

Proposed definition and classification of cerebral palsy, April 2005

INTRODUCTION

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THE DEFINITION OF CEREBRAL PALSY

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THE CLASSIFICATION OF CEREBRAL PALSY

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Because of the availability of new knowledge about the neurobiology of developmental brain injury, information that epidemiology and modern brain imaging is providing, the availability of more precise measuring instruments of patient performance, and the increase in studies evaluating the efficacy of therapy for the consequences of injury, the need for reconsideration of the definition and classification of cerebral palsy (CP) has become evident. Pertinent material was reviewed at an international symposium participated in by selected leaders in the preclinical and clinical sciences. Suggestions were made about the content of a revised definition and classification of CP that would meet the needs of clinicians, investigators, and health officials, and provide a common language for improved communication. With leadership and direction from an Executive Committee, panels utilized this information and have generated a revised Definition and Classification of Cerebral Palsy. The Executive Committee presents this revision and welcomes substantive comments about it.

Introduction

Cerebral palsy (CP) is a well-recognized neurodevelopmental condition beginning in early childhood and persisting through the lifespan. Originally reported by Little in 1861 (and originally called 'cerebral paresis'), CP has been the subject of books and papers by some of the most eminent medical minds of the past hundred years. Beginning at the end of the 19th century Sigmund Freud¹ and Sir William Osler² both contributed important perspectives on the condition. From the mid-1940s the founding fathers of the American Academy for Cerebral Palsy and Developmental Medicine (Carlson, Crothers, Deaver, Fay, Perlstein, and Phelps) in the USA, and Mac Keith, Polani, Bax, and Ingram of the Little Club in the UK, were among the leaders who moved the concepts and descriptions of CP forward, and caused this condition to become the focus of treatment services, advocacy, and research efforts.

It has always been a challenge to define CP, as documented by the number of attempts that have been made over the years. For example, Mac Keith and Polani³ defined CP as 'a persisting but not unchanging disorder of movement and posture, appearing in the early years of life and due to a non-progressive disorder of the brain, the result of interference during its development.' In 1964, Bax⁴ reported and annotated a definition of CP, suggested by an international working group, that has become a classic and is still widely cited. It stated that CP is 'a disorder of movement and posture due to a defect or lesion of the immature brain.' Although this brief sentence is usually all that is cited by authors, additional comments were added by Bax: 'For practical

See end of paper for list of abbreviations.

purposes it is usual to exclude from cerebral palsy those disorders of posture and movement which are (1) of short duration, (2) due to progressive disease, or (3) due solely to mental deficiency.' The group for which Bax was the reporter felt that this simple sentence could be readily translated into other languages and hoped that it might be universally accepted. They felt that it was wiser at that time not to define precisely what they meant by 'immature brain', as any such definition might limit services to those in need. Like its predecessors, this formulation of the CP concept placed an exclusive focus on motor aspects, and also stressed the specific consequences of early- as opposed to late-acquired brain damage. Sensory, cognitive, behavioural, and other associated impairments, though very prevalent in people with disordered 'movement and posture due to a defect or lesion of the immature brain', and often significantly disabling, were not formally included in the concept.

The heterogeneity of disorders covered by the term CP, as well as advances in the understanding of development in infants with early brain damage, led Mutch and colleagues⁵ to modify the definition of CP as follows: 'an umbrella term covering a group of non-progressive, but often changing, motor impairment syndromes secondary to lesions or anomalies of the brain arising in the early stages of development.' This definition emphasized the motor impairment and acknowledged its variability, previously underscored in Mac Keith's and Polani's definition, and excluded progressive disease, a point introduced in Bax's annotation.

An International Workshop on Definition and Classification of Cerebral Palsy was held in Bethesda, Maryland, July 11–13 2004, co-sponsored by the United Cerebral Palsy Research and Educational Foundation in Washington and the Castang Foundation in the UK, with special support provided by the National Institute of Health/National Institute of Neurological Disorders and Stroke. The task of the attendees was to revisit and, if possible, update the definition and classification of CP in the light of emerging understanding of developmental neurobiology and changing concepts about impairments, functional status, and 'participation'. Reassessment of the definition of CP was prompted by a host of factors: changes in delivery of care to children with disabilities; recognition that children with slowly progressive inborn errors of metabolism can present with motor difficulties at times indistinguishable from those of children with non-progressive disease; increased availability of high-quality brain imaging to identify impairments in brain structure; acknowledgment that developmental motor impairment is almost invariably associated with a range of other disabilities; and increased understanding about associated antecedents and correlates of CP.

