A new scale for children and adolescents aged 4-18 years, called the Movement Disorder Childhood Rating Scale, was designed to: (1) describe clinical features of different types of movement disorders; (2) evaluate the intensity of movement disorders in different body regions at rest and during specific tasks; and (3) assess the influence of movement disorders on motor function and daily living activities. The scale is divided into 2 parts: Part I for general assessment, and Part II for movement-disorder severity. It includes a 20-minute standardized video protocol of all items. The scale was applied to 61 patients with different types of movement disorder, and was independently scored by 3 observers. Reliability, construct validity, and consistency indexes are reported. High intrarater agreement on each item, and a high degree of internal consistency on several proposed items, were evident. © 2008 Published by Elsevier Inc.


Introduction

Movement disorders represent a common cause of disability in children with neurologic impairment. They result from a wide spectrum of central nervous system diseases and, depending on their etiology, are classified as primary (e.g., DYT1 gene idiopathic dystonia) or secondary (e.g., cerebral palsy or Wilson disease). Movement disorders can be divided into 2 broad groups: (1) dyskinesias (dystonia-athetosis, chorea-ballism, tics, myoclonus, and tremor) and (2) hypokinetic-rigid syndromes (Parkinsonism) [1-5].

The recognition of the pattern of movement disorders and the possibility of grading their severity are relevant for the clinician when planning rehabilitative and pharmacologic interventions, monitoring the results of treatment, and predicting outcomes.

Several instruments and rating scales are currently used to assess movement disorders, such as the Fahn-Marsden Rating Scale [6], the Barry-Albright Dystonia Scale [7], the Unified Dystonia Rating Scale [8], the Unified Huntington’s Disease Rating Scale [9], and the Federal University of Minas Gerais Sydenham’s Chorea Rating Scale [10]. However, they were mainly designed for adult patients, and are limited to the assessment of a single type of disorder (e.g., dystonia or chorea).

The prevalence of movement disorders in children, their clinical presentation and course, and their prognosis and management substantially differ from those of adults. The presentation is frequently insidious, and may be characterized at onset by mild hypotonia. The clinical picture may be more complex, rapidly changing, and often characterized by the association of different types of movement abnormalities. The pattern of movement disorders may be highly influenced by age at onset and by the stage of development at which the disease occurs. Moreover, the occurrence of movement disorders affects the course of neuromotor and adaptive development.

Accordingly, an assessment scale specifically designed for movement disorders in pediatric populations is crucial. This scale should take into account and measure movement-disorder severity, and also the impact of the disorder on child development and functioning, aspects that are highly age-specific and relevant to all types of movement disorders.
This study presents a new comprehensive rating scale for movement disorders in developmental age called Movement Disorder-Childhood Rating Scale, with the following aims: (1) to describe the clinical features of different types of movement disorders; (2) to evaluate the intensity of movement disorders in different body regions at rest and during specific tasks; and (3) to assess the influence of movement disorders on daily living activities and motor function.

Study Design and Methods

Development of the Scale

We sought to develop a comprehensive test for the assessment of function and disability in different types of movement disorders in children and adolescents. The literature on this topic was reviewed, and items were selected that in our experience were more informative and potentially applicable in childhood. We divided our scale into 2 parts: General Assessment (Part I) and Movement-Disorder Severity (Part II).

In Part I, 4 areas were included: motor function, oral/verbal function, self-care, and attention/alertness, for a total of 15 items. In respect to existing assessments, some items were added for a better characterization of the impact of movement disorders on different functions, in a developmental perspective. In the area of motor function, items related to head control, sitting and standing posture, and hand function were added. In the area of oral/verbal function, we included an item assessing the presence of drooling, an important clinical sign in children with movement disorders. A completely new area was added to explore attention and alertness, which are particularly vulnerable in developmental disorders and potentially sensitive to therapeutic interventions.

Part II assesses the intensity of the prevalent movement abnormality in 2 conditions, at rest and during the execution of specific tasks, in 7 body regions: eye and periorbital region, face, tongue and perioral region, neck, trunk, upper limb, and lower limb. The specific activities are performed after verbal instruction and visual demonstration.

