Diagnosis and management of cerebral palsy

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The interest of clinicians in cerebral palsy has fluctuated markedly over the years and has been influenced serially by diagnostic considerations, classifications, epidemiological studies, aetiological and pathogenetic controversies, neonatal practice and follow up, and by the variety of patterns of care, treatment, and support available for children with neurodevelopmental disabilities and their families. It is against this evolving background that this review summarises the availability and significance of information that can lead to a diagnosis of cerebral palsy in young children and to its appropriate management.

Diagnostic considerations
Historically cerebral palsy has consistently been described as an evolving disorder of motor function secondary to there being non-progressive pathology of the immature brain. However, this begs important clinical, pathological, and aetiological questions.

From the clinical viewpoint it is unsatisfactory to apply a label of cerebral palsy when the movement disorder is either relatively subtle or alternatively is swamped. The former is exemplified by children who are clumsy. This is an important finding in its own right and in some cases may well have an aetiological and clinical continuum with cerebral palsy. This is not true for all clumsy children, however, and it is much more to their advantage to have their disabilities, including accompanying learning difficulties, accurately delineated as a prelude to appropriate help being provided.

Similarly it is not particularly useful to label a child as having cerebral palsy if there is, for example, some degree of hypotonia or spasticity but with this overwhelmed by there being a profound or severe degree of mental handicap. Within this context it needs to be kept in mind that severe hypotonia of central origin in infancy is a non-specific marker of neurological dysfunction and that for older children it almost certainly requires a significant degree of cortical function to be acquired before persisting hypotonia can evolve into spasticity. Thus in practice the description of hypotonic cerebral palsy, if it is to be used at all, will only be seen in association with very severe degrees of mental handicap.

It is preferable therefore to reserve the diagnosis of cerebral palsy to conditions where motor abnormalities dominate or at the very least are a prominent part of the clinical picture. Within the umbrella of this label it is also important that the details of the motor impairments be described as precisely and accurately as possible.

An increasing number of descriptive tools, for example, the motor assessment inventory, what has been described as the limb by limb approach, and the gross motor function measure are now available to do this. Their use in monitoring the progress of individual children is discussed presently.

Classification
From the perspective of early diagnosis classifications are of limited benefit as those which are of most value are based on defined rather than evolving syndromes. Nevertheless it is advantageous to appreciate the part played by an agreed system of classification in correlating epidemiological and clinical studies. The classification used in Sweden and reviewed recently by Hagberg and Hagberg is now widely accepted and is summarised as diplegia, tetraplegia, hemiplegia, dyskinetic and ataxic cerebral palsy.

Synonyms for tetraplegia include quadriplegia and double hemiplegia. Children are assigned to this grouping when they have severe spasticity of all four limbs, the upper limb disability being the same or worse than that in the lower limbs. In diplegia by contrast, there is lesser involvement of the upper limbs, although when present this may have an ataxic as well as a spastic component. In practice it may be difficult to decide where diplegia ends and tetraplegia begins and this difficulty is compounded by North American practice where many children who are labelled quadriplegic would in Europe be considered to have a diplegia.

The dyskinetic group includes those with fluctuating dystonia as well as involuntary movements of choreoathetoid type. Where there are mixed forms of motor disorder, the predominating one is used in classification.

Epidemiology
Studies from Sweden, Australia, and the UK have been very helpful in setting the current scene with regard to the prevalence of the various cerebral palsy syndromes and how this is changing. What is particularly noteworthy is that there is an increasing proportion of children born very prematurely within the total cerebral palsy population and this is a
direct consequence of there being increasing numbers of preterm survivors, a constant proportion of whom have impairments. Moreover, within that population are many children with severe motor disabilities and additional neurodevelopmental problems and this has very major implications for future resource and service provision. What is not yet known is whether the increased morbidity of preterm infants is a consequence of their surviving with prenatally determined abnormalities or whether their problems are purely or primarily a consequence of prematurity.

The epidemiological relationship between birth asphyxia and cerebral palsy also requires elucidation. Case studies series commencing with that of Little have linked perinatal events with cerebral palsy in a non-specific way and often with systematic errors built into the research.10 Cohort studies have also been used extensively in which markers of asphyxia, for example, birth acidosis have in general failed to yield increased numbers of children with cerebral palsy. This might well have been expected given that only a small increase could have been anticipated, although it is of interest that an Apgar score of 3 or less at 20 minutes is associated with a 250-fold increased risk of cerebral palsy.11

Alternatively case-control studies have been used in which children with cerebral palsy are compared with controls for markers of asphyxia,12,13 and it is from these that the figure of approximately 10% for an asphyxial cause for cerebral palsy in children born at term has been derived. It is relevant that a much smaller proportion than this can be demonstrated to have received suboptimal perinatal care.

The other epidemiological method used is that of randomised controlled trials, for example, that of intrapartum monitoring used in Dublin.14 These hitherto have failed to show that differences in obstetric care are reflected in reduced numbers of children with cerebral palsy but again they have the disadvantage of not having been large enough for any measurable change in outcome to be likely.