The group agreed that CP as conceptualized previously had proved to be a useful nosologic construct, but that previous definitions had become unsatisfactory. They underlined that CP is not an etiologic diagnosis, but a clinical descriptive term. Reservations were expressed about the exclusive focus on motor deficit, given that persons with neurodevelopmental disabilities may present with impairments of a wide range of functions that may or may not include severe motor manifestations, thereby calling for the need for an individualized, multi-dimensional approach to each affected person's functional status and needs. However, it was proposed that the concept 'cerebral palsy' should be retained to serve diagnostic,

management, epidemiological, public health services, and research purposes. It was felt that an updated definition of CP, taking into account the advances in the understanding of physiological and pathological brain development as well as changes in terminology, should be proposed for international use to meet the needs associated with these purposes, as well as to enhance communication among clinicians and scientists. As in the original concept, the motor disorder is emphasized, while it is recognized that other developmental disorders can accompany it. This emphasis is justified by phenotypic differences in motor disorder according to whether pathological processes occur early or late with respect to development, with different management and outcome implications. More generally, it is also justified in the context of brain developmental conditions, given the importance of motor aspects in child development. Evidence of the motor impairments of CP is apparent in the first 18 months of life, but many children who are eventually formally diagnosed with CP have received medical attention for neonatal difficulties such as feeding problems before their gross motor function difficulties become apparent.

To underline the idea that a comprehensive approach to CP needs to be multidimensional and that management of patients with CP almost always requires a multidisciplinary setting, disorders commonly accompanying the motor aspects of CP have been identified in the refined definition. This addition reflects the idea that CP is one of a group of neurodevelopmental disorders which involve numerous developing functions. As in other neurodevelopmental disorders, various manifestations of disordered brain function may appear more significant in different persons or at different periods, e.g. some aspects of the motor impairment, intellectual disability, epilepsy, attentional difficulties, and many others may be more prominent, or more problematic, at different stages of the life of a person with CP.

What follows here is an updated definition and classification of CP, an annotated explanation of the terms used, and the thinking behind the choice of those words. It is hoped that this document will spur discussion, and lead eventually to the goal, first envisioned by Bax 40 years ago, of international consensus and adoption of a common set of ideas about this condition.

The definition of cerebral palsy

Cerebral palsy (CP) describes a group of 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, cognition, communication, perception, and/or behaviour, and/or by a seizure disorder.

ANNOTATION

Cerebral palsy (CP)¹ describes a group² of disorders³ of the development⁴ of movement and posture⁵ causing⁶ activity limitation,⁷ that are attributed to⁸ non-progressive disturbances⁹ that occurred in the developing fetal or infant¹⁰ brain.¹¹ The motor disorders of cerebral palsy are often accompanied by¹² disturbances of sensation,¹³ cognition,¹⁴ communication,¹⁵ perception¹⁶ and/or behaviour,¹⁷ and/or by a seizure disorder.¹⁸

