The Dyskinesia Impairment Scale: a new instrument to measure dystonia and choreoathetosis in dyskinetic cerebral palsy

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ABBREVIATIONS

BADS	Barry–Albright Dystonia Scale
DIS	Dyskinesia Impairment Scale
ICC	Intraclass correlation coefficient
MDD	Minimal detectable difference
SCPE	Surveillance of Cerebral Palsy in Europe

AIM The aim of this study was to examine the reliability and validity of the Dyskinesia Impairment Scale (DIS). The DIS consists of two subscales: dystonia and choreoathetosis. It measures both phenomena in dyskinetic cerebral palsy (CP).
METHOD Twenty-five participants with dyskinetic CP (17 males; eight females; age range 5–22y; mean age 13y 6mo; SD 5y 4mo), recruited from special schools for children with motor disorders, were included. Exclusion criteria were changes in muscle relaxant medication within the previous 3 months, orthopaedic or neurosurgical interventions within the previous year, and spinal fusion. Interrater reliability was verified by two independent raters. For interrater reliability, intraclass correlation coefficients were assessed. Standard error of measurement, the minimal detectable difference, and Cronbach's alpha for internal consistency were determined. For concurrent validity of the DIS dystonia subscale, the Barry–Albright Dystonia Scale was administered.
RESULTS The intraclass correlation coefficient for the total DIS score and the two subscales ranged between 0.91 and 0.98 for interrater reliability. The reliability of the choreoathetosis subscale was found to be higher than that of the dystonia subscale. The standard error of the measurement and minimal detectable difference values were adequate. Cronbach's alpha values ranged from 0.89 to 0.93. Pearson's correlation between the dystonia subscale and Barry–Albright Dystonia Scale was 0.84 ($p<0.001$).
INTERPRETATION Good to excellent reliability and validity were found for the DIS. The DIS may be promising for increasing insights into the natural history of dyskinetic CP and evaluating interventions. Future research on the responsiveness of the DIS is warranted.

Cerebral palsy (CP) is worldwide the most common neuromotor disorder in children, with an incidence of 2 to 3 per 1000 live births.[1,2] CP can be categorized into spastic, dyskinetic, and ataxic groups. Dyskinetic CP is further differentiated into dystonia and choreoathetosis.[1,3]

Spastic CP is by far the most common type of CP, with a prevalence of approximately 80%,[4] and is followed by dyskinetic CP with a prevalence between 6.5%[5] and 14.4%.[4] According to the Surveillance of Cerebral Palsy in Europe (SCPE),[6] dyskinetic CP is characterized by involuntary, uncontrolled, recurring, occasionally stereotyped movements, in which the primitive reflex patterns predominate and muscle tone varies.[6] The SCPE described dystonia in CP as dominated by abnormal postures that may give the impression of hypokinesia and muscle tone that is fluctuating (but with easily elicitable tone increase). Characteristics are involuntary movements, distorted voluntary movements, and abnormal postures due to sustained muscle contractions. Choreoathetosis in CP is dominated by hyperkinesia and tone fluctuation (but mainly decreased). Chorea refers to rapid, involuntary, jerky, often fragmented movements. Athetosis means slower, constantly changing, writhing, or contorting movements.[6,7] These SCPE descriptions are in accordance with the recently published definitions of dystonia, chorea, and athetosis by the Taskforce on Childhood Movement Disorders.[8,9] The Taskforce defines dystonia as a movement disorder in which involuntary sustained or intermittent muscle contraction causes twisting and repetitive movements, abnormal postures, or both,[8] chorea as an ongoing, randomly appearing sequence of (one or more) discrete involuntary movements or movement fragments, and athetosis as a slow continuous, involuntary writhing movement that prevents maintenance of a stable posture.[9] The definitions of the SCPE and the Taskforce describe dystonia and choreoathetosis in a very similar way and are essentially

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descriptive, based on consensus emerging from experts from different clinical and basic fields of science.

Over the last few years, there has been continuing development of interventions in children with dyskinetic CP, including intrathecal baclofen,[10–12] deep brain stimulation,[13,14] oral medication,[15–17] ventral rhizotomies,[18] and botulinum toxin injections.[19] However, objective evidence supporting these interventions is only preliminary. Specific assessment of dystonia has mostly relied on the Barry–Albright Dystonia Scale (BADS).[20] Operationally, the BADS has become a criterion standard for scoring dystonia in CP, but several studies[10–17] have reported the difficulty of measuring dystonia reliably and/or questioned the sensitivity of the BADS.

