Using Technology to Assess Upper Extremity Function (Reachable Workspace) in Neuromuscular Disorders

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Advances in Neuromuscular field

• Recent advances in therapeutic developments
  – Neuromuscular field has been active
  – Drug therapies & pharmacologic interventions
  – Gene and cell therapies
  – But also assistive devices and robotics

• Highlight the need for effective outcome measures
  – Improve monitoring of disease severity and progression
  – Better characterization for natural history studies
  – Identify clinical outcome measures for planned efficacy trials
Traditional tools

Brooke Upper Extremity Rating Scale

<table>
<thead>
<tr>
<th>Grade</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Starting with arms at the sides, the patient can abduct the arms in a full circle until they touch above the head.</td>
</tr>
<tr>
<td>2</td>
<td>Can raise arms above head only by flexing the elbow (shortening the circumference of the movement) or using accessory muscles.</td>
</tr>
<tr>
<td>3</td>
<td>Cannot raise hands above head, but can raise an 8-oz glass of water to the mouth.</td>
</tr>
<tr>
<td>4</td>
<td>Can raise hands to the mouth, but cannot raise an 8-oz glass of water to the mouth.</td>
</tr>
<tr>
<td>5</td>
<td>Cannot raise hands to the mouth, but can use hands to hold a pen or pick up pennies from the table.</td>
</tr>
<tr>
<td>6</td>
<td>Cannot raise hands to the mouth and has no useful function of hands.</td>
</tr>
</tbody>
</table>
Traditional Motion Capture Lab

- Large cost
- Space & equipment requirements
- Not very portable
- Marker-based (active or passive)

- Development of algorithm and methods to reconstruct an individual’s reachability (Reachable workspace).
- Graphical visualization of reachable workspace (3D)
Steps to determining the Reachable Workspace

1. Area obtained
2. Normalized by the area of the hemisphere \((2\pi r^2)\)
3. Allows comparison between individuals

Relative Surface Area (RSA)

Upper Extremity 3D Reachable Workspace Analysis in Dystrophinopathy Using Kinect

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Set up
(sensor + display + computer)

- Simple
- Quick
- Low-cost
- Unobtrusive (no markers)
- Intuitive (visualization of reachable workspace)
Kinect Reachable Workspace
RSA - Reliability testing

Kinect vs. Motion capture

Test and retest

Also, test re-test reliability in DMD

ICC = 0.935

Reachable Workspace Differences between: DMD, BMD, FSHD, and ALS
RSA vs. Brooke (DMD)

RSA vs. Strength (FSHD)

Elbow Flexion

Shoulder Abduction

Combined

No Weight (no loading)
500 Gram wrist weight (~1 lb loading)
Using simple wrist-weight to detect subtle differences in Reachability

FSHD:

Reachable workspace by QMT

- Shoulder Abduction
- Elbow Flexion

* one-way ANOVA with Tukey post hoc analysis, cohort significantly different than controls (p<0.05)
RSA correlates well with PUL

Mapping ADL functions (PRO – NeuroQOL) to Reachable workspace Quadrants

Hygiene

Dressing

Feeding

Shampoo Wash Body

Shampoo Wash Body

Shampoo Wash Body

Shampoo Wash Body

Shirt On/Off Zipper Button Shirt Pants On

Shirt On/Off Zipper Button Shirt Pants On

Spoon to Mouth
Reachable workspace correlation with ADLs

FSHD: Longitudinal study (18 subjects: 8mo-5yrs, ave 2.5yrs)

A

subject 201005: left side, No weight

8/29/2012

subject 201005: left side, 0.5kg weight

8/24/2016

subject 201005: left side, No weight

8/29/2013

9/19/2014

4/3/2015

FSHD: Longitudinal (8mo-5 years follow up)

FSHD Longi: 500g weight

FSHD Longitudinal: 18 subjects followed 8mo-5yrs (ave. 2.5 years)

