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## Optic Nerve Decompression for Pseudotumor Cerebri

<b>Type</b>	Technology Assessment
<b>Category</b>	Surgical
<b>Sub-Category</b>	Ophthalmology
<b>Number</b>	2006T0191C
<b>Approved By</b>	<b>Approval Date</b>
Medical Technology Assessment Committee	6/15/2006

### Description

**After evaluating relevant benefit document language (exclusions or limitations), refer to Coverage sections of this document to determine coverage.**

This policy describes a surgical procedure, optic nerve decompression, used to relieve pressure on the optic nerve.

### Search Terms

Intracranial hypertension, optic nerve sheath decompression

### Background

Pseudotumor cerebri, or idiopathic intracranial hypertension, is a condition characterized by increased intracranial pressure, normal ventricular imaging, no evidence of intracranial tumor or lesion, and normal cerebrospinal fluid (CSF) composition (without infection).<sup>1</sup> Usually the etiology is unknown; however, the syndrome may encompass several causative processes. In children, pseudotumor cerebri may follow acute corticosteroid withdrawal, or vitamin A or tetracycline toxicity. In adults, the disorder is more common in women aged 30-50, particularly in those who are obese. Papilledema (swelling of the optic nerve head caused by increased intracranial pressure) may progress to partial or complete monocular vision loss in a small percentage of cases.

Medical therapy and weight loss are successful treatments in most patients. Visual fields should be monitored. In rare cases, visual defects or acuity may worsen despite medical treatment, and surgical treatment may be necessary. Surgical interventions to treat pseudotumor cerebri may include lumboperitoneal shunting (LPS) or optic nerve decompression or fenestration.

Optic nerve decompression involves cutting slits in the optic nerve sheath to allow cerebrospinal fluid to escape. Optic nerve decompression may be accomplished through a variety of approaches including craniotomy, extranasal transthemoidal, transorbital, transantral, and intranasal microscopic.<sup>4</sup>

## Audience

### Targeted Population

Enrollees, in all benefit plans, with pseudotumor cerebri (idiopathic intracranial hypertension).

### Coverage

**All reviewers must first identify member eligibility, any federal or state regulatory requirements and the plan benefit coverage prior to use of this policy.**

### Medicare Coverage

*Centers for Medicare & Medicaid Services (CMS):* CMS has not issued a National Coverage Determination for optic nerve decompression for pseudotumor cerebri.

### Coverage Rationale

Optic nerve decompression is proven for treatment of pseudotumor cerebri.

## Clinical Information

## Clinical Recommendations

**Note:** This section provides detailed information about the clinical intended use for the treatment that is the topic of this Technology Assessment. The detailed information provided in this section is NOT used to decide whether or not a service is paid for. Rather, it provides background information and rationale about the scientifically appropriate use of the treatment, for discussion purposes with providers. See "Coverage" section to determine what procedure(s) are covered/non-covered (i.e., paid for where such benefits are available).

Clinical evidence supports the use of optic nerve decompression for treatment of pseudotumor cerebri.

Clinical indications for optic nerve decompression include:

- 1) patient has worsening visual symptoms, AND
- 2) patient is unresponsive to medical management.

## Clinical Precautions

Complications of optic nerve decompression include failure, transient blindness, and ischemic optic neuropathy.<sup>1</sup>

## Setting(s)

**Outpatient**

## Research Evidence

Pseudotumor cerebri is usually managed medically, but when visual acuity continues to deteriorate despite medical treatment, surgical intervention with optic nerve fenestration (decompression) may be indicated.

A study by Villain et al. evaluated 5 patients who had severe visual field loss despite medical treatment.<sup>6</sup> At an average follow-up of 16 months, 4 patients showed significant improvement, and one showed improvement, but not significant in degree. The authors concluded that on the basis of this very small sample, optic nerve decompression is a safe therapeutic procedure for patients with severe visual loss due to elevated intracranial pressure and optic neuropathy.

Another study examined 14 patients (23 eyes) at a mean interval of 62 months after surgery.<sup>7</sup> Improvement or stabilization was noted in 17 eyes. Six eyes showed a recurrence of papilledema without functional worsening of symptoms. Three eyes showed optic atrophy and extensive visual loss. The authors believe that optic nerve fenestration can prevent further visual loss in most patients with pseudotumor cerebri, unless the eye has already become nearly blind.

