Classification Systems in Cerebral Palsy

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• Cerebral palsy • Classification • Impairment
• Activity limitation

The past decade has seen significant progress made in the evaluation of cerebral palsy (CP) and treatments for its sequelae. Because of advances in neonatal care and increased survival rates for preterm and low birth weight infants, efforts are being made to document the incidence and prevalence of CP through registries in Europe and Australia. Advances in orthopaedic care for children with CP have also been significant. Computerized gait analysis has led to refinements of orthopedic surgeries performed in these patients. Single event, multilevel surgery is now considered the standard of care in areas where gait analysis testing is available. New treatments have emerged, such as botulinum toxin injection and intrathecal baclofen, to treat spasticity and other types of hypertonia directly.

Because of increasing interest in conducting large-scale, multicenter investigations into the epidemiology of CP and its prevention and treatment, efforts have been made to establish a standard definition and classification systems for CP. In recent years there has also been increased focus on measurement of functional status of patients, and new classifications for gross and fine motor function have been developed.

The purpose of this article is to update the orthopaedic community on the current classification systems for patients with CP. This information will be of value to surgeons in determining patients' suitability for certain treatments and will also assist them in reviewing current literature in CP.

DEFINITION OF CEREBRAL PALSY

In 2007, the results of an International Workshop on Definition and Classification of CP were published.[1] The group included experts in the field of CP and developmental disorders from around the world. The purpose of the workshop was to update the existing definition and classification of CP to incorporate current knowledge about the disorder, and to improve communication among clinicians, researchers and epidemiologists. The following definition of CP was agreed upon:

> *Cerebral palsy describes a group of permanent disorders of the development of movement and posture, causing activity limitation, that are attributed to non-progressive disturbances that occurred in the developing fetal or infant brain. The motor disorders of cerebral palsy are often accompanied by disturbances of sensation, perception, cognition, communication and behavior, by epilepsy, and by secondary musculoskeletal problems.*[2]

This definition improves upon previous ones by emphasizing that CP involves a variety of disorders caused by various factors acting at different points in fetal development, and also highlights the importance of comorbidities that accompany the orthopaedic and neurologic manifestations. The definition excludes neurodevelopmental disabilities in which movement and posture are unaffected, as well as progressive disorders of the brain. The

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definition does not specify an upper age limit for onset of disorders, but inclusion of the phrase "fetal or infant" implies that it refers to insults occurring before full development, before specific milestones, such as walking, would have been achieved.

There remains some disagreement about this definition, but it is generally accepted and being used. Current issues with it include the lack of definition of an upper limit for age at onset in postnatally acquired cases, the need for definition of a lower limit for severity of involvement for a case to be classified as CP, and the need for a decision regarding whether to categorize syndromes, genetic disorders, or brain abnormalities resulting in static encephalopathy as CP.

INTERNATIONAL CLASSIFICATION OF FUNCTIONING, DISABILITY, AND HEALTH

In 2001, the World Health Organization published the International Classification of Functioning, Disability, and Health (ICF) for member states to use to standardize health and disability data worldwide.[3] The ICF is increasingly being incorporated into research in developmental disabilities. The ICF describes disability as dysfunction at 1 or more of 3 levels: *impairment* of body structures (organs or limbs) or functions (physiologic or psychological), limitations in *activities* (execution of tasks or actions by the individual), and restriction of *participation* (involvement in life situations). Researchers frequently design studies addressing these various domains of disability. Currently, classification schemes exist for CP at both the impairment and activity limitation levels, and these are the focus of this article. No classification systems exist to date for restriction of participation.

CLASSIFICATION OF IMPAIRMENTS

Motor Abnormalities

It has been estimated that about 80% of children with CP have some type of movement disorder.[4] CP is most often classified as either spastic, dyskinetic, or ataxic.[5] Although spasticity is often the dominant disorder, many children with CP have mixed spasticity and dystonia. When more than 1 type of movement disorder is present in patients, experts recommend classifying patients by the predominant disorder, for epidemiologic purposes,[1] with listing of secondary disorders as well.[6] Secondary movement disorders should be noted because this may impact treatment decisions. In particular, the results of soft-tissue surgeries are often less predictable in children with movement disorders.

