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Optic nerve disorders: role of canal and nerve sheath decompression surgery

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Abstract

Optic nerve sheath decompression (ONSD) maintains a role in the management of visual loss complicating papilloedema in selected patients primarily with idiopathic intracranial hypertension. The evidence base for this intervention is reviewed and audit data on visual outcomes for patients with acute, chronic, and atrophic forms of papilloedema are contrasted. Optic canal decompression has a role in the management of compressive optic neuropathies complicating mass lesions arising from paranasal sinuses and intracranially and can be achieved by transethmoidal, transbasal, and open craniotomy routes. The evidence base supporting this intervention in traumatic optic neuropathy and in primary bone disease causing canal stenosis (in particular fibrous dysplasia) is reviewed where the indications are more controversial.
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Keywords: optic nerve sheath decompression; optic canal decompression; papilloedema; compressive optic neuropathy

Introduction

Surgical decompression of the optic nerve within its canal, and of the anterior optic nerve sheath within the orbit continues to generate debate regarding indications and efficacy. To some extent, both operations share a background of technical progress arriving in advance of clinical scientific understanding. In the case of nerve sheath decompression, the technical progress took place a long time ago in 1872 with a report by De Wecker[1] who worked without direct visualisation of the nerve and without a clear understanding of the distinction between papilloedema due to raised intracranial pressure and disc swelling from local causes.[1] It is noteworthy that at the fourth international ophthalmological congress where De Wecker presented his paper in London, during the same session there was a vigorous debate regarding the safety of chloroform *vs* ether *vs* no anaesthetic at all for cataract surgery. Little did he know that both minimal anaesthetic cataract surgery and the role of optic nerve sheath decompression (ONSD) would still be debated 130 years later.

Optic nerve sheath decompression

Pathophysiology

In raised intracranial pressure, the intrasheath cerebrospinal fluid (CSF) pressure of the anterior optic nerve rises due to transmission of the raised intracranial CSF pressure through the trabecular meshwork of the optic canal. Interrupted fast and slow axoplasmic transport at the lamina cribrosa results in ophthalmoscopically visible papilloedema and eventual loss of vision. Decompression of the sheath by cutting a window in the dura and arachnoid of the bulbous part of the sheath of the optic nerve leads to resolution of the disc swelling on the operated side in both the animal model and in patients[2] (Figure 1). In some instances, headaches and contralateral disc swelling are relieved, suggesting that there is a prolonged lowering of intracranial pressure via a persistent orbital fistula. More typically, however, the operation only achieves a local effect and the headaches and contralateral papilloedema deteriorate even if the ipsilateral

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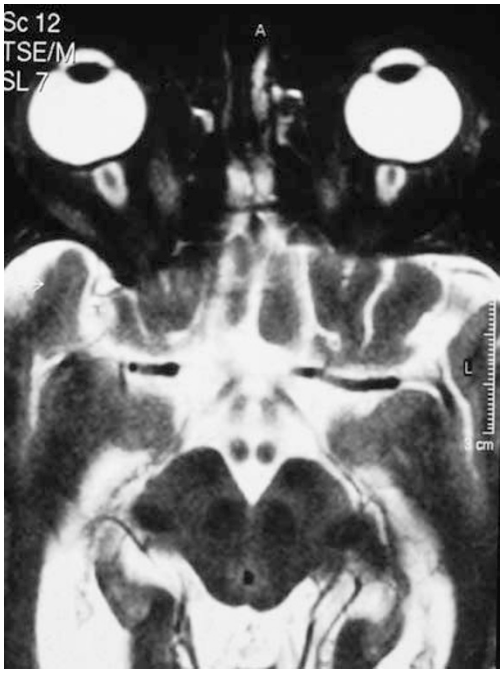

Figure 1 Orbital MRI scan showing dilated optic nerve sheaths and indented posterior globes in chronic papilloedema due to raised intracranial pressure.

disc and visual function improve. Reparative fibrosis at the site of fenestration may prevent transmission of raised intracranial pressure past the subarachnoid scars and protect the ipsilateral optic nerve but not reduce the intracranial pressure (Figure 2a and b). In some instances, a CSF-filled cyst may form in the orbit at the site of sheath decompression and this may serve as a filtration bleb. However, accurate direct measurement of intrasheath CSF pressure *in vivo* has yet to be achieved and so the question of continuing CSF leak into the orbit remains speculative.[3–5]

ONSD for visual failure associated with raised intracranial pressure

Resolution of papilloedema is often, but not always, accompanied by improvement in the visual field and acuity. Numerous reports have testified to the role of ONSD for papilloedema when visual function fails and medical measures are inadequate to restore or stabilise vision.[6–11] This applies not only to idiopathic intracranial hypertension but also to selected other causes of refractory chronic raised intracranial pressure such as cerebral venous sinus thrombosis and other causes of dural venous sinus outflow obstruction.[12,13] Some failures can be attributed to delayed intervention when secondary optic atrophy and loss of axonal population will be irreversible.

