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Using both electromyography and movement disorder assessment improved the classification of children with dyskinetic cerebral palsy

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Short title: Systematic assessment of dyskinetic cerebral palsy

Abbreviations: BFM, Burke-Fahn-Marsden Dystonia Rating Scale; CFCS, Communication

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Abstract

Aim: Children with dyskinetic cerebral palsy (CP) are often severely affected and effective treatment is difficult, due to different underlying disease mechanisms. Comprehensive systematic movement disorder evaluations were carried out on patients with this disorder.

Methods: Patients born from 1995-2007 were identified from the Danish Cerebral Palsy Register and referrals to the neuropaediatric centre, Rigshospitalet, Copenhagen. They were classified by gross motor function, manual functional ability, communication ability, dystonia and spasticity. Electromyography was carried out on the upper and lower limbs. Magnetic resonance imaging scans were revised and aetiological searches for underlying genetic disorders were performed.

Results: We investigated 25 patients with dyskinetic CP at a mean age of 11.7 years. Dystonia, spasticity and rigidity were found in the upper limbs of 21, four and six children, respectively, and in the lower limbs of 18, 18 and three children. The mean total Burke Fahn Marsden score for dystonia was 45.02 and the mean Disability Impairment Scale level was 38% for dystonia and 13% for choreoathetosis. Sustained electromyography activity was observed in 20/25 children. Stretching increased electromyography activity more in children with spasticity. There were 10 reclassifications.

Conclusion: The children had heterogenic characteristics and 40% were reclassified after systematic movement disorder evaluation.

Key words: cerebral palsy, dystonia, electromyography, genetics, imaging

Key notes

- We carried out systematic movement disorder evaluation, including electromyography, on 25 children with dyskinetic cerebral palsy at a mean age of 11.7 years.
- Dystonia, spasticity and rigidity were found in the upper limbs of 21, four and six children, respectively, and in the lower limbs of 18, 18 and three children.
- The children with dyskinetic cerebral palsy had heterogenic characteristics and 40% were...
reclassified after evaluation.

**INTRODUCTION**

Cerebral palsy (CP) is caused by damage to the developing brain early in life. It occurs in about two out of 1,000 infants$^{1,2}$ and 10% of cases are classified as dyskinetic CP. Individuals with dyskinetic CP are often severely affected by the condition and the treatments used for other types of CP often have no, or a limited, effect because the disease mechanisms are not the same.$^{3,4}$ Therefore, correct classifications and precise descriptions of dyskinetic movements are essential for systematic tests of relevant treatment.$^4$ This systematic approach to the patient’s movement disorders can be difficult, due to the inherent fluctuating symptoms of dyskinetic CP. Furthermore, it is vital to describe whether the movement disorder changes over time, as some studies have shown that a changing phenotype may imply that there is an unrecognised underlying, and possibly progressive, disorder.$^{5,6}$

The causes of dyskinetic CP and other types of CP, including the more prevalent spastic CP and the equally rare ataxic CP, are profoundly different. In some cases, the cause is obvious, such as asphyxia during labour, cerebral bleeding or infection, but in many cases the cause is unknown. Despite this, knowledge gained during the last decade, and improved diagnostic methods, have enabled us to identify the underlying causes in an increasing number of individuals with dyskinetic CP. For example, it has become clear that several treatable metabolic and genetic diseases, such as dopa-responsive dystonia, may be misdiagnosed as CP.$^{7,8}$ Identifying the cause makes it possible to offer genetic counselling and specific treatment, such as nutrient-specific diets or medication, which can interrupt disease progression and avoid increasing damage.$^{6,9}$ Imaging methods can localise damage in the basal ganglia and make it possible to use specific anti-dystonic medication. It can also be possible to use invasive neuromodulation with deep brain stimulation in cases with basal ganglia damage and severe motor disorder symptoms. However, the symptoms also fluctuate in these cases and classifying and evaluating treatment is difficult. One way to evaluate disease severity over time is to use a combination of imaging, genetic testing and systematic video-based
evaluations of the motor manifestations of dyskinesia.