Aetiologies and pathogenesis

DIPLEGIA

The underlying mechanisms are those of periventricular leukomalacia and periventricular haemorrhagic venous infarction.15 The diplegic syndromes are characteristically seen in infants born prematurely with haemorrhagic infarction of the periventricular areas being seen especially in the increasing numbers of very preterm survivors. Radiological support for this concept is available from magnetic resonance imaging studies.16

Thus diplegia does not correlate with birth asphyxia; rather its likely cause is hypoperfusion of periventricular structures at the time in gestation when these are most vulnerable.

TETRAPLEGIA

Within the terms of the classification detailed above, these are the most catastrophically disabled of individuals with cerebral palsy, the vast majority being severely mentally handicapped and epileptic with bulbar palsies, microcephaly, growth failure, and visual defects being common. The syndrome is seen in both preterm and term infants and in the former is often accompanied by a haemorrhagic hydrocephalus. A wide variety of pathologies including malformations, intrauterine infections, fetal encephalopathies, and perinatal hypoxic ischaemic encephalopathy can all manifest themselves ultimately in this way. So also can genetically determined disorders and particularly when a child presents with a symmetrical tetraplegia in the absence of significant perinatal abnormalities, it is appropriate to estimate an empirical recurrence risk of one in eight to one in 10.

Neuropathology and neuroimaging may show a widespread multicystic encephalopathy,17 often with cortical and subcortical atrophy and gliosis,18 when there is a history of perinatal adversity.

HEMIPLEGIA

This is the second commonest syndrome seen in children born preterm and the commonest in term infants.2 Preterm hemiplegia has a non-specific association with birth problems.19 By contrast term hemiplegia most commonly results from events early in the third trimester involving hypoperfusion,20 although malformations and late events leading to infarction within the distribution of a middle cerebral artery are not uncommonly seen.

DYSKINETIC CEREBRAL PALSY

In current paediatric practice, dyskinetic cerebral palsy occurs most often in term infants and has a close association with obvious perinatal adversities. Most frequently this is unforeseen acute circulatory failure with severe but short lived birth asphyxia, although often only a mild or moderate hypoxic ischaemic encephalopathy.21 From the neuropathological and neuroradiological perspectives there is ample evidence of basal ganglia pathology as being the basis of the clinical abnormalities that result.22,23

These characteristics are of the dystonic type producing a very severe degree of motor disability with preservation of primitive neonatal reflex patterns. In other survivors of severe and prolonged birth asphyxia, there can be both basal ganglia and cortical and subcortical damage demonstrated with a resulting mixed clinical picture.24

The choreoathetoid form of dyskinetic form of cerebral palsy is, by contrast, now less common and this parallels the virtual eradication of bilirubin encephalopathy.

ATAXIC CEREBRAL PALSY

As might be predicted there is often clinical difficulty in distinguishing congenital ataxic syndromes from the heredodegenerative ataxias in older children. Most congenital ataxias
have a prenatal origin that is often genetic, although acquired haemorrhagic cerebellar lesions have been described.15

Diagnosis of cerebral palsy
Neurological abnormalities should always be routinely sought as part of neonatal care and this applies especially when infants are significantly premature or are recognised as having been subject to other perinatal hazards. Clinical changes that are found can often be correlated with concurrent neuropathological, neurophysiological, and radiological findings while the newer techniques of magnetic resonance spectroscopy and near infrared spectroscopy25 offer the prospects of additional non-invasive assessment measures of cerebral oxidative metabolism. However, currently available neonatal evaluation techniques have a less well established relationship with neurological and developmental abnormalities seen in later infancy and childhood.

So far as specific clinical findings are concerned, these have been well summarised by Dubowitz who has detailed the common observations seen in infants who are small for their gestational age, those who survive significant intraventricular bleeding, the evolving picture seen in periventricular leukomalacia, and the clinical stages of hypoxic ischaemic encephalopathy.26 She rightly emphasises the requirement to use age appropriate techniques and for examinations to be performed repeatedly with nevertheless there being continuing difficulty in correlating focal lesions with localising signs. Nevertheless, persistent generalised disturbances of tone, seizures, continued irritability or decreased alertness, persistent asymmetries of posture and movement, and delay in establishing efficient feeding are all in favour of affected infants continuing to be neurologically abnormal beyond the neonatal period.

A combination of clinical and investigative features in the neonatal period should determine the details of follow up. Crucially the aims of this are to ensure optimal progress and health of infants, to detect adverse sequelae including all types of neurodevelopmental disorders, and to initiate appropriate help. The details of relevant professional involvement require to be determined locally.

The role of investigations
Irrespective of neonatal findings a low threshold of suspicion is required for infants who show evidence of developmental delay (corrected when necessary for prematurity), persistent hypotonia, evolving dystonia or spasticity, focal abnormalities of movement, posture or tone, a decreased rate of head growth, abnormal visual or auditory behaviour, and seizures.

