The Definition and Classification of Cerebral Palsy

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Foreword

This supplement is centred on the final version of the Report on the Definition and Classification of Cerebral Palsy from the group chaired by Murray Goldstein and Martin Bax. We have devoted a Supplement to it for several reasons, including the importance of the topic and the advantage of having a separate stand-alone section to use for reference. It also allows the Report to be seen in its context.

This final version of the Report is based on the discussion paper published last year, which was accompanied by commentaries, [1-3] and followed by an extensive discussion on the Castang website (www.castangfoundation.net/workshops_washington_public.asp) as well as correspondence in the Journal. [4] These comments have been taken into account in the revised version. It is followed by a section summarizing most of the presentations at the workshop in Bethesda in 2004 which provided the background to the present Report. At that meeting selected international experts discussed specific aspects. These are very informative and reflect a wide range of considerations and perspectives, both on the difficulties involved and on the value and use of classification in terms of diagnosis, prognosis, management, and clinical trials. The presentation by Krägeloh-Mann has since been expanded into a review of the role of neuroimaging in cerebral palsy (CP), which is published separately in the accompanying issue of the journal (DMCN 2007; 49: p 144–151).

The Report is preceded by a paper giving a brief history of the concept of CP, which is later also covered by Gilles from a pathological perspective. In a subsequent section are three papers describing the definitions and classifications currently in use by the European (SCPE) and the Australian research groups, and those of Mutch et al., as it is instructive to compare the different methods used in formulating these. The final section has brief articles looking forward to the implications of the report on clinical practice and the provision of health care.

I hope that this Supplement will be useful. It illustrates the difficulties inherent in trying to agree what we mean by the terms we use and that a classification that suits one purpose, such as a diagnostic approach, may not always be ideal for others, such as therapy issues. Defining and classifying CP is far from easy, so the group who have produced the Report deserve applause. We do need a consensus that can be used in all aspects of day-to-day care and for future research on CP.

Peter Baxter

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Definition and classification of cerebral palsy: a historical perspective

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The definition of a diagnosis identifies explicitly which cases are to be recorded under that term and, by implication, which are to be specifically excluded. The definition is the basis for planning treatment and for counting cases in a population. Classification within a diagnosis categorizes those cases with similar characteristics together and distinguishes those cases with diverse features apart. The design of a classification system, for instance whether it is organized into nominal or ordinal categories, will vary depending on the concept being classified and intended purpose for which classification is being made. The most frequently cited definition of cerebral palsy was published by Bax (1964) as 'a disorder of posture and movement due to a defect or lesion in the immature brain'. The label does however encompass a variety of syndromes and some, therefore, prefer the term *cerebral palsies*.

Cerebral palsy (CP) is now familiar to most health and social service professionals, as well as to many members of the general public, as a physically disabling condition. In fact, although CP only affects between 2 and 3 per 1000 live births, it is thought to be the most common cause of serious physical disability in childhood (Surveillance of Cerebral Palsy in Europe 2000). Historically, CP was predominantly studied in relation to the pathology and aetiology of the impairment. Discussion regarding the definition and classification of CP was first recorded in medical literature during the nineteenth century, predominately in French, German, and English language publications. However, what exactly the term 'cerebral palsy' describes has been debated for more than 150 years, and discussions about how the different manifestations of CP can be best classified continue to the present day.

Before 1900

The quest to correlate brain lesions with their clinical manifestation began with early French publications by pathologists debating the association of hemiplegia of the body with hemiatrophy of the brain identified by post-mortem (Lallemand 1820, Cazauvieilh 1827 [as cited in Ingram 1984]). However, the seminal work describing *cerebral paralysis*, and particularly the related musculoskeletal issues, was elucidated by an English orthopaedic surgeon named William Little in one of a series of lectures in 1843 entitled 'Deformities of the Human Frame'. Whilst his lectures focused on joint contractures and deformities resulting from long-standing spasticity and paralysis, Little clearly indicated that the cause of the spasticity and paralysis was often damage to the brain during infancy, and specifically preterm birth and perinatal asphyxia (Little 1843). Little also noted that behavioural disorders and epilepsy were only occasional complications and not central to the condition.

At about the same time, a German orthopaedic surgeon, von Heine, was reporting similar clinical syndromes as a result of infections such as scarlet fever and vaccinations (von Heine 1860). He cited the work of his compatriot Henoch, who had written his dissertation several years earlier, describing hemiplegia in children (Henoch 1842). It has been suggested that it was actually von Heine, rather than Little, who first distinguished CP from the flaccid paralysis caused by poliomyelitis (Osler 1889, Bishop 1958). However, Little was known to have spent some years studying in Germany during the 1830s and it is possible that there was some cross-fertilization of ideas, although this is not formally recorded. Regardless, CP was known for many years after as 'Little's Disease'.

In his best known work, published in 1862, Little expands on the association between a large number of his patients' clinical presentation and their birth history as recalled by the family (Little 1862). Little differentiated between the congenital deformities observed at the time of birth, such as falipes equinovarus, and the limb deformities that developed subsequent to preterm, difficult, or traumatic births, due to what he termed spastic rigidity. He demonstrated his familiarity with the work of French, German, and Irish pathologists in constructing his theory. Little grouped the clinical presentation of 47 cases as either: (1) hemiplegic rigidity affecting one side only, although lesser impairment of the apparently uninvolved limb was frequently observed; (2) paraplegia affecting both legs more than arms; and (3) generalized rigidity. Little showed careful consideration for his audience in the

published discussion by conceding to the President of the Obstetrical Society of London that for every 'one (case) that depended on abnormal or premature labour there were twenty or more from other causes incidental to later life'. Sarah McNutt, an American physician, continued to raise the profile of the risks of long-term disability arising from birth trauma (McNutt 1885). Notably, the American Neurological Association admitted her as their first female member; but the content of her lectures apparently made her unpopular with some eminent obstetricians whilst she was on a tour in the UK (Ingram 1984).

At the time he was resident in America, the eminent Canadian William Osler published articles in 1886 and 1888 before his more notable monograph was published in London in 1889. 'The Cerebral Palsies of Children' comprehensively described his study of a case series of 151 patients (Osler 1889). Osler acknowledged the contributions from his German, French, English, and American colleagues and stated that he would 'for clearness and convenience adhere to custom and classify cases according to the distribution of the paralysis, whether hemiplegic, diplegic or paraplegic'. In fact, he classified his cases into the three categories but used the terms: (1) infantile hemiplegia; (2) bilateral spastic hemiplegia; and (3) spastic paraplegia. Osler references the synonym *spastic diplegia* for bilateral spastic hemiplegia to Samuel Gee at St Bartholomew's Hospital in London. William Osler later moved from Pennsylvania to become Regius Professor of Medicine at the University of Oxford and was knighted in the UK for his contributions to medicine.

In the year following Osler's seminal book, the neurologists Sachs and Peterson published their series of 140 cases (Sachs and Peterson 1890). They contrasted the comprehensive understanding that had then been achieved regarding the clinical symptoms and pathology of poliomyelitis with the dearth of understanding about CP. Sachs and Peterson followed the convention of the time by using the same classification system as Osler: hemiplegic, diplegic, or paraplegic. Where possible, they investigated aetiology using postmortem examinations but concluded that any of the three clinical presentations could result from a variety of causes. Despite this lack of correlation they advocated that classification should include 'special reference to the pathology of the disease'.

Sigmund Freud was of the opposite opinion (Freud 1893). Despite his background in neuropathology, he advocated classifying CP using only clinical findings. Freud recognized that, even with post-mortem examination, the pathological findings resulted from a combination of the initial lesion and repair process and, therefore, were only partially related to the clinical manifestation. His classification system combined previously separate categories under the single term 'diplegia' for all bilateral disorders, as distinct from hemiplegia. The term diplegia was used to describe generalized rigidity of cerebral origin, paraplegic rigidity, double spastic hemiplegia, generalized congenital chorea, and generalized athetosis. Athetosis had already been described, initially by Hammond, as involuntary writhing movements in adults affected by hemiplegia (Hammond 1871), and it would later be more clearly differentiated from other movement disorders by Gowers (1876). Freud's observations regarding aetiology identified three groups of causal factors: (1) maternal and idiopathic congenital; (2) perinatal; and (3) post-natal

causes. He noted that it was difficult to know whether later problems resulted from birth trauma, as described by Little, or whether in fact there were predisposing factors that may have caused these infants to have difficult births. He thought the task of separating congenital from acquired cases impossible in some cases and generally unhelpful. Freud was aware that children with ataxic symptoms might require a separate group, as became the case after the work of Batten (1903), but at the time of his writing he had not seen enough cases of non-progressive ataxia to be sure.

Freud lost interest in CP and instead focused on his study of psychoanalysis (Accardo 2004). Nevertheless, his influence was such that his lasting statements regarding the futility of attempting to associate clinical syndromes with neuropathology may have predisposed to the dearth of research about CP during the first half of the twentieth century. Also, at that time, poliomyelitis and tuberculosis were more common causes of disability and, therefore, attracted greater attention from medical researchers.

From 1900 to 2000
In the early 1920s, some 30 years after Freud's comments, an American orthopaedic surgeon made the next major contribution to our understanding of CP (noted by Mac Keith and Polani 1959). Winthrop Phelps pioneered modern approaches to the physical management of children with CP advocating physical therapy, orthoses, and nerve blocks. In a later article Phelps identified his four treatment goals: locomotion, self-help, speech, and general appearance (Phelps 1941). His approach to surgery was conservative. Phelps acknowledged the need for a neurological classification system for diagnostic purposes but preferred to use his own classification system as a basis for treatment. He proposed that classification should be made on a functional basis including both mental and physical ability, and that a social assessment should precede treatment. Phelps grouped all movement disorders under the term dyskinesia, and used spasticity, athetosis, overflow or synkinesia, incoordination or ataxia, and tremor as sub-categories. He noted that these five varieties rarely occurred in pure form. Phelps helped to found the American Academy for Cerebral Palsy in 1947 and was elected its first president. The Academy's mission remains 'to foster and stimulate professional education, research, and interest in the understanding of these conditions and in improving the care and rehabilitation of affected persons' (American Academy for Cerebral Palsy and Developmental Medicine 2005).

American neurologist Myer Perlstein recognized the prevailing confusion regarding classification of CP and contributed a lucid account of the various systems that existed in the 1940s and 1950s (Perlstein 1952). He recounted methods for classifying children according to the anatomical site of the brain lesion, clinical symptoms, degree of muscle tone, severity of involvement, and aetiology. Thus, he suggested that a modular description using components from each category can be assembled. Minear conducted a survey with the members of the American Academy for Cerebral Palsy in 1953 and published the resulting classification system based on their majority opinion (Minear 1956). He defined CP simply as any 'symptom complex' arising from non-progressive brain lesions. Minear's system is similar to Perlstein's in that it is more of a comprehensive listing of all clinical symptoms

with categories for motor impairment, topography, aetiology, supplemental, neuro-anatomical, functional capacity, and therapeutic requirement. A separate dimension for functional capacity with four levels is included in the classification but used undefined terms such as mild and moderate limitation of activity.

Meanwhile in the UK, the classification systems used to describe case series by Evans (1948) and Asher and Schonell (1950) comprised different combinations of topography and motor impairment. Wyllie (1951) used a confusing combination of neurological and aetiological criteria to define categories which were: (1) congenital symmetrical diplegia; (2) congenital paraplegia; (3) quadriplegia or bilateral hemiplegia; and (4) hemiplegia. The selected category was supplemented with a statement of the type of motor disorder: spastic, flaccid, mixed, athetoid, or ataxic. Harking back to Freud's argument that it was not possible to classify using aetiology, Ingram preferred a system using neurological and topographical categories, supplemented with an indication of the severity using the terms mild, moderate, and severe (Balf and Ingram 1955). The Ingram classification separated hemiplegia, double hemiplegia, and diplegia from ataxic and dyskinetic categories. Ingram grouped involuntary movement disorders, such as dystonia, chorea, and athetosis, under the term dyskinesia. Ingram pointed out that transient changes in muscle tone seen consistently in children with diplegia would require their continual reclassification if the terms 'rigidity' or 'spasticity' were used as categories.

Again in the UK, in 1957 Mac Keith and Polani convened an informal group called the Little Club that was dedicated to thinking through the terminology for describing CP. The Little Club published its definition of CP as 'a permanent 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' (Mac Keith and Polani 1959). The Little Club classification uses the term 'spastic' with sub-categories of hemiplegia, double hemiplegia, and diplegia; the other categories were dystonic, choreo-athetoid, mixed, ataxic, and atonic CP. Ingram continued his aforementioned criticism citing the changes observed in the series of 1821 patients by Bronson Crothers (Crothers 1951) that would require cases to be moved continually between classification categories (Ingram 1984). Some of the original Little Club members refined the definition of CP as 'a disorder of posture and movement due to a defect or lesion of the immature brain' and 'for practical purposes disorders of short duration, due to progressive disease or due solely to mental deficiency were excluded' (Bax 1964). The group noted the inconsistent interpretation of terms such as 'spastic' between different professional and country cultures. These inconsistencies precluded further progress which led to their conclusion that, at that time, it was 'impossible to proceed definitively with classifying cerebral palsy' (Bax 1964).

In the 1980s, another expert group commissioned by the Spastics Society (now SCOPE) discussed how to classify CP from an epidemiological perspective (Evans and Alberman 1985; Evans et al. 1986, 1987). Evans' group were particularly interested in monitoring rates of CP in populations as public health markers of perinatal and neonatal health care. Their approach built upon earlier work by Fiona Stanley and others in Western Australia for a 'limb-by-limb' classification system.

The subsequent 'Evans form' recorded details of central motor deficits in terms of the neurological type: (1) hypotonia; (2) hypertonia (including stiffness, spasticity, and rigidity); (3) dyskinesia; and (4) ataxia (Evans et al. 1987). A decision was made to record details of each limb and the head and neck separately. The 'Evans form' also enabled recording of functional mobility and manual dexterity in one of four ordinal levels, the presence of intellectual and sensory impairments, communication difficulties, seizures, congenital and acquired malformations, as well as genetic and other disorders. Some effort was made to validate this system, with repeated meetings showing videos to test inter- and intraobserver, and within and between patient variations. However, details of the reliability and validity of their classification were not widely disseminated.

A summary of several meetings held in Europe and America between 1987 and 1990 was published by Mutch et al. (1992) resulting in a further revised definition to underline the heterogeneity of the condition: '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'. Notably this annotation also included a revised Swedish classification system which, whilst still not perfect, offered simplicity as its major asset. The three neurological categories were spastic, ataxic, and dyskinetic; these were subcategorized in mixed ways as hemiplegia, tetraplegia, or diplegia for spastic cases; as either diplegic or congenital for ataxic cases, and as either mainly chorioathetotic or mainly dystonic for dyskinetic cases. Whilst noting that at the time it remained beyond their capability, the authors resuscitated the yearning for an aetiologically-based classification system (Mutch et al. 1992).

The Gross Motor Function Classification System (GMFCS) was developed in response to the need to have a standardized system for classifying the severity of movement disability among children with CP (Palisano et al. 1997). Previous descriptive systems had included three levels, such as: (1) *mild, moderate,* or *severe*; or four levels such as (2) *non-ambulatory or physiological, household* and *community walkers* (Hoffer 1973); and (3) the Evans system: *not walking, restricting lifestyle, functional but not fluent*, or *walks fluently* (Evans and Alberman 1985). A five level description of children's ambulatory ability was reported by Hutton et al. in their study of factors affecting life expectancy, though they collapsed the data into only two categories of 'walking' and 'not walking' for their analyses (Hutton et al. 1994). However, there was no evaluation of the validity and reliability of any of these systems until the development of the GMFCS.

Palisano and his colleagues used the underlying construct of self-initiated functional abilities in sitting and walking and the need for assistive devices, such as walkers or wheelchairs, to develop the GMFCS and systematically tested its validity and reliability (Palisano et al. 1997, Wood and Rosenbaum 2000). The GMFCS describes movement ability of children with CP in one of five ordinal levels. The GMFCS currently includes descriptions of children's abilities for each level across four age bands: less than 2 years, 2 to 4 years, 4 to 6 years, and 6 to 12 years, with an adolescent age band currently under development. Children in Level I can perform all the activities of their age-matched peers, albeit with some difficulty with speed, balance, and coordination; children in Level V have difficulty controlling their head and trunk posture in most positions and

achieving any voluntary control of movement. The GMFCS has now become the principal way to describe the severity of motor disability for children with CP. The system has had good uptake internationally and across the spectrum of health care professions for use in research and clinical practice by providing a system for clearly communicating about children's gross motor function (Morris and Bartlett 2004).

From 2000

Following a survey of practice across the continent, the group for the Surveillance of Cerebral Palsy in Europe (SCPE) published their standardized procedures for ascertaining and describing children with CP for registers and databases (SCPE 2000). The definition was largely a reiteration of that proposed by Mutch and colleagues (Mutch et al. 1992) and included five key points. CP is: (1) an umbrella term; (2) is permanent but not unchanging; (3) involves a disorder of movement and/or posture and of motor function; (4) is due to a non-progressive interference, lesion, or abnormality; and (5) the interference, lesion, or abnormality is in the immature brain.

The system adopted by SCPE provides a decision flow chart to aid classification into neurological and topographical categories including spastic (unilateral or bilateral), ataxic, dyskinetic (dystonic or choreo-athetotic), or not classifiable. Clearly defined symptoms and requirements are provided for each neurological category. Despite careful planning of the system, there has been little work to demonstrate the validity and reliability of classification. The lack of any defined criteria for recording functional limitations in the SCPE definition was noted by Lenski et al. (2001). Subsequently, SCPE, along with other research groups, demonstrated that the inclusion of a description of functional ability markedly improved the reliability of diagnosing children with CP (Paneth et al. 2003). Consistent application of the diagnosis is of paramount importance when the prevalence of CP from different sources and places is being compared.

There has also been further progress in classifying children's motor abilities. The Manual Ability Classification System (MACS) now provides a method analogous to the GMFCS for classifying the ability of children with CP to handle objects (Eliasson et al. 2006). The Functional Mobility Scale (FMS) has been devised as an evaluative system to measure changes in walking ability, such as might be seen following intervention (Graham 2004). The FMS enables a child's performance over three distances (5, 50, and 500 metres) to be classified by their need for assistive devices such as a wheelchair or walking aid. In contrast to the GMFCS, where a child's level would not be expected to change, significant changes in FMS levels have been observed following orthopaedic surgery. This joins the battery of outcome measures to evaluate treatment for children with CP such as the Gross Motor Function Measure (Russell et al. 2003).

With rapidly improving imaging technology there is renewed interest in aetiological classification systems correlating clinical syndromes and neuroanatomy, challenging Freud's 100-year-old statement that this task was futile. Progress has been made using ultrasound and magnetic resonance imaging (MRI) to detect structural impairments of the brain before they manifest as movement disorders (Accardo et al. 2004). MRI can also be used to approximate the timing at which the brain was damaged, based on normal neurodevelopmental stages (Barkovich 2002, Krägeloh-Mann 2004). Only partially explained to date, Krägeloh-Mann (2004) summarizes some of the correlations that are emerging between the timing and location of the lesion and functional, cognitive, and sensory impairments.

