SOME OCULAR MANIFESTATIONS OF PREMATURETY*

BY

N. L. McNEIL

Barnsley, Yorks.

It is not proposed to add further to the literature on retrolental fibroplasia by describing further cases of the fully-developed disease. The picture presented by those cases not showing the fully-developed cicatricial end-stages has not been described to nearly the same extent in Great Britain.

In a classification of the cicatricial end-stages of retrolental fibroplasia produced by Reese, King, and Owens (1953) for the National Society for the Prevention of Blindness (U.S.A.), five grades were described, ranging from a small peripheral opaque mass in the fundus without visible retinal detachment, to retrolental tissue covering the entire pupillary area. Szewzyk (1953) also discussed a terminology and classification. Both systems were primarily based on variations in fundus appearances. There was less emphasis on functional variations. In the cases described below the functional capacities as well as the ophthalmoscopic appearances are recorded.

Case Histories

Case 1, a male infant, born in July, 1951, birth weight 4 lb., 8 weeks premature, oxygen therapy after birth (duration and concentration not known). Now aged 4 years, and appears to be a normally intelligent child of normal size and weight.

Right Eye.—Anterior synechiae present 7 to 10 o'clock, broad retinal fold running from the optic disc downwards and temporally becoming flatter as it merges into an indistinct peripheral mass. Retina atrophic in central area with some shiny crystals.

Visual Acuity.—Nil or virtually so. Refraction +2 D sph.

Left Eye.—No synechiae, disc enlarged and thickened by excess formation of glial tissue, vessels run over the disc, mostly temporally in a broad sheath (Fig. 1, opposite). No nystagmus.

Visual Acuity.—Impossible to assess, but the patient can see reasonably well with spectacles, though he gazes to the left whilst looking at an object straight ahead. Refraction -9 D sph., 3 D cyl. at 90°.

Case 2, a male infant, born in December, 1951, birth weight 3 lb. 6 oz., about 8 weeks premature, oxygen therapy for an unknown period. The child is definitely backward in physical and mental development.

Right Eye.—No synechiae, a large grey raised band runs from the disc temporally into a large peripheral grey mass. The remainder of the retina is atrophic with mottled pigmentation.

Visual Acuity.—Perception of light.

Left Eye.—Posterior synechiae, complete retrolental membrane present, early cataractous changes occurring in the lens, nystagmus. Refraction not known.

Case 3, a male infant, born in June, 1952, birth weight 3 lb., 8 weeks premature, oxygen therapy for an unknown period. The child is backward in development.

Right and Left Eyes.—No synechiae, a large broad band runs from optic disc temporally

*Received for publication March 28, 1955.

FIG. 1.—Case 1, left eye, showing thickened raised disc, abnormal vessel pattern, and depigmented myopic retina.

in both eyes and disappears peripherally and anteriorly, the remainder of the retina shows atrophic changes with some crystalline deposits in the left eye, nystagmus.

Visual Acuity.—Perception of light. Refraction not known.

Case 4, a female child, born in November, 1947, birth weight 3 lb., 8 weeks premature, oxygen therapy for an unknown period. The child is normal physically and mentally.

Right Eye.—Disc pale and appears to have been pulled temporally, a narrow raised fold runs from the disc temporally into a peripheral hyaline mass, and a non-branching vessel runs in the free edge of the fold; the pattern of the remainder of the retinal vessels is abnormal (Fig. 2).

Visual Acuity.—Hand movements. Refraction -2 D sph.

FIG. 2.—Case 4, right eye, showing distorted disc, non-branching vessel running on free edge of fold, abnormal retinal vessel pattern, and peripheral mass.

Left Eye.—Distortion of disc as in right eye. An indefinite thickened greyish retina runs temporally for about three disc diameters, the retinal vessels form a narrow horizontal V pattern. Bilateral rotatory nystagmus.

Visual Acuity.—6/24 pt. Refraction $-4\text{ D sph.}, -2\text{ D cyl. at } 90^\circ$.

Case 5, a female infant, born in February, 1953, birth weight 2 lb. 12 oz., about 8 weeks premature, in oxygen for an unknown period. For the first 18 months of life, this child was backward both mentally and physically, but recently there has been a marked improvement and she is now developing well physically, and visually is taking much more notice.

Right and Left Eyes.—A well-marked fold runs temporally from the disc into a peripheral mass. The vessels over the remainder of the retina are few in number and attenuated, a large vessel runs in the free edge of the fold in each eye. Coarse nystagmus. Eccentric gaze. Refraction -3.5 D sph.

Case 6, a male child, born in May, 1950, birth weight 2 lb. 6 oz., 8 weeks premature, was in oxygen for an unknown period. A physically normal child.

Right Eye.—Distorted pale disc with dragging over of the vessels towards the temporal side, atrophic-looking retina, no fold seen.

Visual Acuity.—About 6/60. Refraction $-11\text{ D sph.}, -3\text{ D cyl. at } 180^\circ$.

