

Clinical Policy: Optic Nerve Decompression Surgery

Reference Number: LA.CP.MP.128

Date of Last Revision: 02/24

[Coding Implications](#)
[Revision Log](#)

See [Important Reminder](#) at the end of this policy for important regulatory and legal information.

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 Louisiana Healthcare Connections that ON sheath decompression surgery is **medically necessary** for treatment of the following conditions:
 - A. Papilledema accompanying idiopathic intracranial hypertension (IIH), cerebral venous sinus thrombosis, or intracranial tumors causing intracranial pressure elevation from decreased cerebrospinal fluid outflow with either of the following:
 1. Visual function that is severely impaired or continues to deteriorate, despite aggressive medical management (e.g., Diamox, furosemide, and corticosteroids);
 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. Intra-neural 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 Louisiana Healthcare Connections 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), cerebral venous sinus thrombosis, or intracranial tumors causing intracranial pressure elevation from decreased cerebrospinal fluid outflow 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. Optic nerve sheath fenestration (ONSF) has been shown to be effective for the treatment of papilledema due to cerebral venous thrombosis (CVT) after the failure of medical treatment, including anti-coagulation medication.

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 mucocoeles.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 mucocoeles.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

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 2022, American Medical Association. All rights reserved. CPT codes and CPT descriptions are from the current manuals and those included herein are not intended to be all-inclusive and are included for informational purposes only. Codes referenced in this clinical policy are for informational purposes only and may not support medical necessity. 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	Optic nerve decompression (eg, incision or fenestration of optic nerve sheath)

ICD-10-CM Diagnosis Codes that Support Coverage Criteria
+ Indicates a code requiring an additional character

ICD-10-CM Code	Description
G93.2	Benign intracranial hypertension
H47.021	Hemorrhage in optic nerve sheath, right eye
H47.022	Hemorrhage in optic nerve sheath, left eye
H47.11	Papilledema associated with increased intracranial pressure
I67.6	Nonpyogenic thrombosis of intracranial venous system
M85.08	Fibrous dysplasia (monostotic), other site
M85.09	Fibrous dysplasia (monostotic), multiple sites
Q78.1	Polyostotic fibrous dysplasia
S04.011A	Injury of optic nerve, right eye, initial encounter
S04.011D	Injury of optic nerve, right eye, subsequent encounter
S04.011S	Injury of optic nerve, right eye, sequela
S04.112A	Injury of optic nerve, left eye, initial encounter
S04.112D	Injury of optic nerve, left eye, subsequent encounter
S04.112S	Injury of optic nerve, left eye, sequela

Reviews, Revisions, and Approvals	Revision Date	Approval Date
Converted corporate to local policy.	08/15/2020	
Revised language in II from “investigational” to “insufficient evidence to support...” References reviewed, updated and reformatted. 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.	1/22	1/22
Annual review. References reviewed and updated. Specialist review. Background updated with no clinical significance.	9/22	11/28/22
Annual Review; Added nonpyogenic thrombosis of intracranial venous system as a medical necessity indication; Removed diagnosis code H47.019 due to unspecified eye; Updated References.	02/24	4/26/24

