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REVIEW ARTICLE

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Current behavioral assessments of movement disorders in children

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Summary

Pediatric movement disorders (PMDs) are common and have recently received increasing attention. As these disorders have special clinical features, the selection of appropriate behavioral assessment tools that can clearly distinguish movement disorders from other diseases (eg, epilepsy and neuromuscular disorders) is crucial for achieving an accurate diagnosis and treatment. However, few studies have focused on behavioral assessments in children. The present report attempts to provide a critical review of the available subjective and objective assessment tests for common PMDs. We believe that the principles of objectification, multi-purpose use, and simplification are also applicable to the selection and development of satisfactory pediatric behavioral assessment tools. We expect that the development of wearable sensors, virtual reality, and augmented reality will lead to the establishment of more reliable and simple tests. In addition, more rigorous randomized controlled trials that have been specifically designed to evaluate behavioral testing in children are also expected in the future.

KEYWORDS
behavioral assessments, dystonia, pediatric movement disorders, tics

1 | INTRODUCTION

Pediatric movement disorders (PMDs), which are fairly common, have received increasing attention over time. A discussion of PMDs should consider several characteristics: (a) PMDs are paroxysmal; abnormal movements are often observed with specific conditions or at specific time points. (b) PMDs exhibit varied clinical manifestations consequent to mixed movement disorders with different pathophysiological processes, which may overlap and thus complicate the signs and symptoms.[1] For example, children suffering from cerebral palsy (CP) often exhibit simultaneous dystonia and spasticity. (c) PMD is associated with difficulties in clinical examinations. For example, some children might not cooperate with the physical examination and may even refuse further communication. Therefore, the selection of effective behavioral assessment tools that can distinguish a specific movement disorder from other diseases is crucial to

ensuring an accurate diagnosis and the pursuit of appropriate treatment. To date, however, rigorously designed randomized controlled trials (RCTs) of the efficacies of therapeutic strategies for PMDs have provided little evidence.[1] Accordingly, the various new RCTs for PMDs expected in the near future will further enable the selection of satisfactory tools for improving diagnostic accuracy.[2]

An effective behavioral assessment tool that could provide strong evidence for the investigation of changes in symptoms would also be indispensable to an experimental design. Our previous studies appealed to the principles of objectification, multi-purpose use, and simplification (OMS) when selecting and developing appropriate behavioral assessments of Parkinson disease in adults.[3,4] Objectification means that a more objective test, or objective indexes in a scale, should be developed and adopted to avoid observation bias. Multi-purpose use means that during a single test, multiple indexes should be measured simultaneously to enhance experimental efficiency and reduce the burden on the patient. Simplification means that the testing processes should be simple and easily accepted by the patients and clinicians. We believe that these principles are also applicable to the selection and development of satisfactory behavioral assessments for pediatric patients.

Several important systematic reviews on this topic have been read and reviewed. For example, Delgado and Albright published a systemic review of the definitions, classifications, and grading systems.[5] Although this document provides general information regarding the subjective scales of PMDs, this information needs to be upgraded. Pietracupa et al.[6] systematically reviewed hyperkinetic movements, including tremors, dystonia, chorea, tics, myoclonus, and drug-induced dyskinesia (Pietracupa et al.[6]). However, that study did not focus on children and included only subjective scales. Koy et al.[1] summarized progress in the management of PMDs; although several instruments were mentioned, this article did not focus on behavioral assessments. Martino et al.[2] provided a systematic review of subjective scales for tics. That study evaluated a number of recommended tic severity rating scales and advocated the development of a scale comprising all pathological behaviors involving tics. Note that all these previous studies discussed only subjective scales; by contrast, no reported study has summarized objective instruments. Given the complexity of PMDs and the frequent overlap and confusion between signs and diseases, a critical review concerning the progress of behavioral assessments, particularly objective assessments, is extremely important. Therefore, we have reviewed the available subjective and objective behavioral assessments of PMDs and have attempted to briefly and critically summarize the existing tools and establish a prospective based on our insights and experience.

2 | SEARCHING STRATEGY

We searched the PubMed, EMBASE, Web of Science, and Google Scholar databases for articles published from Jan 1, 2015 to August 1, 2017, using the terms "movement disorders" "spasticity" OR "stereotypies" OR "dystonia" OR "athetosis" OR "myoclonus" OR "tics" OR "Tourette" OR "tremor" OR "ataxia" OR "ballism" AND "children" OR "pediatric" OR "childhood". English-language peer-reviewed original studies and review articles were considered. Articles obtained by searching the above-mentioned databases were read to identify additional reports. All behavioral assessments involving PMDs were included. The final references were established using citations in the context of the present review.

3 | BEHAVIORAL ASSESSMENTS OF PMDS

We attempted a critical review of the available subjective and objective assessments of common PMDs. We have described comprehensive scales in the sections where they were most commonly used. We included a discussion regarding spasticity, which is not always considered as a "movement disorder," with reference to a previous study.[5]

3.1 | Spasticity

Spasticity is the most common symptom of CP in affected children and has the most limiting effects on the performance of motor skills and activities of daily living (ADLs). The Task Force on Childhood Motor Disorders defines spasticity as hypertonia resistant to movement or movement acceleration.[7]

The Modified Ashworth Scale,[8] which was developed by Ashworth in 1964 and modified in 1987 and 2008,[9] is most commonly used subjective scale of spasticity. Over time, this investigator-reported 5-item scale has been adopted as a standard of practice for spasticity assessment.[10] Although a recent report described an attempt to objectivize this subjective scale in adult spastic patients,[11] its appropriateness for children remains unknown.