1. 'Cerebral palsy' (CP) – it was generally agreed that the CP concept, essentially a clinical formulation based on phenomenology, remains useful in the current state of nosology. Although the word 'palsy' has become largely obsolete in medical nosography and it has no univocal connotation, the term 'cerebral palsy' is entrenched in the literature and it is used universally by clinicians, therapists, epidemiologists, researchers, policy makers, health care funding organizations, and lay persons. The term 'CP' has, however, been variably used, with poor comparability across different places and times, indicating the need for a consensual definition. Epidemiologists in particular require consistent terminology and concepts across time and space in order to identify changing patterns of diseases and disorders. It was proposed to retain the term to relate future research in CP to existing published work, but to clarify several aspects of the definition in this report.
2. 'a group' – there is general agreement that CP is a heterogeneous condition in terms of etiology as well as in types and severity of impairments. Several groupings are possible and warranted to serve different purposes. These groupings may show overlap. Therefore, the singular form 'CP' is used (as opposed to 'cerebral palsies') as an umbrella term.
3. 'disorders' – this refers to conditions in which there is disruption of the usual orderly processes of child biopsychosocial development. The disorders are persistent.
4. 'development' – the notion of alteration in development is essential to the CP concept. It distinguishes CP from phenotypically similar disorders in children or adults due to late-acquired lesions, at a time when motor development is relatively well developed. The 'developmental' aspect of CP is also important with regard to management strategies that may include interventions that address the developmental consequences of the functional limitations associated with CP, and interventions that are directed at the underlying neurobiological processes. The developmental nature of CP almost always implies impacts on the developmental trajectories of the people who have CP. The motor impairments of CP manifest very early in child development, usually before 18 months of age, with delayed or aberrant motor progress. The clinical picture of CP evolves with time, development, learning, training, therapies, and other factors.
5. 'movement and posture' – abnormal motor behaviour (reflecting abnormal motor control) is the core feature of CP. It is characterized by various abnormal patterns of movement and posture related to defective coordination of movements and/or regulation of muscle tone. Patients with CP may also have other neurodevelopmental impairments that can affect adaptive functioning, sensory function, learning, communication, and behaviour, as well as seizures. Abnormal motor control may be further impaired by features that are associated with CP. However, patients with neurodevelopmental disabilities that do not primarily affect movement and posture are not considered to have CP.
6. 'causing' – activity limitations are presumed to be a consequence of the motor disorder. Thus disorders of movement and posture that are not associated with activity limitations are not considered part of the CP group.
7. 'activity limitation' – the World Health Organization's International Classification of Functioning, Disability and Health⁶ speaks of 'activity' as '...the execution of a task or action by an individual', and identifies 'activity limitation' as '...difficulties an individual may have in executing activities'. This term amplifies the previous concept of 'disability' to recognize changing international concepts and terminology.
8. 'attributed to' – understanding of developmental neurobiology (including the effects of genetic, chemical, and other influences on brain development) is increasing rapidly, such that it is becoming possible to identify structural and other evidence of brain maldevelopment in people with CP. As a consequence, structural-functional connections and correlations are becoming more clearly delineated than has previously been possible. It must, however, be acknowledged that at the present time a full understanding of causal pathways and mechanisms leading to CP remains elusive in many cases.
9. 'disturbances' – this term refers to processes or events that in some way interrupt, damage, or otherwise influence the expected patterns of brain maturation, and result in permanent (but non-progressive) impairment of the brain. In a proportion of cases it is currently not possible to identify a specific 'disturbance' or a specific timing of the events that appear to impact on maturation. These disturbances may include cerebral dysplasia.
10. 'fetal or infant' – the specification 'fetal or infant' reflects the idea that disturbances that occur very early in human biological development impact differently on motor function than disturbances that occur later, even those that occur in early childhood. There is no explicit upper age limit as, depending on aspects of motor functioning, the first two or three years of life may be concerned. Therefore, the notion of early lesion would appear more useful clinically than arbitrarily specified time limits. In practical terms, disturbance resulting in CP is presumed to occur before the affected function has developed (e.g. walking, manipulation, etc.).
11. 'brain' – the term 'brain' includes the cerebrum, the cerebellum, and the brain stem. It excludes motor disorders of spinal, peripheral nerve, muscular or mechanical origin. (Note, however, that alterations in the neuromuscular and musculoskeletal systems may occur in CP as a consequence of the chronic motor impairment. These alterations may restrict further motor function of patients with CP, and be associated with 'secondary' changes in skeletal alignment and/or functional capacity.)
12. 'accompanied by' – in addition to the disorder of movement and posture, people with CP often show other disorders or impairments. These may be caused by the same disturbances as those that caused CP and/or represent indirect consequences of the motor impairment and/or be caused by independent factors (hence the term 'accompanied by' as opposed to 'associated with').
13. 'sensation' – vision, hearing, and other sensory modalities may be affected.
14. 'cognition' – both global and specific cognitive processes may be affected, including attention. Note, however, that when a child has severely delayed cognition and no motor signs (except perhaps for some degree of hypertonicity or hypotonicity) it is not usual to include them within the concept of CP.

