An Introduction to Idiopathic Intracranial Hypertension

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Introduction

The constellation of raised intracranial pressure without ventricular enlargement along with an elevation of cerebrospinal fluid pressure with a normal composition of the same constitutes Idiopathic Intracranial Hypertension (IIH). The many names it bears viz. pseudotumor cerebrii, serous meningitis, otitic hydrocephalus benign intracranial hypertension underscores the enigma that it is in terms of etiology and pathogenesis apart from it's uncertain clinical course. Owing to it's significant morbidity in terms of possible visual loss and chronic headache IIH needs early diagnosis and management.

Pathogenesis of Idiopathic Intracranial Hypertension

Though modern imaging studies have demystified our understanding of this disease, much is yet to be known about the exact pathogenesis. As per the Munro Kellie doctrine (1), there is a constant relationship between blood, CSF and brain contents with one increasing at the cost of raised pressure beyond a point of physiological limit. An increase in CSF production, proposed in the pathogenesis of IIH by Quincke (2) has since been discounted. Dandy (3) proposed increased blood volume though later investigators could not find evidence for the same. The recent studies with MR imaging reveal increased white matter water with an increase in apparent diffusion coefficient of the same (4).

The more acceptable theory includes one of brain's venous out flow obstruction at the large cortical venous sinuses. IIH involves elevated venous pressure, leading to increased resistance to CSF absorption and subsequently increased ICP (5). Though an obstruction is not always evident, the pressures in these venous channels are found to be elevated in patients with IIH. Here again controversy is courted as to whether the sinus obstruction or resistance to venous outflow is secondary to elevated intracranial pressure affecting the cortical venous system (6).
Clinical features

The common symptom that brings the patient to the physician is headache, which is often more in the early hours of the morning (7). Vomiting though this is not a common feature at times relieves this. Pain along the occiput and trigeminal regions of the face also has been reported.

It is the onset of visual obscuration with the headache that often necessitates a comprehensive ophthalmological evaluation for the victims of IIH. Another visual phenomenon is the presence of double vision due to abducens nerve involvement (8).

Tinnitus is present in some due to augmented blood flow through the venous sinuses is often experienced by its sufferers (9). Ataxia and vertigo also accompany the visual disturbances. Often there may be features of endocrine disturbances in the form of pituitary deficiency due to empty sella turcica.

It is important to obtain a detailed history of previous drug therapies as many a time the IIH may be secondary to pharmacological interventions. This list includes Vitamin A (10,11) and its derivatives, Tetracyclines(12), Fluoro quinolones, sulfa derivatives, growth hormone(13), and steroids including oral contraceptives(14).

The list of systemic disorders associated with IIH includes SLE (15), anemia(16), underlying malignancies, (17), Addison's disease (18), thyroid disorders(19), uremia (20) etc. It is important to rule out the same prior to embarking on surgical treatment.

The Dandy's criteria (21) modified subsequently holds good to identify cases of IIH as it seeks out those with features of raised intracranial pressure in an alert patient without neurologically localizing features or false localizing features. The CT/MRI subsequently rules out mass lesion and the CSF study shows elevated pressure without other causes being found for the same.

Investigating a case of IIH

The investigations into a case of suspected IIH includes a visual evaluation including optic fundus examination and perimetry to evaluate the severity of papilledema and extent of visual loss. Rarely papilledema may be absent too (22). The Snellen’s visual (23) chart may not reveal the extent of visual loss. Visual Evoked Potential often show disturbances. Endocrine evaluation involving cortisol and thyroid evaluation is also required (18,19).

CT scan is often the commonest investigation sought for IIH as the physician tries to rule out a case mass lesion in the brain. The widened perioptic space and the presence of empty sella turcica (arachnoid membrane hernia ting into the pituitary fossa) are often present. A slit-like ventricle, though classically described, is seldom found in patients with IIH (25).

