Development of the Hypertonia Assessment Tool (HAT): a discriminative tool for hypertonia in children

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LIST OF ABBREVIATIONS

HAT	Hypertonia Assessment Tool
KR-20	Kuder–Richardson Formula 20
PABAK	Prevalence-adjusted bias-adjusted kappa

AIM The aim of this study was to develop a tool to identify paediatric hypertonia subtypes.
METHOD Items generated by experts were subscaled (spasticity, dystonia, rigidity). The tool was administered to 34 children (19 males, 15 females, mean age 8y 2mo, range 2y 5mo–18y 7mo) with hypertonia and cerebral palsy (CP) in Gross Motor Function Classification System (GMFCS) levels: I, *n*=7; II, *n*=5; III, *n*=7 level IV, *n*=7; and level V, *n*=8 level. Kuder–Richardson Formula 20 determined internal consistency. To assess reliability, two physicians administered the tool to 25 additional children with CP (15 males, 10 females; mean age 10y 8mo; GMFCS levels I, *n*=4; II, *n*=3; III, *n*=7; IV, *n*=4; and V, *n*=7) on two occasions, 2 weeks apart. To evaluate validity, a third physician diagnosed the hypertonia by neurological examination.
RESULTS The internal consistency of the spasticity items was moderate (α=0.58), and dystonia was high (α=0.79). Item reduction eliminated seven of the 14 original items. The agreement of the spasticity and rigidity subscales was adequate (prevalence-adjusted bias-adjusted kappa [PABAK] ranging from moderate [0.57] to excellent [1.0]) for validity, test–retest reliability, and interrater reliability. For dystonia agreement was lower, with PABAK ranging from fair (0.30) to good (0.65). Eighty-seven per cent had spasticity and 78% had dystonia.
INTERPRETATION The Hypertonia Assessment Tool has good reliability and validity for identifying spasticity and the absence of rigidity, and moderate findings for dystonia.

Hypertonia is defined as 'abnormally increased resistance to externally imposed movement about a joint'.[1] Hypertonia is observed in a variety of paediatric neurological conditions, most commonly cerebral palsy (CP), defined as 'a group of disorders of the development of movement and posture, causing activity limitations that are attributed to non-progressive disturbances that have occurred in the developing fetal or infant brain'.[2]

There are three subtypes of neurologically mediated hypertonia: spasticity, dystonia, and rigidity. Spasticity is hypertonia in which 'resistance to externally imposed movement increases with increasing speed of stretch and varies with the direction of joint movement and/or in which resistance to externally imposed movement rises rapidly above a threshold speed or joint angle'.[1] Dystonia is 'a movement disorder in which involuntary sustained or intermittent muscle contractions cause twisting and repetitive movements, abnormal postures or both'.[1] Rigidity is a velocity-independent bidirectional resistance which may involve simultaneous co-contraction of agonists and antagonists. Mixed tone occurs when two subtypes of hypertonia are present.

Numerous scales to quantify the severity of hypertonia, spasticity, or dystonic postures have been developed, including the modified Ashworth Scale,[3] the Tardieu scale,[4,5] the Burke–Fahn–Marsden Scale,[6] and the Barry–Albright Dystonia Scale.[7] However, a standardized clinical tool to differentiate between the subtypes of hypertonia is not available. The development of a discriminative tool is required to serve this purpose. In this context, a discriminative tool differentiates between groups that share a common characteristic, in this case hypertonia.[8]

Neurological examination is currently the only method used to differentiate subtypes of hypertonia. The neurological examination, however, lacks standardization, and the outcome is often influenced by the experience of the clinician. Identifying and distinguishing the subtypes of hypertonia is becoming increasingly important in both the clinical and research settings. For example, treatment strategies, including medication type and dosing, may differ depending on the type of hypertonia present. In addition, a standardized discriminative clinical tool would allow research studies to recruit and report on individual outcomes more specifically. The objective of the

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current study was to develop a standardized discriminative tool to differentiate between spasticity, dystonia, and rigidity in the paediatric population.