It has been difficult to quantify muscle overactivity in clinical settings due to the lack of objective quantification methods.\textsuperscript{10} Objective assessment instruments for spasticity,\textsuperscript{11} contractures\textsuperscript{12} and spastic dystonia\textsuperscript{13,14} are now available in clinical settings and these use electromyography and biomechanical evaluation.

As far as we know, the relationship between objective measurements of these phenomena and the classification of dyskinetic movements has not previously been investigated. Our aim was to provide a comprehensive insight into children classified with dyskinetic CP. We assessed movement disorder by using systematic video-based evaluation tools and electromyography. Our hypothesis was that this evaluation would lead to improved descriptions of children diagnosed with dyskinetic CP. In addition, we revised magnetic resonance imaging (MRI) scans and carried out an aetiological search for underlying genetic disorders.

**METHOD**

**Participants**

Participants were identified from the national Danish Cerebral Palsy Register\textsuperscript{15} and cases referred to the tertiary neuropaediatric centre at Copenhagen University Hospital, Rigshospitalet, for a dyskinetic CP assessment. The Register is based on CP cases validated by experienced child neurologists when children are 4-5 years of age, as defined by the Surveillance of Cerebral Palsy in Europe (SCPE)\textsuperscript{1}. The CP subtype was assessed by the clinician involved in the assessment for the CP register or the clinician who referred the patient to our specialist hospital centre. They had all been instructed to use the SCPE definition of topography and the most prevalent motor feature, namely spasticity, dystonia, choreoathetosis, ataxia or non-classifiable. Participants were identified and included in the study if they were born between 1995-2007 and were living in the Copenhagen region at the time of the study. Their medical files were re-evaluated and updated, as necessary, during the study.

**Assessment**
After parental consent, the families were assessed by a child neurologist (PB or CHH) and a neurological physiotherapist (JL). It took approximately 1.5 hours to administer the assessment battery to the patient and a further 30 minutes for the two observers to score the video.

CP impairments were classified using the Growth Motor Function Classification System (GMFCS),\textsuperscript{16} the Manual Ability Classification System (MACS)\textsuperscript{17} and the Communication Function Classification System (CFCS).\textsuperscript{18} The GMFCS describes functional abilities for independent sitting, walking and the need for assistive technology. The MACS classifies manual handling of objects and is based on the child’s self-initiated ability and need for assistance or adaptations. The CFCS classifies ease of communication and effectiveness in everyday life.

**Movement disorder rating**

The movement disorder was rated by systematically examining the videos and consensus-based non-blinded scoring by the first author (JL) and another colleague (CHH or PB). We used several classification systems: the Burke-Fahn-Marsden Dystonia Rating Scale (BFM), the Dyskinesia Impairment Scale (DIS) and the Hypertonia Assessment Tool (HAT). The BFM protocol was used to evaluate dystonia during rest and during action, such as high velocity stretches.\textsuperscript{19} Scores on the severity of nine individually assessed body regions, and the impact that action had on them, were rated from 0-4. The total BFM raw score ranged from 0-120, with a higher score denoting more severe dystonia. The DIS scale consisted of two subscales, dystonia and choreoathetosis, which evaluated duration and amplitude in 12 body regions, during rest and during action. Adding the scores for these regions together provided a total action score of 0–192 and a total rest score of 0–96 for both subscales. The total DIS score was the sum of the dystonia and choreoathetosis subscales.\textsuperscript{20} The HAT scale identified the paediatric hypertonia subtypes of spasticity, dystonia and rigidity.\textsuperscript{21} There were seven items in total. A positive score in at least one item in a subgroup defined the presence of hypertonia for that particular subtype.

**Aetiological classification**

The children were classified according to SCPE and evaluated by a child neurologist who carried out a full neurological examination and evaluated the signs of metabolic or syndromic disorder. MRI was systematically evaluated by a trained neuroradiologist (ARL) who systematically scored
the features listed in Table 3. Genetic and metabolic testing and imaging were performed if they had not previously been carried out or if they were clinically indicated.

