Correspondence

Adult vitelliform macular degeneration

SIR, We believe there is confusion concerning the definition of 'adult vitelliform macular degeneration.'

Gass described important features of this condition in his original report²: 'symptoms usually begin during the fourth or fifth decade of life, are mild, and progress of visual loss is slow.' There is a characteristic ophthalmoscopic appearance which bears a resemblance to typical vitelliform degeneration, is autosomal dominant, and the EOG may be reduced in the presence of good visual function. Gass originally described this lesion as 'peculiar foveomacular dystrophy' and, because of its resemblance to Best's disease, subsequently labelled the condition 'foveomacular vitelliform dystrophy: adult type.'³ However, the visual loss occurs later in life and the lesions are smaller than seen in typical Best's disease. 'Pseudovitelliform macular degeneration,'⁴ 'adult-onset foveomacular pigment epithelial dystrophy,'⁵ and 'adult vitelliform macular degeneration'¹ have been applied to disorders simulating that originally described by Gass.

The subsequent lack of agreement as to terminology is illustrated well by a recent article by Epstein and Rabb⁶ in which cases labelled 'adult vitelliform macular degeneration' were published and yet autosomal dominant inheritance was proved in none. Only one published case had an

Fig. 2 Macula, right eye, of patient IIIb showing hypopigmented ring with central pigmentation.

affected relative and none had an abnormal light-induced rise in ocular potential bilaterally. Only 2 were under the age of 60 and most appeared to have a lesion falling within the spectrum of senile macular degeneration.

While we accept that the lesions may resemble ophthalmoscopically those seen in Best's disease, the authors have used a term originally proposed for a well defined nosological entity which is illustrated by the following family and which confirms the accuracy of Gass's original description (Table 1).

The pattern of transmission appears to be autosomal dominant with 3 consecutive generations affected (Fig. 1). The ophthalmoscopic appearance of the macula is characteristic (Fig. 2); EOGs are normal or slightly reduced even in the presence of good visual function (Table 1); and ophthalmoscopic abnormalities can be identified as early as 20 years. This last patient has normal visual acuity, colour

Table 1 Affected family members

Patient	Age/sex	VRE	VLE	EOG (RE, LE)	Ophthalmoscopy
IIb	74 M	6/6	6/18	Not performed	Multifocal hypopigmented areas, some with clumped hypopigmentation. Drusen present
IIIa	47 M	6/6	6/6	161, 185	Typical ring lesion. No drusen
IIIb	47 M	6/24	6/36	155, 155	Typical ring lesion. No drusen
IIIc	44 F	6/6	6/9	165, 165	Typical ring lesion. No drusen
IVc	20 M	6/24*	6/6	200, 200	Hypopigmented incomplete ring No hyperpigmentation No drusen

*Amblyopia.

Fig. 1 Pedigree.

vision, EOG light rise, and fluorescein angiography, but a definite small lesion can be seen bilaterally at the level of the pigment epithelium with the appearance of an incomplete, hypopigmented ring around the fovea.

The disorder in this family is similar to Best's vitelliform dystrophy in that it is autosomal dominant, there is a resemblance between the 2 conditions ophthalmoscopically, and the EOG can be reduced in the presence of good visual function, but the 2 conditions differ in their severity, as shown by age of onset, extent of visual loss, size of lesion, and consistency of EOG abnormality. There is no doubt that the term 'adult vitelliform dystrophy' may lead to confusion, but it has now been widely used in ophthalmic literature as a condition quite distinct from Best's disease. We would make a plea for this term to be restricted to the specific genetically determined dystrophy described by Gass in his original article.²

