LP Shunt, the Treatment of Choice in Idiopathic Intracranial Hypertension. (A 5 Years Experience at Lahore General Hospital, Lahore)

AMIR AZIZ, YASER-UD-DIN, MUHAMMAD IRFAN
Bashir Ahmed, Tariq Salahuddin
Department of Neurosurgery Unit II, Lahore General Hospital, Lahore

ABSTRACT
Objective: The history, diagnosis, and therapy of idiopathic intracranial hypertension (IIH) are reviewed. Theories of pathogenesis are considered, the clinical presentation is described, and potential diagnostic and therapeutic challenges are explored. Keeping in view all these things efficacy of L.P Shunt is observed.

Material and Methods: A total of 35 patients were diagnosed as IIH according to the Modified Dandy’s criteria in our department from August 2007 to October 2012. An extensive literature review of IIH and related conditions was performed. The history of and rationale for the diagnosis and medical and surgical approaches to treatment are reviewed. Outcome of the patients’ with L.P shunt is observed.

Study Design: Prospective and analytic study.

Results: LP shunt addresses the cause of both headache and papilloedema more directly by effecting a global reduction of intracranial pressure. All patients underwent clinical, imaging, and CSF manometry evaluations. All patients showed failure or noncompliance to medical treatment and necessitated placement of an LP shunt. Among 35 patients who underwent LP shunt placement for IIH, 14 (40%) patients had severe and fulminant opening CSF pressures with values of more than 400 mm H2O. While in 21 (60%) patients opening CSF pressure was 260 to 400 mm H2O. Symptoms of headache and papilloedema and visual obscuration improved in most of the patients. Shunt complications included two (0.7%) cases of shunt dislodgement and one (0.35%) case of shunt over drainage. Multiple potential contributing causes of intracranial hypertension must be excluded. On the basis of the advantages and disadvantages of different treatment modalities, we found L.P Shunt to be more effective and advantageous.

Conclusion: Idiopathic intracranial hypertension is the diagnosis of exclusion. Medical therapy has a very little role in relieving signs and symptoms of this disease. L.P Shunt should be the treatment option in IIH.

Key Words: Lumboperitoneal shunt, Headache, Idiopathic intracranial hypertension, Papilloedema.

Abbreviations: IIH = Idiopathic Intracranial Hypertension, LP = Lumboperitoneal, CSF = Cerebrospinal fluid, ICP = Intracranial pressure, H2O = Water.

INTRODUCTION
Idiopathic intracranial hypertension (IIH) was originally described by Quincke in 1893. He reported several cases of increased intracranial pressure (ICP) without a brain tumor. Nonne described the syndrome more fully and coined the term pseudotumor cerebri in 1904. In 1937, Dandy described 22 patients with increased ICP without brain tumor. In 1955, Foley attempted to simplify the nomenclature by describing this condition as benign intracranial hypertension. However, in view of the potential for devastating loss of vision associated with papilloedema, Corbett and Tho-
mpson removed the adjective “benign” and substituted “idiopathic.”

Idiopathic intracranial hypertension denotes the condition of increased ICP without an obvious underlying brain pathological condition (and no evidence of venous thrombosis).

The diagnosis of the idiopathic intracranial hypertension is based on the Dandy’s criteria being modified by Smith in 1985. Dandy originally described the presence of symptoms of increased ICP, documented cerebrospinal fluid (CSF) pressures of 250 to 550 mm H₂O (18.11 to 39.85 mmHg) with normal CSF findings and excluded the possibility of brain tumors with ventriculography.

