Letters to the editor

‘Visual impairment in children with brain damage: towards a diagnostic procedure?’

SIR—The discussion in this journal (Vol. 44: 356–357, 2002) between the Vancouver group (Carey Matsuba, James Jan, Christopher Lyons, and Roberta Heaven) and the Lausanne group (Eliane Roulet-Perez and Thierry Deonna), concerning the terms cortical versus cerebral visual impairment in children, and whether eye motility problems may or may not be covered by the term, has highlighted an important issue. Children with visual impairment due to damage to the retrogenticulate visual pathways constitute an increasing group among visually impaired children in the Western world. Clinicians in the paediatric field (neurologists, psychiatrists, psychologists, and ophthalmologists) need to define criteria which would include a child within this functional diagnosis, and consent on grading and description of the pattern of the visual disability.

Since the 1980s, groups from USA, Canada, UK, the Netherlands, Italy, and Scandinavia, have presented studies of children with cerebral visual pathway lesions and visual function problems. In 1985, Whiting and colleagues demonstrated that children with permanent cortical visual impairment rarely had complete lack of vision, did not have manifest nystagmus, and that it often coexisted with abnormal optic disc appearance, presumably the result of a retrograde transsynaptic degeneration. In 1987, Jan et al. described cortical visual impairment as severe visual loss, normal or minimal ocular finding, and clinical, electrodiagnostic, and computed tomographic (CT) evidence of post-geniculate lesions involving the visual cortex. In 1994, the same group defined cortical visual impairment as ‘bilateral loss of vision, with normal pupillary responses and an eye examination that shows no abnormalities’.

Research groups in Europe have chosen terms such as cerebral visual disturbance and cerebral visual impairment for children with retrogenticulate causes of visual dysfunction. The manifestations of this kind of visual impairment include subnormal acuity and crowding, affected visual fields, a characteristic behavioural profile, and associated disorders of higher visual processing. It also includes ocular motor problems, such as strabismus, neurological nystagmus, deficient accommodation, fixation, saccades, and smooth pursuit movements. Aetiological and morphological causes of cerebral visual impairment in young children have been described and may be malformation, hypoxia-ischemia, infection, or trauma. The pattern of pathology found depends on the stage at which the immature cerebral visual system was injured.

The lesions are seldom restricted to the visual pathways but often also engage other parts of the CNS, and result in more pervasive developmental disorders. A wide range of disorders have been described in children who are visually impaired due to brain lesions, these include: uneven cognitive performances, visual-spatial and visual-motor difficulties, attention problems and autistic-like behaviours, and cerebral palsy. Varying degrees of learning problems are reported within a spectrum from severe learning disability, to more limited areas of dysfunction in children with an intellectual ability within the normal range. Therefore, a child with brain damage may present many functional deficits, one of which is visual impairment. The label of the functional deficit will then depend on which dysfunction is the most conspicuous, and in whose office the child was seen. In a pre-verbal child it may not be possible to know whether observed behaviour and practical problems are the consequence of visual impairment or learning disability, or both.

Although the terminology differs, there is a fair amount of agreement about the functional core deficits in children with visual impairment due to brain damage. There is a need to reach consensus on the diagnostic terminology, as well as on inclusion and exclusion criteria, and grading of the severity of the functional disability.

We suggest that the term cerebral visual impairment, in keeping with the term cerebral palsy, is chosen to describe the clinical condition and functional deficit due to damage to the posterior visual pathways and/or visual cortex. The term cerebral visual impairment has been preferred to cortical visual impairment because only rarely is the cortex of the brain damaged in isolation. Furthermore, we put forward a proposal that the functional diagnosis of cerebral visual impairment should be suspected in a child who in the absence of ocular pathology, other than mildly abnormal appearance of the optic disc, demonstrates subnormal visual acuity for age (as measured with gratings or with linear optotypes with the best correction), and/or constricted visual fields found with confrontation, Goldmann or computerized perimetry in combination with a disabling visual perceptual-cognitive dysfunction, as demonstrated with neuropsychological assessment and/or a structured history taking. This condition may be aggravated by eye motility problems, such as poor fixation and/or eye movement disability. Cerebral visual impairment may be graded as mild, moderate, or severe, based on assessment of these functions.

Visual perceptual-cognitive dysfunction may be suspected early but cannot be accurately assessed until the age of 4–5 years. Cerebral visual impairment may therefore be a tentative clinical diagnosis at early ages. Later in life the visual acuity may improve but the visual cognitive problems persist or even increase.

The diagnostic procedure demands a clinical examination of the eyes and ocular motility, and an assessment of visual perception-cognition by psychological testing and/or by structured history taking. The diagnosis may be strengthened by neuroimaging of the brain in order to demonstrate lesions to the visual pathways/visual cortex. Other clinical signs of brain damage, such as cerebral palsy or epilepsy, may also indicate a cerebral cause of visual dysfunction.

The four children described below, illustrate how function can be evaluated with a battery of different standard techniques as a base for diagnoses.

Child 1 was 9 years old, with grating acuity 12 c/d, binocular linear optotype acuity 0.15. Fields: low neural capacity (15%) measured with computer-assisted high-pass resolution perimeter, but near normal outer limits measured with Goldmann perimeter. WISC: Verbal IQ 120, Performance IQ 60, Full Scale IQ 100. Eye motility: esotropia, manifest nystagmus, problems with holding fixation, occurrence of paroxysmal deviations, inability to perform adequate saccades, and

*US usage: mental retardation.

poor smooth pursuit. However, these problems with eye motility can be fairly well compensated for with head movements. Eyes: normal fundi. Neuroimaging: moderate periventricular leukomalacia documented with MRI. Functional diagnosis: severe cerebral visual impairment.