The search for a single internationally accepted definition of CP continues. Another international multidisciplinary group met in 2004 and some of those participants then revised the oft-cited definition by Bax (1964) to recognize that the key motor deficit is often accompanied by other neurodevelopmental impairments. Their new definition is:

> Cerebral palsy describes a group of permanent disorders of the development of movement and posture, causing activity limitation, that are attributed to non-progressive disturbances that occurred in the developing fetal or infant brain. The motor disorders of CP are often accompanied by disturbances of sensation, perception, cognition, communication, behaviour, by epilepsy and by secondary musculoskeletal problems. (Modified after Bax et al. 2005)

Whilst welcoming the debate and the desire for consensus, the new definition received mixed reviews in the accompanying editorials. Carr (2005) described how the proposed definition and classification would affect clinical practice and the challenge of shifting from traditional modes of thinking; Blair and Love (2005) considered the precision of the definition to be flawed in the same way as previous attempts, particularly from an epidemiological perspective. Chiefly, they point out that the term 'non-progressive' was no more clearly defined than before, neither were the age limits and lower limit of severity for inclusion, or what syndromes should specifically be excluded. However, Blair and Love did not themselves provide any suggestions of how to address these issues. Whilst the precision with which the definition is applied by clinicians may have negligible consequences for treatment, the implications for measuring rates of CP over time are more profound.

So, in summary, after more than 150 years of debate we do not yet have a universally accepted definition of CP; nor do we have an agreed method for classifying the impairment that has been shown to be robust in terms of validity and reliability. It would be ungracious, however, not to pay a respectful tribute to those illustrious and often remarkable people who have all in their own way strived to further the scientific study of CP. In contrast, there has been more progress in classifying children's movement and manual abilities as these are probably easier to observe and categorize. The GMFCS has been adopted widely to classify movement ability and perhaps demonstrates that testing the fundamental properties of the validity and reliability of classification systems vastly enhances their credibility. To move the scientific study of CP forward we now need to examine how well the recent definitions and classifications proposed by SCPE and Bax's group actually perform in practice.

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A report: the definition and classification of cerebral palsy April 2006

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For a variety of reasons, the definition and the classification of cerebral palsy (CP) need to be reconsidered. Modern brain imaging techniques have shed new light on the nature of the underlying brain injury and studies on the neurobiology of and pathology associated with brain development have further explored etiologic mechanisms. It is now recognized that assessing the extent of activity restriction is part of CP evaluation and that people without activity restriction should not be included in the CP rubric. Also, previous definitions have not given sufficient prominence to the non-motor neurodevelopmental disabilities of performance and behaviour that commonly accompany CP, nor to the progression of musculoskeletal difficulties that often occurs with advancing age. In order to explore this information, pertinent material was reviewed on July 11–13, 2004 at an international workshop in Bethesda, MD (USA) organized by an Executive Committee and participated in by selected leaders in the preclinical and clinical sciences. At the workshop, it was agreed that the concept 'cerebral palsy' should be retained. Suggestions were made about the content of a revised definition and classification of CP that would meet the needs of clinicians, investigators, health officials, families and the public and would provide a common language for improved communication. Panels organized by the Executive Committee used this information and additional comments from the international community to generate a report on the Definition and Classification of Cerebral Palsy, April 2006. The Executive Committee presents this report with the intent of providing a common conceptualization of CP for use by a broad international audience.

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 one hundred years. At the end of the 19th century, Sigmund Freud and Sir William Osler both began to contribute 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 United States, and Mac Keith, Polani, Bax and Ingram of the Little Club in the United Kingdom, 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 'cerebral palsy', as documented by the number of attempts that have been made over the years. For example, Mac Keith and Polani (1959) 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, Bax reported and annotated a definition of CP suggested by an international working group that has become a classic and is still used. It stated that CP is 'a disorder of movement and posture due to a defect or lesion of the immature brain.' Though this brief sentence is usually all that is cited by authors, additional comments were added by Bax: 'For practical 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. At that time, it was felt that it was wiser 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. Not formally included in the concept were sensory, cognitive, behavioral and other associated impairments very prevalent in people with 'disordered movement and posture due to a defect or lesion of the immature brain', and often significantly disabling.

The heterogeneity of disorders covered by the term CP, as well as advances in understanding of development in infants with early brain damage, led Mutch and colleagues to modify the definition of CP in 1992 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 continued to emphasize the motor impairment and acknowledged its variability, previously underscored in the MacKeith and Polani definition; it also excluded progressive disease, a point introduced in Bax's annotation.

In response to the emerging need to evaluate the status of information about cerebral palsy and revisit the language presently used to describe it, an International Workshop on Definition and Classification of Cerebral Palsy was held in Bethesda, Maryland (USA), on July 11–13 2004, co-sponsored by United Cerebral Palsy Research and Educational Foundation in the USA and the Castang Foundation in the United Kingdom: support was provided by the National Institutes of Health/ National Institute of Neurological Disorders and Stroke and the Dana Foundation. The task of the participants (listing follows) was to revisit and update the definition and classification of cerebral palsy in light of emerging understanding of developmental neurobiology and changing concepts about impairments, functional status and personal '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 nonprogressive 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 Workshop participants 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 often present impairments of a wide range of functions that may or may not include severe motor manifestations, thereby calling for the need of an individualized, multidimensional approach to each affected person's functional status and needs. However, it was suggested that **the concept 'cerebral palsy' be retained** to serve diagnostic, management, epidemiologic, public heath, and research purposes. It was felt that an updated definition of CP, taking into account recent advances in the understanding of the physiology of and pathology associated with brain development, as well as changes in terminology, should be developed for international use. The updated definition needed to meet the requirements associated with these purposes, as well as to enhance communication among clinicians, scientists and the public. As in the prior concept, it was agreed that **the motor disorder needed to be emphasized; however, recognition should be provided that other developmental disorders of performance and behaviour can and often do accompany it**. This emphasis on the motor disorder is stipulated in that children with CP most often present for medical attention because of motor abnormalities, even if they have other developmental problems.

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, classes of disorders commonly accompanying CP have been identified and included in the revised definition. This addition reflects the concept that CP is one group of neurodevelopmental disorders which involve numerous developing functions. As in other neurodevelopmental disorders, various manifestations of the disordered brain may appear more significant in different persons or at different life periods, e.g. some aspects of the motor impairment, sensory loss, intellectual disability, attentional difficulty, epilepsy, musculoskeletal dysfunction and many others may be more prominent or more problematic at different stages of the life of a person with CP.

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What follows is: The Definition and Classification of Cerebral Palsy, April 2006, an annotated explanation of the terms used, and the thinking behind the choice of those words. This material was authored by the members of the Executive Committee functioning in panels enriched with expertise from consultants and by comments and suggestions from many reviewers responding to drafts provided to the international community. The Definition and Classification of Cerebral Palsy, April 2006 document is offered for international consensus and adoption, with the intent of providing a broad spectrum of audiences with a common conceptualization about cerebral palsy.

I. Definition of cerebral palsy

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

ANNOTATION

Cerebral palsy (CP)[1] describes a group[2] of permanent[3] disorders[4] of the development[5] of movement and posture[6] causing[7] activity limitation,[8] that are attributed to[9] non-progressive[10] disturbances[11] that occurred in the developing fetal or infant[12] brain.[13] The motor disorders of cerebral palsy are often accompanied by[14] disturbances of sensation,[15] perception[16], cognition,[17] communication, and behaviour, by epilepsy[20], and by secondary musculoskeletal problems.[21]

COMMENTARY ON THE TERMS AND CONCEPTS

It is hoped this annotation of the 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.

1. 'Cerebral palsy (CP)' – It is generally agreed that the CP concept, essentially a clinical formulation based on phenomenology, remains useful in the current state of nosology, insofar

as the term describes a prevalent, clinically important and identifiable group of persons with neurodevelopmental disabilities. Although the word 'palsy' has become largely obsolete in medical nosography and has no univocal connotation, the term 'cerebral palsy' is established in the literature and is used universally by clinicians, therapists, epidemiologists, researchers, policy makers, health care funding organization and lay persons. The term 'CP' has, however, been variably used, with poor comparability across different places and times, indicating the need for an internationally acceptable definition. The term cerebral palsy (CP) has been retained to relate future research in CP to existing published work.

The following explanations are offered to clarify several aspects of the definition of CP:

2. 'a group' – There is general agreement that CP is a heterogeneous condition in terms of aetiology 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').

3. 'permanent' – This definition excludes transient disorders, but recognizes that children and adults have changing patterns of clinical manifestations.

4. 'disorders' – This refers to conditions in which there is disruption of the usual orderly processes of child development.

5. 'development' – The notion of alteration in children's early development is essential to the CP concept. It distinguishes CP from phenotypically similar disorders in children due to later-acquired lesions, at a time when basic motor development is relatively well established. 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, as well as 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 children eventually diagnosed with CP begin to manifest very early in child development, usually before 18 months of age, with delayed or aberrant motor progress; other neurodevelopmental and functional difficulties that often accompany the motor signs can appear throughout childhood or later. The clinical picture of CP evolves with time, development, learning, activities, therapies, ageing, and other factors.

6. 'movement and posture' – Abnormal gross and fine motor functioning and organization (reflecting abnormal motor control) are the core features of CP. These motor problems can lead to difficulties with walking, feeding and swallowing, coordinated eye movements, articulation of speech, and secondary problems with behaviour, musculoskeletal function, and participation in society. **However, people with neurodevelopmental disabilities that do not primarily affect movement and posture are not considered to have CP.**

7. '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.

8. 'activity limitation' – The World Health Organization's (WHO) International Classification of Functioning, Disability and Health 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 WHO concept of 'disability' to recognize changing international concepts and terminology.

9. 'attributed to' – Understanding of developmental neurobiology (including genetic, biochemical, 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 cerebral palsy remains elusive.

10. 'non-progressive' – The term non-progressive is used to denote that the pathophysiological mechanisms leading to CP are presumed to arise from a single, *inciting event or* discrete *series of events* which are no longer active at the time of diagnosis. This inciting event(s) produce(s) a disruption of normal brain structure and function which may be associated with changing or additional manifestations over time when superimposed on developmental processes. **Motor dysfunction which results from recognized progressive brain disorders is not considered CP**.

11. 'disturbances' – This term refers to processes or events that in some way interrupt, damage or otherwise influence the expected patterns of brain formation, development and 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.

12. 'fetal or infant'– The specification 'fetal or infant' reflects the idea that disturbances that occur very early in human biological development impact differently on the development of motor function than disturbances that occur later, even those that occur in early childhood. There is no explicit upper age limit specified, although the first two or three years of life are most important in the timing of disturbances resulting in CP. In practical terms, disturbance resulting in CP is presumed to occur before the affected function has developed (e.g. walking, manipulation, etc.).

13. 'brain' – The term 'brain' includes the cerebrum, the cerebellum and the brain stem. It excludes motor disorders solely of spinal, peripheral nerve, muscular or mechanical origin.

14. 'accompanied by' – In addition to the disorder of movement and posture, people with CP often show other neurodevelopmental disorders or impairments.

15. 'sensation' – Vision, hearing and other sensory modalities may be affected, 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.

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. 'cognition' – Both global and specific cognitive processes may be affected, including attention, 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. **A child who has severely impaired cognition and no motor signs**

(except perhaps for some degree of hypotonicity) is not included within the concept of CP.

18. 'communication' – Expressive and/or receptive communication and/or social interaction skills may be affected, 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.

19. 'behaviour' – This includes psychiatric or behavioural problems such as autistic spectrum disorders, ADHD, sleep disturbances, mood disorders and anxiety disorders.

20. 'epilepsy'– Virtually every seizure type and many epileptic syndromes may be seen in persons with CP.

21. 'secondary musculoskeletal problems' – People with CP may develop a variety of musculoskeletal problems, such as muscle/tendon contractures, bony torsion, hip displacement, spinal deformity. Many of these problems develop throughout life and are related to physical growth, muscle spasticity, ageing and other factors.

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II. Classification of cerebral palsy

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

The above definition of cerebral palsy covers a wide range of clinical presentations and degrees of activity limitation. It is therefore useful to further categorize individuals with CP into classes or groups. The purposes of classification include:

1. Description: providing a 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 healthcare 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 (e.g., hemiplegia, diplegia) with an added modifier describing the predominant type of tone or movement abnormality (e.g., spastic, dyskinetic). However, it has become apparent that additional characteristics must be taken account of for a classification scheme to contribute substantively to the understanding and management of this disorder.

INFORMATION REQUIRED FOR CLASSIFICATION

The information available to provide 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 accompanying impairments, such as cognitive deficits, is age-dependent, and in young children the type of motor disorder may be hard to characterize. Some young children diagnosed as having CP may in fact have as yet undiagnosed neurological disorders that are very slowly progressive. While progressive disorders are not included in the CP rubric by definition, a period of observation that includes serial examinations of the child may at times be needed before their exclusion can be assured.

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, it may not be possible to completely exclude progressive disorders and underlying pathology, as described by neuroimaging and other laboratory findings, **However, all classification documentation should include the age of the child, the nature of the information available from clinical history (e.g. whether from clinical notes, maternal recall or period of observation of the child), and the extent to which metabolic and neuroimaging investigation 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, while determining service delivery needs will require consideration of accompanying impairments.

No classification system is useful unless it is reliable. Thus it is 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. Providing such definitions is, however, beyond the scope of this document. For example, the term 'spastic diplegia' is problematic because its existing definitions are variable and imprecise, and because evidence is lacking 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, while 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 since they perform very different functions. **Discontinuation of the term 'spastic diplegia' is recommended;** however, if the term is used, the user should define exactly what is meant, and what characteristics the term describes.

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, hypotonia) as well as the diagnosed movement disorders present, such as spasticity, ataxia, dystonia, athetosis.
   B. FUNCTIONAL MOTOR ABILITIES: The extent to which the individual is limited in his or her motor function, including oromotor and speech function.
2. Accompanying impairments
   The presence or absence of later-developing musculoskeletal problems and/or accompanying non-motor neurodevelopmental or sensory problems, such as seizures, hearing or vision impairments, or attentional, behavioral, communicative and/or cognitive deficits, and the extent to which impairments interact in individuals with cerebral palsy.
3. Anatomical and neuro-imaging findings
   A. ANATOMIC DISTRIBUTION: The parts of the body (limbs, trunk, bulbar region, etc.) affected by motor impairments or limitations.
   B. NEURO-IMAGING FINDINGS: The neuroanatomic findings on CT or MRI 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 post-natal CP (e.g. meningitis, head injury) or when brain malformations are present, and the presumed time frame during which the injury occurred, if known.

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 neuro-imaging and clinical features of cerebral palsy. These advances will continue to improve our ability to classify children and adults with cerebral palsy more accurately. **For classification of CP, use of the four major dimensions of classification listed in Table I is recommended.** Each is elaborated upon in the text that follows.

1. Motor abnormalities
1.A. NATURE AND TYPOLOGY OF THE MOTOR DISORDER
The type of abnormal 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 etiologic circumstances, as in kernicterus. Individuals with cerebral palsy 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),[i] 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 etiologic utility, as recommended by the 2001 NINDS workshop on childhood hypertonia.[ii] **It is recommended 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. For a recent review of the terminology of motor disorders, see Sanger et al.[iii,iv]

1.B. FUNCTIONAL MOTOR ABILITIES
The WHO International Classification of Functioning, Disability and Health (ICF),[v] along with several other recent publications, have 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 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.[vi] A parallel classification scale, the Bimanual Fine Motor Function Scale, or BFMF, has been developed for assessing upper extremity function in cerebral palsy, but has not been as extensively studied as the GMFCS.[vii] A newer instrument for assessing hand and arm function – the Manual Ability Classification System or MACS – has been shown to have good inter-rater reliability between parents and professionals, and will shortly be published.[viii] Concurring with SCPE, it is recommended that a functional classification system be applied to hand and arm function in children with CP. Bulbar and oromotor difficulties are common in cerebral palsy and can produce important activity limitation, but there is as yet no activity limitation scale for such functions. A high research priority is the development of a scale for speech and pharyngeal activity limitation in cerebral palsy. In the meantime, the presence and severity of bulbar and oromotor involvement should be recorded.

While 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, evaluation of participation restriction (formerly termed "handicap") in CP is not well developed, and reliable categorization of children based on this aspect of daily life is therefore not yet possible.

2. Accompanying impairments

In many individuals with cerebral palsy, 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 cerebral palsy. These impairments may have resulted from the same or similar pathophysiologic 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, emotional and behavioral issues, and later-developing musculoskeletal problems. These impairments should be classified as present or absent, and 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. In concurrence with the SCPE recommendation, **the presence or absence of epilepsy (defined as two or more afebrile, non-neonatal seizures) be recorded, and IQ, hearing and vision be assessed.** While SCPE provides terminology for describing different degrees of cognitive, hearing and visual impairment, **the IQ score, corrected vision in each eye, and decibel loss (if any) in each ear be recorded** 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, mild mental retardation*) have come to be generally accepted.

3. Anatomical and neuro-imaging findings

3A. ANATOMIC DISTRIBUTION

The pattern and extent of the motor disorder in CP with respect to different anatomical areas should be specified. Previous classification schemes included only the extremities and required a subjective comparison of severity in the arms and the legs. The inherent validity of making this comparison has been questioned since the arms and legs are so structurally and functionally diverse. Notably missing from current anatomical classification schemes is description of truncal and bulbar involvement. **All body regions – trunk, each limb, and oropharynx – need to be described individually in terms of any impairments of movement or posture.** A scale for describing truncal posture in cerebral palsy has recently been developed.[ix] Separate objective classification schemes have also been developed for the upper and lower extremities.

It is acknowledged that the terms "diplegia" and "quadriplegia" have been extensively used for determining the anatomic distribution of the motor disorder and have become firmly entrenched in research and clinical practice, The severity of involvement in the arms (ranging from 'none' to 'less that that of the legs') has been used as the main characteristic for making this distinction which is problematic as stated above. Gorter et al. have documented the imprecision of these terms in clinical practice.[x] **It is recommended that the terms diplegia and quadriplegia not be used until more precise terminology evolves and gains similar acceptance.**

*UK usage: learning disabilities.

Those who continue to use these terms should define exactly what is meant by them and the characteristics the terms describe.

A promising alternative approach that has been recommended, and which is being utilized currently by the SCPE, is the differentiation of unilateral versus bilateral motor involvement. Categorization based on this distinction has shown good reliability (SCPE manual[1]). Even this distinction can still be blurred since 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. This distinction should be considered as part of a multiaxial classification scheme, thus it should be coupled with a description of the motor disorder and functional motor classification in both upper and lower extremities.

3.B. NEURO-IMAGING FINDINGS

Until recently, correlations between neuroimaging findings and clinical presentation in cerebral palsy were weak. However, advances both in imaging technology and in quantitative motor assessments are changing this picture. The goal of categorizing all patients based on specific neuroimaging findings will require more development before implementation. **The recommendation of the American Academy of Neurology to obtain neuroimaging findings on all children with cerebral palsy should be followed whenever feasible.**[xi] At present, information is insufficient to recommend any specific classification scheme for neuroimaging findings.

4. Cause and timing

It is increasingly apparent that cerebral palsy may result from the interaction of multiple risk factors, and in many cases, no identifiable cause may be found. Therefore, while 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, a more salient method of classification may be developed. For the present, **timing of insult should only be noted 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 post-natal meningitis in a previously well infant. While recording adverse events in the prenatal, perinatal and post-natal life of a child with CP is necessary, clinicians 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.

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Workshop Presentations

Classification of cerebral palsy: paediatric perspective

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Epidemiologists and clinicians should use the same broad definition and classification of cerebral palsy (CP). As clinicians make the diagnosis, they should lead on classification but the epidemiologist must temper such ideas so they are applicable to the whole population, not just a subgroup which present to one type of paediatrician or neurologist. Where clinicians want detailed sub-classification, this also needs epidemiological advice if we are to be confident that different clinicians, when using the same word, mean the same thing.

