Contemporary diagnosis and approaches toward optic nerve decompression

K. Christopher McMains, MD, a Stilianos E. Kountakis, MD, PhD

From the aDepartment of Otolaryngology–Head and Neck Surgery, University of Texas–San Antonio, San Antonio, Texas; and the
bDepartment of Otolaryngology–Head and Neck Surgery, Medical College of Georgia, Augusta, Georgia.

Indirect traumatic blindness is often associated with closed head injuries and specifically with blows to the cheekbone and the forehead area above the eyebrow. Mainstay therapy includes corticosteroids with surgery reserved for persistent blindness and when appropriate instruments and experienced surgeons are available. Advanced endoscopic techniques offer trans-sphenoid access to the optic canal and can assist in the management of traumatic blindness in selected cases while minimizing patient morbidity.

© 2006 Elsevier Inc. All rights reserved.

KEYWORDS Traumatic blindness; Endoscopic surgery; Optic neuropathy; Optic canal; Decompression

Walsh1 defined indirect optic nerve injury as “traumatic loss of vision which occurs without external or initial ophthalmoscopic evidence of injury to the eye or its nerve.” The optic nerve is injured in 0.5% to 1.5% of cases of closed head injury and often involve the optic canal.2,3 Common injuries resulting in traumatic optic neuropathy (TON) include a blow to the ipsilateral brow or forehead, most often as a result of a motor vehicle or bicycle accident, fall, or assault.3-6 Although widely recognized and much investigated, the precise pathophysiology of and optimal treatment modalities for TON continue to be the subject of debate.

Anatomy/mechanisms of injury

The optic nerve is a fiber tract of brain white matter, with afferent fibers coursing posteriorly from the retina to the bony optic canal located at the posterosuperior aspect of the bony orbit. Posterior to that, the optic nerve, ophthalmic artery, postganglionic sympathetic fibers, and meningeal extensions are contained within the optic canal. Although the intraorbital segment of the optic nerve is permitted limited motion, the intracanalicular portion of the optic nerve is tethered within the optic canal, where it fuses with the periorbita.7 Thus, the intracanalicular nerve is at risk for both primary and secondary ischemic injury caused by shearing and swelling within the fixed cross-sectional area of the bony canal.8 Although 4% of optic nerves are dehiscent within the sphenoid sinus,9 the optic nerve may be exposed in up to 25% of cases in which an Onodi cell is present (Figure 1).10

The role of excitatory amino acids and their involvement in free radical production have been explored as another factor contributing to TON. Glutamate is an excitatory neurotransmitter present in high concentration within neurons. It is released in response to cell damage. Ultimately, this process induces nitric oxide synthase, produces free radicals, and degrades cell membranes.11 The end result of these pathways is secondary neuronal damage. The mechanism of injury in cases of intracanalicular optic nerve edema or hematoma may involve vascular ischemia and interruption of neuro-feedback mechanisms that contribute to nerve dysfunction.12

Diagnosis

TON is a clinical diagnosis and, as such, a thorough history and physical examination remain the cornerstones of diagnosis. History should comprise the mechanism of injury, including direction and relative force of the trauma, and history of progressive visual compromise. Physical examination should include complete primary and secondary surveys, as well as thorough ophthalmologic examination. Several investigators4,5,12 have shown the prognostic importance of visual acuity after injury. The most commonly
associated optic abnormality is subconjunctival hemorrhage. Patients with a lucid interval of unaffected sight before deterioration have a better prognosis than those with no light perception from the time of injury. In patients with isolated TON, the integrity of the globe is maintained. In the acute phase of posterior segment injury, the optic nerve head appears normal.

Despite normal funduscopic examinations at initial presentation, later in the course, optic atrophy may be noted. The most common findings include afferent papillary defect with normal retina and disc. The swinging flashlight test is a good examination tool for afferent papillary defect in the trauma setting; however, analgesic opioids can inhibit this test. When visual field loss develops, in 75% of patients with visual loss, the loss is limited to the lower visual fields. This limitation is caused by higher adherence of the pia and arachnoid in the upper canal. Contrasted high-resolution computerized tomography of the orbits and sphenoid sinus is critical to assess the integrity of the optic canal and identify bony impingement. Carotid-cavernous fistulae have been reported in cases of shown canal fracture. Failure to show canal fracture does not exclude TON. An intrathecal metrizamide study can provide further delineation of the intracanalicular subarachnoid space.

