Asymmetric Papilledema and Visual Loss in Pseudotumour Cerebri

Charles E. Maxner, Mark I. Freedman and James J. Corbett

ABSTRACT: We report the case of a 26 year old obese woman who presented with intermittent headaches and blurred vision in her left eye (OS) and on clinical examination had an enlarged visual field blind spot OS with OS disc edema. After an extensive neurologic work up including two nondiagnostic lumbar punctures, a clinical diagnosis of OS anterior ischemic optic neuropathy was made. Gradual progression of visual field loss OS prompted reassessment of the diagnosis and intracranial pressure was confirmed to be markedly elevated by usage of a subarachnoid monitoring bolt, thus establishing the diagnosis of pseudotumour cerebri. An optic nerve sheath fenestration was performed OS with subsequent reversal of the progressive visual field loss.

RÉSUMÉ: Asymétrie de l'oedème papillaire et perte d'acuité visuelle dans le syndrome d'hypertension intracrânienne bénigne. Nous rapportons le cas d'une femme obèse âgée de 26 ans qui a consulté pour des céphalées intermittentes et une vision embrouillée de son oeil gauche (OS) et qui, à l'examen clinique, avait une tache aveugle élargie au champ visuel OS avec un oedème papillaire OS. A la suite d'une investigation neurologique extensive incluant deux ponctions lombaires qui ne permirent pas d'orienter le diagnostic, un diagnostic clinique de neuropathie optique antérieure d'origine ischémique OS a été posé. La progression graduelle de l'atteinte du champ visuel nous a incités à réévaluer ce diagnostic et nous avons constaté une élévation importante de la pression intracrânienne par monitorage au moyen d'un capteur de pression sous-arachnoïdien, posant ainsi le diagnostic de syndrome d'hypertension intracrânienne bénigne. Une fenestration de la gaine du nerf optique OS a été pratiquée entraînant une régression ultérieure de l'atteinte progressive du champ visuel.

Can. J. Neurol. Sci. 1987; 14:593-596

Pseudotumour cerebri is a common condition of uncertain etiology characterized by raised intracranial pressure (ICP) in patients with normal cerebrospinal fluid (CSF) biochemical profile and normal neuroradiographic studies. The patients are usually obese young women who present with headaches, blurred vision and bilateral papilledema. Visual field testing shows enlarged blind spots and elevated ICP is confirmed by lumbar puncture.[1] We report the case of a young woman who presented with markedly asymmetric and progressive ophthalmological symptoms and signs. Lumbar puncture revealed normal CSF pressure and invasive monitoring was required to identify raised ICP.

Case Report

A 26-year-old right handed white female noticed an abrupt onset of a "black circle" in the temporal field of vision of her left eye (OS) associated with intermittent headache. She had a history of common migraine. She also had a 20 pack-year smoking history, was using oral contraceptives, was obese (Ht = 157 cm, Wt = 88 kg), and had essential hypertension. Confrontation visual fields confirmed a temporal visual field scotoma OS and ophthalmoscopy revealed an edematous disc OS. The following studies were normal: CBC, ESR, ANA, VDRL, cranial CT-scan, echocardiogram, and CSF profile. She had a mild thrombocytosis. A lumbar puncture was performed and the opening pressure was 130 mm CSF. A presumptive diagnosis of anterior ischemic optic neuropathy (AION) was made. Coumadin anticoagulation was initiated and the patient discontinued smoking and oral contraceptive usage. Several weeks later she was seen in the University of Iowa Neuro-Ophthalmology Clinic for further assessment. In the interim, her headaches had persisted and she had some enlargement of the OS scotoma. On examination, visual acuity was 20/20 in both eyes with symmetric optic nerve function test results although a 0.3 log unit relative afferent pupillary defect was seen OS. Goldmann perimetry revealed an enlarged blind spot OS with a shrunken I2 isopter (Figure 1a). Ophthalmoscopy (Figure 2) showed distinct OS disc edema with the right (OD) disc appearing clinically normal although subsequent review of stereo disc photographs obtained at the time suggested that there was some OD disc elevation. We continued to believe that the patient had AION. Coumadin was discontinued and replaced with aspirin.

