The term diplegia should be abandoned

A F Colver, T Sethumadhavan

Use of the term has served to confuse classification and obscure interpretation of epidemiological and clinical studies

From the writings of Freud onwards there is broad agreement on the definition of cerebral palsy (CP) but attempts to classify it, based on brain pathology, timing of postulated insult, aetiology, or clinical syndrome, taking account of one or more of neurological findings, distribution, and associated impairments, have been less successful. The term must be clearly defined, meaningful, reliable, and used consistently by different people. A few CP syndromes such as choreoathetosis with deafness caused by bilirubin encephalopathy and ataxia caused by hydrocephalus have stood the test of time and are reviewed by Ingram. However, we think that diplegia is not a description of a valid category or syndrome and use of the term has served to confuse classification and obscure interpretation of epidemiological and clinical studies. We draw on historical papers and our own new analyses of recent published epidemiological papers to argue that the term diplegia should be abandoned.

HISTORICAL OVERVIEW OF USE OF TERM DIPLEGIA

Table 1 summarises important classifications of the past 150 years which we shall discuss with respect to their use of the term diplegia.

William Little first described the syndrome complex of cerebral palsy in 1862. His paper proposed a link between abnormal parturition, difficult labour, premature birth, asphyxia neonatorum, and physical deformities, which he described lucidly. He did not use the term diplegia.

In 1890, Sachs and Peterson proposed a classification which linked clinical syndromes to timing of the insult, and introduced diplegia and paraplegia as separate categories. In 1893, Freud considered cerebral palsy to be caused not just at parturition but also sooner in pregnancy because of “deeper effects that influenced the development of the foetus”. Freud was the first to use the term “cerebral diplegia”, which covered all bilateral cerebral palsies, including non-spastic types.

Further classifications appeared in the 1950s. As table 1 shows, Minear’s was more a descriptive grid than a classification, and for him diplegia was “paralysis affecting like parts on either side of the body”.

In Ingram’s classification of 1955, spastic diplegia was described “as a condition of more or less symmetrical paresis of cerebral origin more severe in the lower limbs than the upper and dating from birth or shortly thereafter”. He qualified the disorder with an association with prematurity and lesser incidence of mental retardation, pseudobulbar palsy, and seizures compared to quadriplegia. He also described its clinical evolution through the stages of hypotonia, dystonia, and rigidity. Later in his 1966 review article “The neurology of cerebral palsy”, he re-emphasised what he regarded as the unequivocal distinction between spastic diplegia and other bilateral cerebral palsies.

In 1959 in England, The Little Club presented a definition and classification of cerebral palsy: “In diplegia there is affection of the muscles of all four limbs. The lower limbs are the more affected.” The classification had an additional category “atonic diplegia”, but the reasoning presented in the article for this category and other aspects of the classification are difficult to follow. By 1964 the problems were apparent, and an annotation recommended that description should be based on clinical features and that attempts to define certain syndromes combining clinical aetiological and pathological features should be avoided. In particular the idea of a diplegic syndrome should be avoided.

In their first epidemiological report in 1975, the Hagbergs’ used a classification with a definition of diplegia similar to Ingram’s. Hypertonic cases were diplegic where the lower limbs were more affected than the upper ones. Cases were regarded as diplegic even when they exhibited severe generalised damage and “might have been classified as tetraplegia by other investigators”. Their classification had an additional category of “ataxic diplegia” where the children had diplegia with ataxic traits, especially dysmetria and intention tremor in the upper limbs. However, by 1989, Hagberg appreciated the limitations of his own classification—particularly the unsatisfactory nature of the dividing line between diplegia and quadriplegia and the fact that many children change categories as they grow older.

In 2000, the European Collaboration SCPE did not include diplegia in its classification because of the inherent ambiguities.

Not surprisingly, all this confusion is mirrored in the standard paediatric textbooks. Forfar and Arnell regard diplegia as a clear entity in which the legs are more affected than the arms; the more affected the upper limbs, the lower is intelligence. It is linked to specific cerebral pathologies, which are themselves linked to premature delivery or asphyxia. In Nelson, diplegia is spasticity of just the legs, with good prognosis for intelligence and seizures. The lesions in the brain are similar in diplegia and quadriplegia, except for more extensive necrotic degeneration of white matter which coalesces into cystic cavities in quadriplegia.

In Rudolph, diplegia requires the legs to have greater spasticity and weakness than the arms. The children are frequently preterm and the deficit is more apparent in wrist extensors and activities of daily living such as self feeding, drawing, or writing. Quadriplegia is determined by symmetric impairment of all four extremities; growth retardation, mental retardation, language disorders, and seizures are common. Avery describes spastic diplegia as restricted to bilateral spastic involvement of the lower extremities, often associated with normal cognitive function. Quadriplegia is spastic involvement of the extremities, often with orobucal muscle atrophy, swallowing and talking difficult.