METHOD

The Guyatt framework for measure development was followed in the development of this discriminative instrument.[8] The components of measure development include item generation and reduction and assessment of reliability and validity.

Ethical approval for this study was obtained from Bloorview Kids Rehab Research Ethics Board and participants and/or their caregivers provided informed consent for both the research and the publication of the results.

Item generation

The purpose of the item generation stage was to develop a comprehensive list of items that discriminate between spasticity, dystonia, and rigidity in children. A preliminary pool of items for the Hypertonia Assessment Tool (HAT) was generated from discussion with members of the National Institutes of Health Task Force on Childhood Motor Disorders.[1] This panel of experts included neurologists, physiatrists, orthopaedic surgeons, developmental paediatricians, physical therapists, and occupational therapists. The items generated were grouped into three subgroups – spasticity, dystonia, and rigidity – based on expert opinion. Further item generation was achieved through small group sessions and individual telephone interviews with professionals experienced in paediatric hypertonia (n=14). The structured interview format included open-ended questions and specific probes. Interviews were conducted until item saturation was reached (defined as three consecutive interviews failing to produce any new items).

Item reduction

The objective of the item reduction stage was to decrease the number of items in the assessment tool to make it more practical to administer, while retaining the tool's discriminative ability. A research assistant consecutively approached families for recruitment in a tertiary-level hypertonia clinic at a paediatric rehabilitation centre. The children varied in age, type of hypertonia, limb involvement, and functional ability. An examiner administered the three subgroups of items on one randomly chosen hypertonic limb in each of 34 children. The Kuder–Richardson Formula 20 (KR-20), a test of homogeneity of items for dichotomous variables, was used to determine the internal consistency of items within the subgroups of spasticity, dystonia, and rigidity. Items were eliminated if removal of the item increased the KR-20 alpha level for the subgroup to 0.7–0.9.

Evaluation of reliability and validity

A research assistant recruited a separate sample of children with hypertonia, of different ages and varying in type of hypertonia, limb involvement, and severity of physical disability, by consecutively approaching families presenting to a tertiary-level hypertonia clinic at a paediatric rehabilitation centre. Three physicians examined the children independently and were blinded to each other's scores. Two of the physicians (AJ and DF) administered the HAT to the same randomly chosen limb. The third physician (JM), a paediatric neurologist with expertise in movement disorders, administered a paediatric neurological examination on the same limb and designated the type of hypertonia present. After 2 weeks, the same group of children were re-examined using the HAT by the first physician (AJ).

Individual item validation

Each item in the original HAT tool was assessed for its ability to agree with the type of hypertonia diagnosed by the paediatric neurological examination. Items that had >50% agreement were retained as part of the final HAT.

Reliability and validity testing

To evaluate interrater reliability, a comparison of the HAT diagnoses by the two physicians (AJ and DF) was undertaken. To evaluate test–retest reliability, a comparison of the HAT diagnoses by AJ 1 at time 1 and time 2 was carried out. Criterion validity was assessed by comparing the HAT diagnosis of physician 1 and 2 with the neurological diagnosis made by physician 3 (JM, paediatric neurologist). Statistical agreement was assessed with analysis of the two by two tables with positive and negative agreement, and prevalence-adjusted bias-adjusted kappa (PABAK).[9,10] For the indices, the strength of the agreement was defined as slight (0–0.20), fair (0.21–0.40), moderate (0.41–0.60), substantial (0.61–0.80), or excellent (>0.80).[11]

RESULTS

The Task Force on Childhood Motor Disorders generated an initial eight items during a meeting held at the National Institutes of Health in 2001. Six additional items were added after the completion of 14 semi-structured interviews with experts in the field. At the completion of the item generation phase, the HAT had 14 items (Table I).

