Traumatic optic neuropathy is a rare but devastating cause of partial or complete visual loss caused by deformational forces that injure the optic nerve. Injuries can be broadly classified as direct or indirect. Direct injuries usually involve direct anatomical disruption of the optic nerve caused by injuries to the head, face and orbit due to projectiles. Indirect injuries result from deforming forces applied to the bony orbit, or by motion of the globe, where the optic nerve absorbs energy concentrated at the orbital apex. These injuries are seen in the absence of open wounds but with a positive history of blunt frontal trauma. Incidence of traumatic optic neuropathy in facial trauma is 0.7 to 5 percent.

Causes of traumatic optic neuropathy can be as varied as motor injuries, falls, falling debris, assaults trivial causes such as weightlifting as well as following endoscopic sinus surgery.

Relevant Anatomy

The optic nerve is the neural conduit linking inputs received from the retina to the brain (consisting of approximately 1.2 million axons) originating from the retinal ganglion cells. The nerve also contains oligodendrocytes which provide axonal myelination; microglia which are immunocompetent phagocytic cells and modulate apoptosis; and astrocytes. The nerve is enclosed within all three meningeal coats - a perineural sheath from the pia mater which also contains the blood vessels, an intermediate sheath from the arachnoid, and an outer sheath from the dura mater, which is also connected with the peristeme as it passes through the optic foramen. Optic nerve fenestration involves incision of the outer two coats surrounding the optic nerve.

The total length is about 50 mm in length and is anatomically divided into:

- Intraocular (about 1 mm)
- Intraorbital (20-30 mm)
- Intracanalicular (5-11 mm)
- Intracranial (3-16 mm)

The optic nerve travels superomedially and passes through the annulus of Zinn to the entrance of the optic canal at the optic ring, which is located medial to the superior orbital fissure. The canal lies within the lesser wing of the sphenoid bone and is approximately 9 mm in length. The intraorbital portion of the nerve extends 18 mm from the posterior aspect of the globe to the orbital apex. The nerve measures between 20 and 30 mm, and, therefore, takes a sinuous course, allowing for a range of movements by the eye. However, as the nerve enters the optic foramen its dural sheath becomes continuous with that lining the orbit and the optic foramen, rendering it immobile. This portion of the nerve is the focus of forces encountered in head trauma and is the most common site of optic nerve injury.

The intracranial portion of the optic nerve may suffer trauma when it is displaced superiority against the sharp edge of the falciform dural fold. In addition, the anterior clinoid process lies lateral to the nerve, and, when fractured, it can crush the nerve.

Presentation, Examination and Diagnosis

Traumatic optic neuropathy is a clinical diagnosis and it usually follows head trauma with or without a history of loss of consciousness. Patients of traumatic optic neuropathy usually present with reduced visual acuity which may be as profound as only light perception or even no light perception. In patients with relatively better visual acuity on presentation, loss of colour vision is a common finding.

Check list for examining a patient with suspected Traumatic Optic Neuropathy:

- Vision Check
- Colour vision (if vision permits)
- Pupillary evaluation
- Check for globe rupture, IOFB, fracture
- Fundus examination
- Neuroimaging (in case of suspected fracture)
- VEP
- ERG

Patients who have suffered extensive head trauma must be neurologically examined and an ophthalmological examination should be performed only after stabilizing the patient. The eye and the adnexa must be examined looking specifically for globe rupture, foreign bodies and fractures. Pupillary reflexes should be evaluated. The presence of an afferent papillary defect (APD) indicates the possibility of unilateral traumatic optic neuropathy. Ocular motility and visual fields should be evaluated, as possible. Fundus examination typically reveals a disc of normal appearance unless an avulsion or an anterior optic neuropathy is present. Further more, a ring of hemorrhage at the site of injury is indicative of partial or complete avulsion of the optic nerve head. Optic atrophy ensues usually after a period of 2 weeks to 3 months. In comatose on unresponsive patients, a visual evoked potentials may be needed in diagnosing traumatic optic neuropathy, especially when pupillary evaluation is not helpful, as in bilateral traumatic optic neuropathy.

