Classification and Definition of Disorders Causing Hypertonia in Childhood

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ABSTRACT. Objective. This report describes the consensus outcome of an interdisciplinary workshop that was held at the National Institutes of Health in April 2001. The purpose of the workshop and this article are to define the terms “spasticity,” “dystonia,” and “rigidity” as they are used to describe clinical features of hypertonia in children. The definitions presented here are designed to allow differentiation of clinical features even when more than 1 is present simultaneously.

Methods. A consensus agreement was obtained on the best current definitions and their application in clinical situations.

Results. “Spasticity” is defined as hypertonia in which 1 or both of the following signs are present: 1) resistance to externally imposed movement increases with increasing speed of stretch and varies with the direction of joint movement, and/or 2) resistance to externally imposed movement rises rapidly above a threshold speed or joint angle. “Dystonia” is defined as a movement disorder in which involuntary sustained or intermittent muscle contractions cause twisting and repetitive movements, abnormal postures, or both. “Rigidity” is defined as hypertonia in which all of the following are true: 1) the resistance to externally imposed joint movement is present at very low speeds of movement, does not depend on imposed speed, and does not exhibit a speed or angle threshold; 2) simultaneous co-contraction of agonists and antagonists may occur, and this is reflected in an immediate resistance to a reversal of the direction of movement about a joint; 3) the limb does not tend to return toward a particular fixed posture or extreme joint angle; and 4) voluntary activity in distant muscle groups does not lead to involuntary movements about the rigid joints, although rigidity may worsen.

Conclusion. We have provided a set of definitions for the purpose of identifying different components of childhood hypertonia. We encourage the development of clinical rating scales that are based on these definitions, and we encourage research to relate the degree of hypertonia to the degree of functional ability, change over time, and societal participation in children with motor disorders. Pediatrics 2003;111:e89–e97. URL: http://www.pediatrics.org/cgi/content/full/111/1/e89; spasticity, dystonia, rigidity, movement disorders, hypertonia, pediatric, childhood.

ABBREVIATION. CP, cerebral palsy.

Abnormalities of tone are an integral component of many chronic motor disorders of childhood. These disorders result from dysgenesis or injury to developing motor pathways in the cortex, basal ganglia, thalamus, cerebellum, brainstem, central white matter, or spinal cord. When the injury occurs in children before 2 years of age, the term cerebral palsy (CP) is often used; when it occurs in older children, a variety of descriptive labels have been applied, depending on the cause. Childhood motor disorders are commonly classified into hypertonic or hypotonic groups on the basis of the abnormality of muscle tone.

At least 3 descriptive terms are associated with different forms of childhood hypertonia: “spasticity,” “dystonia,” and “rigidity.” Although some research laboratories have developed precise definitions for these terms, there has not been general agreement on the definitions as used in clinical situations. Current definitions have been based on adult disorders and the manifestations of spinal cord injury and therefore have not always led to consistent labeling of pediatric signs and symptoms by clinicians and researchers in different fields. Studies of appropriate rehabilitative interventions in chronic motor disorders of childhood have been hampered by the difficulty in establishing homogeneous cohorts for study as a result of varying classification systems. This is in large measure attributable to imprecision in the classification of abnormalities in tone as well as in categorizing the severity of functional impairments. There is therefore a need for a clear and consistent set of definitions that will allow accurate communication between clinicians as well as appropriate selection of children for medical therapy and clinical research trials. The ultimate purpose is to minimize disability and promote independence and full participation in society for children with motor disorders.

The goal of treatment of children with motor disorders mirrors the management of other forms of chronic disease and disability. The World Health Organization separates the issues of chronic diseases...
into 3 categories: impairment, functional ability, and societal participation. The National Center for Medical Rehabilitation Research model encourages those who evaluate outcome for disabling conditions to use a model of outcome that encompasses 5 axes: pathophysiology (underlying disease), impairment (clinically observable abnormality), functional limitations (effect on task performance), disability (effect on daily living), and societal limitations (effect on lifetime opportunities). Major obstacles to evaluation of outcomes within this model include limitations of measurement tools and a lack of objective criteria. In this context, this article provides specific clinical definitions of 3 types of hypertonia that are thought to cause specific impairment of movement.

