Journal of Neurology, Neurosurgery, and Psychiatry 1986;**49**:243–250

Benign intracranial hypertension: visual loss and optic nerve sheath fenestration

RSG KNIGHT, AR FIELDER, JL FIRTH

From the Derbyshire Royal Infirmary, Derby, UK

SUMMARY Five patients with serious ocular complications of benign intracranial hypertension are described. Optic nerve sheath fenestration resulted in resolution of papilloedema with, in three instances, improvement in vision. Benign intracranial hypertension may not always be benign for vision and fenestration operations may prevent or reverse visual deterioration by an effect on the optic nerve rather than by reducing intra-cranial pressure.

Benign intracranial hypertension is characterised by raised intracranial pressure in the absence of a space-occupying lesion, with a normal or small ventricular system and normal CSF constituents. The widely accepted name clearly implies a good prognosis and whilst this is generally true,[1 2] severe and permanent visual complications can occur. The reported incidence of such complications varies between 2% and 24%[3–5] and the most important consideration in the management of benign intracranial hypertension is the prevention of disabling visual sequelae. Many medical and surgical treatments have been employed including corticosteroids, chlorthalidone, acetazolamide, multiple lumbar punctures, subtemporal decompression, CSF shunting procedures and optic nerve sheath decompression.[6]

This paper describes five patients who presented over a four year period with severe ocular complications of benign intracranial hypertension and who underwent optic nerve sheath fenestration.

Patients

Case 1

This 35-year-old man presented after several months of headaches and focal sensory fits with a few generalised convulsions. Bilateral mild papilloedema was present with enlarged blind spots but with otherwise normal visual fields and visual acuities of R6/5 L6/5. A dynamic isotope brain scan and CT scan were normal. Lumbar puncture yielded normal CSF with a pressure of 240 mm water. An EEG showed no significant abnormality. In view of the epilepsy, a presumptive diagnosis of intra-cranial venous thrombosis was made but angiography was not performed. Despite treatment with dexamethasone and frusemide, monthly LP pressures varied from 130 to 330 mm water (mean 265), headaches persisted and the papilloedema worsened with the appearance of haemorrhages (fig 1a). Six months after presentation, he developed transient visual obscurations and although visual acuity was unchanged with full peripheral fields, the blind spots were much enlarged, on the right to within one degree of fixation (fig 2a). Two months later, the right visual acuity had fallen to 6/9 and a right optic nerve sheath fenestration was performed. Postoperatively, the left disc oedema remained (fig 1b) and twelve days later a left optic nerve sheath fenestration was performed. The papilloedema took five months to resolve completely (fig 1c) with concomitant improvement in vision. One year after surgery, vision was R6/5 L6/5 and, apart from moderately enlarged blind spots, the only field defect was a small (two degree) paracentral scotoma on the right (fig 2b). After operation, the LP CSF pressures remained high ranging from 220 to 310 mm water. Repeat CT scans have been normal and there have been no further fits on treatment with phenytoin.

Case 2

This 25-year-old obese woman presented with headaches and blurred vision in the right eye. Vision was R6/18 L6/6; there was bilateral papilloedema and visual fields were constricted with enlarged blind spots. Blood pressure was 160/100 mm Hg. Carotid angiography and ventriculography were normal. A single LP CSF pressure of 130 mm of water was recorded. With dexamethasone and frusemide therapy her symptoms and signs settled over three months. Her weight increased although initially her blood pressure settled. Later, sustained hypertension developed and was successfully treated with atenolol and bendrofluazide. Four years later, she returned after three weeks of headaches and blurred vision. Corrected acuities were R6/9 L6/5, both optic discs were swollen, marked retinal arterial attenuation was present and the visual fields were constricted. Blood pressure was 120/80 mm Hg. A CT scan was normal and

Address for reprint requests: Dr RSG Knight, Department of Neurology, Radcliffe Infirmary, Oxford OX2 6HE.

Received 16 October 1984 and in revised form 22 May 1985. Accepted 28 May 1985.

lumbar puncture produced normal CSF at a pressure of over 300 mm water. Despite treatment with dexamethasone, frusemide and intravenous mannitol, the raised CSF pressure, papilloedema and headaches persisted. After twelve days, vision deteriorated to R6/18 L6/12 with further field constriction. Bilateral simultaneous optic nerve sheath fenestration operations were therefore performed. On the following day, her vision was reduced to counting fingers in each eye with, on the right, occlusions of the cilio-retinal and long posterior ciliary arteries. CSF pressure at cisternal puncture was unrecordably low and at lumbar puncture was 20 mm water. Significant conjunctival and lid chemosis rapidly resolved over the ensuing few days. Irregularity of both pupils settled over a period of four months. A post-operative esotropia occurred and a small deviation persists. Disc swelling resolved over two weeks with an increase in retinal arterial calibre and the appearance of retinal venous pulsation; vision then being R6/18 L6/12. Five years later, vision is 6/9