The most current and comprehensive set of classifications for motor disorders has been published by the Task Force on Childhood Motor Disorders.[7,8] The group is an interdisciplinary panel of experts in the field of movement disorders and cerebral palsy, including pediatric neurologists and neurosurgeons, orthopaedic surgeons, pediatricians, physical and occupational therapists, and other specialists. Their aims included establishment of definitions and classifications of motor disorders, with an ultimate goal of allowing improved communication among clinicians and researchers, and improving classification of patients for clinical and research purposes. To date, definitions have been established for hypertonic and hyperkinetic movement disorders, as well as negative motor signs in children.

Hypertonia

Hypertonia is defined as "abnormally increased resistance to externally imposed movement about a joint."[8] Hypertonicity can be caused by spasticity, dystonia, or rigidity (though rigidity is rare in children and not associated with cerebral palsy).

Spasticity

Spasticity is hypertonia in which resistance to passive movement increases with increasing velocity of movement (or exhibits a spastic catch), and "varies with direction of the movement, and/or rises rapidly above a threshold speed or joint angle."[8] Spasticity is often a component of upper motor neuron syndrome, along with hyperreflexia, clonus, reflex overflow, positive Babinski sign, and pyramidal distribution weakness (upper extremity extensors, lower extremity flexors). Spasticity is caused by a hyperactive stretch reflex mechanism and is amendable to treatments, such as botulinum toxin, baclofen, selective dorsal rhizotomy, and orthopaedic surgery, for resultant contractures or balancing of muscle/tendon forces about the joints.

Dystonia

Dystonia is defined as "a movement disorder in which involuntary sustained or intermittent muscle contractions cause twisting and repetitive movements, abnormal postures, or both."[8] When dystonic postures are such that they are present at rest and do not relax upon attempts at passive movement, they cause hypertonia. Dystonia can also be classified as hyperkinetic (see the following). Dystonic hypertonia is present in cases where the resistance to passive movement does not change with changes in speed of passive movement or joint angle (is present at low and high speeds with no spastic catch), may be

associated with simultaneous agonist and antagonist contraction (equal resistance when the direction of passive movement is reversed), the limb tends to return to a fixed involuntary posture, and is triggered or worsened by voluntary movements at distant joints.[8] Dystonia is not associated with hyperreflexia and often disappears when the child is asleep. Because myelination is needed for development of dystonia, it typically occurs later in life than spasticity (around 5–10 years of age).[4]

It is postulated that a significant proportion of patients with cerebral palsy have a secondary component of dystonia, resulting in mixed hypertonia. Dystonia is associated with disruption of the basal ganglia and therefore is not improved by selective dorsal rhizotomy. In fact, what was previously considered recurrent spasticity after rhizotomy is now thought to be unrecognized dystonia.[4] It is generally accepted that tendon lengthening and transfer procedures are contraindicated in cases of dystonia, because of the risk for recurrence of deformity or development of reverse deformities. Although this is the conventional wisdom, evidence in the literature is limited. Occasionally, surgery may be required despite optimal medical management of the dystonia. This outcome is most commonly seen with deformities of the foot and ankle, particularly varus deformities, which may make shoe wear and bracing problematic. In such cases, surgery with split tendon transfers may be considered to address the varus foot. Whole tendon transfers should be avoided in children with dystonia. Dystonic hypertonia is responsive to botulinum toxin as well as intrathecal baclofen, which generally weaken overactive muscles or muscle groups. For patients in whom the primary movement disorder is hypertonic dystonia (vs spasticity), evaluation by a specialist in movement disorders is recommended before considering orthopaedic surgery for tendon lengthening or transfers. However, bony deformities in these patients, such as femoral anteversion, tibial torsion or bony foot deformities, are appropriate and beneficial when indicated.

Hyperkinetic Movements

Hyperkinetic movements are defined as “any unwanted excess movement”[9] that is performed voluntarily or involuntarily by the patient, and represent what have traditionally been referred to as extrapyramidal symptoms. The hyperkinetic movements most commonly seen in CP include dystonia, chorea, athetosis, and tremors.

