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Clinical Classification of Cerebral Palsy

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Abstract

The classification of cerebral palsy (CP) remains a challenge; hence the presence of so many classifications and a lack of consensus. Each classification used alone is incomplete. Therefore, a multiaxial classification gives a more comprehensive description of a child with CP. The recent WHO International Classification of Functioning, Disability and Health (ICF) emphasizes the importance of focusing on the functional consequences of various states of health and has stimulated the development of newer functional scales in CP. It is widely accepted that the functional classification is the best classification for the patient because it guides management. The objectives of this chapter are to review the various classifications of CP, to highlight the clinical features used in the various classifications, to outline the recent functional classifications of CP and to highlight how these recent classifications guide current management. It is expected that at the end of this chapter, the reader should be able to understand the difficulties in classifying CP, enumerate and discuss the various classifications of CP, understand the merits and shortcomings of each classification scheme, clinically evaluate and classify a child with CP multiaxially and understand how functional scales predict current and future needs of children with CP.

Keywords: clinical classification, cerebral palsy, functional scales, management, spastic, extrapyramidal, SCPE, GMFCS, MACS, CFCS, EDACS, multiaxial

1. Introduction

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The categorization of children with cerebral palsy (CP) into clinical groups remains a challenge, hence the presence of so many classifications that are not comprehensive and the continued search for a holistic classification [1]. The clinical manifestations of CP are heterogeneous as rightly pointed out in the most current definition of CP [1, 2]. This implies that children with CP differ clinically in many aspects. Therefore, different groupings (classifications) are possible [1].

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These classifications (groups) differ in the characteristic(s) used and their individual uses or purposes. A classification may be used for describing the nature of the disability, for predicting current and future management needs, comparing cases in different areas and assessing change following an intervention [1]. Generally, it is desirable that any classification used should be reliable, valid, quantitative, and objective and most importantly assist management [1].

Besides early identification and intervention, the current trend in neurodevelopmental pediatrics is a focus on functional effects of different states of health [3, 4]. This is the outcome of the recent WHO International Classification of Functioning, Disability and Health (ICF) which in the field of CP led to the development of newer measures of functional abilities (functional scales) [3, 4]. There are functional scales for a number of functions impaired in CP. It is widely accepted that the functional classification remains the best classification for a patient with CP because it guides management [1, 5, 6].

2. Overview of clinical classification of CP

Some factors that influence the clinical classification of CP are the age of a child, reliability of the medical history, and extent of diagnostic investigations [1]. This means that the same child may be classified differently at different times (due to changes in peripheral manifestations with age), by different people (due to variable historical data from maternal recall or case notes), and in different regions (due to differences in availability and affordability of neuroimaging and metabolic studies). Therefore, Bax et al. [1] in 2005 proposed that all classification results should indicate these factors at the time of classification.

Children with CP differ clinically in the following characteristics: type/nature of motor disorder, distribution of motor impairment, etiology, presence/number of accompanying impairments, structural brain abnormalities on neuroimaging, degree of severity of impairments, and individual therapeutic needs. These clinical variables form the basis of the traditional classifications of CP. In 1956, Minear [7] and the Nomenclature and Classification Committee of the American Academy for cerebral palsy classification put forward an early classification system that presented seven classification axes based on the aforementioned features.

Subsequent classification systems originated from the Minear classifications and are either a combination or an expansion of the categories. Such classification systems based on multiple variables include the Swedish classification system [8], Edinburgh classification [9] and classification by the Surveillance for Cerebral Palsy in Europe (SCPE) [10].

The current emphasis on the functional consequences of different health states increased interest and research on the functional classification of CP [1, 3, 4]. The result is an evolution of newer measures (functional scales) that objectively and reliably measure and quantify functional abilities. A number of these functional scales have been validated by multiple studies [11–16]. They include Gross Motor Function Classification System (GMFCS) [11] (functional mobility/ambulatory function), Manual Ability Classification System (MACS) [14] (hand and

arm function/manual dexterity), Communication Function Classification System (CFCS) [16] (speech/communication function) and Eating and Drinking Ability Classification System (EDAC) [17] (eating and drinking/oropharyngeal function). They are mainly used for predicting current and future management needs of children with CP, and their use agrees with current thinking in management of CP.

Advances in management of CP including the biopsychosocial method of service delivery that recommends liberal use of assistive devices require additional characteristics or variables to be added to traditional classifications in order to assist management and satisfy other important purposes like clinical description and research [1, 4, 5]. Such a classification would be called holistic, comprehensive or standardized. A consensus on what characteristics/components such holistic classification should incorporate is yet to be reached by experts in the field of CP.

3. Traditional classifications of CP based on single characteristics

The traditional classifications of CP are basically the Minear [7] classifications in seven axes namely:

- **1.** Physiological
- 2. Topographic
- 3. Supplemental
- 4. Aetiologic
- 5. Neuroanatomic (radiologic)
- 6. Therapeutic
- 7. Functional

3.1. Physiologic classification

This is based on the type/nature of motor or movement disorder (quality and changes in tone) and classifies CP into two types: spastic (pyramidal) and non-spastic (extrapyramidal). Generally speaking, neuromotor findings in spastic CP are consistent and persistent while variability is the rule in extrapyramidal CP [6, 18, 19].

The clinical features of spastic CP are as follows [6, 18, 19]:

• Tone is invariably increased (hypertonia), that is, persistently increased with little or no variation in the awake (movement, tension and emotion) or sleep states. This is further confirmed by asking caregivers whether their child feels stiff when touched or held most times of the day even during sleep. The answer is usually a "yes."