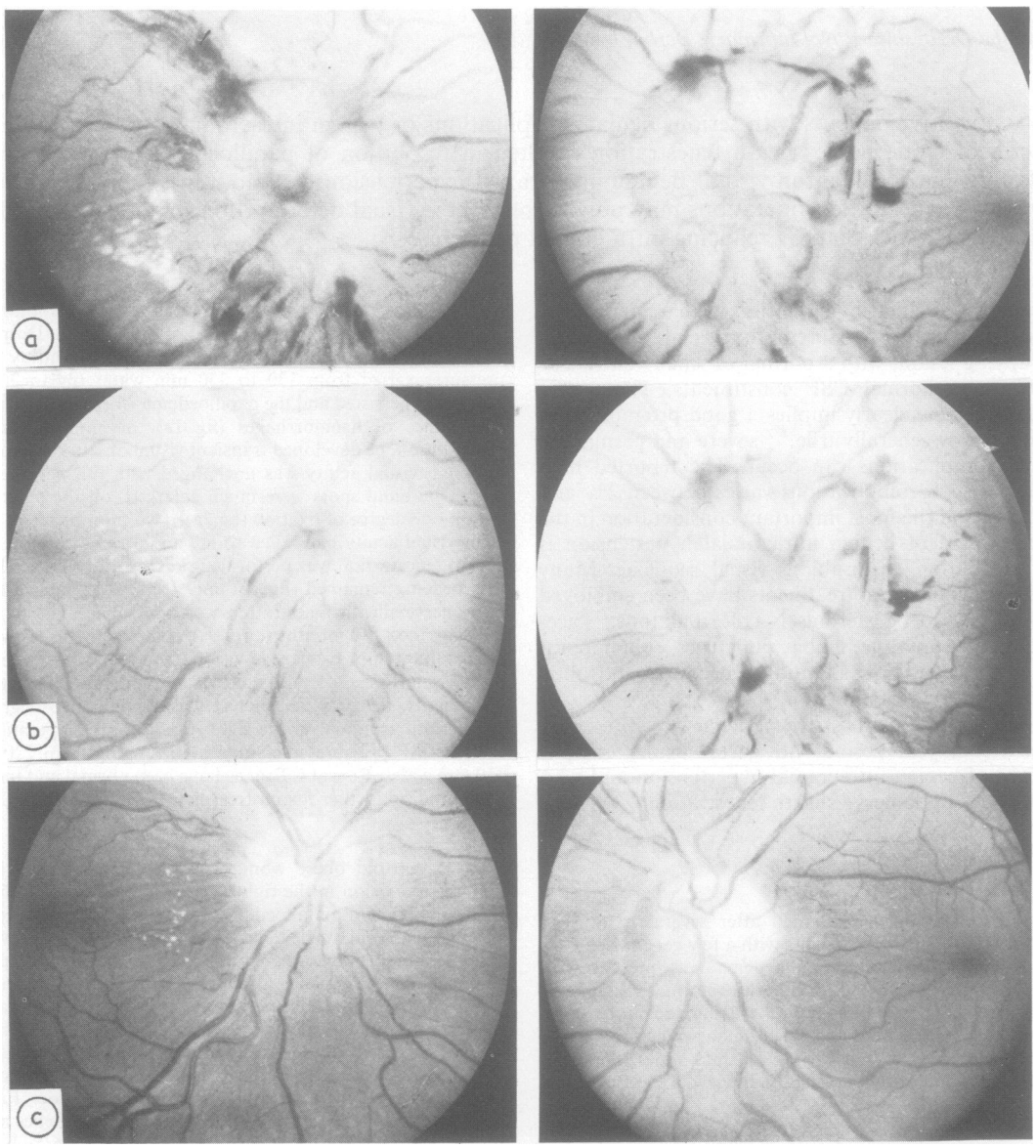

Fig 1 *Case 1: Fundal appearances (a) after six months of medical treatment, (b) twelve days after right optic nerve sheath fenestration showing resolution of only the right disc oedema, (c) several months after left optic nerve sheath fenestration showing complete resolution and consequent optic atrophy.*

in each eye, both discs are atrophic and there are peripheral retinal pigmentary changes corresponding to the posterior ciliary arterial occlusions. Visual fields remain constricted.