Hyperkinetic dystonia is characterized by “abnormal postures that are superimposed upon or substitute for voluntary movements.”[9] These are repeated postures that are unique to each patient, although some common patterns exist, such as foot inversion and wrist ulnar deviation. They can be of varying durations, and can be triggered by volitional movement. Chorea is defined as “an ongoing random-appearing sequence of one or more discrete involuntary movements or movement fragments.”[9] It is similar to hyperkinetic dystonia except that it involves brief extraneous movements rather than postures, which imply maintenance for a length of time. Choreiform movements are also random, can appear continuous and jerky, and can be difficult for patients to relax. Athetosis is defined as “a slow, continuous, involuntary writhing movement that prevents maintenance of a stable posture,”[9] where discrete, repetitive movements or postures cannot be identified. It usually involves the hands or feet, and perioral muscles. Athetosis is not common as an isolated movement disorder in CP and is most often found in combination with chorea. The taskforce recommends that the term *dyskinetic CP* is used instead of *athetotic CP*, because athetosis is rare as an isolated finding, and when present is not often the primary movement disorder.[9]

Orthopaedic surgery in cases of predominantly hyperkinetic movement disorders associated with cerebral palsy is most often limited to bony procedures, because fixed contractures are rare because of the often nearly continuous movements of the extremities and joints. Such patients should be referred to a movement disorders specialist for management, because this type of movement disorder may be best managed through medications.

As noted, bony surgery is more commonly performed in such patients than is tendon surgery. Tendon lengthening in patients with hyperkinetic movement disorders are unreliable and may result in a reverse deformity compared with that seen preoperatively (eg, a posterior tibial tendon lengthening may result in a previously varus foot being positioned in valgus postoperatively). If tendon transfer surgery is contemplated, it should be remembered that split tendon transfers are more successful in patients with dystonia than are whole tendon transfers. Isolated osseous surgery has more reliable results in these patients, but casting should be minimized when possible because these patients often do not tolerate casts well and their dystonia may be exacerbated following cast removal.

Hypertonia in CP is most often rated using the Modified Ashworth Scale (MAS).[10] The Tardieu scale[11] is preferred by some clinicians and researchers, because it assesses resistance to

both fast and slow stretches, the angle at which resistance is felt initially (R1), as well as the end of passively available range of motion (R2). Neither test is able to distinguish spasticity from hypertonic dystonia (or contracture, in the case of the MAS). There are no pure measures of spasticity available. Dystonia in children with CP (hypertonic and hyperkinetic) is assessed using the Barry-Albright Dystonia scale,[12] which was adapted for use in CP from the Fahn-Marsden Movement scale used in adults with primary dystonia. A new scale is being developed to quantify both spasticity and dystonia in the same patient, because this is a frequent occurrence, and determine the primary disorder (Hypertonia Assessment Tool-Discriminant).[13]

Negative Signs

Hypertonicity and dyskinetic movements constitute positive motor signs of increased activity. Negative signs include characteristics that are decreased or insufficient and include weakness (insufficient muscle activation), poor selective motor control (inability to activate a specific pattern of muscles in an isolated fashion), ataxia (inability to activate the correct pattern of muscles during a movement), and apraxia/developmental dyspraxia (inability to activate the correct pattern of muscles to perform a specific task, either because of loss of ability or lack of acquisition of the skill).[7] These problems often coexist with positive signs and can be more disabling than positive signs in some patients. Negative signs should be recognized because their presence may contribute to poor surgical outcomes. These problems are best addressed through physical and occupational therapy.

Topography or Limb Distribution

The traditional classifications of limb distribution for the hypertonic (primarily spastic) form of CP, hemiplegia, diplegia, and quadriplegia/tetraplegia (and occasionally triplegia), continue to be used clinically. However, these classifications have shown poor inter-rater reliability and have been the source of discrepancies in proportions of CP subtypes reported by registries in different countries.[14] Inconsistencies arise because of lack of definition of how much upper extremity impairment is needed to classify patients as quadriplegic versus diplegic. In addition, children with hemiplegia often have some motor signs on the contralateral side, which could put them in a category of asymmetric diplegia, quadriplegia, or triplegia. Some experts recommend abandonment of these labels[2,6] and advocate simplified classifications, such as *unilateral* or *bilateral*, with an indication of upper and lower extremity function (such as Gross Motor Function Classification System level, or Manual Ability Classification System level) as is done in the Surveillance of Cerebral Palsy in Europe registry.[14] This change in classification is controversial, however, because there are suggestions in the literature of etiologic, radiological, and functional distinctions between diplegia and quadriplegia.[15–20] If the traditional terms (diplegia, quadriplegia, hemiplegia) are used, complete description of the motor impairments in all body regions (including the trunk and oropharynx) is recommended.[2] The term *paraplegia* is no longer used with respect to CP, because all children with diplegia have some level of impairment of fine motor upper extremity skills. If no upper extremity involvement is seen in a child with spasticity in the lower extremities there should be a suspicion of hereditary (familial) spastic paraparesis, tethered cord, or spinal cord tumor.[5,21] Some experts suggest a limb-by-limb description of motor impairment and tonal abnormalities seen in each limb, such as that used in the Australian CP register.[1] Their thought is that a description of the clinical presentation yields more valid and reliable information than placement of patients into categories, such as diplegia and quadriplegia.