- The quality of the increased tone is described as "clasp-knife" spasticity and is elicited clinically by a rapid passive movement at a joint (as rapidly as the time taken to say "one thousand and one"). This produces the classic "clasp-knife" resistance followed by a sudden "give." Spasticity refers to hypertonia due to a velocity-dependent increase in tonic spinal stretch reflex.
- Deep tendon reflexes are markedly increased (more commonly grade 3+ or 4+)
- A positive Babinski sign (extensor planter response), that is, lightly stroking the lateral aspect of the sole and across the foot pads/ball of the foot, results in extension/dorsiflexion of the hallux (up-going big toe) and fanning out/spreading of the other toes.
- Sustained ankle clonus, that is, when the ankle is briskly dorsiflexed on a flexed knee, a rhythmic contraction is observed.
- Non-positional contractures (due to persistent hypertonia)
- Decreased movement
- Localization/limb distribution of neuromotor impairment varies from one child to another and so spastic CP can be further classified topographically.

In contrast, the clinical features of extrapyramidal (non-spastic) CP are [6, 18, 19]:

- Tone is variably increased (varies from hypertonia to hypotonia) depending on the state, that is, tone is increased by activity, agitation, tension, and emotions like crying, but tone is decreased in sleep and when relaxed. Caregiver usually tells the clinician that their child limbs feel normal when asleep or quiet.
- The quality of the increased tone is "lead pipe" rigidity or "candle wax" type and is elicited clinically by a slow passive flexion and extension of a limb. The increased resistance to this passive movement is felt all through the movement. Besides, extrapyramidal hypertonus can be diminished by repetitive movement and this is called "shaking it out."
- Deep tendon reflexes are usually normal or mildly increased (grade 1+ to 3+).
- A negative Babinski sign.
- Unsustained ankle clonus.
- Positional contractures (the variable tone is protective against contractures and so contractures like hip/knee flexion contracture only occur after prolonged periods on a wheelchair).
- Movement is disordered. Thus, extrapyramidal CP is also called dyskinetic CP.
- There is a four-limb functional impairment that precludes further topographic classification. However, extrapyramidal or dyskinetic CP is further subdivided based on the different manifestations of abnormal/involuntary movements (dyskinesia) and tone. The subtypes are choreathetoid CP—characterized by excessive and rapid movements involving the proximal body parts (trunk) (chorea) combined with slow writhing movements of

the distal parts of the body (extremities) (athetosis) and usually with reduced tone. This is the commonest type of extrapyramidal CP. Dystonic CP is characterized by extrapyramidal hypertonia and decreased movement (hypokinesia). Dystonia occurs when there is simultaneous contraction of both agonist and antagonist muscles. Ataxic CP occurs when there are signs of incoordination and hypotonia caused by damage to the cerebellum. This is a rare form of CP [6, 18, 19].

One merit of the physiological classification is that it can suggest the areas of brain damage and possible etiological factors. For instance, spasticity would suggest damage to the cortical neurons (pyramidal cells) due to hypoxic ischemic encephalopathy (HIE) from severe perinatal asphyxia and postnatal central nervous system (CNS) infections like meningitis [19]. In addition, dyskinetic CP points to damage to the basal nuclei by bilirubin encephalopathy and severe perinatal asphyxia at term [19]. Therefore, the physiological classification is still clinically useful.

However, the physiological classification is not reliable [6, 18, 20]. The terms spastic (pyramidal) and extrapyramidal CP are strictly incorrect [6, 18, 20]. It is more accurate to refer to these as "predominantly spastic" and "predominantly non-spastic." Due to the complex interactions of the upper motor neuron system (the pyramidal, extrapyramidal, and cerebellar pathways) with the anterior horn cells to control posture and movement, lesions causing CP in real life usually involve both pyramidal and extrapyramidal pathways [21]. Strictly speaking, pyramidal lesions induce spasticity as a result of concomitant damage to extrapyramidal pathways [21]. This explains the clinical combination of motor/movement abnormalities, for example, spasticity and dystonia, and spasticity and choreoathetosis. This is the so-called mixed CP subtype. From the explanation above, this CP subtype should actually be very common but from published studies [22, 23], spastic CP remains the commonest type thereby exposing the subjectivity and imprecision in assessment of patients based on this classification. Additionally, the physiological classification does not aid therapy or inform management of patients with CP, and this inability to indicate functional abilities remains a major drawback [6, 18, 20].

3.2. Topographic classification

This classification relies on the localization/limb distribution of neuromotor impairment in spastic CP [19]. It subdivides spastic CP into: quadriplegia (symmetric/equal and severe spasticity of all four limbs), diplegia (involvement of the four limbs but greater spasticity and weakness in the lower limbs) and hemiplegia (involvement of the upper and lower limbs on one side of the body) [19]. Other types of spastic CP such as tripegia (three-limb spasticity) and monoplegia (one-limb spasticity) are rare, and double hemiplegia (four extremity involvement with greater spasticity of the upper limbs) is no longer in use [6, 20].

An advantage of this classification is that these topographical subtypes can be linked to some etiological factors. For instance, diplegia suggests periventricular leukomalacia due to prematurity/low birth weight; hemiplegia suggests perinatal stroke, periventricular hemorrhagic infarction or neonatal cortical infarction while quadriplegia suggests severe perinatal

asphyxia at term, postnatal infection (bacterial meningitis) and metabolic/genetic disorders [19]. However, the descriptive terms in the topographic classification cannot be used reliably [6, 20]. One notable source of confusion is distinguishing spastic diplegia from quadriplegia. This distinction is highly subjective since it is unclear how much upper limb spasticity is needed to separate a diplegia from a quadriplegia [6, 20]. Recall that there is involvement of the four limbs in both subtypes. The arm and leg naturally perform different functions, and assessing the relative severity of involvement is difficult [1]. Moreover, the imprecise use of these terms in clinical practice has been reported by Gorter et al. [24] Many experts agree that the use of these terms in classification should be stopped [1]. Furthermore, the topographic classification does not consider functional abilities and so does not aid therapy or inform management of these children [6, 20]. Therefore, the topographic and physiological classifications share similar merits and demerits.

