Do we have a practical method for monitorization of pulmonary functions in non-ambulatory patients with neuromuscular disease?

Gokhan Ayik, Ebru Yalcin, Ebru Kutukcu, Deniz Inal Ince, Senol Bekmez, Muharrem Yazici

Hacettepe University, Ankara, Turkey
• No disclosure!
GFT outcome

• EOS patients should be assessed not only clinically and radiologically but also pulmonologically capacity @
  – The time of diagnosis
  – During the disease process
  – The end of treatment
• The 6-min walk test for ambulatory patients
  – Practical
  – Easily applicable
  – Reproducible standard test
• A similar test for non-ambulatory patients
  – Not described yet!
Aim

• To investigate the applicability of respiratory muscle and cough strength tests (RMCST) in non-ambulatory neuromuscular EOS patients
• To correlate with standard PFT
Methods

• Ten low-tone collapsing spine patients scheduled for deformity surgery
  – 5 SMA2
  – 3 DMD
  – 1 neuropathy
  – 1 CMD

• 9 M, 1 F

• Mean age 12.7(10-17) years
Methods

• Standard PFT tests (Vyntus Spiro, Carefusion, Germany)
  – Large and small airway functions
Methods

- Respiratory muscle and cough strength tests (RMCST)
  - Maximal Inspiratory Pressure-MIP
  - Maximal Expiratory Pressure-MEP
  - Peak Cough Flow-PCF
  - SNIFF
    - Portable mouth pressure device (Micro MPM; Micro Medical Ltd, England)
    - Mini Wright peak flowmeter (Clement Clark International, Edinburgh, UK)

Correlation between RMCST and PFT tests was statistically investigated
## Results

<table>
<thead>
<tr>
<th></th>
<th>FVC (L)</th>
<th>FVC PRED (%)</th>
<th>FEV1 (L)</th>
<th>FEV1 PRED (%)</th>
<th>FEV1/FVC</th>
<th>MEF 25-75 (L)</th>
<th>MEF 25-75 PRED (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>1,355 (0.44–1.99)</td>
<td>52.6 (23–95)</td>
<td>1.194</td>
<td>55.5 (22–99)</td>
<td>88.547</td>
<td>1,525</td>
<td>59.9 (14–116)</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th></th>
<th>MIP (cmH2O)</th>
<th>MEP (cmH2O)</th>
<th>PCF (ml/mn)</th>
<th>SNIFF (cmH2O)</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>48.77 (23–85)</td>
<td>29.8 (13–43)</td>
<td>186 (110–270)</td>
<td>51 (24–82)</td>
</tr>
</tbody>
</table>
Results

<table>
<thead>
<tr>
<th></th>
<th>MIP</th>
<th>MEP</th>
<th>PCF</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>FVC</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Sig. (2 tailed)</td>
<td>0.015</td>
<td>0.032</td>
<td>0.001</td>
</tr>
<tr>
<td>Correlation coefficient</td>
<td>0.770</td>
<td>0.675</td>
<td>0.881</td>
</tr>
<tr>
<td><strong>FVC PRED</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Sig. (2 tailed)</td>
<td>0.050</td>
<td>0.013</td>
<td>0.034</td>
</tr>
<tr>
<td>Correlation coefficient</td>
<td>0.832</td>
<td>0.746</td>
<td>0.671</td>
</tr>
<tr>
<td><strong>FEV1</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Sig. (2 tailed)</td>
<td>0.019</td>
<td>0.066</td>
<td>0.002</td>
</tr>
<tr>
<td>Correlation coefficient</td>
<td>0.753</td>
<td>0.602</td>
<td>0.850</td>
</tr>
<tr>
<td><strong>FEV1 PRED</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Sig. (2 tailed)</td>
<td>0.017</td>
<td>0.026</td>
<td>0.022</td>
</tr>
<tr>
<td>Correlation coefficient</td>
<td>0.762</td>
<td>0.693</td>
<td>0.710</td>
</tr>
</tbody>
</table>
PFT

• Sophisticated and relatively expensive
• Needs a well-trained technician
• Relatively longer time is required

• Usability/ Practicality/Reliability ???
  – Young
  – Mentally retarded
  – Mouth disfunction
Conclusion

• Respiratory muscle and cough strength tests (RMCST) are significantly correlated with PFT in non-ambulatory patients with neuromuscular scoliosis
• RMCST is a fast, practical and easy-to-run test
• Measuring the RMCST at EOS outpatient clinics gives an idea about respiratory functions of non-ambulatory neuromuscular patients without need for sophisticated PFT tests
• They can be used for monitorization