Goh, et al. retrospectively reviewed the records of 29 eyes of patients who underwent optic nerve sheath fenestration (ONSF).<sup>8</sup> The patients had extensive visual loss despite medical therapy. Visual fields were compared pre- and postoperatively (within 1 and 6 months), and during a mean follow-

up period of 15.7 months. Visual acuity improved in 4 eyes (14%), did not change in 22 eyes (76%), and worsened in 3 eyes (10%). Visual fields improved in 10 eyes (48%), were unchanged in 8 eyes (38%), and worsened in 3 eyes (14%). Six eyes were lost to follow-up. Four eyes had repeated surgery with vision lost in one eye. The authors concluded that ONSF improved or protected visual function in patients with deteriorating vision despite medical intervention.

Minimal data is available on the pediatric population. Lee, et al. reported on 2 children (under age 16 years) who underwent optic nerve sheath decompression.<sup>5</sup> Optic disc edema resolved in both patients postoperatively. Both children had visual improvement in one eye each. One child had no changes in the other eye, and the other child had worsening of visual acuity in the other eye. The authors also reviewed the available literature and found reports of 12 children treated with optic nerve decompression for pseudotumor cerebri. Of these children, 66% experienced improvement in visual acuity following optic nerve decompression, 33% had improvement in visual field, and 17% experienced worsening of visual acuity and/or visual field. The authors concluded that optic nerve decompression is relatively safe and effective in adults, but accompanied by risk for significant complications. They admitted that the experience with this procedure in children is limited and based on the retrospective data suggest that the safety and efficacy of the procedure may be similar in children and adults.

Published peer reviewed medical literature was reviewed on optic nerve decompression for pseudotumor cerebri and very few relevant new studies were identified that were published after 2000. A retrospective chart review conducted by Thuente and Buckley evaluated 12 children (17 eyes) younger than 16 years of age who underwent optic nerve decompression for pseudotumor cerebri.<sup>2</sup> All patients showed improvement in optic nerve edema, and visual acuity improved or remained the same in all surgical eyes.

Banta and Farris reported on a retrospective case series of 158 eyes in 86 patients with pseudotumor cerebri who underwent optic nerve decompression.<sup>3</sup> Visual fields improved or stabilized in 88% of the surgical eyes. One patient had permanent severe visual loss in one eye secondary to an operative complication. The investigators concluded that optic nerve decompression is safe and effective in most patients.

Given the rarity of the condition, the urgency of treatment when medical therapy is unsuccessful, and the serious consequences of the disorder, prospective randomized trials are difficult to design and conduct. At this time, the limited literature seems to indicate that optic nerve decompression has some efficacy in the treatment of pseudotumor cerebri with worsening visual symptoms despite medical therapy. The risks and benefits of the surgery should be considered with each individual patient's circumstance.

## Definitions

<b>CSF</b>	Cerebrospinal fluid.
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<b>LPS</b>	Lumboperitoneal shunt; a surgical procedure designed to relieve increased intracranial pressure, and in the case of pseudotumor cerebri, to provide relief from visual symptoms caused by pressure on the optic nerve.
<b>ONSF</b>	Optic nerve sheath fenestration; a surgical procedure designed to relieve pressure on the optic nerve.

## References and Resources

### References

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5. Lee, A., Patrinely, J., and Edmond, J. Optic nerve sheath decompression in pediatric pseudotumor cerebri. *Ophthalmic Surg Lasers.* 1998(June);29(6): 514-517.
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## History/Updates

<b>6/15/2006</b>	Policy update with coverage rationale reworded to conform to proven language. CMS information updated. Policy 2000T0191B archived.
<b>10/28/2004</b>	CPT codes 31294 and 67570 added to Coding Section per direction from the Reimbursement Medical Policy Operations Manager.
<b>2/28/2002</b>	Policy Reformatted

## Coding

The Current Procedural Terminology (CPT) codes and HCPCS codes listed in this policy are for reference purposes only. Listing of a service code in this policy does not imply that the service described by this code is a covered or non-covered health service. Coverage is determined by the benefit document.

### CPT Code Section

31294	Nasal/sinus endoscopy, surgical; with optic nerve decompression
67570	Optic nerve decompression (eg, incision or fenestration of optic nerve sheath)