**Neurological examination**

The children were assessed by an experienced neurological physiotherapist (JL). Assessments were performed with the children lying on an examination table as comfortably as possible. The ankle, knee and elbow joints were evaluated for range of movement, reflexes and spasticity using the modified Ashworth scale.\(^{22}\) The range of movement was evaluated by slowly moving the joints through the passive movement range, noting both the end positions and any resistance. Fast stretches were applied to evaluate the presence of a spastic catch and, or, clonus. The presence and possible exaggeration of the Achilles, patellar and biceps tendon reflexes were evaluated using a tendon reflex hammer, while gently supporting the limb to enable relaxation.

**Electromyographic recording**

Electromyography recordings were obtained from either the biceps brachii muscle or the soleus muscle using N-10-A/25 Ambu Blue bipolar surface electrodes (Ambu Ltd, Cambridgeshire, UK). The recording area was 0.5cm\(^2\) and the inter-electrode distance was 2cm. We chose to record electromyography from the child’s most affected biceps or soleus muscles, based on modified Ashworth scale scores. Two types of electromyography recordings were obtained from the investigated limbs: electromyography recordings during two minutes of attempted rest and electromyography recordings during high velocity stretches of the assessed limb. When possible, the two minutes electromyography recordings during rest were obtained using three different positions. For the elbow joint these were a fully flexed, a 90-degree and a fully extended position of the elbow joint and for the ankle joint these were a fully plantarflexed, a 90-degree and a fully dorsiflexed position of the ankle joint. Five high velocity stretches of the assessed limb were performed with an angular velocity of more than 300 degrees per second in accordance with the method described by Yamaguchi et al.\(^{11}\) Electromyography recordings were obtained during rest in order to objectively investigate signs of dystonia and high-velocity muscle stretches were used to investigate signs of spasticity. Furthermore, the recordings during rest were lengthy and performed on different joint positions, in order to quantify the patient’s response to maintained muscle stretch and joint positions.
During measurements of the biceps muscle, a set of electrodes was placed proximally and medially on the short head of the biceps brachii and a reference electrode was placed over the lateral epicondyle of the humerus. For measurements of the soleus muscle, a set of electrodes was placed over the soleus muscle belly distal to the medial head of the gastrocnemius muscle and a ground electrode was placed on the distal part of tibia. To minimise electromyography noise factors, the skin was softly sanded with a very fine grain 3M red dot sandpaper (3M A/S, Glostrup, Denmark). Electromyography recordings were made using the Movotec Portable Spasticity Assessment Device (Movotec A/S, Charlottenlund, Denmark) fitted to a forearm orthosis during biceps measurements and an ankle foot orthosis during soleus measurements. The device contained two electromyography channels and was sampled at 1024 Hz. The technical properties of the device and method have previously been explained more thoroughly by Yamaguchi et al.¹¹ and Forman et al.¹⁴

**Analysis of electromyographic recordings**

All two-minute electromyography recordings of attempted rest were visually examined and recordings that contained continuous muscle activity for at least 30 seconds were identified. To separate muscle activity from electrical background noise, we sought to identify a steady presence of electromyography spikes with a high amplitude and firing frequency and considerable variations in the firing pattern. All the electromyography recordings of sustained involuntary muscle activity that were identified were rectified and a root mean square of the full recording was calculated. A linear regression analysis was fitted to each individual recording to investigate whether the level of sustained involuntary muscle activity tended to increase or decrease during the recordings. A slope coefficient of the linear regression was then calculated. A positive slope coefficient indicated a linear increase in root mean square electromyography over 120 seconds and a negative slope coefficient indicated a decrease in root mean square electromyography. To assess the effect of the joint position on sustained involuntary muscle activity, all recordings were individually normalised to the recording obtained in the maximally flexed position. This was carried out to enable us to group individuals together and to evaluate stretch sensitivity across the groups, despite differences in raw electromyography amplitudes. Further details of the analysis have previously been described by Forman et al.¹⁴

The stretch-induced ratio of the electromyography and background electromyography was calculated from the electromyography recordings during high-velocity stretches. Background
electromyography was defined as the average rectified electromyography amplitude in the 500ms period before the stretch. Stretch-induced electromyography was defined as the average rectified electromyography amplitude in the period immediately following the stretch (Fig. 1). A high ratio described a large electromyography response following the stretch. Statistical tests were not performed due to the small sample size.

