CSF SHUNTING PROCEDURES IN IDIOPATHIC INTRACRANIAL HYPERTENSION (IIH)

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LEARNING OBJECTIVES
1. To become familiar with the different CSF shunting options for patients with IIH
2. To identify the advantages and disadvantages of using VP vs. LP shunts
3. To discuss the steps involved in diagnosing shunt malfunction

CME QUESTIONS
1. What are the potential challenges associated with LP shunt insertion and management?
   a. Body habitus makes positioning and insertion difficult
   b. The absence of a valve increases the risk of low-pressure signs and symptoms
   c. High rate of obstruction and difficulty assessing an obstructed LP shunt
   d. Difficult to evaluate infection
   e. All of the above
2. List the different steps involved in assessing VP shunt malfunction
   a. Bedside valve interrogation
   b. Shunt series
   c. Tapping the reservoir
   d. Shuntogram
   e. Intraoperative interrogation
   f. All of the above
3. Identify the challenges associated with VP shunt insertion in IIH patients and ways to mitigate them.
   a. Normal or small ventricles: use neuronavigation for insertion
   b. Signs and symptoms of over-drainage: use anti-siphon device or programmable valve
   c. A and B are correct
   d. Neither A or B are correct

KEYWORDS
1. Idiopathic Intracranial Hypertension
2. Ventriculoperitoneal Shunt
3. Lumboperitoneal Shunt
4. Lumbar Drain

INTRODUCTION
A 48 year old obese female, presented with worsening of her longstanding holo-cephalic headaches, nausea and transient visual obscurations. Her physical examination on two different occasions revealed progressive worsening of her bilateral papilledema. Her MRI revealed significant narrowing of the transverse sinuses bilaterally without evidence of venous sinus thrombosis. A lumbar puncture revealed an opening pressure of 31 cm of water. A trial of acetazolamide was not successful. The patient underwent insertion of a ventriculo-peritoneal shunt. Post-operatively, her headaches and visual signs and symptoms improved significantly.

Despite medical treatment of IIH and repeated lumbar punctures, some IIH patients have persistent papilledema with visual loss, chronic headaches, increased LP opening pressure, prompting the need for more aggressive treatment (1,2). Furthermore, even after “full recovery” or prolonged interval of clinical stability, the symptoms may recur in up to 38% of patients after a time period of up to 7 years (12,13). All of these factors suggest a need for a systematic approach to CSF diversion when conservative measures fail. In this review, we will focus on the surgical strategies for CSF diversion, in hopes of providing a practical overview.

CSF DIVERSION PROCEDURES
Several procedures result in CSF diversion, thereby decreasing intracranial pressure (ICP). They are sometimes indicated in IIH patients with refractory headaches related to persistent elevated ICP or those with visual loss.

INTERMITTENT PROCEDURES

SERIAL LUMBAR PUNCTURES
Lumbar punctures can be used to decrease the ICP in patients with medically refractory IIH or in cases with contraindications to carbonic anhydrase inhibitors (such as early pregnancy). In
a minority of cases, however, a single lumbar puncture, usually the diagnostic one, may suffice to improve or even reverse the symptoms (15). It is hypothesized that in these cases, the subtraction of CSF from the intracranial compartment, is compensated for (in line with the Monro-Kellie hypothesis) by enlargement of collapsible central venous segments which in turn reduces venous hypertension, promotes CSF absorption, and causes further venous enlargement and pressure reduction in a cyclic manner until a new steady state is attained (2, 16, 1). While this hypothesis of a “causative self-limiting venous collapse” can explain the clinical improvement after single or serial lumbar punctures and during continuous CSF drainage, it still cannot predict which patient would require continuous shunting as opposed to serial punctures. Finally, the benefit of serial lumbar punctures must be weighted against it potential side effects, such as infections, and patient’s discomfort. Furthermore, lumbar puncture may be particularly difficult and hence less appealing in patients with high BMI.

