The relationship of dystonia and choreoathetosis with activity, participation and quality of life in dyskinetic CP children

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Disclosure Information

Disclosure of Relevant Financial Relationships:

No financial relationships to disclose.

Disclosure of Off-Label and/or investigative uses:

We will not discuss off label use and/or investigational use in my presentation.
Introduction

*Cerebral palsy* describes a group of *permanent disorders of the development of movement and posture*, causing *activity limitation*, that are attributed to non-progressive disturbances that occurred in the developing fetal or infant brain. The motor disorders are often accompanied by disturbances of sensation, perception, cognition, communication, and behaviour, by epilepsy, and by secondary musculoskeletal problems.

*Rosenbaum et al. 2007*
Introduction

CEREBRAL PALSY
3/1000°

Spasticity 79.1%
Dyskinesia 14.4% (0.3/1000°)
Ataxia 3.9%

Choreo-athetosis
Dystonia

Chorea
Athetosis

SCPE 2000, 2007 *Dev Med Child Neurol*
SCPE 2005, *R&TM of the SCPE*
Bax e.a. 2006 *JAMA*
Rosenbaum e.a. 2006, 2007 *Dev Med Child Neurol*
Sanger e.a. 2010 *Mov Disord*
Dyskinetic CP

Complex movement disorder
• Little is known about the impact of dystonia and choreoathetosis on activities and participation
• Difficult for targeted therapy

*International Classification of Functioning, Disability and Health (WHO)*
Objectives

To gain more insights in the relationship between the presence of dystonia and choreoathetosis & the level of activity, participation and quality of life
## Methods: participants

<table>
<thead>
<tr>
<th>Characteristics</th>
<th>N=55 (30 male; 25 female)</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>- age 5-22 yrs</td>
</tr>
<tr>
<td></td>
<td>- Mean age=14y6mo ; SD=4y1mo</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Inclusion criteria</th>
<th>predominant dyskinetic CP</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>able to understand test instructions</td>
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</tbody>
</table>

<table>
<thead>
<tr>
<th>Exclusion criteria</th>
<th>orthopaedic or neurosurgical interventions &lt; 12 months</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>spine fusion</td>
</tr>
</tbody>
</table>
Methods: measurement & classification

Assessment dystonia and CA: Dyskinesia Impairment Scale (DIS)

Monbaliu et al 2012, Dev Med Child Neurol
Methods: measurement and classification

Activity measures

Gross motor

- Gross Motor Function Measurement
  - Lying and rolling
  - Crawling and kneeling
  - Sitting
  - Standing
  - Walking, running and jumping
- Functional Mobility Scale

Upper limb

- Jebson-Taylor Test of Hand Function
  - Measures movement, speed and manual dexterity in 6 unimanual tasks
  - Expressed in seconds needed for the task execution
- Abilhand-Kids Questionnaire
  - Assesses manual ability on 21 manual activities
  - Perceived by parents/caretakers
Methods: measurement and classification

Participation measures

- **Assessment of Life Habits Kids (LIFE-H)**
  - Assesses daily activities and social roles over 12 domains
  - 0 (lowest participation) ... 10 (maximal participation)

- **Quality of Life Questionnaire for children with CP (CP-QOL)**
  - Measures physical well-being, social well-being, school, access to services and acceptance by others
  - 0 (lowest QOL) ... 100 (maximal QOL)
  - Parent proxy-form
### Results

**Gross Motor Activity Measures**

<table>
<thead>
<tr>
<th></th>
<th>Gross Motor Function $r_s$</th>
<th>Functional Mobility Scale $r_s$</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Dystonia</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Total %</td>
<td>-0.65**</td>
<td>-0.71**</td>
</tr>
<tr>
<td>Leg %</td>
<td>-0.58**</td>
<td>-0.69**</td>
</tr>
<tr>
<td><strong>Choreoathetosis</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Total %</td>
<td>-0.05</td>
<td>-0.27*</td>
</tr>
<tr>
<td>Leg %</td>
<td>0.12</td>
<td>-0.14</td>
</tr>
</tbody>
</table>

**Moderate to good relationship**

**No to very weak relationship**

No correlation = 0.00 < $r_s$ < 0.25
Fair correlation = 0.25 < $r_s$ < 0.50
Good correlation = 0.50 < $r_s$ < 0.75
Excellent correlation = 0.75 < $r_s$

$r_s$: Spearman’s rho correlation coefficient
Results

upper limb activity measures

<table>
<thead>
<tr>
<th>Condition</th>
<th>Jebson-Taylor Test $r_s$</th>
<th>Abil-Hand Kids Questionnaire $r_s$</th>
</tr>
</thead>
<tbody>
<tr>
<td>Dystonia</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Total %</td>
<td>0.64**</td>
<td>-0.67**</td>
</tr>
<tr>
<td>Arm %</td>
<td>0.76**</td>
<td>-0.72**</td>
</tr>
<tr>
<td>Choreoathetosis</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Total %</td>
<td>-0.17</td>
<td>-0.09</td>
</tr>
<tr>
<td>Arm %</td>
<td>0.24</td>
<td>-0.11</td>
</tr>
</tbody>
</table>

- No correlation = $0.00 < r_s < 0.25$
- Fair correlation = $0.25 < r_s < 0.50$
- Good correlation = $0.50 < r_s < 0.75$
- Excellent correlation = $0.75 < r_s$

$r_s$ : Spearman’s rho correlation coefficient
## Results

**participation and quality of life**

<table>
<thead>
<tr>
<th></th>
<th>LIFE-Habit (n=48)</th>
<th>CP-QOL (n=45)</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>$r_s$</td>
<td>$r_s$</td>
</tr>
<tr>
<td><strong>Dystonia</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Total %</td>
<td>-0.42**</td>
<td>-0.32**</td>
</tr>
<tr>
<td>Mouth</td>
<td>-0.39**</td>
<td>-0.31**</td>
</tr>
<tr>
<td>Arms</td>
<td>-0.60**</td>
<td>-0.44</td>
</tr>
<tr>
<td>Legs</td>
<td>-0.23</td>
<td>-0.11</td>
</tr>
<tr>
<td><strong>Choreoathetosis</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Total %</td>
<td>0.13</td>
<td>-0.21</td>
</tr>
<tr>
<td>Mouth</td>
<td>-0.14</td>
<td>-0.08</td>
</tr>
<tr>
<td>Arms</td>
<td>0.05</td>
<td>-0.21</td>
</tr>
<tr>
<td>Legs</td>
<td>0.14</td>
<td>-0.29*</td>
</tr>
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Excellent correlation = 0.75 < $r_s$

$r_s$: Spearman’s rho correlation coefficient

*No relationship*
Discussion

**Impact of dystonia** on activities and participation/QOL is **higher** compared with **choreoathetosis**

**Why?**
- < hallmark characteristics of dystonia and CA?
- < dystonia is masking the CA?
- Further research!
Discussion

Impact of dystonia in **upper limb** and **mouth regions** on participation and QOL

**Why?**

- < use of mobility aids (often operated using upper limbs)
- < communication as an important factor in participation and QOL
Conclusion

• First study to examine relationship between dystonia/CA and activities, participation and QOL
  – Importance of dystonia
  – Importance of upper limb and mouth region

• Future **targeted intervention studies** are needed: insight in importance of dystonia and CA in children with dyskinetic CP.
Thank you for your attention