A randomly chosen hypertonic limb of 34 children with CP (19 males, 15 females; mean age 8y 2mo; range 2y 5mo to 18y 7mo) was examined in the item reduction stage of the study. The children's functional abilities ranged across all Gross Motor Function Classification System (GMFCS) levels (n=7 level I, n=5 level II, n=7 level III, n=7 level IV, and n=8 level V). The two history items were eliminated because caregivers and children had difficulty understanding and answering the questions posed. The KR-20 for each item is outlined in Table I. The initial KR-20 for spasticity items demonstrated moderate internal consistency (α=0.58). Item 12 was eliminated and the internal consistency of the spasticity subgroup increased (α=1.0). The dystonia items demonstrated high internal consistency (α=0.79) and no dystonia item was eliminated at this stage of the tool development. The rigidity items were not evaluated with the KR-20, as more than two items are required for this statistical analysis.

A further 25 children with CP (15 males, 10 females; mean age 10y 8mo; range 4–19y) were recruited for the reliability and validity evaluation. Functional abilities ranged across all

Table I: Internal consistency (measured by KR-20) of items generated after expert interviews, categorized into dystonia, spasticity, and rigidity, and agreement of items with the neurological diagnosis

Item description	Type of hypertonia identified by item	KR-20α[a]	Agreement (%)
(1) Caregiver history of variability in tone with sleep compared with awake time	Dystonia	0.77	NA
(2) Caregiver history of an increase in tone with activity/movement	Dystonia	0.75	NA
(3) Involuntary twisting movements	Dystonia	0.74	43
(4) Variable abnormal postures	Dystonia	0.73	43
(5) Increased involuntary movements/postures with purposeful movement of a distant body part	Dystonia	0.72	78
(6) Increased involuntary movements/postures with tactile stimulus of a distant body part	Dystonia	0.78	60
(7) Fluctuation of tone with multiple passive stretches	Dystonia	0.76	26
(8) Intermittent low tone during a passive stretch of the muscle	Dystonia	0.72	47
(9) Increased tone with purposeful movement of a distant body part	Dystonia	0.82	78
(10) Velocity-dependent resistance to passive stretch	Spasticity	0.38	65
(11) Presence of a spastic catch	Spasticity	0.38	78
(12) Hyperreflexia	Spasticity	1.0	NA
(13) Equal resistance to passive stretch in bidirectional passive movement of a joint	Rigidity	NA	96
(14) Maintenance of limb position after passive movement	Rigidity	NA	100

[a]The number reflects the Kuder–Richardson 20 (KR-20) index when the item is eliminated. NA, not applicable.

GMFCS levels (*n*=4 level I, *n*=3 level II, *n*=7 level III, *n*=4 level IV, and *n*=7 level V). Eighty-seven per cent of the children had evidence of spasticity and 78% had evidence of dystonia, as determined by the neurological examination. No child was diagnosed with rigidity. Individual item validation (evaluating agreement of each item with the neurological diagnosis) led to four items (items 3, 4, 7, and 8 from Table I) with <50% agreement being eliminated. The final revised version of the HAT is outlined in Appendix I, with administration guidelines for each item. There were seven items in total: two spasticity items, two rigidity items, and three dystonia items. Each item was scored 'yes' or 'no'. A positive score for at least one item of the subgroup confirmed the presence of the subtype of hypertonia.

Agreement matrices were created to evaluate test–retest/interrater reliability and validity, with results reported in Table II. An imbalance in the marginal totals was noted in 92% of the matrices (prevalence index >0.2). For example, in the spasticity and dystonia matrices there was a significantly higher proportion of 'yes' responses, and in the rigidity matrix there was a higher proportion of 'no' responses. To adjust for this imbalance, as recommended by Byrt et al.,[10] PABAK statistics were reported. In addition, positive and negative agreement indices were reported.[9]

For the identification of spasticity, test–retest reliability was excellent (1.0), interrater reliability was substantial (0.65), and validity was found to be moderate to good (0.57–0.74). For dystonia, test–retest reliability was moderate (0.43) and interrater reliability was fair (0.3). Results for validity were mixed, with agreement ranging from fair to substantial (0.3–0.65). For the absence of rigidity, the test–retest, interrater reliability, and validity agreement were all excellent (0.91–1.0). The HAT demonstrated higher positive agreement for identifying the presence rather than the absence of spasticity and dystonia, with the reverse pattern occurring for rigidity.