△-1.63%/yr

△-1.82%/yr

### A. Entire cohort

<table>
<thead>
<tr>
<th>Quadrant</th>
<th>Intercept + S.E.</th>
<th>Slope + S.E.</th>
<th>Calculated Rates (% per year)</th>
<th>Slope P-value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Quadrant 1</td>
<td>0.05511 ± 0.01764</td>
<td>-0.00001 ± 0.0541e-04</td>
<td>-6.62</td>
<td>0.063</td>
</tr>
<tr>
<td>Quadrant 1 weighted</td>
<td>0.04417 ± 0.01765</td>
<td>-0.0871e-04 + 0.0426e-04</td>
<td>-7.20</td>
<td>0.041*</td>
</tr>
<tr>
<td>Quadrant 2</td>
<td>0.10415 ± 0.00815</td>
<td>0.054500e-04 ± 0.0546e-04</td>
<td>+1.91</td>
<td>0.319</td>
</tr>
<tr>
<td>Quadrant 2 weighted</td>
<td>0.09185 ± 0.00997</td>
<td>0.035200e-04 ± 0.0459e-04</td>
<td>+1.40</td>
<td>0.443</td>
</tr>
<tr>
<td>Quadrant 3</td>
<td>0.08285 ± 0.01661</td>
<td>-0.000021 ± 0.0566e-04</td>
<td>-9.25</td>
<td>0.000*</td>
</tr>
<tr>
<td>Quadrant 3 weighted</td>
<td>0.06316 ± 0.01650</td>
<td>-0.000014 ± 0.0413e-04</td>
<td>-8.09</td>
<td>0.001*</td>
</tr>
<tr>
<td>Quadrant 4</td>
<td>0.21091 ± 0.00665</td>
<td>-0.0426e-04 ± 0.0442e-04</td>
<td>-0.74</td>
<td>0.334</td>
</tr>
<tr>
<td>Quadrant 4 weighted</td>
<td>0.20123 ± 0.00848</td>
<td>-0.0421e-04 ± 0.0359e-04</td>
<td>-0.76</td>
<td>0.241</td>
</tr>
<tr>
<td>Total</td>
<td>0.44781 ± 0.01417</td>
<td>-0.00002 ± 0.00001</td>
<td>-1.63</td>
<td>0.144</td>
</tr>
<tr>
<td>Total weighted</td>
<td>0.40010 ± 0.04379</td>
<td>-0.00002 ± 0.000011</td>
<td>-1.82</td>
<td>0.039*</td>
</tr>
</tbody>
</table>

### B. w/o High-function

<table>
<thead>
<tr>
<th>Quadrant</th>
<th>Intercept + S.E.</th>
<th>Slope + S.E.</th>
<th>Calculated Rates (% per year)</th>
<th>Slope P-value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Quadrant 1</td>
<td>0.03252 ± 0.00759</td>
<td>-0.000012 ± 0.0500e-04</td>
<td>-13.47</td>
<td>0.016*</td>
</tr>
<tr>
<td>Quadrant 1 weighted</td>
<td>0.01952 ± 0.00553</td>
<td>-0.0844e-04 ± 0.0424e-04</td>
<td>-15.78</td>
<td>0.047*</td>
</tr>
<tr>
<td>Quadrant 2</td>
<td>0.09956 ± 0.00838</td>
<td>0.0678e-04 ± 0.0527e-04</td>
<td>+2.48</td>
<td>0.198</td>
</tr>
<tr>
<td>Quadrant 2 weighted</td>
<td>0.08876 ± 0.01034</td>
<td>0.0860e-04 ± 0.0490e-04</td>
<td>+3.54</td>
<td>0.861</td>
</tr>
<tr>
<td>Quadrant 3</td>
<td>0.06344 ± 0.01030</td>
<td>-0.0000231 ± 0.0625e-04</td>
<td>-13.20</td>
<td>0.000*</td>
</tr>
<tr>
<td>Quadrant 3 weighted</td>
<td>0.04094 ± 0.00785</td>
<td>-0.0000143 ± 0.0454e-04</td>
<td>-12.75</td>
<td>0.002*</td>
</tr>
<tr>
<td>Quadrant 4</td>
<td>0.20982 ± 0.00731</td>
<td>-0.0582e-04 ± 0.0497e-04</td>
<td>-1.01</td>
<td>0.243</td>
</tr>
<tr>
<td>Quadrant 4 weighted</td>
<td>0.19855 ± 0.00920</td>
<td>-0.0522e-04 ± 0.0402e-04</td>
<td>-9.60</td>
<td>0.194</td>
</tr>
<tr>
<td>Total</td>
<td>0.40036 ± 0.02649</td>
<td>-0.000027 ± 0.000016</td>
<td>-2.47</td>
<td>0.093</td>
</tr>
<tr>
<td>Total weighted</td>
<td>0.34853 ± 0.02660</td>
<td>-0.000029 ± 0.000012</td>
<td>-3.04</td>
<td>0.016*</td>
</tr>
</tbody>
</table>

ALS longitudinal data


Pompe patients: pre- & post-ERT

Subject 105

Pre

3 months post

6 months post

Subject 107
Clinical Trial & in-clinic RWS system

• >20 systems deployed around the world (NA, South America, Asia, Europe)
  – Neuromuscular conditions
  – Neurological
  – Orthopedic & musculoskeletal
  – Post breast cancer rehab
  – Drug trial

• Portable system can be taken to patient’s home (laptop, tripod+Kinect)

• Cloud-based system
  – Internet sign-on
  – Automatic data upload to cloud server
Acknowledgement

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