

Short Reports

Delayed visual maturation

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SUMMARY Three infants, recognised as blind during the first 4 months of life, were found to be normal on neurological and ophthalmological examinations. Visual electro-diagnostic studies showed normal retinal responses, but delayed conduction velocities and impaired visually-evoked responses over the occipital cortex. After age 6 months, normal vision developed gradually and all abnormalities disappeared.

Maturational delay in several fields of development (for example motor and language) is a well-known phenomenon.¹ Delay in visual maturation is less known to paediatricians, paediatric neurologists, ophthalmologists, and psychologists. Illingworth was the first to draw attention to it.¹ Little has been published on the subject.¹⁻⁶

This paper reports 3 children with such delay. A possible aetiology is suggested by data from electroretinograms and visually-evoked responses performed at the time of referral and on follow-up.

Case reports

Three boys aged 3, 3, and 4 months were referred to us by ophthalmologists as blind. Each had a strikingly similar history. They were born to healthy, unrelated families after normal pregnancies and deliveries. Their parents had remarked that they did not follow objects with their eyes, did not focus, and were unresponsive to visual stimuli, while readily reacting to voices and touch.

Physical and neurological examination showed no abnormalities except a mild roving nystagmus. Diagnostic studies, including electroencephalograms and brain computerised tomography, were normal. Ophthalmoscopic examinations were normal.

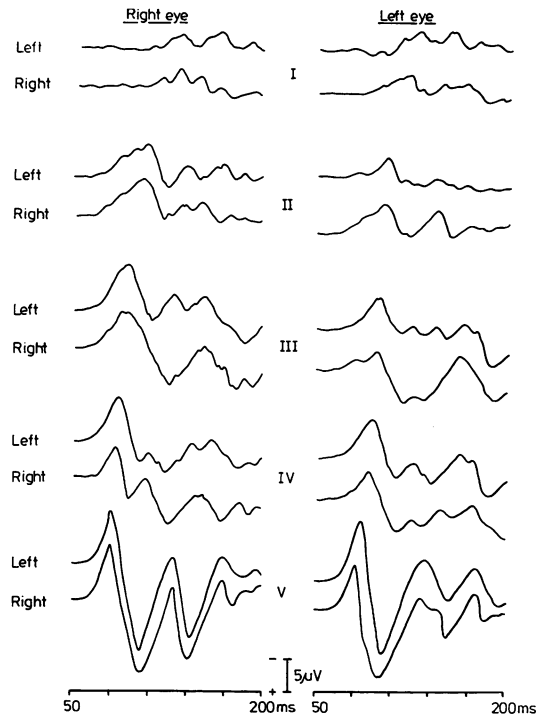


Figure Visually-evoked responses taken at 3-monthly intervals demonstrating the improvement in the brain response to photic stimuli. In the first tracing (I) there is evidence for prolonged latency of the first negative wave. This delay becomes shorter with time until it reaches normal values (50 msec) after a year (tracings IV and V). Concomitantly there is a gradual development of the W-shape wave that follows the initial negative deflection which may correlate with processing of visual information. Recordings are made from over the left and right hemispheres.

Visual electro-diagnostic studies were performed on referral and at 3-monthly intervals.

Methods

Our patients were examined with dilated pupils (Mydracil 12) in a darkened room. One hundred, one per second flashes at 20 cm from the eyes were delivered by a xenon discharge lamp activated by a Grass photostimulator PS-22 at intensity 4. Grass gold-plated electrodes were affixed to the scalp with electrode paste. The visually-evoked response was recorded between the active O_1 and O_2 electrodes, and the linked ears served as reference (10–20 international nomenclature). The resistance between the active and reference electrodes was 2000 to 5000 Ω . The scalp electroencephalogram was amplified and averaged by a Nicolet CA-1000 instrument. The averaged evoked responses to uniocular stimulation were displayed by an x-y plotter.

Results

At referral, the electroretinograms were normal. The visually-evoked responses were of prolonged latency, and only the first negative deflection could be identified (Figure, trace I). Repeated visually-evoked responses at 3-monthly intervals showed constant improvement in the latency and wave forms (Figure, see legend for details).

Clinical follow-up. At 6, 6½, and 8 months respectively the babies showed improving vision, following objects, focusing, and responding to visual stimuli. The roving nystagmus disappeared. At age 2½ years, both eyesight and psychomotor development were normal.

Discussion

Doyne³ was the first to report that the development of sight is delayed in some children. He stressed the common relationship between delay in sight development and mental retardation or albinism. Doggart⁴ reported that some children showed no sign of vision in the early months of life but subsequently developed normal vision. Illingworth¹ was the first to emphasise the problem, described by him as 'delay in visual maturation'.

Our patients are almost identical with the children described by Illingworth.¹ Each of them had a normal perinatal history, and normal neurological and ophthalmoscopic examinations. Each initially had a roving nystagmus that gradually disappeared with visual improvement. Illingworth mentioned the absence of roving nystagmus as an important

negative feature, but this was not the case in our patients. We believe that the roving nystagmus suggests that the children were unable to see, rather than unable to interpret what they saw (visual agnosia). All children had a normal psychomotor development, indicating a picture compatible with a pattern of developmental dissociation.²

Doggart⁴ suggested a possible delay in myelination of the optic nerve. He also referred to the frequency with which optic atrophy (pale discs) in children were wrongly diagnosed. Repeated normal electroretinograms could rule out the possibility of Leber's amaurosis. Mellor and Fielder⁶ in their clinical and electro-diagnostic study of 4 cases, reported that in their infants the initial visually-evoked response showed impaired and immature waveform rather than delayed latencies.

Therefore, they favoured an aetiology based on delayed dendritic and synaptic formation in the occipital cortex rather than unmyelinated pathways. Initial visually-evoked responses from our patients (Figure) clearly demonstrate evidence for prolonged latencies as well as immature waveform responses. These responses became rapidly normal as vision improved. It seems to us that a combination of a transient delayed myelination and synaptic development could better explain the delay in visual maturation. The clinical implication from this assumption could be that an early abnormal visually-evoked response in a blind infant should not necessarily suggest a poor prognosis for vision but rather be seen as an immature response of possible good prognostic value.

The early recognition of this visual phenomenon is not only imperative for prognostic value, but could also prevent unnecessary and sometimes invasive diagnostic studies.

References

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