**Treatment**

Between 20% and 38% of patients with TON will have spontaneous recovery of vision. Corticosteroids were initially used to reduce edema and vasospasm in an effort to limit ischemic nerve cell death. The National Acute Spinal Cord Injury Studies (NASCIS I, II, and III) showed benefit to patients with spinal cord injuries who...
received high-dose corticosteroids within 8 hours of injury.\textsuperscript{21-23} Methylprednisolone is thought to be superior to dexamethasone in patients with trauma because it does not interact with anticonvulsant medications.\textsuperscript{24} The dosing regimen used in the NASCIS involved bolus delivery of 30-mg/kg body weight of methylprednisolone, followed by 5.4-mg/kg/hour infusion for 23 hours. The results of the NASCIS have been used as justification for high-dose corticosteroids for treatment of TON; however, important differences exist between the spinal cord and optic nerve. The spinal cord consists of both gray and white matter, whereas the optic nerve consists exclusively of white matter. These tissue types differ in their response to trauma.\textsuperscript{25} Clinical data remain inconclusive as to the role of corticosteroids with indirect TON.

Research suggests that glucocorticoids are neurotoxic, with concentrated effect on the hippocampal CA1 cells.\textsuperscript{26} In a rat model, methylprednisolone has caused a dose-dependent reduction in optic nerve myelin sheaths after crush injury.\textsuperscript{27} Further analysis of the NASCIS data also showed a harmful effect of high-dose corticosteroids when begun later than 8 hours after injury.\textsuperscript{28} In a recent review, the evidence supporting the use of corticosteroids in TON is described as “weak at best.”\textsuperscript{29} New interventions such as lazaroids, 21-aminosteroids that possess free-radical scavenging capabilities, may have equal or superior neuroprotective effects as corticosteroids without the attendant risks.\textsuperscript{23} Further study should delineate the role of newer therapies.

Some investigators recommend surgical intervention alone or in conjunction with corticosteroids for certain presentations of TON. Several criteria for surgical intervention have been put forward for different procedures and different clinical circumstances.\textsuperscript{8,12,30,31} Most reports show measurable improvement in vision in 31% to 82% of patients undergoing surgical intervention with or without corticosteroid therapy.\textsuperscript{12,32} Caution is advised in the repair of LeFort III fractures in patients with concomitant orbital apex or optic canal fractures because of possible secondary optic nerve damage.\textsuperscript{33} Some investigators note no relationship between the timing of trauma and surgery, while others show benefit from undertaking surgery within a specified time frame.\textsuperscript{34,35}

The benefit of intervention of any kind is not clearly established. The International Optic Nerve Trauma Study aimed to provide conclusive data regarding the best pathway for TON treatment. Unfortunately, it was discontinued secondary to insufficient enrollment. Data from 127 enrolled patients failed to show clear benefit from either corticosteroid therapy or optic nerve decompression.\textsuperscript{14} Surgical decompression should be carefully considered, given the serious potential surgical risks.

**Surgical approaches**

Dandy\textsuperscript{36} described the frontotemporal craniotomy approach to the optic nerve. This procedure became the most frequently used surgery for TON for several decades. Takahashi\textsuperscript{37} described the first endonasal approach in 1951. Niho et al\textsuperscript{38} explained a transethmoidal approach to the optic canal in cases of TON. This approach involved complete extirpation of the mucosal lining of the antrum, ethmoid, sphenoid, and frontal sinus, as well as the medial orbital wall. Kennerdell et al\textsuperscript{39} described a transantral-ethmoidal approach, which involved a Caldwell-Luc, elevation of an inferiorly based mucosal flap, and ethmoidectomy with mucosa stripping. Microscopic removal of the poste-
rior lamina papyracea, removal of the anterior sphenoid, and dissection of the canal followed.

Sofferman\(^40\) introduced the microscopic sublabial sphenoidethmoid approach, which increased the visual angle of incidence with respect to the canal. In this technique, a wider resection of bone is performed, and a posterior septal mucosal flap is preserved for reconstruction. A buttress of bone is preserved between the optic canal and internal carotid artery, while the canal is dissected inferomedially.\(^41\) Call\(^42\) introduced microscopic transorbital-sphenoid decompression, dividing and reflecting the medial canthal tendon, retracting the periorbita laterally. To prevent enophthalmos, defects were reconstructed with silicone. With advances in fiberoptic technology and instrumentation, sinus endoscopes are now almost exclusively used.

