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]

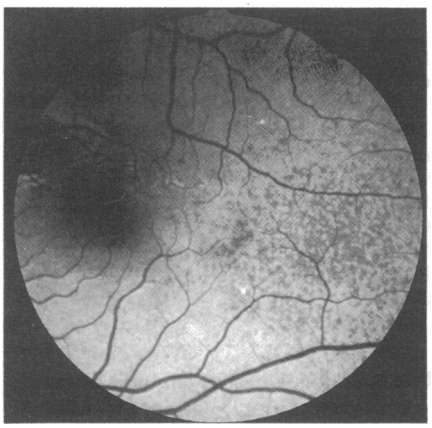

Figure 1 Typical ophthalmoscopic appearance of peau d'orange in one of the patients studied with pseudoxanthoma elasticum.

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

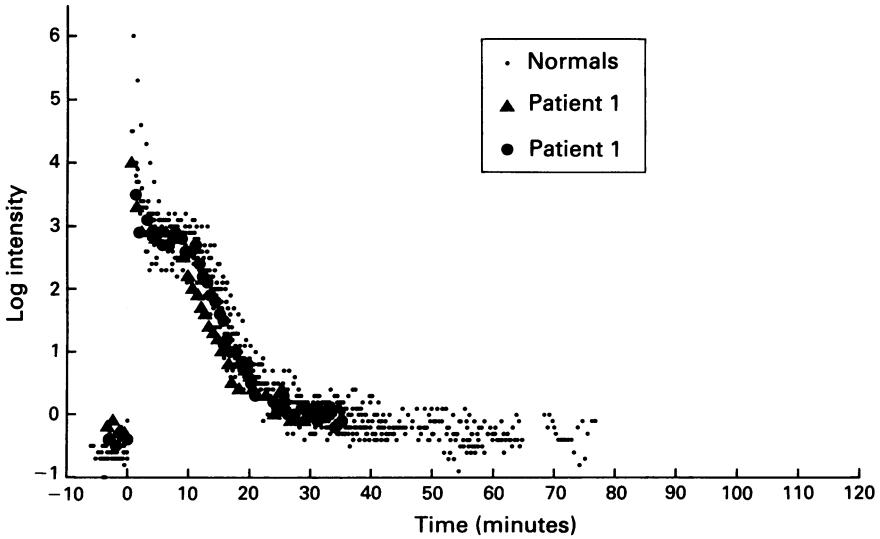

Figure 2 Dark adaptation curve from two locations in patient 1 over areas with peau d'orange compared with five controls.

patients with Sorsby's fundus dystrophy and age-related macular degeneration was thought to be caused by thickening of Bruch's membrane interfering with metabolic exchange across Bruch's membrane between the choriocapillaris and the retinal pigment epithelium. The results may indicate that Bruch's membrane changes associated with peau d'orange in patients with PXE have little effect on function and seem not to interfere significantly with delivery of the metabolic substrates necessary for normal function to the photoreceptor cells.

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1 Pope FM. Historical evidence for the genetic heterogeneity of pseudoxanthoma elasticum. *Br J Dermatol* 1975; 92: 493–509.
2 Shields JA, Federman JL, Tomer TL, Annesley WH. Angioid streaks. I Ophthalmoscopic variations and diagnostic problems. *Br J Ophthalmol* 1975; 59: 257–66.
3 Smith JL, Gass JDM, Justice J. Fluorescein fundus photography of angioid streaks. *Br J Ophthalmol* 1964; 48: 517–21.
4 Bischler V. Le fond muchté multicolor, manifestation fruste de la maladie de Groenblad-Strandberg. *Bull Mem Soc Fr Ophtalmol* 1955; 68: 287–9.
5 Shimuzu K. Mottled fundus in association with pseudoxanthoma elasticum. *Jap J Ophthalmol* 1961; 5: 1–13.
6 Krill AE, Klien BA, Archer DB. Precursors of angioid streaks. *Am J Ophthalmol* 1973; 76: 875–9.
7 Steinmetz RL, Polkinghorne PC, Fitzke FW, Kemp CM, Bird AC. Abnormal dark adaptation and rhodopsin kinetics in Sorsby's fundus dystrophy. *Invest Ophthalmol Vis Sci* 1992; 33: 1633–6.
8 Steinmetz RL, Haimovici R, Jubb C, Fitzke FW, Bird AC. Symptomatic abnormalities of dark adaptation in patients with age-related Bruch's membrane change. *Br J Ophthalmol* 1993; 77: 549–54.

NOTICES

Medical Screening: The Way Forward

Medical screening provides many opportunities for the prevention of disease and handicap. What can it offer and what are its limitations? Based on several case studies, a one day conference entitled Medical Screening: The Way Forward, organised jointly by *BMJ* and *Journal of Medical Screening*, will be held on 26 January 1994 at the QE2 Conference Centre, London to examine the medical, scientific, ethical, social, psychological, and economic aspects of screening. For details: Pru Walters, BMA Conference Unit, BMA House, Tavistock Square, London WC1H 9JR. (Tel: 071–383 6605; Fax: 071–383 6400.)