Two weeks later the patient reported increasing transient visual obscurations (TVOs) and darkening of the OS scotoma. Goldmann

From the Department of Ophthalmology (Drs. Maxner, Freedman, Corbett) and the Department of Neurology (Dr. Corbett), University of Iowa Hospitals and Clinics, Iowa City

Received August 7, 1986. Accepted in final form May 28, 1987

This paper was presented in part at the XXIst Canadian Congress of Neurological Sciences, London, Ontario, June 24-28, 1986

Reprint requests to: C.S. O'Brien Library, University of Iowa Hospitals and Clinics, Iowa City, Iowa, U.S.A. 52242

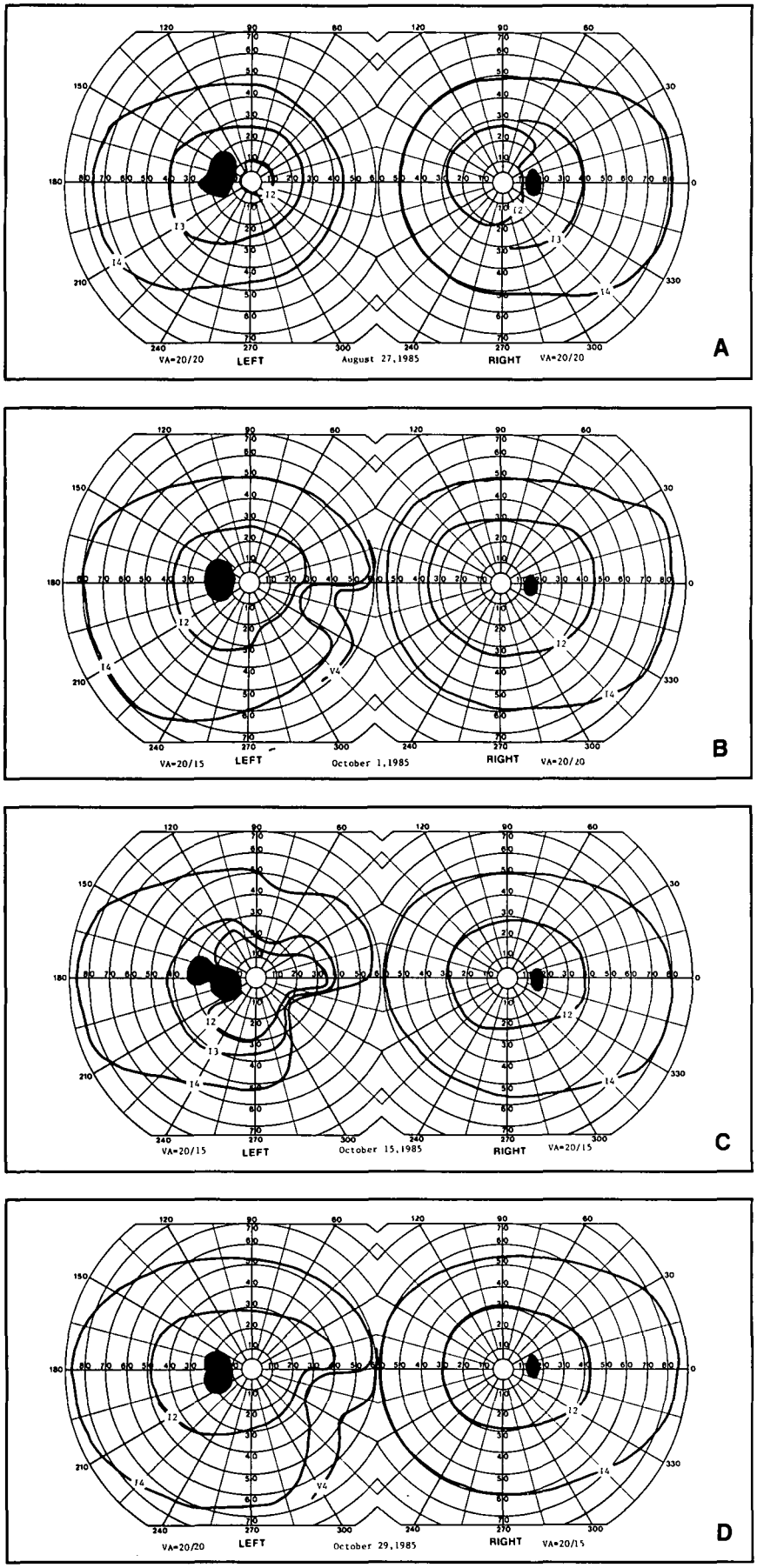

Figure 1 — Sequence of visual fields as determined by using the Goldmann Perimeter. A) Visual fields on the first visit with the enlarged blind spot and shrunken I2 isopter evident in the left eye. B) Perimetry findings on the first follow-up visit with the inferonasal field defect noticeable in the left eye. C) Visual fields on the second follow-up visit demonstrating further blind spot enlargement and inferonasal field loss in the left eye. D) Two weeks following optic nerve sheath fenestration in the left eye with the visual field defects having significantly resolved.