15. 'communication' – expressive and/or receptive communication and/or social interaction skills may be affected.
16. 'perception' – the capacity to incorporate and interpret sensory and/or cognitive information may be impaired both as a function of the 'primary' disturbance(s) to which CP is attributed, and as a secondary consequence of activity limitations that restrict learning and perceptual development experiences.
17. 'behaviour' – this also includes behavioural problems in the context of psychiatric disorders, such as features of autism, ADHD, mood disorders and anxiety disorders.
18. 'seizure disorder' – virtually every seizure type and many epileptic syndromes may be seen in patients with CP. Rarely, the seizure disorder may be the cause of CP (e.g. as a consequence of prolonged infantile status epilepticus), or it may result in further motor impairment.

It is hoped that this definition will clarify the CP concept and allow unified use of the term both within and across the concerned fields. As it relies essentially on clinical aspects and does not require sophisticated technology, it should be possible to apply this definition very widely.

The classification of cerebral palsy

CP describes a group of 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 CP may be accompanied by disturbances of sensation, cognition, communication, perception, and/or behaviour, and/or by a seizure disorder. This proposed definition of CP covers a wide range of clinical presentations and degrees of activity limitation, and it is, therefore, useful to further categorize individuals with CP into classes or groups. The purposes of classification include the following.

1. Description: providing the level of detail about an individual with CP that will clearly delineate the nature of the problem and its severity.
2. Prediction: providing information that can inform health care professionals of the current and future service needs of individuals with CP.
3. Comparison: providing sufficient information to permit reasonable comparison of series of cases of CP assembled in different places.
4. Evaluation of change: providing information that will allow comparison of the same individual with CP at different points in time.

Traditional classification schemes have focused principally on the distributional pattern of affected limbs (for example hemiplegia or diplegia) with an added modifier describing the predominant type of tone or movement abnormality (e.g. spastic or dysknetic), but it has become apparent that additional characteristics must be taken into account for a classification scheme to contribute substantively to the understanding and management of this disorder.

INFORMATION REQUIRED FOR CLASSIFICATION

The information available for providing an adequate classification of the features of CP in any individual will vary over the age span and across geographic regions and settings. The role of aging in changing the clinical phenomenology of CP has been little studied, and the possibility of classification changes over time cannot be completely dismissed. Defining the

presence or degree of associated impairments, such as cognitive deficits, is age-dependent, and in infants the type of motor disorder may be hard to characterize. Some young children diagnosed as having CP may in fact have very slowly progressive disorders that have not yet been diagnosed.

Factors other than age will affect classification. Historical data, especially about the course of pregnancy, will vary in reliability and validity. Where neuroimaging facilities, diagnostic specialists, and biochemical laboratories are not available, exclusion of progressive disorders cannot always be ensured, nor can underlying pathology, as described by radiological findings, be incorporated into classification. All classification results should, therefore, indicate the age of the child, the nature of the information available from clinical history (e.g. whether from clinical notes or maternal recall), and the extent to which diagnostic investigation (metabolic or radiological) has been performed.

USES AND LIMITATIONS OF A CLASSIFICATION SYSTEM

Classification often requires making difficult decisions about where to draw the boundaries within ordinal or quantitative measures. Some degree of arbitrariness is inevitable. Assignment of individuals with the diagnosis of CP to distinct clinical groups is not straightforward and will differ depending on the characteristic(s) chosen as the basis for classification. No one single approach has emerged as definitive; depending on the purpose of the classification, certain characteristics or combinations of characteristics may be more useful than others. For example, in assessing the effectiveness of a new treatment for a specific type of tone abnormality, the nature of the motor disorder and the level of functional motor ability are likely to be paramount, whereas determining service delivery needs will require the consideration of associated impairments.

No classification system is useful unless it is reliable. It is, therefore, not enough to specify the characteristics to be used in classification; they must be operationally defined so that, in general, competent examiners will classify the same individual in the same way given identical information. However, providing such definitions is beyond the scope of this document. For example, the term spastic diplegia is problematic for classification because its existing definitions are variable and imprecise, and because we lack evidence that the term can be used reliably. Some use the term to describe children with spastic CP whose only motor deficit is in the legs, whereas others include children who have arm involvement of lesser severity than leg involvement. However, determining the relative severity of arm and leg involvement can be challenging because they perform very different functions.