The project was approved by The National Committee on Health Research Ethics (H-16028528), The Danish Data Protection Agency (journal number SUND-2017-27) and The University of Southern Denmark (journal number 16/3508).

RESULTS

Participant characteristics
We studied 25 children (17 female) with a mean age of 11.7 (range 3.9-17.9) years who had previously been diagnosed with dyskinetic CP (Table 1). The assessments led to 10 reclassifications: three of mixed dystonic/spastic CP, one of ataxic CP and three of unclassifiable CP. In three cases, Aicardi-Goutiere syndrome had been diagnosed, which did not fulfil the criteria for CP, as it is a progressive disorder. The frequency distribution of the GMFCS levels was: I (n=2), IV (n=9) and V (n=14). The MACS levels were: II (n=2), IV (n=5) and V (n=18). The CFCS levels were: I (n=1), II (n=2), IV (n=9) and V (n=13). The aetiology was: birth asphyxia (n=11), chromosomal abnormality (n=7), kernicterus (n=1), congenital cytomegalovirus infection (n=1), prematurity (n=1) and unknown aetiology (n=4).

There was one set of twins and the remaining children were from singleton pregnancies. Their height at the time of the evaluation, according to Danish height curves (www.vækstkurver.dk), was ≤ 2 SD in 18/24 participants. Data were missing for one participant. Vision was normal in 11 children, moderately affected in four and severely reduced in 10. Hearing was normal in 23 children. The other two had severe impairment and one had a cochlear implant. Epilepsy was present in 10 children and being treated with an antiepileptic drug. Five other children had a history of epilepsy. Comorbidities comprised six cases of respiratory insufficiency, two cases each of scoliosis, autism, reflux and dyspraxia and one case of hypophyseal insufficiency.

Medication
Attempted treatments with muscle relaxants, antispasmodic or antidystonic drugs were highly
variable and six children had not received any of these treatments. Oral baclofen was continuously used by seven children and intermittently by two. An intrathecal baclofen pump was being used by four children and another had the pump removed due to infection. Treatment with botulinum toxin was used continuously by nine children and had previously been attempted, but proved ineffectual in another nine cases. Orthopaedic procedures had been performed on six of the 25 participants.

**Video-based evaluation**

The HAT assessment demonstrated upper limb dystonia as a solitary symptom in 13/25 children or in combination with spasticity or ataxia in 8/25 participants (Table 2). Lower limb dystonia was the only symptom in nine children and combined with spasticity or rigidity in 9/25. None showed signs of upper limb spasticity as the only symptom, but five showed lower limb spasticity as the only symptom. Spasticity combined with dystonia and, or, rigidity was found in 4/25 of the children’s upper limbs and 13/25 of their lower limbs. Signs of rigidity were found in the upper limbs of six cases and in the lower limbs of three cases, but only combined with dystonia and, or, spasticity.

The median BFM movement score was 47 (range 0-85) and the median disability score was 28 (range 0-29). The median limb score for the limb selected for electromyography recording was 12 (range 0-16).

The mean DIS level for dystonia was 38% (range 0-72) and it was 13% (range 0-60) for choreoathetosis (Table 2).