Cerebrospinal Fluid (CSF) evaluation

Once the presence of mass lesion is ruled out the neurologist often embarks to rule out any chronic meningitis by doing a lumbar puncture in a guarded fashion due to raised intracranial pressure. The CSF often shows very high pressure but significantly the chemical and cytological composition does not show any significant alteration suggestive of absence of chronic meningitis, which often mimics IIH.

MRI in IIH

MR imaging especially with angiography has become the cornerstone of any investigation into IIH.

Magnetic resonance Imaging of the brain may reveal (not always) empty sella turcica with arachnoid invagination, patulous optic nerve...
sheaths, flattening of the posterior sclera of the globe and papillary projections on the optic nerve sheath in varying frequencies (26,27,28).

The presence of dural venous sinus stenosis is being increasingly identified in cases of IIH thanks to improvements in the Magnetic Resonance Imaging techniques and technology. One report yielded 27 cases of sinus stenosis in a series of 29 cases imaged using the auto-triggered elliptic-centric-ordered three-dimensional gadolinium-enhanced MR venography (ATECO MRV)(6). This has often changed the management principles from CSF diversions and other surgeries to endoluminal procedures dilating the narrowed venous systems.

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Medical management of IIH

Carbonic anhydrase inhibitors (e.g., acetazolamide) are the only effective medications for treatment of IIH. Acetazolamide was originally demonstrated to variably decrease CSF production by 6 to 57% among human subjects [1]. A standard dosing is a 500-mg time-release capsule twice daily. The effective dose must be individualized; a dose of 0.5 to 1 g per day usually suffices for resolution of papilledema. Caution must be exercised in prescribing Acetazolamide in pregnancy for reported teratogenicity, including limb malformations and cortical dysgenesis in animals [2]. Case reports have documented neonatal metabolic acidosis and electrolyte abnormalities [3]. Furosemide, another diuretic, has little effect on CSF production; it may be used for patients who cannot tolerate acetazolamide.

The role of corticosteroids in the treatment of papilledema is controversial. A short course of high-dose corticosteroid therapy may be helpful for patients with acute visual loss resulting from fulminant papilledema [4]. However, corticosteroids should not be used chronically for treatment of papilledema.

Weight loss can be beneficial for patients with papilledema resulting from IIH. It is difficult to prove that weight loss improves papilledema in IIH because

1) acetazolamide administration is often initiated concurrently [5]

2) few patients lose enough weight to test the theory, and

3) the disease can remit spontaneously.

However, a history of recent weight gain often accompanies the initial presentation of IIH.

Some neurologists advocate serial lumbar punctures (e.g., twice weekly) as an alternative to
surgery for patients with papilledema that cannot be controlled medically. However, serial lumbar punctures are a poor approach because

1) most obese patients with chronic papilledema are difficult to treat with lumbar punctures and
2) patients generally dislike serial lumbar punctures.

At best, lumbar puncture and drainage of a large volume of CSF are useful emergency measures for patients with severe papilledema and sudden deterioration of vision. In some cases, it is necessary to hospitalize patients with lumbar subarachnoid drains until surgery can be scheduled. This is a good strategy if lumboperitoneal (LP) shunting is planned, although it may increase the risk of infection. Optic nerve sheath fenestration is easier to perform when the sheath is turgid; therefore, the drain should be removed or clamped before surgery.

Surgical Treatment

Surgical intervention is warranted when medical treatment fails. A common management error is to delay too long before recommending surgery. Corbett [6] emphasized that “there is no ‘acceptable’ level of visual field or acuity loss which one should wait for—visual loss which continues despite optimum medical therapy is sufficient reason to turn to decompression”. Surgery should also be considered for patients who are unlikely to return for follow-up visits or who are unable to cooperate with medical therapy. Surgery should be considered not only for treatment of visual loss but also for treatment of intractable headaches.