This article presents a set of classifications and operational definitions that are designed to build the foundation for understanding how childhood hypertonia relates to other impairments and how it has an impact on function, disability, and societal participation. We consider definitions of 3 important types of hypertonia. The immediate goals of these definitions are 1) reliable communication between clinicians, 2) accurate distinction of diagnostic groups for clinical research, and 3) appropriate selection of patients for medical or surgical interventions. On the basis of these goals, the definitions must meet the following criteria:

- Utility: the ability to test a child easily in a routine clinical setting and assign appropriate labels that differentiate between spasticity, dystonia, and rigidity even when more than 1 feature is present simultaneously, as well as the eventual ability to confirm the findings by quantitative methods
- Reliability: the likelihood that different examiners will assign the same label to the manifestations of any given child (interobserver reliability) and that the same label will be assigned at different times by the same examiner (intraobserver reliability)
- Validity: the likelihood that the clinical definitions will agree with the assessments of expert clinicians (face validity), predict quantitative measurements (criterion validity), and predict the response to therapy (construct validity)

The definitions will draw on current knowledge of the pathophysiology of neuromuscular systems and on data resulting from objective and quantitative measures, when they are known. We recognize that hypertonia may be attributable to a wide range of underlying pathophysiology and will be associated with varying degrees of impairment, functional limitations, disability, or societal limitations. Multiple types of hypertonia may be present in the same child. Many motor syndromes may include hypertonia, and thus we acknowledge the frequent association between multiple impairments in affected children.

**CURRENT DEFINITIONS**

Current definitions of spasticity are based on velocity-dependent resistance or on presumed properties of increased sensitivity in the tonic stretch reflex response. The definitions sometimes incorporate possible anatomic localization, such as the “upper motor neuron syndrome,” or related clinical observations, such as a “spastic catch,” “clasp-knife response,” or clonus. Furthermore, the term “spasticity” is often used interchangeably with the term “upper motor neuron syndrome.” Current definitions of dystonia are based on the observation of particular abnormal postures or movements with sustained twisting qualities that are often associated with injury to the basal ganglia. Current definitions of rigidity are based on a constant resistance to passive motion that has a “plastic,” “malleable,” or “lead-pipe” quality. Although the current definitions provide a set of useful guidelines, we believe that they are insufficiently specific to distinguish between different findings, particularly when more than 1 is present simultaneously.

We use the term “motor disorder” to include disorders of multiple neural components, including basal ganglia, cerebellum, cerebral cortex, brainstem, and descending spinal tracts, because the term “movement disorder” is often used specifically to refer to disorders associated with presumed basal ganglia or cerebellar dysfunction. Hypertonia is a component of many motor disorders. In common clinical usage, motor disorders are often divided into pyramidal and extrapyramidal types. These terms have strong historical bases and have proved to have clinical utility, but it is increasingly recognized that the pyramidal and extrapyramidal motor systems are highly interconnected and interdependent. Pyramidal motor disorders result from injury to the corticofugal projections to the brainstem (corticobulbar) and spinal cord (corticospinal) at any point along their course. The corticospinal tracts were previously believed to be responsible for all aspects of the motor dysfunction, but recent evidence suggests that other regions must be involved.

Injury to these pathways often is associated with a combination of weakness and increased stretch reflexes. The weakness often occurs in a particular pattern referred to as “pyramidal” or “upper motoneuron” weakness. The pattern of pyramidal weakness can be position and state-dependent. Extrapyramidal motor disorders result from injury to the basal ganglia, cerebellum, or non-primary motor cortical areas, which often leads to abnormal motor control without weakness or changes in spinal reflexes. In children, both pyramidal and extrapyramidal motor disorders are most commonly seen as part of the syndrome of CP.

