Choroidal folds and papilloedema

Lorraine M Cassidy, Michael D Sanders

Abstract

***Aims*—To assess the clinical and fluorescein angiographic features of choroidal folds seen in association with papilloedema.**

***Methods*—In a retrospective study, the clinical data from a database on patients with choroidal folds (1963–97), including fundus photography and fluorescein angiography, from 32 patients (64 eyes) with choroidal folds in association with papilloedema were reviewed. The clinical and fluorescein angiographic features and the clinical course of choroidal folds in these patients are described.**

***Results*—32 patients had choroidal folds associated with papilloedema. Folds of two distinct categories were observed, either coarse folds or wrinkles. The folds persisted in all cases, even after resolution of papilloedema. Follow up ranged from 1 month to 20 years. Only one patient suffered permanent visual impairment as a result of a choroidal fold.**

***Conclusions*—Choroidal folds exist in two forms, coarse folds and wrinkles. They persist even after papilloedema has resolved. Final visual acuity did not appear to be affected by the presence of choroidal folds in the majority of patients.**

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Nettleship first described choroidal folds in 1884 in a patient with atrophic papilloedema as a result of an intracranial mass.[1] The folds consist of linear grooves in the posterior pole of the globe, and may be horizontal, vertical, or oblique, and rarely ever extend beyond the equator.[2 3] A characteristic fluorescein angiographic pattern of alternating fluorescent and hypofluorescent bands has been described in choroidal folds.[4–6] The hyperfluorescent lines correspond to the top of the convex bulge of the fold, and the hypofluorescent lines correspond to the troughs. Histological sections have shown that both Bruch's membrane and the retinal pigment epithelium are involved in a choroidal fold,[5 7 8] and the overlying retina is normal.[3] The fluorescein angiogram is an important tool in distinguishing choroidal folds from retinal folds[2 9] as the latter do not show up on fluorescein angiography.

Choroidal folds can be seen in association with orbital masses, orbital inflammation, dysthyroid eye disease, hypermetropia, and following scleral buckling.[1 2 10–15] Idiopathic choroidal folds have also been described.[16] We describe the clinical and angiographic features of choroidal folds observed in a series of patients with papilloedema.

Department of Neuro-ophthalmology, National Hospital for Neurology and Neurosurgery, London WC1N 3BG
L M Cassidy
M D Sanders

Correspondence to:
Miss Lorraine Cassidy, Department of Ophthalmology, Great Ormond Street Hospital for Children, Great Ormond Street, London WC1N 3JH

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Methods

The clinical details of all patients who attended the ophthalmic photographic department between January 1963 and December 1997 with choroidal folds were retrospectively examined. All patients had either fundus photography and fluorescein angiography. The notes, photographs, and fluorescein angiograms of all patients were retrieved for analysis. The age at presentation, sex, presenting symptoms, visual acuity at presentation and at the final follow up visit, diagnosis, and treatment were recorded. From the fundus photographs and fluorescein angiograms the papilloedema was graded as being early, acute, or chronic. Choroidal folds were described as being peripapillary, perimacular, or macular depending on their location. If the folds involved all three of these regions they were described as posterior polar choroidal folds. Choroidal folds were also categorised according to their appearance as being either coarse folds or wrinkles (fine folds).

Results

Of 52 patients recorded as having choroidal folds, 32 had folds as a result of papilloedema. In 28 cases (56 eyes) the folds were bilateral (Table 1). The mean age at presentation was 45 years (range 21–65 years). The male to female ratio was 1.2:1.

Follow up ranged from 1 month to 20 years.

The aetiology of papilloedema in these patients (Table 2) included benign intracranial hypertension (20 cases), intracranial tumour (seven cases), dural arteriovenous malformation (one case), cerebellar ectopia with secondary hydrocephalus (one case), aqueduct stenosis (one case), and an intracerebral haematoma (two cases). The papilloedema was early in one case, acute in 14, chronic in 17 cases.

