Double CSF to Areolar Connective Tissue Shunting. An Efficient and Minor Procedure in Idiopathic Intracranial Hypertension. A Prospective Comparative Study.

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Abstract: Study Design: A prospective study to evaluate the outcomes of 2 complimentary minor surgical procedures for shunting CSF to loose areolar connective tissues in both the eye and the scalp as an ensured, effective and definitive treatment to idiopathic intracranial hypertension.

Objective: To explore a more effective and less invasive technique without extensive tissue injury to idiopathic intracranial hypertension (IIH).

Summary of Background Data: The optic nerve sheath decompression (ONSD) is an established minimally invasive operation for IIH. The latter operation usually fails to completely eliminate headache as its effect is largely potent as regard the ophthalmological manifestations of IIH. Thus a shunt operation is frequently needed thereafter. In the other hand; the traditional neurosurgical operation subtemporal decompression proved itself for decades as an effective surgery for resistant cases of IIH, however, it was plagued by the temporal-subtemporal extensive tissue injury. We found the newly revived minimally, invasive ventriculo-subgaleal shunt (VSS) that proved effective in our Kasr El Aini hospital and actually divert the CSF to the scalp areolar connective tissues, a minimally invasive alternative to the old style operation subtemporal decompression and a very beneficial and effective operation that compliment the ONSD.

Methods: This prospective study included 18 consecutive patients, 8 patients underwent ONSD and the rest ten patients needed in addition VSS to effectively manage all the manifestations of IIH.

Results: The evaluation of the manifestations of IIH; the headache, papilledema-visual field defects and the abducent nerve paresis were performed before surgery and after surgery. Both maneuvers had an excellent-to-fair operative result, with a low incidence of complications. Thus according to our data the double CSF shunting is a minimally invasion technique with a high efficiency and ensure a very satisfactory result.

Conclusion: Our novel operation of double CSF to areolar connective tissue shunting is a minimally invasive and exceptionally effective operative management to IIH and can be a standard procedure for the surgical treatment of this insidious and dangerous vision threatening disease.

1. Introduction:
Idiopathic intracranial hypertension (IIH) remains an enigmatic diagnosis of exclusion. It is a vision-threatening disease of unknown aetiology that affects predominantly obese women of childbearing age. Prompt diagnosis, thorough evaluation and efficient treatment are crucial for improving the presenting symptoms. IIH is a syndrome of increased intracranial pressure (ICP) without hydrocephalus or mass lesion and with normal chemical and cellular CSF composition. It is usually presented by headache (75%), papilledema (95%), and sixth nerve palsy (25%) with otherwise normal neurological examination.

To start with; Medical therapies should be tried which include:
Weight control and non-ketotic diet.
Discontinuation of contributing medications.
Serial lumbar punctures.
Carbonic anhydrase inhibitors and diuretics.
Steroids, and certain analgesics.
Some authorities believed in digoxin.

Neurosurgical procedures previously used to alleviate ICP include subtemporal decompression, lumboperitoneal shunts, parietal flaps, and recently the venous sinus stenting trials. Lumboperitoneal shunt is considered the traditional method for providing prompt reduction of ICP. However, other surgical option is optic nerve sheath decompression ONSD,
which has an established role for reversal of visual loss. It is a rather simple procedure without great morbidity and it has a long term stability of the result without such complications as shunt obstruction, kinking or recurrence of IH. It actually addresses the nearest subarachnoid tissue that lacks any bony coverage. It simply involves cutting slits in the dura surrounding the optic nerve; which allows CSF flow into the orbital areolar fat to be absorbed into venous circulation. Previous researchers showed that neither headache is reliably relieved by this approach nor aggressive papilledema. We herein in this paper assessed and tested its combination with another simple surgical and minor manoeuvre that recently showed efficiency in Kasr El Aini neurosurgical department Cairo University and promise after its revival in the past two decades, the ventriculo-subgaleal shunt VSS; as to manage successfully all the complaints of the patients including headache and aggressive papilledema. This latter surgery adds only about 15 minutes to the surgical time without any drawbacks. THE HYPOTHESIS WAS CLEAR patients failing medical therapy have ONSD performed if visual deterioration is the only morbidity. Adding VSS procedure is considered if headache is a major symptom, failure of ONSD and/or the papilledema is aggressive.

Aim of work
Assessment of the outcome of patients with idiopathic intracranial hypertension comparing patients adopted and persisted on medical therapy to those who underwent either optic nerve sheath decompression alone or combined with VSS.

