EDITORIAL

CORTICAL VISUAL DYSFUNCTION IN CHILDREN

Cortical visual dysfunction is a relatively uncommon diagnosis in most adult ophthalmology clinics and this diagnosis is often a prelude to no further diagnostic or therapeutic action by the ophthalmologist. However, the paper by Dutton *et al.* in this issue of *Eye* reminds us that patients with cortical dysfunction deserve our clinical attention and have indeed profound lessons to teach us about the organisation of the visual system.

The striate visual cortex area V1 is the key cortical area responsible for conscious vision, and bilateral destructive lesions in this region cause cortical blindness. However, it is clear that this is not the only part of the brain responsible for visual function. For example, the retention of reflex and navigational vision by patients with occipital blindness (often associated with denial of blindness) suggests a pathway for reflex activities outside the geniculostriate pathway.[1] Moreover, a host of other curious visual dysfunctions have been described in association with cortical lesions outside area V1.[2] Two of the most fascinating of these are visual agnosia, the inability to understand or recognise objects by visual clues alone, and prosopagnosia, a specific form of visual agnosia in which the patient is unable to recognise faces but can identify people by voice, bodily movements, prominent items of dress, etc.

Although the functional neuroanatomy of these two disorders is still controversial a combination of clinical observation and laboratory work in animals has elucidated the pathophysiology of some other forms of cortical visual dysfunction. Riddoch[3] first observed residual movement perception in the blind visual field of patients with hemianopia in 1917. Subsequent to this, patients were reported with the converse disability, cerebral motion akinetopsia, in which there is an inability to detect movement. Clinical and neuroimaging description of these patients and the demonstration in primates of areas of cortex containing cells responsive only to moving visual stimuli have established that area V5, lying in the region of the occipitoparietal temporal junction and receiving an input from the striate cortex, is responsible for processing movement perception.[4]

Homonymous hemianopia following stroke in elderly patients is commonly caused by involvement of the optic radiation close to the internal capsule. However, where there is cortical damage, more bizarre visual dysfunctions may not be mentioned by the patient because they are difficult to describe, and may not be sought by the ophthalmologist because the therapeutic value of recognising these symptoms is not fully appreciated.

Cortical visual dysfunction in children has been much less studied than in adults. It is more difficult to investigate because of the plasticity of the child's evolving visual system and because of greater difficulties in communication. However, specific cortical dysfunctions in children are particularly interesting because of the developing arrangement of the vascular supply of the brain, with the watershed areas most susceptible to ischaemia moving from the periventricular area in preterm infants to the posterior parasagittal region in full-term infants. In addition, these disorders are even more important in children because of the very common presentation of children to paediatric ophthalmology clinics with cerebral palsy, microcephaly and other causes of developmental delay that may be associated with cortical blindness and because children are even more dependent on their carers' understanding of their disabilities in creating an environment in which they can flourish.

In this issue of *Eye*, Dutton *et al.* describe their observations of 90 children referred to them with cortical visual dysfunction. Just over one quarter of their patients had either complete blindness or severe visual impairment and 30% had what appeared to be straightforward visual field defects. However, the most interesting patients they describe are the 20 whose visual dysfunction was more than would be expected from the assessments of visual acuity and visual fields.

This paper shows that cortical visual dysfunction

affecting recognition, orientation, depth perception, motion perception and simultaneous perception are by no means uncommon in children, and it suggests how understanding these problems can assist in the everyday life of affected children. Every paediatric ophthalmologist should strive to recognise and understand these dysfunctions. Dutton *et al.* devoted a very large amount of time to each patient (an average of just more than two children were seen in each multi-disciplinary clinic) and others may question whether this is feasible in their own practice. However, much of the information was readily volunteered by parents and a better understanding of these disabilities will allow them to be much more rapidly recognised. This work in children confirms previous findings in adults that pure isolated specific cortical dysfunctions are rare, but dramatically demonstrates that certain dysfunctions commonly occur together in children while others do not. Further study of patients with these disorders should not only help us understand our patients' needs better, but should also provide clinical insights into laboratory work on the functional organisation of the visual system.

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References

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