The final visual acuity ranged from 6/4 to perception of light (PL) (Table 3). In those eyes with a final best corrected visual acuity of 6/12 or less (n=10), optic atrophy was the cause in nine eyes, and a choroidal fold was

Table 1 Laterality of choroidal folds and papilloedema

Choroidal folds	Unilateral	Bilateral
No of patients	4	28
Papilloedema	Unilateral	Bilateral
No of patients	3	29

Table 2 Aetiology of papilloedema

Aetiology of papilloedema	*No of patients*
Benign intracranial hypertension	20
Intracranial tumour	7
Dural arteriovenous malformation	1
Cerebellar ectopia and hydrocephalus	1
Aqueduct stenosis	1
Intracerebral haematoma	2

Table 3 Clinical details

Patient/sex	*Age (years)*	*Papilloedema*	*Diagnosis*	*Choroidal folds*	*Visual acuity*		*Follow up*
					Right	*Left*	
1/F	55	Bilateral Chronic	BIH	Bilateral peripapillary and perimacular wrinkles not seen clinically	6/9 Bilateral optic atrophy	6/9	2 months
2/F	47	Bilateral Acute	BIH	Bilateral Posterior polar Coarse	6/5	6/5	1 year
3/F	44	Bilateral Acute	BIH	Bilateral Macular coarse Perimacular wrinkles	6/4	6/4	3 years
4/M	21	Bilateral Acute	ICT	Bilateral Macular and perimacular Coarse	6/6	6/6	7 years
5/M	57	Bilateral Chronic	BIH	Bilateral Posterior polar coarse Perimacular wrinkles	6/9 Bilateral optic atrophy	6/9	2 months
6/M	44	Bilateral Acute	ICT	Unilateral, right superior peripapillary and perimacular wrinkles	6/6	6/6	1 year
7/M	38	Bilateral Chronic	BIH	Bilateral Peripapillary and perimacular wrinkles	6/6	6/6	6 months
8/F	44	Bilateral Acute	BIH	Unilateral, left Coarse posterior polar Macular wrinkles	6/5 Hyperpigmented fold through fovea	2/60	6 months
9/M	32	Bilateral Chronic	Aqueduct stenosis	Unilateral, right Inferior peripapillary and perimacular wrinkles	6/18 Right optic atrophy	6/6	4 years
10/M	52	Bilateral Acute	Parasagittal meningioma	Bilateral Posterior polar wrinkles	6/4	6/5	1 month
11/M	43	Bilateral Acute	BIH	Bilateral Posterior polar right, coarse at macula, rest wrinkles Macular wrinkles, left	6/9	6/6	20 years
12/F	55	Bilateral Chronic	BIH	Bilateral Macular coarse	6/6	6/9	1 month
13/M	49	Bilateral Chronic	BIH	Bilateral Coarse Perimacular and macular	6/6 Left optic atrophy	6/36	5 years
14/F	52	Bilateral Acute	ICT	Bilateral Posterior polar Coarse macular Rest, wrinkles	PL Right optic atrophy	6/5	6 months
15/F	47	Bilateral Chronic	BIH	Bilateral Posterior polar Coarse	6/6	6/6	2 months
16/M	50	Bilateral Acute	Intracerebral haematoma	Bilateral Posterior polar Coarse	6/6	6/6	1 year
17/M	51	Bilateral Acute	BIH	Bilateral Macular coarse	6/4	6/4	2 years
18/M	40	Bilateral Chronic	Intraventricular glioma	Bilateral Posterior polar Coarse at maculae Rest, wrinkles	6/9 Bilateral optic atrophy	6/9	9 months
19/F	55	Bilateral Chronic	BIH	Bilateral Posterior polar Coarse	6/12 Bilateral optic atrophy	6/24	3 months
20/F	47	Bilateral Chronic	BIH	Bilateral Posterior polar Coarse	6/18 Bilateral optic atrophy	PL	6 years
21/F	42	Unilateral Acute	BIH	Bilateral Posterior polar coarse, left Wrinkles above disc, right	6/6	6/6	4 months
22/F	47	Bilateral Early	Pituitary adenoma	Bilateral Posterior polar Coarse Not seen clinically	6/6	6/6	11 years
23/F	47	Bilateral Chronic	Tentorial meningioma	Bilateral Coarse macular	6/6	6/6	2 years
24/M	29	Bilateral Chronic	BIH	Bilateral Coarse macular	6/5	6/5	1.5 years
25/M	38	Bilateral Chronic	Cerebellar ectopia	Bilateral Posterior polar Coarse	6/9	6/9	15 months
26/F	65	Bilateral Chronic	BIH	Unilateral, left Coarse macular Not seen clinically	6/6 Left optic atrophy	6/18	1 month
27/M	56	Bilateral Chronic	Dural AVM	Bilateral Macular and perimacular Wrinkles	6/12 Right optic atrophy	6/9	4 years
28/F	49	Bilateral Chronic	BIH	Bilateral Right inferior peripapillary and perimacular wrinkles Left coarse peripapillary and perimacular	6/5	6/5	6 years
29/M	55	Bilateral Acute	BIH	Bilateral Perimacular wrinkles Not seen clinically	6/6	6/6	9 months
30/M	27	Bilateral Acute	Cerebral haemorrhage	Bilateral Right posterior polar and left macular coarse	6/6	6/9	3 months
31/M	32	Unilateral left Acute	BIH	Bilateral posterior polar Coarse macular and peripapillary wrinkles	6/5	6/5	7 years
32/M	56	Unilateral left Chronic	BIH	Bilateral posterior polar Coarse	6/5	6/5	3 years