DEVELOPMENT OF A STANDARDIZED CLASSIFICATION SCHEME

The state of the science underlying the proposed classification has evolved in recent years and continues to progress at a rapid pace, particularly in the area of quantitative assessment of the radiographic and clinical features of CP.

These advances will continue to improve our ability to classify children and adults with CP more accurately. Table I indicates the four major dimensions of classification we propose, which are elaborated upon below.

1. Motor abnormalities

A. Nature and typology of the motor disorder: The type of abnormal resting muscle tone or involuntary movement

disorder observed or elicited is usually assumed to be related to the underlying pathophysiology of the disorder, and may also reflect etiological circumstances, as in kernicterus. Individuals with CP have traditionally been grouped by the *predominant* type of motor disorder, with a 'mixed' category available in those cases when no one type dominates. This strategy has been adopted by the classification system described in the Reference and Training Manual of the Surveillance of Cerebral Palsy in Europe (SCPE),⁷ which divides CP into three groupings based on the predominant neuromotor abnormality: spastic, dyskinetic, or ataxic, with dyskinesia further differentiated into dystonia and choreoathetosis. However, an argument can be made that many children have mixed presentations, and that identifying the presence of *each* of the tone and or movement abnormalities may be of greater clinical and etiological utility, as recommended by the 2001 NINDS workshop on childhood hypertonia.⁸ We take a compromise stance here and recommend that cases continue to be classified by the dominant type of tone or movement abnormality, categorized as spasticity, dystonia, choreoathetosis, or ataxia, but that any additional tone or movement abnormalities present should be listed as secondary types. The term 'mixed' should not be used without elaboration of the component motor disorders.

B. Functional motor abilities: The World Health Organization International Classification of Functioning, Disability and Health,⁶ along with several other recent publications, has sensitized health professionals to the importance of evaluating the functional consequences of different health states. The functional consequences of involvement of the upper and lower extremities should, therefore, be separately classified by using objective functional scales. For the key function of ambulation, the Gross Motor Function Classification System (GMFCS) has been widely employed internationally to group individuals with CP into one of five levels based on functional mobility or activity limitation.⁹ A parallel classification scale, the Bimanual Fine Motor Function (BFMF) Scale, has been developed for assessing upper extremity function in CP but has not been as extensively studied as the GMFCS.¹⁰ A newer instrument for assessing hand and arm function – the Manual Ability Classification System (MACS) – has been shown to have good interrater reliability between parents and professionals, and will shortly be published.¹¹ We follow SCPE in recommending that a functional classification system be applied to hand and arm function in children with CP. Bulbar and oromotor difficulties are common in CP and can produce important activity limitation, but there is as yet no activity limitation scale for such functions. A high priority in research is to develop a scale for speech and pharyngeal activity limitation in CP. In the meantime, the presence and severity of bulbar and oromotor involvement should be recorded.

Although activity limitation is important, the extent to which motor disorders affect the ability to participate in desired societal roles is also an essential consideration. However, at present the evaluation of participation restriction (formerly termed 'handicap') in CP is not well developed, and reliable categorization of children on the basis of this aspect of daily life is, therefore, not yet possible.

2. Associated impairments

In many individuals with CP, other impairments interfere with the ability to function in daily life and may at times produce

even greater activity limitation than the motor impairments that are the hallmark of CP. These impairments may have resulted from the same or similar pathophysiological processes that led to the motor disorder, but they nonetheless require separate enumeration. Examples include seizure disorders, hearing and visual problems, cognitive and attentional deficits, and emotional and behavioral issues. These impairments should be classified as present or absent; if present, the extent to which they interfere with the individual's ability to function or participate in desired activities and roles should be described. SCPE recommends, and we agree, that the presence or absence of epilepsy (defined as two or more afebrile, non-neonatal seizures) be recorded, and that IQ, hearing, and vision be assessed. Although SCPE provides terminology for describing different degrees of cognitive, hearing, and visual impairment, we recommend recording IQ score, corrected vision in each eye, and decibel loss (if any) in each ear whenever this information is available. Standardized instruments are available to measure IQ, vision, and hearing, and categories describing specific levels of dysfunction (e.g. visual impairment, profound hearing loss, and mild mental retardation*) have come to be generally accepted.