A classification must be agreed, precise, and reliable. From 1990 in Northern England, the functional difficulties and type of CP a child has have been recorded by us on a population-based register of children with CP. We realized that we were using the conventional CP classification terms differently. I shall illustrate this with respect to the term 'diplegia'. For some clinicians, diplegia meant there should be no functional loss in upper limbs. For others, functional loss in upper limbs was allowed but severe 4-limb involvement with mental retardation could not be diplegia, even if lower limbs were more involved than upper limbs. For more still, truncal control was a defining feature. We undertook a literature review and realized that definitions have changed over the decades, some being descriptive, some syndromic; and that there were large variations between countries in their propor-tions of diplegia, suggesting the term was used differently (Table I). Also, there was no overall association between preterm birth and diplegia; in some countries there was an association but one wondered if being preterm in say Sweden meant that the label diplegia was more likely to be used because of their belief in a characteristic syndrome of preterm birth and diplegia – the association therefore being self-fulfilling.

The issue of severity is important in studying trends in rates of CP. We need to be confident that cases are being counted in the same way in different years and in different places. In any non-deteriorating condition there may be mild cases that are never ascertained or where there is diagnostic uncertainty. These could yield larger differences in apparent rates over time or between places than any change in underlying rate. For instance, in CP there may be diagnostic uncertainty between mild CP and clumsiness or children with very mild hemiplegia may never present or may present at a much older age and would not be counted by a register ascertaining up to 5 years of age. It would be more reliable to compare rates of CP over time or between places by functional severity of Gross Motor Functional Classification System (GMFCS) of level II or worse, for example, than just counting cases of CP.

How should we represent and measure 'severity'? Although felt tone or briskness of reflexes are essential for diagnosis, they cannot be used for severity grading as they are subjective and vary considerably at different times or in different settings. What does 'lower limb more involved than upper limb' actually mean when upper and lower limbs are responsible for different functions? A simple classification of spastic CP determined by the limb's functionally involved is attractive because it will not make assumptions about cause or constellations of features that are so prone to subjectivity. To this should be added a functional severity grading and the GMFCS and Bi-Manual Fine Motor Function have been developed for lower and upper limb function respectively.

Table I: Proportions of cerebral palsy subtypes

Study	Duration	Number of cases of CP	Number of spastic cases of CP	Spastic cases as percentage of all cases	Unilateral spastic as percentage of spastic cases	Diplegia as percentage of spastic cases	Bilateral spastic as percentage of spastic cases	Diplegia as percentage of bilateral spastic cases
North Italy	1980–89	–	–	–	35	45	65	69
Denmark	1979–90	908	734	81	23	62	77	81
North England	1991–96	537	499	93	37	23	63	36
Northeast England	1980–96	380	355	93	42	23	58	40
Avon, England	1979–88	237	196	83	39	39	61	63
Mersey, England	1984–89	497	460	93	39	23	61	38
Atlanta, USA	1985–87	204	166	81	34	19	66	28
Sweden	1979–90	545	470	86	40	51	60	84
England & Scotland	1984–89	1649	1334	81	35	22	65	33
Slovenia	1981–90	768	651	85	33	43	67	64
Western Australia	1980–94	819	639	78	45	37	55	68
Rome, Italy	1977–96	282	213	76	33	27	67	40
Northern Ireland	1977–92	960	572	87	43	21	57	36
Norway	1980–89	46	39	85	49	33	51	65
Oxford, England	1984–95	967	806	83	40	–	60	–
Averaged percentage	–	–	–	85	38	33	62	53
Range	–	–	–	76–93	23–49	19–62	51–77	28–84

In respect of therapy, function has long been the goal but here there is also a need for a change in outlook. Physiotherapy, botulinum toxin, and surgery have not brought the substantial improvements that had been hoped. Indeed what do we mean by 'useful' or 'significant improvement'? Just as physicians use the term 'diplegia' in different ways, physicians, surgeons, therapists, teachers, children, and parents also use terms such as 'useful' in different ways, with different perspectives and no common language. Botulinum may make a limb less stiff but should reduced stiffness be a primary aim? How does one balance the effort to improve the function of a limb against the improvement in lifestyle achievable by providing a wheelchair or ensuring all parts of a school are on the ground floor or have lift access? The International Classification of Function, Disability and Health (ICF) provides the necessary conceptual framework to explore such issues. There are four 'components' to the classification: Body structure and function, Activity, Participation, and Environmental factors. It defines 'Participation' as involvement in life situations. This concept applies to all people, not just those with disabilities. It has positive, rather than negative, connotations and the difficulties are understood to reside in the interaction between the individual and their environment and not in the individual alone. The ICF recognizes that improvement may be achieved through manipulation of a child's environment and therapy requiring a change in the child's body. Therefore, the classification is in agreement with the social model of disability. We need to establish the level at which each child is participating and in which areas they would most like to see improvement. The ICF also recognizes the importance of Quality of Life (QoL), a person's subjective account of how they feel about their life, including their view of their own Participation. There are now instruments such as KIDSCREEN, KINDL, TACQOL, and PEDSQL which are capable of capturing this subjective QoL in childhood.

So a classification of children with CP should have:

– The CP type
– Associated impairments
– The functional effect across trunk and limbs
– The child's participation
– The child's quality of life.

Classification of cerebral palsy: clinical therapist's perspective

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Therapists are not responsible for diagnosing cerebral palsy (CP); rather they are most concerned with recommending or providing the best treatment for individual patients and their families from the standpoint of motor functioning and participation. Therefore, the challenge posed to the group on the behalf of therapists was this: to develop a more accurate and informative diagnosis (better definition) and classification of the motor disability in CP that would ultimately lead to improved treatment prescriptions and functional outcomes.

Physical therapy has been at the forefront of the management of CP for several decades. The present day clinical reality is that most children with non-specific, non-progressive motor disabilities qualify for and receive at least some thera-

py services. However, a 'one-size fits all' approach belies the tremendous heterogeneity in this population in terms of clinical presentations and levels of disability. Changes in definition and classification are likely to not only affect what types of services are delivered, but also who receives services, how many services they may receive, how frequently, and for how long. In this time of increasing accountability and limited resources, it is important to everyone that therapy services are more judiciously allocated, and that those delivered are more efficient and effective.

From a therapist's perspective, the term 'cerebral palsy' and its definition as a disorder of motor coordination resulting from an injury to the developing brain are too vague and inclusive. It is disconcerting as a non-diagnosing clinician to find that, in some cases, the diagnosis of CP may be uncertain (e.g. called CP because no other cause identified), unjustifiably delayed which may delay services, or even perhaps incorrect (e.g. no evidence of brain injury). Since the diagnosis is directly related to prognosis, a more timely and accurate diagnosis would lead to earlier development and implementation of more realistic treatment plans. Most existing definitions also fail to acknowledge the very clinically pertinent fact that CP is not a single disease with a clear etiology, such as Down syndrome and Duchenne muscular dystrophy, but is instead a collection of often diverse movement disorders, each of which may require different intervention strategies. This reality underscores the importance of developing a classification system that differentiates these disorders more accurately for both clinical and research purposes.

Also with respect to classification, a therapist's perspective is that it would be most beneficial if patient groupings were more closely linked to treatment paradigms and ultimately to outcomes. Existing classification schemes are helpful but inadequate. For example, the anatomical classification potentially indicates where to focus treatment efforts. The physiological classification of the tone disorder can also be informative for treatment decisions, but begins to break down if the identified tone abnormality is not contributing to the motor disability or if mixed hypertonia exists. A more recently established classification scheme, the Gross Motor Function Classification System (GMFCS)[1] was a major step forward for the field, perhaps especially for physical therapists since it focuses primarily on differentiating children with CP based on functional mobility irrespective of the type or distribution of the motor disorder. The GMFCS also predicts future mobility and thereby facilitates more realistic goal setting with therapists and families. It is important to note that since GMFCS levels are based on mobility and not physiology, knowing the level alone is insufficient for making specific treatment recommendations. Several biomechanical classifications have also been published in CP whereby patients are grouped by motor patterns in an explicit attempt to link these to treatment.[2,3] The assumption that similar motor patterns may have similar etiologies and responses to treatments is arguable but plausible and may have considerable relevance to physical therapy practice, but more research is needed to validate and refine these.

In summary, this therapist's perspective is that definitions and classifications of CP should be refined to reflect the rapidly accumulating new knowledge in the areas of brain imaging, neurophysiology, biomechanics, outcomes research, and views on disability and health. More precise diagnoses

and links from pathology to patient's priorities for treatment need to be established. While motor disability is the hallmark of CP, therapists are well aware that other concurrent conditions can confound or even eclipse the motor disorder and must be included in these schemes. Finally, a futuristic hope is that progressive refinement of definitions and classifications will pave the way for new thinking in the treatment of childhood brain injury. Greater understanding of the brain disorder and its capacity for development and recovery are critical for any chance of restorative therapies or potential cures.

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'Cerebral palsy' – rejected, refined, recovered

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The 2004 Bethesda Conference on the Definition and Classification of Cerebral Palsy (CP) was clearly a milestone in CP research administration in that it not only gathered a large number of clinicians and researchers to discuss CP issues, but also sparked quite some discussion afterwards, both on the Castang Foundation website (www.castangfoundation.net/workshops_washington_public.asp) and in these pages. As one of the participants who did not make a formal presentation at the meeting (OD) and one who was invited but could not attend (KK), we are grateful for the opportunity to offer our thoughts in this essay.

In what follows, we discuss some of the most difficult issues in CP definition and classification, which make us believe that one remarkable result of the meeting is that we still have the term 'CP'.

'CP' rejected: Part of the discussion on the definition of CP at the Bethesda conference revolved around the question, 'Is there one such thing as CP?' Yes, although some of us use the term 'CP' rather loosely in everyday developmental neuro-speak, we still need to define our terminology rather exactly.

In light of the considerable difficulties to agree on a simple and straightforward definition of CP, at the 2004 Bethesda meeting one of us light-heartedly suggested to just drop the term 'CP' altogether, while adding that this might be almost impossible given the longstanding tradition of the term. One might anticipate that what would happen to CP would be just like what happened to the American pop artist who in 1994

officially abandoned his name and was subsequently called 'The artist formerly known as Prince'.

Thus, we feel (seriously) reluctant to give up the term entirely, although we clearly subscribe to the view that CP is <u>not</u> one disease[1]. Still, we would like to offer the following thoughts how the concept of CP could be refined.

'CP' refined: The etymology of the term 'CP' is much more straightforward than the current discussion about the concept CP would make one think. 'Cerebral' stands for 'related to the cerebrum', i.e., the larger one of the two brains we have. 'Palsy' is an abbreviation for 'paralysis', which in turn refers to impaired motor function. Thus, the actual meaning of the term 'CP' is rather descriptive, general, and overarching – maybe a bit too descriptive, general, and overarching for some.

Indeed, the oft-quoted Martian arriving on earth being confronted with this meaning of CP might be prepared to use it for anyone with a brain-related movement disorder or even the exceptionally clumsy child or the child with extremely lax ligaments. Better descriptors are needed, such as those that indicate topography and character of the motor disability, and conditioners, such as 'non-progressive'.

Assuming that the term 'CP' will continue being used, is there a way to make the **definition** (and, thereby, the diagnosis) more homogeneous and more meaningful? If CP is purely based on characteristics of the motor impairment, regardless of when the impairment occurred (prenatal, postnatal, and childhood), a broad diversity of underlying etiologies will render CP a 'grab bag' of disorders.

Alternatively, we propose that the appellation 'CP' should be reserved for disorders that (1) affect the motor system and (2) are acquired prior to completion of the neonatal period. Post-neonatal acquired motor disability ought to be relegated to a non-CP category specific to a cause and an outcome (traumatic injury, cerebrovascular accident, meningitis, HIV, etc., leading to quadriplegia, hemiplegia, etc.).

We are aware that it may be rather difficult to establish that a particular case of CP 'occurred' prior to the end of the neonatal time period, particularly when the symptoms or signs that lead to the diagnosis may occur only weeks, months, or years later. However, the absence of a post-neonatal sentinel event, particularly with appropriate support from neuroimaging, should help make the proper diagnosis.

Keeping CP (of fetal/neonatal onset) and (post-neonatal) non-CP motor disability separate might not only help clinicians, but it might also help promote research into preventive strategies. As an overarching term for both, one might start using 'acquired developmental encephalopathy'.

We have three reasons to suggest that the current **classification** recommendations are likely to diminish descriptive precision, which will, in turn, lead to oversimplification of different CP forms, make it more difficult to compare research study populations, and make discussions with parents about prognosis and potential comorbidities more difficult.

Firstly, the classification system for CP offered in the April 2006 consensus paper recommends that the clinical description begins with detail about the character of the motor impairment (e.g. spastic), followed by a description of the severity, followed by a description of the comorbid and epiphenomological findings (e.g. musculoskeletal problems), and finally by a description of the parts of the body involved (topography). We think that the order of description should follow the order of clinical evaluation. Thus, we suggest that

the topography of the disability should be described first, not last. The qualitative aspect of the motor disorder should then be detailed, followed by a statement of severity and, finally, the presence of comorbidities and complications of the motor disorder (e.g. orthopedic-musculoskeletal problems).

Secondly, the issue of topography was de-emphasized by the panel with a recommendation that the term spastic diplegia (and quadriplegia) be dropped from the CP lexicon. The panel recommended that CP topographic description should be limited to a statement of either 2 or 4 extremity involvement. The neurologist relies, at least in part, on the gradation, severity, and symmetry of the topography in order to localize the lesion and infer structure-function relations. Severity of CP, as assessed by the Gross Motor Functional Classification System, is significantly associated with topography.[2] Moreover, there are ample data that enable the clinician to prognosticate about the risk of developing comorbid conditions and the likelihood of resolution of CP symptoms depending on whether lower extremities are affected more than upper extremities (diplegia), are affected to the same degree as upper extremities (quadriplegia), or if one upper extremity is more substantially involved than its lower counterpart (hemiplegia).[3] Why do away with a system that enhances the ability to prognosticate?

Lastly, the consensus panel recommends that the character of the motor impairment be described by only a single dominant type of tone or movement abnormality. This suggestion serves to perpetuate imprecision in CP description and diagnosis, and is likely to inhibit consideration of alternative diagnoses and treatment options. Co-occurrence of spasticity and dystonia is common, and when they co-occur, comorbidities are more severe, likelihood of normalization is reduced,[3] and there is diminished response to some treatments, such as dorsal rhizotomy.[4] Spasticity and dystonia are distinguishable, even when occurring in the same individual, and the presence of each informs about lesion localization, which, in turn, may help in understanding antecedents and pathophysiology.

Postscript: 'CP' recovered: Could we do without the term 'CP'? Decades of clinical and scientific work in the field have yielded a vast literature and experiential discourse about CP. We run 'CP clinics', perform 'CP research', and consider ourselves 'CP epidemiologists'. What would we do without the term 'CP' and what might be a better one that could serve as its replacement, in light of the longstanding history of concept and terminology?

In essence, we believe that replacement would leave us stuck with 'the disorder formerly known as CP'. When 'the artist formerly known as Prince' more recently returned to calling himself just 'Prince', some responded to the confusion by referring to him as 'the artist formerly known as the artist formerly known as Prince'. Both the concept of CP and the term 'CP' deserve a better fate.

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The brain imaging perspective

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The power of neuroimaging in revealing the cause of cerebral palsy (CP) is now well accepted. Imaging using various imaging modalities shows pathology in 77%, when computed tomography (CT) is used, and in 89% when magnetic resonance imaging (MRI) is employed.[1] Neuroradiology is capable of defining different kinds of brain pathology including various congenital malformations and different destructive lesions in white and grey matter.

The traditional way to define the timing of an insult and subsequent brain injury responsible for CP is to define an injury as being either pre-, peri-, or postnatal in origin. However, neuroradiology has demonstrated that the morphology of a certain lesion is dependent on the maturation of the brain at the time of the insult. Selective vulnerability in different parts of the brain during different stages of brain development is of greater importance in determining the brain pathology than the type of insult. Thus, it is more logical to relate a certain type of brain injury to the known time window during which this particular lesion is known to occur, than to relate the injury to the time of birth. Thus, it is possible that neuroradiological definition of a specific lesion and the time window during this lesion is known to occur, contradicts the clinical impression of cause and effect in CP. This may have important medical legal implications as well as being of importance in the treatment and rehabilitation of a child with CP.

Although timing of an insult is the most important factor in determining the pattern of pathology, the duration and severity of the insult are other important factors. Thus profound asphyxia causes lesions different from those due to partial hypoxia in the mature brain but also in the immature brain before 34 gestational weeks.

Timing and pathology: The finding of a congenital malformation by MR is usually indicative of an injury during the first half of the pregnancy. Detailed classification of the malformation may further limit the period during which the insult has operated. An abnormality of cleavage, e.g. holoprosencephaly is a very early lesion, 4th to 6th week,[2] while an abnormality of cortical organization, e.g. polymicrogyria is an example of a very late lesion which may occur as late as 20 gestational weeks or later, depending on specific type.[3]

Neuroradiological demonstration of primary white matter damage, e.g. periventricular leukomalacia (PVL) or periventricular haemorrhagic infarction (PVH) represents residual from insults operating between 24 and 34 gestational weeks. While the lower limit, 24 weeks, may be difficult to define, it appears as if the later limit 34 gestational weeks is unusually

well defined. It is not difficult to find statements in the literature, textbooks in particular, saying that PVL can occur even later than 34 gestational weeks. However, the scientific support for this opinion is weak and most reports refer to cases in which the findings were detected and the diagnosis of PVL made after a full-length pregnancy but without solid evidence when the damage indeed occurred. When found in a neonate born at term, PVL should be considered as having occurred in utero.

It has been proposed that PVH is more common in the more immature fetus or the very prematurely born neonate while PVL, particularly if posterior in location, is more common closer to 34 gestational weeks. Hence, anterior white matter damage should be a sign of an earlier insult than posterior damage. This hypothesis has been tested against another independent way to determine the level of maturity at the time of injury and found to probably be valid.[5]

When the brain reaches a maturity close to term, grey matter is more sensitive to injury than white matter. Two principally different patterns are recognized, damage to central grey matter structures and cortical damage. Bilateral symmetrical damage to thalami, posterior putamen, and Rolandic cortex has been described as being caused by profound asphyxia in the mature brain.[6] Diffuse damage to cortical structures is thought to be the result of partial hypoxia close to term.[7] Focal cortical damage, most often in the territory of the middle cerebral artery, is on the other hand thought to be related to hereditary or acquired thrombophilias and environmental factors.[8,9]

Motor dysfunction and associated disability: The diagnosis of CP always involves a motor deficit. However, some of the lesions found to be responsible for the motor deficit in CP may also cause other problems, either associated or in isolation without concurrent motor deficit. Thus, knowledge about the underlying brain lesion may define individuals with a certain kind of destructive brain lesion with associated handicap but without CP. This observation highlights the importance of considering expanding the concept of CP to not only include children with motor deficit but also include children with cognitive disability and no motor disability.

Systematic use of neuroimaging in populations at risk for developing CP have shown that children born preterm have neuroradiological findings of PVL in 32% while only 9% had CP.[10] Jacobson and her group have shown that children with PVL may have significant symptoms with visual cognitive defects from their brain injury even without motor deficits.[11,12] At the same time precise relationships between morphological lesions and motor disability has been shown using sophisticated analysis of conventional and functional MR.[13,14]

It is important to recognize the strength of using neuroimaging not only in establishing the lesion responsible for the motor dysfunction of CP but also to establish the precise relationship between morphology and function, being motor or more cognitive functions.[15] Neuroimaging will be of great importance when the concept of CP has to be expanded to include other kinds of disability than those of pure motor dysfunction.

