Disclosure Information
AACPDM 67th Annual Meeting October 16-19, 2013

Speaker Name: MONBALIU ELEGAST

Disclosure of Relevant Financial Relationships:
I have no financial relationships to disclose.

Disclosure of Off-Label and/or investigative uses:
I will not discuss off label use and/or investigational use in my presentation

Overview

• Introduction
• Evaluation
• Clinical cases

Evaluation and insights in secondary dystonia and choreoathetosis in dyskinetic CP

Monbaliu E, PhD PT
AACPDM 2013-10-1 Milwaukee

Definition and classification

CEREBRAL PALSY

Spasticity
79.1%

Dyskinesia
14.4%

Ataxia
3.9%

Choreo-athetosis

dystonia

Chorea

Athetosis

Dyskinetic CP

• Involuntary, uncontrolled, recurring, occasionally stereotyped movements
• Primitive reflex patterns predominate
• Muscle tone is varying

SCPE (2005, 2007)
– Involuntary movements, distorted voluntary movements and abnormal postures due to sustained muscle contractions (e.g., rotation, extension, flexion of body parts)
– Sanger et al. (2003, 2010)
– A movement disorder with involuntary, uncontrolled, occasionally stereotyped movements and abnormal postures due to sustained muscle contractions
Definition and classification

- **CEREBRAL PALSY**
  - Spasticity: 79.1% (0.3/1000°)
  - Dyskinesia: 14.4% (0.3/1000°)
  - Ataxia: 3.9%
  - Chorea
  - Athetosis

- **SCPE (2005, 2007)**
  - Chorea = rapid, involuntary, jerky, often fragmented movements
  - Athetosis = slower, constantly changing, writhing or convulsive movements

- **Sanger et al (2010)**
  - Chorea = a rapid, fluctuating, ill-defined, incoordinate movement
  - Athetosis = a slow, continuous, involuntary writhing movement that prevents maintenance of stable posture

Prevalence

<table>
<thead>
<tr>
<th>Study</th>
<th>Participants</th>
<th>Spastic CP</th>
<th>Dyskinetic CP</th>
<th>Ataxic CP</th>
<th>Other</th>
</tr>
</thead>
<tbody>
<tr>
<td>Bax e.a. 2006 Cross sectional</td>
<td>558</td>
<td>79.1%</td>
<td>14.4%</td>
<td>3.9%</td>
<td>2.6%</td>
</tr>
</tbody>
</table>

Pathogenesis

- Hyperbilirubinemia
- Neonatal asphyxia & brain damage in the **thalamus** and **basal ganglia** during the 3rd trimester of pregnancy are strongly associated with dyskinetic CP.

Kyllermann, 1982 Acta Pediatr Scand
Himmelmann, 2009 Dev. Med. Child Neurol
**Pathogenesis**

![Brain Lesions in Basal Ganglia & Thalamus]

**Clinical picture**

**Motor Severity**
Gross Motor Function Classification System Scale (GMFCS)

Dyskinetic Population study (N=48)

- Level I & II → 8.3% (n=4)
- Level III → 12.5% (n=6)
- Level IV → 20.8% (n=10)
- Level V → 58.3% (n=28) 79.1%

Himmelmann 2007, Dev Med Child Neurol

**Clinical picture**

Accompanying impairments

![Chart showing GMFCS levels](chart.png)

Himmelmann 2007, Dev Med Child Neurol

**Therapy**

**Medical interventions**

- Oral medication
  - Silber 2003, J Child Neurol
  - Albright 2006, Neuroscience Focus
- Intrathecal baclofen
  - Albright 2006, Neuroscience Focus
- Deep Brain stimulation (DBS)
  - Halley 2006, Neuroscience Focus
  - Vidailhet 2009, Lancet Neurology
  - Marks 2011, Mov Disord
  - Lumsden 2013, DMCN
- Botulinum Toxin
  - ↓ pathologic movements