THE PRESENT SITUATION

The most recent International Classification of Diseases has a category “spastic diplegia” which it does not further define and another category “diplegia of upper limbs”. However the common ground in the papers we have just discussed appears to be that diplegia is a spastic form of cerebral palsy with lower limbs more affected than upper limbs. It is however unclear whether presence or absence of prematurity, seizures, or mental retardation is relevant to the definition or just an association. For example, if a child has severe four limb involvement with upper limbs slightly less affected than lower limbs, does presence or absence of the above features such as mental retardation determine whether this is classified as diplegia or quadriplegia?

Even more importantly, what does “more affected” mean and should the comparison be based on clinical signs or function? If comparison is based on...
<table>
<thead>
<tr>
<th>Reference</th>
<th>Year</th>
<th>Classification</th>
</tr>
</thead>
<tbody>
<tr>
<td>Little²</td>
<td>1862</td>
<td>Hemiplegic rigidity</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Paraplegic rigidity</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Generalised rigidity</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Disordered movements without rigidity</td>
</tr>
<tr>
<td>Sachs and Petersen³</td>
<td>1890</td>
<td>Paralysis of intrauterine origin</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Birth palsies</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Acute acquired palsies</td>
</tr>
<tr>
<td>Freud⁴</td>
<td>1893</td>
<td>Unilateral disorders—hemiplegia</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Bilateral disorders—diplegia</td>
</tr>
<tr>
<td>Wyllie⁵</td>
<td>1951</td>
<td>Congenital symmetric diplegia</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Congenital paraplegia</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Quadriplegia or bilateral hemiplegia</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Hemiplegia with additional qualifications referring to all categories</td>
</tr>
<tr>
<td>Minear⁶</td>
<td>1956</td>
<td>A. Physiological</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Spathicity, athetosis, rigidity, ataxia, tremor, atonia, mixed, unclassified</td>
</tr>
<tr>
<td></td>
<td></td>
<td>B. Topographical</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Monoplegia, diplegia, paraplegia, hemiplegia, triplegia, quadriplegia</td>
</tr>
<tr>
<td></td>
<td></td>
<td>C. Aetiological</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Prenatal, natal anoxia, postnatal, cause described</td>
</tr>
<tr>
<td></td>
<td></td>
<td>D. Trauma</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Cause described</td>
</tr>
<tr>
<td></td>
<td></td>
<td>E. Supplemental</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Psychological evaluation</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Physical status, convulsive seizures, posture and locomotive behaviour pattern, eye-hand behaviour pattern, visual status, auditory status, speech disturbances</td>
</tr>
<tr>
<td></td>
<td></td>
<td>F. Neuroanatomical</td>
</tr>
<tr>
<td></td>
<td></td>
<td>G. Functional capacity</td>
</tr>
<tr>
<td></td>
<td></td>
<td>H. Therapeutic</td>
</tr>
<tr>
<td>Ingram⁷</td>
<td>1955</td>
<td>Hemiplegia</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Right or left</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Diplegia</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Hypotonic</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Dystonic</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Rigid or Spastic</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Ataxia</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Cerebellar</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Vestibular</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Ataxic diplegia</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Hypotonic</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Spastic</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Dyskinesia</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Dystonic</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Chorea-thetoid</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Athetoid</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Tension</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Tremor</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Other</td>
</tr>
<tr>
<td>Little Club⁸</td>
<td>1959</td>
<td>Spastic cerebral palsy</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Hemiplegia</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Diplegia</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Double hemiplegia</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Dystonic cerebral palsy</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Chorea-thetoid cerebral palsy</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Mixed forms of cerebral palsy</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Ataxic cerebral palsy</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Atonic diplegia</td>
</tr>
</tbody>
</table>
clinical signs, tone, tendon reflexes, and power may be easy to separate at the extremes but very difficult when similar. Signs also vary day to day and with the mood of the child. Further, there is a considerable inter- and intraobserver diagnostic variability in the assessment of clinical signs.\(^2\) Ashworth developed a scale, tested for reliability—but this validation was in adults and just for hemiplegia.\(^2\) If comparison is based on function, again this may be straightforward at the extremes but otherwise very difficult. For example, does “walking with difficulty and needing walking aids” mean legs are more or less affected than arms where the child “is not able to write tidily and needs assistance to go to the toilet”?\(^2\)

The North of England Collaborative Cerebral Palsy Survey\(^3\) found such confusion in the application of clinical signs and assessment of functional severity. A local audit of classification of cases of cerebral palsy revealed 98 with diplegia and 130 with quadriplegia. Of those with diplegia, 58 had moderate to severe functional involvement of upper limbs at age 5 but lower limb function was always the same or worse. Five of these children had severe mental retardation. Of those with quadriplegia, six had nil to mild involvement of the upper limbs, but these six had severe mental retardation, suggesting that the clinician used the presence of severe mental retardation to assign the cerebral palsy to the category quadriplegia.

