DELAYED VISUAL MATURATION

BY

R. S. ILLINGWORTH

From the Department of Child Health, University of Sheffield

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In view of the fact that there may be delay in maturation in any of several fields of development (Illingworth, 1961) it would not be surprising if some children showed delay in visual maturation. Having studied the visual development in two such children I searched the literature for information on the matter. The search included all volumes of the Quarterly Cumulative Index Medicus and standard textbooks on ophthalmology. I found very few references to the subject.

Doyne (1930), writing from his experience at The Hospital for Sick Children, Great Ormond Street, London, wrote that 'In some cases the development of sight is only delayed'. He described a child of 3 months of age who showed no sign of vision. Ophthalmoscopic examination showed pigmentary deficiency of the retina and choroid. Sight began to develop from the seventh month, and was normal by 18 months. He referred to the frequency with which mentally subnormal children showed no apparent evidence of vision for some months, and wrote that the commonest cause of delayed sight development was albinism.

Doggart (1957) mentioned the problem of infants who showed no sign of vision in the early weeks, and who subsequently saw normally. He suggested that this might be due to delayed myelination of the optic nerve, described by several French authors as 'La pseudoatrophie grise du nerf optique'. Doggart also referred to the frequency with which optic atrophy and other lesions were wrongly diagnosed. He wrote: 'For every genuine abnormality that escapes detection, at least a dozen imaginary ones are described'.

Duke-Elder (1949) described visual agnosia as the 'inability to recognize or orientate objects perceived, although the reception of their sensory impressions is intact'. He felt that though cases of visual agnosia are seen, the condition is not well recognized.

My colleague, Mr. A. B. Nutt (1960), also referred to the frequency with which fundus changes were suspected in infants referred because of apparently defective vision. He mentioned the fact that the optic disc was paler than that of older children, and that some degree of retinal hypoplasia was normal, so that the macula had a stippled appearance. He wrote that the assessment of sight of an infant did not depend on the appearance of the disc, but on the correlation of the child's visual behaviour with his behaviour in other fields of development.

Law (1961) wrote at length on the problem of the visually defective infant, on the basis of material at the Moorfields Hospital, London. I am indebted to him for allowing me to see the script of the paper before publication. He referred to the difficulty in determining whether a pale optic disc in an infant was normal or abnormal, and emphasized the need for caution in diagnosing optic atrophy in infants. He thought that 80% of infants suspected of having defective vision would prove to have normal or near normal sight, but the evidence for this view was not strong. He described 28 infants referred on account of defective vision. Ten of them, all thought in infancy to have pale optic discs, proved to have normal or almost normal vision. They were followed up for periods ranging from two months to nine years. The others were not adequately followed up, or had gross optic atrophy on examination. Law referred to the frequent finding of mottling of the macula, and of the 'pepper and salt' appearance of parts of the fundus. He rightly referred to the importance of relating the infant's visual perception to the level of mental development. Unfortunately it is not possible to determine from Law's paper the age at which the children began to see or respond to what they saw.

Sorsby and Williams (1960) described blindness in infants with normal eyes or minimal fundus anomalies. The anomalies included the diffuse fine pigmentation called the 'pepper and salt' fundus. They described two families in which children showing these changes developed progressive blindness. The need for caution in interpreting these

minimal changes is obvious in the light of the papers by Law and Sorsby and Williams.

I have not found other papers relevant to the subject, either in French or Italian, nor other papers on blindness in infants without ophthalmoscopic signs; neither have I found a description of cases similar to those below. I have, however, discussed the problem with many paediatric colleagues, and most of them have seen one or two infants presenting features similar to those described.

Below are the case reports of two children seen by me at the Children's Hospital, Sheffield.

Case Reports

Case 1. I saw this boy at the age of 3½ months, through the courtesy of Dr. S. Yudkin of London. He had been born two weeks prematurely, weighing 5 lb. 8 oz. (2,500 g.), after a normal pregnancy and labour, and a normal delivery. There had been no significant infection in pregnancy; and he was well in the newborn period. There were no siblings. There was no family history of delayed development or of other relevant abnormalities.

His parents, who were both intelligent, complained that he could not see. They said that he took no notice of anything, did not focus his eyes and did not smile at them, and that he did not watch them as they talked to him. He had, however, begun turning his head to sound at 12 weeks, which is the average age for this 'milestone', and he had been vocalizing from the age of 7 weeks, which is again average. He was said to smile at nothing in particular when lying in his cot.

On examination his appearance was normal and he was vocalizing normally. The head circumference was average for a small baby—15⅞ in. His weight was 12 lb. 7 oz. In the prone position he bore the weight on the forearms, with his chest well off the couch, and when he was pulled from the supine to the sitting position, there was only minimal head lag. He bore good weight on the legs. The hands were open, and the knee, ankle jerks and muscle tone were normal. He held a rattle placed in the hand and played for a long time with it. In motor and manipulative development he was entirely average, and the vocalizations, though difficult to assess, appeared to be normal. The urine did not contain phenylpyruvic acid.

He appeared to take no notice of his surroundings. He did not watch his mother or father. He did not follow the dangling ring. He took no notice of bright objects, such as a metal hammer. I thought that he appeared to see light, when using the method described by André Thomas and Saint-Anne Dargassies (1952), and the pupils reacted to light. There was no nystagmus and ophthalmoscopic examination was entirely normal. My colleagues, Mr. G. Mackie, and Mr. A. B. Nutt, agreed with these findings.

The baby showed the picture of developmental dissociation (Illingworth, 1958). The diagnosis made was delayed visual maturation, and I told the parents that although it was impossible to be sure, I thought that the outlook was good and that he would see normally in time, and that mental development was normal.