Table II: Results for test–retest reliability, interrater reliability, and validity, presented as positive and negative agreement and prevalence-adjusted bias-adjusted kappa (PABAK)

	Positive agreement	Negative agreement	PABAK
Spasticity			
Test–retest	1.00	0.10	1.00
Interrater	0.90	0.33	0.65
Validity (physician 1)	0.93	0.40	0.74
Validity (physician 2)	0.87	0.40	0.57
Dystonia			
Test–retest	0.80	0.50	0.43
Interrater	0.75	0.43	0.30
Validity (physician 1)	0.75	0.43	0.30
Validity (physician 2)	0.89	0.60	0.65
Rigidity			
Test–retest	0	1.00	1.00
Interrater	0	0.98	0.91
Validity (physician 1)	0	1.00	1.00
Validity (physician 2)	0	0.98	0.91

DISCUSSION

The HAT is a seven-item clinical assessment tool used to differentiate the various types of paediatric hypertonia, namely spasticity, dystonia, and rigidity. To date, there has not been a clinical tool which has allowed for the differentiation or discrimination of hypertonia in children. Expert knowledge of the salient features of each type of tone was used to establish the items.

We showed that the HAT has good interrater and test–retest reliability, as well as validity for the identification of

spasticity. For dystonia, the HAT demonstrated levels of agreement in the fair to moderate range. A hallmark feature of dystonia is its variability in clinical presentation, making the reliable identification of the presence or absence of dystonia more challenging. The tool demonstrated higher positive than negative agreement for spasticity and dystonia, indicating that the HAT is stronger in identifying the presence of, rather than the absence of, spasticity or dystonia. The reverse pattern was found for rigidity. We were able to demonstrate that the rigidity items were negative when rigidity was not present; however, we were unable to validate the ability of the rigidity items to identify rigidity as there was no child with rigidity in our study population. These findings concur with the imbalance in the prevalence in all three subtypes of hypertonia.[9]

Having a tool such as the HAT allows for standardization of the clinical examination used to identify the subtypes of hypertonia. This serves both a clinical and research purpose. Clinicians can improve their decisions regarding management when they are aware of the nature of the impairment. For instance, botulinum toxin for the treatment of hypertonia may require different doses and injection patterns in individuals with dystonia than for those with spasticity. Oral medications can be tone specific; for example, tizanadine is used for spasticity and trihexyphenydyl for dystonia. Within the clinical research setting, the ability to differentiate between types of hypertonia will allow increased specification of participant recruitment, which will aid in the interpretation and generalization of research studies.

An interesting finding from our study was the frequent identification of the presence of both spasticity and dystonia in the majority of children with CP. This was evident across all GMFCS levels, which were evenly distributed in our study population. The literature suggests that the majority of children with CP have 'spastic' CP; however, without a standardized way of assessing the various types of hypertonia present, the presence of dystonia can be underrecognized or overshadowed by the presence of spasticity. In addition, although paediatric rigidity is rarely confirmed clinically, it is not assessed in a standardized way. Rather than eliminating rigidity items, we have retained these items to allow for a more systematic assessment of rigidity in future population-based studies.

The items that described the classic observations of dystonia, specifically twisting and abnormal postures at rest, were not retained in the HAT. They were found to identify children with severe dystonia but were less useful for children with milder forms of dystonia.

For the validation component of the study we used a diagnosis by a paediatric neurologist with expertise in movement disorders as our criterion standard. Although this reflects current clinical practice, there is little information on the interrater reliability of diagnoses by neurologists. Currently, there are standardized quantitative methods to assess spasticity, such as the ramp and hold test,[12] but there are no validated quantitative objective tests for dystonia. As objective laboratory tests become available, validating the HAT against these measures will be helpful.

