

Definition and classification of cerebral palsy

The International Workshop on the Definition and Classification of Cerebral Palsy have proposed a fresh definition and classification of cerebral palsy (CP; p 571). In their accompanying paper they explain their reasoning behind this. Why is there the need for revision of the widely accepted and comfortably familiar definitions of Bax¹ and Mutch²? These definitions are beautifully succinct and clear but perhaps not inclusive enough, making no mention of pathogenesis or of the functional and 'non-motor' features of CP which are most significant when it comes to everyday life. There has been debate in the past as to whether a more aetiological definition should be adopted but we are not yet ready for this. CP remains unexplained in around 15% of children, even after good neuroimaging and metabolic investigation³ and we recognize that a wide range of CNS disorders can result in a similar clinical picture. Thus, a predominantly phenotypic definition and classification remains the most appropriate. The workshop clearly deliberated long and hard over every word of the revised definition producing a much more detailed description of CP that now encompasses the effect on function (activity limitation) and the comorbid features. As a result, the definition is longer and it does not 'trip off the tongue' in the way the old ones did.

I think a fresh classification of CP is most welcome. With the standard classification there is considerable confusion in describing the neurological abnormalities of tone and the anatomical distribution of the disorder. When does hemiplegia become an asymmetric diplegia or diplegia become quadriplegia and when does a spastic disorder become mixed spastic/dystonic? Causation is not always described and functional consequences are not systematically recorded. This makes it difficult to document significant changes for an individual (particularly important when it comes to any therapeutic interventions) and is unsatisfactory for scientific research.

The Workshop has set out a useful and detailed template listing the important components of a classification system. There is much greater emphasis on timing and causation of injury and I welcome and fully endorse their advice that, whenever possible, the diagnosis should be confirmed with neuroimaging. It is also entirely appropriate that functional assessments should be routinely employed for which suggestions are given in the accompanying document. For clinicians and therapists the most provocative suggestion is that CP should now be classified as unilateral or bilateral with the old terms of diplegia, quadriplegia, etc being abandoned, and that there should be specific description of all body areas affected (including trunk and oromotor involvement). The predominant abnormalities of tone should also be recorded and the presence of a hyperkinetic movement disorder documented.

Certainly the old system was confusing and inaccurate. This new system will undoubtedly be helpful for research purposes and is clinically more useful than the old system. The challenge for clinicians and therapists is how to re-educate ourselves and incorporate this system into our everyday practice. Can we extract working 'rules of thumb' that will evoke in our minds an immediate and accurate impression of the dominant problems that the child or adult with CP is facing? The challenge for the Workshop is how to disseminate these new guidelines, collect feedback, and further refine them. Our comments are invited.

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Revisiting and updating the definition and classification of cerebral palsy (CP) is not just an academic task for the developmental paediatrician and neurologist but an exercise aimed at providing a framework for multidisciplinary management by all professionals and care providers involved in the care of children with developmental disabilities.

The overall aim of any classification of health-related states, and an internationally acceptable definition of the same, is to provide a standard language for purposes of identification (diagnosis) and provision of services (management) to meet the different needs of those affected, in addition to providing a systematic coding for Health Information Systems. In the case of CP, a paradigm for childhood disabilities and a health-related condition which is as much a sociological subject, the need to have a consensual definition is even greater because of the multidisciplinary management the condition calls for. There is also need to improve communication between different users such as health care workers, educators, researchers, policy makers, and the public (including persons with disabilities).