In a recent study,[21] the reliability and validity of the BADS was reassessed and special attention was given to the sensitivity of the scale. This study showed reliability results similar to those of Barry et al.[20] but also revealed limitations in the sensitivity of the BADS.

Content analysis showed that the BADS included several dystonia characteristics over eight body regions. However, the items are a combination of several different dystonia characteristics within one score (e.g. duration and amplitude) and no differentiation is made between rest and activity. Also, for the first time, the measurement error of the BADS was assessed and a high standard error of measurement (SEM) and minimal detectable difference (MDD) were found, respectively 6% and 18%. In clinical practice, this means that a score difference of 18% is necessary to ascertain that 'true' improvement has occurred, as lower values might be ascribed to measurement errors. Also in this study,[21] two primary dystonia scales were evaluated in dyskinetic CP, namely the Burke–Fahn–Marsden Movement Scale[22] and the Unified Dystonia Rating Scale.[23] For these scales, even higher MDDs were found, 27% and 25% respectively.[21] Finally, several groups have emphasized that dystonia and choreoathetosis often occur concurrently in dyskinetic CP[9,21,24] However, to our knowledge no standardized tools for measuring choreoathetosis in CP have been validated.

For these reasons, we have strived to develop a new assessment tool to score dystonia and choreoathetosis at rest and during activity in individuals with dyskinetic CP. We attempted to enhance the sensitivity of this tool in comparison with the commonly used dystonia scales. In this paper, we describe how we developed the DIS and assessed its reliability and validity.

What this paper adds

- Good to excellent reliability and validity was found for a new clinical scale evaluating dyskinesia in cerebral palsy.
- This is the first scale that independently measures dystonia and choreoathetosis in dyskinetic cerebral palsy.
- The reliability of the choreoathetosis subscale was found to be higher than that of the dystonia subscale.

METHOD

Development of the dyskinetic impairment scale

One of the first steps in the development of the DIS consisted of a content analysis of the three available secondary and primary dystonia scales.[20,22,23] In accordance with Sanger et al.,[9] movements can be described by the context in which they occur, for example postural, rest, action, or associated with specific tasks.[9] Dyskinesia characteristics can be assessed at rest and during activity and in terms of duration, amplitude, and influence on functional activities. From this point of view, content analysis revealed that the three scales analysed made limited or no differentiation between action and rest or duration and amplitude, and combined several dyskinesia characteristics within one score, which may limit the sensitivity of the scales (see Table I). Additionally, we explored the content and scale construct of the Toronto Western Spasmodic Torticollis Rating Scale[25] and the Unified Parkinson's Disease Rating Scale[26] (Movement Disorder Society). Based on this analysis and the SCPE definitions of dystonia, choreoathetosis, and dyskinetic CP,[3,6,7] the DIS was developed according to the methodological framework of Kirshner and Guyatt.[27] Its content was thoroughly discussed with a clinical expert team (EO, JD, HF, PD, and FR) from the Cerebral Palsy Reference Centre (University Hospital Pellenberg, Leuven, Belgium).

In a second step, the interrater reliability of this scale was assessed in a pilot study. Four physical therapists with extensive clinical experience of children with CP (UH, IV, ES, and ED) underwent a training session with the reference and training DVD of the SCPE[6] and were instructed on how to use the preliminary constructed scale. They then scored 10 videotaped children with dyskinetic CP independently. Afterwards, the content of the scale, the included items, and the scoring criteria were discussed with these four raters and the clinical expert team. Subsequently, the discussion together with (1) the number of participants able to accomplish the task, (2) the reliability of the item scores, and (3) the participants' clinical experience, ensured that an item reduction was obtained and that the scoring criteria and instructions were revised.

The final DIS (Appendix I, supporting information published online) consists of two subscales, one for dystonia and one for choreoathetosis (see Fig. 1). Both subscales evaluate duration and amplitude in 12 body regions including the eyes, mouth, neck, trunk, and limbs. For the limbs, a distinction is made between the proximal and distal region and between the right and left side. For each of the assessed body regions, the duration refers to the amount of time that dyskinesia is present, whereas the amplitude aspects refer to the range of motion of the dyskinetic movements. All body regions are scored during two activities (action) and one resting posture (rest). Summation of the region scores gives a total action score (range 0–192) and a total rest score (range 0–96) for both subscales. The action and rest scores add up to a total score for dystonia and choreoathetosis, each with a range from 0 to 288. The total DIS score is the sum of the dystonia and choreoathetosis subscale.