While neonatal imaging is often difficult to interpret and may give false impressions about the final pathology, clinicians and epidemiologists must accept and integrate the objective information available from neuroimaging in late follow-up of CP. Such imaging represents 'in vivo pathology' and will, in most cases of CP, give clear information about the lesion behind CP and may also in some lesions demonstrate a clear relationship between pathology and functional deficit. Knowledge about underlying pathology is also vital for the therapist when choosing the most appropriate rehabilitation efforts and when discussing with parents the prognosis of therapeutic interventions

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Classification of cerebral palsy: neuropathologist's perspective

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Wallenberg probably first used the term 'cerebral palsy' (CP) in 1886 when he summarized clinical and anatomic aspects of 'Infantile Cerebral Palsy'. Thereafter, Lovett, Osler, and Freud used the term. The necessary underlying anatomical knowledge had been accumulating for many centuries, but was brought together in the 18th century after Malpighi

introduced the microscope to medicine in the 17th century. Early in the 19th century, many pathologists recognized the specific lesions underlying CP, but it wasn't until late in the 19th century that the association between clinical deficit and specific variety of CP was made.

Anatomical investigation of cerebral lesions resulting in diplegia, hemiplegia, and tetraplegia, with or without a movement disorder, didn't become serious until the first quarter of the 19th century when Cazauvielh and Cruveilhier separately recognized bulk lesions involving both gray and white matter. For instance, an overall small brain was associated with CP or cerebral hemiatrophy with contralateral hemiplegia. Cavitary lesions, such as single large cysts or numerous small cysts, were soon set apart.

In the middle of the 19th century, lobar sclerosis was recognized. Türck, Turner, and Cotard described anterograde degeneration in the crus, pons, and pyramids and crossed cerebellar atrophy. They also distinguished lobar sclerosis and lesions in the distributions of specific arterial beds. Little (1862) ascribed abnormalities in the upper half of the brain to neonatal asphyxia. Parrot, Moebius, Vivius, Herschfeld, Hlava, and Schmorl completed many studies of focal white matter necroses between 1862 and the end of the century. Herschfeld identified infection as an antecedent of focal necroses. In the last half of the century, Heschl and Kundrat used the name 'porencephaly' and distinguished between two varieties of pori: (1) those that cut across previously formed gyri; and (2) those in which gyri pointed in a radial fashion into the defect. The walls of some pori contained hemosiderin and a role for bleeding into the brain was identified. Sarah McNutt distinguished bilateral paracentral gyral atrophy. Orth described bilirubin staining of brain, but the anatomic details of kernicterus were not identified until Schmorl in 1903. Regions of multiple small cysts were labeled multicystic encephalomalacia. Freud separated neurological anomalies following preterm birth from those following birth at term. He said that preterm birth predisposes to paraplegic forms of cerebral diplegia three times more than general rigidity.

In the first half of the 20th century, cortical dysplasias (migration abnormalities, e.g. ectopias, heterotopias [nodular or laminar]), pachygyria, lissencephaly, polymicrogyria, sclerotic microgyria, borderzone lesions, basal ganglia status marmoratus, and thalamic sclerosis were added, as well as additional information concerning the residue of infarcts in specific arterial supply beds. Polymicrogyria in the walls of the second variety of porencephaly was recognized. Upper spinal cord lesions mimicking diplegia clinically were found. In the second half of the 20th century, a new class of lesions was differentiated. These lesions were predominantly located in hemispheral white matter with absent or minimal neuronal damage and consisted of widespread proliferation of astrocytes. Less prevalent are focal white matter necroses described a century earlier. Both sets of lesions are followed by hypoplasia of white matter. However, hypoplasia of white matter sometimes occurs without either of these abnormalities. Rubella, cytomegalovirus, herpes simplex, and toxoplasmosis left cavitary or other cerebral defects.

Table I: Classification of lesions associated with cerebral palsy

Upper spinal cord lesions
Bulk abnormalities:
  Microcephaly
  Hemimegalencephaly
  Lobar sclerosis
Acquired abnormalities of gray and white matter:
  Pori
  Multicystic encephalomalacia
  Embolic/thrombotic lesions in specific arterial distributions
  Borderzone lesions
  Girdle atrophy (paracentral cortex)
  Sclerotic microgyria (ulegyria)
Acquired abnormalities of white matter alone:
  Focal necroses
  White matter astrocytosis
  Hypoplastic white matter
  Delayed myelination
Malformations:
  Some cases of holoprosencephaly
  Nodular or laminar heterotopias
  Pachygyria
  Pori with gyri pointing to the defect
  Micropolygyria
  Lissencephaly

Table II: Central nervous system abnormalities in individuals with 'cerebral palsy'

Year	Investigator	
		'Cerebral Palsy'
1886	Wallenberg	Summarized clinical and anatomic aspects of 'infantile cerebral palsy' and used the term
1888	Lovett	Used term cerebral palsy
1888	Osler	Used term cerebral palsy
1893	Freud	'Infantile cerebral diplegia'
1897	Freud	History of cerebral palsy; emphasized prematurity in association with diplegia; Spent much of his book discussing lesions acquired during development and failure of development and developmental retardation, but apparently did not recognize malformations of the brain as we understand them today. Explained CP spasticity by pyramidal tract secondary atrophy in brainstem and cord
Anatomical Knowledge		
1700s	Malpighi	Introduced microscope to medicine
1717	Leeuwenhoek	Nerve fiber and axon
1761	Morgagni	Lesions of one hemisphere result in contralateral paralysis
1781	Fontana	Nerve fiber and axon
1799	Bichat	Brought histology to pathology
1809	Rolando	Cerebrum controls motor function
1810–19	Gall	Pyramid. Functional localization in brain;
1820s		Achromatic Compound Microscope introduced
1833	Ehrenberg	Microscopic structure of nerve cell and fiber
1836	Valentin	Nerve cell, its nucleus and nucleolus
1839	Schwann	Cell theory and serious use of microscope in neurologic disease
1849	Koelliker	Nerve fibers originate from nerve cells
1849	Waller	2nd degeneration

Year	Investigator	
1851	Türck	2nd anterograde atrophy in spinal cord after cerebral lesions
1856	Turner	2nd anterograde atrophy in crus, pons, and pyramids. Crossed cerebellar atrophy. Understood trophic dependence of thalamus upon its ipsilateral cerebral cortex
1875	Erb	Described quadriceps reflex; arrest of development of pyramid
1876	Raymond (Charcot trainee)	Special localization of motor function in internal capsule
1877	Flechsig	Delineated pyramidal tract in posterior limb of internal capsule
1887	His	Each nerve cell is a distinct unit
1891	Waldeyer	Neuron

Acquired defects: central nervous system cerebral gray and white matter

Year	Investigator	
1818	Abercrombie	Parturitional spinal cord injuries
1822	Pinel (the son)	Lobar sclerosis
1826	Denis	Hemorrhages in newborn brain caused by mechanical birth injuries
1827	Cazauvielh	Multiple small cysts; Two cases with larger cysts; decreased pallium
1829–37	Cruveilhier	Small brain. hemiatrophy of brain
1830	Delpech	Small Brain
1842	Henoch	Infantile cerebral atrophy
1853–70	Little	Three papers. Abnormalities of birth and neonatal asphyxia. Little recognized that the majority of stillborn infants, when resuscitated, recover unharmed
1855	Friedleben	65% of newborn autopsies contained cerebral lesions; Brain sclerosis and atrophy
1862	Parrot	Intracerebral hemorrhages; cervical spinal cord injury
1859–68	Heschl	Porencephaly; vascular affection in fetus
1868	Cotard	Partial atrophy of brain; diffuse lobar sclerosis; Lesions in specific arterial beds
1870	Parrot	Upper spinal cord lesions
1882	Ross	Lobar sclerosis results from arterial emboli
1884	Strümpell	Acute infantile hemiplegia 'Polioencephalitis'
1885	McNutt	Convolutional sclerosis; bilateral paracentral atrophy
1886	Wallenberg	Summarized anatomic aspects of 'Infantile Cerebral Palsy' and used term
1887	Abercrombie	Found embolus in artery leading to cerebral defect
1888	Lovett	60 cases of CP: embolism and parenchymal hematomas important
1888	Osler	50 cases of CP: partial or lobar sclerosis; paracentral in all; arterial occlusion
1888	Gowers	Birth palsies: arterial thrombosis and embolism
1896	Schultze	Parturitional cysts and scars in medulla and spinal cord
1899	Bresler	Ulegyria
1890	Sachs, B	Cerebral palsies of early life
1891	Freud	Choreatic form of CP
1893	Anton	Status marmoratus
1958	Crome	Multicystic encephalopathy

Year	Investigator	
Acquired defects: cerebral white matter only		
1850	Bednar	Focal cerebral necroses newborn brain are mostly deep white matter
1862	Parrot	Focal necroses in white matter – metabolic disorder
1865–7	Virchow	Focal necroses in white matter – infection
1904	Schmorl	Described 280 cases of focal necroses in white matter; summarized previous studies from 1862 (Parrot) – 1903 (Moebius, Vivius, Herschfeld, and Hlava)
1940	McClelland	White matter hypoplasia; (separately)
1945	Benda	Delayed myelination
1954	Crome	

Malformations of Brain

Year	Investigator	
1865–7	Virchow	Nodular heterotopias.
1868	Owen	Lissencephaly/pachygyria
1882	Kundrat	41 cases of porencephaly. Two varieties: 1) order of gyri undisturbed by defect; 2) Gyri radiate towards defect. Polymicrogyria. Results from parturitional circulatory disturbances. Described holoprosencephaly and coined term arhinencephaly
1885	Giacomini	Microcephaly
1899	Bresler	Polymicrogyria
1915	Bielschowsky	Polymicrogyria
1936	Jacob	Laminar and nodular heterotopias

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Classification of cerebral palsy: the surgeon's perspective

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Introduction: The Victorian Cerebral Palsy Cohort Study (VCPCS) follows the progress of children born in the Australian state of Victoria between 1990 and 1992. The birth cohort was identified using the Victorian Cerebral Palsy Register (VCPR) and is updated by information from hip surveillance clinics, the gait laboratory, and other sources. It has given the opportunity for a multidisciplinary team to study the prevalence of cerebral palsy (CP) in a defined population and to study topographical

distribution and motor types. Other key areas of interest are gross motor function, gait disorders, and musculoskeletal issues. A cohort study from Sweden based on the same birth years has been published, allowing interesting comparisons, as outlined below, and covered in more detail in the relevant publications.[1,2,3]

1.1 Classifications of Cerebral Palsy: Motor Type – Movement Disorder: Movement disorder and motor type evolve with time. Children who were classified as being hypotonic at the time of registration often developed hypertonia at long-term follow-up. Some children, who were classified as spastic, developed dystonic patterns in later childhood. Evolution of the movement disorder with time has implications for CP registers and in clinical management. We find selective dorsal rhizotomy (SDR) useful in a highly selected group of children with spastic diplegia of prematurity. SDR is contraindicated in dystonic CP.

When we compared our cohort to a contemporaneous Swedish cohort, considerable variation in the description of motor types was evident.[1,2]

1.2 Classifications of Cerebral Palsy: Topography: We have found the separation of topographical types into unilateral or bilateral CP to be valid, reliable, and stable, in long-term follow-up. The subdivision of bilateral types into spastic diplegia and spastic quadriplegia is arbitrary and unsatisfactory. The use of simple body diagrams, to illustrate the location of involvement and its severity, are useful shorthand, yet to be tested in both clinical practice and in the context of data entry to CP registers.

When we compared our cohort study to the contemporaneous Swedish study, marked variation was evident in classifying bilateral CP.[1,2]

1.3 Classifications of Cerebral Palsy: Gross Motor Function: We think that the Gross Motor Function Classification System (GMFCS) represents the biggest step forward in recent years, in providing a common language to classify gross motor function in CP.[4] In clinical practice and research, and in our cerebral palsy register, the GMFCS is a remarkably useful tool. It has a level of validity and reliability, which the movement disorder and topographical classifications do not. The contemporaneous Australian and Swedish cohort studies found a very similar distribution of GMFCS levels, in contrast to the lack of agreement in the description of motor types and topography.[1,2] In addition we found a direct relationship between GMFCS level and hip displacement.[3] This suggests to us that classification of CP by GMFCS gets to the heart of the major musculoskeletal issues in CP.

1.4 Classifications of Cerebral Palsy: Responsive Measures of Gross Motor Function: The GMFCS is reportedly stable over time and is not responsive to change after intervention. Classifying children who use several types of assistive devices and wheelchairs can also be difficult using the GMFCS alone. The Functional Mobility Scale (FMS) addresses some of these issues, by classifying the level of support required in the home, school, and community settings. The FMS describes functional mobility in six grades, over three distances, 5 metres, 50 metres and 500 metres. This increases its sensitivity compared with the GMFCS but maintains both validity and reliability.[5]

2.1 Classifications of Cerebral Palsy – Gait Patterns: Quantitative Classifications of Gait Using Instrumented Gait Analysis: Gait deviations are a key component of the CP motor disorder and are the result of the interaction between the effects of the brain lesion and the acquired musculoskeletal

pathology. Most clinicians with an access to a gait laboratory find instrumented gait analysis of great value as a tool for describing involvement in the individual child, planning intervention, and assessing the outcome of intervention. Relatively little work has been done on using three-dimensional kinematics as a tool for classifying gait patterns in CP.[6]

2.2 Classifications of Cerebral Palsy – Gait Patterns: Unilateral Cerebral Palsy: Spastic Hemiplegia: The classification described by Winters and colleagues in 1987 has been widely adopted as a useful template for the description of gait patterns in spastic hemiplegia and a useful basis for management algorithms.[7] Rodda and Graham have expanded the original classification, based on sagittal gait parameters to include the transverse plane and recent work reported good reliability.[8,9]

2.3 Classifications of Cerebral Palsy – Gait Patterns: Bilateral Cerebral Palsy: Spastic Diplegia/Quadriplegia: Gait patterns in bilateral spastic CP are much more complex, variable and less fixed than those seen in unilateral spastic cerebral palsy. However a number of studies have developed classifications based on both qualitative and quantitative kinematic parameters. Those studies based on cluster analysis have given some mathematical evidence for the validity of this approach. We have recently developed and published a combined quantitative-qualitative classification scheme based on sagittal plane kinematic parameters. We have provided evidence that this classification is valid, reliable, intuitive, and clinically useful.[10]

3.1 Classifications of Cerebral Palsy – Musculoskeletal Pathology: The majority of children with CP do not have musculoskeletal deformities at birth. These develop with time because of the combined effects of the movement disorder and impaired gross motor function. We have found a linear relationship between GMFCS level and hip displacement.[3] The incidence of musculoskeletal deformities in CP is very high, suggesting that a practical classification system should take account of musculoskeletal issues.

3.2 Classifications of Cerebral Palsy: Musculoskeletal Pathology – Contractures: Contractures may be defined as dynamic, when they are the result of the movement disorder but fixed shortening of muscle-tendon units is absent. Dynamic contractures include spastic equinus and flexion posturing of the elbow.

Fixed, or myostatic contractures refer to shortening of a muscle-tendon unit, in relation to the length of the accompanying long bone. Fixed equinus is present when the gastroc-soleus-Achilles tendon complex is too short to allow adequate dorsiflexion at the ankle during walking.[11,12]

3.3 Classifications of Cerebral Palsy – Musculoskeletal Pathology – Torsional Deformities: Long bones develop torsional deformities in the majority of children with CP. The most common torsional deformities are medial femoral torsion and lateral tibial torsion. In spastic diplegia of prematurity, intoeing is usually caused by bilateral medial femoral torsion, also referred to as increased femoral anteversion or 'inset hips'.

Torsional deformities contribute to gait dysfunction and may predispose to joint instability and degenerative arthritis.[11,12]

3.4 Classifications of Cerebral Palsy – Musculoskeletal Pathology – Joint Instability: The most common and clinically most important joint instability in children with CP is hip displacement. In a large population-based study we found the incidence to be 35%. Instability of the subtalar and midtarsal joints is also common.[11,12]

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Classification of cerebral palsy: collaborative study perspective

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The objectives of clinical trials are to determine the benefits and risks of interventions or therapies. This may involve comparison of one treatment with another existing treatment, with a placebo, or with a larger or smaller dose. Protocols are designed to minimize possible bias in favor of one of the treatment arms. The most ideal designs include the prospective comparison of active treatment to a control, random allocation to treatment assignment, masking of study personnel and patients – especially with regard to outcome assessment – a representative population, and adequate power to answer the question asked. Inclusion and exclusion criteria should be clearly defined. Numbers of those dropping out and crossing over between treatment groups should be low and relevant baseline characteristics should be reasonably equivalent among treatment groups.

Classifying children enrolled in a clinical trial at baseline with objective and reproducible criteria is required for determination of homogeneous subgroups, understanding of the natural history and prognosis, and generalizability of results to other samples or individual children. For the purpose of clinical trials, the classification of children with cerebral palsy (CP) has usually been based on physiological or technical parameters. These include weakness, spasticity or muscle hyperactivity,

joint mobility, and gait analysis. Often either the severity is not given or no details are given as to how to reproducibly determine the severity. Anatomical distribution is an essential part of the description, and etiology may be included.

Although assessments that provide measurements in physical parameters, such as tone or spasticity, can be used to compare different trials, they do not offer functional information. For children with CP and their parents, the goal of treatment is to improve motor skills and functional performance. It is assumed that improvement in physiological or technical parameters, such as joint range of motion or gait pattern, will improve function, but this may not always be the case. The use of true functional measures in clinical trials should be considered essential[1] both at baseline and as outcome measures. As detailed in a 1997 review,[2] these were not consistently used in trials but recently have become common, particularly with the development and validation of quality of life scales and gross motor function scales.[3]

The Gross Motor Function Classification Scale (GMFCS)[4] was developed to group children with CP into five levels of functional mobility based on the key function of ambulation. This outcome measure is currently being used in a large randomized clinical trial for the primary prevention of CP (The 'BEAM' trial – Beneficial Effects of Antenatal Magnesium). First, the presence or absence of CP at age 2 is determined by definite findings in any two of the following areas: (1) delay in motor milestones; motor quotient of 70 or less; (2) abnormalities of tone, deep tendon reflexes, co-ordination and movement; and (3) aberration in primitive reflexes, positive support reflex, tonic labyrinthine reflex, and/or postural reactions. If CP is determined to be present, then the GMFCS is used at the final visit at age 2 years to determine level of severity of CP. In order to incorporate hand and arm function, it has been amended in this trial by adding the ability to be able to grasp and release with both hands as a requirement for scoring above level III.

Clinical trials in CP therapies are not useful if they fail to define adequately the baseline characteristics of the sample enrolled. Participants may be described by distribution of weakness only, by abnormality of tone or spasticity with or without a level of severity, and with or without an anatomical distribution. If the functional level is described, it may be vague, such as ambulatory. This makes it difficult to interpret changes due to the intervention being tested, both within the specific trial and with regard to other children with CP. To determine the benefit of an intervention or therapy, baseline and outcome measures that are reproducible, complete, and relevant to improvements in functional abilities are essential.

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Better description of spastic cerebral palsy for reliable classification

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Cerebral palsy (CP) is not an aetiological diagnosis, but a clinical descriptive term. Children present with a wide range of impairments that must include motor manifestations, although these motor impairments may not be the most obvious nor have the greatest impact on function. There is a need for a multidimensional or multilayered classification and description system for use by clinicians with a view to indicating appropriate management, referring for therapy, justifying prognostic advice, communicating with other clinicians, as a record for later comparison for clinical or research purposes, or even to render a child eligible for services. Ideally the condition should be classified by aetiology (often unknown); by central nervous system lesion (if suitable neuroimaging is available); by identification of associated impairments (which often declare themselves at a later age), by description of movement disorder, and by functional status.