**LUMBAR DRAIN**

Rarely, IIH patients presenting with a fulminant form and severe visual loss might benefit from a lumbar drain while awaiting a more definitive treatment. A lumbar drain can be inserted by the bedside or in the interventional radiology suite. Fluoroscopic guidance is useful especially in IIH cases where patients’ body habitus may make the identification of osseous landmarks particularly challenging. The lateral decubitus position is used; with the head flexed, chin on the chest, and legs maximally flexed toward the head. The L4–5 interspace is identified in a perpendicular line from the iliac crest or via fluoroscopy. After appropriate prepping, draping, and injecting the entry site with 1% lidocaine, the spinal needle is inserted into the interspace with the bevel up. The sylet is removed intermittently every 3 mm as the needle is advanced in order to check for CSF flow. Once flow is established and CSF samples are taken if needed, an epidural catheter is threaded and the needle is withdrawn. The catheter is then connected to a drainage system. The zero reference point is set at the level of the external auditory meatus. The amount of CSF drained per hour is regulated by the height of the drain in relation to that reference point.

**SURGICAL IMPLANTS**

**SHUNTING**

Severe intractable headaches, progressive visual loss in spite of maximal medical therapy constitute the major indications for shunting in patients with IIH (2, 17–22). The alleviation of headaches is achieved in most patients soon after shunting (3, 17–22). Long term relief is sustained in about 50% of the patients (30). Negative predictive factors for shunt failure in the treatment of IIH associated headaches include lack of papillaedema at presentation and long-standing (> than 2 years) symptoms (36). The visual outcome following shunting, however, is less uniform. Some have reported improvement and stabilization of visual symptoms in 95–100% of patients (17–20,31). Others have shown that vision continued to worsen in up to 30% of the cases (21).

**LUMBAR PERITONEAL SHUNTS**

Historically, lumbar peritoneal (LP) shunts were the mainstay CSF diversion therapy for IIH. The traditionally cited advantage of LP shunts over ventriculoperitoneal (VP) shunts is the ease of insertion in IIH patients who usually have small and sometimes difficult to catheterize ventricles (23, 24, 25). This is particularly true with the percutaneous method pioneered by Spetzler (26). Multiple studies have shown that when functional, LP shunts are effective in alleviating headaches and improving or stabilizing visual symptoms in patients with IIH (27–29, 33). In 1997, Burgett et al. published a series of 30 patients with IIH who underwent LP shunting. The mean follow-up period was 35 months. Seventy one percent of the examined eyes improved by at least two chart lines and only 1 eye experienced a decline in vision. Visual field improved in 64% of eyes with abnormal fields, and no eyes exhibited any worsening (27). Almost simultaneously, Eggenberger et al. (28) retrospectively reviewed 27 patients with IIH, who were observed for a median of 47 months post-shunting. Fourteen patients presented with visual loss and 13 with headaches. Vision improved or remained the same in all 14 patients, and headaches improved in all patients. In both studies and in many others, the most common complication was shunt obstruction. Eighteen of 30 patients in the Burgett et al. study (27) required shunt revision at a rate of 2.5 revisions/ patient. Four patients required exceptionally high number of revisions (38, 29, 10, 10 revision, respectively).

The revision rate due to obstruction in the Eggenberger et al study (28) was 65%. Radiological imaging is not helpful in detecting these obstructions since the ventricular size does not usually change. LP shunt obstruction can be detected by nuclear medicine studies that may reveal tracer flow into the abdomen and provides half-life time of radionuclide clearance. Other less frequent yet significant complications of LP shunts include infection, radiculopathy, shunt migration, syrinx, low pressure headaches, tonsillar herniation, subdural hematomas.

The primary advantage of a lumbar peritoneal (LP) shunt over a ventriculoperitoneal (VP) shunt is the ability to cannulate the CSF space, in this case the thecal sac, as opposed to having to cannulate the very commonly found slit ventricles associated with IIH when considering a VP shunt. However, there are also as series of challenges associated with LP shunts that can be categorized based on insertion, perioperative immediate and delayed.

