LETTERS TO THE EDITOR

Congenital rubella syndrome

EDITOR,—We would like to compliment Givens *et al*[1] on their paper drawing attention to the persistence of severe ocular and systemic complications of the last rubella pandemic of 1963 to 1965. They note an association between glaucoma and cataract, and also microphthalmia in congenital rubella syndrome. However, they do not note the occurrence of a type of glaucoma with congenital rubella syndrome characterised by marked hypoplasia of the iris but without cataract or microphthalmia. This glaucoma is usually overlooked at an early age presenting relatively late when visual loss has already progressed to a serious degree. This type of glaucoma is commonly associated with deafness, retinopathy, and cardiac defects. As most patients are deaf mutes their visual disturbance is easily overlooked. The iris hypoplasia is accompanied by marked hypoperfusion contributing we believe to the progressive nature of the condition so that intraocular pressure (IOP) becomes more difficult to control with increasing age.

We have previously reported four female patients with hypoplasia of the iris due to rubella embryopathy and accompanying glaucoma.[2 3] An additional male patient[3] with iris hypoplasia did not have a raised IOP but we have not seen him since 1980. We have now an additional three female patients with typical iris findings, two with glaucoma, but no further male patients.

We feel it is important that this condition of subtle onset is recognised before visual loss is too severe.

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1 Givens KT, Lee DA, Jones T, Ilstrup DM. Congenital rubella syndrome: ophthalmic manifestations and associated systemic disorders. *Br J Ophthalmol* 1993; 77: 358–63.
2 Brooks AMV, Gillies WE. Glaucoma associated with congenital hypoplasia of the iris stroma in rubella. *Glaucoma* 1989; 11: 36–41.
3 Gillies WE. Hypoplasia of the iris stroma in Gregg's syndrome. *Aust NZ J Ophthalmol* 1980; 8: 189–92.

Reply

EDITOR,—We thank Anne M V Brooks and W E Gillies for highlighting the association of iris hypoplasia with glaucoma in patients with congenital rubella syndrome. Iris hypoplasia is easy to overlook and would be an important risk factor for the development of glaucoma later in life. We also note that iris hypoplasia has been reported in association with juvenile onset glaucoma with an autosomal dominant inheritance pattern. This particular type of familial iris hypoplasia is characterised by hypoplasia of the anterior iris stroma, a prominent pupillary sphincter, trabeculodysgenesis, and glaucoma.[1–3] We did not observe this particular pattern of iris hypoplasia in our glaucoma patients with congenital rubella syndrome. Perhaps the pattern of iris hypoplasia as described by Brooks and Gillies is different. With the multitude of developmental abnormalities associated with congenital rubella syndrome, iris hypoplasia with associated trabecular meshwork and outflow abnormalities would be consistent. We thank Brooks and Gillies for describing this important clinical finding in patients with congenital rubella syndrome, which would put them at risk of development of glaucoma.

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1 Jerndal T. Dominant goniodysgenesis with later congenital glaucoma. *Am J Ophthalmol* 1972; **74**: 28.
2 Martin JP, Zorab EC. Familial glaucoma. *Br J Ophthalmol* 1974; **58**: 536.
3 Weatherill JR, Hart CT. Familial hypoplasia of the iris stroma associated with glaucoma. *Br J Ophthalmol* 1969; **53**: 433.

Dark adaptation and scotopic perimetry over 'peau d'orange' in pseudoxanthoma elasticum

EDITOR,—Pseudoxanthoma elasticum (PXE), a systemic disorder of elastic tissue involving the eye, is transmitted in either an autosomal dominant or autosomal recessive fashion.[1] In addition to angioid streaks ocular findings in patients with PXE include 'peau d'orange', which may precede angioid streak formation.[2] The term was introduced by Smith and coworkers[3] and appears to be synonymous with the 'fond muchté' of Bischler,[4] and the 'mottled fundus' of Shimuzu.[5]

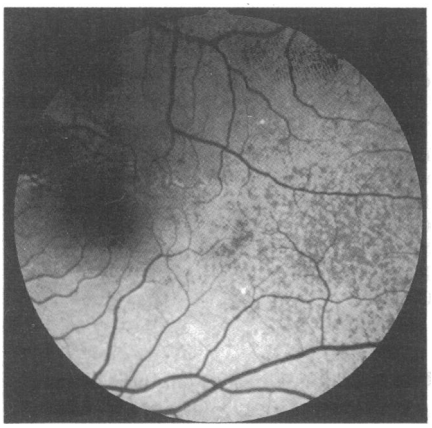

Figure 1 Typical ophthalmoscopic appearance of peau d'orange in one of the patients studied with pseudoxanthoma elasticum.