Traditional classification systems have focused principally on the distributional pattern of motor impairments (e.g. diplegia, hemiplegia) and the type of tone or movement abnormality (e.g. spastic, ataxic). This may be because it is the most readily available information and the usual reason that the child presents for medical attention. Whilst it is clear that ideally any classification or descriptive system needs to take into account the causation and timing, the anatomical and radiological findings, and the associated impairments, validity is also extremely important but has not yet been achieved with traditional classification systems. Whilst we develop better multidimensional classification, there is still a great need to further define and refine the characteristics used in the description of the motor abnormality.

CP classification and description has been under scrutiny in Western Australia since the 1970s. Since the late 1980s, this register has used an adapted version of the *Standard Recording of Central Motor Deficit* (Evans et al. 1989). Reliability was maintained by regular clinician meetings, which included activities to assess and improve inter- and intrarater reliability. As the population grew and many more clinicians, including remote and rural clinicians, were entering data into the register, it became harder to retain this superimposed reliability. In 2002, work began to establish a nationwide Australian Cerebral Palsy Register and to identify a minimum dataset that would be consistent with current knowledge, could continue to link with previously collected data, and would be reliable. Reliability of clinical description had to be achieved across the whole of Australia without the constant energy and resource input of face-to-face meetings. The classical terms proved problematic as the existing definitions are variable and imprecise. To operationally 'define'

Figure 1: *Description of Cerebral Palsy Part 1: Motor Impairments*

these terms (as we initially set out to do, via numerous national conversations, workshops, and questionnaires) did little more than add more definitions to the literature and only proved that reliability would not be achieved whilst using these terms.

We therefore concluded that it is more appropriate to aim to achieve reliability in describing the clinical features of CP rather than attempting to classify according to terms that are variably defined by clinicians from different backgrounds, centres, and eras. For entry into a national register, we believe it is necessary to specify each element of the clinical description and to operationally define them using unambiguous criteria that avoid the use of jargon terms that may be variably interpreted. It seems appropriate to ask clinicians for their primary observations (e.g. the degree of tonal abnormality in each muscle group) rather than their summary in specialized terms (e.g. the term 'diplegia') for which there is no generally agreed definition. This should allow competent examiners to describe the child with CP and produce identical information: i.e. the child's impairments will be reliably described. Although reliability is our primary goal, another important factor is the ability to compare our observations with those of other data collections. We were very aware of the manner in which other CP data collections, particularly SCPE (Cans 2000), describe and classify their cases and we aimed to have a system within which we could choose groups comparable to the classification categories of other collections.

The Australia Cerebral Palsy Register is proposing a limb-by-limb approach and Figure 1 is the final incarnation of many criticisms, feedback sessions, and suggestions over the past few years. The aim of this form is to convey an accurate clinical picture of the child with CP: to provide a reliable description. As function is so well described by the GMFCS and MACS and is influenced by associated impairments, particularly cognition, we have asked that motor impairments be separately described. We provide a separate stick figure for spasticity, dystonia, and for athetosis, recognizing that each of these disorders can have different anatomical distributions which may coexist. The flow of the chart requires the clinician to consider each type of motor impairment and then to rank their predominance, with provision being made for the possibility of equal ranking.

This system of describing primary observations allows clinical facilities and registers to compare *like* with *like*; and to group and compare children in many ways, e.g. based on primary type of motor disorder or on distribution of impairments. It also allows analysis of clusters of signs: exactly which features do cluster? What is the frequency of combinations of clinical signs?

This format still gives the clinician the opportunity to provide a verbal description, as we have found they like to do, and it helps us to understand how the traditional classification terms are being currently used, e.g. how do the clusters of signs relate to the the way that the traditional and classical terms of diplegia and triplegia are assigned?

To this end, we have asked that spasticity be quantified, and for this we use the Australian Spasticity Assessment (Love et al. Forthcoming; Table I). We are continuing to modify this to improve its reliability by conducting Australia-wide studies. Initially we used the Bohannon and Smith modification of the Ashworth Score (1987) to document degree of spasticity, but became aware that there was ambiguity in some criteria.

Table I: The Australian Spasticity Assessment

0	No catch on rapid passive movement (RPM) (no spasticity).
1	Catch on RPM followed by release. There is no resistance to RPM throughout the rest of the range.
2	Catch occurs in second half of available range (after halfway point) during RPM and is followed by resistance throughout remaining range.
3	Catch occurs in the first half of range (up to and including halfway point) during RPM and is followed by resistance throughout remaining range.
4	RPM is difficult; there is resistance to movement throughout the range.
5	RPM is not possible; body part appears fixed in flexion or extension during RPM but moves when passive movement is slow.

The ASA is based largely on the muscle response to passive movement and this is documented relative to the muscle length. The ASA is reliable in that it has tessellated criteria – unambiguous, mutually exclusive criteria – that between them cover every possibility. This means that every participant can fit into one, and only one category (Blair 2006). The ASA borrows heavily from Ashworth's (1964), Bohannon and Smith's (1987), and Tardieu's(1954) systems but removes the ambiguities.

It is not well understood how these variations in distribution and relative degree of spasticity relate to aetiology or outcome, or if indeed they do. It would be tragic to find in later years that the information which could quite easily have been collected by inserting a simple number may have enabled relationships to be made with pathology. Are these distinctions important? They are certainly important to the persons themselves and to planning for the services they require.

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Classification of cerebral palsy: clinical genetic perspective

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Cerebral palsy (CP) may be best understood as a group of clinically-defined, heterogeneous, motor impairment syndromes with onset in early life, which are not due to known progressive disorders and which have at least four dimensions of variability. These include: divergent patterns of motor dysfunction, varying levels of functional impairment, differing comorbidities, and multiple etiologies. Only the first of these is included in the most frequently used current definitions by Bax[1] and Mutch et al.[2] and it may be worthwhile to consider other categories, including etiology, in a revised classification scheme.

As applied to CP, etiology is the initiating or inciting cause which operates through pathophysiological mechanisms, either individually or as part of a causal sequence, to produce the clinical manifestations of CP. Etiology has not been traditionally included in the classification of the cerebral palsies, although Osler discussed its central importance in his initial monograph in 1889,[3] and modern definitions of CP have also emphasized etiology – at least to the extent of excluding known progressive brain disorders.

Extensive epidemiological research over the past 25 years has led to an understanding of etiology based on appreciation of risk factors for CP, particularly with respect to prenatal, intrapartum, and immediate postnatal circumstances.[4,5] It is now clear that most children with CP do not have a single cause of their condition and that a series of contributing factors, termed causal pathways, interact in complex ways to produce the final outcome[6]. In addition, this research has also made it clear that we do not yet know the cause of CP in the majority of affected individuals. As a result, etiological concepts are still evolving and the concept of serial or interrelated causal pathways is germane to any etiological classification.

Despite significant limitations in understanding the etiologies of CP, advances in neuroimaging have had a significant impact on our thinking in this regard. Current studies indicate that between 85 and 90% of individuals with CP will have abnormalities on magnetic resonance imaging (MRI).[7]

Despite the sensitivity of these imaging techniques, however, the neuroimaging patterns are not necessarily etiologically or pathophysiologically specific – a point which has perhaps been underemphasized. For example, MRI findings of periventricular white matter abnormalities are commonly associated with risk factors for prenatal, perinatal, and postnatal ischemia and inflammation.[8,9] Nonetheless, similar MRI findings are associated with genetic conditions, a variety of neurometabolic disorders, and even nutritional deficiency states.[10,11,12] The same can be said for MRI patterns of selective basal ganglia injury which are found not only following severe perinatal cerebral ischemia but also in an increasing number of inherited metabolic disorders.[13,14]

With this understanding, the admonition to consider CP a symptom complex (or phenotype) which requires a thorough search for underlying diagnosable and perhaps treatable diseases seems particularly apt – a point also discussed in the recent practice parameter statement regarding diagnostic assessment of CP.[7,15,16] Modern neuroimaging will need to be coupled with diagnostic testing for a growing number of genetic and metabolic disorders in order to identify those few individuals with CP caused by rare diseases. Although current data indicates that less than 5% of cases of CP are caused by these conditions, the lack of systematic studies of populations affected by CP leaves questions regarding their true prevalence. The current challenge is to recognize those with CP who are likely to benefit from these expensive and technologically-challenging diagnostic studies. Additional research in this area is critically needed.

Whether individual patients with specific neurometabolic/genetic diagnoses are ultimately included or excluded from the CP diagnostic group is also a complex issue, in view of the practical difficulties in recognizing very slowly or intermittently progressive diseases and the staggering variability of genotypic and phenotypic expression in these disorders. An approach to standardizing criteria for inclusion of a large number of genetic and metabolic conditions has been suggested[17] and the concept of 'non-progressive' brain disorders is an important issue in any revised classification scheme.

Notwithstanding the importance of diagnosing biologically-treatable causes of motor impairment, and the importance of genetic counseling when possible, these are very rare conditions which, even as a group, do not affect most individuals with CP. Nonetheless, it is readily apparent that etiological classification has much broader implications. Understanding etiology and causal pathways lies at the root of prevention strategies and treatment opportunities for all individuals with CP. Currently the vast majority of cases of CP are not preventable and very few are biologically treatable. Defining etiologies and their underlying pathophysiologies will provide the best route to identifying effective biomedical therapies in the future.

Additionally, the importance of etiological formulations can hardly be overestimated in terms of the current medical-legal climate. Inclusion of an etiological subclassification could be extremely beneficial in reducing current misunderstanding on issues of causation which are so destructive and expensive when filtered through adversarial judicial systems.[18] Emphasizing etiology could also provide the basis for further research and additional evidence-based recommendations regarding the diagnostic studies required for etiological evaluation of CP, both for clinical care and medical-legal complaint.

Including a category for presumed etiologies in a new classification scheme would more clearly indicate what we know and do not yet know about the causes of CP on a case-by-case basis. Even if many individuals with CP have their etiology classified as 'unknown', the inclusion of this category could foster shared understanding and accurate communication among all concerned and potentially serve to stimulate further research on issues of targeted management and interventions for specific populations.

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Definition of cerebral palsy: clinical perspective

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The term 'cerebral palsy' (CP) enjoys considerable familiarity in the professional and lay literature. Most health professionals and many lay people think they know what it means. The term is frequently misused and, like other medical terminology used to describe disabilities, it can take on a pejorative connotation. There is much human anguish, financial expense, and professional time tied to the stubbornly level prevalence of 'cerebral palsy.' The best minds tell us that the problem is not going to go away.[1] An era in health care is arriving wherein much more precision can be introduced to the discussion of motor disability in children thanks to major conceptual and technical advances.

Definitions of CP in recent reviews[2] are all derived from the elegant annotation published by Bax in 1964:[3] 'A disorder of movement and posture due to a defect or lesion in the immature brain.' Previous generative work on definition, as reviewed by Ingram goes back to Little and Freud[4]. CP, like mental retardation, autism, and attention deficit disorder, is a recognizable pattern of altered neurological development, to borrow a phrase from the dysmorphologists, or, one of a set of 'syndromes of cerebral dysfunction' to quote the Bax annotation. The heterogeneity of CP is emphasized by some who prefer to use the plural 'cerebral palsies'[5].

'Cerebral palsy' is a deeply rooted term in the US. The United Cerebral Palsy Association (UCPA) is one of the most respected nonprofit consumer advocacy and service organizations for persons with disabilities in the US. The philanthropic power of UCPA supporters created the United Cerebral Palsy Research and Educational Foundation which supports innovative research. CP is enshrined in federal legislation as one of the official 'developmental disabilities' that qualify persons for certain government programs. Despite long-standing recognition of the limitations of 'CP' as an umbrella term, no viable alternatives have been proposed. Multi-syllabic neoconstructions seem destined to add only further confusion.

A useful clinical label and an accompanying classification system should help drive the diagnostic workup and choice of treatment, as in the example of the recent development of practice parameters.[6] The clinical label should be useful in discussing recurrence counseling and prognosis. The prognosis encompasses survival, function, and health status. A useful clinical label drives health care services and will influence more functionally-based services such as education and social support.

Degenerative diseases have been excluded from the etiologies of CP even though, at any given point in time, a child with a degenerative disorder may have motor disability that is indistinguishable from that present in a child whose 'lesion' fits the definition of CP. There is an inherent pathophysiological difference between a central nervous system that has been injured or is malformed from one that is subject to an inexorable degenerative process. Careful history, imaging techniques, and laboratory evaluations help make this essential distinction.

Conceptual advances regarding health are reshaping terminology. The concept of disability is giving way to a health status construct in which biological, social, and personal attributes determine activity and participation in society. These concepts are embodied in the International Classification of Function, Disability and Health (ICF) developed by the World Health Organization.[7] The ICF is linked to causes of impairments and disabilities through the International Classification of Diseases.[8] This conceptual model has been driven in part by individuals with disabilities, especially CP, asserting their rights to self-determination. The development of valid and reliable measurements of previously subjective characteristics, such as motor performance and quality of life, have accelerated the development of the broader concepts.

Definition and classification should be the same for clinical and research purposes, otherwise, research, whether at the bench, clinical, epidemiological, or health services levels cannot be easily applied, either at an individual or public health level. Research definitions need to be more precise and carefully parsed than those in daily clinical use to satisfy the demands of logic inherent in rigorous study design.

Application of the results of research findings to a single individual requires an extrapolation back to a more comprehensive, individualized view of the human being facing the clinician. The increasing specificity of interventions demands a precise means of defining and classifying the disorders that fall under the rubric of CP.[9]

There are practical reasons to retain 'CP' as a familiar umbrella term and to require specific descriptions across multiple dimensions including:

1. Clinical evidence, supplemented by imaging whenever possible, that the motor disability is due to a malformation or acquired lesion of the motor control areas of the brain.

2. Firm evidence of the nonprogressive nature of the central nervous system process causing the disorder derived from history, serial clinical observation, and imaging.

3. Timing of onset may be preconceptional, prenatal, perinatal, postnatal but during the time that the CNS is overtly immature. One possible definition for the demarcation between 'CP' and 'brain injury' is 36 months of age when the motor characteristics defined by instrumented gait analysis approximates maturity.[10]

4. Precise clinical description regarding body segment involvement, tone abnormalities, and movement disorder. Allow multiple descriptors that correlate with anatomically accurate brain localization.

5. Severity of the neurological impairments graded using valid and reliable clinical measures such as the Tardieu scale.[11]

6. Severity of the functional consequences graded using valid and reliable clinical measures such as the Gross Motor Function Measure[12] and the Gross Motor Function Classification System.[13]

7. Rigorous identification of etiology should be carried out relying on medical history, a minimum three-generation family genogram and, wherever possible, CNS imaging. Laboratory evaluation will be driven by the history. A specific, clear history of hypoxic-ischemic encephalopathy must be documented from primary sources and confirming imaging must be present to invoke perinatal anoxic injury as a cause.

8. Associated impairments due to injury to the CNS and secondary impairments due to CP itself should be catalogued separately.

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Classification of cerebral palsy: behavioural perspective

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The classification of behaviour as disorder is a fraught and complex field. Lessons learned from this experience are here applied to the challenges inherent in the diagnosis and classification of cerebral palsy (CP).

In psychiatric diagnosis and classification, a number of approaches are taken. Their aim is to take account of the fact that human behaviour is complex, and to give a coherent and pragmatic approach to the vexed issue of labelling behaviour as 'disorder'.

One major issue concerns whether to adopt a 'dimensional' or a 'categorical' approach to the labelling of behaviour as a disorder. In the dimensional approach, there are continuities from 'normal' to 'abnormal': this is appropriate for most types of behaviour, where the definition of disorder is a matter of degree. Mood and anxiety are good examples of normal traits and aspects of normal experience, which lie along a continuum, at the extreme end of which disorder can occur. The categorical approach recognizes that there are some behavioural patterns that are different in nature from anything within normal experience, labelling these as disorders. The experience of realistic hallucinations, perceived to be real by the individual in the absence of any fever, toxin, or hallucinogenic drug, is an example of such a categorically different, or deviant behaviour. Both the dimensional and the categorical approaches rely on observations of the actual manifest behaviour, in all its complexity, and also in the developmental trajectory and natural history seen in the person. Putting it another way, the hallmarks of disorder can be seen in *quantitative* terms – i.e. being in excess of some level of normality on a dimension; or in *qualitative* terms – wherein there is something about the nature of the behaviour which is not within normal experience.

Because human behaviour is complex, and because psychiatric diagnosis endeavours to make sense of the interplay between normal reactions to normal experience and abnormal disorder, most approaches to psychiatric classification include elements of both dimensional and categorical approaches.

In so doing, psychiatric classification usually employs a number of dimensions – often arranged in a hierarchy. The hierarchy aids in identifying which is/are the dominant, or clinically most important, aspect(s) of behaviour.

Furthermore, the delineation of sets of behaviours aids in the delineation of syndromes. Diagnosis by observation and systematic measurement of behaviour is essentially the delineation of clinical syndromes, where the syndrome is defined according to sets of behaviour, best viewed along their developmental trajectory. The same term – syndrome – is often applied to the putative aetiology of the set of behaviours, where there is a discrete genetic cause, and the behaviours are essentially the manifestation of the behavioural phenotype of the condition in question. In this latter context, classification can then be made according to aetiology. While some attempts to develop psychiatric diagnosis and classification according to aetiology have been made, (e.g. 'post-traumatic stress disorder') the majority of psychiatric disorders are not classified by aetiology.

So, where does CP sit, if we apply these approaches? Overall, CP emerges as a mixed concept, combining aspects of observations of the presenting state, and a putative aetiology. In the observations, we see some, such as motor power and muscle tension, which might appropriately be assessed along a continuous dimension. Others, such as the occurrence of abnormal involuntary movements, are qualitatively different from normal experience but can be measured on a scale of severity. Once the various aspects of the clinical manifestations of CP are delineated, the utility of a multidimensional approach becomes apparent. When we come to consider the issue of hierarchy, the relevance of a similar approach to that employed in psychiatry – wherein the dominant dimension is typically the most important or functionally relevant – is readily apparent. For in CP, the challenge is to describe and classify according to the impact of the disorder on the functioning of the individual.

Contemporary approaches to the classification of CP are therefore, in many ways, similar to those employed in psychiatric practice. Dimensions of experience are recognized – and in CP the issue of their continuity with normal experience is importantly emphasized. Multiple observations along the various dimensions comprise syndromes, which have long been employed in the recognition of different types of CP. Where possible, the aetiology is described, along with groupings of sets of observations, especially those which typify such discrete causalities, in a manner which informs treatment and case management.

In conclusion, the approaches to diagnosis and classification employed in psychiatry are empirically applicable to CP. As always in medicine, the ultimate aim is to classify disorder in a manner which reflects both measurable observations and causality, and especially – and ultimately – informs clinical management in a manner relevant to the individual affected patient.

Definition and classification of cerebral palsy – an epidemiologist perspective

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Most epidemiological studies of cerebral palsy (CP) describe the prevalence or outcome of CP, estimate the strength of association with antecedents of CP, or estimate the effect of an intervention to prevent or treat CP. For each of these goals a somewhat different definition may be preferred.[1–2] For example, if the objective of the research is to estimate the need for education and rehabilitative services, the most useful definition of CP might include the criterion that an individual's neurological abnormality results in activity restriction, so that individuals without such limitation would not be counted as cases, despite their having an obvious neurological abnormality.