The Tardieu Scale[11,12] is another important scale of spasticity that is always used in combination with the Ashworth scale. This scale measures the effects of spasticity on position, movement, speed, and angle. In contrast to the Ashworth Scale, the Tardieu scale assesses two different velocities related to passive stretch and can therefore distinguish spasticity from contracture. This scale includes measurements of muscle reaction quality and angle and is considered valid and reliable, although its use is limited by the requirement for an experienced investigator. More recently, Jethwa et al.[13] developed a Hypertonia Assessment Tool (HAT) for the discrimination of spasticity, dystonia, and rigidity. Recently, researchers evaluated the validity and reliability of HAT and confirmed to be valid for the assessment of spasticity.[14]

Additionally, the Gross Motor Function Measure has been adopted for the evaluation of gross motor functions in children with CP.[15] This scale measures both spasticity and muscle strength. The 66- and 88-item versions of the Gross Motor Function Measure evaluate the abilities of lying and rolling; sitting; crawling and kneeling; standing; and walking, running, and jumping. As these items are complex, however, this scale also requires an experienced investigator. In

addition to the commonly used scales listed above, researchers have recently begun to include the Pediatric Balance Scale,[16,17] in evaluations of active range of movement, and the Manual Muscle Test in evaluations of children with CP. We have listed the main subjective assessments in Table 1.

Electromyography (EMG) might be considered the most classical spasticity evaluation method among the available objective assessment tools.[11] Most notably, EMG offers the benefits of objective and visual results. However, EMG requires a special device and must be performed by experienced staff. It is also invasive and may be resisted by some children. Sloot[18] reported a battery of manual instrumentation-based tests, including measurements of joint velocity, imposed force, and muscle activity, used to objectively measure spasticity in CP.[18] The authors of that study verified that the measurements matched the subjects' gait profiles. Furthermore, Russian investigators have reported the use of an ultrasonographic technique to objectively evaluate the degree of muscle degeneration related to spasticity.[19] Additionally, van Hedel et al. developed the YouGrabber measurements, which evaluate arm and hand movements while playing a game. Using that system, which was readily accepted by children, the researchers could differentiate between compensatory and physiological motor performance.[20] Developments in the fields of computer science and sensors have yielded trends with regard to objectification and quantification during evaluations of spasticity.[4,11,20] We therefore expect the emergence of more objective tests and modifications of classic scales.

3.2 | Dystonia

Dystonia is a hyperkinetic PMD defined as the presence of involuntary muscle contractions (intermittent or sustained). This condition might be multi-factorial, with etiologies including genetic (eg, mutations in genes encoding DYT1/TOR1, PRRT2, MR-1, and SLC2A1) and acquired factors (eg, vascular or metabolic complications, infectious disease, or drug toxicities). Dystonia may also occur concomitantly with symptoms such as spasticity and tremor. Clinically, the manifestation of dystonia is complicated, and misdiagnosis is common. For example, although some children have been subjected to selective lumbar rhizotomy for a diagnosis of recurrent spasticity, the cases actually involved undiagnosed dystonia.[5] Therefore,

TABLE 1 Tools for the subjective assessment of spasticity

Scales	Developers	Contents	Behavior assessed	Brief commentary	
				Strengths	Weaknesses
Ashworth Scale	Ashworth (1964)[8] Naghdi (2008)[9]: modification	Five-item scale Investigator reported	Spasticity	A classic and widely used scale Simple, valid, and convenient Allows a rapid impression of severity	Cannot distinguish among types of hypertonia (rigidity, spasticity, or dystonia)
Tardieu Scale	Tardieu (1954)[11] Boyd (1999)[12]: modification	Quality of muscle reaction Angle of muscle reaction	Spasticity	Valid and reliable Can be used to distinguish spasticity and contracture; Can be modified according to disease and affected limbs	Complex Requires experienced investigators
Hypertonia Assessment Tool	Jethwa (2010)[13]	7-item, 3-level scale	Distinguishes spasticity, dystonia, and rigidity	Good validity for spasticity	Validity for dystonia and rigidity has not been confirmed
Gross Motor Function Measure	Palisano (1997)[15]	Four-class scale Two versions: 66- and 88-item Investigator reported	Gross motor performance	A reliable and valid classification system Comprehensive	Some children cannot complete this complicated scale Complex Requires experienced investigators
Pediatric Balance Scale	Franjoine (2003)[17]	Modified from the classic Berg Balance Scale 14-item, 5-class scale Investigator reported	Balance	Particular optimization for children Valid and reliable	Subjective
Manual Muscle Test	Wints (1959)[11]	Includes no movement, test movement, and test positions	Function and strength of individual muscles	Valid and reliable	Complicated Requires a highly experienced therapist

assessments of dystonia are essential to ensure the provision of sufficient information to clinicians.