https://doi.org/10.1017/S0317167100037422 Published online by Cambridge University Press

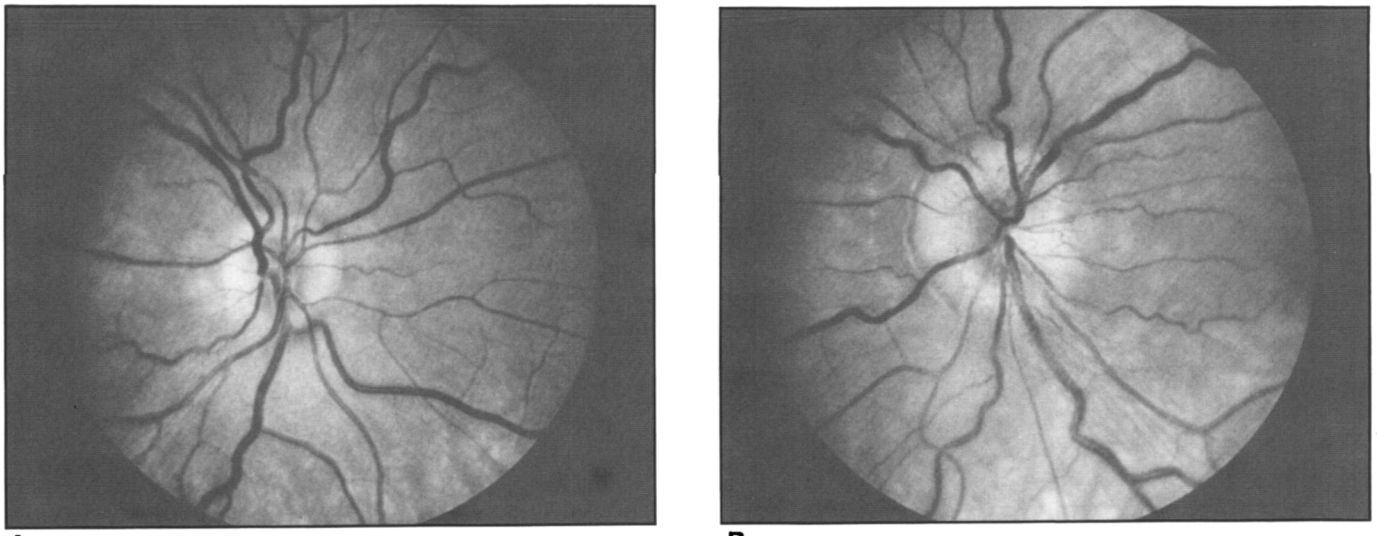

Figure 2 — Fundus photographs of the right eye (A) and left eye (B) demonstrating marked disc edema in the left eye with an essentially normal disc in the right eye.

perimetry (Figure 1b) revealed new inferonasal notches in several isopters OS. Examination was essentially unchanged except for slight disc edema OD in addition to the marked disc edema OS. Given the somewhat anomalous vasculature, pseudopapilledema was considered in the differential diagnosis however, no disc drusen were identified ophthalmoscopically. No autofluorescence was noted on the earliest phases of the fluorescein angiogram again tending to rule out disc drusen. Fluorescein angiography confirmed bilateral, markedly asymmetric papilledema (Figure 3). The lumbar puncture was repeated but again was not diagnostic[2] with an opening pressure of 220 mm CSF.