Most often the goal of epidemiological research is to estimate the strength of association between an exposure, characteristic, or intervention, and a disease (or some other health outcome). In general, this goal is best served by a definition that minimizes misclassification. Misclassification refers to either classifying an individual as having CP when in fact he or she does not, or classifying an individual as not having CP when in fact he or she does. One form of misclassification, referred to as over-ascertainment, is classification of individuals without CP as having CP. If the frequency of over-ascertainment is similar among exposed and unexposed individuals, the misclassification is referred to as non-differential. Non-differential over-ascertainment in clinical trials results in a bias towards the finding of no treatment effect and, in observational studies, towards the finding of no association between exposure and disease. Differential misclassification has the potential to distort our perception of reality either towards no association or by magnifying the apparent association.[3]

The epidemiologist's strategy for limiting over-ascertainment includes specifying a clinically significant degree of abnormality, such as the magnitude of hypertonia and hyperreflexia, as a part of the definition of CP. A definition that includes individuals with questionable abnormalities increases over-ascertainment. Recognizing the inherent imprecision in assessments of tone and reflexes, some epidemiologists have excluded cases where examiners might be less confident about the diagnosis.[4–5]

In an analysis from the Collaborative Perinatal Project, children with 'non-handicapping motor impairments' were excluded, and in studies from the California Cerebral Palsy Project,[6] children with mild CP were excluded. In the Neonatal Brain Hemorrhage study, associations were analyzed between risk factors and both 'disabling' as well as non-disabling CP. The odds ratios of CP associated with mechanical ventilation were 6.9 for disabling CP and 3.6 for non-disabling CP. Such differences in odds ratios for disabling and non-disabling CP could be attributable, in part, to more frequent over-ascertainment among those classified as having non-disabling, as compared to disabling, CP.[7] As one of the investigators subsequently wrote: 'it is difficult to ascertain non-disabling forms of CP reliably'.[8] Low reliability implies low validity.

Non-differential under-ascertainment (incorrectly classifying

an affected individual as not having CP) does not bias risk ratios, even though the precision of the estimated risk ratios and statistical power are decreased.[3] Thus, in an ongoing trial of antenatal magnesium sulfate to prevent CP in preterm infants (Benefits of Antenatal Magnesium Sulfate Trial, sponsored by the National Institute of Neurological Disorders and Stroke), the outcome of interest is 'moderate or severe CP', thereby excluding children with 'mild' CP, whose abnormalities are more likely to be over-ascertained.

A useful definition of CP for epidemiological studies is one that identifies as cases individuals with similar dysfunctions. A definition that includes individuals who share some, but not all, aspects of a phenotype, increases the likelihood that these cases arise from a heterogeneous group of causative pathways. As with over-ascertainment due to inclusion of individuals with 'borderline' abnormalities, the result of phenotypic heterogeneity may be an attenuation of exposure-disease (or treatment/outcome) associations.[3]

The argument for increasing phenotypic homogeneity applies also to decisions about classification (sub-grouping) of CP cases. The greater the heterogeneity within a subgroup of cases, the less likely it is that valid estimates will be obtained for the strength of causal associations.[3] An illustration of this concept is provided by a study of maternal infection and CP, in which associations were analyzed for three subgroups of CP – quadriplegia, diplegia, and hemiplegia. In these subgroups, the odds ratios for the association with maternal infection were 19, 6.7, and 2.3,[9] respectively. While random variability might explain the heterogeneity of these odds ratios, it is also plausible that some of the variation was due to differences in the causal pathways leading to specific subtypes of CP.

In attempting to define homogenous groups of cases, with respect to the specific neurological abnormality(ies), severity of the functional impairment, associated neuroimaging findings, or some other attribute, the epidemiologist may be left with subgroups of cases with only a small number of individuals, resulting in imprecise measures of association or prevalence. However, if one or more subgroups have a very similar profile of risk factors, it is reasonable to consider combining these subgroups to obtain more precise estimates.

In summary, from the epidemiologist's perspective, no single definition or classification system is likely to be optimal for all research studies. Instead, epidemiologists should collect information, when available, about each potential case's neurological abnormalities, the level of certainty about each abnormality, the functional severity related to the abnormality, and associated neuroimaging findings. With these data, the researcher can explore multiple strategies for assigning individuals to case and control groups, and among the cases, to subgroups, and thereby obtain the most information about CP.

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A neurological perspective on definition and classification

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We are very grateful to Martin Bax, Murray Goldstein, and the executive committee of the new 'Definition and Classification of Cerebral Palsy' (Rosenbaum et al. 2007) for providing a clear and useful document that will be helpful to many specialties involved in the care of children with chronic disorders of the motor system. The definition and its commentary emphasize the variety of features of the disorder and its associated disturbances, and, therefore, the report supports the need for multidisciplinary assessment and treatment of this complex syndrome. To this we would add an equally important need for multidisciplinary *research* on the diagnosis, treatment, prevention, epidemiology, and social impact of CP.

Children with CP face many challenges, but each challenge provides an opportunity for treatment. Interventions that change society or provide appropriately accessible buildings and tools can affect the success of specific activities and participation in life situations, even when the motor disorder remains stable over time. Interventions that protect from injury, change the underlying pathophysiology, or minimize the severity of the resulting motor disorder can reduce impairment and improve function and participation. Neurologists are particularly concerned with this latter group of interventions. Unfortunately, our currently available treatments take only small steps in this direction, and neurological practice is often based on only small studies or anecdotal observations.

As neurologists, we are therefore acutely aware of the need for research on the neuroscience of CP. The tremendous strides over the past decades in the treatment of the musculoskeletal consequences of neurological injury have not been matched by similar progress in the treatment of the neuro-

logical injury itself. Many promising results on neuroprotection, neurotrophic factors, oligodendrocyte development, programmed neuronal death, cortical neurophysiology, and basal ganglia neurochemistry have yet to be translated into clinical trials in CP. As a result, there are few data on the use of treatments that could improve abnormal brain function. There are almost no data on the use of treatments that could alter the neurological development of children with early acquired brain injury and thereby delay or prevent the clinical expression of the disorder.

If we are to make progress toward treatment of the neurological deficit, what might a neurologist require from a definition or a classification? Definition and classification must allow neurologists to identify relatively homogeneous groups of children for whom diagnosis and treatment can be tested and ultimately standardized based on reliable clinical trials. Ultimately, a definition and classification must allow a neurologist, faced with an individual child, to predict with some degree of confidence the likely response to different treatments so that the most appropriate and safe interventions can be chosen. Features of a child that will contribute to the correct choice of treatment include the child's goals and all of the personal and environmental factors that influence success in achieving those goals. But equally important is the relation between the intervention and that child's particular pathology.

Therefore, the most useful *classification* for a neurologist is one that aligns with pathology and the response to treatment. The relation between categories of pathology and the response to neurological treatments can be discovered by research. In CP, the pattern of injury to a child's brain is believed to be influenced by neural development, vascular anatomy, and the many other factors that affect injury and recovery (Keogh and Badawi 2006, for review). Neuroimaging studies suggest that there are particular patterns of injury that occur with relatively high frequency, including periventricular white matter injury, cerebrovascular occlusion, and selective neuronal injury (Zimmerman and Bilaniuk 2006, for review). Each of these patterns of brain injury appears to be associated with a particular clinical syndrome such as spastic diplegia, hemiplegia, or dystonia (Volpe 2001, for review). Improvement in treatment will depend upon further research into such associations. For example, whether anti-spasticity or anti-dystonia treatments are effective may depend upon the presence or absence of injury in regions associated with spasticity or dystonia. The opinions of expert clinicians provide an essential starting point for classification, but opinions must subsequently be verified by research.

The most useful *definition* for a neurologist is one that is both sensitive and specific; it must include all children with similar pathophysiology while excluding other disorders. For example, the effectiveness of a particular treatment in children with hemiplegia may differ between children with prenatally acquired lesions and those with postnatal embolic stroke. Whether such children are best defined for neurologists as 'hemiplegic cerebral palsy' or 'pediatric stroke' will ultimately depend on the mechanism, the resulting pathology, and the response to treatment. Therefore a definition that will affect neurological management must be scientifically tested, and we encourage both basic and clinical research to do so. Any future modifications of the definition can then be guided not by changes in opinion but by new scientific discoveries.

There are many purposes for a 'Definition and Classification of Cerebral Palsy,' and we acknowledge with gratitude that we were given the opportunity to participate in this important effort. Uniform terminology will have great benefits for the many disciplines that are concerned with the care of children with motor disorders. As neurologists, it is our sincere hope that exploration of pathophysiology, specific patterns and mechanisms of injury, and the relation between injury and development will allow progress toward prevention, treatment, and eventually cure of the neurological components of this complex disorder.

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Other current definitions and classifications

Commentary on the revised versions of the definition and classification of cerebral palsy

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In cerebral palsy (CP) and other common conditions including asthma,[1] the name reflects only the clinical signs, and not the aetiology. This may give the misleading impression that all cases stem from the same cause, and many regard such names as obsolete. However, the advantage of retaining names familiar to the public and the professions has been generally recognized, but the development of medical knowledge brings with it a need to update and expand these names. The papers in this Supplement are a welcome attempt to improve and clarify currently used definitions of CP.

The definition suggested by Mutch et al.[2] was produced at an international meeting which revealed that some clinicians were still labelling cases due to spinal trauma as CP. The method of recording proposed then was simplified to make it accessible in situations where such children were not seen by neurologists.[3] We have moved forward from this, and this is reflected in the new proposed definition. However, in our view it fails to stress the aetiological heterogeneity enough. This is particularly serious in CP, where its attribution to birth injury remains much overrated, an impression that has had profound policy implications. It was to put stress on this heterogeneity that the wording 'umbrella group' was used in the 1992 definition, and it also led to the use of the plural 'cerebral palsies'.[4] We would like to see the authors of this revised definition include a phrase acknowledging that the clinical condition, although always non-progressive, and stemming from the period of brain development, is largely determined by the type and precise timing of the initiating cause.

The proposed classification broadens and brings an increased stringency to the clinical descriptions of individual cases of CP. The inclusion of measurements on orobulbar and truncal function is welcome. Although it is unlikely that all eligible cases will be seen by physicians familiar with this classification, the knowledge of its existence, and training in its use, should gradually improve the quality of recording. The tests of validity by Palisano et al.,[5] Bartlett et al.,[6] and Eliasson et al.[7] are an essential basis for the common use of the proposed codes. Nevertheless, our own experience, and that of others, is that the integration and analysis of such very detailed and voluminous data for epidemiological purposes is far from simple, even given the recent explosion in computer and statistical packages.

Moreover, the authors' commendable caution about making assumptions about the timing and nature of the causal incident risks 'throwing the baby out with the bath water'. Data on birthweight, gestational age, and the presence of malformations must be included in any comprehensive data analysis as they may provide basic clues to aetiology, although preterm birth can be an event that lies on the causal pathway, rather than initiating it. Regrettably, there is still no general agreement on the type and quantity of investigations that each affected child should receive, or on their classification. The authors state that every effort should be made to investigate causal pathways, but fail to stress the urgency of such attempts, which are essential both for the sake of the families and to help plan specific preventive interventions. Neuroimaging results are an important extension of those produced by clinical examinations, and may indicate the likely timing of the initial cause as well as the site and type of damage. However, metabolic, virological, and haematological investigations and molecular genetics are more likely to give an explanation of the origin of the condition, and their use is burgeoning. It is regrettable that the report by Ashwal et al.[8] from the USA suggests that the latter are performed only if MRI results are not informative.

All in all, these papers provide a much needed step forward in the management of these conditions, but we should ensure that the concentration on the minutiae of clinical findings is accompanied by an increase in the resources and effort to be put into research into their causes.

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Definition and classification of the cerebral palsies: the Australian view

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Background: The Australian Cerebral Palsy Register (ACPR) is a collaboration involving all Australian States and Territories that commenced in 2002. With the aim of pooling data to conduct research, the definition of cerebral palsy (CP) used for the ACPR is, most importantly, a means of producing consistency of case selection between centres.

Western Australia (WA) has the longest standing CP register in Australia, established in the late 1970s with the inclusion of cases born from 1956 onwards. Clear diagnostic criteria (Badawi et al. 1998) and methods of improving interobserver agreement (Blair and Stanley 1985) in the classification of CP have been under scrutiny in WA since the early 1980s.

Definition of CP: Publications arising from the earlier years of the WA Register quoted the concise definition of Bax (Bax 1964) and later the more precise wording offered by Mutch et al. (Mutch et al. 1992). Defining a condition for which there is no definitive test, only a clinical description, remains problematic (Stanley et al. 2000).

When the methodology of the ACPR was considered by the national collaboration in 2003, it was decided that rather than subscribe to any one definition of CP or produce yet another, we would support the decision made by the Surveillance of Cerebral Palsy in Europe (SCPE 2000) to accept any definition that includes the following five key elements: (1) CP is a group of disorders, i.e. it is an umbrella term; (2) It is permanent but not unchanging. (3) It involves a disorder of movement and/or posture and of motor function. (4) It is due to a non-progressive interference/lesion/abnormality; and (5) this interference/lesion/abnormality is in the developing/immature brain.

These criteria do not provide sufficiently specific guidelines with which to reliably select the cases that meet them. In order to obtain a reliable definition, a number of other factors must be specified. These additional factors relevant to criteria 3 to 5 above are explained below based on the experience of the WA CP Register.

Criterion 3: The lower limit of severity of the disorder of movement/posture must be specified with respect to a standard. The standard chosen in WA is the presence of neurological signs of the motor impairment. These neurological signs may or may not be associated with limitations in normal activities of daily living but are likely to limit the potential for physical performance. Those in whom the motor impairment is so minor that it may not be apparent to an untrained eye are classified as 'minimal CP'. However, a label of CP may be given to children who do not demonstrate neurological signs, such as idiopathic toe walkers, in order to make them eligible for services that may benefit them. Therefore, the criteria that define the lower limit of severity for clinical purposes do not necessarily agree with those required for CP registers. The ACPR has overcome the subjectivity of such a severity classification by grading severity according to the Gross Motor Function Classification System (GMFCS; Palisano et al. 1997), and the Manual Ability Classification System (MACS; Eliasson et al. 2006) but still requires the presence of neurological signs to define a case.

Several conditions that meet Criterion 3 have been excluded historically. This originated in the concept of CP as a diagnosis, to the exclusion of other diagnoses. A diagnosable condition could not, therefore, include CP as a component of the diagnosis. Thus, easily recognized syndromes tend to be excluded from the CP rubric even if they are associated with non-progressive cerebral defects/lesions that result in disorders of movement and/or posture (e.g. Angelman syndrome; Badawi et al. 1998). Furthermore, individual research groups have their own idiosyncrasies for including or excluding certain conditions. This does not present a problem provided they are identified so that consistency between registers can be achieved.

Criterion 4: It can be difficult to identify progression or resolution of a neurological abnormality in a very young child since the clinical picture is constantly changing as a result of development. In Australia 5 years was chosen on pragmatic grounds as the upper age limit for deciding to include or exclude a case, as most often by that age those that are going to resolve will have done so and syndromes that are progressive will have come to light.

This also raises the question of the earliest age at which the label of CP can be reliably conferred. Infants with brain defects/damage typically exhibit abnormal behaviour and development, the degree of abnormality correlating with the severity and extent of the cerebral damage. Severe CP can often be recognized during the first months of life. However, infants with brain damage are at increased risk of mortality – the more severe the damage the greater the risk of mortality – and the rate of mortality is highest in the earliest months of life. If in WA we were to exclude those with an obvious motor abnormality who die under the age of 5 years, this would represent a loss of 17% of those children with severe CP (i.e. unlikely to achieve independent ambulation with or without aids). Excluding deaths under 5 years of age would not only underestimate the rate of CP, it would also truncate the distribution of severity of impairment.

Children in Australia suspected of having neurological abnormalities are examined by an experienced neurologist or developmental paediatrician soon after the abnormalities are noted. Both the WA Register and the ACPR accept a description of CP as soon as the attending specialist is convinced that this is the correct label. However, if the child is alive at the age of 5 years, clinical information is reviewed, and if necessary amended, at that time.

Criterion 5: At what age can the brain be considered mature? Some people maintain that the human brain continues to develop for decades. The acquisition of motor function is also a continuous process that peaks after decades and so does not provide a rationale for specifying any particular age as the age of cerebral maturity. Therefore, the specification of 'maturity' for this criterion must be an arbitrary one. In Australia we consider any child acquiring a motor disorder as a result of a brain-damaging event before the age of 5 years to

have CP. However, those whose neurological impairment follows a well-documented causal event after the age of 28 days and before the age of 5 years are grouped separately as post-neonatally acquired CP. The age of 28 days defines the end of the neonatal period and usually differentiates events related to gestation and delivery from those largely independent of it.

Summary of additional criteria: To summarize, in order to refine the specificity of the generally accepted criteria for CP, we feel it is necessary to: (1) define the lower limit of severity together with the standards on which that definition is based; (2) specify known syndromes that are included in or excluded from the data set; (3) define the age of ascertainment at which progression or resolution is decided; (4) define the minimum age of inclusion and the criteria which must be met should the child die before the age of ascertainment; and (5) specify the upper age limit of acquired brain injury to be included.

It is not necessary for everyone to agree on these further criteria; it is only necessary that each register define their criteria and specify them when reporting on their samples so that data can be compared or pooled appropriately.

Classification: The term CP applies not so much to a *group* of neurological disorders as to a continuum of pathologies and clinical descriptions which can result from a wide variety of aetiological pathways, many of which are as yet imperfectly understood. If it were a group of discrete disorders it might be anticipated that classification by pathology, clinical description, and aetiology would each identify the same groups: one group for each disorder. However with some exceptions, choreoathetosis following kernicterus being the most notable, correlations between pathology, aetiology, and clinical description appear to be rather weak. For the time being then, pathology, clinical description, and aetiology must be considered independently, and correlations between them examined. Although some authors are attempting to incorporate the results of magnetic resonance imaging (MRI) into the classification of CP, the relationship between motor impairment and the brain injury seen on MRI is not yet clear. In Australia, while an MRI is now considered to be part of the routine work-up on all children with CP, there are still many who never undergo such imaging studies. Therefore, since the aetiology is unknown or uncertain in a large proportion of cases, the primary basis for classification is clinical description.

However, the clinical manifestations of CP are almost infinitely variable. Motor impairment may be accompanied by sensory and/or cognitive impairments and/or epilepsy, any of which may be more disabling than the motor impairment. Classification systems are often designed to meet a specific need. For example, one might want to classify in order to identify children suitable for a particular therapy or service, or to identify suitable comparisons in a clinical trial. The classification categories appropriate for each purpose may well differ. We, therefore, believe that there is no ideal CP classification system that will meet all the reasons for which one might want to classify. The poor reliability of classification systems for people with CP to date may be the result of variation in the goals of the classifiers when making the classification.

The lack of interobserver agreement in classifying CP is of considerable concern when the aim is to identify suitable groups for comparison in research. For the ACPR we therefore concluded that it is more appropriate to aim to achieve reliability in describing the clinical features of CP rather than attempting to classify according to terms that are variably defined by clinicians from different backgrounds, different centres, and different eras. Achieving reliability of clinical description will assure reliability of any classification system based on any combination of elements of that description, since classification could then be performed by computer software. This has the added advantage of having the flexibility required to consider differently defined groupings for specific purposes.

To this end we have devised and are currently testing the reliability of a form for collecting data concerning the clinical features of CP that includes the type, bodily distribution, and degree of impairment of each type of movement disorder; function as measured by the GMFCS, the MACS, speech and swallowing ability, and the existence and severity of any associated visual, auditory, or cognitive impairments.

