Cardiac MRI in Muscular Dystrophies

Daniel Wallihan MD
Muscular Dystrophy

Group of inherited muscular disorders characterized by progressive muscle weakness/wasting

**Dystrophinopathies** – absent, abnormally low levels or abnormal configuration of the dystrophin protein in the muscle cell membrane

- Duchenne MD
- Becker MD
- X-linked cardiomyopathy
Duchenne Muscular Dystrophy

X-linked inheritance

Incidence of ~1 in 3500 male newborns

Progressive weakness of trunk and extremity skeletal musculature starting in early childhood

Life expectancy 25-30 years
  - Respiratory
  - Cardiac

- Walking problems
- Wheel chair - skeletal deformity
- Very limited use of arms
- Ventilation at night
- Ventilation 24hrs
- Death
12 years old

14 years old
Cardiac Involvement

- Absent or abnormal dystrophin
- Unstable cell membrane
- Myocardial cells more susceptible to injury during normal function
- Micro-ischemia?
- Inflammation/Necrosis
- FIBROSIS
CMR: Functional Imaging

- Cardiac output/index
- Ejection fraction
- End diastolic volume
CMR: Strain Imaging

• Strain = change in length of a segment of myocardium in systole relative to its resting length
  – Harmonic phase imaging (HARP)
  – Displacement Encoding with Stimulated Echoes (DENSE)
  – Feature-tracking

• Circumferential, radial, longitudinal
CMR: Strain Imaging
Circumferential Strain Analysis Identifies Strata of Cardiomyopathy in Duchenne Muscular Dystrophy

A Cardiac Magnetic Resonance Tagging Study

Kan N. Hor, MD,† Janaka Wansapura, MD,‡ Larry W. Markham, MD,§ Wojciech Mazur, MD,§ Linda H. Cripe, MD,∗ Robert Fleck, MD,† D. Woodrow Benson, MD, PrtD,∗
William M. Gottliebson, MD∗
CMR: Tissue Characterization

Fibrosis imaging - Late Gadolinium Enhancement (LGE)

LGE ≠ Systolic Dysfunction

<table>
<thead>
<tr>
<th>Parameters</th>
<th># Patients</th>
<th>LGE negative</th>
<th>LGE positive</th>
<th>NPS with LGE</th>
<th>Odds ratio (95% CI)</th>
<th>P value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age &lt; 10 years</td>
<td>83</td>
<td>69 (83%)</td>
<td>14 (17%)</td>
<td>4.4 ± 3.0</td>
<td>1.0</td>
<td>&lt; 0.006</td>
</tr>
<tr>
<td>Age 10–15 years</td>
<td>149</td>
<td>98 (66%)</td>
<td>52 (34%)</td>
<td>4.8 ± 2.5</td>
<td>2.6 (1.3-5.0)</td>
<td>&lt; 0.001</td>
</tr>
<tr>
<td>Age &gt; 15 years</td>
<td>82</td>
<td>34 (41%)</td>
<td>48 (59%)</td>
<td>5.9 ± 3.3</td>
<td>7.0 (3.4-14.3)</td>
<td>&lt; 0.0001</td>
</tr>
<tr>
<td>LVEF ≥ 55%</td>
<td>277</td>
<td>195 (70%)</td>
<td>82 (30%)</td>
<td>4.1 ± 2.4</td>
<td>1.0</td>
<td></td>
</tr>
<tr>
<td>LVEF &lt; 55%</td>
<td>37</td>
<td>6 (16%)</td>
<td>31 (84%)</td>
<td>7.8 ± 3.4</td>
<td>12.3 (4.9-30.6)</td>
<td>&lt; 0.0001</td>
</tr>
</tbody>
</table>

Hor et al. JCMR 2013
CMR: Tissue Characterization

Hor et al. JCMR 2013
CMR: Tissue Characterization

2006
EF = 63%
EDV = 67 ml/m²

2009
EF = 52%
EDV = 82 ml/m²

2013
EF = 39%
EDV = 140 ml/m²
Why CMR?

- Early initiation of treatment can delay onset and halt progression of LV systolic dysfunction
- Decreased LVEF by echo or standard CMR indicates onset of cardiomyopathy
- Myocardial strain and fibrosis imaging can serve as an earlier, more sensitive indicator for deciding when to start cardioprotective treatment