**TECHNICAL INSERTION RELATED CONSIDERATIONS**

Generally these patients are positioned in the lateral position to provide simultaneous access to the lumbar spine and flank. Given the often associated obesity this positioning itself creates some real challenges on a standard bed to maintain the position. The insertion site of the proximal catheter into the thecal sac can be very challenging given the body habitus. Percutaneous cannulation of the thecal
Ventriculoperitoneal Shunt

Technical Insertion Related Considerations

Proximal Catheter Insertion
The positioning on these patients is supine and represents an anatomic orientation. The primary challenge has been the ability to canulate normal or smaller than normal size ventricles in this patient population. Traditionally, predetermined landmarks were used to provide for trajectories based on historical percutaneous passages. Specifically, this involved taking measurements from the Bregma (the intersection of the coronal and sagittal sutures) and then measuring the distance between the nasion aligning with the mid-pupillary line. While these measurements often lead the surgeon to the ventricle, they can be less than reliable when the margin of error is very low, as in the case of IIH patients. In essence, in IIH patients, the surgeon used to balance the risk of a less than ideal proximal catheter placement, and hence the associated relatively high rate of proximal revision, with the benefit of having a regulated system with a valve to improve the immediate and long-term perioperative course.

The rapid evolution of frameless stereotactic navigation over the last 20 years has clearly addressed this issue. Using a thin acquisition CT scan, a 3D volume of the individual patient is recreated in an operating room using computer assisted systems. An individualized target for the catheter placement can now be created and ideal trajectories generated. This allows for the surgeon to select the optimal canulation and dynamically place the catheter under precise stereotactic guidance; thus, making VP shunting a valid, and in our opinion, a better alternative to LP shunts in patients with IIH. This comes with the added advantage of a decreased rate of proximal obstruction by specifically selecting the target for the proximal catheter.

Moreover, this technology allows the surgeon to take advantage of the use of valves while minimizing the risk of mal-position of the proximal catheter. In addition to the primary advantage of the valve (i.e. the ability to interrogate by the bedside), the availability of programmable valves allows the surgeon to externally manipulate the opening pressure of the valve and hence increases or decreases the drainage in response to each patient’s signs and symptoms. This particular setting, helps decrease the rate of symptomatic over-drainage, subdural hematomas, and tonsillar herniation.

Distal Catheter Insertion
Once the proximal catheter is in place, it is connected to the specific valve selected. The distal catheter is tunneled subcutaneously (often using an intervening incision) to the epigastric region. Given that the patient is in a supine anatomic position, this facilitates the tunneling and
abdominal insertion. While the catheter can be placed in any quadrant of the abdomen, we prefer a direct midline approach. The midline raphe between the muscles is opened and the peritoneum visualized and catheter inserted.

Again the patients’ body habitus can prove to be a significant challenge in the placement of the abdominal catheter. As a strategy to compensate for this we have used a minimally invasive laparoscopic approach to place the peritoneal catheter. In collaboration with laparoscopic surgeons, a percutaneous needle is passed into the peritoneum and the abdomen is insufflated. In this era, this is a standard and well established technique for abdominal surgery. This distal catheter is then simply inserted under direct visualization with the endoscope into the peritoneum.

This technique serves the primary role of positioning the distal catheter in a reliable fashion, and at the same time, doing so in a minimally invasive manner. (31). Both of these are valuable especially in this patient population.

SHUNT MALFUNCTION
The early signs and symptoms associated with shunt malfunction can be difficult to assess, given that they are often non-specific and commonly found in this population. Obviously more delayed clinical presentations, such as, recurrent vision loss are more specific but maybe end stage. Imaging with a conventional CT scan is not always helpful as the patient usually has normally small ventricles but still experiencing shunt malfunction. Therefore the reliance on the valve becomes an important consideration. Incremental investigations providing greater specificity and increasing invasiveness are helpful.