The term diplegia continues to be used in studies which assume its meaning is understood and uniformly applied. For instance, it is claimed that diplegia is associated with and may be caused by premature birth.\(^2\) In this study, Powell et al even postulate the existence of a “diplegia factor” which tends to cause premature labour; and impair growth and development if the fetus remains in utero. Their study does not define diplegia and uses the clinical diagnoses from cases notes of children looked after by many paediatricians. In another more recent study, it is suggested that chorioamnionitis may be the cause of spastic diplegia in low birth weight infants—especially in babies with a normal neonatal ultrasound—but again diplegia is not defined.\(^3\)

NEW ANALYSES OF EPIDEMIOLOGICAL STUDIES

We shall show that the confusion, which is clear from our review of papers describing the definitions and practical problems in making sense of them, is born out by our review and further analysis of recent epidemiological studies.

Table 2 shows the overall numbers of cases of cerebral palsy and percentages of different subtypes in studies reported from about 1980—which we take as the date by which neonatal intensive care had been introduced in the reporting regions. We have only included studies where all types and severities of cerebral palsy are reported, and where a whole population is reported either on the basis of birth cohort prevalence or population prevalence. It shows that spastic cases as

### Table 1 continued

<table>
<thead>
<tr>
<th>Reference</th>
<th>Year</th>
<th>Classification</th>
<th>Hemiplegia</th>
<th>Tetraplegia</th>
<th>Diplegia</th>
<th>Ataxic</th>
<th>Congenital</th>
<th>Diplegia</th>
<th>Dystonic—mainly</th>
<th>Choreo-athetotic</th>
</tr>
</thead>
<tbody>
<tr>
<td>SCPE(^1)</td>
<td>2000</td>
<td>Spastic</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td>Ataxic</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td>Dysskinetic</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

\(^2\) Reference Study Duration

<table>
<thead>
<tr>
<th>Reference</th>
<th>Study</th>
<th>Duration</th>
<th>Number of cases of CP</th>
<th>Number of spastic cases</th>
<th>Spastic cases as % of all cases</th>
<th>Unilateral spastic as % of spastic cases</th>
<th>Diplegia as % of spastic cases</th>
<th>Bilateral spastic as % of spastic cases</th>
<th>Diplegia as % of bilateral spastic cases</th>
</tr>
</thead>
<tbody>
<tr>
<td>26</td>
<td>North Italy(^*)</td>
<td>1980-89</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>27, 28</td>
<td>Denmark</td>
<td>1979-90</td>
<td>908</td>
<td>734</td>
<td>81</td>
<td>35</td>
<td>45</td>
<td>65</td>
<td>69</td>
</tr>
<tr>
<td>29</td>
<td>North England</td>
<td>1991-96</td>
<td>537</td>
<td>499</td>
<td>93</td>
<td>37</td>
<td>23</td>
<td>63</td>
<td>36</td>
</tr>
<tr>
<td>23</td>
<td>Northeast England</td>
<td>1980-96</td>
<td>380</td>
<td>355</td>
<td>93</td>
<td>42</td>
<td>23</td>
<td>58</td>
<td>40</td>
</tr>
<tr>
<td>31</td>
<td>Mersey, England</td>
<td>1984-89</td>
<td>497</td>
<td>460</td>
<td>93</td>
<td>39</td>
<td>23</td>
<td>61</td>
<td>38</td>
</tr>
<tr>
<td>32</td>
<td>Atlanta, USA</td>
<td>1985-87</td>
<td>204</td>
<td>166</td>
<td>81</td>
<td>34</td>
<td>19</td>
<td>66</td>
<td>28</td>
</tr>
<tr>
<td>33, 34, 35</td>
<td>Sweden(^†)</td>
<td>1979-90</td>
<td>545</td>
<td>470</td>
<td>86</td>
<td>40</td>
<td>51</td>
<td>60</td>
<td>84</td>
</tr>
<tr>
<td>36</td>
<td>England and Scotland</td>
<td>1984-89</td>
<td>1649</td>
<td>1334</td>
<td>81</td>
<td>35</td>
<td>22</td>
<td>65</td>
<td>33</td>
</tr>
<tr>
<td>37</td>
<td>Slovenia</td>
<td>1981-90</td>
<td>768</td>
<td>651</td>
<td>85</td>
<td>33</td>
<td>43</td>
<td>67</td>
<td>64</td>
</tr>
<tr>
<td>38</td>
<td>Western Australia</td>
<td>1980-94</td>
<td>819</td>
<td>639</td>
<td>78</td>
<td>45</td>
<td>37</td>
<td>55</td>
<td>68</td>
</tr>
<tr>
<td>39</td>
<td>Rome, Italy</td>
<td>1977-96</td>
<td>282</td>
<td>213</td>
<td>76</td>
<td>33</td>
<td>27</td>
<td>67</td>
<td>40</td>
</tr>
<tr>
<td>40</td>
<td>Northern Ireland(^‡)</td>
<td>1977-92</td>
<td>960</td>
<td>572</td>
<td>87</td>
<td>43</td>
<td>21</td>
<td>57</td>
<td>36</td>
</tr>
<tr>
<td>41</td>
<td>Norway</td>
<td>1980-89</td>
<td>46</td>
<td>39</td>
<td>85</td>
<td>49</td>
<td>33</td>
<td>51</td>
<td>65</td>
</tr>
<tr>
<td>42</td>
<td>Oxford, England</td>
<td>1984-95</td>
<td>967</td>
<td>806</td>
<td>83</td>
<td>40</td>
<td>60</td>
<td>60</td>
<td>53</td>
</tr>
</tbody>
</table>

