Dystonia, spasticity & choreoathetosis

This instructional course will rely on interactivity

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Questions will be displayed viewed on the screen

How to recognise, discriminate and measure them in cerebral palsy?
Disclosure

• No conflict of interests
• No relations that could be relevant for the meeting

Learning objectives

• To describe definitions & classification of dystonia/spasticity/choreoathetosis
• To understand the pathophysiology of dystonia/spasticity/choreoathetosis
• To recognize clinical characteristics of dystonia/spasticity/choreoathetosis
• To acquire a practical framework for discriminating and evaluating dystonia/spasticity/choreoathetosis

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
Cerebral Palsy - Definition

- Cerebral Palsy (CP) 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 of CP are often accompanied by disturbances of sensation, perception, cognition, communication, and behaviour, by epilepsy, and by secondary musculoskeletal problems

Rosenbaum et al. (2007), DMCN

Cerebral Palsy - Motor abnormalities

A. NATURE AND TYPOLOGY OF THE MOTOR DISORDER:
- The observed tonal abnormalities assessed on examination: hypotonia, hypertonia
- Diagnosed movement disorders present: spasticity, ataxia, dystonia, athetosis

B. FUNCTIONAL MOTOR ABILITIES:
- The extent to which the individual is limited in his or her motor function, including oromotor and speech function.

Rosenbaum et al. (2007), DMCN

Challenge...

- Spasticity
- Chorea-athetosis
- Dystonia

Rosenbaum et al. (2007), DMCN
Cans et al. (2000), SCPE
Bax et al. (2006), JAMA
Overview

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

Questions & answers

• Is there consensus re definitions?
• What is necessary in a motor disorder definition?
• Is a functional criterion required (e.g. walking...)?
• Should the definition be medical condition-specific (e.g. stroke/CP/...)?
• Should the definition be age-specific (e.g. childhood)?
• Should it include a physiological parameter?
• Should it include an anatomical parameter (e.g. MRI)?
• Should comorbidity be included?

Part 2: spasticity/dystonia/choreoathetosis

Definition & classification

• Definitions are international & consensus based:
  • Expert panels: e.g. Surveillance of Cerebral Palsy in Europe (SCPE), National Institutes of Health (NIH)
  • Neural basis – upper motor neurone syndrome
  • Several features are mostly incorporated, depending from clinical/physiological perspective
    • clinical tone, kinesia, active/passive (resistance, stretch), reflexes, cocontraction, overflow, trophic changes, aetiology, presence during sleep, ...
    • Notion from neurology: ‘+’ (excessive); ‘-’ (too low)
  • Terms spasticity, dystonia, choreoathetosis are used in CP classification => what is the content?
Part 2: spasticity/dystonia/choreoathetosis

Spasticity – definition

- Historical overview
  - Good (1829)
  - Lance e.a. (1988)
  - SCPE (2000, 2007)
  - Sanger (2003, 2010)
- Old concepts live on => new definitions are updated reformulations
  - Sometimes emphasis on clinical aspects
  - Sometimes emphasis on physiological aspects
- Consensus definitions?
  - Children/adults – neurology
  - Currently no specific consensus spasticity definition for children or for CP

Part 2: spasticity/dystonia/choreoathetosis

Spasticity – definition

- Two most used definitions in CP:
    - 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.
  - Sanger (2003/2010)
    - Spasticity is hypertonia in which 1 or both of the following signs are present:
      1. Resistance to externally imposed movement increases with increasing speed of stretch and varies with the direction of joint movement.
      2. Resistance to externally imposed movement rises rapidly above a threshold speed or joint angle.

Part 2: spasticity/dystonia/choreoathetosis

Spasticity – resistance to externally imposed movement
Part 2: spasticity/dystonia/choreoathetosis

Spasticity – typical pathological posturing

Cave: abnormal posture is not per se spasticity!

Part 2: spasticity/dystonia/choreoathetosis

What spasticity is not!

- dynamic hypertonia/co-activation
- musculo-skeletal alterations
- muscle weakness
- abnormal motor control/lack of selectivity

Take home message

Part 2: spasticity/dystonia/choreoathetosis

Spasticity – Pathophysiology
Part 2: spasticity/dystonia/choreoathetosis

Spasticity – Tonic stretch reflexes

Motoneuron excitation (direct and indirect)
Motoneuron inhibition
Control of fine finger movements
Security of monosynaptic connection between distant neurons
Selection, control and modulation of spinal reflexes
Long-term plasticity of spinal circuitry
Descending control of afferent input, incl. noceception
Control of somatosensory and proprioceptive input to sensory and associative cortex (afferent control of sensory exploration)
Control of autonomous nervous system
Intermediary pathway between subcortical structures (BG, cerebellum), cortical areas (premotor, associative), sensory afferences from peripheral sensory receptors and spinal motor neurons
Trophic role during development
Part 2: spasticity/dystonia/choreoathetosis

Questions & answers

What is necessary in the dystonia definition?

- Should it include tone?
- Should it include movement?
- Should it be age-specific?
- Should the definition be medical condition specific?
- Should it include an anatomical parameter (e.g. MRI)?
- Should it include functional aspects?
Dystonia – definition

- Historical overview
  - Oppenheim (1911)
  - Kyllermann (1975)
  - Marsden (1984)
  - Sanger (2003/2010)
  - Albanese (2013)

- Consensus definitions?
  - Children/adults – neurology
    - Currently definition of SCPE (2000,2007) and Sanger et al (2003, 2010) are most often used in CP

Two most used definitions in CP:

  - involuntary movements, distorted voluntary movements and abnormal postures due to sustained muscle contractions (slow rotation, extension, flexion of body parts)

  - a movement disorder in which involuntary sustained or intermittent muscle contraction cause twisting and repetitive movements, abnormal postures or both

Challenge of dystonia:

- Leads to abnormal posture but also abnormal movements => it is a tone problem but also a movement (kinesia) disorder
Part 2: spasticity/dystonia/choreoathetosis

What dystonia is not!

- Spasticity
- Dystonia
- Abnormal posture or movement outside of pathology, e.g. due to physiological ‘immaturity’
- Do not use the terminology dystonia for these posture/movements, e.g. in normal development

Take home message

Part 2: spasticity/dystonia/choreoathetosis

Dystonia - pathophysiology

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

Kyllermann, 1992 Acta Paediatr Scand
Kägeloh-Mann, 2007 Dev. Med. Child Neurol
Himmelmann, 2009 Dev. Med. Child Neurol
Subcortical structures important for motor control

• Basal ganglia
• Cerebellum
• Dopaminergic mesencephalic system

Lesions do not lead to loss of movement but to incoordination and disorganisation of movement

Dystonia - pathophysiology

- remains largely unclear how brain abnormalities produce dystonia
- role of cerebellum, other different important regions?
- role of direct (facilitatory) and indirect (inhibitory) pathways?
- no significant association between severity of dystonia and lesions confined to basal ganglia and thalamus

Dystonia => network disorder

Quartarone et al (2013), Neurobiol Dis
Monbaliu et al (2016), JNCW

Choreoathetosis – classification

Spastic CP
Dystonic CP
Ataxic CP
Choreo-athetosis
Cerebral Palsy

Surveillance of Cerebral Palsy in Europe (2000, 2007)