CLASSIFICATION OF ACTIVITY LIMITATION

Gross Motor Function Classification System

In the past, patients' gross motor functional limitations were categorized as mild, moderate, and severe. Alternatively, some were characterized using the descriptors published by Hoffer and colleagues[22] for myelomeningocele (ie, household and community walkers). Although these descriptions conveyed information regarding the patients' ambulatory function, they were not standardized or validated. In 1997 a hallmark paper was published by Palisano and colleagues[23] that provided a new classification system for gross motor function in children with CP, the Gross Motor Function Classification System (GMFCS). This system rated patients' ambulatory function, including use of mobility aids and performance in sitting, standing, and walking activities. The original GMFCS had some limitations. These limitations included an upper age limit of 12 years (before adolescence) and the necessity of using a single rating to describe a child's ambulatory performance across different terrains and distances, resulting in a tendency of parents and therapists to rate a child based on their best capability rather than their typical performance when forced by the rating scale to choose a single category.[24] These issues

were considered and addressed in an updated version of the scale.[25] The GMFCS-Expanded and Revised includes children up to 18 years of age. The descriptions of gross motor function were also revised to incorporate aspects of the framework of the ICH and recognizing that a child's environment and other factors may affect gross motor performance.

The GMFCS-ER provides a method for communicating about gross motor function, based on the use of mobility aids and performance in sitting, standing, and walking activities. It is intended to classify a patient's level of gross motor function based on his or her typical performance, rather than their best capability. It classifies gross motor function on a 5-point ordinal scale, with descriptions of skills provided for 5 age groups: less than 2 years of age, 2 to 4 years of age, 4 to 6 years of age, 6 to 12 years of age, and finally 12 to 18 years of age. In general, the levels are as follows (**Fig. 1**):

Level I: Walks without limitations
Level II: Walks with limitations
Level III: Walks using a hand-held mobility device
Level IV: Self-mobility with limitations; may use powered mobility
Level V: Transported in a manual wheelchair.

The validity and reliability of the GMFCS-ER have been demonstrated repeatedly in multiple studies.[26–29] Gross motor reference curves have been developed using GMFCS level data to allow clinicians to compare patients' status to that of children at the same age and GMFCS level, as well as to enable them to give patients and families a prognosis for gross motor progress over time.[30,31] The GMFCS has also been used to study and document the age at which peak gross motor function is achieved for each level (approximately 5 years of age for GMFCS levels I and II, 8 years of age for level III, and 7 years of age for levels IV and V), and to document the stability or decline in gross motor skills through adolescence (no decline for levels I and II, approximately 5%, 8% and 6 % decline in GMFM scores by 21 years of age for levels III, IV, and V, respectively).[30] Further, the GMFCS has been useful in categorizing patients for orthopaedic prognostic and experimental studies, both short and long term. Hip surveillance data in children with CP have shown that the incidence of hip dislocation increases linearly with GMFCS classification from level I (0% incidence) to level V (>90% incidence).[32] Patients at GMFCS levels IV and V have been shown to have greater acetabular dysplasia and hip subluxation than those at levels II and III.[33] Satisfaction with the functional and cosmetic outcome of multilevel orthopaedic surgery has been shown to be higher among parents of patients classified at GMFCS level I than those whose children function at levels II and III.[34] Stability of GMFCS classification in patients over time has also been documented. McCormick and colleagues[27] demonstrated that the GMFCS level observed around 12 years of age is highly predictive of adult gross motor function. Children who are independent walkers at 12 years of age (GMFCS levels I and II) have an 88% chance of having a similar functional status as an adult and children who use a wheelchair as their primary mode of mobility have a 96% chance that they will continue to use the wheelchair into adulthood. Single-event multilevel surgery (SEMLS) can affect the stability of the GMFCS over time as patients at all levels, especially levels II, III and IV, have been shown to experience an improvement in GMFCS level after such surgery.[35] Although children functioning at GMFCS level IV can show improvement in ambulatory function after SEMLS, they have been found not to benefit from the addition of distal rectus femoris transfer to multilevel surgery.[36] All of this information is beneficial to orthopaedic surgeons, patients, family members, and caregivers in preparing, administering, and planning for long-term care.