2. Methodology
Study design:
A prospective (longitudinal) comparative study on therapeutic modalities for IH.
It included 29 patients; they were followed up for at least 18 months.

Criteria for diagnosis:
Presence of papilledema and headaches, documented raised CSF pressure (>250 mm/CSF), with normal CSF analysis.

Exclusionary criteria:
(1) Ventriculomegaly or an intracranial mass on imaging.
(2) Evidence of venous sinus thrombosis.
(3) Abnormal CSF constituents.
(4) Patients with irregular follow-up.
Patients were considered irregular when two follow-up visits were missed.

Initial Evaluation:
Initially 43 patients were included, (14 were irregular) and were not subjected to any surgical intervention.
All the other 29 patients were submitted to:

Neurological Assessment:
History taking [headache characters and intensity, visual symptoms including transient visual obscurations (TVOs) and double vision].
Funduscopic assessment, visual acuity (VA), pupillary reactions, ocular motility, other cranial nerves, and signs of lateralization.
BMI was calculated.

Ophthalmological Assessment:
58 eyes were evaluated for:
Degree of papilledema with fundus photographs.
Intraocular pressure measurement.
Visual Acuity.
Pupillary reaction.
Clinical assessment of field of vision.

According to severity of papilledema:
Early papilledema.
Acute fully developed papilledema.
Chronic papilledema.
Atrophic papilledema: divided into:
Atrophic on top of chronic papilledema.
Atrophic on top of acute papilledema.
Post-papilledemnic optic atrophy.

Visual Field Testing:
The test strategy: full threshold test.
It evaluates the mean deviation of retinal sensitivity (MD) in each eye, and the size of blind spot.
The mean deviation (MD) was used to evaluate the deterioration and improvement, taking (-/+ 2) of MD as a significant worsening or improvement (Spoor and McHenry).

Radiological Assessment:
All patients had had brain CT, MRI and MRV.

Laboratory Tests:
Include CBC, fasting and post-prandial blood sugar, hormonal profile when clinically indicated, coagulation profile and parallel blood sugar level just before spinal tap. For patients receiving diuretics, serum K+ level was periodically checked.
Spinal tap.
Medical Management.
Modest dieting and a low-salt regimen.
Carbonic anhydrase inhibitor [acetazolamide] (4 weeks).
Repeating of spinal tap for 9 patients.
If no clinical improvement steroids were added, in addition to furosemide as a 2nd line agent (for 8 weeks). Thereafter, patients were reassessed neurologically and ophthalmologically, and by using visual field testing.

Surgical management:

1-ONSD
- Operative procedure: a single incision over the insertion of the medial rectus, extending from the upper to the lower fornices without release incisions was done.
- Disinsertion of the medial rectus muscle with a double armed 6-0 Vicryl leaving a muscle stump of about 1mm for the intrascleral suture.
- Traction on the globe to obtain adequate exposure; anterior traction then lateral traction on the globe was done by vertical recti silk sutures looped beneath the insertion of the muscle and tied. This was combined with intrascleral traction suture (5-0 non-absorbable), (which is a suture passed through the scleral stump of the medial rectus muscle taking partial thickness of the sclera). This suture was mainly used to obtain lateral traction of the globe. The traction on the globe was limited to 30 to 60 seconds and the interval between traction periods was 15 to 30 seconds. Because of the frequent cutting of the suture through the muscles stump, one suture is passed through the stump until the middle and another suture is passed from the other end of the muscle stump to meet the other suture in the middle. Now we have 4 points of traction instead of only two points. A retractor was used to retract the medial rectus and the medial orbital contents medially. The intrascleral course of the long posterior ciliary artery was used as an easy and constant guide for orientation in finding the nerve sheath. The operating microscope was focused along this line to reach the sheath.
- Multiple incision technique was used in 19 surgeries and 18 eyes by the fine microvitreo-retinal (MVR) blade, starting from 2mm behind the globe and extending for about 5mm, not exceeding 8mm behind the globe making 3 incisions. An initial CSF gush was seen in most of the cases, also secondary gushes or leaks were noticed with the second and sometimes the third incisions. Window removal technique was used in 4 surgeries. The window removed was always rectangular, and in the least vascular area of the nerve sheath. It measured 2 x 4 mm.
- The medial rectus was reattached with closure of the conjunctiva. Subconjunctival gentamicin and decadron were given in the lower fornix in all cases. The operation is minor, easy and straightforward with minimal manipulations. The following figures show crucial steps in ONSD; in a respective manner:

