Australian Orthoptic Journal 1990; 26: 42-44

CORTICAL BLINDNESS IN MULTIHANDICAPPED CHILDREN

HEATHER CROSSMAN, DipAppSc(Cumb), DOBA
Royal NSW Institute for Deaf and Blind Children, North Rocks

Abstract

Three case studies of diagnosed cortical blindness are presented, and their patterns of recovery are compared, with particular emphasis on length of recovery time and current level of vision. All three were found to progress to a level of "panoramic vision", with any further improvement being limited to within these boundaries, that is, vision primarily concerned with spatial relationships and movement, by orienting the eyes and limbs to sudden peripheral movements.

Key Words: *Permanent cortical visual impairment, panoramic vision.*

Cortical blindness is a phenomenon commonly encountered when working in the field of developmental disabilities. Hoyt[1] found that on examination, the cortically blind child failed to show any visual fixation or following movements retained pupillary responses to light, had no nystagmus and no abnormalities of the ocular structures. These findings were generally associated with widespread neurologic disease.

Denny Brown and Chambers[2] concluded that two distinct visual systems may exist in humans. This conclusion was reached through experiments on Macaque monkeys and through the review of literature on cortical blindness in adult patients. The first system which they labelled "object vision" involves the macula, the lateral geniculate body and the striate cortex, it also requires interaction with the superior colliculus. The second system which they labelled "panoramic vision" is primarily concerned with spatial relationships and movement, orienting the eyes and limbs to sudden movements registered by the peripheral retina. This second visual pathway includes the peripheral retina, the inferior pulvinar nucleous and areas 18 and 19. They found that each system can function without the other, though normally they must be integrated at all levels. This research would indicate therefore that destruction of the striate cortex (area 17) alone should not cause complete loss of vision.

Whiting, Jan, Wong, Flodmark, Farrell and McCormick[3] have identified two forms of cortical blindness which they prefer to call cortical visual impairment (CVI). The first they call transient CVI and the second permanent CVI where little or incomplete recovery occurs. They have also reported that the general pattern of recovery from cortical blindness is that the patient perceives and follows light, then moving objects. Later, visual acuity improves but poor vision and visual-perceptual difficulties may remain permanently.

It is this pattern of recovery that has been observed in a series of multihandicapped children. The conclusions reached by Whiting et al[3] would therefore have implications regarding the extent of visual recovery that can be expected in these children.

Address for correspondence: H. Crossman, Royal NSW Institute for Deaf and Blind Children, 361-365 North Rocks Road, North Rocks, NSW 2151.

They also found that transient CVI most often is reported following meningitis and minor head injury, with the major causes of permanent CVI being perinatal asphyxia and shunt malfunction.

Hodgson[4] in her study of 22 children with "pure" cortical blindness (ie. with no optic nerve or retinal pathology) found that seventeen showed apparent improvement in vision before the age of two years with no improvement occurring after that. Hoyt[1] in his study of 43 children found that improvement was variable but generally quite slow with visual recovery continuing to occur in some patients more than two years after the original insult.

This study shows how "permanent CVI" has presented in three multihandicapped children currently being seen at the Royal New South Wales Institute for Deaf and Blind Children. All three children have shown improvement in their functional vision with improvement occurring up to six years after the initial insult.

CASE 1

The first child contracted encephalitis at four months of age resulting in a diagnosis of cortical blindness, abnormal muscle tone and developmental delay. He was first seen at 15 months where he was noted to be aware of light by turning towards a diffuse light source such as a window, he did not however, respond to a torchlight. A small to moderate right exotropia was present, optokinetic nystagmus (OKN) was elicited when the drum was rotated with the stripes moving from right to left and when it was rotated in a downwards direction. He was then seen at six monthly intervals, during which a steady improvement was evidenced with the child being able to locate brightly coloured objects that were held at one third of a metre (1/3 m). This progressed to his being able to follow a slowly moving object when he was two and a half years old. At this stage no central fixation was evident, optokinetic nystagmus was now present in all directions. When last assessed at three and a half years he saw and reached for the stycar toys when they were held fifteen to twenty (15-20) centimetres from his eyes. Responses were more consistent if the object was moved slightly. When tested with the stycar balls he was again noted to be more reliable when the balls were moving rather than stationary. Cover testing showed that both eyes were in a divergent position with a right hypertropia. All smooth pursuit movements were irregular with the object of regard having to be moved slowly for him to be able to follow it.

