Clinical Policy: Optic Nerve Decompression Surgery

Reference Number: PA.CP.MP.128
Effective Date: 01/18
Date of Last Review: 02/21/2023

Coding Implications
Revision Log

Description
Optic nerve (ON) sheath decompression involves direct decompression (fenestration) of the ON sheaths just behind the globe. The approach and technique for an ON sheath fenestration varies. This policy describes the medical necessity requirements for ON decompression surgery.

Policy/Criteria

I. It is the policy of Pennsylvania Health and Wellness® (PHW) that ON sheath decompression surgery is **medically necessary** for treatment of the following indications:

A. Papilledema accompanying idiopathic intracranial hypertension (IIH) for either of the following:
   1. Visual function that is severely impaired or continues to deteriorate, despite aggressive medical management (e.g., Diamox [acetazolamide], furosemide, and corticosteroids); or
   2. Incapacitating headaches;

B. Traumatic optic neuropathy (TON) with radiologic evidence of any of the following:
   1. Optic canal fracture with impingement of the ON by a fracture fragment;
   2. Intraneural edema;
   3. Sheath hematoma;

C. Facial fibrous dysplasia, and either of the following:
   1. Cystic degenerations and optic canal narrowing if intent is prophylactic, risk of ON damage is clearly explained;
   2. Vision loss.

II. It is the policy of PHW that there is insufficient evidence in the published peer-reviewed literature to support the use of ON sheath decompression surgery for the treatment of nonarteritic anterior ischemic optic neuropathy (NAION).

Background
ON sheath decompression surgery is typically performed in instances of papilledema due to idiopathic intracranial hypertension (IIH), in which the main symptom is rapid and/or progressive vision loss rather than headache. The effect is normally limited to the ipsilateral ON, although occasionally the procedure appears to have a filtration effect, resulting in improvements in headaches and contralateral disc edema as well.

Idiopathic Intracranial Hypertension
Idiopathic intracranial hypertension (IIH), also known as pseudotumor cerebri, is a disorder defined by clinical criteria that include symptoms and signs isolated to those produced by increased intracranial pressure (e.g., headache, papilledema, vision loss), elevated intracranial pressure with normal cerebrospinal fluid composition, and no other cause of intracranial hypertension evident on neuroimaging or other evaluations.17 The incidence of IIH in the general population is thought to be about 1-2 per 100,000. In obese, young females between the ages of 15-44, the incidence of IIH is higher (4-21 per 100,000). IIH occurs in men and children

as well, but with substantially lower frequency. Weight is a risk factor for men but is less prevalent than in women and is not usually a factor in prepubertal children.20 Many individuals suffer from intractable, disabling headaches, and there is a risk of severe, permanent vision loss. Recommendations for the treatment of IIH are limited due to a lack of randomized controlled trials. In addition, the natural history of untreated IIH is uncertain.

The goals of treatment are to detect and prevent vision loss, reduce intracranial pressure, and relieve headache. Medical treatment consists of first line treatment with Diamox (acetazolamide), which inhibits choroid plexus carbonic anhydrase and reduces cerebrospinal fluid production by 50 to 60%. Furosemide (Lasix®) and corticosteroids can be added. Surgery is reserved for patients whose visual function is severely impaired or continues to deteriorate despite aggressive medical management. Those who suffer incapacitating headaches may also be candidates for surgery.

Two main surgical options include ON sheath decompression and cerebrospinal fluid (CSF) shunting. The overall rate of visual improvement seems to be equivalent across both surgical treatment modalities and an individualized approach is recommended when choosing a surgical procedure.20 In one of the largest case studies, ON sheath decompression stabilized or improved visual acuity in 94 % of patients and visual fields in 88% of patients. Visual function is greatly improved in patients with acute rather than chronic papilledema. Thus, in patients with significant visual loss, waiting a prolonged period for a response to medical therapy may not be warranted. ON sheath decompression also may improve visual function in patients with progressive visual loss despite a functioning shunt.

Traumatic Optic Neuropathy
Traumatic optic neuropathy (TON) is an important cause of severe visual loss following blunt or penetrating head trauma. Following the initial insult, ON swelling within the ON canal or compression by bone fragments are thought to result in secondary retinal ganglion cell loss. ON decompression with steroids or surgical interventions, or both, have been advocated to improve visual prognosis in TON.

