Clinical Policy: Optic Nerve Decompression Surgery

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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 ON sheath decompression surgery is **investigational** for the treatment of non-arteritic 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
IIH, also known as pseudotumor cerebri, is a disorder defined by clinical criteria which 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 per 100,000. In obese, young females the incidence of IIH is about 20 per 100,000. IIH occurs in men and children as well, but with substantially lower frequency. Weight is not usually a factor in men and in children under 10 years of age. Many individuals suffer from intractable, disabling headaches, and there is a risk of severe, permanent vision loss. Individuals with mild vision loss have an associated reduction in quality of life. 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, to reduce the intracranial pressure, and to 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.

Surgical options include ON sheath decompression and lumboperitoneal shunting. However, prevailing opinion seems to favor the former. This procedure has been found to be highly effective for relief of papilledema. In fact, following a unilateral procedure, most patients have improvement in bilateral disc swelling and in severity of headache. Stabilization or improvement of vision occurs in an estimated 85 to 100% 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 functioning lumboperitoneal shunts.

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 Dypslasia
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. 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
This clinical policy references Current Procedural Terminology (CPT®). CPT® is a registered trademark of the American Medical Association. All CPT codes and descriptions are copyrighted

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	Date	Approval Date
References reviewed and updated.	09/18	10/18

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