In unilateral traumatic optic neuropathy, flash VEP amplitudes that are at least 50% of the normal eye are critical for a good visual outcome. An absent VEP response indicates that visual loss is complete, and recovery of vision may be unlikely, although reports have shown that patients with persistently negative VEPs may also show visual improvement.

Address for Correspondance: Department of Neuro Ophthalmology, Sankara Nethralaya, Chennai
absent electroretinogram (ERG) is associated with a poor potential for visual recovery. A CT scan of the brain with fine cuts (axial sections of 1 to 1.5 mm) through the orbits should be sought. Coronal images are necessary to evaluate the optic canal properly and rule out a fracture. In patients with traumatic optic neuropathy, orbital fractures especially canal fractures have been associated with poorer visual acuity and a poor prognosis. MRI of the orbit may reveal focal edema of the optic nerve or optic nerve sheath enhancement with gadolinium.

Pathophysiology of traumatic optic neuropathy
It is believed that damage in traumatic optic neuropathy is caused by a primary and secondary mechanism of injury. While the optic nerve can be injured anywhere along its course, the most common site of injury is the intracanalicular part followed by the intracranial portion. Blunt trauma to the frontal bone result in forces being transmitted to the fixed intracanalicular segment of the optic nerve which can result in a fracture of the optic canal. As mentioned earlier, the tight adherence of the optic nerve's dural sheath to the periosteum within the optic canal causes the optic nerve to be vulnerable to the impact of skull injuries. Haemorrhage either within the optic nerve sheath or in the orbital cavity can cause loss of optic nerve function. Mechanical forces are considered to be the primary mechanism of injury. These forces cause lacerations, partial or complete avulsion of the retrobulbar nerve, contusion necrosis, and disruption of the nerve's vascular supply, resulting in hemorrhages, and thereby cause permanent damage.

Once the vascularity of the optic nerve is disturbed, the secondary mechanisms of injury come into play. Oedema sets in soon after and this in turn further compromise the vascular supply by causing a rise in intraluminal pressure. Secondary mechanisms that have been studied include numerous pathways for the generation of free radicals and arachidonic acids, lipid peroxidation, production of inflammatory mediators such as bradykinin, loss of calcium homeostasis with disruption of cellular function, glutamate-induced excitotoxicity, cell-mediated inflammation, and initiation of neuronal apoptosis. Most treatment modalities revolve around limiting the secondary injury with the hope of rescuing those axons which have survived the initial trauma. Direct traumatic optic neuropathy is less common because the laxity of the intraorbital optic nerve allows for both absorption and deflection of the penetrating object. The resilience of the dura to penetration also offers further protection.

Treatment of traumatic optic neuropathy
The management of traumatic optic neuropathy remains controversial owing to a general lack of understanding of the pathophysiology involved and uncertainty about clinical results after therapeutic intervention.

Three major therapeutic options for traumatic optic neuropathy exist:

- Corticosteroids
- Surgical optic nerve decompression
- Combination of the two.

Steroids in Traumatic Optic Neuropathy:
The use of systemic corticosteroids in traumatic optic neuropathy is currently thought to be the best form of treatment as opposed to none at all. It is hypothesized that that pathologic free radical reactions are initiated following major central nervous system trauma and that very high doses of corticosteroid functioned as antioxidants to inhibit free radical damage. Steroids as a form of therapy for traumatic optic neuropathy has been accepted after the results of National Acute Spinal Cord Injury Study (NASCIS-2) which showed positive results when systemic corticosteroids were used in patients of acute spinal cord trauma. NASCIS 2 was a multicenter, randomized, double-blind, placebo-controlled study involving patients with acute spinal cord injury, patients. These patients were randomly assigned to receive placebo, naloxone, or methylprednisolone within 12 hours of spinal injury. Intravenous methylprednisolone was given as an initial dose of 30 mg/kg followed by a continuous infusion of 5.4 mg/kg/h. When compared with placebo, treatment with methylprednisolone within 8 hours of injury resulted in a significant improvement in motor and sensory function.