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References

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- 2 Gass JDM. A clinicopathologic study of a peculiar foveomacular dystrophy. *Trans Am Ophthalmol Soc* 1974; **72**: 139-56.
- 3 Gass JDM. *Stereoscopic Atlas of Macular Diseases*. 2nd ed. St Louis: Mosby, 1977.
- 4 Fishman GA, Trimble S, Rabb MF, Fishman M. Pseudovitelliform macular degeneration. *Arch Ophthalmol* 1977; **95**: 73-6.
- 5 Vine AK, Schatz H. Adult-onset foveomacular pigment epithelial dystrophy. *Am J Ophthalmol* 1980; **89**: 680-91.
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Notes

Eye epidemiology

The National Eye Institute will hold a symposium on the 'Epidemiology of Eye Diseases and Visual Disorders' on 10-11 June 1982. Details from Barbara DiSimone, Office of the Director, National Eye Institute, Room 6A-03, Building 31, National Institutes of Health, Bethesda, Maryland, 20205, USA. Investigators wishing to present papers should send abstracts of not more than 200 words by 5 February 1982 to Fred Ederer, Chief, Office of Biometry and Epidemiology, National Eye Institute, Room 6A-10 (address as above).

International Corneal Society

A meeting will be held on 27-28 October 1982 at Las Vegas, Nevada, USA. Topics to be discussed include corneal transplantation, corneal immunology, and inflammation. Details from Dr Stuart I. Brown, Eye and Ear Hospital, 230 Lothrop Street, Pittsburgh, PA 15213, USA.

Book reviews

Documenta Ophthalmologica Proceedings Series 27. Visual Pathways Electrophysiology and Pathology. Eds. H. SPEKREIJSE and P. A. APKARIAN. Pp. 453. Dfl. 195.00. W. Junk: The Hague, Netherlands. 1981.

This book is a collection of papers given at the 1980 meeting of the International Society for Clinical Electrophysiology of Vision. The book is arranged in 6 parts, all dealing with different aspects of the VEP. There are a number of papers on the VEP and binocularity as well as another section on VEP changes in relation to field loss in glaucoma. The main value of this volume is that it provides a useful reference source to the specialist. However, the nonspecialist may be surprised to read about some of the recent advances in this field, many of which have possible clinical applications.

N. R. GALLOWAY

Lecture Notes on Ophthalmology. 6th edn. By PATRICK D. TREVOR-ROPER. Pp. 128. £4.25. Blackwell Scientific: Oxford. 1980.

The author is to be congratulated on another excellent edition of his lecture notes written in his own inimitable style. The chapters are divided in a practical clinical manner to cover the various aspects of ophthalmology with which every doctor will be involved at some time in his career. External eye diseases, injuries, red eye, sudden and gradual loss of sight, squint, field loss, and tropical conditions are all covered. There is a useful chapter of questions to give feed back on assimilation of knowledge, as well as an introduction to multiple choice technique. As with any publication there are some printing errors, but most of these are obvious and not beyond the wit of the average student. One could take issue with the emphasis of a diagram of trephination for glaucoma surgery rather than trabeculectomy and the detailed illustrations of keratoplasty in a book of this size.

This book is ideal to introduce the subject in parallel with slide lectures as well as for revision prior to examination. It will continue as a favourite with medical students and can be recommended by clinical teachers.

G. V. CATFORD

Vitreous Surgery. By RONALD G. MICHELS. Pp. 462. £51.50. C. V. Mosby: London. 1981.

In the past 10 years microsurgical vitrectomy has become a firmly established technique for management of many previously inoperable intraocular disorders, making exciting technical and intellectual demands on those committed to its practice. Dr Ronald Michels, of the Wilmer Eye Institute, is well known for his uniquely eloquent presentations and numerous publications arising from his work in this field. In this the latest in a line of recent books on vitrectomy he presents arguably the most comprehensive and comprehensible work to have appeared to date. Chapters on surgical pathology, clinical assessment, instrumentation, techniques, complications, results, and indications are written in a lucid, if sometimes verbose and repetitive, style, and the book is very generously illustrated, including a host of fine drawings by Timothy C. Hengst and Garry P. Lees.