Left Eye.—Posterior synechiae present, atrophic iris, retrolental membrane. Eccentric fixation. Rapid horizontal nystagmus.

Case 7, a boy, born in July, 1940, birth weight 3 lb., 8 weeks premature, in oxygen for an unknown period. Has a right hemiplegia dating from birth which has begun to recover slightly. First attended at the age of 14 years with a right convergent squint which had been present for years but was ignored because of his other disabilities.

Right Eye.—A narrow retinal fold on the nasal side of the fundus runs from the disc to the ora, no peripheral mass, a vessel runs on the free edge of the fold, the remainder of the retina is atrophic in appearance. Small hypermetropic errors.

Visual Acuity.—Less than 6/60.

Left Eye.—Normal appearance. Small hypermetropic errors.

Visual Acuity.—6/6.

***Case 8, a male child, born in December, 1949,** birth weight 3 lb., in oxygen for an unknown period. The child is backward and has a congenital absence of the left hand. He first attended at the age of 4 years with a convergent squint.

Right and Left Eyes.—Both fundi show a laterally distorted disc with the retinal vessels running directly temporally. Myopic crescent in the left eye with some pigmentation and mottling of the macular area.

Visual Acuity.—Difficult to estimate but with spectacles appears to be reasonably good:

Right $-7.5\text{ D sph.}, -1.5\text{ D cyl. at } 90^\circ$.

Left -8.5 D sph.

Left convergent strabismus with bilateral external rectus palsy, no nystagmus.

Case 9, female infant, born in May, 1950, birth weight 3 lb., 8 weeks premature, oxygen given for an unknown length of time. A normally developed child.

Right Eye.—Depigmented fundus but no other abnormality. Refraction $-1.75\text{ D sph.}, -1\text{ D cyl. at } 10^\circ$.

Left Eye.—Disc full and slightly raised in front of level of surrounding retina. Some distortion of disc and vessels. Refraction -4.25 D sph. No nystagmus.

Case 10, female infant, born in May, 1952, birth weight 2 lb. 13 oz., 8 weeks premature, oxygen for an unknown period. A normally developed child. First attended at the age

* This boy has two brothers, the elder of whom weighed 2 lb. 11 oz. at birth and the other 5 lb. The former was never given oxygen and has refractive errors of about $+1\text{ D sph.}, +1.5\text{ D cyl. at } 90^\circ$ in each eye; his vision is good. The second brother is said to have no visual defect and has not been examined.

of 2 years with a left convergent squint. Neither eye showed any fundus abnormality apart from a myopic appearance. Refraction -9 D sph.

Case 11, female child, born in February, 1948, birth weight $4\frac{1}{2}$ lb., 4 weeks premature, no oxygen given, a normal healthy child. First attended at age $2\frac{1}{2}$ with a right convergent squint.

Right Eye.—Posterior polar cataract, an unusual retinal fold appears to be suspended from a persistent hyaloid artery; the upper free border of the fold extends from the lens to the disc, and the lower free border sweeps down on to the retina to a point half way between the disc and equator at 7 o'clock (Fig. 3). The peripheral retina shows some areas of healed choroidal inflammation.

Left Eye.—Normal appearance, small hypermetropic error.

FIG. 3.—Case 11, semi-diagrammatic fundus painting of right eye, showing lower free edge of fold sweeping on to retina with choroidal lesions lying more peripherally. The details of the surfaces of the fold cannot be seen as they run directly backwards.

Case 12, a boy, born in May, 1940, birth weight 3 lb. 15 oz., 6 weeks premature, whether he received oxygen after birth is not known.

Right and Left Eyes.—A typical myopic appearance and vitreous floaters:

Visual Acuity: Right -11 D sph., -2 D cyl. at $90^\circ = 6/12$.

Left -11.5 D sph., -1.5 D cyl. at $90^\circ = 6/18$.

Case 13, a girl, born in July, 1943, birth weight 4 lb. 5 oz., 8 weeks premature, no oxygen therapy. This child is normal physically.

Right and Left Eyes.—The fundi appear to show severe myopia, the fundi and discs being pale with a large conus but no other abnormality. No nystagmus.

Visual Acuity: Right -22 D sph., -1 D cyl. at $180^\circ = 6/18$.

Left -21 D sph. $= 6/12$.

It seems hardly believable that this child was not found to have defective vision until the age of 10. Since then there has been no appreciable change in the refraction.

Case 14, a male infant, born in June, 1953, birth weight 6 lb., said to be 6 weeks premature, and to have had premature characteristics in spite of the weight. No oxygen therapy. A

normal child who attended for a right convergent strabismus. The vision appeared to be within normal limits, with no fundus changes and no nystagmus.

Refraction: Right -0.75 D sph., -1.5 D cyl., at 180° .
Left -0.75 D sph., -0.5 D cyl., at 180° .

Cases 15 and 16, twin girls, born in 1946, birth weight 5 lb. each (approx.), 4 weeks premature, no oxygen therapy, normal physically. They were first seen at the age of 6 years and were found to have severe myopia. Their fundi show no abnormality other than a myopic appearance. No nystagmus.