References

1. Kumaran AM, Sundar G, Chye LT. Traumatic Optic Neuropathy: A Review. *Craniofacial Trauma Reconstr.* 2015;8(1): 31–41. doi:10.1055/s-0034-1393734
2. Belsuzarri TA, Araujo JF, Melro CA, et al. McCune-Albright syndrome with craniofacial dysplasia: clinical review and surgical management. *Surg Neurol Int.* 2016;7 (Suppl 6): S165-S169. doi: 10.4103/2152-7806.178567.
3. Cohen AJ. Optic nerve sheath fenestration. Medscape. <https://emedicine.medscape.com/article/1891241-overview#showall>. Published November 2, 2021. Accessed June 13, 2022.
4. Dickersin K, Li T. Surgery for nonarteritic anterior ischemic optic neuropathy. *Cochrane Database Syst Rev.* 2015;2015(3):CD001538. Published 2015 Mar 12. doi:10.1002/14651858.CD001538.pub4
5. Levin LA, Beck RW, Joseph MP, et al. The treatment of traumatic optic neuropathy: the International Optic Nerve Trauma Study. *Ophthalmology.* 1999;106(7):1268–1277. doi:10.1016/s0161-6420(99)00707-1
6. Li H, Zhou B, Shi J, Cheng L, Wen W, Xu G. Treatment of traumatic optic neuropathy: our experience of endoscopic optic nerve decompression. *J Laryngol Otol.* 2008;122(12):1325-1329. doi:10.1017/S0022215108002296
7. Lu Y, Yang J, Wu Y, Pan S, Lu J, Mu X. "Well Digging" Subcraniotomy Strategy with Navigation for Optic Nerve Decompression in Frontoorbital Fibrous Dysplasia: Preliminary Experience. *Plast Reconstr Surg Glob Open.* 2016;4(11):e1080. Published 2016 Nov 8. doi:10.1097/GOX.0000000000001080
8. Optic nerve decompression surgery for nonarteritic anterior ischemic optic neuropathy (NAION) is not effective and may be harmful. The Ischemic Optic Neuropathy Decompression Trial Research Group. *JAMA.* 1995;273(8):625-632.
9. Ropposch T, Steger B, Meço C, et al. The effect of steroids in combination with optic nerve decompression surgery in traumatic optic neuropathy. *Laryngoscope.* 2013;123(5):1082-1086. doi:10.1002/lary.23845
10. Sosin M, De La Cruz C, Mundinger GS, et al. Treatment Outcomes following Traumatic Optic Neuropathy. *Plast Reconstr Surg.* 2016;137(1):231-238. doi:10.1097/PRS.0000000000001907
11. Spoor TC, Ramocki JM, Madion MP, Wilkinson MJ. Treatment of pseudotumor cerebri by primary and secondary optic nerve sheath decompression. *Am J Ophthalmol.* 1991;112(2):177-185. doi:10.1016/s0002-9394(14)76698-x
12. Welkoborsky HJ, Möbius H, Bauer L, Wiechens B. Traumatische Optikusneuropathie. Langzeitergebnisse nach endonasaler mikrochirurgischer Dekompression des N. opticus [Traumatic optic nerve neuropathy. Longterm results following microsurgical optic nerve decompression]. *HNO.* 2011;59(10):997-1004. doi:10.1007/s00106-011-2266-3
13. Yang QT, Zhang GH, Liu X, Ye J, Li Y. The therapeutic efficacy of endoscopic optic nerve decompression and its effects on the prognoses of 96 cases of traumatic optic neuropathy. *J Trauma Acute Care Surg.* 2012;72(5):1350-1355. doi:10.1097/TA.0b013e3182493c70
14. Yu-Wai-Man P, Griffiths PG. Surgery for traumatic optic neuropathy. *Cochrane Database Syst Rev.* 2013;6(6):CD005024. Published 2013 Jun 18. doi:10.1002/14651858.CD005024.pub3