Static encephalopathies, disorders of brain function which arise in the fetal or perinatal period and remain relatively stable in later life, can be produced by a variety of mechanisms and varied causes.¹ In light of an increased understanding of

developmental neurology, it appears that the functional limitations in CP (and allied conditions in this group) are related to disturbances in the organization and function of specific neuronal circuits within the brain.² The nature of the developmental problems are dependent on the timing, nature, and magnitude of the disrupting influence (such as hypoxia, drugs, infections). It is, therefore, imperative that a multipurpose definition designed to serve various disciplines should also look at providing a scientific basis for understanding the determinants and outcomes.^{1,2}

Hence, accepting to write a commentary on the proposed new definition was not without some trepidation, considering that all of us would like to retain the concept 'cerebral palsy' as a clinical descriptive term to describe a group of developmental motor disorders with varied aetiological associations and much phenotypic differences in the clinical presentation even as we attempt to refine the definition.

The consensual definition of cerebral palsy emerging from the Washington workshop of July 2004 appears more or less comprehensive in describing the condition. Recognizing 'activity limitation' as an important component of the definition should make the definition more acceptable to all concerned. Another commendable feature is the emphasis on the accompanying disturbances or functional limitation in other domains besides 'motor', as these singly or in concert can affect participation greatly.

I do, however, have some reservations in accepting the definition in full and would like to place before you a couple of suggestions to consider before the consensual definition is declared the International definition of CP.

Firstly, it would perhaps be useful to include the disorder of development of coordination (dysequilibrium) and have the first component of the definition as: 'CP describes a group of disorders of development of movement, posture, and coordination.'

There is a need to widen the scope of the definition to include older children with CP from post-natal causes. It has been our experience while working with the under-6 years' age group in underprivileged communities that a considerable number of children presenting with CP have a history of antecedent infections (e.g. meningoencephalitis, measles, tuberculosis, and their sequelae) or disruption of CNS development in their early childhood. These include causes as varied as near-drowning, exposure to drugs and toxins, trauma from accidents, metabolic disturbances, and dehydration from diarrhoeal episodes.³⁻⁵ This has major implications for epidemiological and public health purposes as well as for clinical research and service coverage. Insults to the developing brain beyond infancy affect many children in the developing countries (including India), hence the need to modify the component that refers to 'fetal or infant brain'.⁶ We may even require to be more specific regarding the timeframe (e.g. developing central nervous system during the childhood years).

As one involved in the development of the World Health Organization's International Classification of Functioning, Disability and Health, I am also partial to the terms 'activity limitation' and 'participation'.⁷ However, I'm not in agreement with the statement that, 'disorders of movement and posture not associated with activity limitations are not considered part of the CP group'. It would be akin to denying the diagnosis of epilepsy in an individual with no activity limitation if his seizures are controlled on AED therapy and he is well adjusted

socially, participation being a social construct and an additional dimension to the classification of disabilities. The outcomes evaluation, so dear to early interventionists, is an area that would be most influenced by this component of the definition.

I, for one, would prefer having the functional component as an additional qualifying statement, perhaps put this way:

'Cerebral palsy describes a group of disorders of the development of movement, posture, and coordination, attributable to non-progressive disturbances affecting the brain in its early developmental phase, in other words, the fetal, infantile, and early childhood development. The motor disorder is often accompanied by disturbances of sensation, cognition, communication, perception, behaviour and/or by a seizure disorder, contributing further to activity limitation and restricted participation.'

Retaining the concept of CP as it is understood today and updating the definition to be more comprehensive is no small task and I hope it will not be long before the experts can come up with the final version of the definition that can gain universal acceptance and wide application.

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'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.'

In defining CP, it is important primarily that the term is kept for historical and cultural reasons, as a label accepted by those who have CP. Any attempt to change the term would result in rejection by users, as opposed to the way the term 'spastic' was used.

In looking at 'activity limitation', 'limitation' may be incorrect, I would replace the former with impaired function, as activity relates to social/cultural activities such as education.

In the list of additional impairments, I would add continence, very common now in more severe CP.

Something needs to be mentioned firstly about the increased wear and tear on the body, and secondly, the additional medical issues related to CP such as chest infections, reflux, trapped nerves, etc. The reason is that although CP is indeed non-progressive, it does affect aging and additional complications can occur during a person's life. For example, an ambulant person may become a wheelchair user.