Case 3

This 31-year-old obese woman presented with a 3 month history of visual obscurations in the left eye followed, 2 months later, by generalised headaches and bilateral obscurations. A week before presentation, the left eye became blind. Visual acuity was R6/9, left light-perception only, there was bilateral papilloedema and the right visual field was constricted particularly infero-nasally. The lateral ventricles appeared small on CT scan and lumbar puncture yielded normal CSF with a pressure of >300 mm water. Two weeks of treatment with dexamethasone, frusemide and serial lumbar punctures failed to produce improvement. A left optic nerve sheath fenestration was therefore performed with resolution within a week of only the ipsilateral papilloedema; the left vision improved minimally but the right acuity fell to 6/18. Right optic nerve sheath fenestration was therefore performed. Resolution of the disc oedema on this side took a month, with improvement in right acuity to 6/9 and expansion of the visual field; left vision remained unchanged. Immediately after the second procedure, the lumbar CSF pressure exceeded 400 mm water; two weeks later it was only 80 mm but rose to 250 mm over a further two week period and remained elevated thereafter. She became pregnant and gave birth to a normal boy ten months after the operations. Four months after delivery, she returned with transient visual obscurations affecting the right eye; vision was unchanged and the lumbar CSF pressure was 240 mm water. Bilateral optic atrophy was now evident. A lumbar thecoperitoneal shunt was inserted without major complications although she suffered from low-pressure headaches and lumbar wound CSF leakage for two weeks. Later, she developed mild root pain in the right leg attributable to the shunt's tip in the subarachnoid space. The obscurations ceased and, a year later, both visual acuity and visual field remain unchanged. However, two years after operation she developed fatal infection associated with the shunt.

Case 4

This 36-year-old man presented with eight months of left visual loss and three weeks of headaches associated with mild right visual disturbance. Two years previously he had suffered from diplopia for 9 months but no details are available. He took regular vitamin A supplements (in unknown quantities). Bilateral papilloedema was present. Visual acuity was R6/9, left hand movements only; the right blind spot was grossly enlarged (being within a degree of fixation) (fig 3a) but the right peripheral field was full; the left field was grossly constricted. The lateral ventricles appeared small on CT scan and lumbar puncture yielded normal CSF with a pressure of 320 mm water. After one week of dexamethasone therapy, his headaches had ceased but papilloedema and raised CSF pressure persisted. The right visual acuity was 6/9 but the visual field loss had extended leaving only a 1–2 degree central island intact (fig 3b). Right optic nerve sheath fenestration was therefore performed with resolution of only the ipsilateral papilloedema over the ensuing two weeks. Neither right field or left vision improved and the lumbar CSF pressure remained elevated (260 mm water). Three months later, he returned with a sudden deterioration in right visual acuity to finger counting only and the previously preserved central island had been lost despite preservation of

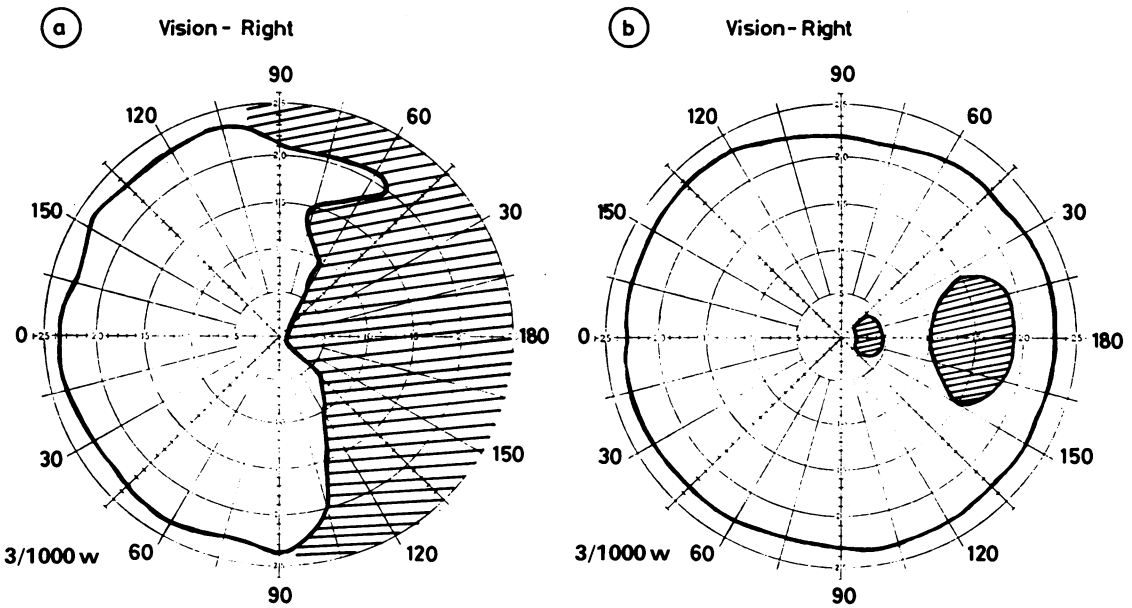

Fig 2 *Case 1: Central visual field (a) right eye: after six months of medical treatment, (b) right eye: one year after right optic nerve sheath fenestration.*

