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## ABSTRACT

**Objectives.** Optic nerve sheath fenestration (ONSF) is commonly used in idiopathic intracranial hypertension (IIH). Here we will present our experiences of ONSF in 26 patients with special attention to indications, surgical techniques and results

**Methods.** The recorded data of patient management (with the result) who underwent ONSF were reviewed and studied retrospectively.

**Results.** The total number of patients who underwent ONSF was 26. The male-female ratio was 1:12.

Indications of ONSF were: 1. Idiopathic Intracranial Hypertension (IIH)-23 cases; 2. Cerebral Venous Sinus Thrombosis (CVST)-02 cases; 3. CNS Tuberculosis-01 case.

All patient underwent bilateral ONSF with post-operative continues lumbar CSF drain for 04 days. After fenestration gush of CSF came out with force in all-first operated eyes whereas 13-second operated eyes showed very little CSF flow after fenestration. Vision improved in different grades in all cases at discharge except in three cases. Preoperatively, visual acuity was either PL&PR or hand movement in 40 eyes where 04 eyes were preoperatively total blind (no PL&PR). Visual acuity improved in 48 eyes (92.3% eyes) where the patient can do his/her daily life activities including self-care. Improvement in IIH is 100% (23 cases i.e-46 eyes) whereas 01 case out of 02 cases in CVST. Though vision was improved dramatically fundal appearances changes very slowly and very less frequently returned to normal appearance.

**Conclusion.** Due to the delicate and technically demanding nature of the surgery, safety is a major concern of the ONSF. Our experience showed ONSF is a technically safe operation with very good results where indicated.

## INTRODUCTION

Orbit contains eyeball, optic nerve (ON), extraocular muscles, fat, lacrimal gland, vessels and nerves (Figure 1). Actually, the optic nerve is an optical system projection white matter tract of the CNS. It is covered by pia mater, arachnoid mater, and dura mater. ON has intracranial, foraminal and orbital part. The subarachnoid space (SAS) of the CNS is continues with SAS of optic nerve. So increased intracranial pressure is transmitted to the optic nerve head and causes papilledema.<sup>1</sup>

## Keywords

optic nerve sheath  
fenestration,  
ONSF,  
IIH,  
CVST,  
TB,  
visual impairment



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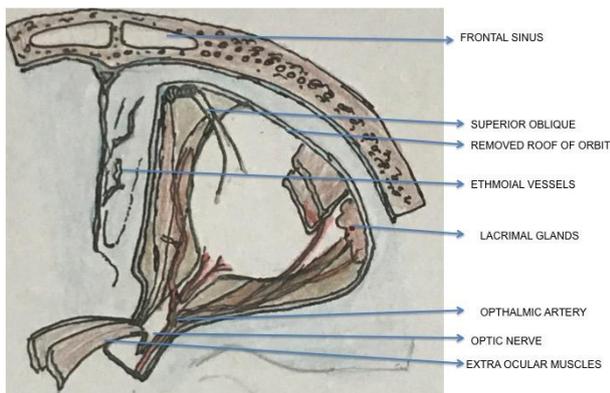


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Optic nerve sheath fenestration (ONSF) was first used by De Wecker in 1872 as an incision in the dura and arachnoid surrounding the optic nerve in order to relieve increased intracranial pressure. 2,3The technique is commonly applied in idiopathic intracranial hypertension (IIH) with rapid and/or progressive vision loss.4

But this technique can also be used in other conditions where CSF pressure in optic nerve SAS is increased locally, compartmentally or generally i.e. cerebral venous sinus thrombosis, CNS infection such as tuberculosis.

Here we will present our experiences of ONSF in 26 patients with special attention to indications, surgical techniques and results.



**Figure 1.** Schematic hand drawing showing the orbital structures including optic nerve after removal of orbital roof.

## METHODS

From January 2011 to December 2018, the patients who underwent ONSF in National Institute of Neurosciences & Hospital and some other private Hospital in Dhaka, Bangladesh were studied. The recorded data of patient management along with follow up were reviewed retrospectively. Clinical pictures, investigations, medical and other failed surgical management/s, ONSF procedures and follow up (clinical and investigations) were carefully studied and presented as results. Post-operative follow ups were done on 1st POD, 3rd POD, at discharge, after one month, at the end of six month and then 01 yearly. If patient failed to attend for follow up then follow up was achieved through voice or videophone call. Perimetry and fundal photograph were done six months after operation. Total follow up period was ranging from 06 months to 72 months (average 22.4 months) except one

patient where patient recovered vision from PL&PR to hand movement at discharge on 7th POD and then she lost from follow up even over telephone.