15. Zhang Q, Lu H, Li G, et al. Long-term efficacy of nasal endoscopic ON decompression for traumatic optic neuropathy. *Lin Chung Er Bi Yan Hou Tou Jing Wai Ke Za Zhi*. 2015;29(12):1082-1085.
16. Zhilin G, Huoni O, Zhihua C, Guorong D. Wide optic nerve canal decompression for the treatment of blindness resulting from an indirect optic nerve injury. *J Craniofac Surg*. 2011;22(4):1463-1465. doi:10.1097/SCS.0b013e31821d184a
17. Wall M, Lee AG. Idiopathic intracranial hypertension (pseudotumor cerebri): Prognosis and treatment. UpToDate. www.uptodate.com. Published September 30, 2021. Accessed June 7, 2022.
18. Tamhankar M, Volpe NJ. Nonarteritic anterior ischemic optic neuropathy: Prognosis and treatment. UpToDate. www.uptodate.com. Published September 22, 2021. Accessed June 7, 2022.
19. Tamhankar M, Volpe NJ. Nonarteritic anterior ischemic optic neuropathy: Clinical features and diagnosis. UpToDate. www.uptodate.com. Published February 11, 2020. Accessed June 7, 2022.
20. Wall M, Lee AG. Idiopathic intracranial hypertension (pseudotumor cerebri): Epidemiology and pathogenesis. UpToDate. www.uptodate.com. Published February 11, 2020. Accessed June 8, 2022.
21. Adesina O, Patel BC. Optic Nerve Decompression. [Updated 2022 Aug 28]. In: StatPearls [Internet]. Treasure Island (FL): StatPearls Publishing; 2022 Jan-. Available from: <https://www.ncbi.nlm.nih.gov/books/NBK538300/>
22. Murdock J, Tzu JH, Schatz NJ, Lee WW. Optic nerve sheath fenestration for the treatment of papilledema secondary to cerebral venous thrombosis. *J Neuroophthalmol*. 2014;34(1):67–69.

Important Reminder

This clinical policy has been developed by appropriately experienced and licensed health care professionals based on a review and consideration of currently available generally accepted standards of medical practice; peer-reviewed medical literature; government agency/program approval status; evidence-based guidelines and positions of leading national health professional organizations; views of physicians practicing in relevant clinical areas affected by this clinical policy; and other available clinical information. LHCC makes no representations and accepts no liability with respect to the content of any external information used or relied upon in developing this clinical policy. This clinical policy is consistent with standards of medical practice current at the time that this clinical policy was approved.

The purpose of this clinical policy is to provide a guide to medical necessity, which is a component of the guidelines used to assist in making coverage decisions and administering benefits. It does not constitute a contract or guarantee regarding payment or results. Coverage decisions and the administration of benefits are subject to all terms, conditions, exclusions and limitations of the coverage documents (e.g., evidence of coverage, certificate of coverage, policy, contract of insurance, etc.), as well as to state and federal requirements and applicable LHCC administrative policies and procedures.

This clinical policy is effective as of the date determined by LHCC. The date of posting may not be the effective date of this clinical policy. This clinical policy may be subject to applicable legal

and regulatory requirements relating to provider notification. If there is a discrepancy between the effective date of this clinical policy and any applicable legal or regulatory requirement, the requirements of law and regulation shall govern. LHCC retains the right to change, amend or withdraw this clinical policy, and additional clinical policies may be developed and adopted as needed, at any time.

This clinical policy does not constitute medical advice, medical treatment or medical care. It is not intended to dictate to providers how to practice medicine. Providers are expected to exercise professional medical judgment in providing the most appropriate care, and are solely responsible for the medical advice and treatment of member/enrollees. This clinical policy is not intended to recommend treatment for member/enrollees. Member/enrollees should consult with their treating physician in connection with diagnosis and treatment decisions.

Providers referred to in this clinical policy are independent contractors who exercise independent judgment and over whom LHCC has no control or right of control. Providers are not agents or employees of LHCC.

This clinical policy is the property of LHCC. Unauthorized copying, use, and distribution of this clinical policy or any information contained herein are strictly prohibited. Providers, member/enrollees and their representatives are bound to the terms and conditions expressed herein through the terms of their contracts. Where no such contract exists, providers, member/enrollees and their representatives agree to be bound by such terms and conditions by providing services to member/enrollees and/or submitting claims for payment for such services.

©2024 Louisiana Healthcare Connections. All rights reserved. All materials are exclusively owned by Louisiana Healthcare Connections and are protected by United States copyright law and international copyright law. No part of this publication may be reproduced, copied, modified, distributed, displayed, stored in a retrieval system, transmitted in any form or by any means, or otherwise published without the prior written permission of Louisiana Healthcare Connections. You may not alter or remove any trademark, copyright or other notice contained herein. Louisiana Healthcare Connections is a registered trademarks exclusively owned by Louisiana Healthcare Connections.