Functional Mobility Scale

The Functional Mobility Scale (FMS) was designed by Graham and colleagues[24] as a measure of ambulatory performance in children with CP. It has been shown to be a better discriminator of differences in ambulatory function among children with CP than the Rancho Scale.[22] The FMS is the only existing functional scale that accounts for the fact that children may demonstrate different ambulatory abilities and use different assistive devices to walk various distances. Intended as an outcome measure, the FMS is also useful as a means of classifying ambulatory ability.

The FMS is administered via parent/patient interview and categorizes the assistance needed (none, canes, crutches, walker, wheelchair) for a child to walk 3 distances (5, 50, and 500 yards, or 5, 50, and 500 m). The distances are not specifically measured, but are used as estimates to represent household, school, and community ambulation. Ratings are given for each distance category: 1, uses wheelchair; 2, uses walker or frame; 3, uses crutches; 4, uses sticks (canes); 5, independent on level surfaces; 6, independent on all surfaces. A rating of *C* is given if the child crawls the designated distance, and an *N* is given if the

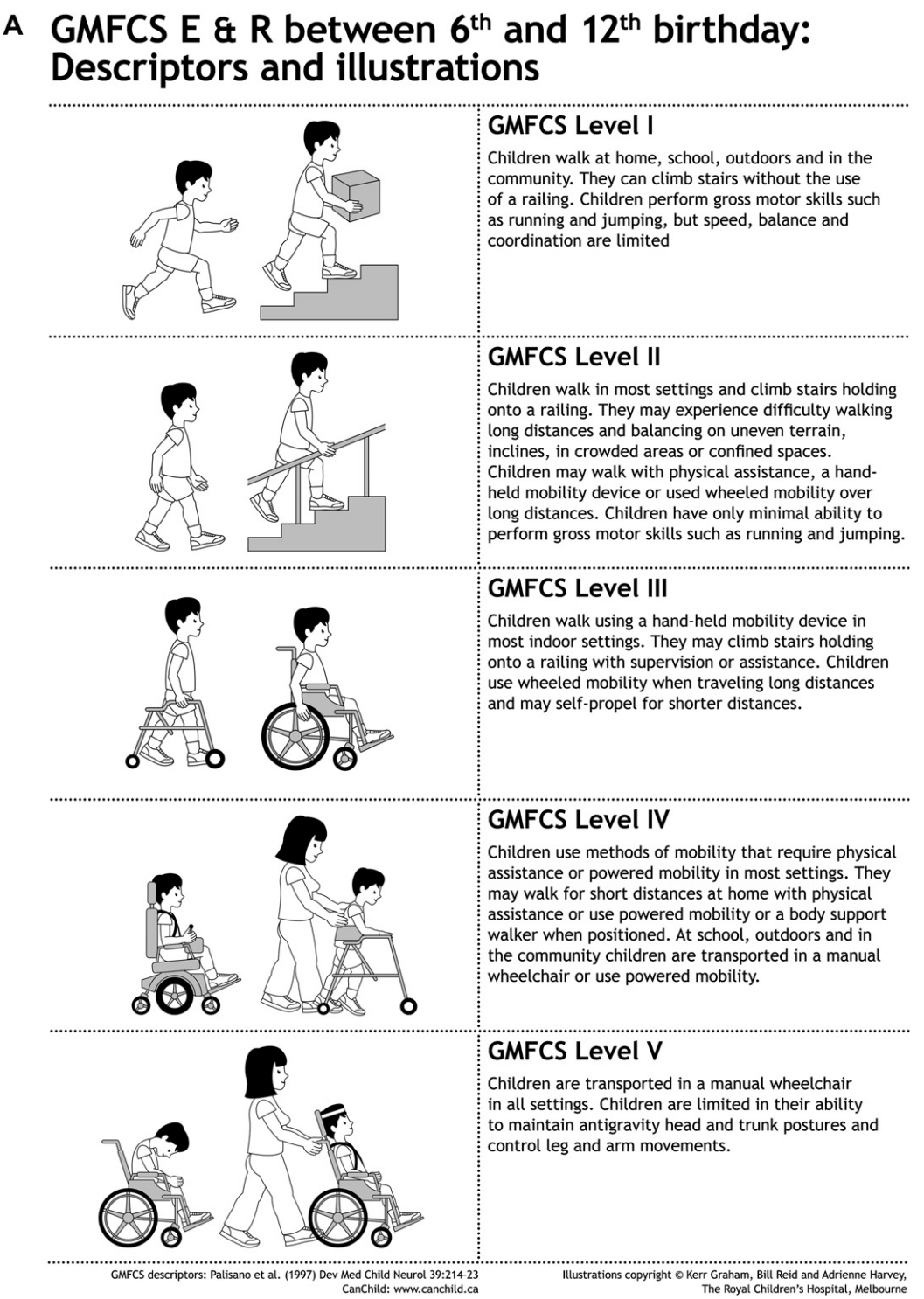
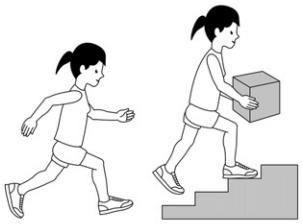