**Electromyography recordings during rest**

Sustained electromyography activity was observed, during rest, in 20/25 children in at least one two-minute recording from the muscle that we assumed was most affected: 16 biceps brachii and four soleus. As illustrated in Figure 1, the sustained electromyography activity demonstrated several different patterns. Subject six had near-constant electromyography activity with frequent large bursts. In contrast, subject nine had constant electromyography activity with few clear bursts of activity, but with a large decrease in the extended joint position. Subject 22 had long periods without electromyography activity, but with sudden large bursts in the 90-degree and extended joint positions.
In total, 43 two-minute recordings from the 20 children contained sustained electromyography activity. The children without observed sustained electromyography activity were cases three, 11, 13, 15 and 22 (Table 2). Recordings were obtained from all three different joint positions in 18/20 children with sustained electromyography. Therefore, these recordings were grouped to investigate whether the position of the joint influenced the sustained electromyography activity on a group level. The root mean square for sustained electromyography activity, which was normalised and logarithm transformed, did not change significantly at a group level from the flexed elbow position to either the 90-degree elbow position (-0.02±0.29, p=0.83) or to the extended elbow position (-0.05±0.39, p=0.62). The linear regression slope of the individual 43 recordings of sustained electromyography activity was positive in 19 recordings (44%) and negative in 24 recordings (56%).

The presence of sustained electromyography activity was related to the individual child’s clinical dystonia ratings (Table 2). Of the 21 children who had dystonia with the HAT rating, either alone or with another feature of hypertonia, 18 had sustained electromyography activity. Two of the four cases who did not have dystonia with the HAT rating had sustained electromyography activity. The DIS scale showed that the 20 children with sustained electromyography activity had higher levels of dystonia of the investigated limb than the five children without sustained electromyography activity (55.7±30.4 versus 37.9±31.8). When the BFM movement scale was used, the average movement scores of the investigated limb were 10.4±5.1 and 8.0±6.7, respectively.

**Electromyography recordings during high-velocity stretches**

We obtained high-velocity stretches of the assessed limb for 18/25 participants and the ratio for stretch-induced electromyography and background electromyography was calculated. Participants were divided into spasticity and non-spasticity groups according to the HAT rating of the assessed limb. The five children with spasticity had an average ratio of 16.3 and the 13 without spasticity had an average ratio of 4.3 (Figure 2). This could indicate that the electromyography response to the stretch was larger for the total spasticity group than the non-spasticity group. The applied velocity of the stretches were all above 300 degrees/s, with a range of 350–456 degrees/s for the non-spasticity group and 359-481 degrees/s for the spasticity group.

**Imaging and genetic and metabolic testing**

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Magnetic resonance imaging (MRI) was performed on 23/25 children and one of those 23 had an additional MRI. Images were available for 16 of the children and were re-evaluated by a trained neuroradiologist (ARL) (Table 3). Basal ganglia signal changes were found alone in two children and in combination with other features in seven children. Malformations were present in 6/7 children: one case of holoprosencephaly with schizencephaly, four cases of corpus callosum agenesis or hypoplasia and one case of a hypoplastic unilateral cerebellar hemisphere. The MRI scans were normal in the other child. There was also sustained electromyography activity and dystonia, identified with the HAT score, in all seven cases with basal ganglia signal changes on their MRI scans.

Metabolic or genetic testing had been performed on 11/25 children prior to our study. Aetiology was found in five children, as three had Aicardi-Goutieres syndrome, one had holoprosencephaly caused by del13q32.3 and one had congenital cytomegalovirus. We carried out additional testing on six children. A whole exome sequencing was performed and one child also underwent array comparative genomic hybridisation. Three children had a normal exome. Three children had their aetiology detected by exome sequencing. One child had a pathogenic variant in GRIN2A: c.1904C>A, p.(ala635Asp), one in SOX2 c.764_767dup, p.(Ser257Tyrfs*54) and one was compound heterozygous for p.Glu2526Asnfs*2 og p.Arg2426Ter in the SACS gene. One other child was offered further genetic testing, but the parents declined.