The Barry-Albright Dystonia Scale (BADS), a modified version of the Burke-Fahn-Marsden Dystonia Rating Scale (BFMDRS), is the subjective assessment most commonly used in children.[5] The BADS, a 5-level, investigator-reported evaluation of the eyes, mouth, neck, trunk, upper extremities, and lower extremities, can be performed either face-to-face or with video assistance. The maximum total BADS score is 32 points, with a higher score indicating more severe symptoms. BADS features the clinical advantages of a concise and convenient design. Additionally, several studies have directly adopted the BFMDRS for children with CP. However, evidence regarding the reliability of the BADS and BFMDRS is limited; these tests exhibit moderate internal consistency and inter-rater reliability but no evidence of test-retest reliability.[21]

The Dystonia Study Group established the Unified Dystonia Rating Scale (UDRS) and Global Dystonia Rating Scale (GDS) to address the limitations of BFMDRS. These scales evaluate dystonia in 14 body areas. The possible UDRS scores range from 0 (no dystonia) to 4 (severe dystonia), whereas the possible GDS scores range from 0 (no dystonia) to 10 (severe dystonia). Although the authors of an earlier study claimed that both scales had many advantages, including excellent internal consistency and simplicity,[22] neither scale has been used routinely in children or established as valid and reliable in this population. In addition to dystonia-specific scales, several comprehensive scales such as the Dyskinesia Impairment Scale (DIS) and Movement Disorder-Childhood Rating Scale (MD-CRS) include dystonia subscales. The DIS includes both dystonia and choreoathetosis subscales and can therefore be used to distinguish these conditions. Both 5-level DIS subscales evaluate durations and amplitudes in 12 body regions and can be assessed using video records.[23] However, only moderate evidence is available regarding the internal consistency and inter-rater reliability of the DIS, and no existing evidence concerns intra-rater or test-retest reliability.[21] The MD-CRS is a comprehensive battery of scales used to evaluate the features of movement deficits and includes symptoms such as hypokinetic/rigid syndrome, chorea/ballism, dystonia/athetosis, myoclonus, tic, and tremor. The MD-CRS can be also conducted using video records and used to distinguish dystonia from other symptoms.

The Toronto Western Spasmodic Torticollis Rating Scale (TWSTRS) is used to evaluate cervical dystonia.[24] This scale includes the three subscales addressing severity, disability, and pain. The TWSTRS-severity subscale is a 6-item, investigator-reported scale that evaluates the maximal excursion, duration, effects of sensory tricks, shoulder elevation/anterior displacement, range of motion, and time. Here, the maximum total score is 35 points, with higher scores reflecting more severe dystonia. The TWSTRS-disability subscale is a 6-item self-reported scale that addresses work performance, ADL, driving, reading, watching television, and leisure activities outside the home. The scores for each item on this subscale range from 0 (no dystonia) to 5 (severe dystonia). Scores for the TWSTRS-pain subscale range from 0 (no pain) to 5 (severe pain). Although the TWSTRS is a valid and reliable tool for evaluating cervical dystonia, it is seldom used in children.

Few objective evaluations of dystonia are available. Only one previous study mentioned the use of hand-drawn kinematic data collected via iPad from two adult patients with dystonia (mean age: 58.5 ± 17.7 years).[25] No pediatric data are available.

3.3 | Chorea and ballism

Chorea, defined as involuntary, jerky, arrhythmic, and abrupt movements, may be caused by many factors, including choreatic disorders and Huntington disease (HD). Chorea can affect the whole body, especially the proximal limbs and face and may sometimes move among parts of the body. Ballism, defined as a series of involuntary but violent movements, usually affects the proximal limbs and may be attributable to a severe form of chorea.

Chorea is a complex condition with variable clinical manifestations. Currently, only 1 subjective scale for chorea is available: the Universidade Federal de Minas Gerais Sydenham Chorea Rating Scale (USCRS)[26] is usually used to evaluate Sydenham's chorea (SC).[27] This is a 27-item, investigator-reported scale with scores ranging from 0 (no symptoms/signs) to 4 (severe chorea) and includes three sections: behavioral, ADL, and motor assessment. The USCRS is considered valid and reliable and can be used to evaluate both children and adults but cannot be used to distinguish tics from chorea. Furthermore, the USCRS evaluation is complicated and requires an experienced investigator. No information is available regarding the clinimetric properties of this scale.

Currently, the Unified Huntington's Disease Rating Scale (UHDRS) is the most commonly used tool to evaluate HD.[28] This scale assesses motor and cognitive performances, behavioral abnormalities, and functional capacity. The motor section includes 15 items with scores ranging from 0 (normal) to 4 (severe), of which one item addresses maximal chorea. Although this scale was not specifically designed to evaluate chorea, it is semi-objective and accompanied by a convenient-to-use teaching video. Furthermore, many studies have proven the reliability of the UHDRS. Accordingly, this scale is considered useful for evaluating the overall function and primary features of HD. The only drawback, however, is the time-consuming nature, as approximately 30 minutes are required to perform a single examination. For a briefer and simpler examination, some reports have applied a modified UHDRS motor score to rapidly evaluate responses to a certain treatment. Additionally, the Abnormal Involuntary Movement Scale is a 12-item clinician-reported scale that evaluates abnormal movements in HD.[29] The scores for this scale range from 0 (normal) to 4 (severe). However, its characteristics for HD have never been reported. Many modified versions of the Abnormal Involuntary Movement Scale are available. Compared with the UHDRS, this scale is concise and requires only 10 minutes per examination. However, the Abnormal Involuntary Movement Scale was designed to assess tardive dyskinesia and cannot distinguish between choreic or dystonic dyskinesia.[6]

Notably, SC has been accepted as a neuropsychiatric disorder and has accordingly been evaluated using many neuropsychological tests.[30] Additionally, comprehensive scales such as the DIS, Gross Motor Function Measure, and MD-CRS have been used to evaluate chorea in patients with different underlying conditions.[21,31] The use of these scales for HD has also been described.

