Optic nerve sheath fenestration (ONSF)

Optic nerve sheath fenestration (ONSF) procedures were first described by De Wecker in 1872 as an incision in the meninges surrounding the optic nerve in order to relieve elevated intracranial pressure.

Indications

An optic nerve sheath fenestration is indicated when papilledema leads to progressive visual loss despite previous, conservative measures and when no cause of increased intracranial pressure can be indentified and eliminated. This rather rare constellation usually occurs in idiopathic intracranial hypertension. The procedure is performed via a medial transconjunctival orbitotomy. If headaches and neurologic symptoms other than visual deterioration prevail, the placement of a ventricular shunt is preferred.

Surgical Technique

The three main surgical approaches for ONSF are superior eyelid, lateral orbital, and medial transconjunctival.

The medial approach for ONSF is most common in American Society of Ophthalmic Plastic and Reconstructive Surgeons (ASOPRS) members respondents; however, the superomedial lid crease incision is gaining popularity for its efficiency and elegance. Three quarters of surgeons remove a window and a quarter fenestrate with slits. Optic nerve sheath biopsy has limited diagnostic value. In the absence of any evidence to suggest a diagnosis other than idiopathic intracranial hypertension, the usefulness of biopsy during fenestration is low.

Superior Eyelid Approach

Approaching the superior and lateral orbit via the anterior superior eyelid crease has been a surgical method preferred by orbital surgeons for removal of intraorbital lesions. In recent years, the anterior superior eyelid crease approach has been adopted by neurosurgery because of the excellent surgical exposure it provides to the anterior skull base. A particular advantage of this approach is the excellent cosmetic outcome because the incision can be hidden within the natural superior eyelid crease.

For the purposes of performing an ONSF, the medial intraconal space is accessed through a superomedial eyelid crease incision.

The orbital septum is opened and the medial horn of the levator aponeurosis pushed laterally. With blunt dissection, a plane is created between the medial rectus muscle and the superior oblique tendon to access the posterior orbit avoiding the superior ophthalmic vein and vortex veins. With further posterior dissection, the optic nerve comes into view and a slit or rectangular window is created within the optic nerve sheath. Advantages of the superiomedial eyelid approach for an ONSF include decreased traction on the globe, decreased operating time, decreased bleeding, limited sharp dissection of normal tissue, and absence of extraocular muscle traction or dissection. Limitations of this approach include an increased distance from incision site to the optic nerve and an external (skin) incision compared with the medial transconjunctival approach (see below).

Lateral Orbital Approach

The approach to the intraconal orbital space for ONSF through the lateral conjunctiva was first
described in 1872.

Subsequently, the lateral approach was modified by a number of surgeons to improve access to the optic nerve. In 1988, Tse and colleagues described a lateral orbitotomy approach for ONSF.

The procedure begins with an en bloc removal of the lateral orbital wall. The periorbita is incised in a T-shaped fashion and blunt dissection of the perimuscular fasical sheaths is performed until the lateral rectus muscle is identified. A traction suture is placed under the insertion of the lateral rectus muscle and the suture is anchored medially, adducting the eye in order to move the optic nerve laterally. Dissection with specially designed orbital-neurosurgical brain retractors is used to gain access to the optic nerve. Once the retrobulbar portion of the optic nerve is adequately exposed, an operating microscope is used to assist in a window incision of the optic nerve sheath. The periorbita is closed with interrupted sutures and the bone fragment is re-approximated to the lateral orbital wall using nonabsorbable suture. Advantages of the lateral approach to ONSF include exposure of a longer segment of the optic nerve. Limitations of this approach include longer operating time, an external incision, and a more complex surgical procedure that requires removal of the orbital rim.

**Medial Transconjunctival Approach**

The medial approach to the retrobulbar optic nerve was first described by Galbraith and Sullivan in 1973.

The following description is the technique performed at the Duke Eye Center.

