Introduction

Liver transplantation is the unique treatment for several end-stage diseases.

Familial Amyloidotic Polineuropathy (FAP) is a neurodegenerative disease related with systemic deposition of amyloidotic fiber mainly on peripheral nervous system, clinically translated by an autonomous sensitive-motor neuropathy with severe functional limitations in some cases. The unique treatment for FAP disease is a liver transplant with a very aggressive medication to muscle metabolism and force production(1).

To our knowledge there are no quantitative characterizations of body composition, strength or functional capacity in this population.

Methods

Sixty-four subjects where assigned in 2 groups: 46 patients FAPTx (27 males, 32±8 yrs and 19 females, 37±5 yrs) and eighteen HG (9 males, 34±7 yrs and 9 female, 36±8 yrs).

• Isometric strength of quadriceps was measured using an isokinetic dynamometer (Biodex) (2).
• Body composition was determined by measuring lean mass of dominant lower extremity in a region of interest (thigh) by dual-energy x-ray absorptiometry (QDR-explorer – Hologic, Waltham, MA: Fan bean mode) (3).
• Muscle quality was ascertainment by taking the ratio of strength to muscle mass (4).
• Functional capacity was determined by the number of meters walked on 6MWT(5).

Results and Discussion

HG showed significant higher values than FAPTx patients for (table 1):
• Peak torque (66.3N 25.2N) vs 40.2N 17.6N respectively p=.000)
• Muscle quality (11.8 2.6 vs 7.9 2.7 respectively, p=.000)
• Functional capacity (675.9 109.1m vs 511.4 139.1m respectively, p=.000)

There are no differences between HG and FAPTx for (table 1):
• BMI (23.8 2.2 kg/m2 vs 22.0 3.8 kg/m2 respectively, p=.07)
• Thigh muscle mass (5.5kg 1.2kg vs 5.04kg 1.1kg respectively p=.14)

Negative correlations were observed for FAPTx patients but not for HG between age and peak torque (p=.013; r= -.363) and age and thigh muscle mass (p=.011; r= -.373)(fig.1)

Table 1 – Body composition variables, strength, muscle quality and functional capacity (Mean ± std; Min-Max; p-value for t-test)

<table>
<thead>
<tr>
<th>Variables</th>
<th>Group FAPTx (n=46)</th>
<th>Group HG (n=18)</th>
<th>p-value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age (years)</td>
<td>35±7</td>
<td>35±7</td>
<td>0.681</td>
</tr>
<tr>
<td>Weight (kg)</td>
<td>62.9±12.5</td>
<td>65.6±9.0</td>
<td>0.406</td>
</tr>
<tr>
<td>Height (m)</td>
<td>1.69±0.08</td>
<td>1.53±1.85</td>
<td>0.185</td>
</tr>
<tr>
<td>Body Mass Index (kg/m2)</td>
<td>22.0±3.85</td>
<td>23.8±2.2</td>
<td>0.074</td>
</tr>
<tr>
<td>Functional Capacity (m)</td>
<td>511.4±139.12</td>
<td>525.7±97.2</td>
<td>0.000*</td>
</tr>
<tr>
<td>Peak Torque (Nm)</td>
<td>40.2±17.65</td>
<td>35.2±12.70</td>
<td>0.000*</td>
</tr>
<tr>
<td>Total lean mass (kg)</td>
<td>45.4±18.08</td>
<td>33.6±60.53</td>
<td>0.834</td>
</tr>
<tr>
<td>% total fat mass</td>
<td>22.5±9.23</td>
<td>11.8±39.42</td>
<td>0.102</td>
</tr>
<tr>
<td>Thigh muscle mass (kg)</td>
<td>5.0±1.1</td>
<td>3.9±2.5</td>
<td>0.138</td>
</tr>
<tr>
<td>muscle quality</td>
<td>7.9±2.4</td>
<td>9.1±7.71</td>
<td>0.000*</td>
</tr>
</tbody>
</table>

* Differences between the means of both groups (p<0.05)

Conclusions

• FAP patients have lower functional capacity, strength and muscle quality than HG.
• The differences between groups for muscle quality and peak torque but not thigh muscle mass seems highlight the importance of the neural component of the disease and show also the importance of training the process of force production specially the sensoriromotor component in FAP patients and probably the importance of a strengthening exercise program.
• Further studies are needed to explore training effects on function after transplantation.

References