Refraction: Joan Right -10 D sph. Left -7.5 D sph., -2 D cyl., at 180° .
Josephine Right -13 D sph. Left -14 D sph., 2 D cyl., at 180° .

Discussion

Cases 1-7 could be classified as cases of abortive retrolental fibroplasia. Cases 8, 9, and 10 might also be included in this group. The remainder would not appear to fit into the retrolental fibroplasia category, though they were born prematurely and have ocular anomalies.

Rubinstein (1952) described five cases of retrolental fibroplasia occurring in this area, and of the ten eyes described five did not show the final end-stage of the disease. In the series of visual defects in premature children recorded in this paper, only two of 32 eyes showed a true retrolental fibroplasia. It would thus appear that the incidence of ocular defects other than a retrolental membrane are more common than the presence of the membrane itself. The following defects occurred in more than one case in this series:

(1) Myopia.—Present in 22 out of 32 eyes. There is no doubt that myopia is a frequent result of prematurity and this fact has not escaped many American observers.

In this small series the defect occurred both with and without an associated anatomical defect and also both with and without the use of oxygen in the immediate post-natal period. The myopia varied in severity. Eight of the children were discovered to have myopia before the age of 5 years—a fact which is in itself unusual. It is the writer's impression that these myopes are not progressive in the same way as "normal" myopes, but the period of observation is too short and the number of case too few for a definite opinion to be expressed.

None of the cases included in the series had a myopic family history. Two other premature myopic children were excluded from the series on the grounds of a strong myopic family history. Case 8 is of interest because of his two premature brothers who are not myopic. It may be that oxygen therapy was an added factor in the development of the myopia in this case.

(2) Retinal Folds.—These are well-known end-results of the oxygenation of premature children, and form Grade 3 of the American classification referred to at the beginning of the paper. Case 11 may be out of context in this series but the unusual fold occurring in a premature child (who did not receive oxygen) warrants its inclusion; apart from the unexplained choroidal (? foetal) lesions there had been definite failure of normal regression of the hyaloid system and the formation of a fold. Mann (1937) described the ophthalmoscopic appearances of congenital retinal folds as follows:

The solid appearance of the fold, the septum-like connection with the retina, the presence of branches of the retinal artery on the fold, and the attachment anteriorly to the ora serrata with strands passing to the lens near the equator are characteristic.

The temporal situation of most folds and the presence upon them of embryonic vessel remnants were also emphasized. This description applies generally to the folds found in premature oxygenated children. Some of the cases in this paper have non-branching vessels on the folds which are possibly hyaloid in origin. It is certain that retinal folds occur in full-term children. Jameson Evans (1954) confirmed that Mann's Case 5 was a normal child of 7 lb. birth weight. It does appear that the formation of retinal folds is dependent primarily on the failure of regression of the hyaloid system, whatever the cause of this latter may be. Ashton (1954), Ashton and Cook (1954), and Ashton, Ward and Serpell (1954) have fully described the further stages in the development of retrolental fibroplasia by a process of vaso-proliferation, mesenchymal proliferation, and contractures.

(3) Distortion of the Disc and Abnormal Vessel Patterns.—These defects are the results of traction on the vessels and disc by a cicatrizing process in the periphery of the fundus.

(4) Gliosis of the Disc.—Excess formation of glial tissue (the mesenchymal cells of Serpell, 1954) is the basis of this appearance.

(5) Eccentric Fixation.—This is associated with a functional displacement of the fixation centre to the temporal side of the normal macular position. Rather unexpectedly one case had no nystagmus.

(6) Nystagmus.—This varied from a coarse searching type to a fine rapid nystagmus; anatomical deformity of the fundus was present in all cases showing this sign.

Summary

The ocular condition of a number of premature children is described. All of them showed some ocular defect not amounting to the typical picture of retrolental fibroplasia. Some of the defects are discussed briefly and enumerated.

I am indebted to Mr. E. G. Mackie and Miss E. Hatherley for permission to report on some of the cases. I wish to thank the Barnsley Hospital Management Committee for the loan of the fundus paintings.

REFERENCES

- ASHTON, N. (1954). *British Journal of Ophthalmology*, **38**, 385.
———, and COOK, C. (1954). *Ibid.*, **38**, 433.
———, WARD, B. A., and SERPELL, G. (1954). *Ibid.*, **38**, 397.
EVANS, P. JAMESON (1954). Personal communication.
MANN, I. (1937). "Developmental Abnormalities of the Eye". University Press, Cambridge.
REESE, A. B., KING, M. J., and OWENS, W. C. (1953). *Amer. J. Ophthal.*, **36**, 1333.
RUBINSTEIN, K. (1952). *British Journal of Ophthalmology*, **36**, 303.
SZEWCZYK, T. S. (1953). *Amer. J. Ophthal.*, **36**, 1336.
SERPELL, G. (1954). *British Journal of Ophthalmology*, **38**, 460.