- The fornix based conjunctival incision.
- Lateral traction on the globe using the single continuous (base ball) suture.
- Clear view of the optic nerve sheath with the lateral traction on the globe and with the retractor in place. (notice the intrascleral course of the long posterior ciliary artery)

2-VSS

It is actually a very simple procedure that can be done even on urgent basis. All that is needed is a Nelaton catheter (size 10) (that was prepared with multiple small elliptical pores done using the scissore), and through a burr hole this tube is used to tap the ventricle. The other end of the catheter is placed in the subgaleal space after its dissection in the areolar connective tissue layer i.e. separation of the galeal layer from the periosteal layer.
The patient is put in the supine position with the head 30 degree flexed and 45 degree tilted to the left side. Linear skin incision of about 3.5 cm is done in the right frontal region, 3 cm from the midline and one cm in front to the coronal suture and is held in its position by a self retaining retractor. Dissection of the sub-galeal space is performed for a circle of a 7 cm diameter. Perfect hemostasis is achieved usually easily. The frontal burr hole is done. Dura opened and cauterized. The lateral ventricle is tapped via a brain canula. A Nelaton catheter size N10 (that was prepared with multiple small elliptical pores done using the scissors) is introduced using a suitable stillet after removal of the brain canula and in the same tract. Sudden gush of the CSF is avoided to prevent the occurrence of subdural hematoma. The Nelaton catheter is then fixed to the surrounding galea. The distal end is inserted in a smooth curve in the subgaleal space to prevent kinking. The direction of the shunt in the subgaleal space should be posteroinferior. Closure in two layers in a water tight manner is mandatory.

Follow up Assessment:
Patients broadly fell into three categories according to Higgins et al:
Asymptomatic: complete resolution of headache and visual impairment.
Improved with some residual not requiring further intervention.
No change or worsening of headache intensity or visual impairment.
Two weeks Post-operatively Assessment included:
Headache evaluation (change in frequency and severity).
Visual symptoms and visual field test.
Fundus photography.

Long-term Follow-up.
Every 3 months for at least 18 months: patients were evaluated for visual field, detailed headache assessment, and full neurological and ophthalmological examination.

3. Results:
Mean Age: 29.07 + 4.89 yrs (Range: 5 – 43 yrs).
Initial presentations: headache and visual symptoms.
Duration of symptoms: Mean: 5.5 + 2.63 months (Range: 2 – 13 months).
Asymmetric visual affection was detected in 40%.
Automated Field Evaluation:
Bilateral enlargement of blind spot in all patients.
Baseline mean deviation (MD) of retinal sensitivity were:
   OD: 9.31 + 3.77 (range 4.9 – 21.7).
   OS: 8.59 + 3.19 (range 4.8 – 23.1).
The Mean CSF Pressure: 362.5 (range 290 – 470 mm/CSF).
Clinical Presentation:
11 TVO.
8 Blurring and sparkling.
5 Diplopia [6th nerve palsy].
4 VA deterioration, and 3 relative afferent pupillary defect (RAPD).
1 Trigeminal neuralgic – like pains.
Maximum medical therapy was advocated for all patients, and they were re-assessed after 12 weeks:
Responder Group (11/29).
Non-Responder Group (18/29).
Characteristics of Responders:
Shorter duration of illness.
Milder degree of papilledema, lesser affection of field of vision.
Lower basic CSF pressure.
Drugs tolerability.
Less frequently needed repeated spinal tap.
Lower BMI.
Post-operative Assessment [2 weeks].
Measurable improvement was detected regarding visual symptoms and the MD of retinal sensitivity in both surgical groups.
After ONSD, Eight patients underwent ONSD, one patient (1/8) experienced marked improvement of headache intensity (reduction of acetazolamide dose), other 7 patients received the same doses of diuretics.
After ONSD+VSS , Ten patients underwent ONSD+ VSS (none was on diuretics post operative) all patients had major headache improvement.
All patients with preoperative transient visual obscurations TVOs experienced complete recovery of this condition after surgery (ONSD or ONSD+VSS).
Three eyes had RAPD; those patients were subjected to surgery. One of them improved after ONSD surgery, and the remaining two improved after ONSD + VSS with normal pupillary reactions.
In ONSD, the unoperated eye showed improvement in response to surgery in the fellow eye.
All eyes with atrophic papilledema that improved after surgery had residual field defects. No complications were encountered in short-term follow up period.
Figure 4. The pre- (top) and post-operative (bottom) fundus pictures of patient N 9 where atrophic papilledema on top of acute papilledema in the preoperative fundus with rapid diminution of vision to HM, was replaced in the post operative (after ONSD+VSS) fundus with increased disc pallor despite functional improvement to V.A. of 6/36 and 6/60.