Over the next two weeks the patient's TVOs and visual field defects OS worsened. Goldmann perimetry (Figure 1c) revealed further enlargement of the blind spot and an increasing inferonasal field defect OS. Ophthalmoscopic findings were essentially unchanged. A subarachnoid ICP monitoring bolt[3] was inserted and ICP greater than 45 mm Hg (600 mm CSF) was recorded (Figure 4) finally confirming a diagnosis of pseudotumour cerebri.

Because she had progressive vision loss OS an optic nerve sheath fenestration was performed on the left eye. Intracranial pressure continued to be elevated during continuous postoperative subarachnoid ICP monitoring done for 36 hours post-fenestration. Despite persistence of ICP elevation the disc edema OS appeared diminished within two days post-operatively and Goldmann perimetry showed significant resolution of the visual field defect OS (Figure 1d). On follow-up assessment over the ensuing six months further resolution of the disc edema was clinically evident again confirming that indeed the elevation was due to true papilledema and not the elevation associated with pseudopapilledema.

Discussion

Patients with acute pseudotumour cerebri classically present with bilateral papilledema.[4,5] This case and previous reports in the literature[6,7] indicates that, asymmetric or unilateral papilledema may occasionally be seen. Hayreh[8] utilized inflatable intracranial balloons in monkeys and was able to demonstrate that when the intracranial pressure was elevated and papilledema developed, the eye ipsilateral to the balloon commonly demonstrated the earliest changes, particularly when the balloon was in a supratentorial position. More recently, Hayreh[9] demonstrated that an intact optic nerve sheath was necessary for raised intracranial pressure to cause swelling of the optic disc. He suggested that asymmetric or unilateral

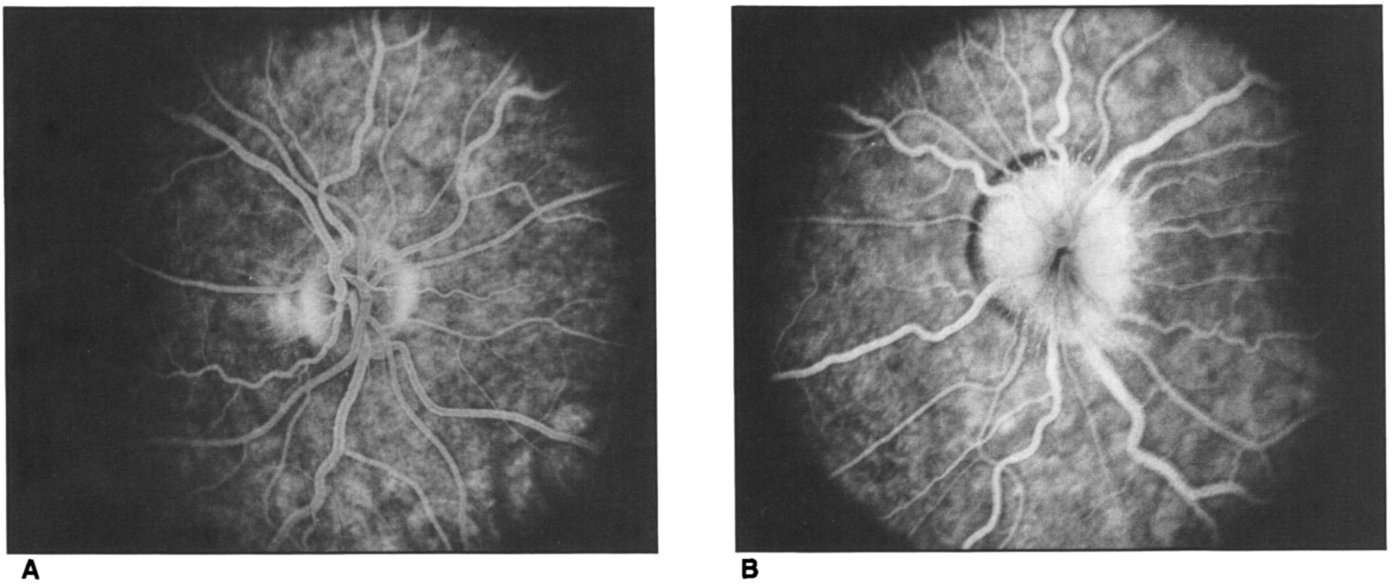

Figure 3 — Fluorescein angiography of the right eye (A) and left eye (B). Although the increased vascularity and dye leakage from the left disc is evident, the bilaterality of the papilledema is confirmed by the slight dye leakage seen most easily temporally from the right disc.