A 2013 Cochrane Review of surgical treatment for TON concluded there is not enough evidence that surgical decompression of the ON provides any additional benefit beyond conservative management, citing a lack of randomized controlled trials (RCTs), and a wide range of surgical techniques that make comparisons difficult.10 Given that it would be quite difficult to conduct an adequately powered RCT of surgical ON decompression for TON, the authors' state ON decompression for TON should be assessed on a case by case basis, taking risks of surgery into consideration.10 A 2015 review of TON investigation and management included 14 articles regarding treatment for TON.1 The authors noted that studies investigating ON decompression for TON are largely small and retrospective, with one larger study- the International Optic Nerve Trauma Study- comprised of 133 patients. Across the studies reviewed, improvement after ON decompression ranged from 27 to 82%, potentially reflecting the poorly defined indications for surgery. The authors argue that surgery should be reserved for instances in which "there is radiological evidence of optic canal fracture (and impingement of ON by fracture fragment), intraneural edema or an ON sheath hematoma." 1

Facial Fibrous Dysplasia
Fibrous dysplasia (FD) is a rare condition involving non-malignant overgrowth of bone; approximately 20% of FD cases involve craniofacial bones. Surgery has been the primary form of management of compression of the optic nerve due to FD, although there is no clear agreement on timing of surgery or in which circumstances the surgery is most beneficial. 6 McCune-Albright syndrome (MAS) is a very rare condition that accounts for about 3% of all FD cases and presents as polyostotic FD (involving multiple bones/foci of disease), café-au-lait skin macules, and precocious puberty.2 Studies have shown that narrowing of the optic canal in MAS is not directly correlated with vision loss, and that acute visual loss is related to aneurysmal bone cysts and mucoceles.2 However, ideal operative management of craniofacial dysplasia in MAS has not been established due to its rarity. Due to the risks of postoperative complications, which occur in 50% of patients, prophylactic surgery to prevent vision loss is only indicated in cases with aneurysmal bone cysts and mucoceles.2 Otherwise, surgery to decompress the ON is reserved for cases of FD with established vision loss.2

Nonarteritic Anterior Ischemic Optic Neuropathy
NAION is the most common form of ischemic optic neuropathy. It is an idiopathic, ischemic insult of the ON head characterized by acute, monocular, painless visual loss with optic disc swelling.18 Visual function can be impaired through decreased central visual acuity or peripheral field loss, or both. The typical presentation is sudden onset of painless monocular vision loss, often upon awakening.

ON sheath decompression surgery was reported in 1989 to be of benefit to patients with NAION. The presumed mechanism of action in ON decompression surgery revolved around restoration of impaired blood flow to the ON through reduction of the pressure around the nerve. Initial results of uncontrolled studies suggested that ON sheath decompression was a promising treatment of progressive visual loss in patients with NAION. Other investigators who evaluated this surgical procedure reported varying degrees of success. To resolve the controversy over the effectiveness of ON decompression for NAION, the National Eye Institute sponsored the Ischemic Optic Neuropathy Decompression Trial, a multicenter, randomized controlled clinical trial of ON decompression surgery for patients with NAION.5,8 The study found no benefit from surgery in NAION patients with progressive visual loss; in fact, significantly more patients in the surgery group had progressive loss of vision than patients who received only careful follow-up. The investigators concluded that ON decompression surgery is not an effective treatment for NAION and, in fact, may increase the risk of progressive visual loss in NAION patients. The trial was stopped early because the surgery was not helping the participants more than careful follow-up alone. Pain and double vision were harms experienced by some participants in the surgery group at one week after the surgery. The trial investigators reported that continued enrollment would be unlikely to produce results in favor of surgery.

Coding Implications
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informational purposes only. Inclusion or exclusion of any codes does not guarantee coverage. Providers should reference the most up-to-date sources of professional coding guidance prior to the submission of claims for reimbursement of covered services.

CPT® Codes	Description
67570	Decompression ON (e.g., incision or fenestration of optic nerve sheath).

HCPCS Codes	Description
N/A	

ICD-10-CM Diagnosis Codes that Support Coverage Criteria

ICD-10-CM Code	Description
G93.2	Benign intracranial hypertension
H47.021	Hemorrhage in ON sheath, right eye
H47.022	Hemorrhage in ON sheath, left eye
H47.11	Papilledema associated with increased intracranial pressure
M85.08	Fibrous dysplasia (monostotic), other site
M85.09	Fibrous dysplasia (monostotic), multiple sites
Q78.1	Polyostotic fibrous dysplasia
S04.011^{+}-S04.019^{+}	Injury of ON

Reviews, Revisions, and Approvals	Revision Date	Approval Date
References reviewed and updated.	09/18	10/18
References reviewed and updated. Specialist review.	12/19	
References reviewed and updated. Specialist review.	06/2021	
Annual review. Revised language in II from "investigational" to "insufficient evidence to support…" Changed "review date" in the header to "date of last revision" and "date" in the revision log header to "revision date." Replaced member with member/enrollee. Background updated with no clinical significance. References reviewed, updated and reformatted. Specialist review.	2/21/2023	

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