Child 2 was 7 years old, with grating acuity 38 c/d, linear optotype acuity RE (right eye) = LE (left eye) 1.0. Fields: normal Goldmann fields. WISC: Verbal IQ 140, Performance IQ 52, Full Scale IQ 98. Eye motility: normal. Eyes: normal fundi. Neuroimaging: CT performed in the neonatal period verified the right hemifield measured with Goldmann perimetry. WISC: Verbal IQ 70, Performance IQ 52, Full Scale IQ 57. Eye motility: esotropia and dysmetric saccades. Eyes: normal fundi. Neuroimaging: CT performed in the neonatal period verified a unilateral brain haemorrhage, resulting in a later documented atrophy of the right occipital lobe. Functional diagnosis: moderate cerebral visual impairment in combination with mild cerebral palsy (hemiplegia) and mild learning disability.

Child 3 was 9 years old, with grating acuity 38 c/d, linear optotype acuity RE 0.63, LE 0.2. Fields: left homonymous hemianopia, increased threshold, and mild restriction of the right hemifield measured with Goldmann21 perimetry. WISC: Verbal IQ 70, Performance IQ 52, Full Scale IQ 57. Eye motility: esotropia and asymmetric saccades. Eyes: normal fundi. Neuroimaging: CT performed in the neonatal period verified a unilateral brain haemorrhage, resulting in a later documented atrophy of the right occipital lobe. Functional diagnosis: moderate cerebral visual impairment in combination with mild cerebral palsy (hemiplegia) and mild learning disability. These children present with a variety of disabilities due to cerebral dysfunction, and their functional diagnoses may be debated. Although they all have visual problems, only two children (1 and 3) fulfil our proposed criteria for the functional diagnosis of cerebral visual impairment. The practical visual problems described in child 2 correspond to the subnormal Performance IQ, but cannot be evaluated or graded by the ophthalmologist, because all tests of vision and the eye examination were normal. In this case, the dysfunction fulfils the criteria for a neuropsychiatric diagnosis. Child 4 represents a group of visually impaired children with lesions of both the eyes and the posterior visual pathways. Here, the severe retinal disease is obviously the main cause of low visual input (i.e. ocular visual impairment), but in daily life, visual problems are aggravated by the inability to interpret visual information. All four children have problems with reading. Child 1 prefers braille to print.

The basis for the functional diagnosis of cerebral visual impairment calls for a multi-disciplinary approach: a task for paediatric ophthalmologists in collaboration with psychologists, paediatric neurologists, paediatric psychiatrists, and neuroradiologists. In the future we need a model to quantify the degree of this functional disorder. A standard assessment procedure that includes visual, ocular motor, cognitive, and behavioural functions is also required. The aim of such a diagnostic procedure is to give support to the child, parents and teachers, and to work out strategies for optimal habilitation and for adapted education.
(sometimes with an epileptic focus in the visual cortex and additional ‘epileptic visual dysfunction’), learning disability, or more complex disorders of communication. This makes the evaluation of the visual problems very difficult. In addition, children with cerebral visual impairment most often have combined peripheral ocular pathology as well as supranuclear eye motility disorders. These latter problems may be either an associated neurological impairment or the consequences of the visual disorder, or both. Each of these dimensions need to be studied separately, require special neurological, ophthalmological and neuropsychological skills, which often need to be repeated over time, and are hard to carry out and interpret in young children often with multiple disabilities. Discrete components within the visual system can be spared (colour, movement) or affected (contour discrimination, and line orientation) in different children, creating puzzling and fluctuating competencies and deficiencies depending on context. For these reasons, it is not surprising that cerebral visual impairment is so difficult to describe in all its components, and to recognize its importance among other visual and non-visual disabilities.

However, one should not feel defeated. There are children with severe brain pathology and associated learning disability and/or cerebral palsy, who have no problem whatsoever with visual perception, indicating that these latter problems are quite specific.

Children with ‘pure’, isolated, cerebral visual impairment are rare. However, there is a newly emerging category of children in whom cerebral visual impairment might be the main or only disability. These are prematurely born children who suffered mild degrees of leukomalacia in the visual pathways, and whose deficit becomes recognized and clinically significant only after the first years of life, when higher levels of visual integration are necessary in different aspects of life, and for certain types of academic skill. These children enlarge the spectrum of cerebral visual impairment, which has indeed been a neglected field. As always, in the domain of developmental disabilities, progress is dependant on the ability of different professionals to work together.

We wonder whether a new diagnostic category, as proposed by Dr Jacobson and colleagues, should be created. This would have specific inclusion and exclusion criteria for children with ‘cerebral visual impairment’, by analogy with cerebral palsy, autism, and attention-deficit disorder. With the present level of available information, it remains difficult to analyze, and particularly to quantify, all aspects of the visual dysfunction in a meaningful way and, therefore, to decide which children to include or exclude. The illustrative cases that are briefly reported by Dr Jacobson show the difficulty in evaluating the clinical importance of the cerebral visual impairment among all other disabilities.

‘Developing Mac Keith “College”’

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SIR—Rarely has the editorial pen slipped so dramatically in my usually nerveless fingers than it did in my December editorial. I and all my colleagues failed to note the misprint of erotic for erratic, so I have ascribed to Hippocrates the view that it was the erotic nature of the gods that led them to the belief that epilepsy was a sacred disease. My colleagues on the journal missed this ‘typo’ too, possibly thinking that I meant erotic. The Greek gods were indeed happily erotic, but Hippocrates’ notion that epilepsy was not divinely inspired was based on his observation of people having epilepsy. It is interesting that he noted the occurrence of temporary paralysis (Todd’s paralysis) after seizures. Perhaps this confirmed him in his view that the biological was occurring and not something divine.

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