After induction of general anesthesia, a medial limbal conjunctival peritomy is performed and the conjunctiva incision is extended superiorly and inferiorly. The medial rectus muscle is isolated and the tendon is secured with a double armed 6–0 vicryl suture. The muscle is detached from the globe using scissors, leaving a small remnant of muscle tendon attached to the globe. A 5–0 dacron traction suture is placed through the muscle tendon, and the globe is retracted laterally. The long posterior ciliary arteries are then identified between the superior and inferior poles of the insertion of the medial rectus muscle. With the aid of small malleable retractors the retrobulbar optic nerve is approached through the posterior reflection of tenon’s capsule and retrobulbar orbital fat. The orbital fat is retracted away from the optic nerve with small strips of cottonoids. A small angled forceps is used to improve exposure of the optic nerve. With the assistance of the operating microscope a sharp blade on a long handle is used to incise the optic nerve sheath approximately 2 mm posterior to the globe with careful attention to avoid any blood vessels on the surface of the nerve. A fine toothed forceps is inserted into the incision site and extended posteriorly with microscissors to a total length of 3–5 mm. A tenotomy hook is sometimes inserted into the SAS and moved in the anterior–posterior direction to lyse any arachnoidal trabeculations and adhesions. On completion of the fenestration, the traction suture is removed, and the medial rectus is reattached to the globe using standard strabismus muscle technique. The conjunctiva is closed with 8–0 vicryl sutures. An antibiotic-steroid ointment is applied to the eye and a protective shield is placed over the eye to prevent any direct external pressure. The advantages of the medial transconjunctival approach include increased width of the operating field, minimal distance from incision to the optic nerve, minimal bleeding, and lack of an external incision.

**Complications**

One patient developed decreased vision and choroidal folds 1 month postoperatively.

Other patient developed proptosis and pain 9 months postoperatively. Neuroimaging showed a cyst-like structure adjacent to the optic nerve in each patient. In each case, symptoms and signs resolved
after surgical excision of the structure\textsuperscript{5}.

After ONSF, if there is a suggestion of further visual loss, shunting should be considered if intracranial pressure is high\textsuperscript{6}.

**Case series**

**2016**

Robinson et al conducted a retrospective chart review of consecutive patients with IIH who failed maximal medical therapy and underwent ONSD between January, 1992 and November, 2014, and were followed at least 3 months postoperatively. The main outcome measure was the relationship between OP on lumbar puncture and ONSD failure. We also investigated the relationship of OP with visual acuity, visual fields, age, and gender.

During this period, 174 patients met inclusion criteria. Of the 40 patients who had an OP $\geq 50$ cm H$_2$O, 6 (15\%) had progressive visual loss after uncomplicated ONSD, vs 6 (4.5\%) of 134 patients with an OP $< 50$ cm H$_2$O ($P = 0.032$, Fisher exact test). Patients with worse visual acuity at presentation also had a higher risk of progressive visual loss after ONSD ($P < 0.001$, Cochran-Armitage trend test), as did men ($P = 0.048$, Fisher exact test).

Patients with IIH and an OP $\geq 50$ cm H$_2$O had a 3-fold increased risk of failure of ONSD to prevent progressive visual loss, requiring a shunting procedure when compared to those with OP $< 50$ cm H$_2$O. Visual acuity at presentation and male sex also were associated with progressive visual decline after ONSD. These risk factors merit closer follow-up in the postoperative period when signs of further visual deterioration would indicate an urgent need for neurosurgical shunting\textsuperscript{7}.

**2015**

A retrospective study of ONSF patients from 2004 to 2011. Patients' symptoms, body mass index, CSF opening pressure, and visual outcomes were analysed.

ONSF's were carried out on 31 eyes of 14 patients. 64\% were female and 36\% were male. The most predominant symptom was a headache (93\%). 71\% of patients had a BMI$>30$. The average CSF opening pressure was 36mmHg (range 22-64). Post ONSF, visual acuity (VA) improved in 24.1\%, remained stable in 62.1\% and worsened in 13.8\% of operated eyes. 6\% were lost to follow up. Visual fields (VF) were reliable in 48\% of operated eyes. Of these 33.4\% improved, 53.3\% remained the same and 13.3\% worsened. Colour vision (CV) improved or remained stable in 87\%, and worsened in 13\% of operated eyes. 4 patients had tertiary procedures (LP or VP shunts). ONSF resulted in statistically significant improvement/stabilisation in visual acuity, visual fields and colour vision. Most importantly, this was not dependent on the body mass index.

ONSF is a safe procedure in experienced hands. It predominantly stabilises visual function in majority of maximally medicated patients but also offers improved visual function to some patients. Colour vision monitoring is a useful adjunct in patient with unreliable visual fields. Unfortunately patients whose visual function deteriorated despite maximal medical and surgical treatment were often those who presented late or had a delay in their clinical diagnosis\textsuperscript{8}.