## RESULTS

The total number of patients who underwent ONSF was 26. Male female ratio was 1:12.

Age range was 11 years to 38 years (average 22.5years). Build of patients was average and no one was obese or morbidly obese patient. Two patients were pregnant and were in 2nd trimester. In all cases bilateral ONSF was done i.e. total number of eyes were 52. No one was taking any drugs, steroid or contraceptive. Indications of ONSF were: 4. Idiopathic Intracranial Hypertension (IIH)-23 cases; 5. Cerebral Venous Sinus Thrombosis-02 cases; 6.CNS Tuberculosis-01case.

## Clinical profile (Table 1)

All IIH patients were diagnosed as IIH in Neurology department according to IIH diagnostic criteria. All underwent MRI of brain and eyeball (Figure 2) including MRV where all ventricles were normal with normal MRV.



**Figure 2.** A&B- MRI of brain and orbit (T2W axial and sagittal image) showing elongated & curvy ON with increased CSF spaces and depression on globe by ON head (arrows marked) in IIH.

When there was severe visual deterioration even after having adequate medical management including serial lumbar puncture and/ continues lumbar CSF drainage for three days. Opening pressure was measured in all cases where CSF pressure was above the normal level ranging from 280 to 330 mm of CSF (Average 295mm). CSF study was normal in all cases except three (one with CNS TB, 01 cases with IIH where CSF protein was just above the normal another case with high CSF

without any known cause) Preoperative visual status of patient was shown in Table 1.

No patient underwent lumbo-peritoneal (LP) or ventriculo-peritoneal (VP) shunt procedure. Preoperatively (Just before ONSF) no patient had nausea or vomiting and in 08 patients (30%) there was only mild to moderate (not severe) headache.

Four patient had diplopia but clinical 6th nerve palsy was found in two cases.

Pre-operatively funduscopy and fundal photograph was taken in all cases and perimetry was done in cases where vision is not badly impaired. Fundal photographic and perimetric findings are shown in Table 1.

**Table 1.** Particulars of patients with pre and postoperative Visual acuity and papilledema outcome.

Age (years), sex & Diagnosis	Pre operative-VA	Post operative last F/U-VA	Pre operative papilledema (Frisén scale)	Post operative papilledema (after six month)
22,F,IIH	Rt-PL&PR, left -HM	6/18 bilaterally	G-4	G-3
31,F,IIH	PL&PR bilaterally	Hand movement-bilaterally (at discharge)	G-4	--
11, M, IIH	PL&PR-bilaterally	6/6 bilaterally	G-4	G-2
35, F, IIH	Finger count- bilaterally	6/6 bilaterally	G-4	G-2
19, F, IIH	PL&PR- bilaterally	6/12 bilaterally (only central field of vision recovered)	G-5	G-4
27,F, IIH	PL&PR- bilaterally	6/6-Rt 6/12-Lt	G-4	G-3
19,F,IIH	Rt-PI&PR Lt-finger count	Rt-6/24 Lt-6/12	G-5	G-4
23,F, IIH	Hand movement-bilaterally	6/18 bilaterally	G-4	G-4
26,F, IIH	Hand movement-bilaterally	6/6 Rt 6/12 Lt	G-4	G-4
15,F,IIH	Left- total blind, Rt-PL&PR	Rt-6/36 Lt-6/24	G-5	G-4
22,F,IIH	PL&PR-Bilaterally	6/36- bilaterally	G-5	G-5
25, F ,IIH	Hand movement-bilaterally	6/6-bilaterally	G-4	G-3
16,F, IIH	PL&PR-bilaterally	6/6-Rt 6/12-Lt	G-4	G-4
26,F,IIH	Hand movement-bilaterally	6/18 -bilaterally	G-4	G-3
33,F, CVST)	PL&PR-bilaterally	Rt-6/18 Lt-6/24	G-4	G-4
27, F, IIH	Hand movement-bilaterally	6/6-bilaterally	G-4	G-3
26, F ,IIH	Rt-hand movement Lt-6/24	Rt-6/12 Lt-6/6	G-4	G-3
14, F, CVST	Blind(No PL&PR)-bilaterally	Blind (no improvement)	Early optic atrophy	Optic atrophy
30, F, IIH	Rt-6/60 Lt-6/36	Rt-6/12 Lt-6/6	G-4	G-4
38, F,IIH	Hand movement-bilaterally	Rt-6/12 Lt-6/6	G-4	G-4
26,M, IIH	Hand movement -Rt Lt- 6/24	Rt-6/12 Lt-6/6	G-4	G-3
20, F, IIH	PL&PR -bilaterally	Total recovery 6/6-bilaterally	G-4	G-2