BIH = benign intracranial hypertension; ICT = intracranial tumour; AVM = arteriovenous malformation; PL = perception of light.

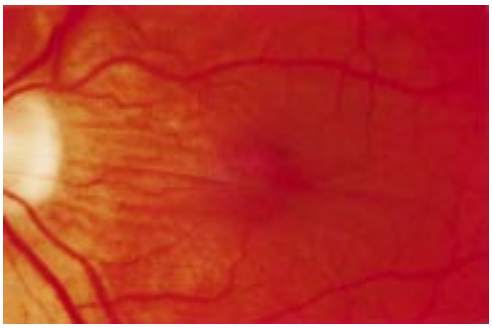

Figure 1 Visual loss due to macular folds. Colour photograph of the left fundus of a patient (patient no 8, Table 3) who developed papilloedema and choroidal folds as a result of benign intracranial hypertension. A heavily pigmented choroidal fold, which can be seen passing through the fovea, is responsible for permanent visual impairment. Visual acuity in the left eye is 2/60, and the patient describes severe left metamorphopsia.

responsible for poor vision in one eye. In this patient the heavily pigmented trough of a fold crossed through the fovea (Fig 1), and she complained of metamorphopsia.

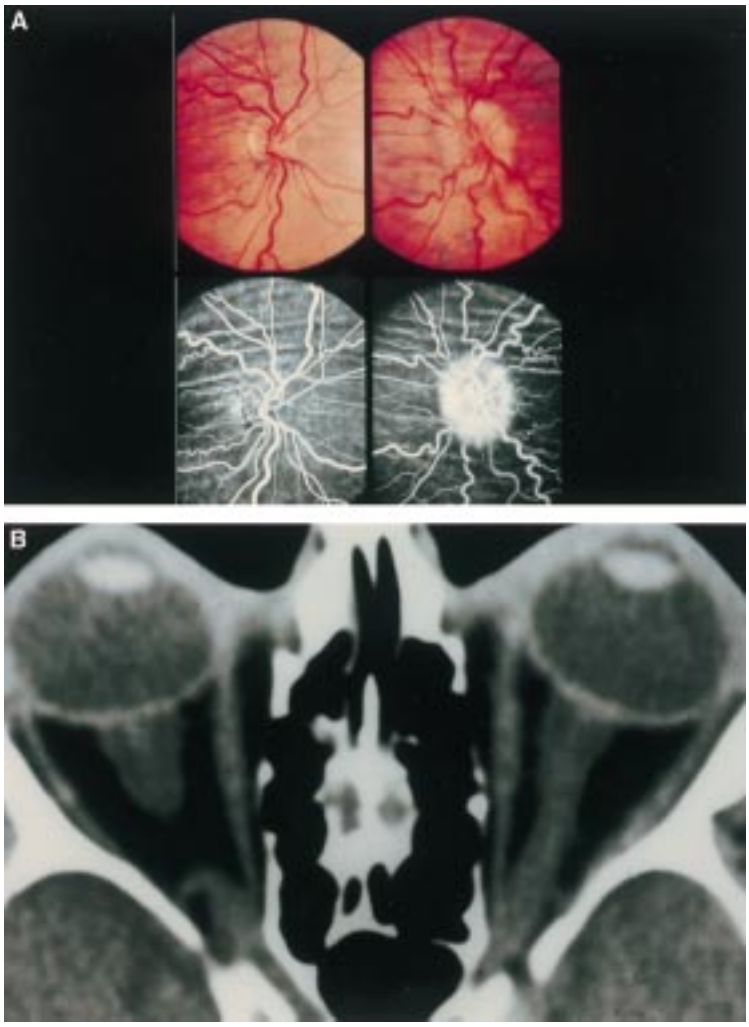

Figure 2 (A) Bilateral choroidal folds and unilateral papilloedema. Colour and fluorescein photographs demonstrating bilateral coarse choroidal folds and unilateral papilloedema in a patient (patient no 32, Table 3) with benign intracranial hypertension. (B) Distended nerve sheath and compressed globes. Computed tomograph scan of the same patient demonstrating dilated optic nerve sheaths. This patient presented with acquired hypermetropia. Examination revealed bilateral choroidal folds and a swollen left disc. The arrow denotes perineural cerebrospinal fluid.

All patients had fundus photography and fluorescein angiography. Choroidal folds were bilateral in 28 cases and all of these patients except three (Fig 2) had bilateral papilloedema. One of the patients, who had unilateral papilloedema and bilateral choroidal folds, had more choroidal folds on the side of the swollen disc. All four patients with unilateral choroidal folds had bilateral papilloedema. Choroidal folds as documented by fluorescein angiography were not observed clinically or on fundus photography in four cases (seven eyes) (Fig 3).