3.3. Supplemental classification

This is an additional grouping that comprises the accompanying impairments in CP and their association with the physiological and topographic classifications [6, 20]. The accompanying physical, mental or physiological impairments in CP include epilepsy, cognitive (intellectual), speech, visual and hearing impairments, behavioral problems and secondary musculoskeletal abnormalities (hip dislocation/subluxation, contractures) [1, 2]. The purpose of linking these supplemental disorders to the physiological and topographic classifications was to identify syndromes with a common etiology that would lead to prevention [6, 20]. Unfortunately, the supplemental disorders correlated poorly with the two earlier classifications. This means that it was only in a few examples like bilirubin encephalopathy that such a link between supplemental disorders, physiology and etiological factor could be established. For instance, the combination of accompanying impairments – vertical gaze palsy, sensorineural deafness and enamel dysplasia—is associated with choreoathetoid CP (physiology) from damage to the basal nuclei by bilirubin encephalopathy (etiological factor) [6, 20].

Though these associations were limited and the aim of the supplemental classification defeated, supplemental disorders (accompanying impairments) remain pertinent to the current management of CP because their presence strengthens the need for multidisciplinary management. This means that the accompanying impairments need to be taken into consideration in planning service delivery. Moreover, the accompanying impairments may cause more functional limitation than the primary motor dysfunction (the core feature of CP) and thus must be addressed to achieve a positive functional outcome. Furthermore, the most recent definition of CP [2] highlights the importance of these accompanying impairments by incorporating them as part of the definition of CP since CP rarely occurs without them. It is generally recommended that the presence or absence of these impairments and the extent to which they interfere with function be recorded in addition to the classifications used [1]. Currently, it is recommended that at least the presence/absence of epilepsy be recorded and intellectual function (IQ), vision and hearing be assessed [1].

3.4. Etiologic classification

The categorization based on the actual cause (etiology) and timing of insult was aimed at prevention, and the association of erythroblastosis fetalis with choreoathetoid CP was the paradigm for this classification [6, 20]. The etiology of CP is multifactorial, and the causal pathways are (mechanisms) multiple and complex. The Collaborative Perinatal Project [25] identified the associated risk factors for CP. Due to the fact that much of the data in these epidemiological studies [25, 26] are still correlational, "risk factors" are more appropriate than etiology. These risk factors or associated etiological factors in CP include genetic abnormalities, cerebral dysgenesis, multiple gestation, intrauterine/congenital infection (TORCHS), maternal infection (UTI), prematurity, low birth weight, perinatal asphyxia (HIE), bilirubin encephalopathy, postnatal CNS infections, etc. [19, 25, 26]. These associated etiological factors can be classified according to the timing of insult as prenatal (commonest), perinatal and postnatal [6, 19, 20].

Identifying both the disturbances or events and causal pathways or processes that led to the damage to the developing motor system remains a challenge [6, 20]. This is compounded by the fact that most of these factors are prenatal in timing. Therefore, the etiological classification was severely limited and failed in addressing prevention [6, 20].

3.5. Neuroanatomic (neuropathologic) classification

This classification correlates specific radiologic findings (brain structural alterations) with types of CP [6, 20]. This implies categorizing CP patients based on neuroradiologic findings. Thus, the neuropathologic classification relies on neuroimaging studies such as magnetic resonance imaging (MRI) and computed tomography scan (CT scan).

Neuroimaging contributes significantly to the understanding of the etiology and pathology of CP and the timing of insults [1, 6, 20]. In a systematic review of neuroimaging for cerebral palsy, Korzeniewski et al. [27] classified abnormal radiological findings and diagnoses into five categories namely: malformations, gray matter damage, white matter damage, ventriculomegaly, atrophy or CSF space abnormalities and miscellaneous findings.

Though the correlations between the neuropathologic substrates and clinical types have been weak and inconsistent, recent advances such as diffusion tensor imaging, magnetic resonance spectroscopy, functional magnetic resonance imaging and fast spin echo imaging have improved greatly the possibility of a comprehensive radiologic classification [6, 20, 27]. A recent study by Hou et al. [28] continues to correlate neuropathologic findings with different clinical types of CP. For example, dyskinesia correlated with lesions detected by MRI in the thalamus and putamen due to HIE and in the globus pallidus and hypothalamus due to kernicterus.

There are also difficulties in estimating the timing of insults in CP using neuroimaging findings. It was initially assumed that the presence of neuronal migrational disorders meant that the insult occurred in the first half of pregnancy while the presence of a glial response indicated insults around the second half of pregnancy [27]. However, there is evidence that cell migrational disorders can occur in the last 2–3 months of pregnancy [27]. Nevertheless, malformations are more likely to occur earlier in gestation, and thus, neuroimaging confirmation of their presence can help establish that the cause of CP is unrelated to perinatal events [27].

Categorizing patients with CP based on neuroradiologic findings implies that neuroimaging studies be carried out on all patients. Therefore, it will be difficult to apply such classification in resource-poor countries where neuroimaging facilities are not readily available or affordable and the professional expertise needed may be lacking. Despite this, the American Academy of Neurology (AAN) recommends neuroimaging studies on all children with CP whenever possible [27]. The bottom line is that neuroimaging can be used to identify the neuropathologic substrates of the various etiologic and risk factors of CP, possibly provide information about timing of insults and detect cerebral dysgenesis or malformations but, at present, a comprehensive neuropathologic classification is still in the pipeline.

3.6. Therapeutic classification

This scheme categorizes CP cases based on treatment needs into four groups namely: nontreatment, modest treatment, need for a CP treatment team, and pervasive support groups [6, 20]. Parents/caregivers want their children to receive treatments that will improve their condition, so any classification that is implicative of treatment is important to the patients and their caregivers and relevant to clinical practice. There is a consensus in the literature that the therapeutic and functional classifications are the most important to the patient [1, 6, 20].

However, the therapeutic classification simply identifies how much treatment or the extent of interventions a given child requires without specifying what is actually needed to improve function. This explains the little emphasis on the therapeutic classification.

3.7. Functional classification

Functionally, CP is classified into levels of severity based on functional (motor) abilities and/ or limitation of activity [1, 6, 20]. Currently, the emphasis on the functional classification is due to its important role in the management of CP. So there is a rekindled interest in this scheme.