In defining CP, it is important to acknowledge that it has a unique appearance as opposed to head injury, and that the type of motor disorder displays a variety of clinical phenotypes or pictures.

In general, I like the definition and would hope to use it to define myself in terms of impairment.

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The occurrence of the International Workshop on Definition and Classification of Cerebral Palsy, in Bethesda, Maryland, USA, July 2004, was an encouraging sign of progress towards a collaborative attempt to reach an internationally accepted definition and classification system for cerebral palsy (CP). The definition given in this issue, apparently submitted by a small subset of workshop attendees, is disappointing in that it does little to address the four areas of imprecision encountered in previous definitions of CP, as described by Stanley et al.¹ These four areas are:

(1) What is meant by non-progressive? This term is one of the few not annotated by the authors, but perhaps one of the few that really needs to be. What steps need to be taken and how long must one wait to exclude the possibility of slow progression?

(2) The age limits for (a) the maximum age at which the 'disturbance' can occur and (b) the minimum age at which the disorder can be recognized in children who die early.

(a) appears to be addressed with the terms 'fetal' or 'infant'. The word infant is not a standardized term, and though it is variously defined in the literature it has not been defined in the annotations appended to this definition. Indeed, Annotation 10 makes it clear that the term infant is not intended to convey an age limit because the authors suggest that the upper age limit for acquisition should be before the affected function has developed. However, many functions may be affected in a person with CP, and they develop at different times throughout childhood. Do the authors have in mind the least skilled affected function (head lifting perhaps), or the most skilled (like playing a musical instrument)?

(3) The lower limit of severity required to warrant the label of CP. The necessity of 'activity limitations' does not set a criterion for the lower limit of severity because neither the activities nor the extent to which they must be limited are defined. For example, people with spastic hemiplegia and excellent unilateral hand function, may experience no real activity limitations: do they not have CP?

(4) There is no mention of the problem of recognized syndromes, exhaustively described by Badawi et al.,² which for historical reasons have not traditionally been included in the

CP group although they meet the inclusion criteria. This may be irrelevant for the types of prospective studies outlined in the Introduction, but are critical to the integrity of retrospective studies and registers from which much of our information about this relatively rare condition has been derived.

The very general discussion on classification is patchy; patches of insight and patches of oversight. The purposes of classification should include an understanding of aetiology, of paramount importance to parents and prevention scientists, and an understanding of pathology which may, in the future, lead to the possibility of 'cure' in some cases and, perhaps in the shorter term, to more accurate prediction. These are the least developed of the possible classification systems, but sufficiently developed to be discussed in sections 3 and 4.

The authors write that 'traditional classification schemes have focused primarily on the distributional pattern of affected limbs...with an added modifier describing the predominant type of tone or movement abnormality'. However severity has also traditionally been considered, although before recent developments in measures of motor function reliability of severity classification has been problematic.

The recommendation of recording of associated impairments and of secondary motor impairments, as is routine in Australian CP Registers, is welcome, but suggestions as to how to overcome the problems of measuring associated impairments in children with multiple deficits when traditional methods rely on other systems being intact would be more welcome still. Nor are any methods of recording multiple motor types suggested although the system currently being devised for the Australian National Register was presented at the Bethesda meeting in 2004.

The last sentence is worth reiterating and extending: 'While recording adverse events in the prenatal, perinatal and postnatal life of a child with CP is recommended, clinicians, lawyers, and families should avoid making the assumption that the presence of such events is sufficient to permit an etiologic classification that implies a causal role for these events in the genesis of CP in the affected individual.' But it has little to do with CP classification. If the aim of an international definition and classification systems for CP is to enable comparative research across international boundaries, both definition and classification systems must be clearly, succinctly, and precisely specified, they must address identified problematic areas and must be arrived at by a process that is both transparent and collaborative.

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