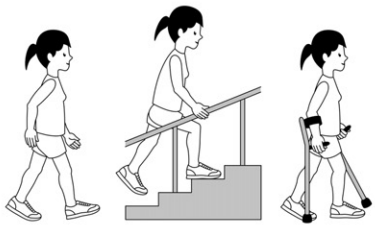
Fig. 1. (*A*) GMFCS expanded and revised, for children aged 6 to12 years. (*B*) GMFCS expanded and revised, for children aged 12 to 18 years. (*Courtesy of* Kerr Graham, MD, The Royal Children's Hospital, Melbourne, Australia.)

B GMFCS E & R between 12th and 18th birthday: Descriptors and illustrations



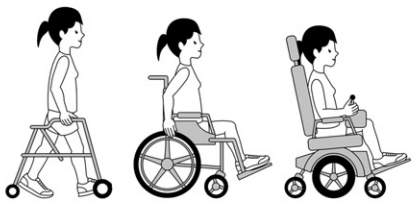
GMFCS Level I

Youth walk at home, school, outdoors and in the community. Youth are able to climb curbs and stairs without physical assistance or a railing. They perform gross motor skills such as running and jumping but speed, balance and coordination are limited.



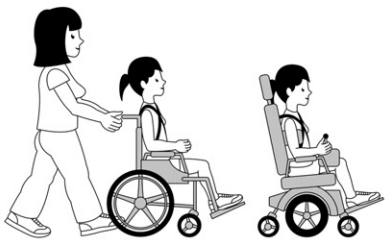
GMFCS Level II

Youth walk in most settings but environmental factors and personal choice influence mobility choices. At school or work they may require a hand held mobility device for safety and climb stairs holding onto a railing. Outdoors and in the community youth may use wheeled mobility when traveling long distances.



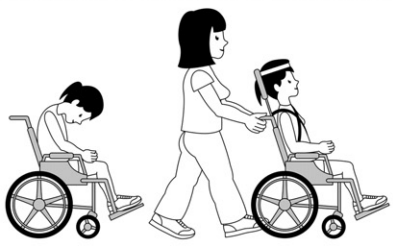
GMFCS Level III

Youth are capable of walking using a hand-held mobility device. Youth may climb stairs holding onto a railing with supervision or assistance. At school they may self-propel a manual wheelchair or use powered mobility. Outdoors and in the community youth are transported in a wheelchair or use powered mobility.



GMFCS Level IV

Youth use wheeled mobility in most settings. Physical assistance of 1–2 people is required for transfers. Indoors, youth may walk short distances with physical assistance, use wheeled mobility or a body support walker when positioned. They may operate a powered chair, otherwise are transported in a manual wheelchair.



GMFCS Level V

Youth are transported in a manual wheelchair in all settings. Youth are limited in their ability to maintain antigravity head and trunk postures and control leg and arm movements. Self-mobility is severely limited, even with the use of assistive technology.