Optics '94

Optics '94, an international exhibition on eye wear, technology, and equipment for optometry and opthalmology will be held on 18–20 February 1994 at the World Trade Center, Singapore. A conference on better eye care will be held in conjunction with the exhibition. Further details: Lines Exposition & Management Services Pte Ltd, 318-B King George's Avenue, Singapore 0820. (Tel: (65) 2998611; Fax: (65) 2998633.)

International Society of Ocular Trauma

The 3rd International Symposium on Ocular Trauma will be held in Cancun, Mexico in March 1994. Further details: Secretariat, PO Box 50006, Tel Aviv, 61500, Israel. (Tel: (972 3) 5174571; Fax: (972 3) 5175674.)

Third Annual Scientific Meeting of the Australian Squint Club

The Third Annual Scientific Meeting of the Australian Squint Club will be held in Melbourne, Australia on 4–6 March 1994. Further details: Dr W E Gillies, 82 Collins Street, Melbourne 3000, Australia (tel: 61 3 654 5860; fax: 61 3 650 4404).

International Ophthalmic Excimer Laser Congress

The first annual United Kingdom International Ophthalmic Excimer Laser Congress will be held on 15 and 16 April 1994 at Redworth Hall Hotel and Country Club, County Durham. Details: Ms Judith Ritchey, Sunderland Eye Infirmary, Queen Alexandra Road, Sunderland, UK SR2 9HP. (Fax: 091-569 9275.)

Fourth Breton Workshop on Autoimmunity

The Fourth Breton Workshop on Autoimmunity will be held on 15–16 April 1994 in Brest, France. Further details: Secretariat, Laboratory of Immunology, Brest University Medical School Hospital, BP 824–29 609 Brest cédex, France. (Tel: (33) 98 22 33 84; Fax: (33) 98 80 10 76.)

European Society of Traditional Ophthalmology and Traditional Chinese Medicine

The 3rd international symposium of traditional medicine will be held on 12–22 May 1994 in Japan. Further details: Dr J Poletti, Société Européenne d'Ophtalmologie Traditionelle, CHIC Tarbes, BP 1330, 65013 Tarbes Cedex, France. (Tel: 62 51 54 55; Fax: 62 51 51 62.)

American Academy of Optometry

A meeting of the American Academy of Optometry will be held on 28–30 May 1994, at the Amsterdam Marriott Hotel, Amsterdam, The Netherlands. Further details: Academy Office, 4330 East-West Highway, Suite 1117, Bethesda, MD 20814, USA. (Tel: (301) 718-6500; Fax: (301) 656-0989.)

International Conference on Biomedical Periodicals

The International Conference on Biomedical Periodicals will be held on 16–18 June 1994 in Beijing, China. Further details: Dr Yongmao Jiang, International Conference on Biomedical Periodicals, c/o Publishing House of Medical Journals, Chinese Medical Association, 42 Dongsi Xidajie, Beijing 100710, China. (Tel: 86–1–5133311 ext 362; Fax: 86–1–5123754.)

XXVIIth International Congress of Ophthalmology

The International Council of Ophthalmology will hold its XXVIIth Congress in Toronto, Canada on 26–30 June 1994. Further details: Secretariat, 275 Bay Street, Ottawa, Ontario, Canada K1R 5Z5. (Tel: (613) 563–1994; Fax: (613) 236–2727.)

Allied Health Personnel – International Congress of Ophthalmology '94

The first Allied Health Personnel Conference will be held in conjunction with the XXVIIth International Congress of Ophthalmology on 26–30 June 1994 in Toronto, Canada. Further details: Congress Canada, 191 Niagara Street, Toronto, Canada M5V 1C9. (Tel: (416) 860–1772; Fax: (416) 860–0380.)

Welsh Cataract Congress 1994

The Welsh Cataract Congress 1994 will be held on 8–10 September 1994. Details from: Eula Mae Childs, coordinator, Cullen Eye Institute, Baylor College of Medicine, 6501 Fannin, NC200, Houston, TX 77030, USA. (Tel: (713) 798–5941; Fax: (713) 798–4364.)

Third International Symposium on Ocular Inflammation

The 3rd international symposium on ocular inflammation will be held on 22–25 October 1994 in Fukuoka, Japan. Further details: Registration Secretary, c/o JTB Communications Inc, New Kyoto Center Building, 5F, Shiokoji, Shinmachi, Shimogyo-ku Kyoto 600, Japan.

Correction

We regret that, in the perspective in the August issue (1993; 77: 515–24), reference 134 was incorrect. The correct reference is:

134 Yang JL, Neufeld AH, Zorn MB, Hernandez MR. Collagen type I mRNA levels in cultured human lamina cribrosa cells; effects of elevated hydrostatic pressure. *Exp Eye Res* 1993; 56: 567-74.