Commonly used classification schemes of CP divide the disorders into pyramidal (spastic) and extrapyramidal (dystonic, athetoid) types, but it is widely recognized that most children with CP have both pyramidal and extrapyramidal features. The coexistence of pyramidal and extrapyramidal signs can make determination of the relative contributions of these systems complex, because types of hypertonia including spasticity, rigidity, and dystonia often are present simultaneously. The complexity of the motor syndromes is superimposed on the unique process of growth (increase in size) and development (maturation of the central nervous system and acquisition of new skills through learning). Because of
growth and developmental plasticity, a static injury to the central nervous system may lead to a dynamically changing clinical picture that can be described as nonprogressive but ever-changing. Thus, children present unique challenges for diagnosis. The primary purpose of this article is to define objectively the terminology used to describe different types of hypertonia and to produce a more consistent use of descriptive terms for the subset of CP in which there is increased muscle tone.

Hypertonia

Innervated muscle exhibits viscous and elastic properties such that force is required to stretch a muscle from its resting position. The components of this muscle force include 1) the force generated by initially active muscle fibers, 2) the force attributable to stretch reflex action, and 3) the force attributable to passive tissue properties.

Accordingly, for clinical use, tone is defined operationally as resistance to passive stretch while the patient is attempting to maintain a relaxed state of muscle activity. Tone therefore in part reflects the state of active muscle contraction, and it may be either increased or decreased at rest. The definition of tone explicitly excludes resistance as a result of joint, ligament, or skeletal properties such as those that may occur with fixed deformities, including connective tissue disease or joint contractures. In many cases, such deformities can be distinguished from neuromuscular tone and they are not classified as hypertonia (examination during sleep or under anesthesia may be helpful in distinguishing such deformities).

Tone is assessed clinically using passive movements about a joint to determine muscular resistance. By our definition, tone is perceived by an examiner but not directly perceived by the patient. Assessment should include palpation of muscles to estimate the resting (baseline) state of muscle activation. Note, however, that tone is not rated by the presence or absence of muscle contraction at rest. Instead, our definition of tone requires an externally imposed movement to make the assessment. Tone may be measured in muscles that are at rest or in those with involuntary active contraction, but it should not be measured during voluntary muscle contraction.

Hypertonia is defined as abnormally increased resistance to externally imposed movement about a joint. It may be caused by spasticity, dystonia, rigidity, or a combination of features. We encourage the use of the terms spastic hypertonia, dystonic hypertonia, or rigid hypertonia to distinguish the primary feature. When hypertonia is so severe that imposed joint movement is not possible, then this subclassification cannot readily be performed. Mechanisms that lead to increased tone may also contribute to poor voluntary motor performance or involuntary muscle contractions, but assessment of tone is independent of strength, dexterity, coordination, or involuntary movements.

Spasticity

We emphasize that in spasticity there is a basic difference between the passive state of muscle tone in the clinical examination and the impairment of voluntary movement that leads to the child’s complaints. Because our task is to provide definitions for clinical use, we define spasticity here in terms of the features of the clinical examination. Nevertheless, we recognize that disability as a result of spasticity is more closely related to any associated deficits such as weakness or lack of coordination, as well as to the fact that the activity of functionally essential reflexes may be reduced or lacking in spastic children.

Spasticity is a velocity-dependent resistance of a muscle to stretch. We therefore define spasticity as hypertonia in which 1 or both of the following signs are present: 1) resistance to externally imposed movement increases with increasing speed of stretch and varies with the direction of joint movement, and/or 2) resistance to externally imposed movement rises rapidly above a threshold speed or joint angle.