Regarding objective assessments, 1 report described the use of surface EMG to record muscular activity,[32] while another used electroencephalography (EEG) to evaluate the processing mode of action.[33] However, these methods are not commonly applied. In other words, a standard objective assessment for chorea remains unavailable.

3.4 | Tics

Tics comprise the most common type of childhood-onset PMD.[5] Tics, which differ from chorea, are repeated, involuntary, intermittent movements that may affect limited groups of muscles and may be accompanied by neuropsychiatric diseases. Tourette syndrome (TS) is the most important disorder involving tics. The Diagnostic and Statistical Manual for Disease, 5th edition (DSM-5)[34] (some reports have used the older version, or DSM-4 TR) is used to rapidly diagnose tics.[35] However, this manual includes global diagnostic criteria without quantitative measurements.

The Yale Global Tic Severity Scale (YGTSS) is the most commonly used subjective tic evaluation scale.[36] The YGTSS evaluates the total severity scores of both vocal and motor tics ranging from 0 to 5, as well as impairment scores ranging from 0 to 50. Higher scores indicate more severe symptoms. Recent studies have widely used various translations of the YGTSS into several languages, which have been proven valid and reliable.[37,38] Therefore, the YGTSS is a satisfactory instrument for the evaluation of tics and can provide a global impression of the associated clinical features. Moreover, this scale can be used to differentiate motor and phonic tics. Many studies have confirmed the good clinimetric properties and especially the reliability and validity of the YGTSS.[6,39] The examination process is brief and can be completed within 15 minutes.

Regarding clinician-reported scales, Robertson[40] developed the TS Diagnostic Confidence Index with which the likelihood of TS is evaluated using scores of 1-100. Although this index is currently used widely,[35] little information is available regarding its validity and reliability. The Tourette Syndrome Clinical Global Impression (TS-CGI) is yet another scale used to evaluate tic severity. This 7-item investigator-reported scale yields scores ranging from 1 (no tics) to 4 (severe).[41] The concise and operative TS-CGI is widely used as an adjunctive instrument with which clinicians can measure both motor and non-motor symptoms related to tic disorders.[42] The Tourette Syndrome Global Scale (TSGS) is an investigator-reported scale used to measure tics and social functioning in patients with TS.[43] It includes eight items intended to measure simple and complex motor and simple and complex phonic disruptions, with scores ranging from 0 (none) to 5 (severe). Another five items on the TSGS assess social functioning related to tics, with scores ranging from 0 (no problem) to 25 (severe). Although this scale has been used in interventional studies, no information is available regarding its reliability and validity. Additionally, the TSGS is complex, and the social functioning scores are disproportionately weighted to the tic symptoms.[6] The Unified Tic Rating Scale (UTRS) comprises both objective and subjective measurements.[44] The former involves a 2-minutes period tic counting, and the subscales include the distribution, types, frequency, and intensity of tics and the degree of interference and suppression. The UTRS also measures symptoms in other psychiatric orders. However, no additional information regarding its reliability and validity is available.

Of the available parent-reported scales, the Child Tourette Syndrome Impairment Scale (CTIM) is used to evaluate the potential impairments caused by tics in home and school settings and during social activities.[45] However, this 37-item scale is somewhat complicated. Recently, Barfell et al. developed a Mini-CTIM comprising only 12 items related to school, home, and social activities with the aim of evaluating parent-child results, group differences, and symptom severity corrections. The authors found that this concise scale is a practical tool for assessing both tic- and non-tic-based impairments.[46] The widely used 14-item Parent Tic Questionnaire (PTQ) evaluates motor and vocal tics and provides information about both frequency (1: weekly, 2: daily, 3: hourly, 4: constantly) and intensity.[47] The PTQ was found to validly and reliably assess tic severity.[42] The TS Severity Scale (TSSS) is an older parent-reported tool[48] comprising 5 original scales with a focus on the social effects of tics, namely the degree to which the tics are noticeable, whether they elicit comments or curiosity, whether the patient is considered odd or bizarre, whether the tics interfere with functioning, and whether the tics lead to incapacitation or to the patient being homebound or hospitalized. The scores for this scale range from 0 (none) to 9 (very severe). The TSSS is considered reliable and valid and is therefore used in interventional studies of TS.[6] Despite the compact and simple nature of the TSSS, however, it provides a weak evaluation of tic severity.[49,50] The Tourette's Disorder Scale (TODS) can be used by either a clinician (TODS-CR) or a parent (TODS-PR).[51] This concise scale includes 15 items, with scores ranging from 0 (no tics) to 10 (severe), and was shown to be valid and reliable.[52] However, a previous report noted that some items of the TODS focused on symptoms other than tics.[49] The Hopkins Motor and Vocal Tic Severity Scale is a 10-item scale (five items each for motor and vocal tics) with scores ranging from 0 (no tics) to 4 (severe). This scale can also be used by both clinicians and parents.[53] Advantageously, the Hopkins Scale can be used to observe changes in specific tics. However, this scale is mainly limited by an inability to separately assess parameters such as tic frequency and intensity.[49] Still, this instrument exhibited excellent reliability and validity when compared with other scales.[54]

