

# Clinical Policy: Optic Nerve Decompression Surgery

Reference Number: CP.MP.128

Last Review Date: 08/20

[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 health plans affiliated with Centene Corporation® that ON sheath decompression surgery is **medically necessary** for treatment of the following conditions:
  - A. Papilledema accompanying idiopathic intracranial hypertension (IIH) 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); 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. 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 health plans affiliated with Centene Corporation 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 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.<sup>17</sup> 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

## CLINICAL POLICY

### Optic Nerve Decompression Surgery

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<sup>®</sup>) 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.<sup>10</sup> 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.<sup>10</sup> A 2015 review of TON investigation and management included 14 articles regarding treatment for TON.<sup>1</sup> 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."<sup>1</sup>

## CLINICAL POLICY

### Optic Nerve Decompression Surgery

#### *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.<sup>6</sup> 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.<sup>2</sup> 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.<sup>2</sup> 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.<sup>2</sup> Otherwise, surgery to decompress the ON is reserved for cases of FD with established vision loss.<sup>2</sup>

#### *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.<sup>18</sup> 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.<sup>5,8</sup> 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 2020, 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

## CLINICAL POLICY

### Optic Nerve Decompression Surgery

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)

### 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 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 <sup>+</sup> - S04.019 <sup>+</sup>	Injury of ON

Reviews, Revisions, and Approvals	Date	Approval Date
Policy adopted from Health Net NMP353 ON Decompression Surgery	08/16	09/16
Reclassified TON as medically necessary with certain criteria; added facial fibrous dysplasia as a medically necessary indication, and updated related background information and codes.	09/17	09/17
References reviewed and updated.	08/18	08/18
References reviewed and updated. Specialist review.	07/19	08/19
References reviewed and updated.	07/20	08/20

### References

- Kumaran AM, Sundar G, Chye LT. Traumatic Optic Neuropathy: A Review. *Craniomaxillofac Trauma Reconstr.* 2015 Mar; 8(1): 31–41. doi: 10.1055/s-0034-1393734
- Belsuzarri TA, Araujo JF, Melro CA, et al. McCune-Albright syndrome with craniofacial dysplasia: clinical review and surgical management. *Surg Neurol Int.* 2016 Mar 11;7 (Suppl 6): S165-9. doi: 10.4103/2152-7806.178567. eCollection 2016.
- Cohen AJ. ON Sheath Fenestration. *Medscape.* July 2016
- Dickersin K, Li T. Surgery to improve vision in people with nonarteritic anterior ischemic optic neuropathy (NAION). *Cochrane Review.* 12 March, 2015.
- 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.
- Li H, Zhou B, Shi J, et al. Treatment of traumatic optic neuropathy: Our experience of endoscopic ON decompression. *J Laryngol Otol.* 2008;122(12):1325-1329.
- Lu Y, Yang J, Wu Y, et al. “Well Digging” subcraniotomy strategy with navigation for optic nerve decompression in frontoorbital fibrous dysplasia: preliminary experience. *Plast Reconstr Surg Glob Open.* 2016 Nov 8;4(11):e1080. eCollection 2016 Nov.

**CLINICAL POLICY****Optic Nerve Decompression Surgery**

8. No authors listed. ON 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 ON decompression surgery in traumatic optic neuropathy. *Laryngoscope*. 2013;123(5):1082-1086.
10. Sosin M, De La Cruz C, Mundinger GS, et al. Treatment Outcomes following Traumatic Optic Neuropathy. *Plast Reconstr Surg*. 2016 Jan;137(1):231-8
11. Spoor, TC, Ramocki, JM, Madion, MP, et al. Treatment of pseudotumor cerebri by primary and secondary ON sheath decompression. *Am J Ophthalmol* 1991; 112:177.
12. Welkoborsky HJ, Möbius H, Bauer L, Wiechens B. Traumatic ON neuropathy. Long term results following microsurgical ON decompression. *HNO*. 2011 Oct; 59(10):997-1004.
13. Yang QT, Zhang GH, Liu X, Ye J, Li Y. The therapeutic efficacy of endoscopic ON decompression and its effects on the prognoses of 96 cases of traumatic optic neuropathy. *J Trauma Acute Care Surg*. 2012 May; 72(5):1350-5.
14. Yu-Wai-Man P, Griffiths PG. Surgery for the treatment of traumatic optic neuropathy Updated. *Cochrane Summaries*. June 18, 2013.
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 Jun;29(12):1082-5
16. Zhilin G, Huoni O, Zhihua C, Guorong D. Wide ON canal decompression for the treatment of blindness resulting from an indirect ON injury. *J Craniofac Surg*. 2011 Jul;22(4):1463-5.
17. Lee AG, Wall M. Idiopathic intracranial hypertension (pseudotumor cerebri): Prognosis and treatment. In:UpToDate, Brazis P (Ed). Accessed July 20, 2020
18. Tamhankar M, Volpe NJ. Nonarteritic ischemic optic neuropathy: Prognosis and treatment. In:UpToDate. Brazis P (Ed). Accessed July 20, 2020  
Tamhankar M, Volpe NJ. Nonarteritic anterior ischemic optic neuropathy: Clinical features and diagnosis. In: UpToDate. Brazis P (Ed). UpToDate, Waltham, MA. Accessed July 20, 2020

**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. The Health Plan 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. “Health Plan” means a health plan that has adopted this clinical policy and that is operated or administered, in whole or in part, by Centene Management Company, LLC, or any of such health plan’s affiliates, as applicable.

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

## CLINICAL POLICY

### Optic Nerve Decompression Surgery



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 Health Plan-level administrative policies and procedures.

This clinical policy is effective as of the date determined by the Health Plan. 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. The Health Plan 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 members. This clinical policy is not intended to recommend treatment for members. Members 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 the Health Plan has no control or right of control. Providers are not agents or employees of the Health Plan.

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

**Note: For Medicaid members**, when state Medicaid coverage provisions conflict with the coverage provisions in this clinical policy, state Medicaid coverage provisions take precedence. Please refer to the state Medicaid manual for any coverage provisions pertaining to this clinical policy.

**Note: For Medicare members**, to ensure consistency with the Medicare National Coverage Determinations (NCD) and Local Coverage Determinations (LCD), all applicable NCDs, LCDs, and Medicare Coverage Articles should be reviewed prior to applying the criteria set forth in this clinical policy. Refer to the CMS website at <http://www.cms.gov> for additional information.

©2016 Centene Corporation. All rights reserved. All materials are exclusively owned by Centene Corporation 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 Centene Corporation. You may not alter or

**CLINICAL POLICY**

Optic Nerve Decompression Surgery

remove any trademark, copyright or other notice contained herein. Centene® and Centene Corporation® are registered trademarks exclusively owned by Centene Corporation.