DISCUSSION
We analysed a cohort of 25 children, who prior to the study had been diagnosed with dyskinetic CP. The extensive evaluation showed that the children were a very heterogenic population with various aetiologies. In general, the participants were severely affected by their movement disorder and, after systematic testing, we were able to measure dyskinesia with the video-based BFM, HAT and DIS scales and by dynamometer electromyography recording. The evaluation was completed by a genetic and metabolic review, additional testing and a systematic review of MRI scans. The thorough assessment carried out on the patients enabled us to evaluate their treatment response and re-classify 10/25 into non-dyskinetic types of CP. A genetic aetiology was found for three participants who previously did not have a known aetiology.
The approach we used in this study was applicable for systematic testing and was based on movement disorder classification scales. It was different to previous studies, as we added electromyography recordings to objectively measure muscle activity in a dynamometer-based assay. This assay has previously been established for measuring spasticity in adults and children with CP.\textsuperscript{11-13} To our knowledge, no one has reported dynamometer-based electromyography measures to the clinical examination of a population with dyskinetic CP. The use of electromyography measures enabled us to find sustained involuntary muscle activity in 20/25 of the investigated children. Stewart et al published an evaluation of papers on dyskinesia and choreoathetosis scales. They concluded that further studies were required to fully examine the specific validity, reliability, responsiveness and clinical use of each scale for children with CP.\textsuperscript{23} We agree with those authors that all tools are designed to classify movement disorders at the level of body functions and structures, rather than activity or restrictions on participation. However, many provide some insight into the impact of dystonia on activities. That is why the current study integrated the different scales with an objective tone measurement to extensively evaluate the patients. These included scales that assessed spasticity and rigidity, which often co-exist with dystonia in many patients. Three children with a HAT rating for dystonia did not have sustained involuntary electromyography activity of the investigated limb. In contrast, two children who did not have a HAT dystonia rating exhibited sustained involuntary muscle activity. Furthermore, comparable DIS and BFM ratings in the limbs that were investigated using electromyography were found in the groups with and without sustained involuntary muscle activity. These findings support the use of electromyography validation in the complex diagnosis and treatment of dystonia. The Barry Albright dystonia scale was not included in this study, as a high correlation with the broader Dyskinesia Impairment Scale has been documented.\textsuperscript{20}

Reid et al published a description of the distribution of predominant and secondary motor types and compared functional profiles, comorbidities, and brain imaging patterns between dyskinetic and spastic CP.\textsuperscript{24} Our findings correlated well with these, as we also found that re-evaluation led to re-classification in many cases or to detecting different movement disorder patterns that were more prevalent at different ages. This paper, and several others, have documented that children with dyskinetic CP have been severely affected. Our nationwide register-based study of dyskinetic CP, published in 2019, found that children with dyskinetic CP were more likely to have a severe GMFCS level, namely level III-V, than children with spastic CP (90.0% versus 66.0%). The same
study showed that children with dyskinetic CP had high rates of reduced developmental quotient (68.1%), visual impairment (39.3%) and epilepsy (51.6%).\(^{25}\)

A limitation of our study, and others, was that we assessed a relatively small cohort with a diverse aetiology. However, this mirrors clinical practice and is an inherent property of dyskinetic CP diagnoses. Other limitations were that patients were treated with various tone reducing medications and were of different ages. These limitations suggest that the electromyography dynamometer tool should be tested on a larger cohort before being used in clinical practice. Another limitation was the classification system that was applied by those who initially assessed the children. This was in line with SCPE, but it implied that the most evident movement pattern was used. We have found that many patients with dyskinesia have also had abundant spasticity. In our study, the \textit{a priori} CP subtype was determined by local neuropaediatricians and therefore lack of training may be the reason for some re-classifications after our more detailed assessments. Evaluations of patients with dyskinetic CP would be strengthened by testing how they perform during real-world daily activities instead of in an outpatient clinic. The aim of testing would preferably be to evaluate treatments where the focus was on goals chosen by patients. The impact of dyskinesia changes considerably according to a patient’s environment and a specific treatment or training can only focus on some tasks. However, it is important to measure the level of movement disorder in the affected regions. Our study found that electromyography recordings could be particularly helpful in specific treatment settings or if a clinician was unsure whether a patient had dystonia or spasticity. However, it could be less suitable for daily clinical practice, due to both the time required and the experience needed by examiners.