Surgical manoeuvres include some form of shunt procedure such as lumboperitoneal shunt, ventriculoperitoneal shunt or optic nerve sheath decompression. Ventriculoperitoneal shunts are quite effective in reducing intracranial pressure but this is technically challenging because of the normal or small-sized ventricles in these patients

LP Shunting

Ventriculoperitoneal (VP) shunts initially advocated for IIH were difficult to insert among patients with IIH, because the ventricles were often small. The development of LP shunting circumvented this problem. Vander Ark et al. [7] published the first description of LP shunting for patients with IIH. Soon thereafter, Spetzler et al. [8] developed a method for percutaneous insertion of the shunt into the lumbar sac, greatly facilitating placement of LP shunts. A lumboperitoneal shunt is considered the preferred neurosurgical procedure for intracranial hypertension. But this procedure has a high failure rate and other complications like sudden severe visual loss and simultagnosia. The mortality from lumboperitoneal shunting, primarily from infection has been reported to be as high as 7% to 16% [9]. In spite of all these limitations, lumboperitoneal shunt remains an acceptable procedure for those patients with severe headache and for those with conjoined visual loss.

After the introduction of LP shunting, few studies documenting its efficacy for the treatment of papilledema were published. Anecdotal reports of success suggested that it was a curative procedure, as long as the shunt functioned properly. In 1981, Johnston et al. [10] published a major review of 134 cases of IIH treated between 1942 and 1979, with a mean follow-up period of 11.6 years. Fourteen patients received shunts (six VP shunts and eight LP shunts). Of the six patients who received VP shunts, four demonstrated resolution of all symptoms within 6 months. One patient developed a shunt obstruction that necessitated revision, and another patient experienced a shunt infection that necessitated removal. Of the eight patients who received LP shunts, all demonstrated resolution of all symptoms within 6 months. One patient developed a shunt obstruction that necessitated revision, and another patient experienced a shunt infection that necessitated removal. Of the eight patients who received LP shunts, all demonstrated improvement within 1 month. One patient experienced a shunt infection, and one patient exhibited severe low-pressure symptoms as a result of overdrainage. In a follow-up study, Johnston et al. [11] reported
on 36 patients who received shunts for treatment of IIH. The patients required a total of 86 shunting procedures, with a complication rate of 52% and a failure rate of 48%; the lowest revision and complication rates were associated with LP shunts.

A multicenter review of the outcomes of shunting for 37 patients was performed in the late 1980s [12]. In that study, 37 patients received a total of 73 LP shunts and 9 VP shunts, and only 14 patients remained “cured” after a single surgical procedure. Sixty-four percent of shunts lasted less than 6 months, with shunt failure (55%) and low-pressure headaches (21%) being the most common reasons for reoperation. The vision of most patients either improved (13 patients) or stabilized (13 patients) postoperatively. That report initially led to a resurgence of interest in optic nerve sheath fenestration among ophthalmologists. However, the finding that many optic nerve sheath fenestrations fail within 1 year, as well as mounting evidence of serious complications, has restored LP shunting as the favored surgical treatment option.

Two major studies demonstrated the efficacy of LP shunting for treatment of IIH. Eggenberger et al. [13] conducted a retrospective study of 27 patients with IIH, who were monitored for a median of 47 months after shunting. Visual loss was the main reason for surgery for 14 patients; headaches were the reason for the remaining 13 patients. Vision improved or remained the same for all 14 patients, and headaches improved for all patients. There were no serious complications, except for shunt failure. Fifteen patients (56%) required shunt revision, sometimes more than once (range, 1–13 revisions). The average number of revisions per patient was 2.4, with one revision being performed every 2.6 years. The authors concluded that LP shunting was a satisfactory treatment for the majority of patients.

