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Sir,

Retinal haemorrhages following Retcam screening for retinopathy of prematurity

We read with interest the article by Adams *et al*[1] describing retinal haemorrhages following Retcam examination for retinopathy of prematurity (ROP). On both visits, they detected no retinal haemorrhages initially by Retcam, which were detected later by indirect ophthalmoscopy, although they do not mention the stage of vessel maturation or presence of ROP. We routinely perform ROP screening by the Retcam and have not observed any retinal haemorrhages. Following the authors report, we performed indirect ophthalmoscopy 60 min after ROP screening with Retcam in 50 eyes of 25 children; however, failed to detect such retinal haemorrhages and it seems to be of rare occurrence. It is possible that immature fragile vasculature in very premature babies as in this case or very vascular ROP may present with retinal haemorrhages by inadvertent ocular pressure during the Retcam examination.

A rise in intraocular pressure is not uncommon as disc pulsations are induced during examination when pressure is applied from the hand piece. Although we use the second-generation 130-degree ROP lens, the presence of small pupils and persistent ocular movement makes it difficult to visualize the periphery; with a need to tilt the head and the hand piece in various configurations to obtain a suitable view, which causes an increase in pressure. It is essential to ensure that the coupling solution is replenished repeatedly as it flows out of the eye during the examination, as a lack of it causes a blurring of image, with more manoeuvers by the observer. Proper immobilization of the head is essential to prevent sudden head jerks and consequent injury.

With modern neonatal care as younger preterm infants survive, such vascular incidents may be more common. Although such cases are rare, this report guides us to take utmost care during Retcam examination.

Acknowledgements

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Reference

1 Adams GG, Clark BJ, Fang S, Hill M. Retinal haemorrhages in an infant following RetCam screening for retinopathy of prematurity. *Eye* 2004; **18**(6): 652–653.

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Sir,

Reply to RV Azad *et al*

We thank Professor Azad and colleagues for their interest in our case report.

At the time of examinations the baby was between 32 and 34 weeks gestation, with no suggestion of abnormalities of retinal vascularisation.

We concur with their view that this is a rare occurrence that we noted during an audit of RetCam screening against conventional indirect ophthalmoscopy. At the time of these events, the manufacturers considered that some 1 million RetCam examinations had taken place with no other similar report. They were not aware of any other Unit undertaking a similar audit process, and we are therefore interested to know that Professor Azad and colleagues have not demonstrated a similar occurrence in their study.

We agree with Professor Azad and colleagues that care must be taken not to apply excessive pressure on the eye when using the RetCam. We advise that all neonatal screening should use the lighter ROP screening head and not the heavier standard

paediatric head which would produce a greater load on the ocular surface.

We agree that our case report is extremely rare but it does emphasise the fragile nature of the immature retinal vascular system.

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Sir,

Stargardt's disease and retinitis pigmentosa: different phenotypic presentations in the same family

The hereditary macular dystrophies are progressive degenerations of retinal and choroidal tissue. Genetic studies have shown that a single mutation or mutations in different parts of the same one gene can result in different macular dystrophies. Mutations in the Stargardt's disease gene (ABCA4) was shown to cause also fundus flavimaculatus, autosomal recessive retinitis pigmentosa (RP), and cone rod dystrophy (CRD).[1,2] Since they are all the result of mutations in genes that are presumed to express in either the photoreceptor cells or the retinal pigment epithelium (RPE), it would not be surprising to find variable presentations in members of the same family.[1–3]

Here, we report an atypical macular dystrophy in a young female who has two brothers with typical RP.

Case report

A 21-year-old woman, presented with gradually progressing poor vision starting about 15 years ago. She denies a significant difference in day and night vision. Her parents were first-degree relatives and she had two brothers complaining of poor night vision. Visual acuity was 20/400 in both eyes. There was no nystagmus. Ophthalmic examination was unremarkable except fundus examination, which revealed a well-circumscribed one-disc diameter area of choroidal and RPE atrophy in the fovea OD (Figure 1a) and a larger area of severe choroidal and RPE atrophy associated with posterior bowing (posterior staphyloma) in the macular area with a whole thickness macular hole and a few pigment clumps OS (Figure 1b). There was minimal or no arteriolar attenuation with a pink optic disc bilaterally. A punctate retina pigment epitheliopathy could be noticed in the midperipheral retina bilaterally, which is evident in fluorescein angiography (FA). FA showed window defect in the fovea surrounded by a hypofluorescent circular area of choroidal atrophy OD and, hypofluorescence of the whole macular area because of severe choroidal and retina pigment epithelial atrophy OS (Figure 2). Electroretinography (ERG) revealed severely affected cone and rod responses in both eyes (Figure 3).

The first brother, 32 years old, had a BCVA of 0.6 OD and 0.7 OS. There were arteriolar narrowing, waxy-pallor optic discs, bone-spicule pigmentation of the peripheral

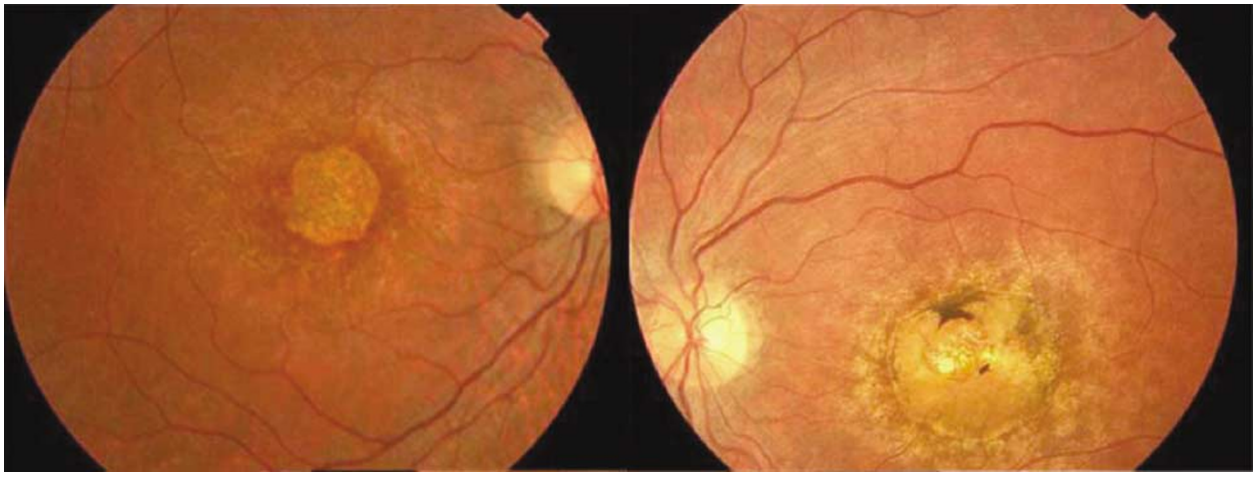

Figure 1 Fundus photographs of (a) the right eye and (b) the left eye of the patient.