Chapter 1

What is cerebral palsy?

Overview

The newly crafted definition and classification of cerebral palsy (CP) represent an international effort to bring coherence to an aspect of childhood neurodevelopmental disability that has often been associated with rather imprecise thinking. This chapter discusses the details of this definition to highlight what we believe are key concepts regarding CP. Whether the current efforts will prove to be more useful than the earlier systems remains to be seen. The authors hope that people will use this definition and classification across clinical and geographical boundaries, thus moving the field forward using consistent concepts and terminology.

In clinical medicine there is a longstanding tradition of labelling and categorizing diseases and disorders. It is always felt to be important to separate conditions based on a combination of factors such as their clinical features (e.g. presentation, manifestations, natural history), their biomedical underpinnings (derived from varied investigations) and even their responses to interventions. The process of diagnosis is important to both physicians and patients, because knowing what ‘it’ is helps us all to focus our attention on the ‘right’ condition (and also to know what ‘it’ is not.).

Any group of professionals working in the field of childhood disability will each have their own working definition of CP. None will be identical but all will have common components, and our aim is to synthesize these various working definitions. Morris (2007) has considered the historical perspectives and definitions of CP, and the interested reader should make reference to his helpful summary.
In the summer of 2004, an international group of clinicians and researchers gathered for 2 days in Bethesda, MD, USA, to consider one of the perennial questions in the field of developmental disability. For the past 40 years, the definition of CP had been the classic 1964 statement that CP is ‘a disorder of movement and posture due to a defect or lesion of the immature brain’ (Bax 1964). Despite some modest but useful enhancement of these ideas by Mutch et al. in 1992 (CP is ‘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’), there remained uncertainty about both these specific definitions and, more generally, whether the term ‘CP’ had outlived its usefulness (Bax et al. 2007).

This 2004 meeting was jointly sponsored by the Castang Foundation of the UK and the United Cerebral Palsy Research and Educational Foundation of the USA. It was recognized that a host of factors in many fields of the clinical and biological sciences had increased our understanding of developmental neurobiology. The classic ‘developmental disability’ known as CP remains prevalent across the developed world, at a rate of 2–2.5 per 1000 people, and the rates are often rather higher in the developing world. For this reason, a reassessment of the concept and definition of CP was considered to be a useful undertaking.

The new definition, published in its final version in 2007, reads as follows:

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.

Rosenbaum et al. (2007)

The authors of this book contributed to this definition. We believe that there is still a place for differentiation of the idea of ‘CP’ as a clinical entity and as a ‘diagnosis’ (Rosenbloom 2007). For these reasons we will offer some thoughts about the ways in which this set of ideas may inform thinking not only about CP but about conditions across the spectrum of developmental disabilities.

We consider that it is important to examine the thinking behind the new definition and offer personal reflections and perspective. We argue that this revised and expanded definition may help people to understand the challenges inherent in trying to define conditions that are as much conceptual and phenomenological notions as they are biomedical entities. We also discuss some of the ‘rough edges’ of the concept of ‘CP’.

To illustrate these concepts, the early histories of two children with CP are briefly recounted. This is to emphasize that children with CP have varied early histories and frequently have more than just a motor disorder.
Child 1
AA was born at 37 weeks’ gestation. There was evidence of maternal hypertension and of restricted fetal growth in the later part of gestation. Labour commenced spontaneously, and during its course there was evidence of suboptimal fetal status from a pathological cardiotocograph trace and acidosis on fetal scalp sampling. Delivery was by emergency Caesarean section.

The infant’s birthweight and head circumference were between the third and tenth centiles. There was cardiorespiratory depression at the time of birth and depressed Apgar scores for the first 10 minutes in spite of prompt resuscitation. Thereafter there was a neonatal encephalopathy with seizures.

During the course of her first 2 years it became apparent that there was slow development of motor abilities, social responsiveness and language. There was uncertainty about visual functioning. There was slowing of head growth, with the circumference falling to below the third centile. Magnetic resonance brain imaging demonstrated extensive acquired white matter signal change in the cerebral hemispheres.

On examination at the age of 2 years, AA presented with a bilateral motor disorder considered to be CP. Motor skills were best classified as being Gross Motor Function Classification System (GMFCS) level V (Palisano et al. 2008) (the full GMFCS is reproduced in Appendix I). Muscle tone was primarily increased, with spasticity affecting all four limbs. Her weight was below the third centile and oral feeding was difficult and prolonged. Overall, a delay in development was confirmed.