<table>
<thead>
<tr>
<th>Modified Dandy Criteria for Idiopathic Intracranial Hypertension</th>
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<tr>
<td><strong>Symptoms of increased ICP</strong> (headaches, nausea, vomiting, transient visual obscurations, or papilloedema).</td>
</tr>
<tr>
<td><strong>No localizing findings</strong> in neurological examinations (except for false localizing signs such as abducens or facial palsies) Awake and alert patient.</td>
</tr>
<tr>
<td><strong>Normal</strong> CT / MRI findings without evidence of dural sinus Thrombosis.</td>
</tr>
<tr>
<td><strong>ICP</strong> of 250 mm H₂O (18.11 mmHg) with normal cerebrospinal fluid cytological and chemical findings.</td>
</tr>
<tr>
<td>No other cause of increased ICP found.</td>
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1 mmHg = 13.8 mmH₂O. Shunt surgery (V.P. Shunt) was introduced in 1949. L.P. Shunt for IIH was introduced in 1971.

### Incidence
The prevalence is approximately 1 case / 100,000 women but increases to 13 cases / 100,000 women of ages 20 to 44 years who are 10% above ideal body weight and 19 cases / 100,000 women of ages 20 to 44 years who are obese 20% above ideal body weight.

Most studies demonstrated an age of onset between 11 and 58 years, with a mean of approximately 30 years.

Men are affected less frequently. The incidence is 0.3 cases / 100,000 men but increases to 1.5 cases / 100,000 obese men 20% above ideal body weight. Female – to – male ratios are approximately 4.3:1 to 8:1.

### Pathophysiology
Mostly unclear. Relative resistance to absorption of CSF across arachnoid villi is widely presumed.

Other theories support an abnormality in cerebral circulation which causes an increase in the brain water content which leads to increased ICP resulting in papilloedema.

Disease is most commonly associated with obese women. Obesity causes increase in the intra-abdominal pressure increasing cardiac filling pressure leading to impeded venous return from brain to heart which produces an increased venous pressure and increased intracranial pressure.

### Clinical Presentation
Headache and visual obscuration is the most common symptoms. Sometimes the patients have pulsatile tinnitus and numbness can also occur. Papilledema is a criterion for the condition, is observed for virtually all patients with IIH and is the most important sign. Abducens palsy, a false localizing sign thought to be attributable to traction of the VIth cranial nerve resulting from intracranial hypertension, is observed in approximately 20% of cases. Patients with IIH have traditionally been thought to have slit ventricles, but a review of computed tomographic findings with IIH demonstrated slit – like ventricles for only 11%; enlarged optic nerve sheaths (47%) and empty sella syndrome (46%) were more common. On L. P manometry, pressure of greater than 250 mm H₂O (18.11 mmHg), measured with the patient relaxed, in the lateral decubitus position, is one of the modified Dandy criteria. The most significant sequela of IIH is blindness or permanent visual impairment caused by prolonged papilloedema with secondary optic atrophy. Because visual loss is usually insidious, patients may be nonchalant regarding vision testing and follow-up monitoring until irreversible damage to the optic nerve occurs.

### Treatment
The treatment goal is to preserve optic nerve function while managing the increased ICP. Treatment indicated when there is severe intractable headache and evidence of progressive decrease in visual acuity. Cessation of the causative agents, weight reduction, carbonic anhydrase inhibitors and corticosteroids are the main stay of the medical treatment.

Surgical options include ventriculo peritoneal shunt, lumbo peritoneal shunt and optic nerve sheath fen-
Anecdotal reports of success suggested that it was a curative procedure, as long as the shunt functioned properly. A multicenter review of the outcomes of shunting showed that the vision of most patients either improved or stabilized postoperatively. The finding that many optic nerve sheath fenestrations fail within 1 year, as well as mounting evidence of serious complications has proved LP shunting as the favored surgical treatment option.

**MATERIAL AND METHODS**

A total of 35 patients were diagnosed as IHH according to the Modified Dandy’s criteria in our department from August 2007 to October 2012. An extensive literature review of IIH and related conditions was performed. The history of and rationale for the diagnosis and medical and surgical approaches to treatment are reviewed. Outcome of the patients’ with L.P shunt is observed.