The increased resistance specified in the first criterion is usually not directly proportional to the speed of stretch, and it may show only a modest dependence. However, the resistance must be different for high versus low speeds of passive movement and for flexion versus extension about the joint. The second criterion defines 1 feature of the “spastic catch” that often is felt on examination and that may represent the threshold for onset of the stretch reflex. The velocity dependence and threshold of the catch may reflect the stretch reflex threshold with initial recruitment of previously quiescent motoneurons. The threshold behavior might then be determined by the excitability of the motoneurons being assessed and by the starting length of the muscle. In this case, if motoneurons are highly excitable and if the muscle is stretched from an elongated initial position, then reflex threshold may be reached almost immediately, and a catch may not be evident.

Spasticity can vary depending on a child’s state of alertness, activity, or posture. Spasticity can be increased by anxiety, emotional state, pain, surface contact, or other nonnoxious sensory input. Spasticity may worsen with movement of the involved muscles or maintenance of the limb against gravity, but it is not specific to particular attempted tasks. The presence of spasticity suggests the presence of hypertonia, thus the terms spasticity and spastic hypertonia may be used interchangeably.

Electrophysiological studies of spasticity show changes in the threshold of the tonic stretch reflex such that resistance increases in magnitude or occurs sooner in the movement as passive speed is increased. The increased tone in spasticity as defined here may be attributable to a combination of the reflex component of muscle elasticity as well as to changes in muscle mechanical properties. Spasticity may be accompanied by a transformation of motor units such that tension development in the muscle occurs with lower levels of electromyograph activity, and such a change, if present, could...
contribute to efficiency of weight bearing. It is therefore possible that spasticity may either worsen or improve motor disability.

Because the features of spasticity included in this definition are measured using externally imposed velocity and joint angle changes, this suggests that spasticity as defined here depends on afferent feedback of proprioceptive information from muscle, joint, and skin receptors. Therefore, although afferent information may also contribute to other motor disorders, spasticity is inherently dependent on afferent feedback from muscle.

Spasticity will often coexist with other motor symptoms. Crothers and Paine enumerated criteria for spasticity based on the criteria of Wagley. Our present definition of spasticity encompasses only 1 of those criteria. Although the syndrome of spasticity as defined by Crothers and Paine has some validity, the multiple meanings of the term “spasticity” have led to significant confusion. Thus, we prefer to use the term “upper motor neuron syndrome” to refer to the clinical finding of spasticity plus at least 1 of the following: 1) hyperreflexia with or without clonus, 2) reflex overflow, 3) presence of a Babinski response, and 4) weakness that may primarily affect lower extremity flexor or upper extremity extensor muscle groups (“pyramidal distribution” weakness).

Confirming the distribution of weakness is the only part of the definition of the upper motor neuron syndrome that requires voluntary cooperation by the child on examination, and it may not be reliable in younger children. The remainder of the definition is dependent only on examination of the passive child.

We have chosen the term “upper motor neuron syndrome” because of its common usage, but we note that damage to the tracts that project to the lower motor neurons of the spinal cord does not in general lead to the findings listed above. The elements of the upper motor neuron syndrome can be divided into positive (hyperreflexia, overflow, and Babinski response) and negative (weakness, loss of dexterity) components. The positive symptoms—increased reflexes, clonus, and tone—may be associated with the release of the intact motor system from control. The negative symptoms—lack of agility, fatigability, and weakness—may be linked with the loss of a specific skill of central nervous system origin. Incoordination, loss of selective motor control, poor motor planning, and abnormal muscle activation patterns may occur, but these cannot be easily differentiated from findings attributable to coexisting ataxia or dystonia. The asymmetric tonic neck reflex is commonly present, but it may not be specifically related to spasticity. It is unclear the extent to which components of the upper motor neuron syndrome may represent the persistence or release of normally suppressed primitive or early developmental patterns of muscle activation.