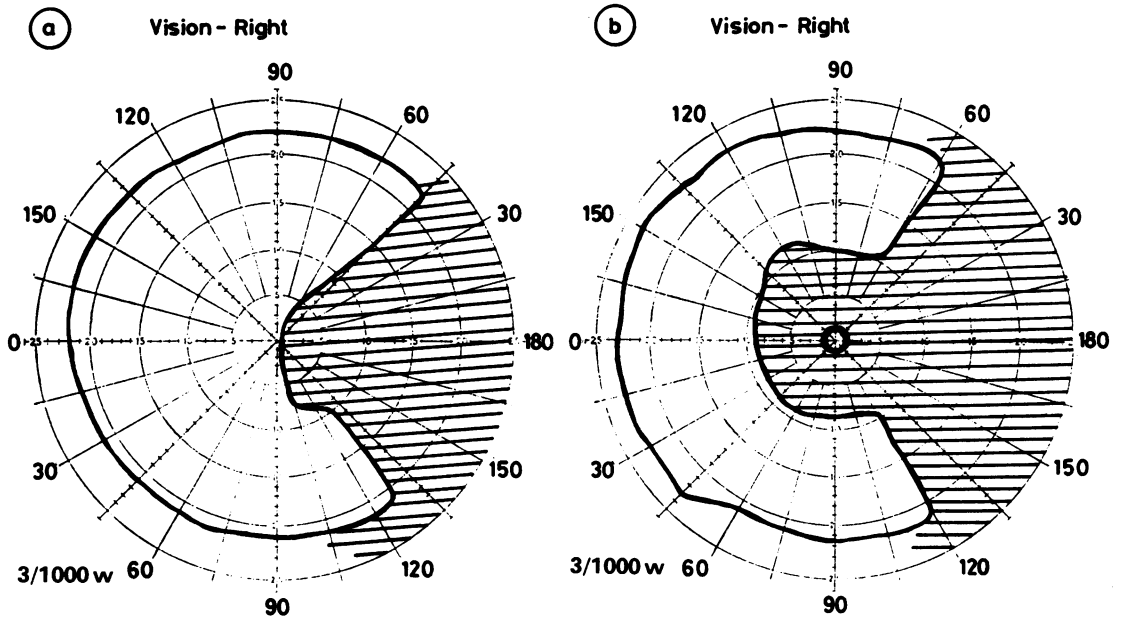

Fig 3 *Case 4: Visual field (a) right eye: central visual field at presentation, (b) right eye: showing deterioration after one week of medical treatment.*

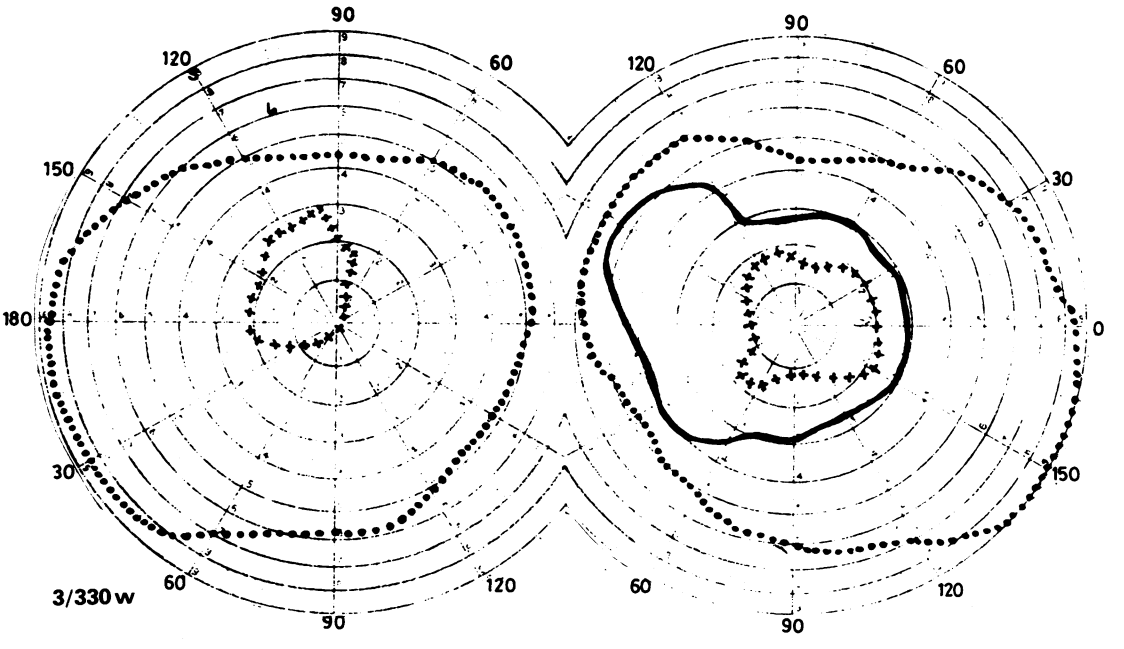

Fig 4 *Case 5: Visual field. (. . . . .) Visual field at re-presentation. (× × × × × ×) Deterioration after a few weeks of medical treatment. (_____) Improvement on the right one week after right optic nerve sheath fenestration.*