**Surgical Procedure**

The patient is placed in the lateral position with his/her knees bend. The surgery is performed in G.A. Under complete aseptic measure a 3-4 cm skin incision is given between the spinous process of L₃ – L₄ or L₄ – L₅. A Tuohy needle can be inserted in the midline after the paraspinal incision. The head end of the table is temporarily elevated 30° to increase the pressure in the lumbar subarachnoid space. A fourteen-gauge Tuohy needle is inserted into the lumbar subarachnoid space with the bevel – pointed cephalad (Fig. 1, 2). The lumbar end of the catheter is passed through the Tuohy needle. The operating table is tilted back to the normal position. The needle is withdrawn over the catheter. A small incision is made in the flank and the subcutaneous catheter passer with the obturator in place is passed through the flank incision. The obturator is removed and the distal end of the peritoneal catheter is inserted into the lumbar opening of the subcutaneous catheter passer. The subcutaneous catheter passer is withdrawn over the catheter leaving the flank portion in place. A small skin incision is made 2 cm below the umbilicus and 3 cm away from the midline. The subcutaneous catheter passer with the obturator in place is inserted from the abdominal incision to the flank incision. The obturator is removed and the peritoneal end is inserted into the flank opening of the catheter passer. The catheter is passed through the passer. The catheter is inserted into the peritoneal cavity under direct vision. The chamber is anchored by stitching it to prevent shunt dislodgement. After homeostasis all the wounds are closed in layers and aseptic bandaging is done.

**RESULTS**

**Sex Incidence**

This study was descriptive study in which we included 35 patients, 30 females and 5 males.

**Age Incidence**

Age ranging from 20 years to 45 years who presented
to us from the outdoor department of Lahore General Hospital, Lahore from August 2007 to October 2012.

In all of our patients Fundoscopy was done which revealed papilloedema in all the 35 (100%) patients, L.P manometry revealed 16 (40%) patients had severe and fulminant opening CSF pressures with values of more than 400 mmH$_2$O, while in 21 (60%) patients opening CSF pressure was 260 to 400 mmH$_2$O. At the same time CSF cytology and biochemistry was done revealing normal reports (Table 1).

C.T Scan brain plain and with I/V contrast, MRI brain plain and with I/V contrast and MRV of all the patients were performed. Most of the CT scans brain and MRI brain revealed normal study except in 16
Table 1: CSF Pressure and Number of Patients.

<table>
<thead>
<tr>
<th>CSF Pressure</th>
<th>Patients</th>
<th></th>
<th>Percentage</th>
</tr>
</thead>
<tbody>
<tr>
<td>mmH₂O</td>
<td>mmHg</td>
<td>No</td>
<td></td>
</tr>
<tr>
<td>More than 400</td>
<td>29</td>
<td>16</td>
<td>40</td>
</tr>
<tr>
<td>260 – 400</td>
<td>18.8 – 29</td>
<td>21</td>
<td>60</td>
</tr>
<tr>
<td>Total</td>
<td></td>
<td>35</td>
<td>100</td>
</tr>
</tbody>
</table>

1 mmHg = 13.8 mmH₂O

(40%) patients showing slit – like ventricles. MRV of all the patients showed that the venous sinuses were patent. Obesity was seen in 28 (80%) patients.

Initially all the patients were offered medical treatment but due to failure of the medical treatment, they underwent L.P Shunt procedure.

Treatment policy of idiopathic intracranial hypertension, include neuroimaging (CT Scan Brain, MRI Brain, MRV), L.P manometry in lateral decubitus position, introduction of touhy needle into L₃ – L₄ space, catheter is directed upwards, CSF cytology is sent, mostly L.P Shunt is done via 2 incisions, post operative X-Rays is done in these patients.