All items are scored on a 5-point ordinal scale (0-4). Zero corresponds to no signs, and 4 corresponds to the most severe findings. The scores for Part I, Part II, and the total are calculated according to the statistical analysis reported below. The entire protocol of Movement Disorder-Childhood Rating Scale 4-18 is described in Appendix 1.

Patients

For the purposes of the present study, 61 patients (37 boys and 24 girls, aged 4.0-17.9 years; mean age, 10.69 ± 4.05 years S.D.) were enrolled from February 2005 to November 2007, from the Department of Developmental Neuroscience, Stella Maris Foundation (Pisa, Italy), and from the Department of Child Neurology and Psychiatry, University of Rome “La Sapienza” (Rome, Italy). All patients were diagnosed with one of the following movement disorders: dystonia (n = 38), ballism (n = 2), chorea (n = 9), hypokinetic-rigid syndrome (n = 6), nonepileptic myoclonus (n = 4), tremor (n = 1), and tic (n = 1). Patients were examined according to a comprehensive diagnostic protocol, to identify the etiology of their movement disorders, and the following diagnoses were rendered: dyskinetic cerebral palsy (n = 28), primary dystonia (n = 4), heredodegenerative disorder (n = 4), neurometabolic deficit (n = 2), posttraumatic disorder (tremor) (n = 1), brain arteriovenous malformation (n = 2), postinfective disorder (complex tic) (n = 1), and iatrogenic disorder because of a chemotherapeutic drug (n = 1). The cause of movement disorder remained undefined in 18 children.

This study was approved by the Ethics Committees of the institutions in Pisa and Rome. Informed consent was obtained from all parents and, when appropriate, from the patients.

Video Protocol

All patients were examined and filmed according to a standardized video protocol (Appendix 2). Videotape sessions lasted about 20 minutes, but in case of limited cooperation, it lasted longer, and sometimes the examination was performed in 2 or more sessions. A parental interview was always performed when scoring a patient’s alertness and attention at home (item D2). It was also used, as necessary, to score swallowing and drooling (items B1 and B2), and self-feeding and personal care (items C2 and C3). Written, informed consent for video recording was obtained for all patients.

The test was scored on the basis of video recordings for all 22 items. The videotapes of all patients were independently rated by 3 clinicians (R.B., E.P., and G.S.) with experience in the field of motor disorders. The use of videotapes in the study ensured that all scorers viewed exactly the same movements and postures.

Statistical Analysis

The Statistical Package for the Social Sciences, version 11.0.1 for Windows (SPSS, Inc., Chicago, IL), was used for data analysis (reliability, construct validity, and internal consistency). To assess rating reliability, the intraclass correlation coefficient was calculated by comparing the variability of different ratings of the same patient to the total variation across all ratings and all patients. To obtain the reliability of all judges averaged together, we calculated the interrater reliability coefficient [11], or average-measure intraclass correlation, by means of the Spearman-Brown correction formula:

$$
\text{icc} = \frac{j \times \text{icc}}{1 + (j - 1) \times \text{icc}}
$$

where j is the number of judges (3 for our study), and icc is the intraclass correlation coefficient.

The intraclass correlation coefficient and interrater reliability coefficient typically range from 0-1, with scores closer to 1 representing greater reliability [12].

To calculate an index for Parts I and II, we performed a standardized value ($X_{\text{stand}}$) for each item with the formula:

$$
X_{\text{stand}} = \frac{X_{\text{assigned}} - X_{\text{min}}}{X_{\text{max}} - X_{\text{min}}}
$$

where $X_{\text{assigned}}$ is the raw value assigned to the patient, $X_{\text{max}}$ is the maximum value of the item (score of 4), and $X_{\text{min}}$ is the minimum value of the item (score of 0). The “method of variables standardization” with variation field was used.