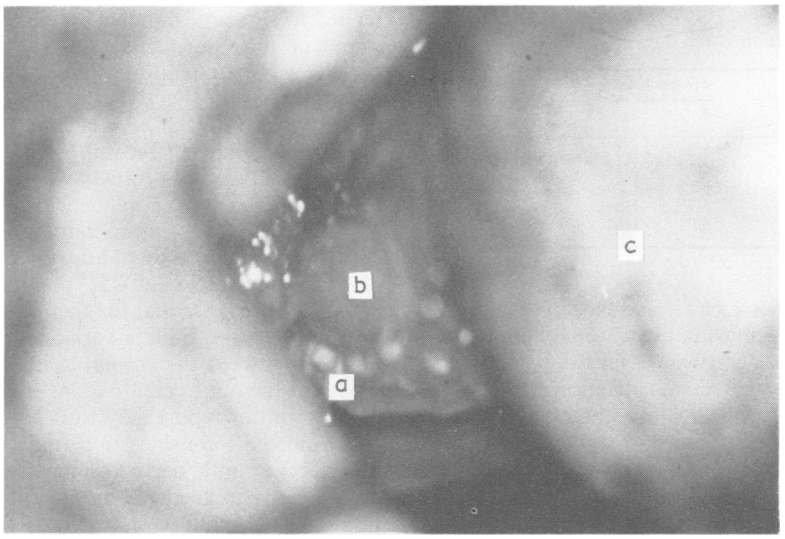

Fig 5 *Operative photograph showing the optic nerve fenestration (a) optic nerve, (b) fenestration, (c) globe.*

the peripheral field. Right optic atrophy was now evident; papilloedema persisted on the left and the lumbar CSF pressure was > 300 mm water. Dexamethasone and frusemide therapy was commenced without visual improvement or fall in CSF pressure. He developed a steroid induced myopathy and became depressed. He was lost to follow up after three months of treatment, with visual acuities of counting fingers on the right and hand movements only on the left.

Case 5

This 20-year-old woman first presented with headaches, bilateral papilloedema and visual acuities of R6/9 L6/9. CT scan and carotid angiography were normal but the CSF pressure was not recorded. Dexamethasone and frusemide therapy produced rapid but incomplete resolution of papilloedema and the visual acuity improved to R6/6 L6/6. Two months later, she was distressed by her cushingoid appearance, treatment was withdrawn and she was lost to follow up. Four years later, she presented again with a 4 day history of visual disturbance. Bilateral papilloedema was present and the lumbar CSF pressure was >350 mm water. A CT scan was again normal but a right carotid angiogram showed delayed filling and poor visualisation of the lateral sinus with narrowed segments in the superior saggital sinus, suggestive of venous sinus thrombosis. Despite four weeks of treatment with frusemide, acetazolamide and anticoagulants, papilloedema persisted with the appearance of secondary optic atrophy and her vision deteriorated to R6/9 L6/60 with gross peripheral field constriction (fig 4). Five weeks after presentation, with acuities of R6/9, left hand movements only, a left optic nerve sheath fenestration was performed with resolution of only the ipsilateral papilloedema over one week but no improvement in the left vision. Although the right visual acuity remained 6/9, the right field loss increased (fig 4) and a right optic nerve sheath fenestration was performed a week after the first operation. The disc oedema settled over two weeks with improvement in the right acuity and field (fig 4). Six years postoperatively, visual acuity is R6/6, hand movements only on the left; there is bilateral optic atrophy and the right visual field is unchanged.

Despite continued high CSF pressure after surgery (still at 260 mm water two years postoperatively), spontaneus retinal venous pulsation was evident within two weeks.

Operative procedure

Essentially, we used the procedure described by Galbraith and Sullivan in 1973 in which the optic nerve sheath is approached by detaching the medial rectus from the globe enabling the eye to be rotated laterally.[7] Using either the retractor described by Galbraith or cotton wool buds on long sticks adequate exposure can be achieved and a window about 5 × 7 mm is excised from the dural sheath of the optic nerve (fig 5). Post-operative complications were minor (except for case 2 as described) and limited to mild conjunctival and lid chemosis. Transient pupillary irregularity was observed in two cases.