AA’s history illustrates the need to recognize the relationship between her brain damage and the circumstances of gestation and labour, to appreciate that her disabilities extend way beyond her impairments of motor function and to deal with the implications of her need for lifetime care.

Child 2
BB was born at term after an uneventful gestation, labour and delivery. His birthweight and head size were close to the 50th centile and the neonatal period was uneventful.

At age 7 months he had a preference for using his left arm. By the time he was starting to stand at 14 months, it was evident that there was increased muscle tone in the right leg. He walked independently at age 15 months and was considered to have a right hemiparesis. Magnetic resonance brain imaging demonstrated focal infarction within the territory of the left middle cerebral artery.

At age 9 years he functions at GMFCS level I, and there is evidence of significant right-sided calf muscle hypertonus: he weight-bears on his toes on the right foot during walking. He is reluctant to use the right arm and hand, and is classified as functioning at level II on the Manual Ability Classification System (MACS) (Eliasson et al. 2006) (the full MACS is reproduced in Appendix II).
There is also concern because his educational progress is uneven, he has some behavioural difficulties and he has had two seizures.

Here the issues for consideration principally relate to understanding the significance of the totality of his motor impairments for independence and adult functioning, and to appreciate that his prognosis in these fields may be adversely affected by cognitive and behavioural difficulties and by epilepsy.

With these case illustrations as background, we present some issues that we believe should be considered when we talk about CP, and certainly when professionals engage in the assessment and management of children and young people with CP.

**Should cerebral palsy be considered a ‘disease’, a ‘diagnosis’, a ‘developmental disorder’ or a simply a ‘condition’?**

The first issue that requires consideration is the question of whether CP should be considered a ‘disease’, a ‘diagnosis’, a ‘developmental disorder’ or simply a ‘condition’. In so far as CP is known to be associated with a host of proven aetiological factors (e.g. brain malformations, kernicterus, maternal iodine deficiency) and established risk factors (e.g. preterm birth, being a twin, maternal genital tract infections, perinatal adversity), it seems clear that CP is not a single ‘disease’ in the way that type I diabetes or Duchenne muscular dystrophy might be so labelled. With today’s sensitive imaging techniques it is increasingly apparent that the nature, timing and distribution of brain impairments are all very varied and that the ‘clinical–pathological’ (in this case clinical–radiological) correlations are also varied and often distinct. Work by Bax and colleagues (2006) in a large European study of CP has shown that there are important relationships between the location of brain impairments (as identified by expert interpretation of magnetic resonance scans) and what might be called clinical ‘syndromes’ involving both motor function impairments and a host of other neurodevelopmental difficulties.

Given both the widely varying manifestations of CP and the many causal pathways thought to be important in its genesis (Stanley et al. 2000), the idea of referring to CP as a ‘disease’ seems somewhat problematic. What does a ‘diagnosis’ of CP mean? As outlined by any of the traditional definitions presented at the start of this chapter, the term describes impairment in the development of motor function and posture presenting early in life. Leaving aside for a moment the specific question of what ‘early’ means, it must be emphasized that all these definitions refer to aspects of a child’s gross motor development rather than to any specific biomedical marker by which the diagnosis can be confirmed or ruled out.

Although we argue that diagnosis is less precise in CP than in conditions with a discrete biomedical ‘cause’, it is certainly important to reach the formulation that this child has ‘CP’, to assess that child carefully as outlined elsewhere in this book, to communicate the findings empathically to the family, to plan and review interventions and to follow the child’s development on a long-term basis.
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There is often debate about how soon one can be comfortable about the ‘diagnosis’. Some argue that one cannot be certain that a child has CP before the age of 2 years, while others (including the authors) believe that in many cases – usually in those whose functional manifestations are more apparent (‘severe’) – one can identify and at least tentatively label the patterns of aberrant motor posture and function at 6 months of age or sometimes even sooner. Certainly a realistic approach for clinicians is to be prepared to identify and act on ‘variations’ in a child’s early motor development that compromise functional development (Rosenbaum 2006a,b). This can be done when there are problems with ‘quantity’ and/or ‘quality’ of motor behaviour by referring the infant or young child to the appropriate developmental services while continuing to follow the child’s progress. This should be done, among other activities, in order to evaluate the natural history of the child’s development and eventually to decide whether the child’s status actually fits the definition of ‘CP’. As Weindling (2008) has pointed out, it follows that it is difficult to predict CP at a presymptomatic level from, for example, the presence of abnormalities on neonatal cranial ultrasound studies. We support the ideas contained in the 2007 definition of CP that there should be some evidence of ‘activity limitation’ and not simply risk factors or the presence of isolated ‘impairments’ (problems in body structure – see Chapter 7) such as may be found in people with CP.

