CP or not CP?

Editorial

'Cerebral palsy' is an emotive term, which to parents often has the same impact as cancer or other dreaded diagnoses. For a child it is a label which can open doors to extra provision in schools and in the community, but which in itself may mean segregation, causing difficulties in participation. For a clinician it can be a diagnostic trap due to the failure to consider other conditions that impair motor function. For both clinicians and researchers it can be difficult to specify. Concerned about limitations of the current concept, a group of selected experts met last year to discuss the definition and classification of cerebral palsy. The preliminary results of their work are printed in this issue (p 571–576), with accompanying invited personal commentaries. We hope that it will provoke vigorous discussion.

The group highlights very clearly the limitations of our current approaches. This is well demonstrated in the section on functional motor abilities where they emphasize current deficiencies in the validated assessment of upper limb function, truncal involvement, and, especially, bulbar and oromotor function. Without doubt this approach will benefit epidemiological studies and, in addition, research and therapy teams will benefit from the focus on these wider effects, which look beyond the frequently overriding concern of whether a child will walk or not.

A major difference between the proposed new definition and the widely cited version[1] annotated by Martin Bax in 1964 is the substitution of 'non progressive disturbances that occurred in the developing fetal or infant brain' for 'defect or lesion of the immature brain'. 'Disturbances', as further defined, appears to include a wider pathological spectrum: in theory, embracing what is currently classified as developmental coordination disorder or motor dyspraxia. If so, this has huge implications. For example, cerebral palsy would be diagnosed in 50–90/1000 children instead of 2/1000 and the prevalence of different aetiologies would change.[2] There would be major effects on the distribution of resources, which should benefit children and families currently excluded from the diagnosis, but if those resources remain finite they will be stretched more thinly. In some countries the reimbursement schemes currently in place for therapy or medical and surgical care of children with cerebral palsy may end up being revised.

The classification retains the classic neurological terms for central motor disorders, spasticity, dyskinesia, and ataxia. These terms have both diagnostic and therapeutic implications, and so have a significance with respect to aetiology and, possibly, prevention. In a diagnostic sense, their original purpose was to help determine from the clinical features where in the central motor pathways a disturbance might be, and also to allow a differential diagnosis of possible causes.

For example, a child with upper motor neuron signs involving their legs but not their arms could have a disorder affecting the apices of the motor strips, white matter, central grey structures, the brainstem (especially at the foramen magnum), or the spinal cord. The disorder could be, for example, malformative, lesional, genetic, or metabolic in its nature. As the group points out, use of the term cerebral palsy can blur diagnostic thinking. More recently these terms have become important in determining which therapy might be indicated, such as botulinum toxin, intrathecal baclofen, or orthopaedic surgery.

More generally it is necessary to emphasize that all children who are considered to have cerebral palsy merit investigation, including neuroradiological studies where these are possible, in order to attempt to determine aetiology, to inform epidemiological studies, and to offer pointers towards prevention. An example is the described correlation between maternal infection, cytokine release, and damage to fetal brain white matter.

However, an important lesson is evident from dopa-responsive dystonia, which can present as 'spastic diplegia' under the current classification even though the pathogenesis appears extra pyramidal.

If this argument is taken to the limit it could even be suggested that in the interests of precision the term cerebral palsy should be abolished. Terms such as the ague, apoplexy, and, more recently, amaurotic idiocy have been surpassed when their limitations have become too obvious. Perhaps a child's disorder could be defined more specifically, as proposed by the group, in terms of the motor features, effects on function, and associated problems. This might also allow more appropriate interventions. Also, it might be easier to tell families that a child has a hemiparesis, and then explain the cause if possible, and avoid the emotive term 'cerebral palsy'.

However this term is an old friend and, through familiarity, still has a role. This includes unlocking facilities for an affected child and obtaining research funding. Originally it also acknowledged the difference between childhood and adult motor disorders, not just in aetiology but in their clinical expression over time due to brain maturation. The authors of this document and the working group deserve our thanks. Now it is up to us not to let '…their currents turn awry, And lose the name of action.'[3]

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References
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2. Kadesjo B, Gillberg C.(1999) Developmental coordination disorder in Swedish 7-year-old children. *J Am Acad Child Adolesc Psychiatry* **38:** 820–828.
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