Two types of choroidal folds were observed on fluorescein angiography—coarse folds, which consist of wide bands of alternating hyperfluorescence and hypofluorescence; and wrinkles, which manifest as fine bands of hyperfluorescence and hypofluorescence. These two distinct variations of choroidal folds may coexist in the same eye (Fig 4A), and are easily distinguishable on fluorescein angiography. Folds were always horizontal or oblique in this group of patients and no vertical folds were observed. In the majority of eyes (67.2%) the choroidal folds formed a characteristic peripapillary/perimacular or posterior polar pattern. This pattern of choroidal folding, where the folds curve around the nasal aspect of the disc and then sweep superotemporally and inferotemporally, is the most common pattern of choroidal folding seen in association with papilloedema (Fig 4A). The folds were seen at the macula only in 17.2%, in the macular and perimacular region in 12.5%, at the perimacular area alone in 1.56%, and in the peripapillary area alone in 1.56% (Table 4).

Choroidal folds persisted in all cases, even after resolution of papilloedema (Fig 5).

Discussion

Choroidal folds in association with papilloedema have been reported as case reports and in a few small series in the literature.[1 14 17–20] We report the largest series to date of choroidal folds resulting from papilloedema.

Papilloedema results from the transmission of elevated intracranial pressure to the perioptic subarachnoid space[21] resulting in elevated pressure in the optic nerve sheath. The elevated sheath pressure holds up axoplasmic flow in the optic nerve head, which results in papilloedema due to axonal swelling.[22] Should the optic nerve sheath become distended it may in turn press on the globe causing distortion and hence choroidal folds.[14 20] This flattening of the globe is often associated with an acquired hypermetropia.[2 5 13 20 23 24] In some cases the choroidal folds precede the development of papilloedema. This suggests that the elevated intra-sheath pressure produces indentation of the globe before there is any reduction in retrolaminar perfusion. Hence it is possible, though rare, to see unilateral papilloedema with bilateral choroidal folds (Fig 2A), or choroidal folds alone as a sign of raised intracranial pressure.[20] However, in our experience, there are cases where severe papilloedema in conjunction with a grossly dilated nerve sheath