Reliability and validity

Participants

This study included 25 participants aged between 5 and 22 years (17 males; eight females; mean age 13y 6mo; SD 5y 4mo). All participants were diagnosed by a paediatric neuro-

Table I: Characteristics of the Burke–Fahn–Marsden Movement Scale (BFMS), the Unified Dystonia Rating Scale (UDRS) the Barry–Albright Dystonia Scale (BADS), and the Dyskinesia Impairment Scale (DIS)

						Discrimination		
Scale	Primary dystonia	Secondary dystonia	Choreoathetosis	Body regions[a]	Video scoring	Action/ rest	Duration/ amplitude	Proximal/ distal limbs
BFMS	+	–	–	9	+	–	–	–
UDRS	+	–	–	14	+	–	+	+
BADS	–	+	–	8	+	–	–	–
DIS	–	+	+	12	+	+	+	+

[a]Number of items. +, present; –, absent.

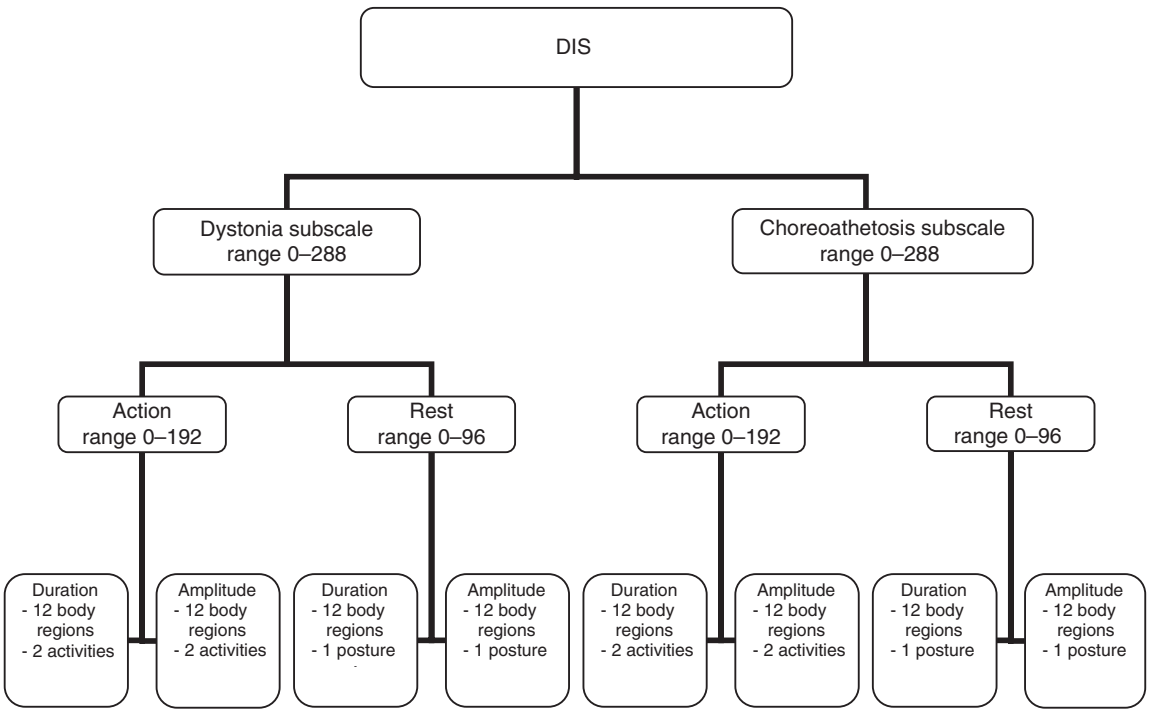

Figure 1: Diagram of the Dyskinetic Impairment Scale.

logist and were recruited from special schools for children with motor disabilities. Individual participant characteristics are presented in Appendix II (supporting information published online). Exclusion criteria were changes in muscle relaxant medication within the previous 3 months, orthopaedic or neurosurgical interventions within the previous year, and spine fusion. Ethical approval was obtained from the Ethical Committee of the Katholieke Universiteit Leuven. All participants and/or their parents provided informed consent.

