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A CASE STUDY IN CORTICAL PLASTICITY

JOANNE MITCHELL, BAppSci(Cumb)DOBA
The Royal New South Wales Institute for Deaf and Blind Children. The Alice Betteridge School, Sydney.

HEATHER CROSSMAN, BAppSci(La Trobe)DOBA
The Royal New South Wales Institute for Deaf and Blind Children. The Alice Betteridge School, Sydney.

Abstract

Much research has gone into establishing the critical or sensitive period for the development of binocular vision in humans. An understanding of this and the plasticity of the visual system has had implications for the treatment of amblyopia in children. Occlusion therapy is thought to be effective until approximately eight years of age and most effective below the age of four years.

This paper presents a case study of an eleven year old boy who is diagnosed with cortical visual impairment, cerebral palsy and epilepsy. At six months of age no visual responses were evident on both observation and VER testing. At eleven months, responses to light were noted and vision continued to slowly develop. Part-time occlusion to decrease right amblyopia began at the age of four years and eight months. Greatest improvement was noted between the ages of nine and ten years when vision improved to approximately 6/45, equalling the vision in the left eye.

Literature in the 80's, describing experiments on cats, found a lengthening of the "critical period" following an early period of no visual attention. The hypothesis is made that in cases of delayed visual development in children there is the possibility of a lengthening of the sensitive period and thus response time to occlusion therapy.

Key Words: *critical/sensitive period, vision, amblyopia, occlusion, development, cerebral palsy.*

INTRODUCTION

The critical or sensitive period, as described by Von Noorden[1] is a time span known to exist in humans and experimental animals during which certain visual functions are modifiable by decreased or abnormal stimulation.

Foundational research into the critical period was carried out by Hubel and Weisel in the 1960's. By studying the receptive fields in the visual cortex of the cat they discovered that by altering the binocular visual experience of kittens through monocular deprivation and artificially induced strabismus, that marked changes in the way in which the visual cortical cells processed information occurred[2,3]. In studies comparing monocular lid suturing at different ages, in the newborn and older cat, they established the critical period which extends from 3 weeks to approximately 3 months of age[4] (see Figure 1).

In similar experiments, a critical period for Macaque monkeys was also discovered. The Macaque monkey is most sensitive to monocular deprivation during the first 6 weeks of life,

Address for Correspondence: Miss J Mitchell, The Alice Betteridge School, The Royal New South Wales Institute for Deaf and Blind Children 361-365 North Rocks Road, North Rocks 2151.

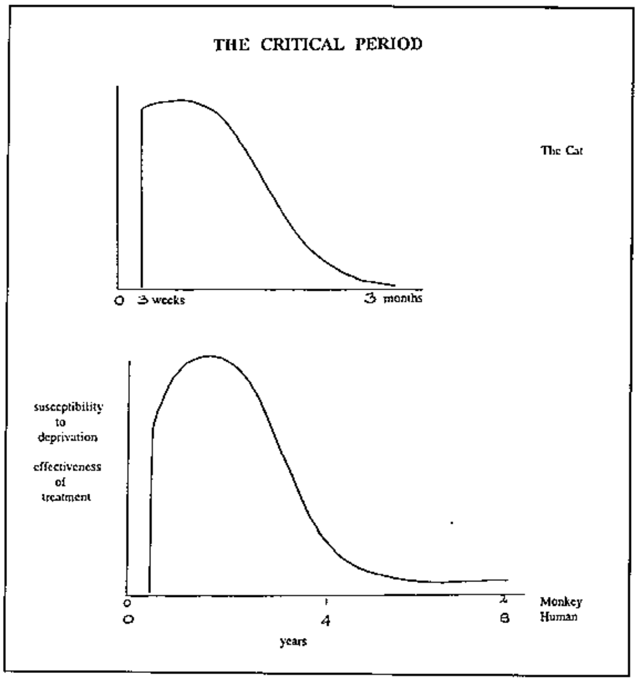

Figure 1.

sensitivity then declines progressively to 1½ to 2 years when the monkey loses this type of cortical plasticity[5].

The development of the visual system in humans is thought to be approximately four times longer than in monkeys, therefore in humans the critical period is thought to extend from birth until approximately 6-8 years, however a precise age has not been demonstrated[1].