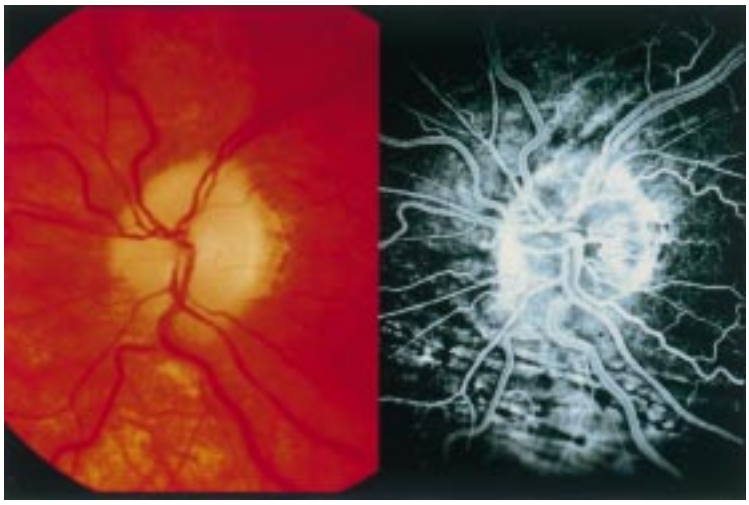

Figure 3 Subclinical choroidal folds. Colour fundus photography showing no evidence of choroidal folds in a patient with benign intracranial hypertension. Fluorescein angiogram of the same patient demonstrating the presence of "subclinical" choroidal folds.

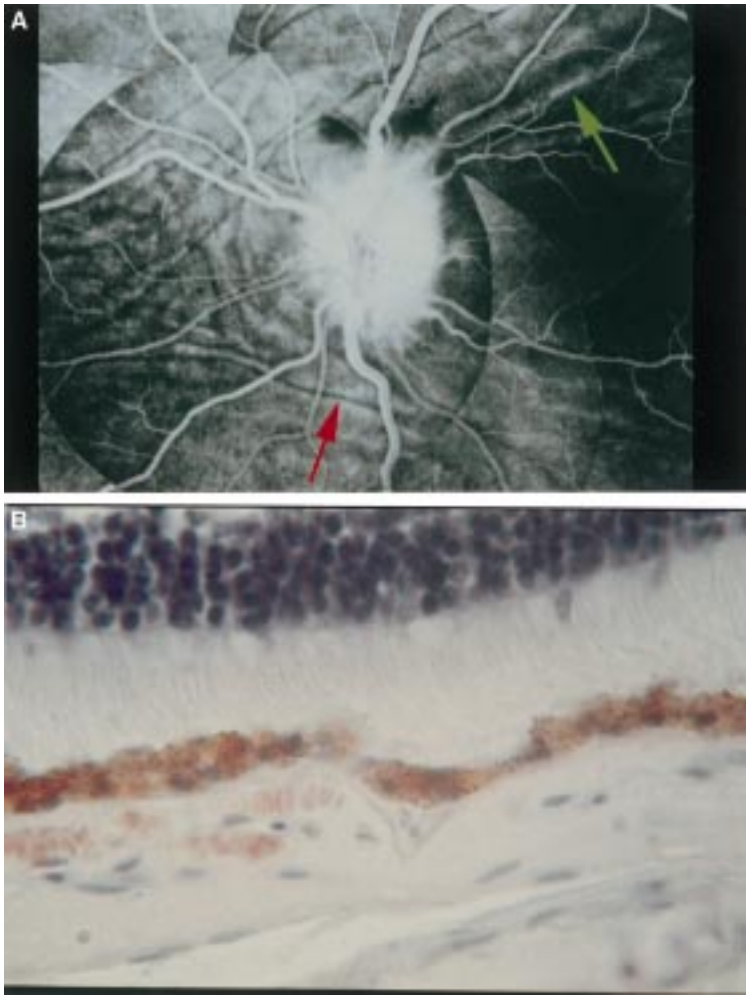

Figure 4 (A) Classic appearance of choroidal folds and papilloedema. This pattern of choroidal folding, where the folds curve around the nasal aspect of the disc, and then sweep superotemporally and inferotemporally, is the most common pattern of choroidal folding seen in association with papilloedema. Note the presence of both choroidal wrinkles (red arrow) and coarse choroidal folds (green arrow). (B) Histological section showing a wrinkle (arrow) involving only the retinal pigment epithelium and Bruch's membrane. The overlying retina is flat.