![Deep Brain stimulation](DBS.png)
Therapy

Rehabilitation

• Physical therapy
• Occupational therapy
• Speech therapy
• Mobility aids
• Orthotics

↓ pathological movements & ↑ function, activity, participation and quality of life

Problem

1. Dystonia and choreoathetosis are complex movement disorders => measurement of therapy effects?

2. Little is known about the clinical presentation of dystonia and choreoathetosis in dyskinetic CP => difficult for targeted therapy

Overview

• Introduction
• Evaluation
• Clinical cases

Evaluation

Clinical Rating Scales

Choreoathetosis Scale

• No scales available

Dystonia Scale

• Primary Dystonia Scales
  – Burke Fahn Marsden Scale, BFMS, Burke e.a. 1985 Neurology
  – Unified Dystonia Rating Scale, UDRS, Comella e.a. 2003 Movement Disorders
• Secondary Dystonia Scales
  – Barry Albright Dystonia Scale, BADS, Barry e.a. 1999 Dev Med Child Neurol

1. How reliable, valid and sensitive are the existing dystonia scales?

2. There is a need for a new evaluation scale to measure dystonia and choreoathetosis in a similar scoring construct.
**Evaluation studies**

**Study 1:**
Rating scales for Dystonia in Cerebral Palsy: Reliability and Validity
published Dev Med Child Neurol 2010

**Study 2:**
The Dyskinesia Impairment Scale: a new instrument to measure dystonia and choreoathetosis in Cerebral Palsy
published Dev Med Child Neurol 2012

**Study 3:**
Can the Dyskinesia Impairment Scale be used by inexperienced raters? A reliability study.
published European Journal of Paediatric Neurology 2013

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**Objectives study 1**

1. To investigate the reliability of the three dystonia scales in dyskinetic CP

2. To assess the validity of the three scales in dyskinetic CP

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**Methods**

**Dystonia Scales**

**Primary Dystonia Scale**
- Burke Fahn Marsden Movement Scale (BFMS), Burke e.a. 1985 Neurology
- Unified Dystonia Rating Scale (UDRS), Comella e.a. 2003 Movement Disorders

**Secondary Dystonia Scale**
- Barry Albright Dystonia Scale (BADS), Barry e.a. 1999 Dev Med Child Neurol

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**BFMS**

- Ordinal scale
- Movement Scale
- Disability Scale
- Provoking Factor
  - 6 body regions range 0-4
- Severity Factor
  - 2 body region range 0-3

Burke e.a. 1985 Neurology
Dystonia Scales

Primary Dystonia Scale
- Burke Fahn Marsden Movement Scale, BFMMS, Burke e.a. 1985 Neurology
- Unified Dystonia Rating Scale, UDRS, Comella e.a. 2003 Movement Disorders

Secondary Dystonia Scale
- Barry Albright Dystonia Scale, BADS, Barry e.a. 1999 Dev Med Child Neurol

Participants
- N=10
- Dominant dystonic CP, > 4 yrs (mean age=14.0 yrs)
- GMFCS representation:
  - GMFCS I: N=1
  - GMFCS II: N=1
  - GMFCS III: N=1
  - GMFCS IV: N=1
  - GMFCS V: N=6

Methods

Procedures
- Extended version of the proposed Video Protocol for Dystonia Study Group, 1997 Comella e.a. 2003
- Standard montage:
  - 12 body regions
  - Rest/activity

Raters
- N=3
  - 2 child neurologists
  - 1 physical therapist

Results
- Intraclass Correlation Coefficients:
  - Moderate to high interrater reliability for the total scores
  - Low to moderate interrater reliability for the itemscores
- Cronbach’s α: Good internal consistency
- Correlation: Good relationship between BFMMS, UDRS, BADS
Results

- Standard Error of Measurement (SEM) and Minimal Detectable Difference
  - High MDD % make the scales insufficiently useful for intervention studies and longitudinal follow-up
- Content analysis:
  - no differentiation between action/rest
  - no differentiation between duration and amplitude
  - no differentiation between proximal and distal limbs