Figure 5. The preoperative (top) and the postoperative (bottom) fundus picture and visual fields of the patient n12 with preoperative acute papilledema that was resolved in both eyes after ONSD+VSS, also the visual field showed marked improvement.

During 18 months Follow-up:

One patient who was on medical therapy has recurrence of symptoms associated with increase in CSF pressure [non-compliance]; and she was subjected to ONSD+VSS.

One patient who underwent shunting procedure had the shunt kinked and not working and it was revised once.

Doses of acetazolamide were reduced in 4 patients out of the responders to 750 mg/d after one year of stabilization.

All patients with ONSD+VSS markedly improved without the need of any therapy.

Two patients of ONSD were on reduced doses of medical treatment.

4. Discussion:

It was proven in the literature that visual loss can happen with a functioning VP shunting procedure (27). Optic nerve sheath decompression (ONSD) alone has very good potential for the reversal of visual loss according to many reports (1,5,35). The procedure is simple, without great morbidity, with a very low rate of complication, and a long term stability of the results (11, 12). However, headache is frequently not improved denoting that the bouts of increased intracranial tension in some patients are still present. Combining the former operation with (VSS) completely resolved all the complaints of the patients and the fundus examination as well as the field of vision follow-up showed marvelous results.

The female dominance was noted in this study as well as in other similar studies. The two children in this study were males, and this may be due the fact that IIH has nearly equal sex incidence in children (9, 38).

Headache was present in all cases in this study; however, visual loss may occur in IIH patients even if medical therapy succeeded in relieving headache (38). ONSD alone is not a surgery for headache relief. Headache relief should be viewed as a bonus on top of visual recovery (25). Thus we combined VSS to it and gave marvellous results.

A very important rule is that in absence of papilledema there is no potential for visual loss. On the other hand, in cases of atrophic papilledema, effective surgery is indicated without waiting for the results of medical therapy because this may jeopardize the remaining vision of the patient. It is very important to know that a white flat disc is not hopeless because inside this atrophic disc there may be viable fibers suffering only axoplasmic flow obstruction and their number may not be sufficient to cause ophthalmoscopically visible disc swelling (35,37).

In acute fully developed papilledema, the visual loss was observed to follow all or none rule. Visual loss is either severe acute and not responsive to medical therapy or there is no or mild visual loss. If visual deterioration is detected, an emergency ONSD +VSS is a must. If visual functions are stable, with or without medical therapy, follow up of cases is indicated. In our cases, as well as in the literature, acute fully developed papilledema and chronic papilledema have the best prognosis of visual recovery after surgery (1, 35).

All the eyes in this work had improved or stabilized V.A. except one eye with transient outer layer retinal ischemia as a complication of ONSD surgery. However, there are other 2 cases with V.A. deterioration after initial improvement. In one case, the V.A. regressed to the preoperative level. However, after a secondary ONSD+VSS, the V.A. improved, but...
less than the level reached after the first surgery. The other eye was improved after few days because of the resolution of macular edema after VSS. The results of this novel work are very satisfactory and in harmony with most of the literature about the successful and more sophisticated manoeuvres.

Preoperative V.A. does not always predict the final postoperative V.A. If we draw a line at the level of 6/60 vision, acute cases even with optic atrophy have the greatest chance of crossing this line of legal blindness. Atrophic chronic cases without disc elevation usually improve, but usually few cases cross this line.

During this work, two problems were especially related to IIH patients going for field examination. The first problem was headache. Field examination should not be done while the patient has a headache because this usually will lead to an unreliable result and time wasting due to repetition.

The second problem is TVOs which can obscure vision for seconds and lead to false negative errors and fixation losses. Field-testing should be done during a TVO-free period. Medical therapy may be intensified before field examination in a trial to decrease both headache and TVOs.

In the results of this work, all eyes improved after ONSD alone or combined with VSS except 2 eyes, which had deterioration of visual field. VSS also resolved the problem of the headache and improvement of all aspects were detected. Thus a higher incidence of eyes had no residual field defects and only 2 eyes had a mild postoperative field defects. This is because of the relatively delayed presentation in our cases.

The effect of ONSD on visual field is far beyond any doubt as proved in this work and many other similar studies. The role of the visual field as the major indicator for surgery is also the cornerstone of any management decision in every patient with IIH and visual deterioration (17, 20, 21). We found in addition that when ONSD was combined with VSS, prompt and excellent solution to all the manifestations were satisfactorily gained and assured.