Dystonia

Dystonia is an involuntary alteration in the pattern of muscle activation during voluntary movement or maintenance of posture. In general, dystonia is diagnosed by the observation of abnormal twisted postures or repetitive movements. Following earlier definitions, we define dystonia in childhood as a movement disorder in which involuntary sustained or intermittent muscle contractions cause twisting and repetitive movements, abnormal postures, or both.

Dystonia is commonly triggered or exacerbated by attempted voluntary movement and may fluctuate in presence and severity over time. The severity and quality of dystonic postures may vary with body position, specific tasks, emotional state, or level of consciousness.

Dystonia may cause hypertonia, but by this definition hypertonia is not always present in dystonia. For example, dystonia may lead to sustained involuntary muscle contraction only during attempts at voluntary movement, with normal or decreased tone and muscle activity when measured at rest. However, if dystonia is present at rest and causes an involuntary posture, then it may be a cause of hypertonia. We encourage the use of the term dystonic hypertonia for this condition. Hypertonia caused by dystonia is the result of tonically contracting muscles that contribute to passive joint stiffness as a result of the force generated by the initially active muscle fibers. A direct consequence is that dystonia is a cause of hypertonia only when there is muscle activity when the child is at rest and the limb is supported against gravity, or when muscle activity begins before the onset of externally imposed passive joint movement.

To diagnose dystonic hypertonia, there must be observable dystonic postures that do not relax during the examination of tone. The body part being examined must be supported against gravity to ensure that postural muscle activity is not contributing to the apparent tone. In dystonic hypertonia we expect to find all of the following: 1) resistance to externally imposed joint movement is present at very low speeds of movement, does not depend on imposed speed, and does not exhibit a speed or angle threshold; 2) simultaneous co-contraction of agonists and antagonists may occur, and this is reflected in an immediate resistance to a rapid reversal of the direction of movement about a joint; 3) the limb tends to return toward a fixed involuntary posture; and when symptoms are severe, the limb tends to move toward extremes of joint angles; 4) hypertonia is triggered or worsened by voluntary attempts at movement or posture of the affected and other body parts and may be strongly dependent on the particular movement or posture attempted or the activity of distant muscle groups; 5) the pattern as well as the magnitude of involuntary muscle activity varies with arousal, emotional and behavioral state, tactile contact, or attempted task; and 6) there is no other detected spinal cord or peripheral neuromuscular pathology causing tonic muscle activation at rest.

Note that these are features of dystonia when it causes hypertonia, but these features are not part of the definition of dystonia per se. Dystonia may be subclassified as action induced or posture induced. In adults, the actions that lead to dystonia may be restricted to certain attempted tasks, although task

Dystonia
specificity is less common in children. When dystonia is present at rest or with posture, certain attempted postures may be impossible to attain. Dystonia may be triggered or worsened by attention, distraction, startling, overuse, fatigue, touch, or pain. It is often exquisitely sensitive to postural and antigravity control, and it must therefore be tested seated, standing, supine, and with nearby joints in both flexion and extension. We emphasize again that dystonia is not necessarily a primary disorder of tone, but it may seem to be because of the inability to relax the muscles fully.

Dystonia and spasticity may occur in the same limb, and distinction requires determining the velocity-dependent, action-induced, and posture-responsive components. In particular, it may be difficult to distinguish dystonia from extensor posturing of the lower extremities, particularly when the extensor posturing is triggered by muscular effort. It may also be difficult to evaluate for spasticity in a muscle that is initially active as a result of dystonia, because the motoneuron pools may be in a suprathreshold state before the onset of externally imposed movement. When both spastic and dystonic hypertonia occur together, this is referred to as mixed hypertonia. In children, mixed hypertonia may be more common than either pure dystonic or pure spastic hypertonia. The term mixed hypertonia is preferred to older terms such as spastic dystonia or dystonic spasticity.