Burgett et al. [14] reported data for 30 patients who underwent LP shunting for treatment of IIH. The mean follow-up period was 35 months. Of 14 eyes with impaired acuity, 10 eyes (71%) improved by at least two chart lines; only 1 eye experienced a decline in vision. Goldmann perimetry documented improvement for 64% of eyes with abnormal fields, and no eyes exhibited any worsening. Again, the only complication was frequent shunt obstruction. Twelve patients required no shunt revision. The remainder underwent a mean of 2.5 revisions/patient. These two studies provided encouraging data regarding the efficacy of LP shunting; the operation seems effective, as long as the shunt remains patent. As in most large reviews [15, 11, 12], obstruction was the most common complication of LP shunts (accounting for 65% of revisions in the study by Eggenberger et al. [13]. In all patients with suspected shunt obstruction, lumbar subarachnoid pressure should be measured. Neuroimaging findings may not be revealing, because the ventricles are not enlarged in IIH [16]. Technetium-99 shunt function studies can provide valuable data by demonstrating tracer flow into the abdomen and providing a halftime for radionuclide clearance [17, 18]. Other complications associated with LP shunting in those studies were less common. Secondary intracranial hypotension caused by CSF overdrainage accounted for 15% of revisions in the study by Eggenberger et al. [13], and lumbar radiculopathy accounted for 4.5% of all revisions. Shunt infections occur in only approximately 1% of cases of LP shunting [19]. Tonsillar herniation (acquired Chiari malformation) [20, 21] and syringomyelia [22] are other recognized complications of LP shunting, but they only rarely necessitate revision [23]. Tonsillar herniation may create a “new” headache syndrome. A problem common to all obese patients is technical difficulty with excessive subcutaneous abdominal fat, which necessitates large incisions. In this respect, the use of laparoscopic techniques for insertion of the peritoneal catheter is potentially advantageous.

LP shunt valve pressure mechanisms with external pressure control are now being developed. The current preference is for LP shunting with an inline
horizontal-vertical valve. For patients with repeated LP shunt obstructions, the option of VP shunting should be considered. Although VP shunting is more invasive, the long-term outcomes may be better [24]. Technical innovations in stereotactic surgery enable accurate targeting of the lateral ventricle. A recent study of seven patients treated with stereotactic VP for IIH demonstrated successful uncomplicated shunt placement in all cases [25]. Five of the seven patients experienced resolution of papilledema and six of the seven experienced resolution of headaches postoperatively. Another study demonstrated the application of frameless stereotaxy and intraoperative fiberoptic endoscopy for precise ventricular catheter insertion [26]. Those studies support the idea of routine stereotactic VP shunting in IIH, with either frame-based or frameless stereotaxy. VP shunting may facilitate noninvasive assessment of shunt function, because it provides a reservoir for isotope shunt function testing; noninvasive analysis of LP shunt function has been limited to radiological findings [27].

**Complications of lumboperitoneal shunting**

- Obstruction
- Infection
- Low-pressure headaches
- Radiculopathy
- Tonsillar herniation (acquired Chiari I malformation)
- Syringomyelia
- Subdural hematoma
- Shunt migration
- Shunt dependency

**Optic Nerve Sheath Fenestration**

Optic nerve sheath fenestration, introduced by de Wecker [28], was the first treatment devised for the surgical relief of papilledema. The operation involved insertion of a guarded neurotome into the orbit to slit the optic nerve sheath via a conjunctival incision. However, subtemporal decompression, which was introduced by Dandy [29] in 1937, became the operation of choice for papilledema. Dandy performed a right subtemporal craniectomy for decompression and reported excellent initial results in alleviating headaches and preventing visual loss. However, the longer-term efficacy was uncertain and morbidity and complications were significant, including seizures, infections, focal brain damage, cosmetic disfigurement, intracranial hematomas, and further visual deterioration [30]. Subtemporal decompression rapidly became obsolete after the introduction of intracranial shunting procedures by Ingraham et al. [31] and Matson [32].

The failure rate associated with LP shunting renewed enthusiasm for optic nerve sheath fenestration among ophthalmologists in the 1980s. The procedure had continued to be performed by a few surgeons [33, 34, 35] but only sporadically. In 1988, three major reports appeared in the ophthalmologic literature, describing the outcomes of optic nerve sheath fenestration for treatment of IIH in large series of patients [36, 37, 38]. The results were surprisingly good; the operation seemed to provide effective treatment of papilledema and maintained or improved visual acuity for 85 to 100% of patients. However, the follow-up periods were short in those studies.