For each of the 2 parts of the scale, the mean value of $X_{\text{stand}}$ was calculated by means of the formula:

$$
\text{Index} = \frac{\sum X_{\text{stand}}}{n}
$$

where n is the number of items. The index for Part I (Index I) was derived from 15 items, and the index for Part II (Index II) was derived from 7 items. Each index could range from 0-1, and was divided into 5 classes, according to severity of movement disorder. Class 1 included an index from 0-0.2, i.e., healthy. Class 2 included an index from 0.2-0.4, i.e., mildly affected. Class 3 included an index from 0.4-0.6, i.e., moderately affected. Class 4 included an index from 0.6-0.8, i.e., severely affected. Class 5 included an index from 0.8-1, i.e., profoundly affected. In each patient, a class for Part I (Class I) and a class for Part II (Class II) was obtained.

To obtain a global measure of the scale, including the information from Indexes I and II, we used the method of weighted means of the two indexes, calculated with the formula:

$$
\text{Global Index} = \frac{(\text{Index}_{\text{I}} \times n_I) + (\text{Index}_{\text{II}} \times n_{II})}{n_I + n_{II}}
$$

where $n_I$ is the number of items in Part I (15 items), and $n_{II}$ is the number...
of items in Part II (7 items). On the basis of the Global Index, a Global Class was obtained in each patient, according to the ranges previously described.

The Spearman correlation coefficient was used to measure the correlation between (1) the 2 indexes, (2) the Global Index and each of the 2 indexes separately, (3) the 2 classes, and (4) the Global Class and each of the 2 classes separately.

Test-retest reliability was used as an index of score consistency, and was obtained by administering the same test a second time, 1 week later, and then correlating the 2 score sets. A test-retest coefficient was determined for 20 patients.

The construct convergent validity of the Movement Disorder-Childhood Rating Scale 4-18 was also determined by correlating performance in the Movement Disorder-Childhood Rating Scale of our 38 patients with dystonic movement disorders, with the Burke-Fahn-Marsden Rating Scale [6]. This scale is one of the most widely used assessment scales for dystonia [13,14], and is based on the examination of a patient in a standard setting (Burke-Fahn-Marsden Movement Scale) and on a patient’s subjective report of disability in activities of daily living (Burke-Fahn-Marsden Disability Scale).

The internal consistency of Movement Disorder-Childhood Rating Scale Parts I and II was assessed by means of Cronbach’s alpha, and by the Spearman rho correlation coefficient between each item of Part I.

### Scale Responsiveness

Scale responsiveness is an index of the capacity of a scale to detect changes. We tested the ability of the Movement Disorder-Childhood Rating Scale 4-18 to quantify adequate correlations in a preliminary longitudinal study of 8 dystonic patients (4 girls and 4 boys; mean age at first scoring, 13.36 ± 3.47 years S.D.; range, 8-17.7 years) before and after 3 and 6 months of a specific drug treatment (an anticholinergic drug, e.g., trihexyphenidyl).

### Results

A large range of results was obtained in our sample for all scores, with the Global Index ranging from 0.17-0.99 (mean, 0.56; S.D. 0.22). No correlation with age at testing was evident (Pearson nonparametric test, \( P = -0.153 \)).

### Reliability

The intraclass correlation coefficient values for each item of Parts I and II ranged from 0.98-1 (median, 1), and from 0.99-1 (median, 0.99), respectively. The interrater reliability coefficient for each item of Parts I and II ranged from 0.97-1 (median, 1), and from 0.98-1 (median, 0.99), respectively (Table 1). We measured the mean and median values of Index I, Index II, and Global Index, and Class I, Class II, and Global Class for the Movement Disorder-Childhood Rating Scale 4-18 are reported in Table 3. The test-retest reliability, measured by Pearson correlation coefficient, ranged from 0.91-0.98.

### Construct Validity

The mean of the Burke-Fahn-Marsden Rating Scale was also measured (Table 2). The correlations between different indexes of the Movement Disorder-Childhood Rating Scale 4-18 are reported in Table 3. The test-retest reliability, measured by Pearson correlation coefficient, ranged from 0.91-0.98.