32, F, IIH	Hand movement-bilaterally	6/18 -bilaterally	G-5	G-4
14, F, IIH	Rt-6/18 Lt-PL&PR	Rt-6/6 Lt-6/12	G-4	G-3
26, F, IIH	Hand movement-bilaterally	6/6-bilaterally	G-4	G-3
30, F, TB,	Rt eye vision loss (no PL/PR) Lt-PL&PR	Rt-No improvement Lt-PL&PR lost (blind)	Early optic atrophy	Optic atrophy

### Operation (ONSF)

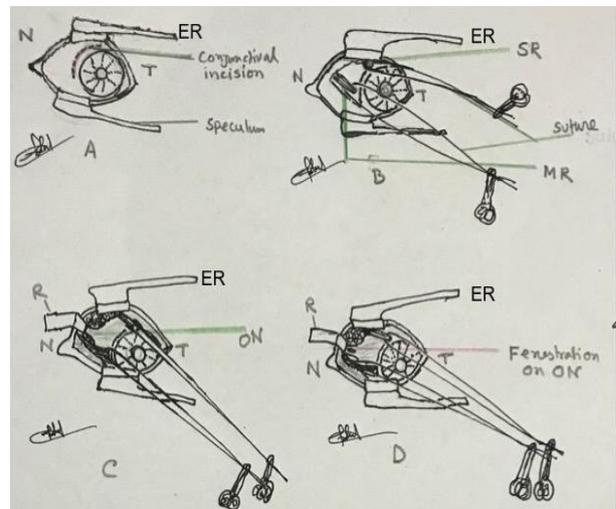
All patient underwent bilateral ONSF with post-operative continues lumbar CSF drain for 04 days.

### Surgical techniques (Figure 3 and Figure 4)

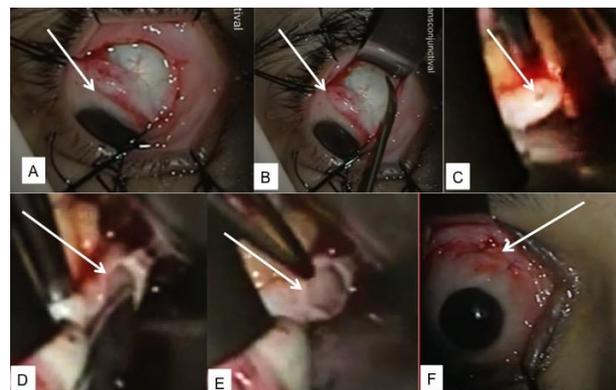
Under general anaesthesia with endotracheal intubation patient was positioned in supine position. Eyelid and periorbital skin was painted carefully (with protection of cornea and conjunctiva) by diluted povidone iodine solution. Then eyelids, corneal and conjunctiva were irrigated with normal solution. Keeping both eye exposed patient was draped. More severely affected eye was operated first. Universal eye speculum was placed to retract the eyelids. Then rest of the procedure was done with operating neurosurgical microscope under high magnification. A perilimbal conjunctival incision was given 12 to 3 o'clock position on right side and 9-12 o'clock position on left side. Scleral insertion of superior and medial rectus muscle were identified and a 2-0 silk was passed under the tendons of SR and MR muscles near its insertion to sclera for controlling the movement of eyeball. The eyeball was rotated downward and outward to make the optic nerve superficial and accessible. By dissecting under the bulbar conjunctiva in between SR and MR ON was reached. Conjunctiva and upper eyelid was retracted upward and medially. ON was identified by its position, color and continuity with the posterior eyeball. Vortex veins and ciliary nerves also help to identify the ON to some extent. If orbital fat prolapse in the field then the situation becomes difficult and more number of spatula's retraction may be needed. Undue pressure on globe was avoided. During globe rotation with MR & SR stay sutures corneal injury was carefully avoided. A linear, parallel to optic nerve incision was given to dural sheath and gush of CSF was noted and aspirated. Then retractor, MR & SR sutures were removed. Conjunctiva was re-sutured with 7-0 vicryl.