The functional classification remains the best classification of CP because it is a useful guide to providing care for patients appropriate for their functional level and helps clinicians set and discuss with parents/caregivers realistic rehabilitation goals [1, 4, 5, 11, 12]. The functional classification satisfies the needs of patients and parents/caregivers by informing the current and future service needs of children with CP [5]. It provides information that will permit comparison of CP cases in different locations. It provides information that will allow evaluation of change at different points in time in the same patient such as after an intervention [1].

However, it falls short of giving full descriptive information about a child with CP that clearly delineates the nature of the problem. It does not indicate the nature of the motor abnormality, the topography, the etiology, or neuropathologic substrates which in their own respects are

important descriptive information. Besides, it does not indicate supplemental disorders that are necessary for assessing the service delivery needs of patients with CP.

Iloeje and Ogoke [29] in 2017 reported that the type of CP (physiology and topography), etiological factors and the number of accompanying impairments (supplemental disorders) were positively associated with the severity of gross motor dysfunction and walking ability of children with CP. In that study [29], children with spastic quadriplegic type, bacterial meningitis as etiological factor or many (five or six) accompanying impairments all had severe gross motor dysfunction and were non-ambulatory. Therefore, the other classifications may suggest functional abilities in children with CP.

4. Traditional classifications of CP based on multiple variables

These are the Swedish classification [8], Edinburgh classification [9] and Surveillance for Cerebral Palsy in Europe (SCPE) classification [10].

4.1. The Swedish classification

CP subtypes based on the Swedish classification (1989) [8] are spastic (hemiplegic, tetraplegic, and diplegic), dyskinetic (dystonic and athetotic), ataxic and unclassified/mixed. It is immediately obvious that this classification combines the Minear's Physiologic and Topographic schema. Thus, it shares the same merits and demerits as the physiological and topographic classifications as earlier discussed.

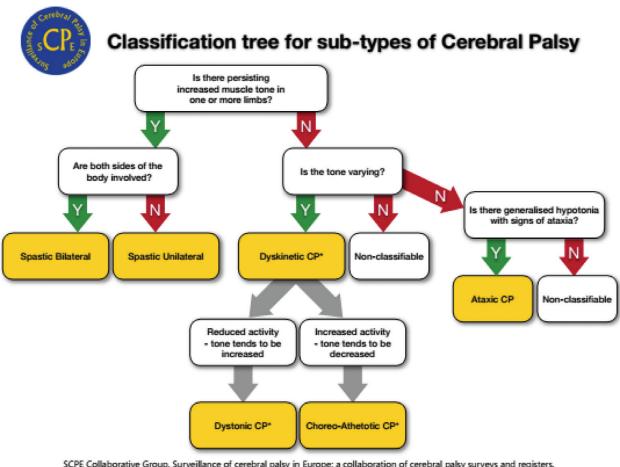
4.2. The Edinburgh classification

According to the Edinburgh classification [9], there are six subtypes of CP namely hemiplegia, bilateral hemiplegia, diplegia, ataxic, dyskinetic and other forms of CP including mixed forms. This classification is a combination of classifications based on topography and physiology and so has the same advantages and shortcomings as the topographic and physiologic classifications.

4.3. The Surveillance for Cerebral Palsy in Europe (SCPE) classification

The SCPE [10] classifies CP into the following four subtype groups: spastic (bilateral and unilateral), dyskinetic (dystonic and choreoathetotic), ataxic, and non-classifiable. This grouping also combines the physiological and topographic classifications. The classification tree of the SCPE for subtypes of CP is shown in **Figure 1**.

Due to the lack of reliability of the terms used in Minear's topographic classification, SCPE [10] introduced two new terms to replace quadriplegia, diplegia, and hemiplegia. These terms are bilateral and unilateral used to describe involvement of both sides and one side of the body, respectively. By this classification, spastic quadriplegia and spastic diplegia are classified as bilateral spastic CP (BS-CP) while spastic hemiplegia is termed unilateral spastic CP. This



SCPE Collaborative Group. Surveillance of cerebral palsy in Europe: a collaboration of cerebral palsy surveys and registers. Developmental Medicine and Child Neurology. 2000;42:816-24.

Figure 1. Classification tree for sub types of CP by SCPE.

classification is easy to apply and is more reliable than the earlier traditional classifications. Therefore, by improving the reliability of the terms used in the topographic component of this classification, the SCPE currently seems to be the best traditional classification for description of patients with CP.

However, the SCPE classification [10] does not include functional abilities and so does not aid therapy for patients with CP. Hence, this classification currently has not had a similar level of advocacy as the functional classifications.

5. Current classifications of CP

Currently, functional classification of each case of CP is internationally advocated due to its important role in management. Thus, current classifications of CP are functional scales for various functions impaired in CP such as communication, gross motor, fine motor, and oromotor/oropharyngeal functions. They are basically ordinal scales to categorize functional abilities or severity of limitation of activity and are not used as outcome measures, tests or assessments [14, 30]. They are simple and easy to apply both by healthcare professionals and care givers and are good for clinical use and patient stratification for research purposes [5, 11, 30]. They have been validated by studies [12, 13, 15] and shown to be objective and reliable clinical classification systems for CP. They have replaced previously used imprecise and subjective functional classifications of CP into mild, moderate and severe.

Their development resulted from the paradigm shift from a focus on body structure and function (impairment-based assessments and treatments) to current emphasis on activity or participation (function and social engagement) [3–5]. These concepts are contained in the ICF [3]. The ICF is a new classification system for health and disease that is universal (for everybody not only people with disabilities) [3]. It is a new way to consider health conditions and posits an interactive relationship between health conditions and contextual factors (environmental and personal factors) in which all components are linked together [3, 4]. It represents a coherent view of health from biological, individual and social perspectives (a biopsychosocial approach to health, functioning and disability) [4]. The ICF model has been used to guide clinical thinking and service delivery to patients with CP [4]. This conceptual change introduced by the ICF is topical.