\(^*\) Derived from rates in fig 4 of reference 26.

\(^†\) Includes 11 acquired cases 1976-82.

\(^‡\) CP type only available in two thirds of cases.

\(§\) Includes 68 acquired cases.
a proportion of all cases average 85% (range 76–93%).

Diplegic cases as a proportion of spastic cases average 33% (range 19–62%); whereas diplegic plus quadriplegic cases as a proportion of spastic cases average 62%, with a much smaller range of 51–77%. If one looks just at bilateral spastic cases, the proportion of diplegic cases varies from 28% to 84%—a more than triplefold difference. Such a large difference is very unlikely to be a real one and is almost certainly a result of the way in which different centres interpret diplegia.

Table 3 shows data from the studies which report by birth weight. In those less than 2.5 kg, diplegic cases as a proportion of spastic cases average 37% (range 30–55%), only very slightly higher than the average of 33% in all birth weights; and for those less than 1.5 kg, the average is 35%. These findings argue against diplegia being especially associated with either low birth weight or very low birth weight.

**DISCUSSION**

Semantically, the word “diplegia” must mean two limbed or two sided weakness. If two limbed, it is bizarre that often three or four limbs are involved. If two sided, it is preferable to use a term such as bilateral with no implication about sided, it is preferable to use a term such as bilateral with no implication about sidedness, it is preferable to use a term such as bilateral with no implication about sided, it is preferable to use a term such as bilateral with no implication about sidedness.

There is no justification for separating diplegia and quadriplegia. While it is possible that the term diplegia as used by Ingram® may have represented a distinct clinical syndrome before the introduction of neonatal intensive care, we are confident that it does not now. Our analysis of published epidemiological studies has shown that confusion about and inconsistent application of the term diplegia is embedded unknowingly in the studies. When diplegia and quadriplegia are treated separately, there are very unlikely differences between countries which largely disappear when treated together. Further, the supposed association of diplegia with low birth weight is not seen.

Recent advances in imaging support this position. In her magnetic resonance imaging (MRI) studies of children with spastic cerebral palsy,™ Kragehol-Mann found that periventricular leucomalacia and parasagittal lesions were the commonest lesion in preterm infants and term infants. Very similar MRI appearances were responsible for quadriplegic, diplegic, and spastic syndromes. Severity of motor involvement was related to severity of white matter reduction but the authors could find no justification for separating these syndromes and therefore used the phrase “bilateral spastic cerebral palsy”.

In epidemiological studies, more will be gained from an agreed simple logical classification which is uniformly applied than from a complex one which may have internal inconsistencies, be inconsistently applied and influenced by clinical judgement. Our own preference is for that developed by the European Collaboration.™

In clinical studies, there may be a need to define more refined groups of children, but these should be described by their neurological and functional deficits, not by shorthand terms such as “more” or “less affected”, when like is not being compared with like. An Oxford group addressed this issue in the 1980s.™ Their systematic description would allow the detail necessary for evaluating the effects of different drugs, operations, or physical therapies. An alternative would be to build on the European classification.™ For example, the description:

“spastic, bilateral, neurological signs in four limbs, minimal functional involvement of upper limbs, all cognitive abilities”

would describe reliably a group of children if the term “minimal” was defined for the age group(s) of children being studied.

**ACKNOWLEDGEMENTS**

We are grateful to all members of Surveillance of Cerebral Palsy in Europe (SCPE) for discussions on this subject over four years.

Arch Dis Child 2003;88:286–290

**REFERENCES**

13 Ingram TS. The neurology of cerebral palsy. Arch Dis Child 1966;41:337.