Procedure

Based on the recommendations of the Dystonia Study Group,[23] the 25 participants were videotaped (by ES and MV) according to a standard video protocol. It contained all postulated activities and rest postures of the DIS (see Appendix III, supporting information published online). An effort was made to provide relaxing surroundings. All participants were filmed in their habitual environment at school and in the presence of their own physiotherapist. The duration of videotaping was similar to the duration in other dystonia scales (e.g. Unified Dystonia Rating Scale, BADS, Burke–Fahn–Marsden Movement Scale), with a maximum of 30 minutes. The passive range of motion of the upper and lower limb joints was measured with a goniometer to serve as a baseline for the amplitude assessment of the DIS. Afterwards, a video montage was made in accordance with the scoring order of the DIS.

To assess interrater reliability, two physical therapists (EM, JV) scored all videos in series within 15 days. The two raters had experience in discriminating dystonia and choreoathetosis in CP and were trained in scoring with the DIS.

To assess concurrent validity, the second rater (JV) scored the BADS for all 25 participants. The BADS evaluates dystonia over eight body regions on a five-point ordinal scale. The video protocol was also used to assess the BADS scale.

Statistical analysis

Rigby's statistical recommendations[28] were applied. For interrater reliability, the intraclass correlation coefficients (ICCs) and 95% confidence intervals (CIs) were used for the total scores and item scores of the DIS. Portney and Watkins[29] considered an ICC higher than 0.90 as excellent, an ICC between 0.75 and 0.90 as good, and an ICC<0.75 as poor to moderate. To interpret the ICC scores<0.75, we considered ICC values between 0.60 and 0.75 as moderate and less than 0.60 as poor. The SEM and MDD were calculated using the formula $SEM=SD\times\sqrt{(1-ICC)}$ and $MDD=SEM\times1.96\times\sqrt{2}$.[29] The internal consistency was evaluated by Cronbach's alpha.[29] Concurrent validity was determined by Pearson correlation coefficients. All statistics were calculated with SPSS 16.0 (SPSS Inc., Chicago IL, USA).

RESULTS

Interrater reliability

The total score of the DIS, the dystonia subscale, and the choreoathetosis subscale showed excellent interrater reliability with ICCs of 0.96 (95% CI 0.91–0.98), 0.91 (95% CI 0.91–0.86), and 0.98 (95% CI 0.95–0.99) respectively.

The ICCs and 95% CIs of the total subscale scores and region scores are presented in Table II.

For the dystonia subscale, ICCs of the total scores of the duration aspect, amplitude aspect, and the summation of both were 0.87, 0.87, and 0.88, respectively, during action and 0.90, 0.94, and 0.93 respectively, during rest. ICCs for the body regions of the duration factor during action were moderate to excellent except for the eyes, neck, and trunk regions. The amplitude aspect showed moderate to excellent reliability for 7 of the 12 regions and lower reliability for the neck, trunk, right proximal arm, and both proximal legs. During rest, moderate to excellent reliability was found for the duration aspect for nine regions and lower reliability for the neck, right proximal leg, and left distal leg. The amplitude aspect presented moderate to high reliability for all regions. For the choreoathetosis subscale, the ICC of the total scores of the duration aspect, amplitude aspect, and the summation of both were 0.97, 0.94, and 0.96, respectively, during action and 0.96, 0.93, and 0.96 respectively, during rest. ICC region scores of the DIS choreoathetosis subscale ranged from moderate to excellent except for the duration of the left distal leg, the eyes amplitude aspect during activity, and the eyes amplitude during rest.

Standard error of measurement and minimal detectable difference

For interrater reliability, the SEM and MDD values for the total DIS were 3% and 9% respectively. The SEM and MDD were 5% and 15% for the DIS dystonia subscale and 3% and 7% for the choreoathetosis subscale.

Table II: Interrater reliability: intraclass correlation coefficients (ICC) with 95% confidence intervals (CI) between raters for the Dyskinesia Impairment Scale

