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 report: '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.'

However, the visual loss occurs later in life and the lesions are smaller than seen in typical Best's disease. 'Pseudovitelliform macular degeneration,' and 'adult-onset foveomacular pigment epithelial dystrophy,' and 'adult vitelliform macular degeneration' 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 Rabb 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 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 the 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

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Table 1  Affected family members

<table>
<thead>
<tr>
<th>Patient</th>
<th>Age/sex</th>
<th>VRE</th>
<th>VLE</th>
<th>EOG (RE, LE)</th>
<th>Ophthalmoscopy</th>
</tr>
</thead>
<tbody>
<tr>
<td>IIb</td>
<td>74 M</td>
<td>6/6</td>
<td>6/18</td>
<td></td>
<td>Multifocal hypopigmented areas, some with clumped hypopigmentation. Drusen present</td>
</tr>
<tr>
<td>IIIa</td>
<td>47 M</td>
<td>6/6</td>
<td>6/6</td>
<td>161, 185</td>
<td>Typical ring lesion. No drusen</td>
</tr>
<tr>
<td>IIIb</td>
<td>47 M</td>
<td>6/6</td>
<td>6/36</td>
<td>155, 155</td>
<td>Typical ring lesion. No drusen</td>
</tr>
<tr>
<td>IIIc</td>
<td>44 F</td>
<td>6/6</td>
<td>6/9</td>
<td>165, 165</td>
<td>Typical ring lesion. No drusen</td>
</tr>
<tr>
<td>IVc</td>
<td>20 M</td>
<td>6/24*</td>
<td>6/6</td>
<td>200, 200</td>
<td>Hypopigmented incomplete ring</td>
</tr>
</tbody>
</table>

*Amblyopia.

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Fig. 1  Pedigree.

Fig. 2  Macula, right eye, of patient IIIb showing hypopigmented ring with central pigmentation.
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,
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
cfusion, 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
1 Epstein GA, Rabb MF. Adult vitelliform macular degeneration:
2 Gass JDM. A clinicopathologic study of a peculiar foveomacular
3 Gass JDM. Stereoscopic Atlas of Macular Diseases. 2nd ed. St.
4 Fishman GA, Trimmel S, Rabb MF, Fishman M. Pseudovitelliform
5 Vine AK, Schatz H. Adult-onset foveomacular pigment epithelial
6 Epstein GE, Rabb MF. Adult vitelliform macular degeneration:

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 trans-
plantation, 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

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. Gallows

Lecture Notes on Ophthalmology. 6th edn. By Patrick
D. Trevor-Roper. Pp. 128. £4.25. Blackwell Scientific:

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 introduc-
tion 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


In the past 10 years microsurgical vitreectomy has become a
firmly established technique for management of many
previously inoperable intraocular disorders, making excit-
ing 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 presenta-
tions 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, instru-
mentation, techniques, complications, results, and indica-
tions 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.
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L H Bloom, D E Swanson and A C Bird

doi: 10.1136/bjo.65.11.800