High-resolution MR Imaging Of Bone-Muscle-Fat
In Glucocorticoid Treated Boys With Duchenne Muscular Dystrophy: Results from the ScOT-DMD study

S Joseph 1,2, H Elsharkasi 3, J Foster 4, I Horrocks 2, M DiMarco 5, C McComb 3, SF Ahmed 1, SC Wong 1
Developmental Endocrinology Research Group, Department of Paediatric Endocrinology, The Royal Hospital for Children, Glasgow, UK;
Paediatric Neurosciences Research Group, Department of Paediatric Neurolgy The Royal Hospital for Children, Glasgow, UK; Clinical Physics, NHS Greater Glasgow & Clyde, Glasgow; West of Scotland Genetic Services, Queen Elizabeth University Hospital, Glasgow, UK

Background

The pathophysiological mechanism of skeletal fragility in Duchenne Muscular Dystrophy (DMD) is unclear.

Objectives

To compare trabecular bone microarchitecture, cortical geometry, muscle inflammation and fat fraction at distal femur and vertebral bone marrow adiposity (BMA) in DMD and controls.

Methods

Bone-muscle and muscle fat fraction (FF) were assessed using 3T MRI and quantitative Dixon technique. BMA was assessed using 1H-MRS. Results expressed as median (range). Cortical parameters were compared following adjustment for femur length, muscle area, and age.

Results

Table 1: Cohort characteristics

<table>
<thead>
<tr>
<th></th>
<th>DMD</th>
<th>Control</th>
<th>p-value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Number of cases</td>
<td>16</td>
<td>25</td>
<td></td>
</tr>
<tr>
<td>Age (years)</td>
<td>11.7 (8.8,18.8)</td>
<td>13.0 (8.1,18.1)</td>
<td>0.694</td>
</tr>
<tr>
<td>Height SDS</td>
<td>-1.5 (-4.9,1.0)</td>
<td>0.5 (0.8,1.6)</td>
<td>0.001</td>
</tr>
<tr>
<td>BMI SDS</td>
<td>2.7 (-1.4, 3.5)</td>
<td>0.0 (-1.0,3.3)</td>
<td>0.004</td>
</tr>
<tr>
<td>Non-ambulant cohort</td>
<td>10/16 (63)</td>
<td>-</td>
<td></td>
</tr>
<tr>
<td>Length of non-ambulant status (years)</td>
<td>2.1 (1.1, 5.3)</td>
<td>-</td>
<td></td>
</tr>
<tr>
<td>GC therapy (n)</td>
<td>16/16 (100)</td>
<td>-</td>
<td></td>
</tr>
<tr>
<td>GC length (years)</td>
<td>5.9 (1.5, 10.5)</td>
<td>-</td>
<td></td>
</tr>
</tbody>
</table>

Comparison of Trabecular Parameters showed significantly lower apparent Bone Volume against Total Volume (appBV/TV) and apparent Trabecular Thickness (app TbTh) in DMD cases

Figure 1: CISS (Constructive Interference in the Steady State) pulse sequence

Comparison of Cortical Bone Parameters showed significantly lower Mean Cortical Thickness and Cortical Area in DMD cases

Figure 2: T1-weighted turbo spin echo (TSE) pulse sequence

Comparison of Muscle Parameters showed significantly higher Fat Fraction and increased Muscle Inflammation

Results continued

Fig 3: 6-point gradient echo Dixon sequence

Muscle Fat Fraction (FF%) is significantly higher and increases with age in DMD cases

Figure 4: T2-weighted multi-echo spin echo pulse sequence

Muscle Inflammation (T2 Tri-exponential model) Is significantly increased and Residual Muscle Contractile Area is significantly reduced in DMD cases

Figure 5: H1- MRS was performed using a PRESS pulse sequence

Bone Marrow Adiposity is significantly increased in DMD cases

Conclusion

High resolution MRI provides novel information on multiple components of musculoskeletal health in DMD.

GC treated boys with DMD have deficits in trabecular microarchitecture, cortical geometry and showed increased bone marrow adiposity.