Dystonia may be limited to specific regions of the body, leading to a more specific dystonic syndrome, such as writer's cramp, blepharospasm, torticollis, or opisthotonus. In general, the location of dystonia is characterized as focal when it affects a single body part, segmental when it affects 1 or more contiguous body parts, multifocal when it affects 2 or more noncontiguous body parts, generalized when it affects 1 leg and the trunk plus 1 other body part or both legs plus 1 other body part, and hemidystonia when it affects only one half of the body. We encourage the use of the same regional classification for dystonic hypertonia.

Children with dystonia commonly have other features, including athetosis, poor dexterity, and abnormal patterns of muscle activation. Eye movement and oromotor abnormalities are frequently associated, but these features do not distinguish dystonic hypertonia from other causes of hypertonia. The anatomic localization of lesions that lead to dystonia has not yet been identified with certainty. It is likely that many forms of childhood dystonia are attributable to lesions in the basal ganglia.

Rigidity

Rigidity is a common movement disorder in adults, frequently diagnosed as a feature of parkinsonism, but only rarely reported in children. It is not known whether the apparent rarity of parkinsonian rigidity in children is attributable to underrecognition or to low incidence. Some practitioners use the term “rigid” to refer to any joint that cannot be moved. We encourage the use of a more specific definition in which the resistance to passive movement is independent of posture and speed of movement. To avoid erroneous inferences that the presence of rigidity equates with the presence of a full parkinsonian syndrome, we advocate use of the adjective “lead-pipe” rather than “parkinsonian” or “cogwheel” (which reflects coexistent tremor). Like spasticity and dystonia, rigidity may be dependent on the state of the child. Unlike dystonia, rigidity is not specific to particular tasks or postures. In adults with Parkinson’s disease, rigidity may result from baseline muscle contraction, hyperactive long-latency stretch reflexes, or both. At this time, there has been relatively little investigation of the features of rigidity in children, and therefore the following definition is based on experience with adults.

We define rigidity as hypertonia in which all of the following are true: 1) the resistance to externally imposed joint movement is present at very low speeds of movement, does not depend on imposed speed, and does not exhibit a speed or angle threshold; 2) simultaneous co-contraction of agonists and antagonists may occur, and this is reflected in an immediate resistance to a reversal of the direction of movement about a joint; 3) the limb does not tend to return toward a particular fixed posture or extreme joint angle; and 4) voluntary activity in distant muscle groups does not lead to involuntary movements about the rigid joints, although rigidity may worsen.

The presence of rigidity suggests the presence of hypertonia; thus, the terms rigidity and rigid hypertonia may be used interchangeably. The distinction from dystonic hypertonia is based on the lack of an associated abnormal posture or extreme position of the joint. This distinction may also be supported by the finding of a lack of muscle activity at rest. In rigidity, muscle activity is brought on by the externally imposed movement. In addition, hypertonia as a result of rigidity is usually not as sensitive to changes in posture. Rigidity may be worsened by movement of distant or contralateral muscles, an effect referred to as “activated rigidity.” Note again that rigidity as defined here is different from the finding of a stiff, immovable, or “rigid limb” that may be attributable to contractures, spasticity, dystonia, or rigidity as defined here. We encourage the use of the term rigidity only in the more specific sense defined in this article.

Rigidity may be associated with bradykinesia, tremor, flexed posture, and gait instability, leading to the syndrome of juvenile parkinsonism. Rigidity may be attributable to disorders of dopaminergic transmission or basal ganglia function, but the present definition does not include an implied localization.