GMFCS descriptors: Palisano et al. (1997) Dev Med Child Neurol 39:214-23
CanChild: www.canchild.ca

Fig. 1. (*continued*)

child is unable to move through a given distance. A child who ambulates independently for all distances and on all types of surfaces would be given a rating of 6, 6, and 6. A child who ambulates independently on level surfaces in the home, uses crutches at school, and a wheelchair for shopping trips and family outings would be given a rating of 5, 3, and 1 (**Fig. 2**).

Like the GMFCS, the FMS assesses a child's average performance in daily life rather than their maximum capability. The FMS has been demonstrated to have good construct and concurrent

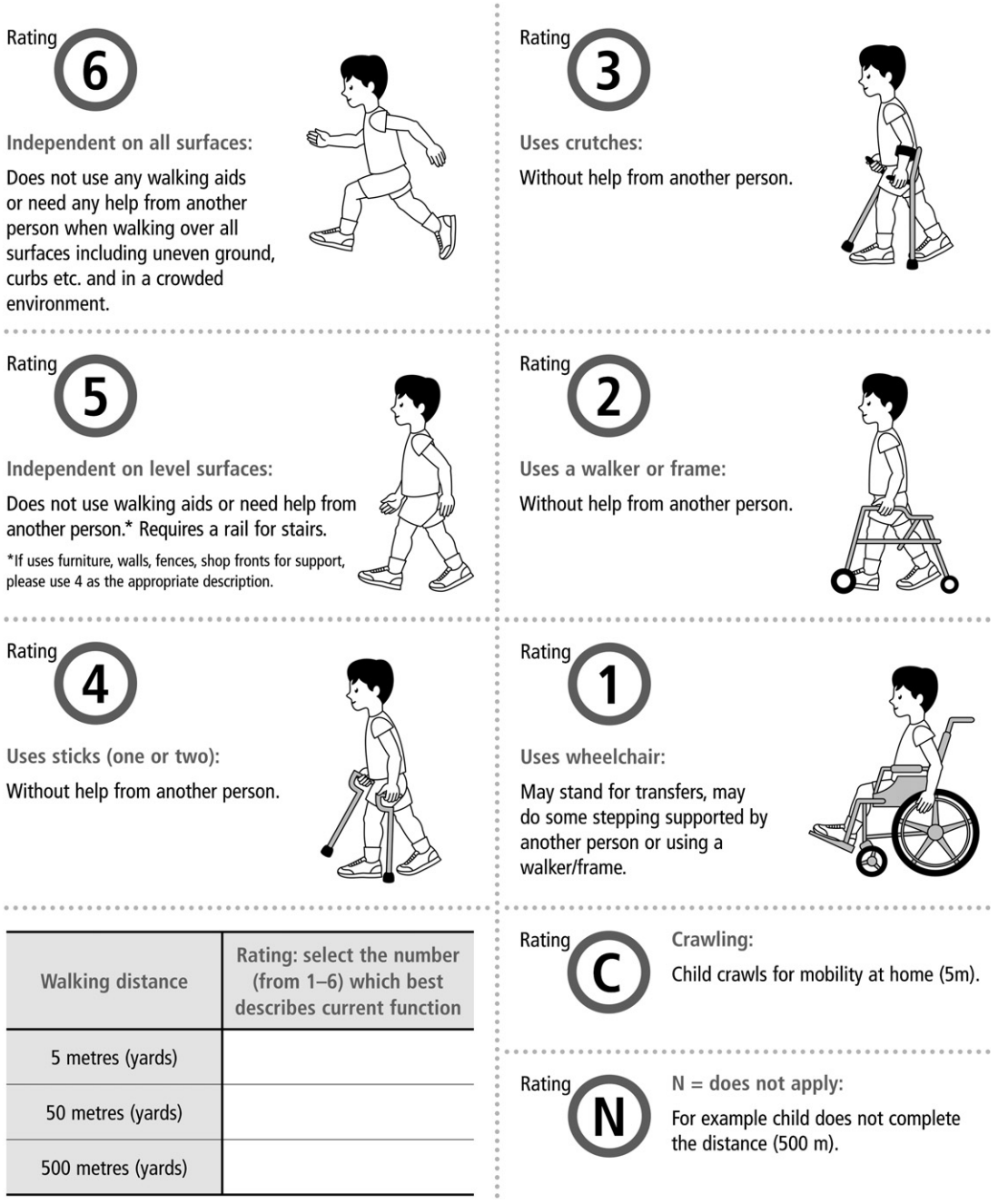


Fig. 2. Functional Mobility Scale. (*Courtesy of* Kerr Graham, MD, The Royal Children's Hospital, Melbourne, Australia.)

validity, good inter-rater reliability (substantial agreement among therapists and orthopaedic surgeons), as well as showing sensitivity to change after surgery (positive and negative change).[24,37,38]

The FMS specifically addresses ambulation and, therefore, is not intended to substitute for the GMFCS, which assesses mobility on a more general level. The FMS should be used as a companion rating scale to the GMFCS.