The excellent Surveillance of Cerebral Palsy in Europe (SCPE) group (Cans 2000) has offered one epidemiologically motivated compromise. Members of the many CP registries that contribute to this database make a tentative ascertainment of CP at any age, but only when the child has passed his or her fourth birthday will the SCPE members confirm for their register that the child has the clinical findings of CP, as outlined in their very useful manual (SCPE Reference and Training Manual). They argue that after age 4 years it should be clear whether the child’s earlier-identified functional problems have disappeared or whether they have progressed in ways that are phenomenologically inconsistent with the definition of CP. In the interim, of course, one would expect such children to be carefully monitored and their families offered appropriate developmental interventions, few of which are specific to CP.

What are the implications of considering cerebral palsy as a ‘developmental disability’?

What we believe to be the most significant contribution to the new definition of CP are the ideas contained in the second sentence (see p. 4). It is here that the various potential disorders of function, in addition to the ‘motor’ manifestations, have been formally identified as part of the spectrum of the CP picture. There is no implication that these functional difficulties are necessarily part of the condition (although frequently they are observed), but by explicitly including them the authors of the new definition have endeavoured to focus attention on the fact that many aspects of a child’s development may be associated with and impacted upon by ‘CP’, both primarily (as part of the impairment in brain structure and function) as well as secondarily (related to developmental challenges associated with the limitations in motor function).
The terminology in the newly crafted definition of CP highlights the *impact* of the condition on a child’s development, function and life trajectory as opposed to emphasizing solely the brain malfunction or ‘disease’ components of CP. This approach directly leads to thinking about CP (and many other early-onset childhood problems) in terms of how it actually or potentially alters the patterns of a child’s development. This idea distinguishes the important biomedical aspects of CP or other developmental conditions (e.g. issues of diagnosis, aetiology, genetic implications, ‘treatments’, etc.) from the challenges associated with ‘management’ and life-course perspectives.

Among these challenges is the question of the extent to which one ‘treats’ CP by addressing the primary ‘impairments’ associated with it (e.g. spasticity, hypertonicity, abnormalities of reflex function) and the extent to which one works to promote functional capability with techniques and equipment that may include ‘augmentative and alternative’ interventions. These latter interventions, when provided at developmentally appropriate times to accommodate children’s emerging interests in those aspects of function, may allow children to be functional despite the fact that progress is atypical, and despite the reality that the underlying neurological impairments may not be altered by these interventions. (Of course, when impairment-directed treatments such as botulinum toxin are indicated to moderate the effects of spasticity they should also be applied for the appropriate indications, without assuming that their use will, by themselves, necessarily change function [Wright et al. 2008].)

Butler’s work (Butler et al. 1984; Butler 1986) illustrates the value of alternative interventions in enhancing function and development. She demonstrated the remarkable influence that powered (‘augmented’) mobility had on children as young as 2 years and 6 months of age. The children’s language, exploration, social function and even their efforts at self-initiated movement were all profoundly impacted when they were provided with an external means to move. Control of powered mobility was something they learned to do independently within a relatively small number of hours of exposure and practice. This intervention did not address the basic impairments at all. Rather, it represented an ‘environmental’ manipulation that impacted upon the children’s lives in ways that no currently known ‘treatments’ could possibly do (Rosenbaum 2008). This work anticipated by almost 20 years the current ideas inherent in the World Health Organization’s International Classification of Functioning, Disability and Health (ICF) (World Health Organization 2001), about which more is written later in this book (see Chapter 7).

At the same time, it must be noted that there are still many traditional ‘orthodox’ impairment-based treatments that eschew the use of alternative interventions, such as walkers for mobility or sign language for communication of children with hearing or language impairments. The proponents of these approaches seem to want to direct therapeutic efforts exclusively at remediating the underlying impairment in order to promote ‘normal’ function. We argue that such thinking is excessively conservative in scope. It is unsupported by sound clinical research to bolster the thesis that promoting ‘normal’ function is a better approach than the more eclectic one recommended here, nor is there convincing evidence that it is even possible to ‘correct’ biomedical
impairments in ways that make a functional difference, thereby making it the right course of action to take.