### Table 1. Movement Disorder-Childhood Rating Scale 4-18: Intraclass correlation coefficient and interrater reliability coefficient for 3 raters

<table>
<thead>
<tr>
<th>Item</th>
<th>Movement Disorder-Childhood Rating Scale 4-18</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>ICC</td>
</tr>
<tr>
<td>Part I</td>
<td></td>
</tr>
<tr>
<td>Head control</td>
<td>0.99</td>
</tr>
<tr>
<td>Sitting position</td>
<td>0.98</td>
</tr>
<tr>
<td>Standing position</td>
<td>0.99</td>
</tr>
<tr>
<td>Walking</td>
<td>1.00</td>
</tr>
<tr>
<td>Reaching</td>
<td>0.99</td>
</tr>
<tr>
<td>Grasping</td>
<td>0.99</td>
</tr>
<tr>
<td>Handwriting</td>
<td>0.99</td>
</tr>
<tr>
<td>Swallowing</td>
<td>1.00</td>
</tr>
<tr>
<td>Drooling</td>
<td>0.98</td>
</tr>
<tr>
<td>Language</td>
<td>1.00</td>
</tr>
<tr>
<td>Self-dressing</td>
<td>1.00</td>
</tr>
<tr>
<td>Self-feeding</td>
<td>1.00</td>
</tr>
<tr>
<td>Personal care</td>
<td>1.00</td>
</tr>
<tr>
<td>Attention/alertness during observation</td>
<td>1.00</td>
</tr>
<tr>
<td>Attention/alertness at home</td>
<td>1.00</td>
</tr>
<tr>
<td>Part II</td>
<td></td>
</tr>
<tr>
<td>Eye and periorbital region</td>
<td>0.99</td>
</tr>
<tr>
<td>Face</td>
<td>0.99</td>
</tr>
<tr>
<td>Tongue and perioral region</td>
<td>0.99</td>
</tr>
<tr>
<td>Neck</td>
<td>0.99</td>
</tr>
<tr>
<td>Trunk</td>
<td>1.00</td>
</tr>
<tr>
<td>Upper limb</td>
<td>1.00</td>
</tr>
<tr>
<td>Lower limb</td>
<td>1.00</td>
</tr>
</tbody>
</table>

**Abbreviations:**

- ICC = Intraclass correlation coefficient
- IRC = Interrater reliability coefficient

### Table 2. Movement Disorder-Childhood Rating Scale 4-18: Values of Indexes and Classes, in Comparison with Values Obtained with Burke Fahn Marsden Rating Scale

<table>
<thead>
<tr>
<th>Index I (Mean ± S.D.)</th>
<th>Class I (Median)</th>
<th>Index II (Mean ± S.D.)</th>
<th>Class II (Median)</th>
<th>Global Index (Mean ± S.D.)</th>
<th>Global Class (Median)</th>
<th>Burke Fahn Marsden Rating Scale* (Mean ± S.D.)</th>
</tr>
</thead>
<tbody>
<tr>
<td>0.45 ± 0.25</td>
<td>2</td>
<td>0.67 ± 0.23</td>
<td>4</td>
<td>0.56 ± 0.22</td>
<td>3</td>
<td>54.75 ± 32.89</td>
</tr>
</tbody>
</table>

* Only dystonic patients.

**Abbreviation:**

S.D. = Standard deviation
Table 3. Movement Disorder-Childhood Rating Scale 4-18: Reliability and construct validity according to Spearman \( \rho \) correlation

<table>
<thead>
<tr>
<th>Class I</th>
<th>Index I</th>
<th>Class I</th>
<th>Index II</th>
<th>Class II</th>
<th>Global Index</th>
<th>Global Class</th>
</tr>
</thead>
<tbody>
<tr>
<td>0.957**</td>
<td>0.717**</td>
<td>0.752**</td>
<td>0.963**</td>
<td>0.920**</td>
<td>0.935**</td>
<td>0.953**</td>
</tr>
<tr>
<td>0.711**</td>
<td>0.753**</td>
<td>0.902**</td>
<td>0.891**</td>
<td>0.906**</td>
<td>0.858**</td>
<td>0.834**</td>
</tr>
<tr>
<td>0.902**</td>
<td>0.910**</td>
<td>0.920**</td>
<td>0.986**</td>
<td>0.906**</td>
<td>0.834**</td>
<td>0.953**</td>
</tr>
</tbody>
</table>

Burke-Fahn-Marsden Rating Scale findings included dystonic patients only.