An understanding of the 'critical or sensitive' period and the plasticity of the visual system has had marked implications for the treatment of amblyopia in children. It has long been recognised, in fact since 1742, that by patching the 'good eye' one can improve the vision in an amblyopic eye[6]. Amblyopia therapy today takes on many forms, from occlusion of the better eye, to penalisation using lenses and the use of cycloplegic drugs, however the general principle remains the same. It is experimental research that has given us an understanding of the mechanisms behind amblyopia and the cortical changes that take place, but more than that, research into the critical period has also given us a time frame within which to work.

Recent clinical studies based on the age of onset of the disruption to binocular vision have estimated the sensitive period for different

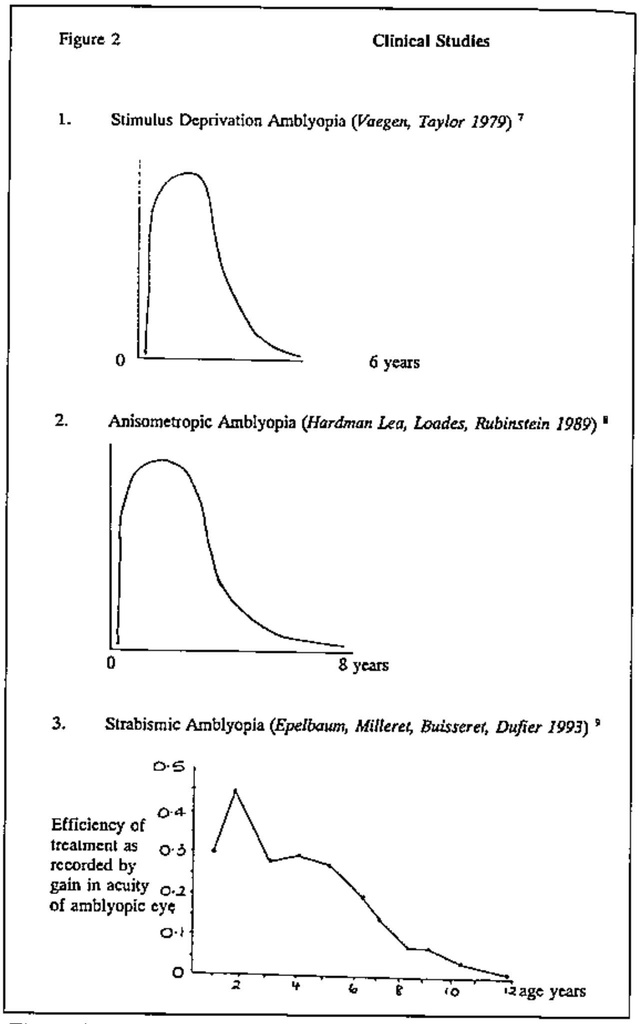

Figure 2.

types of amblyopia[7-9]. For example the sensitive period for stimulus deprivation amblyopia is thought to last to approximately 6 years[7] and anisometropic amblyopia to 8 years[8]. Another study published earlier this year, by Epelbaum, Milleret, Buisseret and Duffier, looked at the sensitive period for strabismic amblyopia[9] (see figure 2). In this paper however, the authors chose to refer to the age of initiation of occlusion therapy rather than try and estimate the age of onset of strabismus. Epelbaum et al discovered, using a population of 407 children with strabismic amblyopia, ranging in age from 21 months to 12 years, that occlusion therapy was most effective between 2 and 3 years of age followed by a progressive decrease in efficiency to 11-12 years of age where therapy was no longer effective. The following case is presented in light of these findings.

CASE STUDY

J. was born at 33 weeks gestation via caesarean section as his mother had suffered chronic renal failure. His birth weight was 1660 grams. J.'s motor development was queried at one month of age and subsequent ongoing investigations led to the diagnosis of cerebral palsy and epilepsy at 9 months.

J.'s visual development was first queried at the age of 6 months, and an ophthalmological assessment at that age found no visual responses. Eyes and fundi were found to be normal. A VER performed at that time failed to elicit a response. A diagnosis of cortical blindness was subsequently made.

Follow up over the next 4 years revealed a gradual development of vision with J. responding to light at 11 months followed by gross movements at 16 months. At 2 years and 8 months J. would reach towards toys held 20cms from his eyes and occasionally roll towards large, brightly coloured toys up to 1m away. By the age of 3 years 4 months it was possible to measure his vision using stycar balls. An acuity of 1.5/60 was achieved, which improved to 6/90 by the age of 4 years 8 months. Part-time left occlusion was commenced at this stage due to J.'s objection to having the left eye covered and his apparent inability to take up right fixation. Occlusion then continued for the next 6 years.