Conclusion

Considering:
- high measurement errors
- limitations in sensitivity (action/rest; duration/amplitude)
- Lack of choreoathetosis scale

Need for a new scale

Evaluation of dystonia and choreo-athetosis in a similar scoring construct

Evaluation studies

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Study 2:
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Scale Development

Rating Scales for Dyskinesia

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<tr>
<th>Scale</th>
<th>Prim. Dyst.</th>
<th>Sec. Dyst.</th>
<th>Chorea Athet.</th>
<th># body areas</th>
<th>Video Scoring</th>
<th>Distinction Action / rest</th>
<th>Distinction Duration / Amplitude</th>
<th>Distinction Proximal / Distal limbs</th>
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<tr>
<td>BFMMDR</td>
<td>-</td>
<td>-</td>
<td>-</td>
<td>9</td>
<td>+</td>
<td>-</td>
<td>-</td>
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<td>+</td>
<td>-</td>
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Evaluation Scale Development

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Scale development

**BMDR**
- Dystonia
- Spasticity
- Hyperreflexia
- Babinski

**USDS**
- Dystonia
- Spasticity
- Hyperreflexia
- Babinski

**DIS**
- Dystonia
- Spasticity
- Hyperreflexia
- Babinski

**DIS**
- Dystonia
- Spasticity
- Hyperreflexia
- Babinski

**Objective study 2**

To assess the **reliability** and **validity** of the DIS in **dyskinetic CP**

**Methods**

**Participants**
- N=25
  - Dominant dyskinetic CP, 5-22y (mean age=13y6mo)
  - GMFCS
    - GMFCS I: N=4
    - GMFCS II: N=4
    - GMFCS III: N=4
    - GMFCS IV: N=3
    - GMFCS V: N=10

**Procedures**
- Extended version of the proposed Video Protocol for Dystonia
- Standard montage
  - 12 body regions
  - Action/rest
- N=2 PTs
Results

1. Intraclass Correlation Coefficients
   - dystonia subscale: very good interrater reliability
   - choreoathetosis subscale: excellent interrater reliability
2. SEM-MDD: low measurement errors
3. Cronbach’s α: very good internal Consistency
4. Pearson’s correlation coefficient (r): good concurrent validity between Dystonia subscale and BADS

Evaluation studies

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Study 3:
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Methods

Procedures

Raters
- N=5 PT
  - 3 senior PTs
  - 2 junior PTs

Results

1. choreo-athetosis subscale => no concurrent validity
2. discriminating between dystonia & choreoathetosis => requires sufficient experience

Objectives study 3

1. To assess the reliability of the DIS in hands of inexperienced raters, new to discriminating between dystonia and choreoathetosis
2. To determine the effects of clinical experience on reliability

Interrater reliability
- Choreoathetosis total scores: good to excellent reliability for all inexperienced raters
- Dystonia: total scores: moderate to good reliability for all inexperienced raters
- no difference in reliability between senior/junior PT

Internal consistency
- similar Cronbach’s alpha between experienced and inexperienced raters
Results

- Interrater reliability inexperienced raters
  - Region scores generally ranged from poor to good
  - For dystonia subscale lower reliability during rest than during activity

- SEM & MDD
  - Higher measurement errors than experienced raters

Conclusion study 2 & 3

- Experienced raters > inexperienced raters
- Choreo-athetosis scale is more reliable than the dystonia scale for inexperienced raters
- No effect of clinical expertise

- Good acquaintance of operational definitions is crucial to enhance the reliability
  - A manual of the DIS and (online) training is necessary

Dyskinesia Impairment Scale

Useful tool to increase further understanding of dystonia and choreoathetosis

Clinical pattern analysis of secondary dystonia and choreoathetosis in dysskinetic CP

Introduction
Evaluation
Clinical cases