** Correlation significant at the .01 level.

Abbreviation:
BFM = Burke-Fahn-Marsden

Table 4. Movement Disorder-Childhood Rating Scale 4-18: Consistency according to Spearman \( \rho \) correlation

<table>
<thead>
<tr>
<th>HC</th>
<th>Sit P</th>
<th>St P</th>
<th>Walk</th>
<th>Reach</th>
<th>Grasp</th>
<th>Gr</th>
<th>Sw</th>
<th>Drool</th>
<th>Lang</th>
<th>S-Dress</th>
<th>S-Feed</th>
<th>P Care</th>
<th>A/A Obs</th>
</tr>
</thead>
<tbody>
<tr>
<td>0.802**</td>
<td>0.763**</td>
<td>0.380**</td>
<td>0.774**</td>
<td>0.835**</td>
<td>0.954**</td>
<td>0.649**</td>
<td>0.775**</td>
<td>0.741**</td>
<td>0.766**</td>
<td>0.661**</td>
<td>0.777**</td>
<td>0.750**</td>
<td>0.759**</td>
</tr>
<tr>
<td>0.518**</td>
<td>0.692**</td>
<td>0.622**</td>
<td>0.741**</td>
<td>0.745**</td>
<td>0.691**</td>
<td>0.710**</td>
<td>0.651**</td>
<td>0.705**</td>
<td>0.698**</td>
<td>0.692**</td>
<td>0.797**</td>
<td>0.808**</td>
<td>0.787**</td>
</tr>
<tr>
<td>0.745**</td>
<td>0.720**</td>
<td>0.875**</td>
<td>0.880**</td>
<td>0.735**</td>
<td>0.747**</td>
<td>0.573**</td>
<td>0.610**</td>
<td>0.835**</td>
<td>0.862**</td>
<td>0.704**</td>
<td>0.654**</td>
<td>0.808**</td>
<td>0.733**</td>
</tr>
<tr>
<td>0.681**</td>
<td>0.989**</td>
<td>0.929**</td>
<td>0.729**</td>
<td>0.949**</td>
<td>0.974**</td>
<td>0.929**</td>
<td>0.974**</td>
<td>0.989**</td>
<td>0.989**</td>
<td>0.989**</td>
<td>0.989**</td>
<td>0.989**</td>
<td>0.989**</td>
</tr>
<tr>
<td>0.445**</td>
<td>0.334**</td>
<td>0.408**</td>
<td>0.395**</td>
<td>0.327**</td>
<td>0.371**</td>
<td>0.472**</td>
<td>0.393**</td>
<td>0.374**</td>
<td>0.415**</td>
<td>0.373**</td>
<td>0.364**</td>
<td>0.352**</td>
<td>0.373**</td>
</tr>
<tr>
<td>0.372**</td>
<td>0.271**</td>
<td>0.323*</td>
<td>0.305*</td>
<td>0.307*</td>
<td>0.274*</td>
<td>0.335*</td>
<td>0.336*</td>
<td>0.304*</td>
<td>0.311*</td>
<td>0.295*</td>
<td>0.256*</td>
<td>0.274*</td>
<td>0.847**</td>
</tr>
</tbody>
</table>

* Correlation significant at the 0.05 level.
** Correlation significant at the 0.01 level.