		Active						Rest					
		Duration (*D*)		Amplitude (*A*)		∑(*D*+*A*)		Duration (*D*)		Amplitude (*A*)		∑(*D*+*A*)	
		ICC	95% CI	ICC	95% CI	ICC	95% CI	ICC	95% CI	ICC	95% CI	ICC	95% CI
Dystonia subscale													
1	Eyes	0.50	0.14–0.74	0.55	0.21–0.76	0.63	0.31–0.82	0.75	0.52–0.88	0.84	0.66–0.93	0.79	0.59–0.90
2	Mouth	0.73	0.48–0.87	0.78	0.57–0.90	0.86	0.70–0.93	0.75	0.50–0.88	0.67	0.38–0.84	0.75	0.51–0.88
3	Neck	0.50	0.14–0.75	0.66	0.37–0.84	0.61	0.29–0.81	0.56	0.22–0.78	0.64	0.34-.083	0.65	0.35–0.83
4	Trunk	0.49	0.12–0.74	0.48	0.11–0.73	0.54	0.19–0.77	0.72	0.45–0.86	0.81	0.62–0.91	0.78	0.57–0.90
5	Arm RP	0.62	0.30–0.81	0.47	0.11–0.73	0.51	0.15–0.75	0.88	0.74–0.94	0.81	0.61–0.92	0.87	0.73–0.94
6	Arm LP	0.86	0.71–0.94	0.70	0.43–0.86	0.79	0.57–0.90	0.91	0.80–0.96	0.83	0.66–0.92	0.88	0.75–0.95
7	Arm RD	0.98	0.97–0.99	0.90	0.78–0.95	0.99	0.97–0.99	0.81	0.61–0.91	0.88	0.75–0.95	0.87	0.72–0.94
8	Arm LD	0.99	0.99–1.00	0.99	0.98–1.00	1.00	0.99–1.00	0.69	0.41–0.85	0.86	0.70–0.93	0.79	0.57–0.90
9	Leg RP	0.64	0.33–0.82	0.47	0.10–0.73	0.60	0.28–0.80	0.55	0.21–0.77	0.71	0.44–0.86	0.64	0.34–0.82
10	Leg LP	0.62	0.30–0.81	0.47	0.10–0.73	0.56	0.22–0.78	0.68	0.39–0.84	0.67	0.38–0.84	0.80	0.60–0.90
11	Leg RD	0.70	0.43–0.86	0.65	0.35–0.83	0.75	0.51–0.98	0.70	0.43-.086	0.87	0.72–0.94	0.81	0.62–0.91
12	Leg LD	0.77	0.55–0.89	0.61	0.30–0.81	0.78	0.58–0.90	0.46	0.08–0.72	0.75	0.50–0.80	0.64	0.40–0.82
Total score		0.87	0.73–0.94	0.87	0.72–0.94	0.88	0.74–0.94	0.90	0.79–0.96	0.94	0.87–0.97		0.85–0.97
Choreoathetosis subscale													
1	Eyes	0.60	0.28–0.80	0.50	0.13–0.74	0.57	0.23–0.78	0.71	0.44–0.86	0.48	0.11–0.73	0.67	0.38–0.84
2	Mouth	0.87	0.72–0.94	0.71	0.44–0.86	0.81	0.62–0.92	0.90	0.80–0.97	0.84	0.68–0.93	0.93	0.85–0.97
3	Neck	0.81	0.62–0.91	0.76	0.53–0.89	0.85	0.69–0.93	0.83	0.65–0.92	0.80	0.60–0.91	0.84	0.68–0.93
4	Trunk	0.87	0.72–0.94	0.64	0.50–0.90	0.80	0.60–0.91	0.81	0.61–0.91	0.77	0.55–0.89	0.81	0.62–0.91
5	Arm RP	0.89	0.74–0.94	0.92	0.82–0.96	0.91	0.81–0.96	0.85	0.69–0.93	0.87	0.72–0.94	0.95	0.79–0.96
6	Arm LP	0.91	0.80–0.96	0.88	0.74–0.94	0.92	0.82–0.96	0.84	0.68–0.93	0.89	0.76–0.95	0.86	0.71–0.94
7	Arm RD	0.81	0.61–0.91	0.89	0.76–0.95	0.89	0.75–0.95	0.85	0.70–0.93	0.94	0.86–0.97	0.94	0.87–0.97
8	Arm LD	0.89	0.77–0.95	0.86	0.72–0.94	0.89	0.76–0.95	0.94	0.88–0.98	0.86	0.71–0.94	0.94	0.87–0.97
9	Leg RP	0.81	0.62–0.91	0.81	0.62–0.91	0.81	0.62–0.91	0.96	0.90–0.98	0.73	0.48–0.87	0.90	0.79–0.96
10	Leg LP	0.73	0.73–0.94	0.78	0.56–0.90	0.86	0.70–0.93	0.83	0.65–0.92	0.75	0.52–0.88	0.86	0.71–0.94
11	Leg RD	0.85	0.69–0.93	0.73	0.47–0.87	0.81	0.62–0.91	0.88	0.75–0.95	0.74	0.50–0.88	0.85	0.69–0.93
12	Leg LD	0.59	0.28–0.79	0.60	0.28–0.80	0.64	0.33–0.82	0.92	0.83–0.96	0.79	0.58–0.90	0.88	0.74–0.94
Total score		0.97	0.92–0.98	0.94	0.86–0.97	0.96	0.92–0.98	0.96	0.92–0.98	0.93	0.85–0.97	0.96	0.92–0.98

RP, right proximal; LP, left proximal; RD, right distal; LD, left distal.