Of the available self-reported scales, the TS Questionnaire is a 35-page survey used to collect information about the history of tics, course of tic behaviors, and effect of tics on daily life.[55] Although this questionnaire includes many potential risk factors for tics, the large-scale nature makes it unsuitable for children. Furthermore, parent-reported items are often subject to recall bias.[49] Other self-reported

scales include the TS Symptom list[56] and the Motor Tic, Obsessions and Compulsions, and Vocal Tic Evaluation Survey.[57] The latter is a 20-item scale intended to investigate the presence of symptoms in the previous 4 weeks. The scores range from 0 (never) to 4 (severe). A previous study evaluated the reliability of this scale and observed that some items exhibited low standard correlation coefficients.[57] However, recent relevant studies have seldom used these tools, given the limitations of self-reporting in children. Although some studies have used the Premonitory Urge for Tics Scale (PUTS)[58] to evaluate tic-related premonitory urges,[42,59] this tool does not assess tics; rather, it is a 10-item scale that assesses 10 somatic sensations related to tics. The scores from this scale range from 0 (never) to 4 (severe). Studies have indicated that the PUTS is only acceptable for use in older children[60] and is not satisfactory for children aged <10 years.[58] The commonly used subjective assessments of tics are listed in Table 2.

Among the objective assessment tools developed for tics, video tape ratings and tic counts are the most classic. Goetz[61] developed the most widely used video-based clinical rating scale for tics, which was later modified.[62] This scale includes five items with scores ranging from 0 (no tics) to 4 (severe). All scoring is based on the counting of tics in a 10-minutes video. The greatest advantage of this test is its objective nature, which avoids observation bias. This scale has therefore been widely used since its development. Another study by Chappell et al.[63] used a tic-counting method as an objective evaluation. There, children were recorded under five conditions: being alone, doing homework, watching television, talking to a stranger, and receiving attention when ticcing.[42] The frequency and number of tics were measured in each condition. Although the subjects are recorded in a free-moving state, this evaluation may occasionally be stressful for the children. Furthermore, this tool requires an expensive device but measures only the frequency of tics. Importantly, the expense associated with video evaluation makes it impossible to record one child for a relatively long time, leading to potential sampling bias among children whose tics fluctuate widely.[49] Recently, Brabson used a mobile phone to perform visual assessments,[64] which could be considered a type of video assessment. Liu et al. employed the SMART Balance Master® 8.2 system to evaluate postural stability in children with tics. The authors of that study found that children with TS experienced difficulties in maintaining postural stability, which may be considered a tic-related impairment.[65]

3.5 | Myoclonus

Myoclonus is defined as a sudden, repeated, involuntary, shock-like muscle jerk that may affect a single region or the entire body. This is the briefest and most rapid of all PMDs. Myoclonus may occur in normal children as a physiologic phenomenon (eg, hiccups) and/or during sleep. Importantly, however, it may also be a prominent symptom of many disorders. Although it is sometimes difficult to distinguish myoclonus from mimics (ie, jerks and twitches), children with myoclonus may require a rapid but appropriate assessment to detect the potential cause, which might be life-threatening. Accordingly, the selection of an effective assessment tool is crucial.

Few reports are available concerning subjective assessments designed specifically for myoclonus, possibly because of the complicated manifestation and pathogenesis of this condition. Currently, the most important myoclonus evaluation tool is The Unified Myoclonus Rating Scale (UMRS), which was developed by Frucht.[66] This scale includes six sections: patient questionnaire (11 items, scores of 0-4), myoclonus at rest (8 items, scores of 0-16), stimulus sensitivity (17 items, scores of 0-1), myoclonus with action (10 items, scores of 0-16), function tests (7 items, scores of 0-4), and global disability score (1 item, scores of 0-4). A higher score indicates more severe myoclonus. This investigator-reported scale has been used in recent pediatric studies.[67] The UMRS is easy to use, and the examination can be completed in 15 minutes. Although no information is available regarding the validity and reliability of this scale, a previous review mentioned its good inter-rater reliability.[6]

Wendy (2015)[68] introduced the Opsoclonus Myoclonus Syndrome Rating Scale for the evaluation of myoclonus in children. This 6-item investigator-reported scale yields scores of 0 (normal) to 3 (several), and the scoring system is concise and clear and thus convenient. However, information is not available regarding the validity and reliability of this scale.[68] Magaudda[69]established a highly simplified 5-point scale to evaluate the severity of myoclonus, which may be suitable for rapid global assessments.[70] Tate[71] developed the concise, semiquantitative Pediatric Myoclonus Scale Scoring Form for use in children. This form measures the frequency, intensity, and distribution of myoclonus during spontaneous (ie, resting), sensory, and action states, and yields scores ranging from 0 (normal) to 4 (severe). However, this investigator-rated scale is seldom used in current studies, and no additional information is available regarding its clinimetric properties.