https://doi.org/10.1017/S0317167100037422 Published online by Cambridge University Press

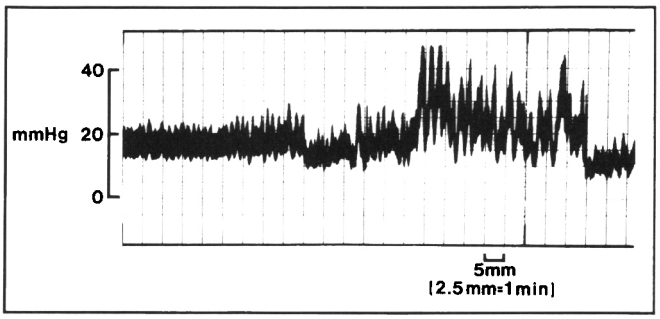

Figure 4 — Portion of the ICP monitoring record demonstrating a period of markedly elevated intracranial pressure with peak values greater than 45 mm Hg, this occurring while the patient was sleeping at approximately 1:30 a.m.

due to differences in the ease with which intracranial pressure was transmitted to the optic nerve sheaths through the naturally formed trabecular meshwork of fibrous adhesions between the dura and pia that cover the optic nerve in the optic canal. This meshwork effectively reduces the subarachnoid space around the optic nerve and is quite variable from specimen to specimen. Thus the optic nerve with the larger subarachnoid space would be expected to demonstrate disc edema earlier and to a greater extent when the intracranial pressure became elevated.

The CSF opening pressure in our patient was below 250 mm CSF on two occasions. This was decidedly unusual given the results of lumbar puncture studies in 116 patients with acute pseudotumour cerebri reported by Corbett and Mehta.[2] In their study, no acute patient had a pressure less than 200 mm CSF and only 14% had CSF pressures in the non-diagnostic 200 to 250 mm range. The ICP monitoring results in our patient along with previously reported studies utilizing intraventricular catheters,[10] extradural sensors,[11] long term subdural sensors,[12] and lumbar spinal canal CSF pressure monitoring[13] show that the ICP may have marked short term (minutes to hours) and long term (days to weeks) fluctuations in patients with pseudotumour cerebri. In occasional patients it is necessary to maintain a high degree of suspicion for a diagnosis of pseudotumour cerebri even if routine lumbar puncture pressure values are normal.

Although the classical visual field defect in pseudotumour cerebri is enlargement of the blind spots,[1] more serious visual defects are recognized.[4,5,14] Corbett et al[14] reported that 25% of a series of 57 patients with long term follow-up developed serious unilateral or bilateral visual loss with the field defects tending to be disc-related with inferior nasal defects, arcuate defects, concentric constriction, and cecocentral scotomas. These visual field defects may be reversible but may progress. Exactly when to undertake surgical intervention is still a matter of personal judgement however, if vision continues to deteriorate on appropriate medical therapy, we recommend surgical treatment. Previous procedures consisted of subtemporal decompression or lumbar-peritoneal shunting.[1] Another approach is the optic nerve sheath fenestration.[15,16] This consists of excision of a rectangle of dura from the optic nerve sheath using either a lateral or medial orbitotomy approach. This procedure was initially reported by deWecker[17] in 1872 and has recently been repopularized. The efficacy of optic nerve sheath fenestration in reducing papilledema and reversing visual dysfunction is not likely to be due to lowering of intracranial pressure via a CSF leak. Our patient and the patient reported by Kaye et al[18] demonstrated continued elevation of CSF pressure with ICP monitoring following optic nerve sheath fenestration. Hayreh[9] has demonstrated in the rhesus monkey, that optic nerve sheath fenestration prevents disc edema from occurring in the operated eye when the intracranial pressure is again raised suggesting that the effect seen in patients with pseudotumour cerebri who undergo optic nerve sheath fenestration is due to a local drop in optic nerve sheath pressure.