In a study of 53 patients (101 eyes), Spoor et al. [39] reported that optic nerve sheath fenestration improved visual function for 69 eyes with acute papilledema and 10 eyes with chronic papilledema. In a later report with longer follow-up periods, Spoor and McHenry [40] described the outcomes of optic nerve sheath fenestration for 75 eyes of 54 patients with IIH. After initial improvement in visual function, 24 eyes (32%) required repeat optic nerve sheath fenestration because of deteriorating visual function. Deteriorating vision was detected a mean of only 10.4 months after surgery, and 25% of eyes continued to lose vision even after repeat surgery.
In 1989, Sergott et al. [41] reported improved visual function for 12 of 14 patients with progressive nonarteritic AION who were treated with optic nerve sheath fenestration. In 1995, the National Eye Institute-sponsored Ischemic Optic Neuropathy Decompression Trial Research Group reported the results of a multicenter study of optic nerve sheath fenestration for treatment of AION [42]. The study found no benefit of optic nerve sheath fenestration for treatment of AION, contradicting the study by Sergott et al. [41], and documented significant complications of the procedure, including optic nerve injury during surgery. The incidence of catastrophic visual complications was 3 cases/115 patients, or 2.6%. Another study reported postoperative blindness for 3 of 200 patients (1.5%) [43]. A 2% risk of outright blindness has discouraged patients and ophthalmologists. Plotnik and Kosmorsky [44] emphasized that the complication rate may be as high as 40%, including vascular compromise (11%, central retinal artery occlusion, branch retinal artery occlusion, or outer retinal ischemia), transient ocular motility disturbances (29%), and papillary dysfunction (11%). Although enthusiasm for optic nerve sheath fenestration has moderated, the procedure remains a viable option for the prevention of visual loss resulting from papilledema. Optic nerve sheath decompression has emerged as the preferred surgical treatment for progressive visual loss in patients with intracranial hypertension when medical therapy fails. Visual field improvement occurred in 55% [37] to 100% [36] of eyes. The appearance of optic disc, particularly in relation to optic pallor, and the magnitude of visual loss should not dissuade the physician from surgical therapy. Marked improvement in visual function occurs after surgery in patients with preoperative disc pallor [38]. Although most patients with intracranial hypertension either improved or stabilised their visual function after optic nerve sheath decompression, some patients seemed to regress after an initially successful operation.

In a few cases, optic nerve sheath decompression needs to be repeated. Technically, this is a difficult procedure due to scarring changes around the optic nerve, mainly involving fat adhesion [47]. Other complications of optic nerve sheath decompression include macular changes such as chorioretinal striae, pigmentary disturbances, exudates and subretinal hemorrhage or scar. These changes are due to transudated fluid that emerates from swollen optic disc and may cause permanent central visual loss. Visual loss can occur despite a technically well-performed optic nerve sheath decompression and can result from hemorrhage or infection. Progressive visual loss despite optic nerve sheath decompression can sometimes be reversed by a lumboperitoneal shunt [45]. The mechanism of optic nerve sheath decompression has not been entirely elucidated but it is likely that a bleb, similar to a trabeculectomy bleb is created during the healing phase and allows a low-grade leak of CSF into the retro-orbital tissues, thereby reducing the amount of fluid mechanical force that is exerted on the neuro-ocular junction.

Success of therapy is judged by the relief of headache, elimination of transient visual obscurations and diplopia, and the reduction and elimination of papilloedema. Stabilisation or improvement of visual field indicates adequate therapy. The discovery of a relative afferent papillary defect is an important finding and may either indicate worsening of a particular eye or improvement in the opposite eye. It should be remembered that as many as 40% of the optic nerve axons may be lost before there is any detectable field defect, implying that once a field defect is found, therapy must be aggressive and effective [46, 47].

**Complications of Optic nerve sheath fenestration**

- Central/ Branch retinal artery occlusion
- Central retinal vein occlusion
- Choroidal infarction
- Traumatic optic neuropathy
- Hemorrhage (intracranial or intraorbital)
- Diplopia
- Pupil dilation resulting from sphincter denervation
- Anterior segment ischemia
- Compressive optic neuropathy resulting from orbital cyst
- Infection

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