Objective studies play a greater role in the diagnosis of myoclonus relative to other PMDs. For example, Espay[72] observed that an electrophysiological examination can be very useful for distinguishing myoclonus from mimics. Moreover, a comprehensive consideration of findings from electrophysiological assessments, including surface EMG, EEG (with stimulation), EEG-EMG (jerk-locked), long-latency reflex, cutaneous reflex, and transcranial magnetic stimulation, can provide clinical information about the source of the myoclonus (eg, spinal, cortical, or subcortical). In recent pediatric studies, EEG[73] and video EMG[74] were commonly used to demonstrate the origin of myoclonus. These electrophysiological methods are completely objective, effective, and specific and are therefore essential to an appropriate diagnosis/differential diagnosis of myoclonus. However, these methods are limited by the requirements for special devices and experienced clinicians, as well as high examination costs and the invasive nature of EMG, which might be resisted by some children.

3.6 | Ataxia

Ataxia is defined as an inability to perform a voluntary movement due to a failure of muscular coordination and is associated with

TABLE 2 Tools for the subjective assessment of tics

Scales	Developers	Contents	Behavior assessed	Brief commentary	
				Strengths	Weaknesses
Yale Global Tic Severity Scale (YGTSS)	Leckman (1989)[36]	Five-point Investigator-reported	Vocal and motor tics	Very classic and widely used scale Multiple language versions Valid and reliable	Subjective scale Requires an experienced investigator
DSM-5	American Psychiatric Association (2013)[34]	Diagnostic criteria for TS, Persistent (also called chronic) motor or vocal tic disorder and Provisional tic disorder	Rapid diagnosis of tic disorders	Simple Investigator can quickly obtain a global impression of the patient	Cannot be used for quantitative observation
Tourette Syndrome Global Scale (TSGS)	Harcherik (1984)[43]	8-items Investigator-reported	Tic symptoms and social functions	Global	Complex, need experienced investigator, poor relationship between tic symptoms and the social functions
Unified Tic Rating Scale (UTRS)	Kurlan 1993[44]	A 2-min tic count Measure of tic Measure of OCD and ADHD	Tic and symptoms of OCD, ADHD	Including objective and subjective measurements	Complicated
Tourette Syndrome Diagnostic Confidence Index (TSDCI)	Robertson (1999)[40]	26-item scale (Coprolalia, echophenomenon, complex tics, temporal tics, subjective and cognitive experiences, tic severity) Total score of 100	Distinguishes spasticity, dystonia, and rigidity	Simple and easy to perform	Validity and reliability are uncertain
Tourette Syndrome Clinical Global Impression (TS-CGI)	Leckman (1988)[41]	7-item, 4-level investigator-reported scale	Severity of tics	Valid and reliable Convenient to use	Subjective scale
Child Tourette Syndrome Impairment Scale (CTIM) and Mini-CTIM	Storch (2007)[45] Barfell (2017)[46] modified mini version	37-item parent-reported scale Mini-CTIM: 12-item scale including four categories (parent tic, parent non-tic, child tic, child non-tic)	Impairments caused by tics or non-tics	Valid and reliable scale Mini-CTIM is easy to use Can evaluate both tic and non-tic impairments	Parent needs to be trained Observation bias may be larger than investigator-reported scale Mini-CTIM requires further verification of validity and reliability
Parent Tic Questionnaire (PTQ)	Woods (2007)[47]	14-item scale including the frequency and intensity of motor and vocal tics Scores range from 0 (no tics) to 8 (severe) Parent-reported	Severity of tics	Valid and reliable Convenient to use	Observation bias as a parent-reported scale

(*Continues*)

TABLE 2 (Continued)

Scales	Developers	Contents	Behavior assessed	Brief commentary	
				Strengths	Weaknesses
Tourette syndrome severity scale (TSSS)	Shapiro (1984)[48]	5-item parent-reported scale Focuses on how much tics are noticed by others and degree of impairment	Social influence of tics	Concise and easy to use Valid	Observation bias as a parent-reported scale Weak evaluation of tic severity
Tourette's Disorder Scale (TODS)	Shytle (2003)[51]	15-item, 10-level scale	Evaluation of motor and phonics tics	Concise Valid and reliable Can be rated by both clinician and patient	Psychiatric symptoms other than motor and phonic tics are also included
Hopkins Motor and Vocal Tic Severity Scale	Walkup (1992)[53]	10-item, 5-level scale	Motor tics and vocal tics	Concise Convenient Can be rated by both clinician and patient Can follow changes in special tics	Does not allow separate evaluation of tic frequency, intensity
Tourette's Syndrome Questionnaire (TSQ)	Jagger (1982)[55]	35-page self-reported or patient-reported survey	Includes information about tic history, behaviors, and effects	Includes many risk factors for tics	Time-consuming Not suitable for some children Subject to recall bias

cerebellar dysfunction. This condition might involve the upper and lower limbs and trunk and may occasionally affect functions such as speech, swallowing, and even respiration.