Same procedure was done on the opposite eye and antibiotic eye ointment was given.

The patient was positioned lateral to insert lumbar CSF drain and 200ml/day CSF was drained for the next four days. Tab. Acetazolamide continued for next six months.



**Figure 3.** Sequential (A, B, C & D) schematic hand drawings of medial trans-conjunctival ONSF on left side. N-nasal side, T-temporal side, ER-eyelids retractor, R-retractor, Sutures-controlling sutures on superior and medial rectus tendon, SR-superior rectus, MR-medial rectus, ON-optic nerve.



**Figure 4.** A, B, C, D, E & F - per operative sequential images showing stages of ONSF on right side. A&B- conjunctival incision and trans conjunctival dissection (arrow marked). C, D&E-Fenestration on sheath (arrow marked). F-Conjunctival closure after ONSF.

## OBSERVATIONS

The total operation time was ranging from 45 minutes to 90 minutes (average 55 minutes). Peroperative corneal abrasion occurred in 02 cases. Single vortex vein injured in one case that needed to be coagulated. Peroperative minor venous bleeding occurred in two cases that needed to be coagulated. In six cases peroperative difficulties occurred due to orbital fat prolapse in the targeted zone. After fenestration gush of CSF came out with force in all first operated eyes whereas 13-second operated eyes showed very little CSF flow after fenestration. There was no scleral injury or bulbar perforation or rupture.

Post-operative transient upper eyelid swelling occurred in three cases. Transient anisocoria with abnormal light reflexes occurred in 18 eyes that recovered within three days. There was no proptosis, retrobulbar hematoma or no ocular cranial nerve palsy occurred in any of the cases.

Headache cured in all cases after the operation. Vision improved in different grades in all cases at discharge except in three cases:

1. Case with CNS TB
2. One CVST case &
3. One case of IIH where at discharge there was no visual improvement but by the end of three month her vision recovered to 6/12 bilaterally.

Case with high CSF protein began to improve from 1st POD.

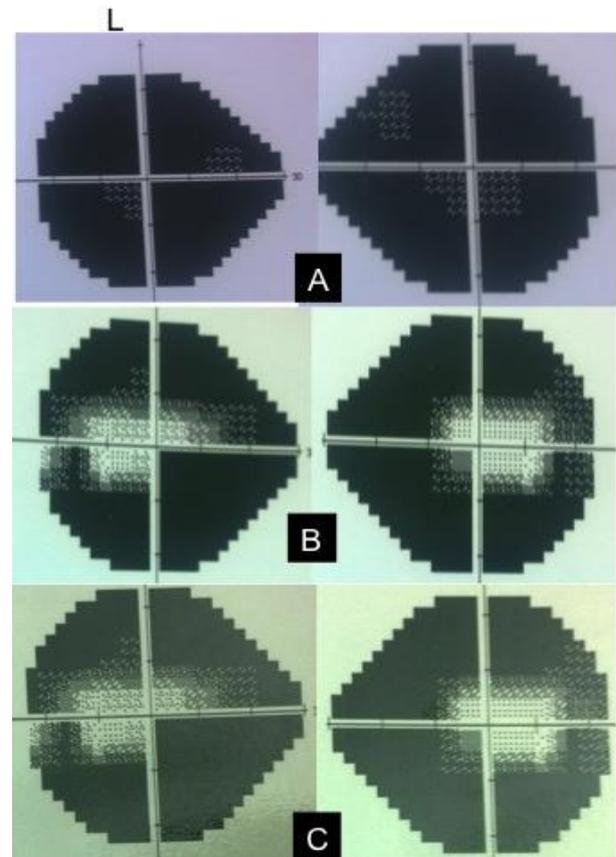
By the end of three month all sixth nerve palsy recovered.

In one case patient's vision recovered from hand movement to near normal but at the end of six month she developed left sided sixth nerve palsy without any return of IIH symptoms. This abducent nerve palsy recovered with steroid therapy for three weeks.