Manual Ability Classification System

In 2006 a classification system similar to the GMFCS was developed for the upper extremity called The Manual Ability Classification System (MACS).[39] The MACS was designed to describe upper-extremity performance in activities of daily living for children with CP. As with the GMFCS-ER, the MACS takes into account the fact that upper-limb function is influenced by personal, environmental, and contextual factors. The MACS is not designed to describe best capacity or the function of individual upper extremities, such as comparing involved to uninvolved sides. It reports on performance of upper-limb tasks in daily living, regardless of how they are accomplished, and the collaboration of both hands together (bimanual tasks).

The MACS is also designed as a 5 category scale and the levels include (Full descriptions and distinctions between levels is available at: www.macs.nu):

- Level I: Handles objects easily and successfully
- Level II: Handles most objects but with somewhat reduced quality or speed of achievement
- Level III: Handles objects with difficulty; needs help to prepare or modify activities
- Level IV: Handles a limited selection of easily managed objects in adapted situations
- Level V: Does not handle objects and has severely limited ability to perform even simple actions.

The MACS is intended to apply to children of all ages and, therefore, does not include age bands. Raters are instructed to consider the child's performance doing age-appropriate tasks and using age-appropriate objects. The MACS has been demonstrated to be both reliable and valid.[39–41] Eliasson and colleagues demonstrated excellent reliability of MACS among children aged 4 to 18 years. Morris and colleagues found good reliability among children aged 6 to 12 years. Finally, Plasschaert and colleagues looked at reliability in children aged 1 to 5 years. Results showed moderate reliability among this age group with overall less reliability in children aged less than 2 years. Imms and colleagues[42] looked at stability of caregiver-reported MACS and GMFCS-ER. Levels were found to be generally stable over 12 months (67% for MACS and 79% for GMFCS). It is important to note that there are other hand classifications in the literature, but these all focus on specific aspects of grasping and not overall functional performance.[43–46]

Carnahan and colleagues[47] and Gunel and colleagues[48] looked at how closely associated the GMFCS-ER and MACS are when classifying children. Overall results showed that the 2 systems often show some discrepancies in children with CP. However, when looking at the 2 systems in relation to CP subtypes some associations were found. In diplegic CP, children were found to have a lower level (higher functioning) MACS score and higher level (lower functioning) GMFCS-ER. The opposite was found for hemiplegic CP; in these children manual ability was generally found to be more limited than ambulatory function. In general, the study found that there were differences in gross motor function and manual ability when looking at different CP subtypes. It is, therefore, essential to combine information on CP subtype with both the GMFCS and MACS systems when evaluating these children.[47,48]

It should be noted that neither the GMFCS-ER nor the MACS are intended to identify the cause of activity limitation (neurologic or musculoskeletal impairments, cognitive or attention deficits), but are simply intended to categorize a child's ability to function in daily life.

The development of both the GMFCS-ER and the MACS has revolutionized the way we describe the gross motor and manual abilities in CP patients. However, there is limited information in the literature on the communication skills in this patient population. In response to this weakness The Communication Function Classification System (CFCS) is currently under development by an international development and research team led by Michigan State University.[49] The CFCS is also a 5-level classification system that is being modeled after the GMFCS and MACS. The goal of this classification system is to be a quick and simple instrument easily used by a person familiar with patients. This system will help parents and clinicians understand how different communication environments, partners, and communication tasks affect the CFCS level, and will also assist in individual goal setting for communication.[49]

Although still evolving, the standardization of definitions and classifications has been an essential step forward for the CP community. Armed with these tools we can communicate more clearly about our patients and evaluate interventions more effectively. This progress has made conducting large-scale multicenter investigations more meaningful. As a result, we will continue to advance our collective understanding of the condition, further advance the care and improve the quality of life for our patients with CP.

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