Internal consistency

Cronbach's alpha for the dystonia subscale during action was 0.91 for the duration aspect and 0.92 for the amplitude aspect. During rest posture, Cronbach's alpha was 0.90 and 0.93 for duration and amplitude respectively. Similar values were found for the choreoathetosis subscale: 0.92 for duration and 0.90 for amplitude during action, and 0.94 and 0.89 for duration and amplitude respectively, during rest posture.

Concurrent validity

Pearson's correlation between the DIS dystonia subscale and BADS was 0.84 (95% CI 0.66–0.92; $p<0.001$).

DISCUSSION

In this study, the DIS was developed to measure both dystonia and choreoathetosis in dyskinetic CP. These movement disorders are known to be mostly simultaneously present in this participant group.[9] The DIS also allows the measurement of dystonia and choreoathetosis separately. This is important for further determining the dominant type of movement abnormality, as recommended by Rosenbaum et al.[3] The description and definitions of dystonia and choreoathetosis[5–7] were the starting point of the DIS. In accordance with the clinical evaluation recommendations of the Taskforce on Childhood Movement Disorders,[9] we have included several components such as action, rest, duration, and amplitude so that dyskinetic movement disorders could be measured in their predominant presence and the context in which they occur. The DIS measures both dystonia and choreoathetosis, thus allowing the possibility of calculating a ratio between these movement disorders in dyskinetic CP and thereby increasing our insight into the full clinical presentation and natural history of dyskinetic CP. It is well known that the expression of dystonia and choreoathetosis is mostly linked to brain lesions in the basal ganglia.[30] However, their pathophysiology is complex and not fully understood.[31] Therefore, it is hoped that a reliable, valid, and sensitive clinical measurement of dystonia and choreoathetosis may result in the recognition of dyskinesia patterns that can be related to the observed brain lesions, and subsequently may enhance our insight into the pathophysiology of CP in the long term. Such a tool should also help in the evaluation of existing and emerging treatments for children with CP. Furthermore, the differentiation of dystonia and choreoathetosis in the DIS will be particularly important in judging the outcome of medical interventions focusing on one or both clinical symptoms.

In this study, we found excellent interrater reliability for the total score of the DIS and the dystonia and choreoathetosis subscales. All ICCs exceeded 0.90 with a small 95% CI. The total score of the dystonia subscale showed higher reliability than the BADS, Burke–Fahn–Marsden Movement Scale, and the Unified Dystonia Rating Scale.[20,22,23]

The dystonia subscale also showed good interrater reliability during action and excellent reliability during rest. The reliability of the region scores during action and rest overall was moderate to good. Reliability for the arms and legs was higher than for the eyes, neck, and trunk regions. Similar results were found in previous studies.[20–23] The choreoathetosis subscale also revealed excellent interrater reliability both during action and during rest. For the body regions, almost all ICCs exceeded 0.60, except for the eyes region. Owing to the lack of other choreoathetosis assessments in CP, comparison of these results with other studies is not possible.

The DIS dystonia subscale generally showed a somewhat lower interrater reliability than the choreoathetosis subscale. This can be explained by the lack of sustained postures in choreoathetosis and the more identifiable nature of choreoathetosis,[5–9] which makes choreoathetosis easier than sustained postures of dystonia to score on videotapes. Nevertheless, the reliability of the majority of the dystonia region scores was sufficient and total scores showed good to excellent ICCs.

The SEM and MDD showed small values. The MDD for interrater was 9% for the total DIS, 15% for the dystonia subscale, and 7% for the choreoathetosis subscale. The measurement errors (MDDs) for the DIS are obviously lower than the measurement errors for the BADS (18%), Burke–Fahn–Marsden Movement Scale (27%), and the Unified Dystonia Rating Scale (25%).[21] In other studies, MDD values for other measurement scales, for example for upper limb function in children with CP, have varied between 9% and 13%.[32] Low MDD values, as presented for the DIS, will benefit the sensitivity.

Also, the internal consistency was high and indicates a stable rating construct in measuring choreoathetosis and dystonia in dyskinetic CP.[29] The high internal consistency and the good MDD values of the DIS support the use of the scale in long-term follow-up and intervention studies, but future studies are needed to assess the responsiveness of the DIS.