The first step is to percutaneously “pump” the shunt as discussed above. If it fails this test it is relatively reliable in suggesting a malfunction; however, if it passes the test it may still be obstructed, i.e., good specificity poor sensitivity. The shunt can be percutaneously tapped under sterile conditions to determine the same information of proximal and distal flow. This improves the sensitivity significantly but still has limitations. To further enhance the sensitivity, a radioisotope can be injected into the shunt and subsequently imaged. This latter investigation-shuntogram—provides the greatest specificity and sensitivity.

Finally, once in the operating room, a similar strategy is used to locate the site of the obstruction during revision surgery. The first step is to disconnect the valve and assess the proximal catheter for flow. If there is adequate proximal flow then this suggests a distal problem. The distal catheter is similarly interrogated by disconnecting it from the valve and connecting it to a manometer and allowing for flow under gravity. This strategy allows for isolation of the malfunction and the appropriate is replaced.

OTHER SHUNT OPTIONS
In cases where the peritoneum represents a contraindication for placement of the proximal catheter, e.g., active infection or open abdominal wound, the distal catheter can be placed within the pleura or atrium of the heart. Both of these represent less than ideal choices but may be an important consideration in selected patients. The option of the ventriculo-atrial shunt (VA) involves cannulating the jugular vein and under fluoroscopy passing the catheter into the atrium. The ventriculopleural shunt requires tunneling to the thorax and through a small incision placing the distal catheter within the pleura.

SUMMARY OF SHUNTING
The higher revision rates in both adults and pediatrics- for LP shunts in comparison to VP shunts has made it, in our opinion, less appealing to us for the treatment of IIH. For example, a large study of 42 patients (115 shunt placement) that retrospectively compared LP to VP in shunts in IIH patients over a 30 years period reported a revision rate of 86% in the former but only 44% in the later (30). The over-drainage rate was similar (14%) for both shunts. Furthermore, there was no significant difference between, the two shunting procedures with respect

| Shunt Type               | Primary Advantage                                      | Primary Concern                                      | Solution                                                       |
|--------------------------|--------------------------------------------------------|-----------------------------------------------------|                                                               |
| Lumbar Peritoneal        | Proximal catheter placement into the thecal sac         | Higher malfunction rate                              | Computer assisted frameless stereotactic Neuronavigation       |
|                          |                                                       | Lack of a valve (Limited regulation, difficult interrogation) |                                                               |
| Ventriculoperitoneal     | Valve (programmable if needed )                        | Placement into slit ventricles                       |                                                               |
|                          |                                                       |                                                     |                                                               |
| Ventriculoatrial         | Alternative to distal placement when peritoneum not available | Placement of distal catheter into vasculature/cardiac system |                                                               |
| Ventriculopleural        | Alternative to distal placement when peritoneum not available | Placement of distal catheter into the pleura         |                                                               |
to distal catheter migration and infection (30). A more recent study by Tarnaris et al. where the outcomes following VP vs. LP shunting for IIH were compared concluded that patients who received LP shunts were more likely to suffer complications and require revisions than those with VP shunts. Furthermore, while not statistically significant, patients with VP shunts survived longer than their counterparts with primarily LP shunts (32). With the advent of neuronavigation for ventricular catheter insertion, the primary disadvantage of VP shunts in this patient population is mitigated. Therefore, in our experience VP shunts are well suited for IIH even in cases with normal ventricles.

CONCLUSION
The absence of randomized controlled trials addressing the different treatment options for IIH limits our ability to define a standard management plan for patients with IIH. Weight-directed therapy and medical management should be the first-line treatment in patients with mild symptoms and without visual deficits. In cases of severe headaches refractory to maximal medical management and where CSF pressures are high, we recommend consideration of lumbar punctures, if appropriate, followed by ventricular shunting if the symptoms did not improve.

CME ANSWERS
1. e
2. f
3. c

REFERENCES