The Scale for Assessment and Rating of Ataxia (SARA)[75] is the most important tool used to evaluate ataxia. This scale comprises eight items: gait (scores: 0-8), stance (0-6), sitting (0-4), speech disturbance (0-6), finger chase (0-4), nose-finger test (0-4), rapid alternating hand movements (0-4), and heel-shin slide (0-4). SARA has been translated into several languages[76] and is used to evaluate both the limbs and trunk; it is therefore considered a comprehensive scale for ataxia and is currently in wide use.[77] However, the reliability of the SARA remains controversial. Some studies have confirmed the validity of this scale for Friedreich's ataxia,[78] spinocerebellar ataxia,[75] and posterior fossa tumor.[79] Although additional reports indicate that the SARA is age-dependent and reliable in normal children,[80,81] another author objected to this conclusion.[82] In short, the SARA requires further assessments of validity and reliability.

The International Cooperative Ataxia Rating Scale (ICARS) is the most comprehensive scale and includes three sections to address posture and gait disturbances (7 items), kinetic functions (12 items), and speech disorders (5 items). The maximum total ICARS score is 100, with higher scores indicating more severe symptoms.[83] Although this scale includes all aspects of ataxia, its reliability and validity have been disputed by many researchers. In addition, the ICARS is complicated and therefore stressful and time-consuming for some patients. The subsequently developed Brief Ataxia Rating Scale (BARS) is based on the comprehensive SARA and ICARS.[84] The BARS includes only five items, gait (scores: 0-8), knee-tibia test (0-4), finger-to-nose test (0-4), dysarthria (0-4), and oculomotor abnormalities (0-2), and omits a test of speech. The maximum total BARS score is 30, with higher scores indicating more severe symptoms. The BARS is concise and easy to use, and subsequent studies verified its reliability and validity. Despite its brief nature, it is as valuable as the more complicated SARS for the evaluation of ataxia severity in children with posterior fossa tumors.[79]

The Friedreich Ataxia Rating Scale (FARS) includes the following measurements: a six-stage evaluation of ataxia and tests that target the upper limbs [finger-to-finger test (scores: 0-3), nose-finger test (0-4), dysmetria test (0-4), rapid alternating movements of hands (0-3), and finger taps (0-4)] and lower limbs [heel along shin slide (0-4) and heel-to-shin tap (0-4)]. This simple scale, which was found to be reliable and valid, can be easily administered by clinicians. The FARS has therefore been identified as a good tool for Friedreich ataxia.[85] Additionally, the ataxia-related subscale itself can be used as a rapid assessment of ataxia.

Objective assessments are usually used to test ataxia in the extremities. For example, gait evaluations are always performed using a camera motion capture system while children walk on a walkway, and a video is recorded for further analysis. Additionally, motor analysis software is used to record a battery of the walking parameters, which is later analyzed to evaluate the gait status.[86,87] This task is easy and minimizes the stress experienced by children but is not suitable for patients with severe gait disabilities. This test also

requires measures to prevent children from falling, as well as expensive instruments and complex data analyses. A 3-dimensional kinematic analysis system[88] and robotic system[89] have also been used to assess ataxia of the upper limbs. However, both assessment systems require special devices and challenging data analyses and are not widely implemented in clinical practice.

3.7 | Other types of movement disorders

3.7.1 | Tremor and stereotypies

Tremors and stereotypies are quite common in children.

Tremor is defined as a rhythmic, involuntary oscillation about a joint axis and can be classified as resting, postural, action, task-specific, or essential tremor.[5] A detailed description of the clinical features of tremor is extremely important, as it is useful for discriminating different tremors and determining an appropriate diagnosis.

The methods used to assess tremor in children differ considerably from those used in adults. Subjective tools commonly used for adults, such as the Clinical Rating Scale for Tremor, are rarely used for children. Instead, pediatric evaluations usually use the 5-spiral-drawing method,[90,91] wherein children are required to draw five spirals. First, an Archimedes spiral is drawn with the dominant hand for practice, followed by two spirals drawn with the dominant hand and two with the non-dominant hand. A blinded investigator then scores the spiral drawings as: 0 (no tremor), 0.5 (subtle), 1.0 (low-amplitude oscillations), 1.5 (low-amplitude oscillations along with oscillations, at times reaching moderate amplitude), or 2 (moderate-amplitude oscillations).[91] This brief and convenient method is suitable for large epidemiological investigations. However, it subjectivity and its reliability and validity are unknown. Objective methods, such as EMS, are invasive and thus seldom used in children. We expect that the development of wearable sensors will facilitate the objective evaluation of tremor in children.[4]

Stereotypies are defined as repetitive, rhythmic movements that may be predictable and can be voluntarily suppressed. Both normal children and those with disorders such as CP and autism spectrum disorders (ASD) may exhibit stereotypies. Recent studies have applied several subjective assessments of stereotypies.[92,93] For example, the Stereotypy Severity Scale (SSS), developed by Miller,[94] is a 5-item, investigator-reported scale that measures motor function and impairments; the scores of the former and latter categories range from 0 to 18 and from 0 to 50, respectively. The SSS can be used to measure self-respect and social acceptance related to stereotypies. However, no information is available regarding the reliability and validity of this scale. The Motor Severity Stereotypy Scale (MSSS), which was developed by Johns Hopkins Hospital, is a 5-item scale that evaluates the number (scores: 0-5), frequency (0-5), and intensity of stereotypies (0-5), as well as interference (0-5) and a global impairment rating (0-50). Higher scores indicate more severe symptoms. Although the MSSS is rapid and easy to use, its validity and reliability are unknown.