Other Forms of Hypertonia

We do not address resistance to passive movement as a result of disorders of spinal cord, peripheral nerve, muscle, or connective tissue. Such disorders include startle syndromes, stiff person syndrome, α motoneuron dysfunction, myotonia, neuromyotonia, myokymia, and others. We also do not address other childhood movement disorders such as athetosis, chorea, ataxia, the hyperkinetic features of dystonia, myoclonus, tremor, and tic disorders.
RECOMMENDED TECHNIQUE FOR CLINICAL EVALUATION OF A HYPERTONIC JOINT

For evaluating a hypertonic joint, the clinician should elicit the parents’ description of abnormal tone and involuntary movements, including whether movements occur with action or at rest, and whether there are particular trigger movements or task specificity. Observe posture at rest and the position of the limbs with respect to gravity. Observe the child lying, sitting, walking, and running, if possible. If complaints include abnormal performance or postures in response to specific activities or tasks, then the child should be observed while performing the affected task. Any abnormal fixed, twisted, or repetitive posture should be noted, as well as the degree of functional limitation.

The following observations should be performed for each joint to be tested. Recognizing the contribution of anxiety to tone, the child should be relaxed as much as possible during the examination and the body part being examined should be supported against gravity. The head should be maintained in the midline to avoid contributions to tone from the tonic neck reflex. In addition, if lying supine, then the head and trunk should be resting comfortably.

1. Palpate the muscles to determine whether contraction occurs at rest.
2. Measure resistance to movement of the affected joint with the child supine, seated, and standing, if possible, as well as while distracted.
3. Measure passive range of motion at very slow (3 seconds to complete the movement), intermediate (0.5 second to complete the movement), and fast (as rapidly as possible) speeds. Note the resistance at the onset of movement, the presence or absence of a “catch” occurring at some time after the onset of movement, and the joint angle at which the catch occurs.
4. Perform sudden reversal in the direction of movement at slow, intermediate, and fast speeds, and note the presence or absence of increased resistance immediately on reversal (suggesting co-contraction) or at some time after (suggesting a spastic catch), as well as any velocity dependence.
5. Instruct the child to move the same joint on the contralateral side and observe for involuntary movement, then test for a change in resistance to slow, passive movement. Instruct the child to move a distant and unrelated joint (eg, by opening and closing 1 fist) on the contralateral side and then the ipsilateral side, and observe for involuntary movement or a change in resistance to passive movement.

MEASURES OF SEVERITY

Although most current scales of severity of impairment do not distinguish between diagnostic categories, they may still be useful once a category has been assigned. Such measures include the pendulum test,33 the modified Ashworth scale,34–36 and others.37 Such scales could be applied to hypertonia as a result of spasticity, dystonia, or rigidity but do not differentiate between them.38 The Tardieu scale explicitly compares the occurrence of a catch at low and high speeds and therefore is effective in measuring

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the velocity-dependent component of hypertonia. Other measures may be useful for rating severity of dystonia and rigidity, including the Barry-Albright Dystonia scale, the Burke-Fahn-Marsden dystonia rating scale, the Unified Dystonia Rating Scale, and the Unified Parkinson’s Disease Rating Scale. Quantitative kinematic and electromyogram analysis provides an additional set of methods for determining the severity of clinical findings, although these techniques have been more often applied to spasticity than to dystonia or rigidity.

MEASURES OF FUNCTIONAL ABILITY

It is essential to recognize that increased tone may have a variable relationship to functional limitation or disability. In many cases, increased tone permits greater functional ability in a child with underlying weakness, and this may be particularly true for mobility. Conversely, severe hypertonia can be associated with severe disabling joint contractures. Hypertonia may also be associated with pain, and it may be difficult to separate any restriction as a result of the primary handicap from possible restriction as a result of pain. Therefore, in addition to assessing the presence and severity of hypertonia, it is important to assess the child’s functional abilities and to recognize that these abilities may be affected by many components of function, including sensory processing, cognitive abilities, alertness, and others.

For childhood disorders, the recent development of quantifiable measures such as the Pediatric Evaluation of Disability Index, Peabody Developmental Motor Scale, Bayley Scales of Infant Development, Functional Independence Measure for Children, Gross Motor Function Measure, Test of Infant Motor Performance, Bayley Scales of Infant Development, and the Child Health Questionnaire has provided some ability to quantify the burden of care, level of function, and quality of life. However, such scales are not designed to distinguish between diagnostic categories, and very different disorders and symptoms may lead to similar scores of functional ability. It will be important in the future to establish whether different forms of hypertonia and tone abnormalities affect the person’s functional abilities and quality of life in different ways.

