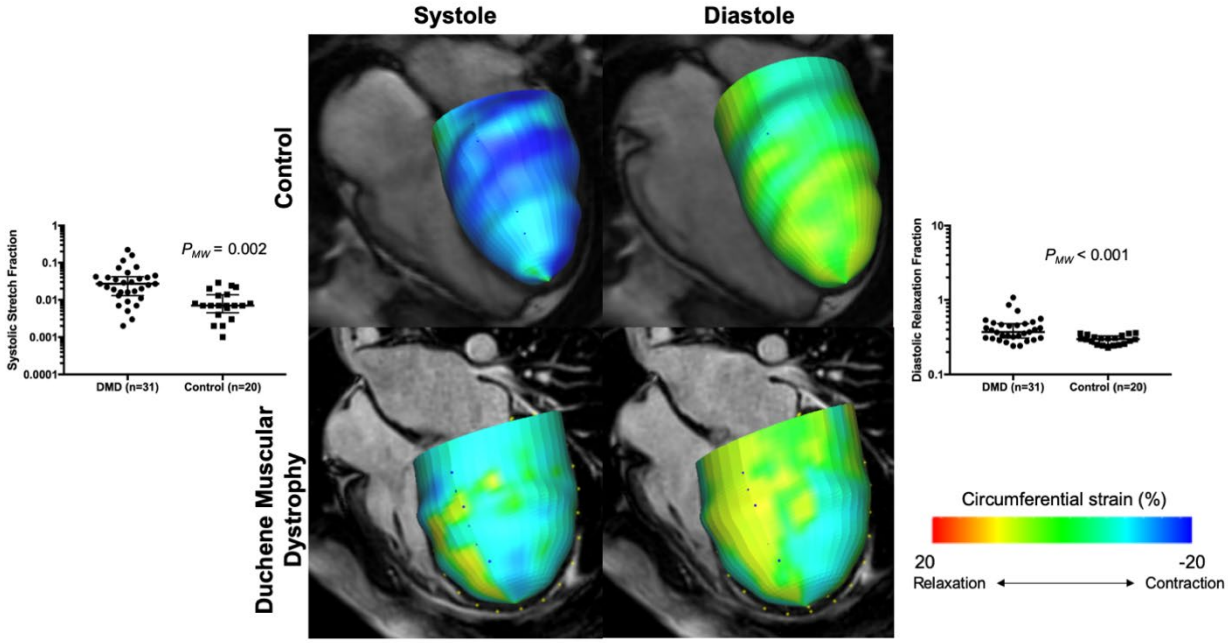
Introduction:
Progressive ventricular dysfunction is a cardinal symptom in Duchenne Muscular Dystrophy (DMD). Some of the earliest signs of cardiomyopathy in DMD are myocardial fibrotic deposition and LV strain defects. Electromechanical discoordination, as measured by Systolic Stretch Fraction (SSF) and Diastolic Relaxation Fraction (DRF), has been shown to be a sensitive marker of ventricular dysfunction. The presence of this discoordination in relation to fibrotic deposition in DMD has yet to be elucidated.

Hypothesis:
Patients with DMD will have abnormal SSF and DRF on cardiac MRI (CMR) tissue tracking analysis and associated with fibrotic deposition.

Methods:
Patients with DMD (n=31)(mean age: 14 ± 4 yrs) and controls (n=20) (mean age: 15 ± 3 yrs) underwent CMR for volumetric and functional analysis as well as Gadolinium (Gd) enhancement to evaluate the presence of fibrosis. Circumferential strain and strain rate indices from each segment were used to calculate electromechanical discoordination. Strain rate data was used to calculate SSF and DRF.

Results:
Patients with DMD showed increased median LV SSF compared to controls [0.027 (IQR: 0.015-0.041) vs 0.007 (IQR:0.005-0.013), $P = 0.002$] as well as increased median LV DRF [0.371 (IQR: 0.310-0.473) vs 0.300 (IQR: 0.264-0.325), $P < 0.001$] (Figure). When comparing Gd(+) (n=14) vs Gd(-) (n=17) DMD patients, there was no difference between groups in either SSF [0.027 (IQR: 0.016-0.042) vs 0.026 (IQR: 0.008-0.040), $P= 0.929$] or DRF [0.371 (IQR: 0.309-0.537) vs 0.379 (IQR: 0.322-0.464), $P= 0.931$]. The SSF was associated with ESVi ($R=0.71$, $P<0.001$), EDVi ($R=0.65$, $P< 0.001$) and inversely associated with EF ($R=-0.63$, $P<0.001$).

Conclusion:
Patients with DMD showed increased levels of LV electromechanical discoordination independent of qualitative presence of fibrosis noted by Gd enhancement. This allows speculation that changes in electromechanical discoordination may precede visible fibrotic change in DMD.


Systole
Diastole
Control
Duchene Muscular Dystrophy
Systolic Stretch Fraction
1
0.1
0.01
0.001
0.0001
DMD (n=31)
Control (n=20)
P_{MW} = 0.002
Diastolic Relaxation Fraction
10
1
0.1
DMD (n=31)
Control (n=20)
P_{MW} < 0.001
Circumferential strain (%)
20
-20
Relaxation
Contraction