The functional classifications are analogous and when used together complete the description of daily functional activities in CP at the activity or participation level of the ICF [3, 30]. They include

- a. Gross Motor Function Classification System (GMFCS) [11]
- **b.** Manual Abilities Classification System (MACS) [14] & Mini–MACS [31]
- c. Communication Function Classification System (CFCS) [16]
- d. Eating & Drinking Ability Classification System (EDACS) [17]

There are other functional scales like the Functional Mobility Scale (FMS), Bimanual Fine Motor Function (BFMF), Functional Assessment Questionnaire (FAQ), the Pediatric Orthopaedic Society of North America Outcomes Data Collection Instruments (PODCI), etc.

However, the first four are more commonly used and will be discussed here.

5.1. Gross Motor Function Classification System (GMFCS)

This is the most widely used clinical functional classification of CP [1]. It is an ordinal scale that categorizes a child's mobility/ambulatory or lower limb function in five levels ranging from walking without restrictions (level I) to inability to maintain antigravity head and trunk postures (level V) [11]. The first version of GMFCS was published in 1997 by Palisano et al. [11] and described gross motor functional abilities and limitations in children aged less than 12 years. The upper limit of 12 years (before end of adolescence) was a limitation of the first version, and the GMFCS was revised and expanded in 2007 by Palisano et al. [32] to include an age group for youths 12–18 years. This current version of GMFCS [32] emphasizes the concepts inherent in the WHO's International Classification of Functioning, Disability and Health (ICF). The GMFCS— ER [32] is shown in **Figure 2**. A summary of the criteria for the GMFCS [11, 32] is as follows:

Gross Motor Function Classification System – Expanded and Revised (GMFCS – E & R) BEFORE 2ND BIRTHDAY

LEVEL I: Infants move in and out of sitting and floor sit with both hands free to manipulate objects. Infants crawl on hands and knees, pull to stand and take steps holding on to furniture. Infants walk between 18 months and 2 years of age without the need for any assistive mobility device.

LEVEL II: Infants maintain floor sitting but may need to use their hands for support to maintain balance. Infants creep on their stomach or crawl on hands and knees. Infants may pull to stand and take steps holding on to furniture.

LEVEL III: Infants maintain floor sitting when the low back is supported. Infants roll and creep forward on their stomachs.

LEVEL IV: Infants have head control but trunk support is required for floor sitting. Infants can roll to supine and may roll to prone.

LEVEL V: Physical impairments limit voluntary control of movement. Infants are unable to maintain antigravity head and trunk postures in prone and sitting. Infants require adult assistance to roll.

BETWEEN 2ND AND 4TH BIRTHDAY

LEVEL I: Children floor sit with both hands free to manipulate objects. Movements in and out of floor sitting and standing are performed without adult assistance. Children walk as the preferred method of mobility without the need for any assistive mobility device.

LEVEL II: Children floor sit but may have difficulty with balance when both hands are free to manipulate objects. Movements in and out of sitting are performed without adult assistance. Children pull to stand on a stable surface. Children crawl on hands and knees with a reciprocal pattern, cruise holding onto furniture and walk using an assistive mobility device as preferred methods of mobility.

LEVEL III: Children maintain floor sitting often by "W-sitting" (sitting between flexed and internally rotated hips and knees) and may require adult assistance to assume sitting. Children creep on their stomach or crawl on hands and knees (often without reciprocal leg movements) as their primary methods of self-mobility. Children may pull to stand on a stable surface and cruise short distances. Children may walk short distances indoors using a hand-held mobility device (walker) and adult assistance for steering and turning.

LEVEL IV: Children floor sit when placed, but are unable to maintain alignment and balance without use of their hands for support. Children frequently require adaptive equipment for sitting and standing. Self-mobility for short distances (within a room) is achieved through rolling, creeping on stomach, or crawling on hands and knees without reciprocal leg movement.

LEVEL V: Physical impairments restrict voluntary control of movement and the ability to maintain antigravity head and trunk postures. All areas of motor function are limited. Functional limitations in sitting and standing are not fully compensated for through the use of adaptive equipment and assistive technology. At Level V, children have no means of independent movement and are transported. Some children achieve self-mobility using a powered wheelchair with extensive adaptations.

BETWEEN 4TH AND 6TH BIRTHDAY

LEVEL I: Children get into and out of, and sit in, a chair without the need for hand support. Children move from the floor and from chair sitting to standing without the need for objects for support. Children walk indoors and outdoors, and climb stairs. Emerging ability to run and jump.

LEVEL II: Children sit in a chair with both hands free to manipulate objects. Children move from the floor to standing and from chair sitting to standing but often require a stable surface to push or pull up on with their arms. Children walk without the need for a handheld mobility device indoors and for short distances on level surfaces outdoors. Children climb stairs holding onto a railing but are unable to run or jump.

LEVEL III: Children sit on a regular chair but may require pelvic or trunk support to maximize hand function. Children move in and out of chair sitting using a stable surface to push on or pull up with their arms. Children walk with a hand-held mobility device on level surfaces and climb stairs with assistance from an adult. Children frequently are transported when traveling for long distances or outdoors on uneven terrain.

LEVEL IV: Children sit on a chair but need adaptive seating for trunk control and to maximize hand function. Children move in and out of chair sitting with assistance from an adult or a stable surface to push or pull up on with their arms. Children may at best walk short distances with a walker and adult supervision but have difficulty turning and maintaining balance on uneven surfaces. Children are transported in the community. Children may achieve self-mobility using a powered wheelchair.

LEVEL V: Physical impairments restrict voluntary control of movement and the ability to maintain antigravity head and trunk postures. All areas of motor function are limited. Functional limitations in sitting and standing are not fully compensated for through the use of adaptive equipment and assistive technology. At Level V, children have no means of independent movement and are transported. Some children achieve self-mobility using a powered wheelchair with extensive adaptations.[®] Palisano, Rosenbaum, Bartlett & Livingston, 2007 Page 3 of 4

Figure 2. Gross Motor Function Classification System-Expanded & Revised (GMFCS-E & R). Reproduced with permission.