He began to show signs of seeing at the age of 5 months, and at the age of 6 months I saw him laughing. He lifted his head from the supine spontaneously (an achievement typical of a 6-month-old baby). He was now following a light, dangling ring or bell, but not as far as an average 6-month-old baby. He followed objects in the lateral and vertical direction, but not for 180°. He repeatedly opened his mouth when the feeding bottle approached his lips. Motor development was normal. He transferred objects from hand to hand.

At 10 months he weighed 22 lb. The head circumference was 17½ in. He made immediate attempts to pick up the Gesell 8 mm. pellet, but could not quite achieve finger-thumb apposition. There was almost an index finger approach to objects. The grasp of one-inch cubes was mature. Vocalizations were good. He bore full weight on the legs, but would not stand holding on to furniture. He was sitting securely, and had begun to sit without support at 7 months. He actively banged and rang the bell. He repeatedly opened his mouth on the approach of a piece of chocolate. He was not able to wave bye-bye or play pat-a-cake.

The boy was normal in his behaviour; in some aspects of development his developmental level was that of a 9-month-old baby, but he was quite up to the average in motor development. His visual pursuit and visual development were normal.

Case 2. This girl, whom I have described in part elsewhere (Illingworth, 1960) was a full-term baby born normally after a normal pregnancy. She was well in the newborn period. At the age of 3 days she first showed the signs of spasmus nutans. I have not heard of this condition in a child so young.

For the first six months she showed no evidence of vision. She did not smile. Her mother, an intelligent woman, was convinced that she was blind. Her family doctor and a doctor at the infant welfare clinic concurred. At 4 months she took no notice of bright Christmas-tree lights, but she was able at that age to hold and play with a rattle placed in the hand, and she had been turning her head to sound from the age of 3 months. At 5 months she was taken to Mr. Muirhead, ophthalmological surgeon at Chesterfield, to be certified as blind. He declined to certify this, though agreeing that she showed no sign of vision. The pupil reactions, optic discs and fundi were normal. At this age she took no notice of the feeding bottle when it was approaching her, and showed no response until it touched her lips. She began vocalizing 'mama'.

At the age of 6 months she first began to smile at her mother and to follow with her eyes. She now began to reach for objects and get them. The spasmus nutans persisted. At 8 months she was able to sit without support, at 9 months to play pat-a-cake, and at 10 months to wave bye-bye, handle a toy and part with it, and to cast objects on to the floor—all average for the age.

At 11 months she held her arms out for clothes,

which is average for the age. At 16 months she was speaking in sentences, and I was impressed with her advanced speech, her interest in surroundings, and her alertness. She had begun to acquire sphincter control. She was somewhat hypotonic in the lower limbs and did not walk without support until she was 2 years old. I regarded this as isolated delay in motor development, and of no significance. She showed spasmus nutans, with the characteristic lateral movements of the head, brought on by concentration on an object, and the usual peculiar way of looking at objects out of the eye corners. Her eyesight and fundi were normal. At 33 months she was speaking in long sentences, she was able to put on her pants, skirt, cardigan, shoes and socks, and sometimes to fasten buttons. On the picture card she named eight objects (Gesell; average for 36 months), and with the uncoloured geometric forms (Gesell) she achieved the level of a child of 45 months. She did not co-operate in drawing nor in repeating digits. There were slight residual signs of spasmus nutans. It was obvious that she was a bright normal child with normal vision and a better than average level of intelligence.

Discussion

When a child is referred for an opinion because he or she does not appear to see, by far the commonest finding is mental subnormality, as indicated by retardation in other aspects of development. A full developmental examination is therefore necessary. Even if the child is retarded in other fields of development, examination of the eyes and the optic fundus is essential, for optic atrophy, cataract and other eye defects are commonly associated with mental subnormality. If, however, the child who appears not to see at the appropriate age, is found to have developed normally in other respects, and no defect is found in the eye, the likely diagnosis would be blindness or delayed visual maturation. The possibility that the child will see normally in time is an important one to remember, because one has to discuss the prognosis with the parents.

The same applies to the retarded child who does not seem to respond to what he sees. If examination of the eye does not reveal any abnormality, the odds are that the child will see normally when he is older. The difficulty which commonly arises, however, is interpretation of the pale optic disc, which may or may not be normal. If there is doubt about such a disc, the odds are that the child will see normally in time.

I am frequently asked to see babies because of suspected blindness, and the great majority are found to be mentally subnormal. I often feel that they are relatively more retarded in visual recognition than in other fields—an example of what I have termed developmental dissociation (Illingworth, 1958), and it may well be that delayed visual maturation is more common in retarded children than in others, just as generalized delay in maturation is more common in mentally subnormal ones than in children who will prove to have a normal level of intelligence (Illingworth, 1961).

An important feature of children with delayed visual maturation may well be the absence of roving nystagmus. It is difficult to say how soon roving nystagmus is seen in blind children, but it is certainly seen by the age of 3 or 4 months.

When faced with an otherwise normal infant who fails to show visual response, there is a remote possibility of autism. Such a child may fail to smile at his mother or to show interest in his surroundings.

It is not possible to say whether the child with delayed visual maturation is unable to see, or is able to see but unable to interpret what he sees (visual agnosia).

The condition of delayed visual maturation in an otherwise normal child is uncommon, but it is clear from discussion with other paediatricians and with ophthalmologists that it is not very rare. It will probably always be impossible to give an accurate prognosis to anxious parents, because it may well be that there are many gradations between blindness which will be permanent and 'blindness' which will be temporary, as in the two children described. Prognosis, therefore, must always be guarded, but it is a great help to parents if reasonable hope of complete normality can be given.

Neither of these children has yet been followed into school life and there is always the possibility that perceptual difficulties will reveal themselves in later years.

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