An audit within our own department clearly illustrates superior visual outcomes when the procedure is

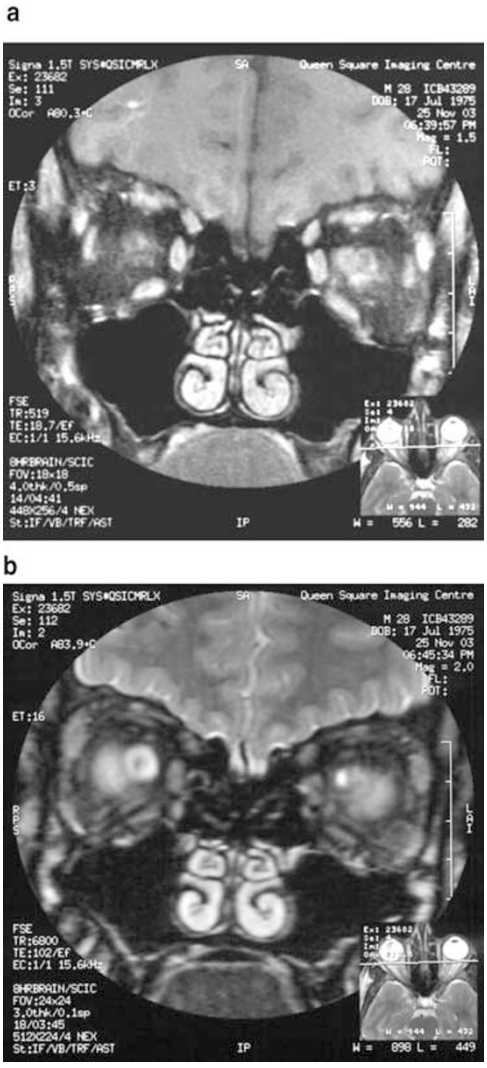

Figure 2 (a) and (b) Early postsurgical orbital MRI following left optic nerve sheath decompression. A dilated distal optic nerve sheath is seen with contrast enhancement medially at the decompression site.

performed in the acute phase of disc swelling compared to the atrophic phase and this is in concordance with reports from elsewhere (Figure 3). The same audit of a series of patients undergoing ventriculoperitoneal shunting for progressive visual loss in papilloedema showed similar and expected superior outcomes in acute patients. This reflects the greater visual potential of the eye without secondary optic atrophy, and also the inherent bias of retrospective studies that fail to account for spontaneous remission rates.

Long-term results from ONSD and revision ONSD

Data on long-term results following ONSD are limited and hard to interpret in the absence of adequate

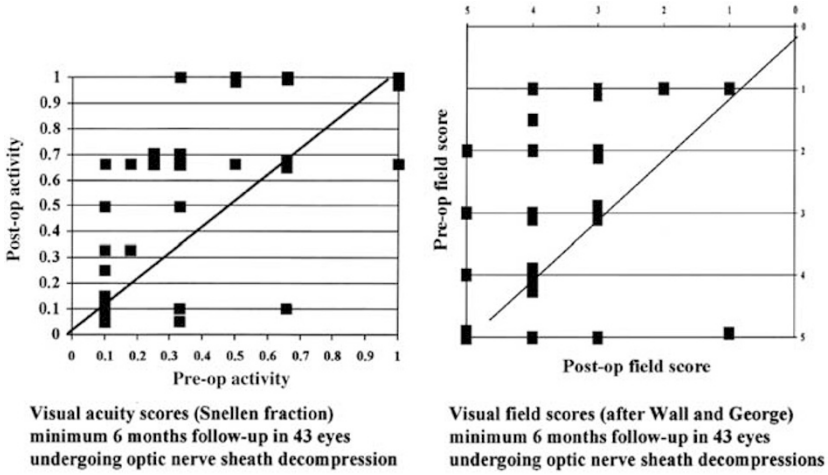

Figure 3 (a Left) Visual acuity scores (Snellen's fraction) minimum 6 months follow-up in 43 eyes undergoing ONSD. (b Right) Visual field scores (after Wall and George) minimum 6 months follow-up in 43 eyes undergoing ONSD.

follow-up of controls.[14] However, Spoor reported deterioration in visual function in 24 out of 75 eyes between 1 and 39 months after initially successful ONSD. In that series, revision ONSD was performed with stabilisation of vision in about 50% of cases.[15] Adjuvant mitomycin C has been used in such cases apparently without adverse effects to the optic nerve,[16,17] although there is a dose-dependent toxic effect on the VEP in a rabbit model. In our clinic, patients with failed ONSD are generally referred for shunting rather than revision surgery.