- **a.** Level I—Walks without limitations.
- **b.** Level II—Walks with limitations.
- c. Level III—Walks using hand-held mobility device.
- **d.** Level IV—Self mobility with limitations; may use powered mobility.
- e. Level V—Transported in a wheelchair.

These general headings or titles for each level represent the method of mobility or highest level of mobility that a child with CP is expected to achieve after 6 years of age [11].

Current management of CP involves a liberal use of adaptive/augmentative equipment in addition to impairment-based treatment approaches to achieve independence [5]. A major goal in the management of CP is to ambulate the children and enable independent living; this gave birth to the changing concepts and the GMFCS. So, how GMFCS is a useful guide to providing care appropriate for the functional level and age of a child with CP?

A child on GMFCS level I will walk independently and so requires no adaptive mobility equipment but appropriate stimulation. The child on level II may need hand-held mobility device when first learning to walk (younger than 4 years) and eventually walks with limitations (after 6 years). Thus, a hand-held mobility device may be provided initially for the child on level II. Therefore, the management of patients on GMFCS levels I and II would focus on appropriate stimulation, preventing complications from occurring and treatment of accompanying impairments. The child on GMFCS level III will require adaptive equipment for low back support for floor and chair sitting and at about 6 years, a hand-held mobility device for walking indoors and a self-propelled manual wheelchair for mobility outdoors and in the community. The management is multidisciplinary depending on the nature and number of accompanying impairments. It is important to note that the child on GMFCS level III may be added to children on levels I and II (walking at least indoors) or to children on levels IV and V (wheeled mobility at least in the community). Nevertheless, GMFCS level III is usually classified as ambulatory because the child is independently mobile in some settings irrespective of the need for assistive mobility device. This need or use of adaptive mobility equipment is acceptable (current thinking) [5]. In addition to multidisciplinary care, the child on GMFCS level IV requires initially a body support walker that supports the pelvis and trunk for floor and chair sitting and later powered mobility and a manual wheelchair for transportation outdoors, at school, and in the community. The management of a child on GMFCS level V involves pervasive supports and a manual wheelchair for transportation in all settings (physical assistance at all times) [11].

5.2. Manual Abilities Classification System (MACS) and mini-MACS

The MACS [14] and the mini-MACS [31] are five-level scales for classifying arm and hand function (manual abilities/manual dexterity) in children with CP aged 4–18 years and 1–4 years, respectively. They classify children's usual performance in handling objects with two hands (not best use or individual hand function) in important daily activities (**Figures 3** and 4).



What do you need to know to use MACS?

The child's ability to handle objects in important daily activities, for example during play and leisure, eating and dressing

In which situation is the child independent and to what extent do they need support and adaptation?

- Handles objects easily and successfully. I. At most, limitations in the ease of performing manual tasks requiring speed and accuracy. However, any limitations in manual abilities do not restrict independence in daily activities.
- П. Handles most objects but with somewhat reduced quality and/or speed of achievement. Certain activities may be avoided or be achieved with some difficulty; alternative ways of performance might be used but manual abilities do not usually restrict independence in daily activities.
- III. Handles objects with difficulty; needs help to prepare and/or modify activities. The performance is slow and achieved with limited success regarding quality and quantity. Activities are performed independently if they have been set up or adapted.
- IV. Handles a limited selection of easily managed objects in adapted situations. Performs parts of activities with effort and with limited success. Requires continuous support and assistance and/or adapted equipment, for even partial achievement of the activity.
- V. Does not handle objects and has severely limited ability to perform even simple actions. Requires total assistance

Distinctions between Levels I and II

Children in Level I may have limitations in handling very small, heavy or fragile objects which demand detailed fine motor control, or efficient coordination between hands. Limitations may also involve performance in new and unfamiliar situations. Children in Level II perform almost the same activities as children in Level I but the quality of performance is decreased, or the performance is slower. Functional differences between hands can limit effectiveness of performance. Children in Level II commonly try to simplify handling of objects, for example by using a surface for support instead of handling objects with both hands.

Distinctions between Levels II and III

Children in Level II handle most objects, although slowly or with reduced quality of performance. Children in Level III commonly need help to prepare the activity and/or require adjustments to be made to the environment since their ability to reach or handle objects is limited. They cannot perform certain activities and their degree of independence is related to the supportiveness of the environmental context.

Distinctions between Levels III and IV

Children in Level III can perform selected activities if the situation is prearranged and if they get supervision and plenty of time. Children in Level IV need continuous help during the activity and can at best participate meaningfully in only parts of an activity

Distinctions between Levels IV and V

Children in Level IV perform part of an activity, however, they need help continuously. Children in Level V might at best participate with a simple movement in special situations, e.g. by pushing a simple button.

Figure 3. The Manual Ability Classification System (MACS). Reproduced with permission [14].



What do you need to know to use Mini-MACS?

Mini-MACS users need to find out what objects the child usually handles and how they handle them: with ease or difficulty, quickly or slowly, with precision or randomly? For example, you can ask about and/or observe how the child uses his or her hands when playing and during meals, or when participating in usual activities of daily living.

Ask questions about the child's self-initiated ability and how much adult help and support the child needs to handle everyday objects, e.g. toys.

Below is a description of the five Mini-MACS levels of children's self-initiated ability and their need for assistance or adaptation when handling objects.

- I. Handles objects easily and successfully. The child may have a slight limitation in performing actions that require precision and coordination between the hands but they can still perform them. The child may need somewhat more adult assistance when handling objects compared to other children of the same age.
- Handles most objects, but with somewhat reduced П. quality and/or speed of achievement. Some actions can only be performed and accomplished with some difficulty and after practice. The child may try an alternative approach, such as using only one hand. The child need adult assistance to handle objects more frequently compared to children at the same age
- Handles objects with difficulty. Performance is slow, with Ш. limited variation and quality. Easily managed objects are handled independently for short periods. The child often needs adult help and support to handle objects.
- IV. Handles a limited selection of easily managed objects in simple actions. The actions are performed slowly, with exertion and/or random precision. The child needs constant adult help and support to handle objects.
- Does not handle objects and has severely limited V. ability to perform even simple actions. At best, the child can push, touch, press, or hold on to a few items, in constant interaction with an adult.