Discussion

All five patients presented comply with the definition of benign intracranial hypertension in that they had symptoms and/or signs of raised intracranial pressure without a space-occupying lesion, with a normal or small ventricular system and normal CSF constituents. One patient had angiographic features of venous sinus thrombosis (case 5), a recognised cause of benign intracranial hypertension,[8] another (case 1) had seizures which may occur with intracranial venous thrombosis; and although this diagnosis was not

Table 1 *Visual acuities (VA) at presentation, prior to optic nerve sheath fenestration*

Case	Initial VA		Pre-op VA		Final VA	
	R	L	R	L	R	L
1	6/5	6/5	6/9	6/5	6/5	6/5
2	6/9*	6/5	6/18	6/12	6/9	6/9
3	6/9	LP	6/18	LP	6/9	LP
4	6/9	HM	6/9	HM	CF	HM
5	6/9	6/9	6/9	HM	6/6	HM

*Re-presentation.

confirmed, no other cause was detected over a two year period. Both female patients were obese but neither had menstrual disturbances (case 3 became pregnant). None had an associated infection and only one had been taking any medication known to cause benign intracranial hypertension; case 4 took regular vitamin A supplements (dose unknown).

Each patient developed serious progressive visual problems despite intensive medical treatment and one patient (case 3) died from treatment complications. Two patients (cases 1 and 3) noted transient visual obscurations at a time when there was objective evidence of a worsening condition and some studies have suggested that obscurations indicate a poor prognosis.[3] However, Corbett *et al*[5] reported that 72% of 57 cases had obscurations as an initial symptom and found no correlation with visual outcome. They found that systemic hypertension and, possibly, raised intra-ocular pressure were significant risk factors for visual loss in benign intracranial hypertension. Case 2 was hypertensive but all of our patients had normal intra-ocular pressures. Two patients (cases 3 and 4) had visual failure on presentation, which is known to be associated with poor visual prognosis.[2 9]

Visual acuity reflects foveal function and does not indicate the state of preservation of either the paracentral or peripheral fields. Case 4 is particularly instructive, presenting simply with an enlarged blind spot in his right (good) eye. In one week, whilst on medical treatment, despite the acuity and peripheral field remaining unaffected, the central field loss extended to surround completely the central 1-2 degree island of remaining field (figs 3a, b). These observations serve to reinforce the recommendations in previous reports that visual acuity, peripheral and central fields must all be sequentially recorded.[5 8] In our five cases, the left eye was generally more severely affected than the right (table 1). Hayreh reported earlier and more severe papilloedema on the left in six out of nine benign intracranial hypertension patients.[10] He has also demonstrated that transmission of cranial subarachnoid CSF pressure into the optic nerve sheath is essential for the production of papilloedema and has found a variable degree of communication between the cranial cavity and optic nerve sheath in the optic canal region.[11] The asymmetrical findings in our report may thus have an anatomical basis and these considerations may have bearing on the effects of optic nerve sheath surgery.

Relief of papilloedema by optic nerve sheath incision or fenestration has been reported several times[7 12-19] but in varied conditions. Galbraith and Sullivan[7] reported on the operation in four cases of benign intracranial hypertension but the surgical success was described mainly in terms of papilloedema resolution. Kilpatrick *et al* described operations on 14 patients with benign intracranial hypertension, but the surgical indication was progressive visual failure in only six cases.[20] In one, visual deterioration continued and in the other five, there was said to be no change but no details of vision were given. Aulhorn reported operations in six cases of benign intracranial hypertension but all were long-standing cases with evidence of significant optic atrophy after regression of papilloedema.[21] Vision remained essentially unchanged or worsened, but again few details were given. A recent review described experience with this form of treatment as inchoate.[8] This paper represents the largest published collection of cases with details of postoperative visual improvement. Of our five surgical cases, four had bilateral and one had unilateral operations. In every instance papilloedema resolved, usually over one or two weeks, although unilateral surgery lead to resolution of the ipsilateral papil-

Table 2 *Effects of optic nerve sheath fenestration in papilloedema and vision*

Case	Side of operation	Papilloedema resolution		Post-op VA improvement	Post-op VF improvement
		R	L		
1	R	+	−		
	L	+	+	+	+ (central)
2	BILAT	+	+		
3	L	−	+		
	R	+	+	+	+ (peripheral)
4	R	+	−		
5	L	−	+		
	R	+	+	+	+ (peripheral)

VA = Visual acuity; VF = Visual field (main area of improvement shown in brackets).