Adequate treatment should not be restricted to correction of specific clinical features but should instead be based on the functional limitations that are of direct concern to the patient. We therefore encourage clinicians and researchers to add rating scales to their clinical evaluations and to seek to determine the relationship of such measures to specific clinical parameters.

FUTURE GOALS

This article has provided a set of definitions for the purpose of identifying different aspects of childhood hypertonia. An important next step is to develop rating scales that are based on these definitions. Scales are needed both to quantify the degree of increased tone and to distinguish between the different types of increased tone. Such scales will most likely be modifications of 1 or more of the many existing scales intended for this purpose. It will be essential to determine the utility, validity, and reliability of such scales. An important component of validation of the scales is to determine the extent to which any measure of increased tone correlates with the degree of functional impairment. This is particularly important when treatment decisions will be made on the basis of the nature and degree of impairment. It will also be important to determine whether definition and distinction of the different types of hypertonia and their causes in fact lead to clinically significant changes in the management of patients.

There may be only a weak relationship between the physical signs obtained during the clinical examination and the impaired neuronal mechanisms in operation during an active movement. It is hoped that through recording and analysis of electrophysiological and biomechanical parameters during a functional movement such as locomotion, the significance of impaired reflex behavior or pathophysiology of muscle tone and its contribution to the movement disorder can reliably be assessed. Therefore, at the same time as clinical rating scales are developed, it is essential to validate quantitative and physiologic measures of function. Such measures can include kinematic and dynamic analysis, electrophysiology, and neuroimaging. These measures would need to be validated by comparison with clinical measures and ultimately determination of functional outcomes.

We therefore expect the following steps to be necessary for the continuing study of childhood motor disorders:

1. Development of impairment rating scales based on the definitions given here.
2. Development of definitions and rating scales for other childhood motor impairments, including athetosis, chorea, ataxia, the hyperkinetic features of dystonia, myoclonus, and tremor.
3. Assessment of the utility of the rating scales in terms of their ease of application in a clinical setting and the ability to detect changes in hypertonia over time.
4. Evaluation of the intrarater and intrarater reliability for diagnosis of hypertonia as well as for determining the relative severity of spasticity, dystonia, and rigidity.
5. Validation of the rating scales against current clinical judgment, electrophysiological measures, quantitative biomechanical measures, and neuroimaging modalities.
6. Validation of the rating scales against functional outcome measures to determine the extent to which hypertonia causes functional limitations and in which situations resolution of hypertonia leads to reduction of disability.
7. Determination of the ability of the rating scales to predict the response to therapy.
8. Determination of the ability of the rating scales to select appropriate patients for different therapeutic options.
9. Selection of standardized rating scales for childhood functional limitation, disability, and societal participation, with the choice of appropriate scale based on the child’s functional ability and degree of impairment.

There is increasing evidence that sensory systems may be abnormal in children and adults with movement disorders.59–64 In particular, abnormalities of proprioception and tactile sensation could potentially contribute to worsening symptoms and might be amenable to new treatment options. We therefore encourage research to determine the extent to which sensory involvement is present in childhood disorders and how it contributes to hypertonia and the success of intervention.

Because of the wide range of causes and symptoms in children with motor disorders, we expect that clinical treatment trials will frequently need to be performed between multiple clinical centers to obtain a sufficiently large and homogeneous population for testing. Organization of a clinical collaborative group thus is an important goal in this field.

This article is intended to provide an initial foundation for discussion of hypertonia in childhood by focusing on definitions of terms and syndromes. The ultimate goal is to provide a reliable method to characterize hypertonia and to establish effective treatment options for affected children.

APPENDIX

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