is not associated with even subtle choroidal folding. Why some patients with distended nerve sheaths develop choroidal folds and others do not is not clear. It may be possible that variation of elastic properties of the sclera/scleral rigidity in different individuals could have some influence on how easily pressure can be transmitted through to the choroid. Variations of insertion of the sheath may also play a role in determining the formation of choroidal folds. While these matters remain unresolved, it is most important for the clinician to be aware that choroidal folds may in themselves be a sign of elevated intracranial pressure.

Table 4 Pattern of choroidal folds

Pattern of choroidal folds	*No of eyes (%)*
Posterior polar folds	43 (67.2)
Macular folds	11 (17.2)
Macular and perimacular folds	8 (12.5)
Perimacular folds only	1 (1.55)
Peripapillary folds only	1 (1.55)

The two categories of choroidal folds we observed may be explained as follows. Choroidal wrinkles, which are fine lines of hypofluorescence with hyperfluorescent borders, appear to be folds which are confined to the retinal pigment epithelium and Bruch's membrane (Fig 4B). Coarse folds, which are broader bands of alternating hyperfluorescence and hypofluorescence. This may result from folding of the full thickness of the choroid. The wrinkles are most often seen in the peripapillary area and coarse folds at the macula and perimacularly. However the reverse situation may occur. The mechanical factors which determine the category of choroidal fold are not known.

It is interesting to note that even after resolution of papilloedema, the choroidal folds persisted in all patients in this series. The final visual acuities in the majority of these patients did not appear to be affected by the persistence of folds, even though most patients with choroidal folds develop an acquired hypermetropia at the time of presentation. This acquired hyperopia is well documented[2 5 13 20 23 24] and is a result of flattening of the globe by a distended optic nerve sheath. It is possible that in the initial stages the globe is flattened together with distortion and folding of the choroid, and that later on when intracranial pressure is reduced and the papilloedema resolves, the mass effect of the distended nerve sheath on the globe is removed, allowing the refractive state of the eye to return to normal, but with the persistence of the choroidal folds. Unfortunately, as this was a retrospective study, it was not possible to include refraction or axial lengths in the clinical details. This is disappointing, as these variables may be associated with scleral rigidity, and therefore such information could shed more light on the possible mechanism of choroidal folding.

Conclusions

Choroidal folds exist in two forms, coarse folds and wrinkles, and can persist after the resolution of papilloedema, but only rarely do they have any long term effect on visual acuity. Choroidal folds can be a sign of raised intracranial pressure in the absence of papilloedema.

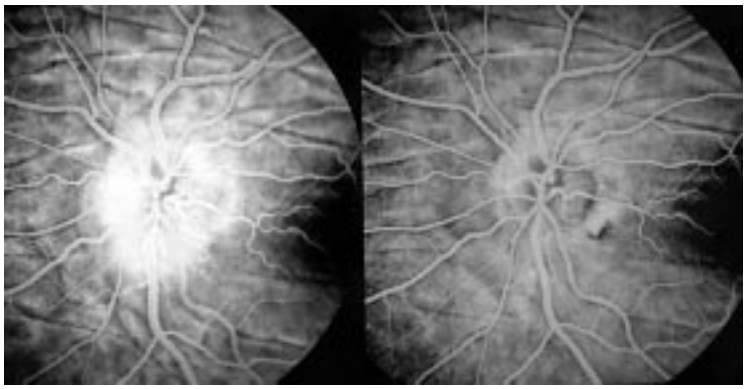

Figure 5 Persistence of choroidal folds. Fluorescein photographs showing the persistence of choroidal folds 7 years after resolution of papilloedema. This patient had a left optic nerve sheath fenestration for benign intracranial hypertension. Left: choroidal folds with papilloedema; right: 7 years after resolution.