CSF diversion surgery by shunting

A shunt will have the additional benefit of headache relief when successful and experience of these techniques is extensive, particularly by the lumboperitoneal route. However, shunts may fail because of disconnection or obstruction and may be contraindicated in instances of cerebellar tonsillar herniation into the foramen magnum.[18] As a result, patient selection is important: as a primary procedure for idiopathic intracranial hypertension, shunting is usually recommended where headache is the dominant complaint, and ONSD initially when visual failure is predominant.[19–21] Surgical failure may then be followed if necessary by a shunt, which, in our practice, is usually preferred to revision ONSD. The selection of CSF diversion surgical technique remains strongly influenced by available local expertise, and multicentre prospective controlled studies are urgently needed.[22–24]

ONSD for other disorders

After a period of enthusiasm, the role of ONSD for the progressive form of nonarteritic ischaemic optic neuropathy has passed out of favour. The concept that decompressing the retrolaminar optic nerve fibres would ameliorate a disease process acting at the level of the lamina cribrosa was not supported by the Ischemic Optic Neuropathy Decompression Trial in 1995.[25] This study underlined the modest but significant spontaneous recovery rate seen when these patients are carefully followed without any intervention. There are also reports of a possible role for ONSD in central retinal vein occlusion, post-traumatic venous obstructive retinopathy, radiation optic neuropathy, optic disc drusen, and optic disc pit-associated central serous retinal detachment. In chronic meningitis, spinal block, and papilloedema associated with Guillain–Barre syndrome, raised CSF protein is strongly associated with limited efficacy of any CSF diversion procedure.[26,27]

Transconjunctival medial orbitotomy approach to biopsy the optic nerve sheath obtain local CSF specimens, and sometimes the optic nerve parenchyma in blind eyes with suspected malignant infiltrative processes may be a useful low morbidity approach to diagnosis in selected patients[28] (Table 1).

Optic canal decompression

Pathophysiology

Visual function may be lost when the optic nerve is impinged on by an extrinsic lesion. Initially, this is the result of reversible physiological axonal conduction block but later processes of demyelination and both retrograde and orthograde (Wallerian) axonal degeneration and ischaemia, all play a less reversible role.[29,30] Decompression surgery within the confines of the optic canal will relieve conduction block, but complex

Table 1 ONCD: evidence for efficacy

A: *Supportive descriptive studies*
Papilloedema complicating idiopathic intracranial hypertension
Papilloedema complicating intracranial dural sinus obstruction
B: *Anecdotal/single case reports*
Central retinal vein occlusion
Post-traumatic venous obstructive retinopathy
Optic disc drusen
Radiation-induced optic neuropathy
Chronic meningitis
Optic disc congenital malformation: associated central serous retinal detachment
C: *Negative randomised control trial*
Non arteritic anterior ischaemic optic neuropathy
D: *Non visual indications*
Nerve sheath biopsy

and unpredictable effects on vascular perfusion may result in disappointing functional results.

Traumatic optic neuropathy

Projectiles and other sharp objects may injure the optic nerve directly, but typically traumatic optic neuropathy is the result of indirect force to the head, most often the forehead. The impact is thought to transmit a shock wave through bone to the optic canal. Optic nerve function is compromised sometimes after relatively minor blows to the frontal skull and in other instances in the context of major orbitocranial injury with fractures across the skull base including the optic canal. Vision may be lost instantaneously, or in a delayed progressive manner with the evolution of an intrasheath haematoma. In the case of delayed secondary visual loss, surgical decompression of the intracanicular nerve has been proposed.[31–33] Extracranial and intracranial approaches have been described, the latter including open and endoscopic ethmoidectomy techniques, as well as a bifrontal anterior skull-base approach. Medical measures are also available in the form of high- and mega-dose corticosteroid therapy. The rationale is that steroids may reduce post-traumatic oedema, contusion necrosis, and vasospasm and aid functional recovery in acute spinal cord trauma, and therefore these effects might be extrapolated to patients with traumatic optic neuropathy.[34] The International Optic Nerve Trauma Study (1999) has provided some welcome guidance.[35]

In this prospective observational study, visual outcomes were compared in patients following observation alone, high-dose steroids given within 7 days of the injury, and optic canal decompression with or without corticosteroids and performed within 7 days of the injury. As expected, an initial acuity of NPL predicted a poor outcome in all groups. No clear benefit was demonstrated for patients undergoing high-dose steroid therapy, or canal decompression surgery compared to observation alone. Importantly, 57% of the untreated group showed three lines or more acuity improvement emphasising a spontaneous remission rate that had not been widely appreciated before the study.

Optic canal decompression surgery may therefore only have a limited role in the management of traumatic optic neuropathy. In practice, unconscious patients and patients with associated injuries to the globe cannot be operated upon and any intervention on the canal is hard to justify without clinically proven progressive visual deterioration in an otherwise fit patient following the injury.