3. Anatomic and radiological findings

A. Anatomic distribution: The pattern and extent of the motor disorder in CP with regard to different anatomic areas should be specified. Previous classification schemes included only the extremities and required a subjective comparison of severity in the arms and the legs. Notably missing from current anatomic classification schemes is a description of truncal and bulbar involvement. We recommend that all body regions – trunk, each limb, and oropharynx – be described in

*UK usage: learning disability.

Table I: Components of CP classification

1. Motor abnormalities

A. Nature and typology of the motor disorder: the observed tonal abnormalities assessed on examination (e.g. hypertonia or hypotonia) as well as the diagnosed movement disorders present, such as spasticity, ataxia, dystonia, or athetosis

B. Functional motor abilities: the extent to which the individual is limited in his or her motor function in all body areas, including oromotor and speech function

2. Associated impairments

The presence or absence of associated non-motor neurodevelopmental or sensory problems, such as seizures, hearing or vision impairments, or attentional, behavioural, communicative, and/or cognitive deficits, and the extent to which impairments interact in individuals with CP

3. Anatomic and radiological findings

A. Anatomic distribution: the parts of the body (such as limbs, trunk, or bulbar region) affected by motor impairments or limitations

B. Radiological findings: the neuroanatomic findings on computed tomography or magnetic resonance imaging, such as ventricular enlargement, white matter loss, or brain anomaly

4. Causation and timing

Whether there is a clearly identified cause, as is usually the case with postnatal CP (e.g. meningitis or head injury) or when brain malformations are present, and the presumed time frame during which the injury occurred, if known

terms of any impairments of movement or posture. A scale for describing truncal posture in CP has recently been developed.¹²

It is clearly important to distinguish unilateral from bilateral motor involvement, and categorization based on this distinction has good reliability.⁷ However, even this distinction can be blurred because many children with primarily unilateral CP may also have some degree of motor involvement on the opposite side and some children with primarily bilateral involvement may have appreciable asymmetry across sides. Although the terms 'diplegia' and 'quadriplegia' have been extensively used in research and clinical practice, we propose that these terms not be used in classification. Gorter et al.¹³ have documented the imprecise use of these terms in clinical practice. We advise that the anatomic distinction between unilateral and bilateral CP be coupled with a description of the motor disorder and functional motor classification in both upper and lower extremities.

B. Radiological findings: Until recently, correlations between radiographic findings and clinical presentation in CP were weak. However, advances both in imaging technology and in quantitative motor assessments are changing this picture. The goal of categorizing all patients on the basis of specific radiographic findings will require more development before implementation, but we concur with the recommendation of the American Academy of Neurology to obtain neuroimaging findings on all children with CP whenever feasible.¹⁴ At present, information is insufficient to recommend any specific classification scheme for neuroimaging findings.

4. Causation and timing

It is increasingly apparent that CP can result from the interaction of multiple risk factors, and in many cases no identifiable cause can be found. Therefore, although every reasonable effort should be undertaken to investigate causes or causal pathways, clear-cut categorization by cause is unrealistic at the present time. It is possible that by looking further downstream from putative cause to common mechanisms of injury, and by grouping cases on that basis, we may ultimately have a more salient method of classification. Timing of insult should be noted only when reasonably firm evidence indicates that the causative agent, or a major component of the cause, was operative in a specific time window, as, for example, with postnatal meningitis in a previously well infant. Although recording adverse events in the prenatal, perinatal, and postnatal life of a child with CP is recommended, clinicians should avoid making the assumption that the presence of such events is sufficient to permit an etiological classification that implies a causal role for these events in the genesis of CP in the affected individual.¹

¹Note: See editorial and commentaries at the beginning of this journal which deal with issues raised in this paper. In addition the editorial in the July issue of DMCN by Ingeborg Krägeloh-Mann discusses areas of interest raised here.

Comments are also invited on the Castang Foundation website www.castangfoundation.net (where some comments are already posted).

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List of abbreviations

GMFCS	Gross Motor Function Classification System
SCPE	Surveillance of Cerebral Palsy in Europe