loedema alone (fig 1 and table 2). The effect on vision was less predictable but there was significant postoperative improvement in three cases (table 2 and figs 2 and 4). In case 4, there was no initial change and a later deterioration, but he had had visual impairment with optic atrophy for eight months prior to surgery. More disturbingly, there was a sudden postoperative visual loss in case 2 despite resolution of papilloedema. This may have resulted from a disturbance of the choroidal circulation either following the sudden fall in intracranial pressure or possibly due to excessive CSF drainage into the orbits. This was the only case in which simultaneous bilateral fenestration was undertaken, with evidence of a marked fall in CSF pressure and notable conjunctival and lid chemosis. Severe visual loss has been reported following ventriculography[22] and surgical relief of raised intracranial pressure including subtemporal decompression for benign intracranial hypertension.[10 16 23] More relevantly, Davidson has described two patients who suffered sudden serious visual loss in one eye after unilateral optic nerve sheath incisions.[16 17] He stated that intraoperative nerve trauma was not the cause and there was no such trauma in case 2 of our report. In our case, the postoperative visual loss significantly but partially resolved and has now remained static for five years. When visual improvement occurred, it was sustained for the period of follow up (which ranged from 1 to 6 years) although in case 3 transient visual obscurations developed in the right eye 14 months after bilateral optic nerve sheath fenestration. Although there was no evidence of deterioration in vision at this time, the development of obscurations might imply that the previous optic nerve sheath fenestration had not provided long-term protection of the optic nerve. On resolution of the papilloedema, optic atrophy became evident in four cases and earlier operation might have produced better results. This was Aulhorn's conclusion concerning his six chronic benign intracranial hypertension cases.[21]

There are two possible explanations for the operation's success in reducing papilloedema. Firstly, that the surgical defect acts as a CSF shunt to lower intracranial pressure and secondly that the procedure somehow protects the optic nerve from the effects of raised pressure.[13 15 16] There is clinical evidence that optic nerve sheath decompression may lower intracranial pressure in some cases, at least temporarily. Davidson reported five cases of papilloedema due to intracranial neoplasm in whom the operation produced immediate relief of headache and vomiting although the mean postoperative survival time was only eleven weeks.[17] Galbraith and Sullivan also reported immediate postoperative headache relief in two intracranial tumour cases but only for ten days while permanent relief resulted in one case of benign intracranial hypertension.[7] In case 2 of our report, despite a marked postoperative fall in CSF pressure it was again elevated within two weeks. In all our cases and in most reports, with the notable exception of Kilpatricks,[20] unilateral surgery produces only ipsilateral effects (table 2).[13 15 24] Kaye *et al*[19] and Kilpatrick *et al*[20] have reported continued high readings on postoperative intracranial pressure monitoring, despite papilloedema resolution. Further studies involving such monitoring are needed especially as CSF pressure is known to fluctuate spontaneously in benign intracranial hypertension.[25] Davidson[16 17] reported two cases which came to necropsy after surgery and the incisions were sealed by granulation tissue within one and three weeks after operation respectively, although Keltner[13] has reported a case with a sheath split still patent at 29 days. Incision-slit and excision-window defects may have different closure rates but there are no pathological reports on fenestration cases and it is unlikely that any defect could remain indefinitely patent. It could be argued that even temporary patency may be satisfactory in benign intracranial hypertension but it is not likely that patients with a truly benign self-limiting illness would come to surgery. As an alternative hypothesis, Davidson[16] has suggested that optic nerve sheath surgery results in adhesions which may protect the nerve from the effects of CSF pressure and the possible relevance of Hayreh's work has already been mentioned. Other surgical treatments are directly aimed at lowering CSF pressure but, as case 3 illustrates, these are not without complications.

Conclusion

Benign intracranial hypertension is an unsatisfactory name for a condition that may often not be benign for vision. There is no attempt, in this paper, to establish the incidence of serious visual loss but our experience contrasts with that of Jefferson and Clark who found only two cases with significant visual impairment over a thirty year period.[26] They stated that vision consistently improved on simple dehydration therapy and concluded that "surgery is never required in the management of benign intracranial hypertension". Medical treatment of benign intracranial hypertension is outside the scope of this paper and remains controversial.[6] However, any therapy must approach either the basic pathogenetic mechanisms (which are uncertain and possibly varied), the resultant raised intracranial pressure or the principal target organ at risk. Optic nerve sheath fenestration always relieves papilloedema although it lowers intracranial pressure only occasionally and then temporarily. The operation will improve vision only if performed before irreversible

optic nerve damage has occurred and, therefore, patients must be carefully and frequently monitored, It is not a technically simple procedure and potential complications exist although the risk of serious sudden visual deterioration may not be unique to this treatment but may result from any therapy that rapidly reduces intracranial pressure. Other surgical approaches, such as lumbar-thecal peritoneal shunting have their own complications. We suggest that optic nerve sheath fenestration should be considered in cases of benign intracranial hypertension which, despite medical treatment, suffer serious, progressive visual impairment. Whatever the treatment employed, lengthy follow up is obviously necessary with regular assessment of visual acuity, central and peripheral visual fields.