However, whether the results of NASCIS can be extrapolated to justify the use of systemic corticosteroids in traumatic optic neuropathy needs to be further investigated. The International Optic Nerve Trauma Study, in which visual outcomes were compared with patients following observation alone, high dose steroids given within 7 days of the injury, and optic canal decompression with or without corticosteroids and performed within 7 days of the injury; showed no clear benefit for either corticosteroids or optic canal decompression in patients of traumatic optic neuropathy.

Other recent studies have also shown that there is no difference in visual acuity improvement between intravenous high-dose corticosteroids and placebo in treatment of recent traumatic optic neuropathy. Most recently, the Corticosteroid Randomization After Significant Head Injury (CRASH) trial, a large randomized, placebo-controlled study, evaluated the effect of early administration of 48 hours infusion of methylprednisolone on the risk of death and disability after head injury. The investigators found a small but statistically significant increase in the risk of death within 2 weeks after head injury in the group allocated to corticosteroids compared with placebo. Furthermore, recently A Cochrane systematic review in 2007 critically analyzed the available evidence for the role of systemic steroids in traumatic optic neuropathy and showed that there were no convincing data of additional benefits of
steroids over observation alone\textsuperscript{16}. The spontaneous improvement seen in many patients makes it difficult to assess the efficacy of any treatment method. Thus the use of corticosteroids in traumatic optic neuropathy continues to be a controversy.

**Surgical Management of Traumatic Optic Neuropathy**

Optic canal decompression surgery has a limited role in the management of traumatic optic neuropathy. This treatment is based on the hypothesis that swelling in the optic canal may lead to a compartment syndrome. The increasing edema would decrease tissue perfusion to cause more postinjury ischemia to the optic nerve. This decompression procedure is believed to decrease edematous pressure in the optic canal to reverse ischemia and axonal conduction block, which can result in irreversible axonal degeneration. A variety of approaches have been described, including transfrontal craniotomy, and transethmoidal, transantral ethmoidal, sphenoidethmoidal, and endoscopic decompression. The surgical approach should be based on the location of the pathology as visualized by CT scanning. The goal of optic nerve decompression is to provide surgical relief of pressure on the intracanalicular segment of the optic nerve\textsuperscript{3}.

Nerve sheath hemorrhage occurs rarely and the images must be looked at together by an experienced neuroradiologist and clinician to avoid calling peri-sheath blood an intrasheath hemorrhage. If intrasheath blood is convincingly and clinician to avoid calling peri-sheath blood an intrasheath hemorrhage. If intrasheath blood is convincingly demonstrated, optic nerve sheath fenestration is indicated\textsuperscript{17}.

Although significant advances have been made in the understanding of neuronal damage and repair, there is still no consensus among practitioners regarding the protocol for managing traumatic optic neuropathy. The diagnosis of traumatic optic neuropathy remains a clinical one. Clinicians must use their discretion to decide with the patient or family whether the implementation of medical or surgical intervention outweighs the risks\textsuperscript{2}.

**Recent advances and current research:**

It has been shown that the innate adaptive T-cell mediated immune response directed against self-antigens located at the site of damage can be neuroprotective after optic nerve or injury. By augmenting this response in individuals at the site of damage can be neuroprotective after optic nerve or injury. By augmenting this response in individuals and repair, there is still no consensus among practitioners regarding the protocol for managing traumatic optic neuropathy. The diagnosis of traumatic optic neuropathy remains a clinical one. Clinicians must use their discretion to decide with the patient or family whether the implementation of medical or surgical intervention outweighs the risks\textsuperscript{2}.

**References:**