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Recommendations from the SCPE collaborative group for defining and classifying cerebral palsy

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Introduction: Several aims of classification can be argued: (1) monitoring subgroups of cerebral palsy (CP) whose prevalence rate is expected to change over time; (2) aetiological research on CP, according to the neurological subtypes and to the birth circumstances; (3) evaluation of interventions in children with CP, according to the severity of the motor impairment; and (4) comparisons with other studies. The act of classifying requires to have some properties, and among them the three most important are the reliability, the validity, and the simplicity.

Reliability is the consistency or repeatability of classifying, i.e. different persons at different time periods will classify a group of CP children in the same way. Accuracy of the reliability (reproducibility) can be assessed through statistical measurement (kappa coefficient), and has to be tested between professionals (observer) and within the same observer at different time periods (intraobserver).

Validity means that we are measuring what we are supposed to measure, i.e. the quality of its effectiveness. In deciding if a classification is valid or not, we come back to the aims of the classification, since its relevance may vary according to its use, e.g. a classification based on etiological circumstances might not be relevant for evaluating intervention therapies.

Simplicity is the quality of being simple or uncompounded or, in other words, 'easy to use' by anybody. It would not be practical or easy to require an expert child neurologist to examine a child for an hour in order to classify that child as having CP and this would, therefore, be difficult to recommend on a large scale.

It has always been difficult to compare data from CP registers and thus be sure that the differences observed reflect actual 'true' differences, or are due to differences in definition, criteria, and classification. This may concern CP prevalence rates, trends in CP prevalence rates, or comparison of characteristics between CP subtypes. For instance, one study (Meberg and Broch 1995) showed a decrease in occurrences of CP during the 1980s whilst all other studies had shown an increase (Hagberg and Hagberg 1996, Pharoah and Cooke 1997, Topp et al. 1997). More recently, there have also been some differences between authors in the one country regarding the changing trends in the severity of CP (Colver et al. 2000, Surman et al. 2003).

In 1998, a collaborative network of CP registers and population-based surveys was established with the help of funds from the European Commission. The reasons for this collaborative effort were: (1) the need for standardization and harmonization of the definition, inclusion/exclusion criteria, and the characteristics used currently for describing children with CP, and (2) the need to get large numbers in order to be able to analyze distinct subgroups of CP and, in particular, their trends over time. The Surveillance of Cerebral Palsy in Europe (SCPE) network started in 1998 with 14 centres from eight countries, and at present there are 22 centres from 15 countries. The aim of this network was to: (1) harmonize CP data collection using a standard CP definition and an agreed minium data set; (2) to develop a central database of children with CP in order to monitor trends in birthweight specific rates, and to provide information for service planning; and (3) to provide a framework for the development of collaborative research projects in CP field (SCPE 2000).

Methods: Agreeing on classification: During the first 3 years of the network, a subgroup of professionals, comprising child neuro-paediatricians, child rehabilitation doctors, and epidemiologists, representatives from different centres, met together several times before being able to propose a consensus on definition, criteria, and classification of CP. This consensus was presented to all participants of the SCPE network during a plenary meeting held in Oxford in 1999. Besides the skills already described above, other professional skills were present at this meeting, i.e. neonatologists, obstetricians, paediatricians, geneticists, and public health doctors. There was more discussion but also a strong desire to reach a consensus, and thus the SCPE criteria and classification system for CP were agreed.

Implementing the classification: A few months after this meeting it appears that difficulties remained when pooling and comparing information from different sources. The persisting problems were mainly due to the matter of language since not all partners from the different countries were English native speakers. Not everyone had derived the same meaning from terms such as 'increased tone' and 'walking fluently'. Thus, during the next 3 years, collaborative efforts were put together, mainly between child neuro-paediatricians, in order to develop a video-based tool, the SCPE Reference and Training Manual (SCPE R&TM). The aim of this tool was to promote a shared understanding of the words and phrases used to describe the clinical, functional, and neurological features of CP. Text and video material were first discussed within the small group of child neuro-paediatricians and then proposed to illustrate these features and to discuss pitfalls in diagnosis and classification. Interobserver exercise has been performed before spreading widely the use of this SCPE R&TM. After a few years of use, the hope is that it will help to improve the harmonization and standardization level between different CP registers/studies, and that it will encourage new registers in new countries to join the SCPE network.

During the latest years of the SCPE, we have been still working on the data quality and also toward the improvement of available information on denominators within the EURO-PERISTAT project.

Results: SCPE CP definition and criteria: Several definitions of CP already exist in the literature. However although these may vary in wording, they are broadly similar, and can be summarized as follows:

Cerebral Palsy is a group of permanent, but not

unchanging, disorders of movement and/or posture and of motor function, which are due to a non-progressive interference, lesion, or abnormality of the developing/immature brain. This definition specifically excludes progressive disorders of motor function, defined as loss of skills previously acquired in the first 5 years of life.

For any study of CP to be valid, there must be agreement on the 'similar characteristics' of the cases eligible for inclusion. SCPE has spent time agreeing on inclusion and exclusion criteria that should accompany CP definition (SCPE 2000).

Since there are great variations in ability of performing diagnosis in different places, and techniques for these diagnosis are improving over time, the CP definition must be simple and rely on phenomenology (clinical picture and history) criteria and not on aetiology criteria. The CP definition must be valuable and logical for both epidemiologists and clinicians, and, by implication, must be independent of the country in which the child lives.

Inclusion criteria: <u>Optimal age</u>: CP is not an easy diagnosis. It needs time to be confirmed. Premature diagnosis might lead to over-ascertainment (because of transient anomalies in preterm babies) or under-ascertainment, e.g. in mild unilateral spastic cases or ataxic cases. CP, as stated above, is not an unchanging condition, with the clinical picture in some cases altering as a child develops. It was agreed that age 5 years was the optimal age for confirmation of diagnosis.

<u>What about children who die early?</u>: It is recognized that some children with severe CP are correctly diagnosed at a young age, but die before their 5th birthday. Exclusion of these children could result in under-estimation of the prevalence of CP in Europe. Also when studying the aetiology, it would be better to include these cases, for instance cases of hypoxic–ischemic encephalopathy who die early. In fact a compromise was needed, and as a group, SCPE had followed the recommendation from Hagberg that we should not include children with CP who die too early, i.e. before the age of 2 years, and that children with clear signs of CP who die between the ages of 2 and 5 years must be included.

<u>No upper age limit</u> of onset of CP (in children with a post-neonatal cause) was identified. But it is useful to isolate CP cases of post-neonatal origin, defined as cases arising from an aetiological event 27 completed days after birth.

Exclusion criteria: All progressive <u>conditions</u> resulting in loss of acquired skills are excluded. However, we recognize that some progressive disorders might be registered wrongly as CP, due to the delay required, in some circumstances, to confirm a diagnosis of progressive disorder. However, the proportion of these misdiagnosed CP cases does not represent more than a few per cent of all CP cases, at least in the SCPE data.

Children with hypotonia as the sole clinical feature and children with isolated spinal neural tube defects should also be excluded from the CP cases.

An interesting paper had suggested a list of conditions that should or should not be considered 'cerebral palsy' (Badawi et al. 1998). This led to numerous discussions between epidemiologists and clinicians, but finally SCPE has agreed not to adopt this classification system but to rely solely on the clinical features of a case to determine eligibility.

SCPE CP classification scheme: Classification means 'the basic cognitive process of distributing children with CP into classes or categories of the same type'. Different classification systems for CP serve different functions, but for epidemiological purposes, classifications systems based on clinical findings are currently the most widely used.

Drawing on published work, SCPE has classified CP into three main groups, which are based on clear neurological signs indicating pathology in the cerebral motor systems, e.g. spastic, ataxic, and dyskinetic CP.

All CP subtypes have an abnormal pattern of movement and posture in common.

<u>Spastic CP cases</u> have increased tone and pathological reflexes, either increased reflexes, e.g. hyper-reflexia or pyramidal signs, such as Babinski response. Increased tone in spasticity is characterized by an increased resistance which is velocity dependent (Sanger et al. 2003). A spastic catch is felt some time after onset of movement. Clonus is often associated with hyper-reflexia. It is considered pathological when it is prolonged or does not stop spontaneously. Pathological posturing of lower limbs is characterized by: (1) internal rotation of the hip; (2) hip adduction; and (3) equinus foot, resulting in a 'scissored' position.

<u>Dyskinetic CP cases</u> present involuntary, uncontrolled, recurring, and occasionally stereotyped movements. The primitive reflex patterns predominate, and the muscle tone is varying. SCPE uses dystonic and choreo-athetotic CP subtypes for subgrouping.

Dystonic CP is dominated by abnormal postures (may give the impression of hypokinesia) and hypertonia (tone fluctuating, but easily elicitable tone increase).

Characteristics are involuntary movements, distorted voluntary movements, and abnormal postures due to sustained muscle contractions (slow rotation, extension, flexion of body parts). Choreo-athetotic CP is dominated by: hyperkinesia and hypotonia (tone fluctuating, but mainly decreased).

Chorea means rapid involuntary, jerky, often fragmented movements. Athetosis means slower, constantly changing, writhing, or contorting movements.

In some cases, however, it may be difficult to delineate these subgroups when features are present from both. Then the term dyskinetic CP should be used.

<u>Ataxic CP cases</u> present loss of orderly muscular coordination, so that movements are performed with abnormal force, rhythm, and accuracy. Abnormal pattern of movement in ataxic CP is characterized by: (1) Loss of orderly muscular coordination, so that movements are performed with abnormal force, rhythm, and accuracy. Typical features are trunk and gait ataxia (disturbed balance) and past pointing (over- or undershooting of goal directed movements). (2) Tremor is another common sign (mainly a slow intention tremor). (3) Low tone is also a prominent feature.

<u>Mixed CP forms:</u> When it is a mixed CP form, i.e. spasticity with ataxia and/or dyskinesia, the child should be classified according to the dominant clinical feature.

Pure dyskinetic movement disorder does not show hyper-reflexia with clonus nor pyramidal signs. But in dyskinetic CP, these signs of spastic disorder may be present. The dominating features should determine subtype classification. Also, in spastic CP, some dystonic features are often present, especially when the upper extremities are involved. A dystonic posturing of the hand would, however, not be sufficient to classify a child as having the dystonic form of dyskinetic CP. The dystonic posturing of the trunk, arms, and face in the presence of lower-limb spasticity would qualify, however, as predominant dyskinetic features, thus, dystonic

CP (Krägeloh-Mann et al. 1993).

Motor function impairment in CP children: SCPE choice was to recommend the scoring of motor function according to: (1) the Gross Motor Function Classification System (GMFCS) for the lower limbs function (Palisano et al. 1997), http://www.canchild.ca; and (2) Bimanual Fine Motor Function (BFMF) for the upper limbs function. This last choice was achieved only very recently, and in order to conform with the S property (simplicity). A study has now shown the possibility to use this BFMF scoring through medical notes (Beckung and Hagberg 2002). However it has not yet been validated.

Since SCPE does not recommend the use of diplegia/quadriplegia terms, and recommends using instead the term bilateral spastic CP subtypes, the two motor function scales can then be used for describing children with CP according to the functional grading given. For instance, a child with bilateral spastic CP may be 'scored' as GMFCS Level IV and BFMF Level II – which for the clinician involved would give the feature of a diplegia – and another child with unilateral spastic CP may be scored as GMFCS Level II and BFMF Level I.

Associated impairments in CP children: The SCPE collaborative group recommends collecting information on four associated impairments. These recommendations are the minimum information that should be collected for those wishing to pool data or to compare it with data from other centres/countries.

Intellectual impairment: The cognitive impairment should be classified according to the thresholds recommended by the World Health Organization. These thresholds are shown in Table I.

Table I: Thresholds of cognitive impairment as classified by the World Health Organization

Normal	IQ>85, attendance of regular school without support
Borderline	IQ 70–84
Mild impairment	IQ 50–69, some basic literacy and numeracy achieved
Moderate to severe impairment	IQ 20–49
Profound impairment	IQ<20

For visual and hearing impairment, the recommendation is to determine the presence or absence of such impairment, and then to classify the impairment as severe or not, according to the visual acuity (<0,1 in both eyes after correction) or hearing loss (more than 70 dB in the better ear before correction).

Epilepsy can be defined as two unprovoked seizures, neonatal seizures being excluded. Firstly it must be known if the child 'had ever' or 'never had' epilepsy. Then it will be grouped as severe epilepsy if the epilepsy is still active.

Discussion: SCPE trees (SCPE 2000) are used for categorizing children with CP. Firstly, the decision tree is based on the presence of disorder of 'movement and/or posture' and of motor function. Secondly, the classification tree relies on neurological signs and topography for distinction between CP subtypes. By doing so, CP cases that are difficult to classify are not so numerous and less than 5% are observed in data from European centres (SCPE 2002).

At the beginning of the SCPE network, it was decided to use the words 'bilateral/unilateral spastic', with, in addition, the numbers of limbs involved, instead of using the words 'diplegia, tetraplegia'. After a while, the disappointment was great when we observed persisting important differences between centres on the 'theoretically' harmonized data. The overlap between the 'diplegia/quadriplegia' groups in CP classifications has been well described in a recent paper (Colver and Sethumadhavan 2003). These differences, between two and four limbs for example, could not be explained by anything else than by coding differences. Despite having agreed on a text definition and classification categories, large variations in classifying CP cases were still shown in a cross-validation exercise. The distinction between the number of limbs affected, used by several centres, in opposition to the number of limbs predominantly affected used by other centres, was the main reason responsible for these differences.

Thus SCPE's recommendation moved to a more simple categorization, i.e. classifying spastic CP cases in unilateral versus bilateral CP cases. Bilateral spastic CP was not further subdivided into arm/leg-dominated, diplegia/quadriplegia, nor 2-limb/3-limb/4-limb dominated, due to the great interrater variability when these terms are not defined using functional scores respectively for upper and lower limbs.

In a different way the Australian group gives an example that harmonization within one country may authorize more detailed description and classification than what is possible when dealing with several different countries. They are using four levels (minimal, mild, moderate, severe) to describe severity of neurological signs in each limb. However, there is still discussion in Australia about the overlap between triplegia, diplegia, and quadriplegia CP subtypes, and the need for an international consultation was expressed (Blair and Watson 2005).

We agree with Eve Blair's recent comment (2005) that, presently, no satisfactory scale is able to classify multiple deficits, i.e. the GMFCS score and, even more so, the BFMF score, are certainly influenced by the presence/absence of associated intellectual impairment, and thus they do not describe only the motor function. However, these scoring systems are very helpful for epidemiological purposes and evaluation of care.

The reasons for SCPE choosing the BFMF scoring system rather than the MACS (Eliasson et al. 2006) are that: (1) BFMF takes into account possible asymmetry in the hand functions, whilst MACS does not; and that (2) BFMF can be retrieved from written medical records whilst MACS cannot. When collecting data on children with CP for CP registers or surveys, the situation of not directly examining the child is quite common.

In the US there was an attempt to classify children with CP according to severity criteria based on the functional ability of the most affected limbs, i.e. severe involvement meaning no useful function, and moderate involvement meaning the preservation of some function with or without the use of assistive devices. In most studies using data from this California survey, the children with mild involvement or pure hypotonia were excluded (Grether et al. 1992).

The results of the workshop in Washington was an agreement on a more accurate CP definition, with details given for each word of the definition (Bax et al. 2005). This constitutes a great step forward, although this is probably not sufficient. There is a need for precision on inclusion/exclusion criteria

so that the definition can be used, not only as a concept, but also in a pragmatic approach. In the same manner that the GMFCS scoring system is proposed with video pieces and written explanations for training, CP definition should be provided with tools that help in applying the definition criteria. For instance within the SCPE network it was clear that there was a need for further training tools to clarify SCPE written classification guidelines with illustrations, i.e. using video description of children with CP, such as in the SCPE R&TM. Now this manual has been already translated into eight different European languages: French, German, Italian, Lithuanian, Swedish, Spanish, Slovenian, and Dutch, and it will soon be available in Portuguese. Also, alongside the decision and classification trees proposed by SCPE, an ideal data collection form might be very useful for harmonizing the available data used to classify the CP cases. The SCPE Data Collection Form (SCPE-DCF) is one of such possible standardized forms. It comprised the minimum common items that normally should be available for registration everywhere in each country. This form is available on the SCPE website (http://www-rheop.ujf-grenoble.fr/scpe2).

A very positive result of the SCPE harmonization work was to highlight interesting characteristics or trends in some subgroups of CP that needed large numbers before any analysis. After application of the inclusion/exclusion/classification criteria for CP cases, pooling data from several centres allows SCPE to show a four-fold increased risk of CP in multiple birth mainly explained by gestational age distribution (Topp et al. 2004), a decreasing trend of infection as cause of postneonatal CP case (Cans et al. 2004), an optimal birthweight associated with a lower risk of CP (Jarvis et al. 2003), and a decreasing CP prevalence in children with a birthweight between 1000 and 1500g (Platt et al. Forthcoming). Pooling data on CP cases already 'harmonized' was also very useful in a research project on quality of life and participation of children with CP (Colver 2006).

With its CP definition, criteria, and classification, SCPE has got agreement on a 'minimum data set or minimum description' of a child with CP, i.e. a common language which enabled us to build up a reliable database throughout Europe. In addition to this basic description, it is of course possible to go into more detail in some countries, and/or for specific substudies, according to different interests. This could concern more detailed description of the disability profile with respect to cognitive functions, or of the quality of life and participation of CP children and their family, or of additional orthopaedic problems or of information on aetiology/pathogenesis (neuroimaging, genetic findings).

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Future directions

From syndrome toward disease

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Syndromes are collections of symptoms, signs, and sometimes historical information, put together to group individuals and, hopefully, identify a disease entity. A disease is not defined only in terms of symptoms and signs but in knowledge, first of the pathophysiology of a condition and then of the cause (e.g. genetic, infections, etc.) of the disease. Identification of disease, hopefully, leads to effective treatment or prevention of disease but in many fields of medicine we are struggling still to identify what has actually caused the condition. Severe learning disability* is clearly a description of a syndrome but now many children, and indeed adults, with severe learning disability have had the cause recognized as a consequence of the great developments in the field of genetics in the past 2 decades. When thinking about cerebral palsy (CP), it should be recognized that some children who used to be diagnosed as having CP syndromically now prove to have a diagnosed genetic condition. An example is Rett's syndrome.

In recent decades, the development of neuroimaging, first CT and now with magnetic resonance imaging (MRI), has allowed more information to be obtained about the pathophysiology of conditions than was possible in the past when the only resource was pathological examination after the individual with the condition had died. While this provided indications of the anatomical nature of the deficits, it rarely helped towards any conclusion about the cause, partly because of the very restricted numbers of cases that could be examined.[1]

In the recent European collaborative study of CP involving eight centres, it was possible to examine over 350 MRI scans from a clinical population of some 430 cases. Bax's classic definition of CP[2] was used and the sub-classifications of CP used classically hemiplegia, spastic diplegia, athetoid CP, and ataxic CP. For a discussion of this classification see Bax and Keith Brown.[3] In this somewhat exploratory paper, the significance of some of the findings is reviewed.

Spastic diplegia

Briefly, this is a child with gross motor problems, particularly marked in the lower limbs with usually partially retained fine motor function in the upper limbs. Significant associated findings are reported. Many of these children had white matter damage of immaturity (WMDI) including periventricular leukomalacia (PVL) and periventricular haemorrhage. This form of damage in CP has been recognized from the time of Banker and Laroch but a clear account of the findings has not been made because of the lack of a suitable format for the systematic description of the findings on MRI. For the European study, Bax et al. devised a classificatory system.[4] Seventy-one per cent of the children with spastic diplegia proved to have WMDI but in many of them this was restricted to the posterior parts of the cerebrum and in relatively few was it extensive in anterior, middle, and posterior cerebrum (thus, although there was

some overlap, the findings in spastic quadriplegia were rather different *infra vide*). We still have to establish the definitive cause of the WMDI but with this pathophysiology it might be thought that we were moving to understanding CP with this established pathology producing a standard syndrome. However, WMDI is actually the cause of another group of disorders in childhood and more commonly causes visual disability rather than motor disability. Thus, Jacobson et al.[5] reported in 1998 that 27% of all visually-impaired children in the county of Varmland, Sweden had PVL as the reason for their visual disability, while a study in Finland[6] showed that 32% of preterm children in a Finnish population had MRI changes of PVL and a visual disability but only 9% had CP.