We thank Dr D Jefferson, Dr RB Godwin-Austin, Mr R Shepherd and Mr J Taylor for allowing us to study their patients.

References

[1] Johnston I, Paterson A. Benign intracranial hypertension I: diagnosis and prognosis. *Brain* 1974;**97**:289–300.

[2] Boddie HG, Banna M, Bradley WG. "Benign" intracranial hypertension—a survey of the clinical and radiological features and long-term prognosis. *Brain* 1974;**97**:313–26.

[3] Weisberg LA. Benign intracranial hypertension. *Medicine* 1975;**54**:197–207.

[4] Foley J. Benign forms of intracranial hypertension—"toxic" and "otitic" hydrocephalus. *Brain* 1955; **78**:1–41.

[5] Corbett JJ, Savino PJ, Thompson HS. Visual loss in pseudotumour cerebri: follow-up of 57 patients from 5 to 41 years and a profile of 14 patients with permanent severe visual loss. *Arch Neurol* 1982;**39**:461–74.

[6] Keltner JL, Miller NR, Gittinger, JW and Burde RM. Pseudotumour cerebri. *Surv Ophthalmol* 1979;**23**: 315–22.

[7] Galbraith JEK, Sullivan JH. Decompression of the perioptic meninges for relief of papilloedema. *Am J Ophthalmol* 1973;**76**:687–92.

[8] Rush JA. Pseudotumour cerebri. *Br J Hosp Med* 1983;**29**:320–5.

[9] Bulens C, Devries WAEJ, Van Crevel H. Benign intracranial hypertension—a retrospective and follow-up study. *J Neurol Sci* 1979; **40**:147–158.

[10] Hayreh SS. Optic disc oedema in raised intracranial pressure 5: Pathogenesis. *Arch Ophthalmol* 1977;**95**: 1553–65.

[11] Hayreh SS. Pathogenesis of oedema of the optic disc. *Doc Ophthalmol* 1968;**24**:289–411.

[12] Gutgold-Glen H, Kattah JC and Chavis RM. Reversible visual loss in Pseudotumour Cerebri. *Arch Ophthalmol* 1984;**102**:403–6.

[13] Keltner J Albert PM, Lubow M. Optic Nerve Decompression: A clinical pathological study. *Arch Ophthalmol* 1977;**95**:97–104.

[14] Smith JL. Optic nerve sheath decompression for relief of chronic monocular choked disc. *Am J Ophthalmol* 1969;**68**:633–9.

[15] Burde RM Karp JS and Miller RN. Reversal of visual defect with optic nerve decompression in longstanding Pseudotumour cerebri. *Am J Ophthalmol* 1974;**27**:770–2.

[16] Davidson SI. The surgical relief of papilloedema. In: Cant JS, ed. *The Optic Nerve*. London: Henry Kimpton, 1972:174–9.

[17] Davidson SI. A surgical approach to plerocephalic disc oedema. *Trans Ophthalmol Soc UK* 1969;**89**:669–90.

[18] Hayreh SS. Pathogenesis of oedema of the optic disc (papilloedema). (A preliminary report). *Br J Ophthalmol* 1964;**48**:522–43.

[19] Kaye AH, Galbraith JE and King J. Intracranial pressure following optic nerve decompression for BIH. *J Neurosurg* 1981; **55**:453–6.

[20] Kilpatrick CJ, Kaufman DV, Galbraith JE and King JO. Optic nerve decompression in benign intracranial hypertension. *Clin Exp Neurol* 1981; **18**:161–8.

[21] Aulhorn E. Ophthalmological features of pseudo-tumor cerebri. Report on 18 cases. *Doc Ophthalmol* 1984; **58**:25–33.

[22] Keane JR. Sudden blindness after ventriculography. *Am J Ophthalmol* 1974;**78**:275–8.

[23] Holmes G. The prognosis in papilloedema. *Br J Ophthalmol* 1937;**21**:337–42.

[24] Lubow M, Kuhr L. Pseudotumour cerebri: comments on practical management. In: Glaser J, ed. *Neuro-Ophthalmology*. St Louis: Mosby and Co., 1977:199–206.

[25] Gucer G. Viernstein L. Long-term intracranial pressure recording in the management of pseodotumour cerebri. *J Neurosurg* 1978;**49**:256–63.

[26] Jefferson A, Clark J. Treatment of benign intracranial hypertension by dehydrating agents with particular reference to the measurement of the blind spot area as a means of recording improvement. *J Neurol Neurosurg Psychiatry* 1976;**39**:627–39.