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The Spectrum of Vision Impairment Caused by Pediatric Neurological Injury

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Injury to the central nervous system is the leading cause of vision impairment in children in Western countries. Known as cortical, or cerebral, visual impairment (CVI), the usual cause is perinatal hypoxia/ischemia, although developmental brain disorders such as lissencephaly, polymicrogyria, and porencephaly can be etiological. Since injury rarely affects the visual cortex or optic radiations alone, afflicted children often experience disruption of other aspects of neurological functioning, with seizures, mental retardation, disturbed attention, and cerebral palsy commonly occurring in tandem with disturbances of vision.

Considerable debate has emerged over the past several years regarding the terminology that should be applied to children with vision impairment caused by neurological injury. Cortical visual impairment, cerebral visual impairment, neurological visual impairment, and retrogeniculate visual impairment—all are terms with enthusiastic supporters. Common to all these terms is the presence of impaired visual acuity. None of these terms addresses other problems that could cause higher disorders of vision processing.

A much healthier debate could center on the term *impairment*. Jim Jan, who did pioneering research on children with CVI, deserves credit for using this word to describe children with vision loss caused by neurological injury. Virtually no child with CVI is completely blind. All experience some degree of preservation of vision, with the potential for at least some recovery. So what constitutes impairment? It certainly indicates impaired spatial resolution, but reduced visual acuity also can be accompanied by other abnormalities of higher vision integration. This edition of the Journal offers two papers that probe the concept of *impairment*.[1,2]

Brain injury in children occurs for the most part by different mechanisms than in adults. Adults can experience global ischemia in rare near-drowning events and with general anesthesia complications. However, the usual insult to the adult brain comes in the form of an infarct to a fully-developed brain. Prosopagnosia in adults (inability to recognize faces) and other disorders that affect higher aspects of visual functioning are the result of an infarct that spares much of the brain. In contrast, children who suffer brain damage do so as the result of a global neurological insult. Perinatal hypoxia affects most of the brain and does not selectively cause isolated disturbances of higher vision processing. Exceptions occur in perinatal stroke, wherein a major cerebral vessel is occluded in the setting of a hypercoagulable disorder.

Children with vision loss caused by neurological injury who are referred to pediatric ophthalmologists therefore are often profoundly neurologically impaired. The examination can be very difficult, since so many of these children have impaired motor control (including eye

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movement control), seizures, and poor attention. Quantitative analysis of visual function is often limited to behavioral assessment of grating acuity or confrontational visual field analysis. As a result, the assessment of these children in many offices involves an almost binary algorithm: does the child have any vision or not? The pediatric ophthalmologist may also be asked to assist with a diagnosis and lesion or disease localization. Seldom is the ophthalmologist asked to evaluate so-called higher aspects of visual functioning, such as face recognition, or aspects of visual functioning described in two excellent papers in this issue of the Journal.

Saidkasimova and colleagues[1] describes cognitive visual impairment in 7 children with posterior periventricular white matter injury. These children, to one degree or another, experienced impaired orientation, impaired perception of movement, and varying degrees of simultanagnosia. Drummond[2] describes possible Balint syndrome in a child with posterior parietal and occipital lobe damage. In this condition, affected individuals have simultanagnosia, impaired hand and feet movements under visual control, and optic apraxia (trouble moving eyes to a desired position or target). Conditions described in these two papers are more characteristic of strokelike syndromes seen in adults.

Or are they? This is the important question. While diagnostic criteria for these higher cognitive and attentional disorders are difficult to satisfy in pediatric cases, there can be little question that CVI is accompanied by various other vision problems and that these problems can occur in the presence of normal visual acuity (therefore not CVI per se). Each child has his or her own constellation of additional visual problems.

It is possible to quibble about certain aspects of these case reports. Most children with nystagmus, no matter the cause, experience some degree of ocular motor apraxia. How does one differentiate a simple attention problem from true simultanagnosia? By what measure are inferior visual fields really reduced or constricted in CVI, since it is so difficult to measure fields even in healthy 5-year-olds? Nevertheless, observations on cognitive visual impairment and Balint syndrome are so detailed and so congruous with observations by families that there can be little doubt about the validity of these observations. Furthermore, the diagnostic problems noted should not dissuade us from attempting to help our patients when they are confronted with these obstacles.

The authors of these publications are to be congratulated for bringing to our attention these important and troubling conditions that accompany CVI. Pediatric ophthalmologists should pay closer attention to problems other than visual acuity deficits in their patients. What remains for the future are better diagnostic tests and treatments for these disorders of vision. It is likely that cognitive visual impairment and conditions such as Balint syndrome are far more common than we have appreciated.

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