Vision in the left eye continued to improve reaching 6/45 at the age of 5 years 8 months. A gradual improvement of the right vision was observed with the most significant improvement being apparent from the age of 8 years. (See Table 1).

Occlusion was commenced using a face patch. Part-time occlusion was favoured over full time due to J.'s multiple disabilities, extremely poor right vision and resultant limited attention. No improvement was apparent in the first 12 months. The type of occlusion was consequently changed to plano lenses with a translucent occluder to eliminate the effect of intermittent latent nystagmus. Over the next 12 months a slight improvement was documented with J. being able to detect hand movements. This improvement continued with a Teller Acuity Card reading of 1/60 being obtained at the age of 8 years. From the age of 8, visual improvement was increasingly evident and J.'s ability to respond to the Teller Acuity Cards improved thus making test results more reliable. The resumption of a face patch at 8½ years due to J. persistently removing the plano lenses further enhanced the visual improvement with a result of 6/45 being obtained at the age of 10 years. Occlusion continued with equal vision in each eye being maintained over the next 12 months. The fluctuation of acuities from 6/45 to 6/60 in either eye are more a reflection of J.'s co-operation during testing than actual visual fluctuations.

Table 1
Pattern of Visual Improvement

Age	Right Eye	Both Eyes Open	Left Eye
11 months		response to light	
16 months		gross movement	
2.08		large objects	
3.04		1.5/60	
4.08		6/90	
5		left occlusion commenced	
6	?light perception		6/45
7	hand movements		3/60
8	1/60		6/24
8.07	5/60		-
9.03	5/60		6/60
9.05	6/60		6/45
10	6/45		6/45
11.02	6/60		6/60

DISCUSSION

Having reviewed J.'s early history and subsequent development it is thought that Delayed Visual Maturation (DVM) is a more appropriate term than the previous diagnosis of cortical

Table 2
Delayed Visual Maturation - classification

Group 1
- I DVM only
  - (i) poor vision on presentation
  - (ii) history of poor vision, improved prior to examination
- II DVM with systemic illness/perinatal problems

Group 2
- DVM with intellectual impairment

Group 3
- DVM with nystagmus/ocular abnormality

Fielder, Russell-Eggitt, Dodd & Mellor 1985

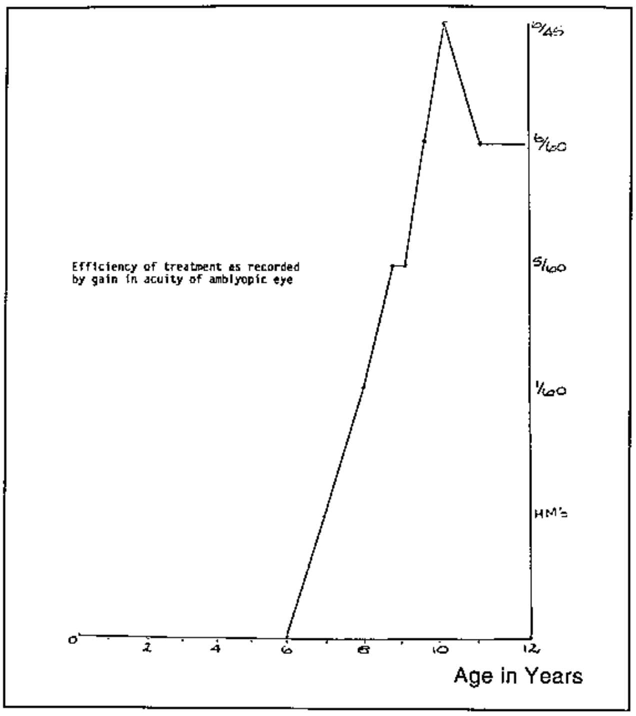

Figure 3. Recorded visual improvement by J.

blindness. Fielder, Russell-Eggitt, Dodd and Mellor reported on three groups of infants with DVM[10] (see Table 2). J.'s visual development is similar to those in group 2 of this classification which consists of children with DVM and a significant degree of mental retardation. These children characteristically have severely reduced visual interest, however, pupil responses and fundal examination are normal. Divergent strabismus is common. The manner in which visual improvement occurs in group 2 infants is characteristically very slow and it can often take many months to decide whether any visual change has taken place. The recovery period has been shown to extend from approximately 6 months to 2 years of age. Mellor and Fielder had also stated in a previous paper that where visual improvement has occurred the possibility of later perceptual problems can not be eliminated[11].

