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CFEH Facebook Case #86

A 32 year old Middle Eastern male has had a right hyper-exotropia and poor vision in his right eye since he can remember. What in his history could have caused the retinal appearance seen below?



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Retinopathy of prematurity.

This patient was born prematurely at 30 weeks gestation. The Optomap and fundus autofluorescence images show vascular sheathing at the disc and peripheral retina in both eyes, tortuous blood vessels, peripheral retinal hyperplasia (more notable in inferiorly in the right eye and temporally in the left), circumferential retinoschisis with vitreomacular traction in the left eye and an infero-temporal ridge in the right eye. The optic disc also showed signs of dragging away from the fovea in the right eye.

Retinopathy of prematurity has 2 main phases – firstly there is delayed growth of the retinal vessels after birth, and a partial regression of vessels that have developed. This is followed by the growth of pathological vessels, stimulated by retinal hypoxia.

The main risk factors for this condition include preamture birth (before 31 weeks), a low birth weight (less than 1.25kg) and the use of oxygen following birth. The smaller the baby and earlier the birth, the higher the risk of ROP, however not all premature babies will develop this condition.

The pathophysiological process is, in brief, as follows. The high oxygen levels suppress VEGF in the early stages of ROP, inhibiting normal vessel growth. This is followed by the second phase of ROP where retinal hypoxia induces high levels of VEGF caucsing pathological retinal vessels to develop. Tractional retinal detachment(s) can occur in the later stages of this disease.

This condition is classified into 5 stages, ranging from stage 1 (mildly abnormal blood vessel growth requiring no treatment) through to stage 5 (tractional retinal detachment and blindness). In severe cases, timely treatment in the form of laser therapy or cryotherapy is required to avoid vision loss.