Post operatively, X-Ray abdomen lateral and A.P views were performed to ensure the proper placement of the L.P shunt.
DIAGNOSIS

History - signs / symptoms of increased ICP (e.g. Headache transient visual obstructions)

Review of past medical history (e.g. Obesity, recent weight gain, uremia) and current medications (e.g. Vitamin A and derivatives, antibiotics)

Thorough neurological examination revealing awake and alert patient, no localizing findings (except false localizing signs such as abducens or facial palsy), ophthalmoscopic examination for papilloedema

Lumbar puncture with opening pressure > 250 mm H₂O (1.18 mmHg) normal CSF cytology

MRI examination with MRV

No mass lesion nor sinus thrombosis

Dural venous sinus thrombosis or mass lesion

Idiopathic intracranial hypertension

Secondary intracranial hypertension

TREATMENT

Ophthalmology consultation
Visual acuity testing and visual field examination via automated perimetry

MEDICAL THERAPY
Cessation of contributory medications
Weight loss (professional counseling, programs, surgical therapy when indicated)
Acetazolamide

Failure of medical therapy (poor control of Headache or declining visual function)

SURGICAL THERAPY
Lumboperitoneal shunt (with inline horizontal - vertical valve)
OUTCOME
With the L.P shunt in all the 35 (100%) patients, headache was relieved. In 28 (80%) patients the visual symptoms resolved completely while 7 (20%) of the patients there was only partial improvement. The visual improvement in our patients depended upon the pre operative visual status of the patients, as some patients presented to us late.

Shunt complications included two (0.7%) cases of shunt dislodgement and one (0.35%) case of shunt over drainage.

DISCUSSION
Two goals must be considered when managing patients with idiopathic intracranial hypertension: prevention of visual loss and resolution of symptoms. Presence of severe visual loss (20/40 or worse) in one or both eyes at time of initial examination is one of the indications for surgery. In one of the case report Levent Sinan Bir et al visual acuity was 20/100 in the right eye and 5/100 in the left eye on admission. It is estimated that up to 17 percent of patients with IIH will progress to develop permanent visual loss. The final visual outcome in fulminant IIH is variable but, up to half of the patients, could become totally blind. Mensah et al. reported that patients with marked visual field constriction and severe visual acuity loss at the time of onset of the IIH showed only partial response to therapy with diuretics and steroids and these patients have suffered clinically malignant course with rapid progression to near blindness. Malomo et al. reported a case with a 4 – week history of persistent of IIH with visual loss and deafness that had a good response to the nonoperative treatments; however, Kidron and Pomeranz reported two cases with malignant IIH and one of these patients underwent lumboperitoneal shunt operation. In fact, it is not well known about malignant form of IIH, and there is no obvious explanation of the disastrous outcome in these patients, but, in agreement with occasional reports in the literature, it is strongly suggested not to squander time so as to protect the patient’s visual acuity.

While in our study In 28 (80%) patients the visual symptoms resolved completely while 7 (20%) of the patients there was only partial improvement, although in some patients presence of visual symptoms were for a longer period of time, even than the result visual improvement after L. P. Shunt in our study is very much comparable to the international studies. At present, no prospective, randomized, clinical trials have compared the outcomes of these different therapies. Therefore, no procedure has been proven to be most efficacious.

Skau M et al. showed that optic nerve fenestration may prevent further visual loss, and has also helped relieve headache in up to one thirds of patients while with L.P Shunt there was improvement in headache in more than two third of the patients. Where as in our study 100% of the patients got relief from the headache symptoms. However, up to 33% of patients receiving fenestration who demonstrates visual improvement initially may manifest later deterioration in visual function.

In a study by Spoor TC et al, it was shown that cerebrospinal fluid shunting may be beneficial in treating acute visual loss and symptoms of intractable headache. Although considered the superior treatment for headache relief, shunting also significantly improved or stabilized visual function (Burgett RA et al). The same is proved in our study. Baker et al. recommended shunting for intractable headaches in the setting of progressive visual loss.

So, we recommend L.P Shunt to be performed as early as the patient is diagnosed.

CONCLUSION
Idiopathic intracranial hypertension is the diagnosis of exclusion. Medical therapy has a very little role in relieving signs and symptoms of this disease. L.P Shunt should be the treatment option in IIH.

Address for Correspondence:
Dr. Amir Aziz
Senior Registrar, Neurosurgery Unit II
PGMI / Lahore General Hospital, Lahore
Cell: 0333-6101244

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