Genetic disorders and treatable metabolic disorders can present with symptoms of CP. Novel treatment options are being tested, including tetrabenazine.\(^ {26}\) This is one example of where a systematic evaluation, preferably with an objective measure such as electromyography, could be applied. There are many reports in the literature of specific single diagnoses underlying dyskinetic CP, but the collective incidence of treatable inborn errors of metabolism mimicking CP is unknown and can only be determined by systematic or large-scale screening studies. However, studies have indicated that genetic testing cannot stand alone, even with extensive genetic sequencing, and that these disorders can only be diagnosed if a thorough clinical evaluation goes hand-in-hand with metabolic and genetic testing.\(^ {9}\) Increasing awareness among clinicians is worthwhile, as timely

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diagnosis and appropriate treatment can improve the primary features or stabilise and prevent further neurological sequelae and decline. An underlying, non-progressive genetic aetiology was found in three participants in this study. This will be of special interest for families, as it can help to explain their child’s condition. It also has implications for what kind of follow up is needed, the patient’s prognosis and genetic counselling if the parents want further children. We strongly support repeated genetic testing, as our knowledge of this area is constantly increasing.

CONCLUSION
Children with dyskinetic CP need to undergo multi-faceted testing that evaluates their current status, as this forms the basis for evaluating their treatment response and helps us to investigate the aetiology of their CP. Using several tests is the best way to evaluate fluctuating and diverse symptoms and our experience shows that electromyography recording is a useful supplementary analysis.

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CONFLICTS OF INTEREST
The authors have no conflicts of interest to declare.

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Legends for figures and tables

**Figure 1.** A. Electromyography during high-velocity stretching. This example is from patient four, with a 12.4 stretch-induced electromyography/background electromyography (EMG) ratio. B. Examples of electromyography recordings from three different participants in the three investigated joint positions. These recordings were all obtained from the biceps brachii muscle of the most affected side. The three participants exemplify some of the different patterns of sustained electromyography activity that can be observed from an individual muscle. The electromyography is measured in microvolts (μV). The duration of each recording was 120 seconds.

**Figure 2.** Stretch-induced electromyography/background electromyography (EMG) ratio in the group with and without spasticity, based on the hypertonia assessment tool (HAT) rating. Black dots depict individual participants and red lines depict group averages.

**Table 1.** Clinical characteristics, background data and associated impairments of the 25 included patients who had a diagnosis of dyskinetic CP before entering the study. Any re-classifications of the revised diagnoses are listed. Abbreviations: BW, birth weight; CP, cerebral palsy; GA, gestational age; It, intrathecal; N, none/normal; na, not analysed; pn, used at need; prev, previously; resp. insuff, respiratory insufficiency; Y, yes.

**Table 2.** Systematic evaluation with movement disorders scales and electromyography results. The degree of gross motor, fine motor and communication impairment is first listed with GMFCS, MACS and CFCS. This is followed by the investigation of contractures and electromyography recordings. The dystonia/hypertonia scale assessments by BFM, HAT and DIS are listed on the right hand side of the table. Abbreviations: CFCS, communication function classification system; BFM, Burke-Fahn-Marsden dystonia rating scale; D, dystonia; EMG electromyography; GMFCS, gross motor function classification system; HAT, hypertonia assessment tool; MACS, manual ability classification system; UE, upper extremity; LL, lower limb; R, rigidity; S, spasticity.

**Table 3.** Systematic re-evaluation of available MRI images with scores for the features listed.

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Abbreviations: DWI, diffusion weight imaging; L, left; MRI, magnetic resonance imaging; N, normal; na, not analysed; R, right; Y, yes.
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**Supplementary Table.** MRI findings. Abbreviations: DWI=diffusion weight imaging; L=left; MRI=magnetic resonance imaging; N=normal; na=not analysed; R=right; Y=yes.

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