Under endoscopic vision, complete ethmoidectomy is performed. A wide sphenoidotomy allows visualization of the optic canal. The canal sits immediately superior and anterior to the carotid imprint on the lateral wall of the sphenoid sinus (Figure 2). The anterior wall of the sphenoid sinus is removed laterally until the optic tubercle is reached. This is the thick buttress of bone formed at the junction of the posterior lamina papyracea and the anterior and lateral sphenoid walls, and it marks the location of the optic nerve foramen and annulus of Zinn (Figure 3). The posterior 1 cm of the lamina papyracea is removed at the level of the optic canal, preserving the orbital periorbita, and the optic tubercle is thinned with either straight diamond burrs or a 15° dacryocystorhinostomy burr (Figure 4). Thin fragments of bone are elevated using a thin curette or an elevator, and are then removed using pediatric straight or angled biting sinus forceps. We prefer using a double-ended size 5 dental excavator to elevate thin bone fragments because it offers easy 360° manipulation within a tight space. It is important that sharp cutting burrs are avoided because of the potential for overly aggressive dissection. It is also important to provide adequate irrigation to avoid compounding thermal injury.

Once the optic tubercle is thinned and removed, drilling ensues along the optic canal, exposing the optic nerve sheath along its full length, beginning anteriorly at the annulus of Zinn and dissecting posteriorly toward the optic chiasm (Figures 5 and 6). The size 5 dental excavator has proved most useful when removing the thinned optic canal wall. We know that the optic chiasm is approached when the bone of the optic canal gets progressively thicker and thinning it becomes more labor intensive. The optic nerve sheath may be incised; however, Sofferman\(^13\) states that the benefit of sheath incision is unclear. Given the high rates of hematoma formation and potential effect of optic nerve edema, incision of this sheath should be entertained (Figure 7). A word of caution when there is a visible transverse optic canal fracture. The fracture may extend over the carotid canal (Figure 8), and, thus, manipulation of a bone fragment should be performed with extreme caution. Avoid extensive packing of the surgical site to allow maximal decompression. Patients should irrigate their nasal cavities copiously with physiologic saline postoperatively.

Results and recommendations

Kountakis et al\(^12\) treated 34 patients with TON using a protocol that allowed megadose steroids with a bolus intr-
venous delivery of 30-mg/kg body weight of methylprednisolone over 1 hour, followed by 5.4-mg/kg/hour infusion for 47 hours. Patients with worsening or not improving visual acuity after 48 hours of treatment were offered endoscopic optic nerve decompression (EOND). Of the 34 patients treated with steroids, 11 (32%) had improvement, and 23 (68%) did not. Of the 23 patients who had steroids fail, 17 elected to undergo EOND, of whom 14 (82%) had improvement, and 3 (18%) did not. Steroid treatment and EOND were superior to steroids alone (chi-square 11.338, P = 0.0007). Improvement was defined as an increase of 2 lines of Snellen visual acuity or if the patient had a 10% improvement in the visual field.

One of the interesting findings in this study was that it offers some guidance as to the benefit of megadose steroids. When the visual acuity was 20/200 or better, all patients had improvement with megadose steroids, and none of these patients elected to undergo EOND. When the visual acuity was 20/400 or worse, the majority of the patients had improvement only when EOND was added to their treatment plan (chi-square = 17.1, P = 0.00004). The data are highly statistically significant and identify a niche for megadose steroid treatment in patients with TON. When the optic nerve trauma is mild, steroids may reduce intracanalicular pressure, and control trauma-related inflammation and damage to assist in the recovery of the optic nerve. Severe optic nerve trauma most often requires surgical intervention for relief of the compartment syndrome within the optic canal. Data in this study were also analyzed to determine if optic nerve sheath fenestration was beneficial, but the numbers were too small for statistical significance.

Based on our data, EOND for indirect TON is offered if vision does not improve or if it worsens despite 48 hours of megadose steroids. The weakness of the study is that it lacked an untreated control group for obvious ethical considerations. No complications were seen in this study. Experienced rhinologists should perform EOND to avoid the serious possible consequences of drilling millimeters away from the internal carotid artery canal.

Conclusion

TON has potentially devastating consequences on the patient’s quality of life. Although high-dose corticosteroids have been widely used, results are mixed. Some basic science research suggests that steroids may have a negative effect on neuron survival. Our data indicate that steroids are beneficial in cases of mild TON, with a visual acuity of 20/200 or better. The role of surgical decompression is not clear, with a lack of controlled studies in the literature. The International Optic Nerve Trauma Study failed to resolve these issues because of slow enrollment. Our experience shows that EOND is beneficial when patients do not respond to megadose steroids for 48 hours. Ultimately, decisions regarding the type of intervention to use would consider all patient-specific factors while weighing risk versus potential benefit.

References

37. Takahashi R: Exposure of the optic canal. Operation 5:300-302, 1951