Abbreviations:
A/A HOME = Attention/alertness at home
A/A Obs = Attention/alertness during observation
Drool = Drooling
Grasp = Grasping
HC = Head control
Lang = Language
P Care = Personal care
Reach = Reaching
S-Dress = Self-dressing
S-Feed = Self-feeding
Sit P = Sitting position
St P = Standing position
Sw = Swallowing
Walk = Walking

Scale Responsiveness

The functional improvement of the 8 dystonic patients was evaluated before and after 3 and 6 months of continuous use of trihexyphenidyl, using a two-sided Student \( t \) test. The mean total Movement Disorder-Childhood Rating Scale 4-18 score changed from 0.53 (SD, 0.11) before treatment, to 0.46 (SD, 0.12) after 3 months (\( P = 0.03 \)) and 0.37 (SD, 0.15) after 6 months of treatment (\( P = 0.0002 \)).
Discussion

The Movement Disorder-Childhood Rating Scale was developed to address the perceived limitations of rating scales that were mainly developed for use in adult patients or for specific subtypes of movement disorders. Some important aspects differentiate childhood movement disorders from those occurring at a later age, in particular the presence of changing clinical patterns (e.g., hyperkinetic forms can evolve into more dystonic or hypokinetic forms), the coexistence of different subtypes of movement disorders, and the influence of a disorder on the achievement of developmental milestones. In designing the Movement Disorder-Childhood Rating Scale, we addressed a wider range of movement-disorder subtypes, and we added items specifically exploring the impact of movement disorders on neurodevelopment.

The scale is divided into 2 parts. The first part explores, in a developmental perspective, the degree of disability in different domains (motor function, oral/verbal area, and daily living activities), and the second part focuses on the topography and severity of specific movement disorders. This multiaxial approach reflects the approach in some of the most diffuse rating scales applied in adult movement disorders, such as the Burke-Fahn-Marsden Rating Scale for dystonia, which separately considers disability and motor disorder [6]. Interestingly, when applied to the subgroup of patients with dystonia (primary and secondary), the Motor Subscale and the Disability Subscale of the Burke-Fahn-Marsden Rating Scale indicated a high correlation with Movement Disorder-Childhood Rating Scale 4-18 Parts I and II, respectively.

The high intraclass correlation coefficients for all Movement Disorder-Childhood Rating Scale 4-18 items and test-retest reliability, demonstrating excellent scores in all items for 3 independent raters, indicate that the assessment can be performed by experienced clinicians with high accuracy, because the scores remained constant and there was no change in the property being measured. We also found a high degree of internal consistency, as expressed by the Cronbach’s alpha. Moreover, in the Spearman correlation analysis, all items were reciprocally correlated, suggesting that they are all tapping the same general construct.

The results of this pilot study on repeated testing of a small group of children, who began to use an anticholinergic drug, seem to indicate the responsiveness of Movement Disorder-Childhood Rating Scale 4-18 and its potential usefulness in the assessment and monitoring of children and adolescents with movement disorders during a specific treatment. However, these results need to be confirmed in larger groups.

One important limit of the present study is the low number of patients with tic or tremor. No conclusions can be drawn about the validity of the scale in these types of movement disorders. Nevertheless, application of the Movement Disorder-Childhood Rating Scale in these children might prove useful during clinical and therapeutic follow-up, particularly for the subscales concerning movement-disorder distribution and severity.

In conclusion, the Movement Disorder-Childhood Rating Scale 4-18 seems to fulfill the statistical requirements for application in the quantitative assessment of movement disorders in childhood. Compared with existing scales, the Movement Disorder-Childhood Rating Scale can be used in children affected by different types of movement disorders. Moreover, this new tool includes items relevant for assessing a child’s impairment and participation, in terms of postural control, manipulation, drooling, and attention. We think that this scale can be useful in assessing movement disorders in childhood. Longitudinal studies are being planned, to explore the sensitivity and reliability of this new instrument in detecting the clinical changes associated with either natural history or therapeutic intervention.

This work was supported by grants from the Italian Ministry of Health (RC1/2006 and FEF 2003). We thank Michele Coluccini, PT for his clinical expertise, and Pietro Patusi, PhD for statistical analysis.