This case illustrates several important features of pseudotumour cerebri. The papilledema can be unilateral or strikingly asymmetric. Single normal CSF pressure measurements do not rule out pseudotumour cerebri. Visual field defects can be more extensive than blind spot enlargement and are potentially reversible. Finally, occasional patients with pseudotumour cerebri, may benefit from surgical intervention.

Acknowledgements

We wish to thank Dr. W. Risk of Cedar Rapids, Iowa for referring this patient. This study was supported in part by an unrestricted grant from Research to Prevent Blindness to the Department of Ophthalmology. Dr. Maxner was supported by the E.A. Baker Foundation for Prevention of Blindness, Toronto, Ontario.

References

1. Corbett JJ. Problems in the diagnosis and treatment of pseudotumour cerebri. Can J Neurol Sci 1983; 10: 221-229.
2. Corbett JJ, Mehta MP. Cerebrospinal fluid pressure in normal obese subjects and patients with pseudotumour cerebri. Neurology (Cleveland) 1983; 33: 1386-1388.
3. Mann JD, Butler AB, Rosenthal JE, et al. Regulation of intracranial pressure in rat, dog, and man. Ann Neurol 1978; 3: 156-165.
4. Aulhorn E. Ophthalmological features of pseudotumour cerebri: report on 18 cases. Documenta Ophthalmologica 1984; 58: 25-33.
5. Orcutt JC, Page NGR, Sanders MD. Factors affecting visual loss in benign intracranial hypertension. Ophthalmology 1984; 91: 1303-1312.
6. Kirkham TH, Sanders MD, Sapp GA. Unilateral papilloedema in benign intracranial hypertension. Can J Ophthalmol 1973; 8: 533-538.
7. Sedwick LA, Burde RM. Unilateral and asymmetric optic disk swelling with intracranial abnormalities. Am J Ophthalmol 1983; 96: 484-487.
8. Hayreh SS. Pathogenesis of edema of the optic disc (papilloedema) — A preliminary report. Br J Ophthalmol 1964; 48: 522-543.
9. Hayreh SS. The sheath of the optic nerve. Ophthalmologica 1984; 189: 54-63.
10. Johnston I, Paterson A. Benign intracranial hypertension II. CSF pressure and circulation. Brain 1974; 97: 301-312.
11. Gücer G, Viernstein L. Long-term intracranial pressure recording in the management of pseudotumour cerebri. J Neurosurg 1978; 49: 256-263.
12. Cooper PR, Moody S, Sklar F. Chronic monitoring of intracranial pressure using an *in vivo* calibrating sensor: experience in patients with pseudotumour cerebri. Neurosurgery 1979; 5: 666-670.
13. Spence JD, Amacher AL, Willis NR. Benign intracranial hypertension without papilledema: role of 24-hour cerebrospinal fluid pressure monitoring in diagnosis and management. Neurosurgery 1980; 7: 326-334.
14. Corbett JJ, Savino PJ, Thompson HS, et al. Visual loss in pseudotumour cerebri: follow-up of 57 patients from five to 41 years and a profile of 14 patients with permanent severe visual loss. Arch Neurol 1982; 39: 461-474.
15. Galbraith JEK, Sullivan JH. Decompression of the perioptic meninges for relief of papilledema. Am J Ophthalmol 1973; 76: 687-692.
16. Burde RM, Karp JS, Miller RN. Reversal of visual deficit with optic nerve decompression in long-standing pseudotumour cerebri. Am J Ophthalmol 1974; 77: 770-772.
17. deWecker L. On incision of the optic nerve in cases of neuroretinitis. Int Ophthalmol Congr Rep 1872; 4: 11-12.
18. Kaye AN, Galbraith JEK, King J. Intracranial pressure following optic nerve decompression for benign intracranial hypertension. Case report. J Neurosurg 1981; 55: 453-456.