The aim of this study was to examine the left ventricular (LV) function of patients with DMD using MRI-derived ventricular electro-mechanical discoordination indices including: 1) systolic stretch fraction (SSF) and 2) diastolic relaxation fraction (DRF).

Background

- Progressive ventricular dysfunction is a cardinal symptom in Duchenne Muscular Dystrophy (DMD).
- The aim of this study was to examine the left ventricular (LV) function of patients with DMD using MRI-derived ventricular electro-mechanical discoordination indices including: 1) systolic stretch fraction (SSF) and 2) diastolic relaxation fraction (DRF).

Methods

- Adolescents with DMD (n=31) and healthy controls (n=20) of similar age underwent MRI for standard volumetric and functional analysis. Segment-specific circumferential strain and strain rate indices were evaluated along with standard mechanical dyssynchrony.
- Patients also underwent MRI with Gadolinium enhancement to evaluate for the presence of fibrosis.

Results

- There were no group differences noted between DMD patients that underwent MRI with and without gadolinium enhancement.
- Future studies will involve echocardiographic evaluation of SSF and DRF for MRI validation and for the assessment of clinical prognosis.

CONCLUSION

- Patients with DMD showed increased levels of LV electromechanical discoordination compared to controls.
- This difference was independent of qualitative presence of fibrosis noted by Gadolinium enhancement.
- This allows speculation that changes in electromechanical discoordination may precede visible fibrotic change in DMD.
- Future studies will involve echocardiographic evaluation of SSF and DRF for MRI validation and for the assessment of clinical prognosis.

Table 1. MRI Hemodynamics (DMD vs. Control)

<table>
<thead>
<tr>
<th>Age (yrs)</th>
<th>BSA (m²)</th>
<th>Sex (Female)</th>
<th>SSF</th>
<th>LV EDV (mL/m²)</th>
<th>LV ESV (mL/m²)</th>
<th>LV CI (L/min/m²)</th>
<th>LV EF (%)</th>
<th>LV CO (L/min)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Control (n=20)</td>
<td>DMD (n=31)</td>
<td>Control (n=20)</td>
<td>DMD (n=31)</td>
<td>Control (n=20)</td>
<td>DMD (n=31)</td>
<td>Control (n=20)</td>
<td>DMD (n=31)</td>
<td>Control (n=20)</td>
</tr>
<tr>
<td>14.79 ± 3.10</td>
<td>1.42 ± 0.31</td>
<td>55%</td>
<td>0.007 (0.005 - 0.013)</td>
<td>0.03 (0.02 - 0.04)</td>
<td>0.002</td>
<td>0.3 (0.26 - 0.33)</td>
<td>0.37 (0.31 - 0.47)</td>
<td>0.001</td>
</tr>
</tbody>
</table>

Table 2. MRI Hemodynamics (Gd- vs. Gd +)

<table>
<thead>
<tr>
<th>Age (yrs)</th>
<th>BSA (m²)</th>
<th>Sex (Female)</th>
<th>SSF</th>
<th>LV EDV (mL/m²)</th>
<th>LV ESV (mL/m²)</th>
<th>LV CI (L/min/m²)</th>
<th>LV EF (%)</th>
<th>LV CO (L/min)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Control (n=14)</td>
<td>DMD (n=17)</td>
<td>Control (n=14)</td>
<td>DMD (n=17)</td>
<td>Control (n=14)</td>
<td>DMD (n=17)</td>
<td>Control (n=14)</td>
<td>DMD (n=17)</td>
<td>Control (n=14)</td>
</tr>
<tr>
<td>12.3 ± 3.6</td>
<td>1.23 ± 0.31</td>
<td>55%</td>
<td>0.03 (0.01 – 0.04)</td>
<td>0.022 (0.016 – 0.042)</td>
<td>0.703</td>
<td>14.3 ± 3.6</td>
<td>1.47 ± 0.38</td>
<td>0.082</td>
</tr>
</tbody>
</table>

All values are reported as means with corresponding 95% confidence interval. SSF, systolic stretch fraction; DRF, diastolic relaxation fraction; LV EDV, left ventricular end diastolic volume; LV ESV, left ventricular end systolic volume; LV SV, left ventricular stroke volume; LV CI, left ventricular cardiac index; LV EF, left ventricular ejection fraction; LV CO, left ventricular cardiac output.

Figure 1. Algorithm for the LV discoordination indices: systolic stretch fraction (SSF) and diastolic relaxation fraction (DRF). From the segment specific strain rate indices were separated into positive and negative components to calculate the ratio of myocardial relaxation and contraction during the ejection and filling phases.

Figure 2. LV endocardial and epicardial segmental propagated 3D circumferential strain model comparing patients with DMD and controls in systole and diastole.

Figure 3. Graphical representation of the LV electromechanical discoordination results. A) Systolic stretch fraction (SSF) was elevated in patients with DMD, B) as was diastolic relaxation fraction (DRF). There were no group differences noted between DMD patients that were Gd- or Gd+ for C) SSF or D) DRF.