Part 2: spasticity/dystonia/choreoathetosis

- Why discriminate chorea & athetosis
  - Necessary for diagnosis?
  - Necessary for prognosis?
  - Necessary for management?
  - Necessary for scientific research (basic research/epidemiology...)?

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Part 2: spasticity/dystonia/choreoathetosis

Choreoathetosis – definition

- Historical overview
  - Phelps (1942) – 12 forms of athetosis (rotary A, tremor A, dystonic A, shudder A, tension A, hemiathetosis, neard&P athetosis, deaf athetoid, balance release, emotional release...)
  - Minar (1956) – 4 forms of athetosis (tension A, non-tension A, dystonic A, tremor A, ...)
  - Bobath
  - Kellermann
  - Camp/SCPE (2000/2007)
  - Sanger (2010)

- Consensus definition?
  - Currently definition of SCPE & Sanger et al. are most often used in CP

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Part 2: spasticity/dystonia/choreoathetosis

Choreoathetosis – definition

- Two most used definitions in CP:
  - SCPE (2005, 2007)
    - Choreoathetosis in CP is dominated by hyperkinesia and muscle tone fluctuation.
      - Chorea, i.e. rapid, involuntary, jerky and often fragmented movements.
      - Athetosis, i.e. slower, constantly changing, writhing or contorting movements.
  - Sanger et al. (2010)
    - Chorea = a ongoing random-appearing sequence of one or more discrete involuntary or movement fragments.
    - Athetosis = a slow continuous, involuntary writhing movement that prevents maintenance of a stable posture.

- Why combining chorea and athetosis?
  - Distinction does not appear to be clinically useful in CP.
Part 2: spasticity/dystonia/choreoathetosis

Choreoathetosis – definition

Choreoathetosis – pathophysiology

What choreoathetosis is not!

- dystonia
- tics
- myoclonus
- stereotypies
- tremor

Take home message
Part 2: spasticity/dystonia/choreoathetosis

Choreoathetosis – pathophysiology

Discussion:
- significant association between severity of choreoathetosis and lesions confined to basal ganglia and thalamus

Monbaliu et al (2016), DMCW

Overview

- Part 1: Why discriminate spasticity – dystonia – choreoathetosis in CP
- Part 2: What is spasticity – dystonia – choreoathetosis in CP?
- Part 3: Clinical discrimination & evaluation overview
- Part 4: Clinical cases & discussion

Part 3: clinical discrimination- evaluation

A. Measures
- Discriminative measure
- Predictive measure
- Evaluative measure

B. Clinimetric criteria
- Clinical relevance & potential responsiveness
- Patient acceptance & time needed to assess
- Reliability and validity of the scale
Discriminative measures for spasticity – dystonia – choreathetosis

<table>
<thead>
<tr>
<th>Choreoathetosis</th>
<th>No scales available</th>
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<tbody>
<tr>
<td>Spasticity/Dystonia</td>
<td>Hypertonia Assessment Tool</td>
</tr>
<tr>
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<td>Jetwa e.a. 2010</td>
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<td>Knights e.a. 2014</td>
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<tr>
<td></td>
<td>Acquaintance of operational definitions</td>
</tr>
</tbody>
</table>

Part 3: clinical discrimination – evaluation

How to discriminate dystonia from spasticity in clinical practice?

Good acquaintance of operational definitions is essential!

- **Dystonia:**
  - *SCPE:* involuntary movements, distorted voluntary movements and abnormal postures due to sustained muscle contractions (slow rotation, extension, flexion of body parts)
  - Sanger e.a. (2003, 2010):
    - a movement disorder in which involuntary sustained or intermittent muscle contraction cause twisting and repetitive movements, abnormal postures or both
- **Spasticity:**
  - is characterized by an increased resistance which is velocity dependent.
  - A spastic catch is felt some time after onset of movement.
  - Clonus is often associated with hyper-reflexia. It is considered pathological when it is prolonged or does not stop spontaneously.
  - Pathological posturing of lower limbs is characterized by: (1) internal rotation of the hip; (2) hip adduction; and (3) equinus foot, resulting in a ‘scissored’ position.
Spasticity: increased tone – velocity dependent

Dystonia: Upper limb

Dystonia: Lower limb
Spasticity:

Dystonia:

Mixed presentation:
How to discriminate dystonia from choreoathetosis in clinical practice?

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**Discrimination between dystonia and choreoathetosis**
Understanding of dystonia and choreoathetosis

- Simultaneous presence of Dystonia & Choreoathetosis
- Dystonia > Choreoathetosis
- Activity/Part/QLQ: D Impact

Rosenbaum (1990)
Kirshner & Guyatt

A. Measures
- Discriminative measure
- Predictive measure
- Evaluative measure

B. Clinimetric criteria
- Clinical relevance & potential responsiveness
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Part 3: clinical discrimination – evaluation

Clinical evaluation measures for spasticity – dystonia - choreathetosis

| Spasticity                                      | Modified Ashworth Scale |
|                                                | Modified Tardieu Scale  |
| Dystonia                                       | Barry-Albright Dystonia Scale |
|                                                | Burke-Fahn-Marsden Scale |
|                                                | Movement Disorder – Childhood Rating Scale |
| Choreoathetosis                                | Dyskinesia Impairment Scale – Dystonia subscale |
|                                                | Dyskinesia Impairment Scale – Choreoathetosis subscale |
### Part 3: Clinical Discrimination – Evaluation

**Clinical Evaluation Measures for Spasticity – Dystonia – Choreoathetosis**

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### Part 3: Clinical Discrimination – Evaluation

**Stationary Qualitative Spasticity Assessment – Modified Ashworth Scale**

- 0 = No increase in muscle tone
- 1 = Slight increase in muscle tone, manifested by catch and release, or by minimal resistance at the end of the range of motion
- 1+ = Slight increase in muscle tone, manifested by a catch, followed by minimal resistance throughout the remainder (less than half) of the ROM
- 2 = More marked increase in muscle tone, easily moved
- 3 = Considerable increase in muscle tone, passive movement difficult
- 4 = Rigid

Scoring the resistance felt in a specific muscle group by passively moving a limb at one velocity

Subjective feeling of resistance

### Part 3: Clinical Discrimination – Evaluation

**Modified Tardieu Scale**

Joint angles measurement (at which muscle reaction is felt) at different velocities of passive muscle stretch

Subjective feeling of resistance!
Co-existence of different types of hypertonia
Spasticity, dystonia and rigidity
Force generated by active muscles/stretch reflexes/active tissue properties
Voluntary/involuntary muscle contraction
State of al terness, activity, posture
Emotional factors, pain, touch

Difficulties in assessing spasticity in children

Clinical evaluation measures for spasticity – dystonia - choreathetosis

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Part 3: clinical discrimination – evaluation

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Questions?
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