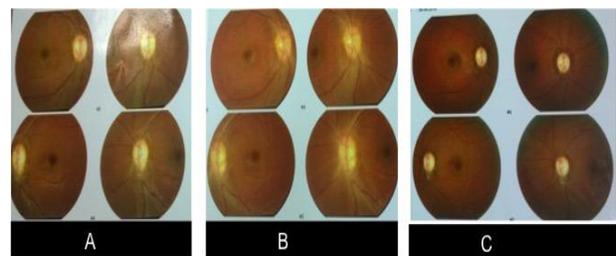
Pre-operative and postoperative (last follow up) visual status is shown in Table 1. In all cases vision were severely affected preoperatively. Preoperatively, visual acuity was either PL&PR or hand movement in 40 eyes where 04 eyes were preoperatively total blind (no PL&PR). Visual acuity improved in 48 eyes (92.3% eyes) where the patient can do his/her daily life activities including self-care. In one case (two eyes), vision recovered only in central part of visual fields where peripheral part did not recovered even after one year (Figure 5).

Improvement in IIH is 100% (23 cases i.e-46 eyes) whereas 01 case out of 02 case in CVST.

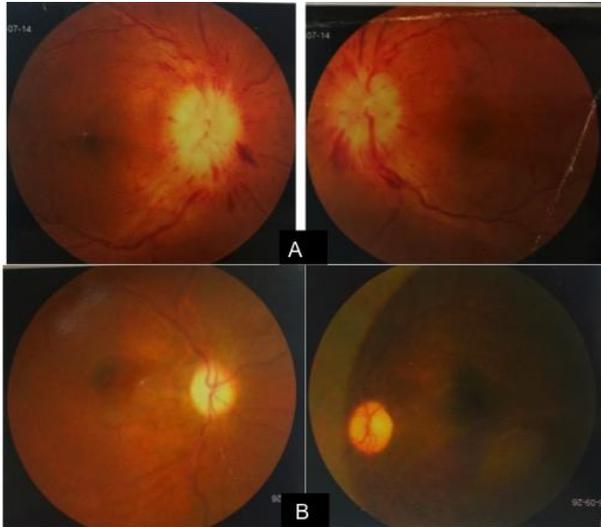
Pre and postoperative fundoscopic findings are also shown in the Table 1. Though vision improved dramatically fundal appearances changes very slowly and very less frequently returned to normal appearance (Figure 6 & 7). In our series, six months after ONSF papilledema improved in 23 cases (88.4%).



**Figure 5.** Visual field analysis (in IIH). A-preoperative. B & C-consecutive postoperative visual field analysis 06 and 12 after operation showed only central fields recovery (though patients' visual acuity was 6/6 bilaterally).



**Figure 6.** A-preoperative fundal photograph, B-post operative (03 months after ONSF) fundal photograph and C- post operative (06 months after ONSF) fundal photograph.



**Figure 7.** A-pre operative fundal photograph and B-post ONSF fundal photograph (after six month).

## DISCUSSION

Pathogenesis of raised intracranial pressure includes increased production of CSF, reduced drainage to circulation, intracranial space occupying lesions, traumatic brain injury, cerebral venous outflow obstruction, CVST and idiopathic intracranial hypertension (IIH).<sup>6,7,8</sup> IIH is a special pathology, a diagnosis of exclusion, diagnosed by papilledema without any identifiable CNS pathology, and usually occurs in obese women of child bearing age.<sup>9</sup> IIH is also referred to as pseudotumor cerebri. It is a disorder of unknown etiology that must meet the following criteria:

- Signs and symptoms of increased intracranial pressure (headache, nausea, transient visual obscurations lasting seconds, double vision, dizziness, emesis);
- Elevated cerebrospinal fluid (CSF) pressure (>200 mmHg in nonobese adults and >250 mmHg in obese adults);
- Normal neuroimaging studies;
- Normal neurologic examination (with the exception of papilledema and/or cranial nerve palsies);
- No other identifiable cause such as medications (including vitamin A, tetracycline, oral contraceptive pills, nalidixic acid, lithium, steroid use, or withdrawal)<sup>5</sup>.

The effects of raised ICP are responsible for disruption of the axoplasmic flow, swelling of axons, leakage of water and proteins resulting

papilledema.<sup>10,11</sup> Papilledema in its severe form (if left untreated), causes significant and irreversible loss of vision, with Visual Field (VF) defects and color vision.<sup>12</sup>

Papilledema treatment is primarily focused on treating identifiable etiologies of raised ICP. IIH is treated by medical or surgical options as indicated. Medical treatment includes Acetazolamide, Steroids, Topiramate, Frousemide Whereas surgical options include Optic Nerve Sheath Fenestration (ONSF) or shunt procedures (lumboperitoneal shunts, ventriculo-peritoneal shunts, or ventriculo-atrial shunt).<sup>9,13,14</sup>

ONSF, first described by DeWecker in 1872 for treatment of papilledema.<sup>14</sup> Hayreh described the blood supply of optic nerve, and thus also described the efficacy of ONSF in resolution of papilledema.<sup>15</sup>

The ONSF is principally used for the treatment of IIH. The ONSF is also used less frequently for the management of other pathological conditions that adversely affect the vision such as progressive non-arteritic ischemic optic neuropathy, and optic disc drusen,<sup>16</sup> cryptococcal meningitis,<sup>17</sup> CVST, and intracranial masses causing raised ICP that causes visual deterioration.