Distinctions between Levels I and II

Children in Level I may have slightly more difficulty handling items that require good fine motor skills compared to children without disabilities of the same age.

Children in Level II handle essentially the same objects as children in Level I, but they may encounter problems performing tasks and/or take longer to perform them, so they often ask for help. Functional difference between hands may cause performance to be less effective. They may need more guidance and practice to learn how to handle objects compared with children in Level I.

Distinctions between Levels II and III

ren in Level II can handle most objects, though they may ta and do so with somewhat less quality, and they may need a lot of guidance and practice to learn how to handle objects.

Level III children manage to use easily handled objects but often need help placing objects in an easy position in front of them. They perform actions with few subcomponents. Performance is slow.

Distinctions between Levels III and IV

Children in Level III manage to use easily handled objects independently for short periods. They perform actions with few subcomponents, and the actions take a long time to perform. At best, children in Level IV can perform simple actions such as

grasping and releasing easily handled objects that are offered in an adapted position. They need constant help.

Distinctions between Levels IV and V

Children in Level IV perform individual actions with a very limited selection of objects and need constant help. At best, children in Level V perform simple movements in special situations. For example, they can press a simple button or hold single simple objects.

Figure 4. The mini-Manual Ability Classification System (mini-MACS). Reproduced with permission.

The MACS, developed in 2006 by Eliasson et al. [14] and modeled on the GMFCS, has been shown by various studies to be valid and reliable. However, a study in 2009 by Plasschaert et al. [33] reported lower inter-rater reliability of the MACS when used in children aged 1–5 years (linear weighted Kappa (k) of 0.67 and 0.55 for 2–5 years and 2 years, respectively). Thus, the MACS was adjusted in 2016 by Eliasson et al. [31] to obtain the mini-MACS which was shown to have excellent inter-observer reliability. The adjustments were simply to obtain descriptions that are applicable to children less than 4 years of age. The mini-MACS differs from the MACS due to the need for assistance in handling objects in children 1–4 years and the nature of the objects they are expected to handle.

The MACS is used to ascertain the child's needs and inform management decisions such as choosing an appropriate upper limb intervention. That is, they are used like the GMFCS to guide functional intervention. For instance, children on MACS levels I and II handle objects independently and do not require any adaptive device to handle objects. The children on level III require some assistance and sometimes adaptive equipment for independent handling of objects. Children on level IV require continuous assistance and adaptive equipment while those on level V need total assistance. Eliasson et al. [31] posited that the mini-MACS is probably not sensitive to changes and should therefore not be used to evaluate development or intervention, but rather to categorize how suspected CP affects the manual abilities of children 4 years and younger.

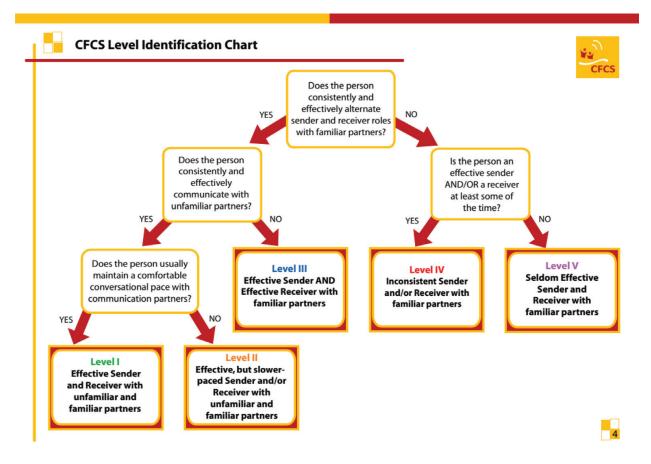


Figure 5. The Communication Function Classification System (CFCS). Reproduced with permission [16].

5.3. Communication Function Classification System (CFCS)

The CFCS was developed and validated by Hidecker et al. [16] in 2011. It classifies everyday communication performance of an individual with CP into five levels ranging from effective communication in all settings (level I) to ineffective communication even with familiar partners (level II). The categorization of the effectiveness of current communication is based on the performance of sender and receiver roles, the pace of communication, and the type of conversational partner. In ascertaining the current level of communication, the CFCS aptly considers and includes use of all methods of communication. This implies that it describes both use of normal verbal and non-verbal communication (speech, gestures, behaviors, eye gaze, and facial expressions) and use of augmentative and alternative communication systems (AACs) (manual sign, pictures, communication books, communication boards and talking devices such as speech generating devices and voice output communication aids) [16]. The CFCS level identification chart is shown in **Figure 5**.

5.4. Eating and Drinking Ability Classification System (EDACS)

The EDACS was developed by Sellers et al. [17] in 2014 and comprises two ordinal scales that describe eating and drinking ability in people with CP from 3 years of age. The five-level scale classifies the safety and efficiency of eating and drinking while the three-level scale classifies level of assistance required to bring food and drink to the mouth. The five-level

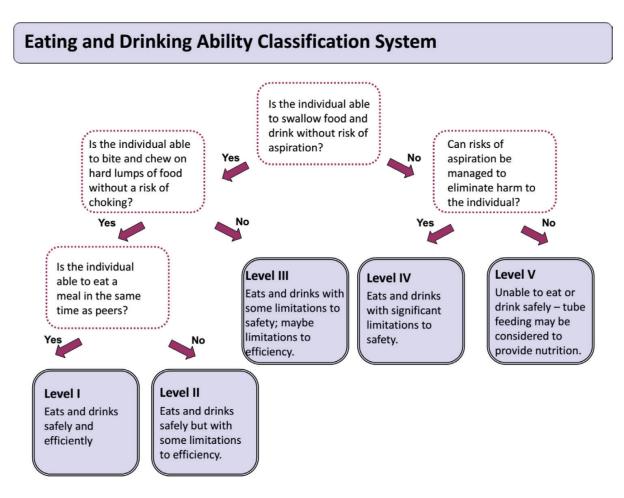


Figure 6. Eating and Drinking Ability Classification System (EDACS) algorithm. Reproduced with permission [17].

scale is based on the range of food textures eaten, the presence of cough and gag when eating or drinking, and the control of movement of food and fluid in the mouth. The three-level scale is categorized into independent, requires assistance, and dependent for eating and drinking. Thus, the EDACS ranges from independent ability to safely and efficiently eat and drink like peers on a wide range of textures (level I) to total dependence for eating and drink-ing and reliance on tube feeding (level V) [17]. The EDACS algorithm is shown in **Figure 6**.