The Repetitive Behavior Scale-Revised (RBS-R), described by Lam,[95] is a 44-item self-reported tool used to measure repetitive behaviors in children. The stereotyped behavior subscale includes six questions with scores ranging from 0 (normal) to 3 (severe). This comprehensive scale assesses self-injurious and compulsive behaviors and is therefore used for psychiatric diseases like ASD. However, the reliability and validity of this scale for movement disorders are unknown. The Stereotypy Linear Analog Scale is a line on which parents can rank the stereotypies exhibited by their child during the past few days from 0 (best) to 10 (severe) with regard to the frequency, intensity, and number of events. This simple tool provides a subjective parental impression and therefore should be used along with other assessment tools. Only one study reported the use of an objective assessment of stereotypies; this video-based study quantitatively assessed the number and frequency of stereotypies in affected infants.[96]

3.7.2 | Rigidity and bradykinesia

Rigidity, defined as an isokinetic increase in resistance to passive movement, is a common hypertonic disorder in adults but is rare in children. The previously described HAL tool (please see the spasticity section) includes a subjective assessment of rigidity.[13] Additionally, rigidity is addressed by other comprehensive assessments of motor deficits in children, including the Modified Melbourne Assessment, Quality of Upper Extremity Skills Test, Pediatric Evaluation of Disability Inventory, Movement Assessment Battery for Children, and MD-CRS. Given the scope of the present study, however, we will introduce these scales elsewhere.

Tremors, rigidity, and bradykinesia can be considered symptoms of Parkinsonism. Numerous conditions present with Parkinsonism in childhood, including HD, Wilson's disease, and neurodegeneration associated with brain iron accumulation disorders. In fact, most of these conditions are considered dystonia-Parkinsonism syndromes.

Athetosis is a controversial condition. Some authors consider it a variant of dystonia,[5] whereas others consider it a form of post-hemiplegic chorea or a point on a continuum between chorea and dystonia.[97] Currently, information regarding assessments of athetosis is limited.

4 | CRITICAL THINKING REGARDING THE DEVELOPMENT AND SELECTION OF PEDIATRIC BEHAVIORAL ASSESSMENTS

The physiological and pathophysiological features of children differ from those of adults; accordingly, these differences should be considered separately when developing and selecting behavioral assessments. Note that many measurements for adults cannot be directly applied to children without modification. The following points should be considered:

1. Invasive (ie, painful), excessively complicated, and time-consuming tests should be avoided when evaluating children. Moreover, self-reported scales should only be used for older children with sufficient writing and comprehension abilities. Observation and selection biases should be considered when applying parent-reported scales. Measures aimed at avoiding potential dangers (eg, fall risk during gait evaluation) should be adopted.
2. Assessment tools should be considered according to the age of the child, which differs from standard practices regarding adult assessment. For example, PUTS is suitable for the measurement of tic-related premonitory urges only in children aged >10 years. Accordingly, some assessments, particularly those regarding ataxia and speech, should be divided into versions for infants, older children, and adolescents.
3. When evaluating children, the principles of behavioral test development and selection summarized in our previous studies,[3,4] namely the OMS principles, are extremely important. Observation bias cannot be avoided when using parent-reported and investigator-reported subjective assessments. Therefore, a trend toward objectification has been observed for future assessments. Classical objective assessments use video cameras to record kinetic parameters and/or the number and frequency of abnormal movements. In the future, wearable sensors and robotic technology may considerably improve objective measurements, particularly in terms of real-time assessments and feedback. Additionally, virtual reality (VR) and augmented reality (AR) can be used to design a battery of game-based assessments that would be easily accepted by children. Particularly, simplification and multi-purpose applications can improve experimental efficiency, as children are clearly more willing to accept a simple test.
4. In some PMDs, the neurologic and psychiatric aspects are deeply intertwined (eg, tics, stereotypies, and chorea). Moreover, it is occasionally difficult to distinguish between complex tics and compulsions in patients with tic disorders.

A satisfactory behavioral test should be able to distinguish among the complicated clinical symptoms and features of PMDs. Such discrimination is extremely important during the development and selection of new assessment tools for PMDs.

5 | LIMITATIONS OF THIS STUDY

The main limitation of this study is that we did not make a systematic (and quantified) review as per the Preferred Reporting Items for Systematic Reviews and Meta-Analyses rules, which would provide stronger evidence for evaluation of these behavioral assessments. In fact, RCTs for evaluating behavioral testing in children have been so limited that we could not "include" or "exclude" any studies according to their quality. Information regarding behavioral testing from all studies (RCT or not) was included in the present study. We will make a systematic and quantified review concerning the behavioral assessments in PMDs in our future studies if sufficient data from RCTs are available. In this regard, we appeal to perform more rigorous RCTs that have been specifically designed to evaluate behavioral testing in children in the future.

6 | CONCLUDING REMARKS

This review has summarized the behavioral assessment tools commonly used in recent and current studies of PMDs. We recommend that assessments of children should fully consider the distinct and characteristic physiological and pathophysiological features of this population. The principles of OMS are crucial to the selection and development of behavioral assessments, especially for children. Furthermore, discrimination is important. We expect that the development of wearable sensors, AR, and VR will lead to more effective and simple tests in the future.

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CONFLICT OF INTEREST

The authors declare that they have no conflicts of interest related to the present study.

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