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Sir,

Lipaemia Retinalis in a Premature Infant with Type I Hyperlipoproteinaemia

A premature infant who was found to have lipaemia retinalis due to hyperlipidaemia of unknown cause was recently reported.[1] We would like to report a premature infant with type 1 familial hyperlipidaemia[2] who developed lipaemia retinalis in association with stage 3 retinopathy of prematurity (ROP).

A premature infant born at 26 weeks gestational age (birth weight 960 g) was found to be markedly lipaemic at 37 weeks gestational age with a triglyceride level of 23.7 mmol/l (normal range 2–4 mmol/l) that rose to a peak of 78 mmol/l before being converted from expressed milk feeds to medium-chain triglyceride (MCT) milk. A diagnosis of hyperchylomicronaemia (Fredrickson type 1 hyperlipoproteinaemia) was made. There was no family history of any lipid disorder and both parents had normal lipid profiles.

Screening for ROP was undertaken from 32 weeks gestational age. Initially, stage 1 ROP in zone III was present in both eyes.[3] At 37 weeks, 2 clock-hours of stage 3 ROP in zone III was observed in the left eye.

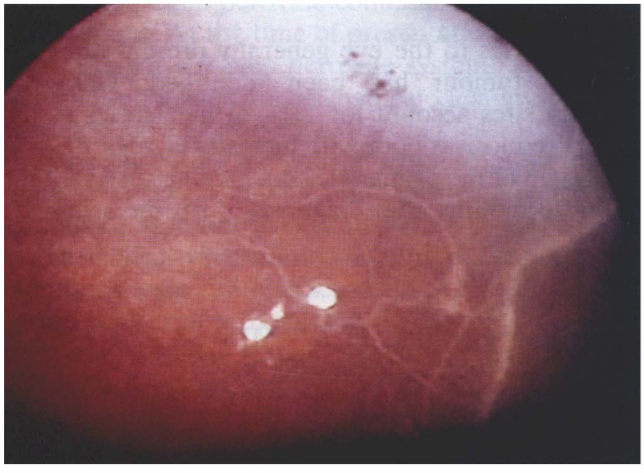

Fig. 1. *Fundus photograph of the left eye at 37 weeks gestational age showing lipaemia retinalis and ROP stage 3 in zone III.*

The retinal vessels remained of normal calibre and there was no 'plus' disease. At 40 weeks the retinal vessels were noted to be uniformly pale pink in appearance (Fig. 1). The plasma triglyceride level at this time was 59 mmol/l. The lipaemia retinalis and ROP both resolved leaving normal retinal vasculature by 43 weeks.

The baby's triglycerides fell to normal on a 25% fat diet made up of 87% MCT lipids. Mother was able to introduce breast feeds at night with MCT lipids by day, with satisfactory growth and triglycerides in the range 5–7 mmol/l. At 2 years he is well, still on the low fat diet but has a mild delay in development and normal triglycerides.

Lipoprotein lipase deficiency, which is the commonest cause of familial hyperchylomicronaemia,[4] was excluded by enzyme assay of post-heparin plasma. It is likely that the patient was deficient in the lipoprotein lipase cofactor apoprotein C-II, but this assay was not available.

Lipaemia retinalis was discovered as an incidental finding during screening for ROP. While it is possible to speculate that an increased plasma viscosity might predispose to abnormal development of the retinal vasculature, the infant was at high risk of ROP in any event and such cases generally resolve spontaneously. Furthermore, in the fully developed retina angiographic evidence suggests that retinal perfusion in lipaemia retinalis is normal.[5]

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Sir,

An Unusual Case of Acute Idiopathic Blind Spot Enlargement Syndrome

We read with great interest reports suggesting multiple evanescent white dot syndrome (MEWDS), multifocal choroiditis (pseudo-presumed ocular histoplasmosis) and acute macular neuroretinopathy (AMN) are linked by the acute idiopathic blind spot enlargement syndrome (AIBES).[1–3] We present a case of AIBES that was preceded by an episode of acute posterior multifocal placoid pigment epitheliopathy (APMPPE) 4 years earlier.