Finally, the validity of the DIS was assessed. Content validity was achieved by analysis of the available measurement scales for dystonia and by the content discussions with the expert group of the CP Reference Centre and the clinical raters of the special schools for children with motor disabilities. Concurrent validity was attained for the dystonia subscale, in which a good correlation was found with the BADS.

This study has some limitations. A first shortcoming is the absence of a concurrent validity assessment for the choreoathetosis subscale. This could not be investigated owing to the lack of available choreoathetosis scales in CP and must be assessed in future studies. Another criticism concerns the duration of scoring the DIS scale on videotape. This varied from 30 to 45 minutes per subscale, which may seem long for application in routine clinical practice. However, because the DIS consists of two subscales, it covers an assessment of both dystonia and choreoathetosis and gives an opportunity to map the dyskinetic movement disorder in a more comprehensive approach. Furthermore, the video time for the children was 30 minutes maximum, which the participants tolerated very well. This is similar to other video-based scales (e.g. the BADS). Item reduction of the DIS may be a possibility for decreasing the duration score of the scale, but this would require a larger study group and its responsiveness to therapy should first be considered. A further consideration involves the complexity of differentiating between dystonia and choreoathetosis for the

different body regions, and therefore application/implementation of the scale requires some clinical experience with dyskinetic CP and careful application of the operational definitions of dystonia and choreoathetosis.

Despite these limitations, this study is the first to present a tool that measures dyskinesia, taking into account the simultaneous presence of dystonia and choreoathetosis in dyskinetic CP. Also, this clinical tool provides a unique contribution to evaluating choreoathetosis in CP, as, to our knowledge, no measurements have previously been available for choreoathetosis in CP. The evaluation of dystonia and choreoathetosis within one scale presents the prospect of including both pathological signs in one dyskinetic score as a ratio between the presence of dystonia and choreoathetosis.

CONCLUSION

This study developed a new measurement tool to evaluate dystonia and choreoathetosis in dyskinetic CP. The DIS showed high internal consistency and proved to be reliable between raters, with a low SEM and MDD. The concurrent validity was established for the dystonia subscale. The DIS is a step towards increasing insights in the clinical presentation and natural history of dyskinetic CP. Therefore, we hope that it will be a promising scale for measuring dystonia and choreoathetosis in long-term follow-up and medical intervention studies. Future research regarding the validity of the choreoathetosis subscale and responsiveness of the DIS is warranted.

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ONLINE MATERIAL/SUPPORTING INFORMATION

Supplementary material for this article may be found online.