References


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Appendix 1. Movement Disorder Childhood Rating Scale for ages 4-18 years

<table>
<thead>
<tr>
<th>Classification of MD</th>
<th>Prevalent MD</th>
<th>Other MD</th>
</tr>
</thead>
<tbody>
<tr>
<td>Hypokinetic-rigid syndrome</td>
<td>□□</td>
<td>□</td>
</tr>
<tr>
<td>Chorea/ballism</td>
<td>□</td>
<td>□</td>
</tr>
<tr>
<td>Dystonia/athetosis</td>
<td>□</td>
<td>□</td>
</tr>
<tr>
<td>Myoclonus</td>
<td>□</td>
<td>□</td>
</tr>
<tr>
<td>Tic</td>
<td>□</td>
<td>□</td>
</tr>
<tr>
<td>Tremor</td>
<td>□</td>
<td>□</td>
</tr>
</tbody>
</table>

Part I. General Assessment

A. Motor function

1. Head control
   - 0 normal
   - 1 mildly affected by the MD and/or control longer than 1 min
   - 2 moderately affected by the MD and/or control between 30 seconds and 1 min
   - 3 severely affected by the MD and/or control shorter than 30 seconds
   - 4 absent

2. Sitting position
   - 0 normal
   - 1 mildly affected by the MD and/or control longer than 1 min
   - 2 moderately affected by the MD and/or control between 30 seconds and 1 min
   - 3 severely affected by the MD and/or control shorter than 30 seconds
   - 4 absent

3. Standing position
   - 0 normal
   - 1 maintained for >1 min, even with abnormal posture
   - 2 maintained for >30 seconds and <1 min, even with abnormal posture
   - 3 maintained for <30 seconds, or for long periods with support
   - 4 absent or for brief periods with support

4. Walking
   - 0 normal
   - 1 walks alone for >5 min, even with abnormal posture
   - 2 walks alone for <5 min, even with abnormal posture or needs support
   - 3 only a few steps with support, and with very abnormal pattern
   - 4 absent

5. Reaching
   - 0 normal
   - 1 possible for age-adequate tasks, even if qualitatively abnormal
   - 2 moderately abnormal, only possible to approach the object
   - 3 severely abnormal, only inefficient efforts
   - 4 absent

6. Grasping
   - 0 normal
   - 1 mildly abnormal, but pincer-grasp possible
   - 2 moderately abnormal, only possible for cubes very close to the hand
   - 3 severely abnormal, only inefficient efforts
   - 4 absent

7. Handwriting
   - 0 normal
   - 1 mild difficulties, but readable
   - 2 moderate difficulty, not completely readable
   - 3 severe difficulties, not readable
   - 4 absent, a functional grasp of the pen is not possible

B. Oral/verbal function

1. Swallowing
   - 0 normal
   - 1 occasional dysphagia
   - 2 frequent dysphagia, swallowing difficulties
   - 3 swallowing of solid food is not possible
   - 4 severe difficulty in swallowing semisolid and liquid food

2. Drooling
   - 0 absent
   - 1 occasional
   - 2 mild
   - 3 moderate
   - 4 severe

3. Language
   - 0 normal
   - 1 mild dysarthria, speech is comprehensible
   - 2 moderate dysarthria, speech is not fully comprehensible
   - 3 severe dysarthria, speech is generally not comprehensible
   - 4 complete or almost complete anarthria

C. Self-care

1. Self-dressing
   - 0 complete autonomy
   - 1 necessity of minimal assistance (not fully efficient, but independent)
   - 2 necessity of assistance in some tasks (e.g., to tie shoes or to button up)
   - 3 partially dependent (can cooperate in self-dressing)
   - 4 totally dependent

2. Self-feeding
   - 0 complete autonomy (can use cutlery)
   - 1 necessity of minimal assistance (not fully efficient, but independent)
   - 2 necessity of assistance in some tasks (e.g., using knife)
   - 3 partially dependent (uses hands for eating)
   - 4 totally dependent