The concept of cerebral palsy as a life-long condition
The third issue to highlight in the definition – implicit in the word ‘permanent’ in the first sentence of the new definition – is the concept that CP is a life-long condition. Like virtually all ‘neurodevelopmental’ disorders, CP has traditionally been thought of as a ‘childhood disease’. This is understandable in so far as CP and related neurodevelopmental conditions present in the very early years of life, and have always been recognized and managed within the child health systems of most countries. The reality, of course, is that mortality associated with these conditions is now very low (Strauss et al. 2007), and the vast majority of children with CP become adults with CP. Indeed, there are more adults with CP than children with CP. This challenging fact has important implications for people who have traditionally thought primarily or exclusively about ‘childhood’ disabilities.

Among the important realities is that in most parts of the world adult-focused services are unfamiliar with these ‘childhood conditions’. While CP is often superficially similar to stroke, acquired brain injury and other neuromotor disorders experienced by adults, people with CP bring to adulthood a life-long experience of development that is fundamentally different from that of people who have lived conventional lives until an adult condition interfered with their function. One of the implications of this difference is that people working in adult services may have little or no appreciation of these unusual life trajectories and ‘cultures’, and may be ill equipped to help adults with CP to fit in to the adult world.

It is also the case that the medical aspects of the lives of people with CP are often much less important than the social, vocational and community-living dimensions for which adults with CP need support. Underemployment and social isolation are often identified as key challenges for adults with CP and other neurodevelopmental disabilities – but the social and counselling services available to adults with CP are usually grossly inadequate in helping them. At the same time, there are important aspects of the processes of ageing with CP that need to be much better understood in order not to assume that anything that happens to an adult with CP can be ascribed simply to their underlying ‘childhood’ neuromotor condition (O’Brien and Rosenbloom 2009).

Another consideration regarding the adult component of the CP story is really a corollary of the first two – namely that traditional therapy efforts directed at the mobility and related ‘motor’ function dimensions of CP are much too narrowly focused to address life-course issues. If one sees CP as both a neurodevelopmental disorder and a permanent condition – as the new definition emphasizes – then it is essential that we reframe our goals for ‘therapy’, ‘treatment’ and ‘management’ during childhood to address the developmental implications of these conditions and that we focus our interventions to ensure that they are developmentally appropriate. This means grounding all our interventions within a broader canvas of life trajectories of children, working
towards promoting functional abilities and a sense of competence and capability, taking a strengths-based approach rather than continuing the tradition of cataloguing what disabled children cannot do. It also emphasizes the importance of continuity of thought and action across the lifespan to create seamless services as young people with CP move into the adult world.

The final implication of these ideas is, of course, that interventions should always be carried out within a family-centred service focus. Beyond the lip-service acknowledgement that most child health professionals would easily pay to this idea is the reality that parents need to be helped to understand that their child with CP has a ‘developmental’ challenge rather than perceive them solely as a child with a ‘disease’ (produced by ‘brain damage’) that must be ‘treated’. Parents need to be well informed about management options, and they need to be listened to with respect to their goals for their child’s development. An important randomized clinical trial from the Netherlands by Ketelaar and her colleagues (2001) showed powerfully that interventions directed at addressing parent- and child-identified functional goals led to better long-term functional outcomes than traditional impairment-based therapies – and with less intervention.

‘Rough edges’ in the definition of cerebral palsy
A fourth issue inherent in the new definition of CP is that there continue to be some ‘rough edges’ – those areas that remained controversial at the end of the 2004 consensus meeting. One question concerns the upper limit of the timing of ‘disturbances that occurred in the developing fetal or infant brain’ (Rosenbaum 2007). Children may acquire impairments of the central nervous system after birth owing to events as varied as brain trauma, central nervous system infections, asphyxia and cerebral malaria (in developing countries). The issue of the upper limit of the timing of causal events is perennially perplexing. This is true in part because from an epidemiological perspective it is important to frame the scope of the events that might be associated with the outcome ‘CP’ and might be relevant to the varied causal pathways that are being identified as potentially important to the genesis of CP. On the other hand, from a clinical and health services perspective, the identification, management and long-term follow-up of children with either early (‘congenital’) CP or so-called late CP (‘acquired’ in the first couple of years in the young developing child) should be similar and should focus on the ‘developmental’ aspects of this quintessentially ‘neurodevelopmental’ condition as these concern children and families.