6. The importance of the current classifications

The final goal of a managing doctor and the final hope of a patient and his family is an ambulatory self-dependent individual. Using the functional classifications to guide management helps the pediatrician, the occupational therapist, the physiotherapist, the speech and language therapist and all involved in the care of children with CP to achieve this goal. For instance, the GMFCS is used to ascertain the requirements for ambulation appropriate for the age of the child and gross motor functional abilities while the MACS helps ascertain appropriate upper limb interventions for independent performance of activities of daily living. The CFCS by classifying communication effectiveness in CP is useful in service delivery. It helps identify those that will require augmentative and alternative communication systems to improve their communication. The EDACS assists in identifying the appropriate food texture to give a particular child, need for assistance, the risks involved in eating and drinking and the appropriate method of feeding (oral/tube feeding). Therefore, in simplistic terms, these current classifications tell us what to do to the child with CP. A summary of all groups of classifications is shown in **Tables 1–3**.

Classification axis	Criterion/characteristic used	Inter rater/ inter observer reliability	Suitability for research (description, comparison/ stratification) (on a scale of 1–5)	Indication of functional abilities	Aiding/ guiding current management
Physiological	Type of motor/movement abnormality (quality and changes in tone)	Poor	++	No	No
Typographic	Distribution/localization of motor impairment	Poor	++	No	No
Supplemental	Accompanying impairments	Not reported	Not reported	No	Yes
Aetiologic	Actual cause and timing of insult	Not available	Not available	Not available	No
Neuroanatomic	Brain structural alterations on neuroimaging	Not available	Not available	Not available	No
Therapeutic	Individual treatment needs	Not reported	Not reported	No	No
Functional	Degree of severity/ activity limitation	Good	+++ (good)	Yes. Its major advantage	Yes. Its strength

Table 1. Comparison of traditional (Minear's) classifications based on single variables.

Classification	Minear's classifications combined	Criteria/characteristics used	Inter rater/ inter observer reliability	Suitability for research (on a scale of 1–5)	Indication of functional abilities	Aiding/ guiding current management
Swedish classification	Physiological and topographic	Type of motor abnormality + localization of motor impairment	Poor	++	No	No
Edinburg classification	Physiological and topographic	Type of motor abnormality + localization of motor impairment	Poor	++	No	No
SCPE Classification	Physiological and modified topographic	Type of motor abnormality + localization of motor impairment as unilateral and bilateral only.	Fair	,	No	No

Table 2. Comparison of traditional classifications based on multiple variables.

Classification	Criterion/ characteristic used	Inter rater/ inter observer reliability	Suitability for research	Indication of functional abilities	Aiding/ guiding current management	Age range included (year developed)	Nature of scale(s)
GMFCS	Gross motor/ ambulatory/ lower limb function (current gross motor abilities/activity limitations)	Good	Yes (valid and reliable)	Yes	Yes	GMFCS (birth– 12 years) (1997) GMFCS- E&R (birth– 18 yreas) (2007)	Ordinal (5-level)
MACS	Manual dexterity/ upper limb function (usual performance in handling objects with two hands).	Good	Yes (valid and reliable)	Yes	Yes	MACS (4–18 years) (2006) Mini-MACS (1–4 years) (2016)	Ordinal (5-level)
CFCS	Communication function (everyday communication performance)	Good	Yes (valid and reliable)	Yes	Yes	≥3 years. (2011)	Ordinal (5-level)
EDACS	Eating & drinking ability/ oropharyngeal/ swallowing function (safe and efficiency of eating and drinking and level of assistance required)	Good	Yes (valid and reliable)	Yes	Yes	2–12 years. (2014)	Two ordinal scales (one 5-level, one 3-level)

Table 3. Comparison of current (functional) classifications.

7. A holistic (standardized) classification of CP: the future

The development of a standardized or holistic classification of CP is topical and in tandem with advances in understanding of CP, imaging techniques and quantitative motor assessments [1]. Bax et al. [1] in 2005 proposed a standardized CP classification scheme with four major components namely:

- **1.** Motor abnormalities (a. nature and typology of motor disorder and b. functional motor abilities)
- **2.** Associated impairments
- 3. Anatomic and radiologic findings
- **4.** Causation and timing.

Currently, there are obvious limitations with categorization of neuroimaging findings and identifying specific causes of CP. Therefore, as we await comprehensive and acceptable neuroanatomic and etiologic classifications, the minimum acceptable multiaxial classification of CP for both developed and developing countries should include:

- 1. Classification of motor abnormalities according to SCPE.
- **2.** Accompanying impairments
- **3.** Functional classification levels for: gross motor/ambulatory function (GMFCS), manual abilities (MACS), communication, (CFCS) and eating and drinking ability (EDACS).

This implies that only the first two components of the standardized classification proposed by Bax et al. [1] are applicable currently. The classification by SCPE provides enough clinical descriptive information about children with CP while the supplemental and functional classifications are useful for management and service delivery. The use of the functional scales in clinical context (to aid management) and in research is in accordance with current thinking and the reconceptualization of the management of CP.

8. Conclusions

Each classification system used in CP has its merits and shortcomings. Therefore, the clinical classification of CP needs to use many axes to be comprehensive. Currently, it must include the functional scales so as to guide management.

The neuropathologic classification is being awaited, and due to its contribution to the assessment of etiological factors and timing of insults in CP, it is critical to the development of a holistic or standardized classification of CP.

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Conflict of interest

None.

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