Fibrous dysplasia and other primary bone disease

Patients with polyostotic forms of fibrous dysplasia and with fibrous dysplasia associated with the McCune–Albright syndrome often have sphenoid bone involvement. This results in encasement of the optic canal with abnormal bone and narrowing of the optic canal. A proportion of patients develop visual loss and prophylactic canal decompression by neurosurgical unroofing has been advocated. This surgery may, however, fail to arrest progressive visual loss and is sometimes associated with acute deterioration in function.[36–38] In a cross-sectional survey of unselected patients with sphenoid and optic canal involvement, Lee *et al*[39] found no correlation between visual loss and canal stenosis on quantitative radiography.[39] Typically, the mechanisms for the optic neuropathy in fibrous dysplasia patients include acute compression from a secondary mass lesion such as a bone cyst with or without haemorrhage, sphenoid–ethmoid mucocoele, or sarcomatous degeneration. Chronic optic nerve traction may also play a role in some cases. In a recent review, Michael *et al*[40] reviewed the literature carefully to reach the same conclusion. Prophylactic canal decompression alone may not be a rational approach to these patients and surgical intervention is best reserved for mass lesions causing acute and subacute loss of function.

Other bone diseases in which visual loss does apparently arise from canal stenosis include renal osteodystrophy, osteopetrosis, and extramedullary haemopoiesis. Canal decompression is usually combined with much more extensive skull-base resections of abnormal bone in these cases. Craniostenoses including diaphyseal dysplasia are likely to also cause

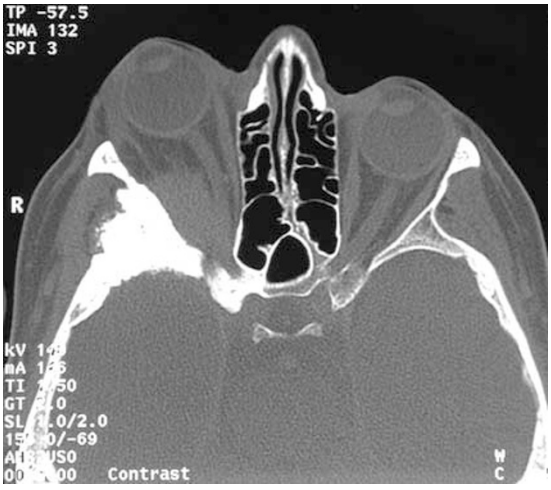

Figure 4 Bone-window orbital CT illustrating displacement and compression of the posterior intraorbital and intracannalicular segments of the optic nerve by a sphenoid meningioma with a significant intraosseous component. Indirect decompression and visual improvement may be achieved via an endoscopic transethmoidal route.

papilloedema due to raised intracranial pressure that will complicate the clinical evaluation.[41–46]

Neoplastic and other compressive lesions

Visual loss caused by extrinsic compression of the intracranial and intraorbital segments of the optic nerve by tumours is likely to respond favourably to well-timed surgery. However, the situation is more complicated when the intracanicular segment is involved. An incomplete list of possible pathologies includes:

- Primary sinus disease (mucocoele, primary sinus carcinoma, invasive aspergillosis).
- Intradural and extradural metastases.
- Invasive skull base tumours (chondrosarcoma, prolactinoma).
- Primary tumours of optic nerve and meninges (glioma, meningioma, and haemangioma).

Since the canal can effectively be approached by transethmoidal, transcranial, and skull-base routes, careful multidisciplinary assessment is required to determine which surgical team or teams can deal most effectively with both the intracanicular compression and major adjacent pathology most effectively. In our unit, truly isolated intracanicular lesions are most likely to be approached via the extradural skull base route. Indirect decompression may also be appropriate: endoscopic transethmoidal decompression of the posterior intraorbital and intracanicular segments can usefully be achieved for otherwise inoperable sphenoid meningiomas impinging on the optic nerve from the superolateral aspect (Figure 4).[47–50]

Table 2 Optic canal decompression: evidence for efficacy

A: *Supportive descriptive studies*	
	Compression due to extrinsic tumour (arising from paranasal sinuses, or intracranially)
	Secondary mass lesions in fibrous dysplasia
B: *Anecdotal/ single case reports*	
	Isolated intracanalicular meningioma
	Traumatic intrasheath haematoma
C: *Negative descriptive studies*	
	Traumatic optic neuropathy
	Canal stenosis due to Fibrous dysplasia

In conclusion, the optic canal may be decompressed as part of the treatment of an extrinsic compressing tumour (Table 2). In the case of fibrous dysplasia, surgery is usually most appropriate for secondary mass lesions and in traumatic optic neuropathy for delayed secondary visual loss in an otherwise fit patient.

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