In cases of normal visual development, visual sensitivity is almost complete at 6 months[12] with the critical period for the treatment of strabismic amblyopia extending to approximately 11 years of age[9]. As previously mentioned, occlusion therapy for this type of amblyopia is most effective between 2 and 3 years of age. But what of children with DVM?

J.'s vision did not appear to begin to develop until he was 11 months of age. Development of vision was slow with the maximum binocular visual acuity being recorded at approximately 6 years of age. Occlusion therapy for J. was most effective between 8 and 10 years of age (see figure 3).

During the course of this study the authors were unable to find any clinical studies that looked at extended critical periods in children. However, there is some experimental research which may shed some light on the subject, particularly one study by Cynader and Mitchell[13] that looked at the effect of dark rearing on the visual plasticity of the cat. They found that by rearing cats in the dark they were able to prolong the critical period. They compared the results of monocular lid suturing in dark reared cats with normally reared cats of different ages and found that the time course of deprivation effects in 4 month old dark reared cats was equivalent to that observed in normal kittens who were sutured at 5-6 weeks of age. Cynader and Mitchell concluded that the cortical age of the dark reared cats may be only 5-6 weeks.

Previous studies of social imprinting in birds[14] came to a similar conclusion that restricted exposure leads to responses in older animals which would normally occur only during clearly defined critical periods early in life.

It is possible that there may be a similar lengthening of the sensitive period and the response time to occlusion therapy in cases of DVM in children. There are of course other factors which may contribute to observed visual improvement. Increasing levels of maturity and cognition, for example, may demonstrate an improved ability to co-operate with formal testing procedures and thus show an apparent improvement in visual acuity results. It must be noted however, that in J.'s case although these factors may have contributed to his overall visual improvement there was also notable monocular improvement coinciding with persistent occlusion therapy.

CONCLUSION

The implications of this paper are that children with a diagnosis of delayed visual maturation or perhaps a general developmental delay may have an extended critical period. It is therfore the responsibility of the Orthoptist to consider continuing to implement occlusion therapy beyond defined critical periods.

References

1. Von Noorden GK, Crawford MLJ. The sensitive period. Trans.Ophthal.Soc.U.K.(1979): 99, 442-446.
2. Hubel DH, Wiesel TN. Receptive fields of cells in the striate cortex of very young visually inexperienced kittens. J. Neurophysiology (1963): 26, 994-1002.
3. Hubel DH, Wiesel TN. Binocular interaction in striate cortex of kittens reared with artificial squint, J.Neurophysiology (1965b): 28, 1041-1059.
4. Hubel DH, Wiesel TN. Extent of recovery from the effects of visual deprivation in kittens. J. Neurophysiology (1965b): 28, 1060-1071.
5. Wiesel TN. Postnatal development of the visual cortex and the influence of environment. Nature (1982): 299, 583-591.
6. Ciuffreda KJ, Levi DM, Selenow A. Amblyopia basic and clinical aspects. Butterworth and Heinenmann 1991: (Ch.1).
7. Vaegan, Taylor D. Critical period for deprivation amblyopia in children. Trans.Ophthal.Soc.U.K. (1979): 99, 432-439.
8. Hardmanlea SJ, Loades J, Rubinstein MP. The sensitive period for anisometropic amblyopia. Eye (1989): 3, 783-790.
9. Epelbaum M, Milleret C, Buisseret P, Dufier JL. The sensitive period for strabismic amblyopia in humans.: Ophthalmology (1993): 100, 3, 323-327.
10. Fielder AR, Russell-Eggitt IR, Mellor DH. Delayed visual maturation. Trans.Ophthal.Soc.U.K. (1985): 104, 653-661.
11. Mellor DH, Fielder AR. Dissociated visual development: electrodiagnostic studies in infants who are 'slow to see'. Develop Med.Child.Neurol. (1980): 22, 327-335.
12. Atkinson J. Human visual development over the first 6 months of life. A review and a hypothesis. Hum.Neurobiol. (1984): 3(2), 61-74.
13. Cynader M, Mitchell DE. Prolonged sensitivity to monocular deprivation in dark-reared cats. J. Neurophysiology (1980): 43, 1026-1054.
14. Gottlieb G. Developmental age as a baseline for determination of the critical period for imprinting. J. Comp. Physiol.(1961): 54, 422-427.