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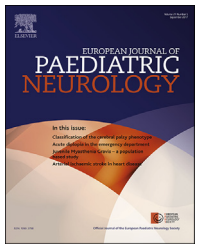

Original article

Spasticity, dyskinesia and ataxia in cerebral palsy: Are we sure we can differentiate them?

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ABSTRACT

Objective: Cerebral palsy (CP) can be classified as spastic, dyskinetic, ataxic or combined. Correct classification is essential for symptom-targeted treatment. This study aimed to investigate agreement among professionals on the phenotype of children with CP based on standardized videos.

Methods: In a prospective, observational pilot study, videos of fifteen CP patients (8 boys, mean age 11 ± 5 y) were rated by three pediatric neurologists, three rehabilitation physicians and three movement disorder specialists. They scored the presence and severity of spasticity, ataxia or dyskinesias/dystonia. Inter- and intraobserver agreement were calculated using Cohen's and Fleiss' kappa.

Results: We found a fair inter-observer ($\kappa = 0.36$) and moderate intra-observer agreement ($\kappa = 0.51$) for the predominant motor symptom. This only slightly differed within the three groups of specialists ($\kappa = 0.33–0.55$).

Conclusion: A large variability in the phenotyping of CP children was detected, not only between but also within clinicians, calling for a discussing on the operational definitions of spasticity, dystonia and ataxia. In addition, the low agreement found in our study questions the reliability of use of videos to measure intervention outcomes, such as deep brain stimulation in dystonic CP. Future studies should include functional domains to assess the true impact of management options in this highly challenging patient population.

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1. Introduction

Cerebral palsy (CP) is commonest motor disorder in children.[1] In clinical practice, CP patients are clinically subdivided according to the predominant motor disorder, e.g. spastic, dyskinetic/dystonic or ataxic. Spasticity is defined as hypertonia where 1) resistance to externally imposed movement increases with increasing speed of stretch and varies with the movement direction and/or 2) resistance to externally imposed movement rises rapidly above a threshold speed or joint angle.[1,2] Dyskinesia or dystonia is a movement disorder in which involuntary sustained or intermittent muscle contractions cause twisting and repetitive movements, abnormal postures, or both.[2] Ataxia is characterized by an impairment of the coordination of goal-directed movements, resulting in gait and trunk disturbances, intention tremor and slurred speech.[3]

Spasticity is the most prevalent form of CP, but dystonic CP is the most common cause of childhood dystonia.[1] Moreover, dystonia may be under-recognized and classified as spasticity in children with CP.[4] Accurate phenotyping is of great importance as management is entirely symptomatic and it is becoming increasingly clear that the different phenotypical subtypes require a specific approach.[4] For instance, (intrathecal) baclofen has been proven effective in spasticity whereas deep brain stimulation (DBS) may ameliorate dystonic symptoms.[5,6] Especially in dystonic CP, intervention studies for dystonic CP primarily use video assessments to measure the extent of dystonia, for instance after DBS treatment.[7] The ability to differentiate between spasticity, dystonia/dyskinesia and ataxia in CP is thus essential to reliably phenotype and follow-up of patients.

This pilot study aimed to determine the agreement on the phenotypical classification of children with CP based on video assessment among and within different clinicians working regularly with young patients with CP.

2. Methods

2.1. Patients

This study was approved by the medical ethical committee of the University Medical Center Groningen (UMCG; the Netherlands – M14.164690). We selected fifteen young CP patients who attended the pediatric rehabilitation outpatient clinic of the UMCG. Informed consent or third party assent was obtained in all participants and/or caregivers.

2.2. Assessment

We videotaped the children according to a standardized video protocol of 5–7 min according to a standard neurological examination, including sitting, standing and walking, and examination of muscle tone, deep tendon reflexes, coordination tasks and simple motor tests.

2.3. Clinicians

Nine clinicians regularly working with young patients with CP participated in this pilot study. The clinicians were selected based upon their background (three pediatric neurologists, three pediatric rehabilitation physicians and three neurologists with an expertise in movement disorders) and worked at different institutions all over the Netherlands. Clinicians were carefully selected to enable a fair comparison between the three disciplines. Firstly, all clinicians spend at least six months to one year of their neurology or rehabilitation training in the pediatric department. Secondly, all nine work with pediatric patients in their daily practice. The pediatric neurologists and rehabilitation physicians only work with children and young adults, and the movement disorder experts work with pediatric as well as adult patients. Every group consisted of one experienced clinician (>15 years of post-training experience) and two younger experts (≤10 years of post-training experience). Together they had a mean of 9.8 years (range 2–29) professional experience in their field.

2.4. Phenotypic classification

The nine clinicians were asked to independently classify the CP symptoms of the fifteen patients based on the videotaped assessment. Except from the videos, no other clinical information was provided. To indicate the phenotype, the assessors divided a total of 100 points between the three motor symptoms, i.e. spasticity, dyskinesia and ataxia. For example, a patient could be classified as 40% spasticity, 60% dyskinesia/dystonia and 0% ataxia. Secondly, clinicians were asked localize the described symptoms in seven different body regions (head, neck, trunk, right arm, left arm, right leg, left leg). Thirdly, overall severity per symptom was indicated, using the global clinical impression (GCI) scale ranging from 1 (symptom absent) to 7 (among the most severe spectrum). After a three months interval, eight randomly selected videos were rated again by the nine clinicians.

We determined the inter- and intra-observer agreement on the predominant symptom, defined as the symptom with the highest percentage, for the whole group and the three subgroups (pediatric neurologists, rehabilitation physicians and movement disorder specialists).

2.5. Statistical analysis

The results were analyzed using descriptive statistics, percentage agreement and Fleiss' kappa and Cohen's kappa for inter- and intra-observer agreement respectively. When describing the results, the common description as provided by Landis and Koch was used: $\kappa < 0.2$ for slight agreement, $\kappa = 0.2–0.4$ for fair agreement, $\kappa = 0.4–0.6$ for moderate agreement, $\kappa = 0.6–0.8$ for strong agreement and $\kappa > 0.8$ for almost perfect agreement.[8]

3. Results

3.1. Patient characteristics

Fifteen children (8 boys, mean age 11.1 y, SD 4.7 y) with an extent of symptoms ranging from gross motor function classification system (GMFCS) 1 (walking without limitations) to 5 (transported in a manual wheelchair) were scored.