Important points and modification of the ONSD technique:

The medial transconjunctival approach, as most of the recent literature states, is much easier, time saving, and without significant difficulty in the nerve sheath exposure, which is the most important goal of the orbitotomy (27, 32). We also found that changing the direction of traction from anterolateral to lateral will be more suitable. As the anterior traction on the globe will consume the redundancy of the optic nerve and the globe will be tethered during the trial to rotate it laterally. This will make lateral traction more difficult and will expose all the vascular structures attached to the nerve sheath to maximum stretch. Conversely, if we used all the redundancy for lateral traction only, the result will be easy rotation of the globe laterally and, also, of the optic nerve that becomes suitably medial to it so as to be readily visible at the surgical site. ONSD is actually a filtration surgery and opening the sheath with the least manipulation, traction and instrument introduction is crucial to surgical success. It is believed that cotton tipped applicators should not be used for exposure because this may provoke further reaction and fibrosis with more risk of surgical failure. If it is a must, an alternative like the surgical microsponges is a much better choice. Using the mentioned modifications, we needed only 2 tractions each of 60 seconds interval to open the sheath by multiple slits.

Important points and modification of the VSS:

As regard the neurosurgical VSS, we think that it is a refined and renewed operative procedure of the same category and idea as the old style operation subtemporal decompression that for many decades was the only available surgical procedure for IIH and proved itself in the old classical literature. Actually; the absorptive surfaces in the subtemporal decompression operation is the areolar connective tissue in the surrounding area. The areolar connective tissue of the subgaleal layer of the scalp is magnificently loose, capacious and extensible. The simple separation of the galea layer of the scalp from the peristium is fast, easy and almost bloodless. In the same time, the heroic blood supply to the scalp actually denotes and reflects our results of efficient CSF absorption in this scalp layer. We preferred the right frontal burr hole and found it suitable; also we found separation of the galea from the peristium for only 7-8 cm is enough. However the operation is extensible and can satisfy and cope with whatever high is the intracranial pressure by increasing the size of the absorptive surfaces if needed. It should be noted that the key of success of VSS is to keep open the CSF filled gap between the peristium and the galea, thus a suitable surface area of the areolar connective tissue layer should be freshened and prepared; if larger than enough; reapposition of the galea upon the peristium will abolish the absorptive ability of the operation. Fixation of the 12-14 cm shunt tube should be properly done to the galea to prevent the shunt tube segment from being driven inside the ventricles which is a complication that occurred in one patient. Water-tight closure in two layers should be considered mandatory. When combined with ONSD the surgeon felt satisfied, and the results were excellent.

Ophthalmologically wise; we consider papilledema is the mirror image of glaucoma, and the
value of ONSD equals the value of trabeculectomy in the glaucoma patient. This subject deserves much more attention because visual loss from papilledema due IIH can be disastrous. Comparing ONSD with the VSS technique of CSF diversion; the former is limited to the orbital area and the intracranial pressure secondarily improve, however headache was frequently unchanged denoting less than optimal management of ICT; yet we found combining both manoeuvres had an assured and long-term satisfactory efficacy.

5. Conclusion:

IIH is a chronic disease that requires long-term maintenance on medical therapy to prevent recurrence. Medical therapy is usually considered satisfactory as long as it is maintained effective and tolerated. ONSD is the surgery of choice for visual deterioration or gradual visual loss in IIH patients. It is not an alternative to a neurosurgical shunting procedure. We found VSS to be effective, easily done, and was indicated mainly for headache and aggressive papilledema. Visual field testing is the main tool for surgical decision and follow-up of cases. Lower surgical manipulations are much correlated to success of surgery. In order to get the best visual results, surgery should not be delayed.

ONSD and VSS were found effective surgical means to reduce the pressure in patients who were intolerant or not responsive to medical therapy or when clinical deterioration was detected. The choice between ONSD or ONSD + VSS is clear and resolved, patients failing medical therapy have ONSD performed if visual deterioration is the only morbidity. Adding VSS procedure is considered if headache is a major symptom, failure of ONSD and/or the papilledema is aggressive; in few cases optic nerve fenestration was required then followed by VSS in another operative setting to satisfy the patient complaint and completely save the optic nerve.

The three modalities (medical therapy, ONSD and VSS) are complementary. A neurosurgeon and an ophthalmologist should work together to provide the optimal and rational decisions in the care of patients with IIH.

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