CASE 2

The second child has diagnosed cortical blindness, bilateral optic atrophy, spastic quadriplegia and epilepsy as a result of placental insufficiency and premature birth (34 weeks gestation). He was first seen at 11 months of age at which stage he was noted to be attracted to diffuse sources of light. Occasional horizontal jerk nystagmus was present.

He was seen again at 19 months of age, at this assessment, he displayed searching eye movements in the general direction of a bright torchlight. He also saw and followed a thin white wooden rod as it was moved across his line of vision. Intermittent horizontal jerk nystagmus was present.

He was not seen again until he was four years of age. At this assessment he followed a penlight torch in all directions. If a brightly coloured object was held approximately 30 cm from his eyes he would reach for it only after it had been moved slightly. He followed a slowly moving object in all directions. On cover testing he was found to have a large left hypertropia, the left eye was also slightly exotropic. Fine horizontal nystagmus and some wandering eye movements were present.

CASE 3

The third child suffered severe anoxic brain damage at birth resulting in cortical blindness, microcephaly, global developmental delay, spasticity and epilepsy. He was first seen when he was four years old, on this assessment he was noted to look in the general direction of a light, both eyes were in a divergent position and horizontal OKN was present. He was reassessed at five years, four months of age, where he was found to be reaching on sight for large bright toys at 33 cm, and showing some irregular

following movements. At the age of seven years four months, he would crawl towards patterned objects (such as the OKN Drum) at approximately 1½-2 m where he would inspect them visually. There was no evidence of central fixation.

The most recent assessment was at seven years and seven months. Any signs of improvement at this stage were minimal. It was noted that he responded to a moving object immediately either by reaching for one that was being moved slightly, or by visually following an object. Central fixation was not present.

DISCUSSION

Of the three cases presented, two were the result of birth difficulties and problems during pregnancy. Both of these children have severe developmental delay and visual improvement of any significance was not apparent until they were over three years of age. In the first case the problems did not commence until four months of age. This child is improving in all areas both cognitively and physically and visual improvement was first noted at the age of two years.

None of the children have firmly established central visual fixation and they have all been noted to respond more readily to moving rather than stationary objects. It would appear that they all have "permanent cortical visual impairment".[3] All three present with a similar pattern of recovery and all have progressed to the stage of "panoramic vision",[2] ie. vision primarily concerned with spatial relationships and movement, where they eyes and limbs are oriented to sudden movements registered by the peripheral retina.

Whilst all three children have an intellectual impairment it is felt that this would not account for the type of recovery that has been documented, ie. the apparent visual improvement was not due to the delayed improvement in other functions. The causes in all three are quite diverse as is the recovery time which ranges from three to seven years after the initial insult. The three subjects, whilst having shown a continual steady improvement, have all reached the stage of detecting movement before stationary objects and have limited, if any, central fixation. Once having reached this stage, improvement has been within these limitations suggesting that improvement in visual acuity may not be a realistic expectation.

CONCLUSION

Cortical blindness is commonly encountered when working with multihandicapped children. This paper has presented three subjects who, whilst making a steady improvement in the use of their vision, have all seemed to plateau at the level of "panoramic vision". The implications of this study are therefore, that in cases of cortical blindness visual improvement can occur up to seven years after the initial insult and that the level of improvement may be limited to "panoramic vision".

References

1. Hoyt CS. Cortical Blindness in Infancy. Paediatri Ophthalmology and Strabismus. Trans of the Nev Orleans Academy of Ophthalmology, Raven Press, Nev York, 1986.
2. Denny Brown D, Chambers RA. Physiological Aspect of Visual Perception. 1. Functional Aspects of Visua Cortex. Archives of Neurology 1976; 33: 219-227.
3. Whiting S, Jan JE, Wong P, Flodmark O, Farrell K McCormick A. Permanent Cortical Visual Impairmer in Children. Developmental Medicine and Chil Neurology 1985; 27: 730-739.
4. Hodgson L. Visual Improvement in the Cortically Blin Child. Student Research Papers, Cumberland College c Health Sciences, Sydney, 1986.