Ophthalmoscopically, affected areas show scattered, subconfluent yellowish lesions in a peculiar stippled pattern at the posterior pole (Fig 1). It is speculated that it is due to a focal degeneration of the elastic portion of Bruch's membrane causing thickening and calcification.[6] A variety of changes at the level of Bruch's membrane associated with thickening and abnormal deposits as in Sorsby's fundus dystrophy and age-related macular degeneration were found to be associated with altered dark adapted retinal function.[7 8]

In a prospective study we investigated six patients with PXE and peau d'orange (aged 24–55 years, mean 35·7 (SD 11·9) years) to determine whether underlying structural changes in peau d'orange are associated with impairment of retinal function. Patients with PXE and peau d'orange underwent routine clinical evaluation. All eyes had angioid streaks, and in two eyes fibrovascular scars from choroidal neovascularisation were present. For psychophysical studies using published techniques,[7] the pupil was dilated with cyclopentolate 1%, and the patient was dark adapted for 45 minutes.

Dark adapted static perimetry was done in all patients to document possible sensitivity loss. After light adaptation sufficient to bleach >95% of the available rhodopsin the modified Humphrey automated perimeter was used to determine dark adaptation curves. Dark adaptation was measured in areas showing peau d'orange and compared with normal controls of the same age group.

In all patients dark adapted sensitivity was normal over areas of peau d'orange using red and blue stimuli. Dark adaptation curves showed a distinct rod-cone break, and both the cone and the rod portion of dark adaptation had normal kinetics. Recovery of retinal sensitivity was achieved within 30 minutes (Fig 2).

Retinal sensitivity and dark adaptation characteristics appear not to be affected in areas in which peau d'orange was detected by ophthalmoscopy. Abnormal dark adaptation in

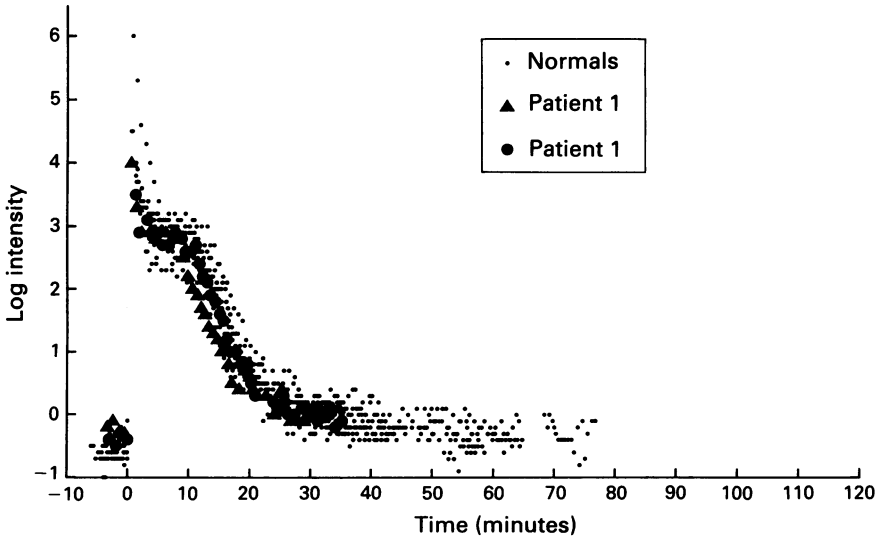

Figure 2 Dark adaptation curve from two locations in patient 1 over areas with peau d'orange compared with five controls.