Future work on the HAT will relate to directing efforts towards improving the administration and interpretation of scoring for the dystonia items. An evaluation of the impact of videotaping of the administration of the dystonia items and review of the videotape before scoring may improve the HAT's psychometric properties. Developing a training video for the administration and scoring of items will also be helpful. Further work needs to be done to assess the impact of the severity of the hypertonia on the ability to 'diagnose' both spasticity and dystonia if both are present. For example, does severe dystonia preclude the ability to assess spasticity accurately? Additional limitations of our study were the lack of children under 4 years of age and the need to further evaluate the HAT in the presence of rigidity. Assessment of the HAT in a larger heterogeneous group of children with hypertonia will need to address these issues.

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APPENDIX I

Final version of the Hyperonia Assessment Tool (HAT) with a description of the administration procedures for each item

Items (in order of administration)	Type of hypertonia	Administration of item	Scoring (fill each box with 0 [negative] or 1 [positive])
Increased involuntary movements or postures of the designated limb with tactile stimulus of a distant body part	Dystonia	With the child at rest, observe involuntary movements of the designated limb as you gently rub a distant body part such as the shin or forearm	Dystonia is present if more involuntary movements or postures are observed in the designated limb with the tactile stimulus
Increased involuntary movements or postures with purposeful movement of a distant body part	Dystonia	Observe movements of the designated limb as the child carries out purposeful movements[a]	Dystonia is present if more involuntary movements or postures are observed in the designated limb with purposeful movement
Velocity-dependent resistance to stretch	Spasticity	Move the limb as described below[b] and assess for a change in muscle resistance between the slow and the fast stretch	Spasticity is present if there is an increase in resistance between the fast stretch compared with the slow stretch
Presence of spastic catch	Spasticity	Note the presence of a rapid rise (spastic catch) in resistance at a particular joint angle when moving the limb as described during the fast stretch[b]	Spasticity is present if a spastic catch is noted
Equal resistance to passive stretch during bidirectional movement of a joint	Rigidity	Assess this item during the fast stretch of the muscle[b]	Rigidity is present if the resistance felt is equal with movement in both directions
Increased tone with movement of a distant body part	Dystonia	Perform two additional fast stretches.[b] During the second stretch ask the child to do a purposeful movement[a] and assess for an increase in tone	Dystonia is present if greater tone is noticed when child is carrying out the purposeful movements
Maintenance of limb position after passive movement	Rigidity	For the arm, note the original position of the elbow; move the elbow by 45° into either flexion or extension and observe if the elbow returns to its original position. For the leg, note the original position of the ankle; move the ankle into 45° further dorsiflexion or plantarflexion and observe if the ankle returns to its original position	Rigidity is present if the limb remains in the final position of stretch rather than returning (partially or fully) to the limb's original position

Before administering the HAT, the child should be supine on the examining table. The child should be as comfortable as possible by having appropriate caregivers present, a roll placed under the knees, a comfortable room temperature, and unrestrictive clothing. Complete all items for the involved extremity being examined before moving on to the next involved extremity. [a]Based on the child's ability, ask the child to carry out two of the following for a 10-second period: (1) count to 10 slowly; (2) open and close one hand (into a fist) repeatedly (chose the hand that is not being examined); (3) open and close eyes (tight blinking) repeatedly; (4) reach for an object placed at least one foot away; and (5) visually track a brightly coloured object (e.g. red-tipped pen) or light source (e.g. flashlight). [b]Support the limb against gravity. Move the joints of the limb through the child's full range starting with the joint in full flexion or adduction, moving to full extension or abduction, and then returning to flexion or adduction, twice slowly and twice as quickly as possible. Upper extremity: shoulder adduction and abduction – begin with shoulder in full adduction; elbow flexion and extension – begin with elbow in full flexion; forearm pronation and supination – begin with forearm in full pronation; wrist flexion and extension – begin with wrist in full flexion. Lower extremity: hip adduction and abduction – begin with hip in full adduction; knee flexion and extension – begin with knee flexed with the hip in 90° flexion; ankle dorsiflexion and plantarflexion – begin with ankle in full plantarflexion.