Under these circumstances and against a background wherever possible of clear explanations to the parents, it is possible for a provisional diagnosis of a neurodevelopmental disorder to be made, for appropriate investigations to be initiated, and for treatment for the child and support for the parents to be offered. Where the putative diagnosis is one of the cerebral palsy syndromes the investigations to be performed are determined by the past history and no further tests at all might be indicated. In other circumstances genetic studies, neuroimaging, and biochemical tests may be relevant especially if there is no certainty that the pathology is non-progressive.

Detailed chromosomal analysis is always indicated when a malformation syndrome is suspected, when the child has dysmorphic features, or when there is a positive family history. This may include the use of specific techniques, for example, to identify fragile X syndrome or Angelman’s syndrome. Computed tomographic or magnetic resonance brain imaging may be indicated if a progressive neurological disorder is possible or to confirm to parents (or sometimes their legal advisers) the nature and timing of acquired brain lesions. The state of this art18 23 24 is considerably in advance of conventional British and European practice because of the limited availability of magnetic resonance imaging and the need for infants to be sedated or anaesthetised for this procedure.

Biochemical studies can be limited to thyroid function tests to exclude occult hypothyroidism and amino acid chromatography unless children present with progressive or fluctuating neurological abnormalities.

Assessment of motor function
Historically the parameters for judging the severity of cerebral palsy have been subjective but a number of more objective measures have become available relatively recently including the motor assessment inventory3 and the limb by limb approach.4 Hallam and her colleagues22 have recently compared both these measures with one another and with the Griffiths’s locomotor development quotient28 in respect of a population of vulnerable children. They concluded that the limb by limb approach, which provides information on the type, distribution, and severity of motor involvement but not on a child’s movement abilities had the greatest sensitivity and was easy to perform. It may very well be, however, that two other novel assessment methods, the gross motor function measure5 and the gross motor performance measure,29 will provide significant advantages.

The gross motor function measure looks at five functional areas of lying, sitting, crawling, standing, and walking on a four point scale for each while the gross motor performance measure, currently a research tool, is endeavouring to examine the quality of movement demonstrated by children with disabilities.

Motor function assessment for children with cerebral palsy has also to be set within a wider developmental context and appropriate measures for looking at cognitive, social, and linguistic skills and sensory functioning require to be applied when the abilities of children with cerebral palsy are examined. However, few standardised tests for these measures are
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available. Clinical pointers such as eye pointing and the rapidity and appropriateness of social responses may be important while the increasing availability and use of computers with specialised switching is likely to become particularly advantageous in the near future.

Physical treatment
The provision of appropriate physiotherapy programmes is a mainstay of treatment for young children with cerebral palsy and sooner or later it is then usual for physiotherapy to be incorporated into the broader curricular requirements of children with physical disabilities. It follows in logic, if not in established fact, that appropriate physical treatment will lessen the effects of these. However, the methodology required to demonstrate this has proved difficult to develop and given also the emotional investment of parent in physiotherapy availability for their affected children it is not surprising that objective measures of its effectiveness have proved elusive.

Available methodologies were described by Scrutton and limited studies of the effectiveness of varieties of neurodevelopmental treatment have been presented by a number of groups of workers. Results in all of these studies have been indeterminate because of the small groups of children involved and limited duration of follow up. Alternative methods of physical treatment have some support often as a consequence of media involvement. In the UK; conductive education is one such approach but in the only published outcome study its effectiveness was not established.

However, it is within the twin contexts of cerebral palsy as an evolving clinical disorder and appropriate multidisciplinary practice, that promoting motor function and limiting motor impairments as a consequence of physiotherapy may require to be placed, rather than that physiotherapy should be regarded as a panacea by either families or therapists. Moreover drug treatments for spasticity with agents such as baclofen or botulinum toxin, the provision of relevant orthoses and other appliances, and the place of gait analysis in determining the appropriateness of orthopaedic and neurosurgical procedures have also to be considered within these contexts.

Current practice and future perspectives
There is now ample evidence that the vast majority of children with cerebral palsy should have this recognised in early infancy. Thereafter parental support and counselling, treatment of reversible conditions, for example, hearing loss and seizure disorders, and limiting the secondary developmental difficulties, for example, in learning and play opportunities or development of contractures are wholly reasonable consequences that should follow early diagnosis.

Specifically, it is now established that there is a necessity for the clinical and radiological surveillance of hip stability, for the early detection and treatment of scoliosis, for the recognition and treatment of drooling, and for there to be an awareness of neurological bladder involvement in cerebral palsy.

It should also follow that more relevant targets for cerebral palsy research and service provision can be set. Appreciable outcomes for services should include evaluation of motor developmental achievements, the prevention of avoidable deformities, achieving satisfactory nutrition and growth for children with cerebral palsy, provision of relevant appliances including communication aids, and the availability of suitable curricula and teaching methods.

Proceeding through childhood and appreciating that the majority of those with cerebral palsy become adults it could reasonably be claimed that it is effective adaptation to adult life, with the provision of appropriate resources for this, that is the ultimate outcome measure for cerebral palsy that requires satisfaction.

It is only in this way that the major advances in knowledge concerning the aetiology and pathogenesis of the cerebral palsy described at the beginning of this review, can serve as a foundation for alleviating the long term disabilities that arise as a consequence of their occurrence.


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