The nature of these serious visual disorders has not been clearly reported in CP but it is of interest, in thinking about the different types of visual problems which might be caused by PVL, to reflect on the recent findings within neuro-ophthalmology of the dorsal and the ventral streams (illustrated in Fig. 1) where we see the different functions of different parts of the visual cortex. The dorsal stream is involved with motor function while the ventral stream disorders affect perception and, for example, visual face recognition. Because of the clinical difficulties, few children with CP have a really thorough examination of the central visual system, in particular, for possible central visual problems. However, the psychologist Landmark in 1962[7] clearly describes the case of a 15-year-old female with spastic diplegia whose problems neatly fit Goodale and Milner's[8] account of the interaction of the dorsal and ventral streams. It is surely not too speculative to think that in the child with spastic diplegia, dorsal stream disorders may play a part in the disturbed motor function, and the ventral stream disorders in the perceptual problems seen in these children. Thus, it seems highly likely that in time we will conclude that at least some of the diplegic CPs have central visual problems.

Spastic quadriplegia

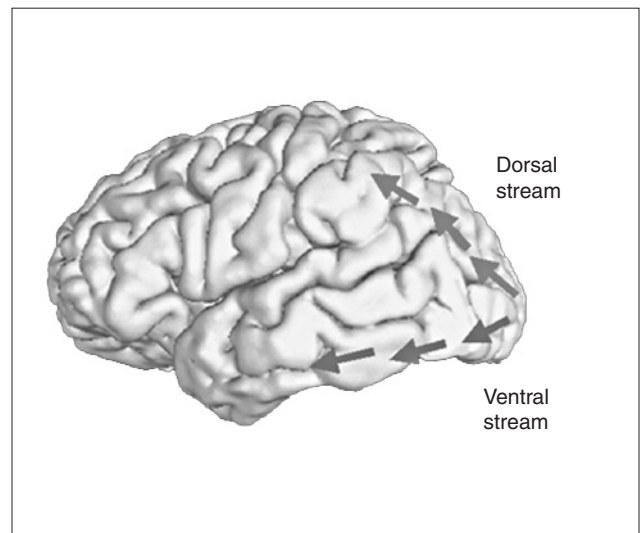In spastic quadriplegia, the MRI findings are rather different.

Figure 1: *Visual disorders.[8]*

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*North American usage: mental retardation.

While approximately 35% of the children had WMDI, this was always extensive insofar that milder posterior lesions did not occur. In addition to the PVL, cortical-subcortical and other lesions were found to cause spastic quadriplegia. In spastic quadriplegia, there are striking clinical differences with spastic diplegia. The children with spastic quadriplegia in general had severe motor involvement, Gross Motor Function Levels IV and V, virtually no hand movements, and many have very little speech and language, which contrasts sharply with the findings in those with diplegia. Communication is affected and these children, as is well known, have severe learning disability. We can sum up our thoughts about the consequences of WMDI and the cortical and subcortical lesions in the schematic diagram (Fig. 2): the WMDI causing spastic diplegia, central visual problems; the more severe WMDI causing spastic quadriplegia, as do cortical-subcortical lesions. But, in addition, these severe lesions cause severe learning disability. This should be regarded as the main or principal diagnosis in these children currently labelled spastic quadriplegia.

Hemiplegia

The child with hemiplegia traditionally has problems restricted to either one side or other of the brain. In fact, in our studies, only 27% of the children with hemiplegia had strokes causing their hemiplegia whereas 34% were caused by asymmetrical PVL. In some, MRI was reported as showing no signs of WMDI on the ipsilateral side and all but one of these had a diagnosis of hemiplegia. Clinically, these two groups were hard to separate. Children with hemiplegia, as previously described by many authors, had some difficulties often with the leg on the affected side, which may be shorter and may develop some contractures. However, with proper care, the child should usually be walking not much later than his normal peer group. Good management should mean that the gross motor problem is not of great severity. Hand function is more affected but the child would have the advantage of normal (or near normal) movement on the non-affected side. Clinically, we could not distinguish between the stroke and asymmetrical WMDI. One can speculate that, apart from the findings of spasticity, perhaps dorsal stream problems were playing a part in the motor disability of the hemiplegic hand. A recent finding in therapy perhaps supports this notion that, if the normal hand is restricted in its movement, improved movements can be achieved in the hemiplegic hand. It is interesting also to note that a number of children have been described clinically a long time ago by Richmond Paine as having a clear-cut hemiplegia at around 6 months but who appeared to recover around the age of 1 year.
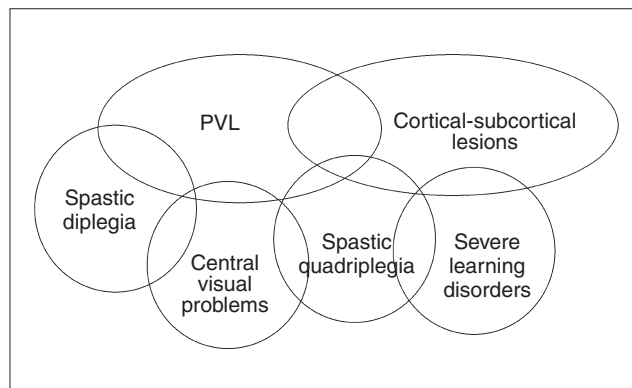
Thus, children with hemiplegia may have a 'relatively minor' motor disability but there are other features of the child. There are quite a lot of visual problems but most significantly there are behavioural problems. These have been well described by Goodman and Yude[9] who found that 50 to 60% of the children with hemiplegia in the study proved to have psychiatric and behavioural problems, stating that, 'Fears and worries are common particularly specific phobias, separation anxiety, and generalized anxiety. Misery was reported in some instances and is related to a depressive disorder. Conduct and oppositional defiant disorders are common. Tension and overactivity are common.' Goodman and Yude[9] stress that 'many children with hemiplegia have their lives, and particularly their education, curtailed by a mixture of fidgetiness, restlessness, poor concentration, and easy distractibility' which occurs at a much higher rate than in a normal population. While a small number fulfil all the criteria for diagnosis of autism, autistic 'features' are more common. Goodman and Yude[9] describe preoccupations and exaggeration of childhood interests, focus on specific cartoon characters to the exclusion of everything else, peculiar preoccupations with things such as a washing machine, lawnmowers, and yogurt pots. These are preoccupations associated with impoverished imaginative play. It is interesting to speculate about the causes of these but Goodman and Yude believe that 'brain factors are as important or more important than social or environmental factors'.

Athetoid cerebral palsy

With the less common athetoid or dystonic CP, the lesion is in the basal ganglia and they have disorganized movement patterns which, if severe, can be totally disabling. Speech production is often affected and some, but not all, individuals have preserved cognitive function. Athetoid CP, caused by kernicterus following rhesus incompatibility, is no longer common.

The pathophysiology found with the four main types of classically recognized CP have been briefly described and schematically represented in Figure 3. But what of the associated findings which are found in CP and which, in the new suggested definition (Rosenbaum et al. 2007), must be seen as very much part of the syndrome?

'Associated' findings

With the emphasis in CP on the motor disorders, many problems which may be causing the most significant difficulties are grouped together as 'associated difficulties'. Epilepsy occurred in our study in 28% of the sample, varying from a 'low' rate of around 16% in spastic diplegia to 50% in spastic quadriplegia. Communication problems occurred in 58%. When the children are at an older age they will no doubt be seen as part of cognitive deficits but again differently in the different diagnoses. Visual problems were noted in 42% and it is virtually certain that these are under-reported because of the difficulties with examination. Dutton and Jacobson[10] have reviewed cerebral visual impairment in children, its diagnosis, and management. We speculate that if good examination of the central visual system was possible, it would also be possible to identify problems with dorsal and ventral streams leading and relating

Figure 2: *Consequences of white matter damage of immaturity and subcortical lesions. PVL, periventricular leukomalacia.*

to both motor problems and perceptual problems. More severe damage involving both anterior and mid-brain structures will lead to severe learning difficulties. Note that behaviour/psychiatric problems are seen in the very different topographical damage found in the children with hemiplegia. Thus, in any single child with the pathologies described, a motor problem may or may not be present, while other problems may predominate. If we are to move ahead, earlier syndromic classifications are becoming less useful. Rather we should look at, for example, the whole group of children who have WMDI and by expanding the samples being studied it will be possible to get more ideas about the processes which lead to these findings, not only visual but motor. As the autistic syndrome is disentangled, we shall want to include children who have hemiplegias to see whether that helps in understanding the causes of that syndromic group, bearing in mind that already many children with autistic syndromes have been shown to have genetic causes such as Fragile X, Prader–Willi, Angelman, etc. In fact, genetics can be put as a cause of all the aspects of syndromic classification we have been discussing.

Rather than keeping to the syndromic classifications we have favoured in the past, we should try and establish the pathophysiology in any one child and look for the causes of that pathophysiology. For the moment we should list these children as having syndromes of cerebral dysfunction (Fig. 3). Although clinicians have talked and emphasized for years the 'multidisciplinary nature of the team' which helps the child with disabilities, it is still possible to visit 'CP Clinics' where the great emphasis is on the motor findings and not on all the functions of the child which we now know may be damaged by the pathophysiology. Similarly, there are Epilepsy Clinics where autism has not been recognized and Visual Disorder Clinics where only the visual disorder is studied. A broader approach will effect the training of health personnel who are involved in the field of care of these children. It will effect, I believe, the thinking of those who are looking for causes of the neurodevelopmental disorders.

In presenting this view we depart from the new definition/classification of CP proposed early in this supplement. We believe, the use of the term CP should be 'downsized'. A child's principal diagnosis is a neurodevelopmental disorder of childhood and within that we look at all aspects of the child's problems, not only motor but visual, perceptual, intellectual, hearing, abnormal behaviours, epilepsy, and autism and present a comprehensive view of the child.

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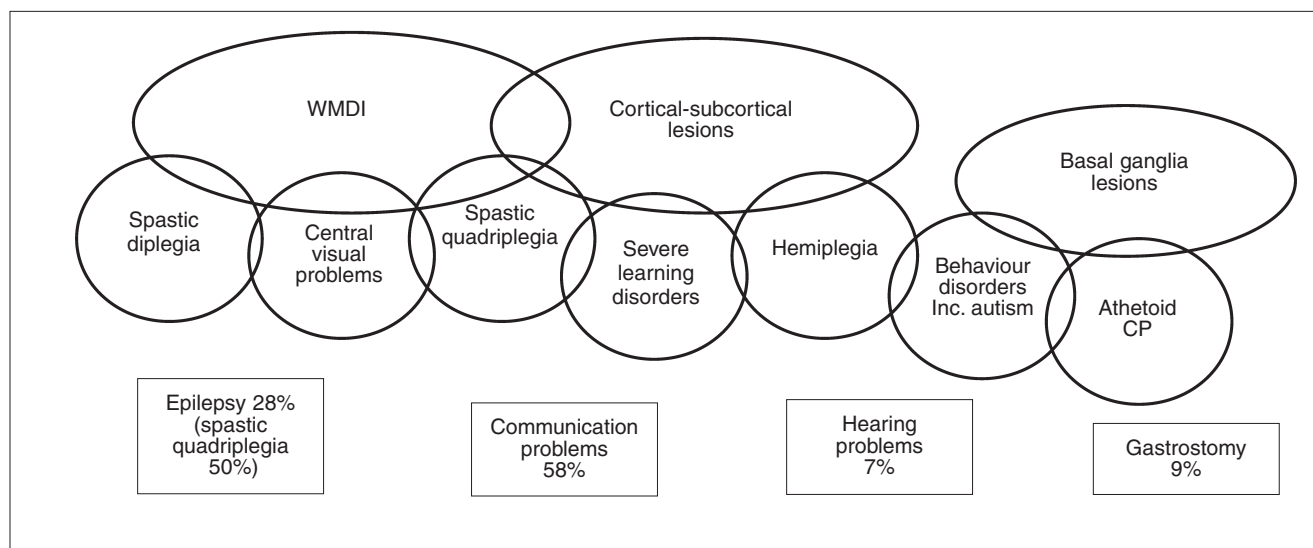10. Dutton GN, Jacobson LK. (2001) Cerebral visual impairment in children. *Semin Neonatol* **6:** 477–485.

Figure 3: *Neurodevelopmental disorders of children and adults. WDMI, white matter damage of immaturity; CP, cerebral palsy.*

Definition and classification of CP: medical-legal and service implications

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The implications of a revised definition and classification system for cerebral palsy (CP) obviously extend well beyond clinical care and research. My commentary will focus on two disparate areas of concern: specific topics related to medical-legal issues and brief comments concerning potential effects on service provision.

Important issues from the medical-legal perspective include the definition itself and issues relevant to causation in the classification section, specifically neuroimaging and etiological understanding. For nearly 150 years, causation theories for CP have been linked to intrapartum events.[1] Despite decades of epidemiological research to the contrary, the causal connection in the public mind between CP, 'brain damage', and obstetrical misadventure continues to encourage litigation with highly significant professional and economic implications for health care and society.[2–4]

Consensus publications identify spastic quadriplegic and/or dyskinetic CP as an essential criterion for consideration of intrapartum hypoxic–ischemic injury as the cause of later disability.[5,6] The revised definition does not affect this perspective. Despite initial concerns that the new term 'disturbances' might broaden the concept of CP to include children with developmental coordination or dyspraxic conditions, the full document clearly specifies patterns of motor disability as spastic, dyskinetic, or ataxic and thereby preserves the traditional designations and relationship between CP pattern and potential etiologies.

In the Classification section, the brief comments on neuroimaging are noteworthy. The committee correctly notes that the classification of CP by imaging criteria is still in development but forcefully endorses the previous recommendation of the American Academy of Neurology on the importance of such testing whenever feasible.[7] This statement cannot be over-emphasized in the medical-legal context. Currently, allegations of 'brain injury' do not require the demonstration of such injury or in fact, any anatomic abnormality. This unfortunately permits CP and other impairments to be presented as evidence of 'brain injury' a seriously uninformed perspective. Such a view fails to acknowledge the value of MRI in identifying injury patterns, and even more importantly, overlooks the significance of brain malformations and genetic/metabolic disorders in producing the CP phenotype.

The critical issues of causation and timing are also addressed directly in the revision document. The annotation on the definition states that 'a full understanding of causal pathways and mechanisms leading to CP remains elusive' and then amplifies this perspective in the section on Cause and Timing. In discussing causation, the current understanding of the importance of multiple interacting risk factors, the lack of a specific etiological relationship between adverse circumstances and CP, and the absence of a known cause in many cases are stated explicitly. This section concludes with the important caveat that, 'clinicians should avoid making the assumption that adverse events in the prenatal, perinatal, and postnatal life of a child with CP [are] sufficient to permit an etiological classification that implies a causal role for these events in the genesis of CP'.

In summary, the committee's statements provide an authoritative reminder about what is actually known and not known about causation and will be useful to those who are asked to render legal opinions in cases involving CP.

With respect to service provision, the diagnosis of CP is generally understood to indicate the presence of significant motor impairments which require educational and community-based services. The new definition maintains the central position of motor impairment but expands the focus considerably by emphasizing the importance of associated neurodevelopmental and musculoskeletal complications. The revision also places these impairments in a functional context by adopting the World Health Organization concept of 'activity limitation' as a defining criterion.[8]

These are significant modifications of the traditional focus on motor impairments alone and should have a major impact on services for those with CP. Care providers, educational institutions, advocacy groups, policy makers and others will need to adapt current approaches by placing increased emphasis on associated impairments as they consider the multiplicity of challenges facing individuals with CP. Such an expansion of the core concept of CP is likely to challenge current systems of care. Carried to their logical extent these changes in definition present rather daunting challenges and are likely to require significant modifications in the application of economic and manpower resources. Despite these practical difficulties, the revised definition presents a welcome opportunity to broaden the focus on CP. I believe that the net effect will be to stimulate progress on multiple fronts including neurobiological understanding, management approaches, and societal participation.

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Definition, classification, and the clinician

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Whilst it is acceptable to question whether the term 'cerebral palsy (CP)' should be discarded, the combination of time-hallowed usefulness and the absence of any form of consensus on a replacement or replacements effectively commits me to continue to use the label – but to be careful in its application.

Against that background and in clinical practice, the starting task for a paediatric neurologist is to provide a diagnostic formulation and within the context of this subject to attempt to answer the question, 'Does this child have CP?'

I, therefore, have to consider whether relevant criteria are satisfied, and in doing so, have to look at the presenting clinical features and then think about aetiology, pathology, investigations, interventions, and prognosis. I then have to communicate effectively with families and colleagues.

Agreed definitions with applicable criteria are enormously helpful and the revisions detailed in this supplement go a long way to at least reaching the clinical starting gate.

Thus, I am comfortable with accepting that we have to consider a heterogeneous collection of conditions which have in common clinically significant motor disorders, occurring as a consequence of non-progressive brain problems, as comprising either CP or the CP syndromes. I am assisted also by the reminder that the motor disorders evolve and have functional consequences in their own right.

However, definitions stand or fall by what they include and what they exclude – and this is where my difficulties start.

Two examples are illustrative. I see many children with global developmental retardation. Their major impairments are cognitive and social but they frequently have motor delay, hypotonia, and motor asymmetries. If I label them as having CP am I missing the point that their motor disorder is not their main problem? And similarly, does developmental coordination disorder – which may or may not be a clinical entity – exclude a diagnosis of CP? And if so, why? Specifically, am I denying that there is a continuum between those that are mildly affected and those with more severe impairments?

I do not think that it is right and proper to be wholly prescriptive on these issues and it follows for me as a clinician that any definition, whatever its degree of precision, has major limitations unless I supplement my identification of a child as having CP by then considering aetiological and other factors.

What this means in practice is that I feel obliged to continue to take into account all aspects of the history and the presentation and investigations including imaging before communicating my opinion that a child has CP with all that that implies. I am aware that not all services and clinicians have the resources to do this but surely this should not exclude the use of these data when they are available.

It follows that if definition goes beyond clinical description of the presentation then so also must classification.

Within that context I do have an anxiety that epidemiological considerations in CP are the tail that wags the clinical dog and that classifications based on topography and tone disorder may hide as much as they reveal. This is not to decry their importance for identifying hypotheses, influencing service development, and illustrating aspects of prognosis.

For example, there are enormous differences between a child born at 24 weeks' gestation and who has extensive cystic periventricular leucomalacia, and one born at term who has extensive cerebral hemispheric infarction of ischaemic origin, and one who has extensive neuronal migration abnormalities. All can present as having a four-limb CP with overt corticospinal tract dysfunction but any classification that masks these differences of pathology is unhelpful, particularly when identifying potential for future prevention.

My longstanding plea is that clinicians and epidemiologists should work more effectively together and that classification should make as much use as possible of all available information from clinicians. In return it is wholly reasonable that not only epidemiologists but also all those who require comprehensible communication from clinicians, not least families, should receive more than brief and often inaccurate diagnostic labels such as diplegia!

For my part, therefore, the re-examination of what constitutes definition and classification in CP is timely and welcome. I see it as an illustration of work in progress and that academic and professional interests are being appropriately applied to include all that is currently being learned in this field.