The patient, a healthy 51-year-old woman, presented in April 1991 with a 6-day history of stationary floaters, described as 'like leaves on a tree', affecting her left eye. She denied any preceding 'flu-like illness. On examination, best corrected Snellen acuities were 6/6 in the right eye and 6/36 in the left. Pupil reactions were normal. A very mild anterior uveitis was noted. Slit lamp biomicroscopic examination of the fundus showed multiple post-equatorial circumscribed, flat, greyish-white lesions at the level of the retinal pigment epithelium. These lesions varied from one-third to one-half of a disc diameter in size. Fluorescein angiography showed early blockage of background choroidal fluorescence with late staining of the lesions (Fig. 1). A diagnosis of APMPPE was made and over the ensuing 18 months left eye acuity improved to 6/6 and the fundal lesions disappeared. The patient re-

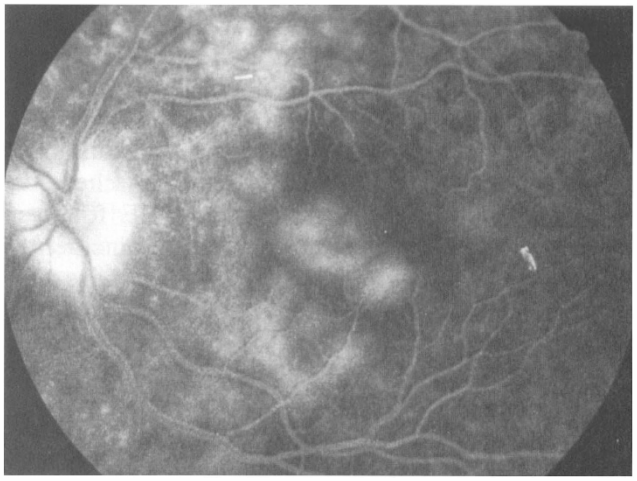

Fig. 1. *Late fluorescein frame, demonstrating late staining of lesions, at initial presentation in April 1991.*

presented in May 1995 complaining of a 7-week history of a 'whirling' photopsia affecting the left temporal field. We noted the absence of preceding 'flu-like illness. Examination revealed corrected Snellen acuities of 6/6 in the right eye and 6/6−1 in the left. Pupil reactions were normal, as was the anterior segment examination. Fundus examination revealed a normal appearance in both eyes. In particular no white dots or macular granularity were noted. We felt that the history of positive visual phenomena coupled with a normal fundal appearance suggested AIBES. Goldmann visual fields revealed a 20° enlarged blind spot with steep margins in the left field (Fig. 2), thus confirming our suspicion. At her last review in February 1996 the patient's symptoms had improved; however, her visual fields remain unchanged.

To our knowledge this is the first case of AIBES preceded by APMPPE in the same eye. Many authors including Gass[2] feel that MEWDS is a subset of AIBES. Various authors describe evidence

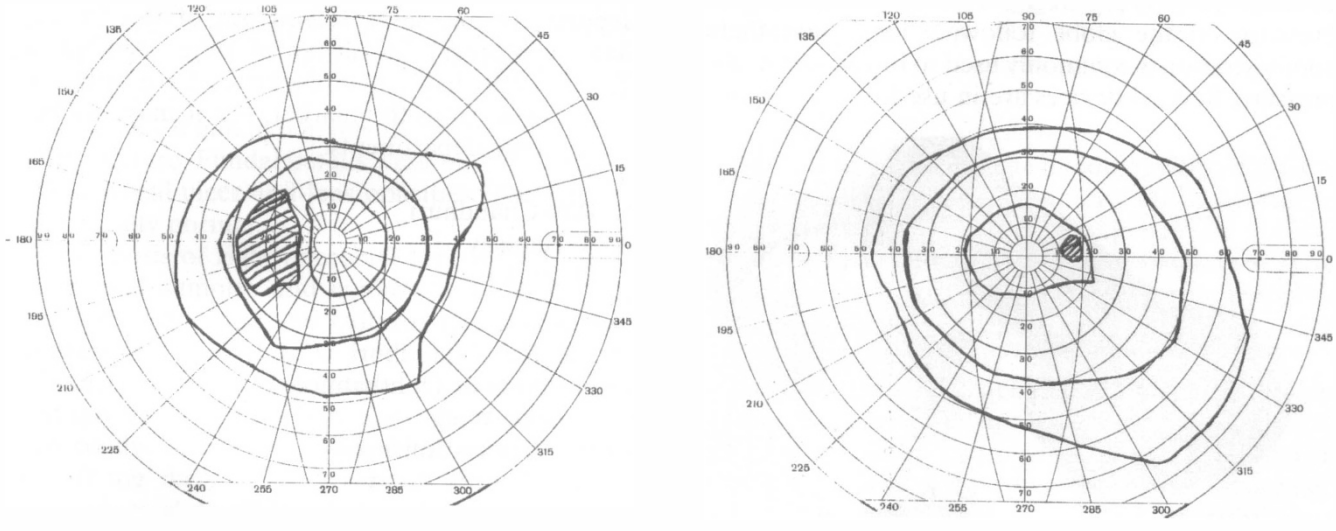

Fig. 2. *Goldmann visual fields at second presentation in May 1995. Note the grossly enlarged blind spot in the left visual field.*