3. Personal care
   - 0 complete autonomy
   - 1 necessity of minimal assistance (not fully efficient but independent)
   - 2 necessity of assistance in some tasks (e.g., bathing)
   - 3 partially dependent (can cooperate in washing face or brushing teeth)
   - 4 totally dependent

D. Attention/alertness

1. Attention/alertness during the observation
   - 0 constantly alert, can answer all age and mental-adequate questions
   - 1 sometimes (1/3 of the observation) inattentive
   - 2 often (2/3 of the observation) inattentive
   - 3 attention needs to be constantly drawn by the examiner
   - 4 constantly a scarce reaction to external stimuli

2. Attention/alertness at home
   - 0 constantly alert during day hours, good response to the environment
   - 1 sometimes (1/3 of day hours) difficult to maintain attention
   - 2 often (2/3 of day hours) the child is hypo-attentive
   - 3 child is constantly sleepy, even during day hours
   - 4 constantly a scarce reaction to external stimuli

Part II. MD Assessment

MD severity

1. Eye and periorbital region
   - 0 MD is absent
   - 1 MD is present only at rest
   - 2 MD is present during one or some of the tasks for the region examined and/or involves one or two of the other regions
   - 3 MD is present during one or some of the tasks for the region examined, and involves three or more of the other regions
4 MD is present during all of the tasks for the region examined and/or involves three or more of the other regions, making completion impossible

2. Face (see item 1)
   - 0
   - 1
   - 2
   - 3
   - 4

3. Tongue and perioral region (see item 1)
   - 0
   - 1
   - 2
   - 3
   - 4

4. Neck (see item 1)
   - 0
   - 1
   - 2
   - 3
   - 4

5. Trunk (see item 1)
   - 0
   - 1
   - 2
   - 3
   - 4

6. Upper limb (see item 1)
   - 0
   - 1
   - 2
   - 3
   - 4

7. Lower limb (see item 1)
   - 0
   - 1
   - 2
   - 3
   - 4

**Abbreviation:**
MD = Movement disorder
* Sign only of type of prevalent MD.

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**Appendix 2. Movement Disorder Childhood Rating Scale 4-18: Protocol of video recording**

<table>
<thead>
<tr>
<th>Video perspective</th>
<th>Activity</th>
<th>Part I Area (Items)</th>
<th>Part II (Items)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Full body view</td>
<td>Ask the child, if possible, to remove shoes and to take off upper/lower limb garments and dresses</td>
<td>Self-care (1)</td>
<td>(4-7)</td>
</tr>
<tr>
<td></td>
<td>Supine position for 1 min</td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>Sitting position for 1 min supporting the trunk, if necessary</td>
<td>Motor (1, 2)</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Standing position for 2 min with support, if necessary</td>
<td>Motor (3)</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Walking for 5 meters with aids of support, if necessary,</td>
<td>Motor (4)</td>
<td></td>
</tr>
<tr>
<td>Upper part of the body (including arms)</td>
<td>Sitting position (on a chair or in his/her own postural system), in front of a table</td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>(a) Put 5 cubes of different size on one side and ask the child to transfer each cube to the contralateral side and back. Repeat the task starting from the other side.</td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>(b) Put a pen on the table and ask the child to draw a human figure and to write his/her own name</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Face Full body view</td>
<td>At rest or during spontaneous behavior</td>
<td>All</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Ask the child to fixate and follow a visual stimulus</td>
<td>Oral/verbal (1)</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Ask the child to smile and open/close eyes</td>
<td>Oral/verbal (2)</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Ask the child to protrude tongue and move it in all directions</td>
<td>Oral/verbal (3)</td>
<td></td>
</tr>
<tr>
<td>Full body view</td>
<td>Ask the name, surname, or observe spontaneous production</td>
<td>Oral/verbal (3)</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Ask the child, if possible, to put shoes back on and upper limb/lower limb garments and dresses</td>
<td>Self care (1)</td>
<td>(4-7)</td>
</tr>
<tr>
<td>Entire video</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Audio recording</td>
<td>Parental interview</td>
<td>Attention/alertness (1)</td>
<td></td>
</tr>
</tbody>
</table>

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