The three main surgical approaches for ONSF are superior eyelid, lateral orbital, and medial transconjunctival approach. In medial transconjunctival approach scleral insertion of MR is cut and after fenestration it is reattached.<sup>1</sup> In our series we used the medial transconjunctival approach but we performed the nerve fenestration without cutting the MR tendon.<sup>1</sup>

Primarily visual acuity (VA) is the function of fovea, and does essentially provide the idea into preservation of central or paracentral visual field (VF).<sup>18</sup> Increased ICP essentially affects the macula, as a result of swelling of retinal nerve fibre layer as well as exudates and subretinal fluid. So resolution of papilledema is expected to improve vision after ONSF.

Analysis the histological features of IIH have shown that vision loss from outer retinal layer changes in the macula is more reversible than vision loss from optic neuropathy and inner retinal layer change.<sup>19</sup> Improvement in VA does not depend upon pre-operative papilledema stage and thus explains the macular function is independence of optic disc swelling alone. It also means change in VA is independent from pre-operative VA, explaining

that poor vision before surgery doesn't necessarily contra-indicates the performance of ONSF.

The range of improvement in VA after ONSF is a debatable and variable subject. Studies have shown wide range in improvement in VA, from as low as 14% to as high as 100%.<sup>20,21</sup>In our series improvement in VA in 24 cases (92.3%), non-improvement in 2 cases (7.7%).In IIH visual improvement is 100% (23cases out of 23 cases of IIH). In one of the largest studies conducted on 578 eyes of 331 patients, improvement or stability was seen in 94.4% and worsening in 5.6% of eyes.<sup>16</sup>

ONSF principally means the improvement of vision. But it has efficacy in improvement of other symptoms also. Published data shows wide range in headache improvement, with as low as 13%, and as high as 90% patients with headache.<sup>22,23</sup> our series also showed the same. So there is indirect evidence that ONSF has role is in controlling chief symptom of IIH or raised ICP.

Post ONSF improvement of papilledema ranges of 71 to 100%.<sup>24,25</sup>It is logical to mention that improvement in papilledema stage does not necessarily mean complete resolution of papilledema. After ONSF complete resolution of papilledema is rare. In our series, six month after ONSF papilledema improved in 23 cases (88.4%). In the largest meta-analysis done on result of ONSF with follow up of 20 months, improvement in headache, visual acuity and papilledema was seen in 26%, 42% and 92% respectively.<sup>9</sup>

The complication rate of ONSF is ranging from 5 to 45% in the literature.<sup>26</sup> Complications of ONSF including visual deterioration (<1%),<sup>22</sup> permanent atonic pupil, retrobulbar hemorrhage and sixth nerve palsy have been reported in the literature.<sup>27</sup> Other less frequently seen complications are transient blindness, choroidal infarction, diplopia and orbital infections.<sup>16,28</sup>In our series we face no major permanent complication.

## CONCLUSION

Due to the delicate and technically demanding nature of the surgery, safety is a major concern of the ONSF. Our experience showed, ONSF is a technically safe operation with very good results where indicated.

## DECLARATION

Ethics approval and consent to participate – Not applicable (NA)  
Consent for publication - Taken from the patients/patient's party.

Availability of data and materials - NA

Competing interests - None

Funding - None

## ABBREVIATIONS

CVST-cerebral venous sinus thrombosis

CNS-central nervous system

CSF-cerebro spinal fluid

ICP-intracranial pressure

IIH-idiopathic intracranial hypertension

LP-lumbo-peritoneal

MR-medial rectus

MRV-magnetic resonance venography

ON-optic nerve

ONSF-optic nerve sheath fenestration

POD-post operative day

PL-perception of light

PR-projection of rays

SR-superior rectus

SAS-subarachnoid space

TB-tuberculosis

VA-visual acuity

VF-visual fields

VP-ventriculo-peritoneal

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