Another element of the definition that has provoked considerable discussion over the years concerns the notion of ‘non-progressive’ disturbances. The issue is complicated in at least three important ways. Children with CP naturally change and develop over time, so that virtually all aspects of their lives and function may look different at different ages. Understanding the natural history of these changes and developments is important in order to distinguish ‘disease progression’ from ‘change and development’ inherent in childhood, even in the face of biological impairments that may constrain or inflect the patterns of developmental progress.
While there are classic neurodegenerative disorders of childhood that are clearly ‘progressive’, it may well be the case that some of the traditional, apparently ‘static’ encephalopathic conditions of infants and children are slowly ‘progressive’ in ways that have not previously been recognized. In addition, it seems likely that adults with CP age differently from other adults, related at least in part to factors such as ‘wear and tear’ on their systems, including the development of secondary conditions of muscles and joints. These accelerated changes in body structure and function may be associated with limitations in fitness imposed by challenges both intrinsic and extrinsic to people with CP (such as the paucity of community-based programmes and services available to these adults). If this is true then there may be important and as yet unrecognized opportunities for secondary prevention of the consequences of CP on adult well-being.

There appears to be no easy answer to this question about ‘progressive’ conditions, but the annotation accompanying the new definition states clearly: ‘Motor dysfunction which results from recognized progressive brain disorders is not considered CP.’ Clinical examples might include ataxia telangiectasia or Rett syndrome, each of which can, in its early manifestations, resemble CP. The previously described SCPE approach to the formal ascertainment of children as having CP at the age of 4 years makes it possible in most cases to identify children whose developmental course reflects a progressive as opposed to static condition, and who by definition are considered not to have ‘CP’. As noted earlier, the authors believe that at a clinical level one can and should identify, and offer help with, problems with a child’s motor development, whatever the specific biomedical diagnosis. Whether and when to intervene to address and to help children experiencing impaired function constitutes a clinical decision separate from whether the problem is eventually labelled as ‘CP’.

Another rough edge is the challenge associated with motor impairments in an affected individual that are considered modest or even subtle. At a diagnostic level, the question can then reasonably be raised as to whether it is still appropriate to use the term ‘CP’ to report the child’s condition. There can be no dogmatic answer to this question, especially if features such as very refractory epilepsy or profound cognitive impairment dominate the clinical picture. In practice we are comfortable with still using the CP label when the motor impairments are clinically apparent, even though they may be functionally of limited importance, but we do recognize that this is a grey area.

Classification of cerebral palsy

Finally, it is appropriate to consider briefly the question of how to classify CP – an issue that the Bethesda group addressed in its 2004 deliberations and which is more fully discussed in Chapter 4. Several basic points were identified. First, classification may be done for one or more of several reasons: to describe a child or a population; to predict future status; or to evaluate change in function. Second, there is a host of dimensions on which classification can be done, so it is important to be clear what purpose(s) one has in mind before applying any specific system. For example, traditional clinical descriptors of CP have focused on (1) topography (which parts of the body are involved); (2) the nature of the motor impairment(s) (whether the motor system is stiff and ‘spastic’, loose and ‘hypotonic’ or experiences fluctuations in motor control – ‘athetotic’, ‘ataxic’ or
'dystonic'); and (3) severity (traditionally characterized as ‘mild’, ‘moderate’ or ‘severe’, whatever those words mean).

For any measurement system to be useful (and classification is a form of measurement), it must be both reliable (provide consistent answers across observers and time when status is stable) and valid (meaningful and a reflection of the ‘truth’). It is important to note that few of the traditional classifications of CP have been shown to be either reliable or valid. Newer purpose-designed systems for people with CP have been created and validated for their ability to describe/discriminate both gross motor function (the GMFCS [Palisano et al. 1997, 2008]) and manual abilities (the MACS [Eliasson et al. 2006]). These systems (reproduced in Appendices I and II, respectively) are known to be acceptable to parents as well as service providers and researchers, and it has been shown repeatedly that the evaluations of parents and professionals are highly consistent with one another (Morris et al. 2004, 2006).

It was demonstrated by Gorter et al. (2004) not only that the GMFCS is the most (perhaps the only) reliable gross motor clinical classification system, but also that the various levels of the GMFCS each include children with a variety of topographic distributions and types of motor impairment. These latter characteristics overlap considerably with each other and seem to be unhelpful with respect to describing the functional aspects of a person’s CP. Thus, once again, it is important to observe that whatever classification systems are chosen, they must meet identified needs and be sound measurement tools if they are to fulfil the purposes to which they are put. These issues are discussed in more detail in Chapter 4.

To conclude, we reiterate our belief that the concept ‘CP’ remains a useful one for both clinical and epidemiological purposes. In particular, this approach allows us to talk with and counsel parents and families, and people with CP, reasonably clearly about a defined group of motor impairment syndromes for which the aetiology, natural history, management and life outcomes are becoming increasingly understood. We believe that abandoning this term would be counterproductive to all concerned.

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