REFERENCES

1. Cans C, Guillem P, Arnaud C. Surveillance of cerebral palsy in Europe: a collaboration of cerebral palsy surveys en registers. *Dev Med Child Neurol* 2000; **42:** 816–24.
2. Koman LA, Smith BP, Shilt JS. Cerebral palsy. *Lancet* 2004; **363:** 1619–31.
3. Rosenbaum P, Paneth N, Leviton A, Goldstein M, Bax M. A report: the definition and classification of cerebral palsy April 2006. *Dev Med Child Neurol* 2007; **49**(Suppl. 109): 8–14.
4. Bax M, Tydeman C, Flodmark O. Clinical and MRI correlates of cerebral palsy. The European cerebral palsy study. *JAMA* 2006; **296:** 1602–8.
5. Cans C, Guillem P, Arnaud C, et al. Prevalence and characteristics of children with cerebral palsy in Europe. *Dev Med Child Neurol* 2002; **44:** 633–40.
6. Krägeloh-Mann I, Petruch U, Weber P-M. SCPE Reference and Training Manual (R&TM). Grenoble: Surveillance of Cerebral Palsy in Europe, 2005.
7. Cans C, Dolk H, Platt MJ, Colver A, Prasauskiene A, Krägeloh-Mann I. Recommendations from the SCPE collaborative group for defining and classifying cerebral palsy. *Dev Med Child Neurol* 2007; **49**(Suppl. 109): 35–8.
8. Sanger TD. Pathophysiology of pediatric movement disorders. *J Child Neurol* 2003; **18**(Suppl. 1): 9–24.
9. Sanger TD, Chen D, Fehlings DL, et al. Definition and classification of hyperkinetic movements in childhood. *Mov Disord* 2010; **25:** 1538–49.
10. Butler C, Campbell S. Evidence of the effects of intrathecal baclofen for spastic and dystonic cerebral palsy. *Dev Med Child Neurol* 2001; **42:** 634–45.
11. Albright A, Barry M, Shafron D, Ferson S. Intrathecal baclofen for generalized dystonia. *Dev Med Child Neurol* 2001; **43:** 652–7.
12. Motta F, Stignani C, Antonello CE. Effect of intrathecal baclofen on dystonia in children with cerebral palsy and the use of functional scale. *J Pediatr Orthop* 2008; **28:** 213–7.
13. Holloway K, Baron M, Brown R, Cifu D, Carne W, Ramakrishnan V. Deep brain stimulation for dystonia: a meta-analysis. *Neuromodulation* 2006; **9:** 253–61.
14. Vidailhet M, Yelnik J, Lagrange C, et al. Bilateral pallidal deep brain stimulation for the treatment of patients with dystonia-choreoathetosis cerebral palsy: a prospective pilot study. *Lancet Neurol* 2009; **8:** 709–17.
15. Rice J, Waugh M. Pilot study of trihexyphenidyl in the treatment of dystonia in children with cerebral palsy. *J Child Neurol* 2009; **24:** 176–82.
16. Vles GF, Hendriksen JG, Visschers A, Speth L, Nicolai J, Vles JS. Levetiracetam therapy for treatment of choreoathetosis in dyskinetic cerebral palsy. *Dev Med Child Neurol* 2008; **51:** 418–9.
17. Shah V, Singh G, Giri V, et al. Dopa-responsive dystonia: correlates in a long-term follow-up. *Dev Med Child Neurol* 2010; **52:**(Suppl. 4) 72–3.
18. Albright AL, Tyler-Kabara EC. Combined ventral and dorsal rhizotomies for dystonic and spastic extremities. *J Neurosurg* 2007; **107:** 324–7.
19. Heinen F, Desloovere K, Schroeder AS, et al. The updated European Consensus 2009 on the use of Botulinum toxin for children with cerebral palsy. *Eur J Paediatr Neurol* 2009; **14:** 45–66.
20. Barry MJ, VanSwearingen JM, Albright AL. Reliability and responsiveness of the Barry–Albright Dystonia Scale. *Dev Med Child Neurol* 1999; **41:** 404–11.
21. Monbaliu E, Ortibus E, Roelens F, et al. Rating scales for dystonia in cerebral palsy: reliability and validity. *Dev Med Child Neurol* 2010; **52:** 570–5.
22. Burke RE, Fahn S, Marsden CD, Bressman SB, Moskowitz C, Friedman J. Validity and reliability of a rating scale for the primary torsion dystonias. *Neurology* 1985; **35:** 73–7.
23. Comella CL, Leurgans S, Wuu J, Stebbins GT, Chmura T. Dystonia study group. Rating scales for dystonia: a multicenter assessment. *Mov Disord* 2003; **18:** 303–12.
24. Himmelmann K, McManus V, Hagberg G, Uvebrant P, Krägeloh-Mann I, Cans C. Dyskinetic cerebral palsy in Europe: trends in prevalence and severity. *Arch Dis Child* 2009; **94:** 921–6.
25. Comella CL, Stebbins GT, Goetz CG, Chmura TA, Bressman SB, Lang AE. Teaching tape for the motor section of the Toronto Western Spasmodic Torticollis Scale. *Mov Disord* 1997; **12:** 570–5.
26. Goetz CG, Stebbins GT, Chmura T, Fahn S, Klawans H, Marsden CD. Unified Parkinson's disease rating scale (UPDRS). Training Video Tape.
27. Kirshner B, Guyatt G. A methodological framework for assessing health indices. *J Chronic Dis* 1985; **38:** 27–36.
28. Rigby A. Statistical recommendations for papers submitted to developmental medicine & child neurology. *Dev Med Child Neurol* 2010; **52:** 299–304.
29. Portney LG, Watkins MP. Foundations of Clinical Research: Application to Practice, 3rd edn. New Jersey: Pearson Prentice Hall, 2009.
30. Himmelmann K, Uvebrant P. Function and neuroimaging in cerebral palsy: a population-based study. *Dev Med Child Neurol* 2011; **53:** 516–21.
31. Thobois S, Taira T, Comella C, Moro E, Bressman S, Albanese A. Pre-operative evaluations for DBS in dystonia. *Mov Disord* 2011; **26**(S1): 17–26.
32. Krumlinde-Sundholm L, Holmefur M, Kottorp A, Eliasson AC. The assisting hand assessment: current evidence of validity, reliability, and responsiveness to change. *Dev Med Child Neurol* 2007; **49:** 259–64.