1 Nettleship E. Peculiar lines in the choroid in a case of post papillitic atrophy. *Trans Ophthalmol Soc UK* 1884;**4**:167–8.
2 Newell FW. Choroidal folds. *Am J Ophthalmol* 1973;**75**: 930–42.
3 Bullock JD, Egbert PR. Experimental choroidal folds. *Am J Ophthalmol* 1974;**78**:618–23.
4 Amalric P. Un nouveau type de plis retiniens congenitaux. *Arch Ophthalmol Rev Gen Ophthalmol* 1968;**28**:507–12.
5 Norton EWD. A characteristic fluorescein angiographic pattern in choroidal folds. *Proc R Soc Med* 1969;**62**:119–28.
6 Francois J, DeLaey JJ. *Fluoro-angiographic aspects of of acquired chorioretinal folds, Photography in ophthalmology.* International symposium on fluorescein angiography. Basle: Karger, 1970:129–30.
7 Birch-Hirschfeld A, Siegfried C. Zur Kenntnis der veraenderungen des bulbus durch druck eines orbitaltumors. *Arch Ophthalmol* 1915;**90**:404–7.
8 Wolter JR. Parallel horizontal retinal folding. *Am J Ophththalmol* 1962;**53**:26–9.
9 Wise GN. Clinical features of idiopathic preretinal macular fibrosis. *Am J Ophthalmol* 1975;**79**:349–57.
10 Hyvarinen L, Walsh FB. Benign chorioretinal folds. *Am J Ophthalmol* 1970;**70**:14–17.
11 Schepens CL, Schwartz A. Intraocular tumours. *Arch Ophthalmol* 1958;**60**:72–83.
12 Gass JDM. Hypotony maculopathy. In: Bellows JC, ed. *Contemporary ophthalmology.* Baltimore: Williams and Wilkins, 1972:343.
13 Cangemi FE, Trempe CL, Walsh JB. Choroidal folds. *Am J Ophthalmol* 1978;**86**:380–7.
14 Bird AC, Sanders MD. Choroidal folds in association with papilloedema. *Br J Ophthalmol* 1973;**57**:89–97.
15 Sanders MD. The Bowman Lecture. Papilloedema: 'the pendulum of progress.' *Eye* 1997;**11**:267–94.
16 Cappaert WE, Purnell EW, Frank KE. Use of B-sector scan ultrasound in the diagnosis of benign choroidal folds. *Am J Ophthalmol* 1977;**84**:375–9.
17 Von Winning CHOM. Fluorography of choroidal folds. *Ophthalmologica* 1973;**167**:436–9.
18 Cairns JD. Disc oedema and choroidal folds. *Aust J Ophthalmol* 1973;**1**:30–5.
19 Gittinger JW, Asdourian GK. Macular abnormalities in papilloedema from pseudotumour cerebri. *Ophthalmology* 1989;**96**:192–4.
20 Jacobson DM. Intracranial hypertension and the syndrome of acquired hyperopia with choroidal folds. *J Neuro-Ophthalmol* 1995;**15**:178–85.
21 Liu D, Kahn M. Measurement and relationship of subarachnoid pressure of the optic nerve to intracranial pressures in fresh cadavers. *Am J Ophthalmol* 1993;**116**: 548–56.
22 Hayreh SS. Optic disc oedema in raised intracranial pressure. V. Pathogenesis. *Arch Ophthalmol* 1977;**95**:1553–65.
23 Atta HR, Byrne SF. The findings of standardised echography for choroidal folds. *Arch Ophthalmol* 1988;**106**:1234–41.
24 Leahey AB, Brucker AJ, Wyszynski RE, *et al.* Chorioretinal folds. A comparison of unilateral and bilateral cases. *Arch Ophthalmol* 1993;**111**:357–9.