 Concurrent use of a lumboperitoneal shunt with programmable valve and ventricular access device in the treatment of pseudotumor cerebri: review of 40 cases

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Object. The authors evaluated the efficacy of treating patients with pseudotumor cerebri (PTC) and headaches due to increased intracranial pressure (ICP) by using a lumboperitoneal (LP) shunt with a programmable valve and ventricular access device (VAD).

Methods. Forty patients in whom PTC was diagnosed were treated using LP shunts with programmable valves and wand-guided placement of a VAD. All patients had papilledema and high opening pressure during spinal tap. The mean follow-up was 18 months (range 3–72 months). When patients complained of headaches that suggested shunt malfunction, the ventricular reservoir was tapped at bedside to assess ICP. The programmable valve was adjusted based on the patient’s headache and ICP.

Results. The VAD was tapped in 21 patients, and the LP valve was readjusted in 14. Shunt malfunction was diagnosed accurately. The 10 patients undergoing revision were all found to have shunt obstruction except 1 whose valve was replaced because it could not be reprogrammed. No patient treated with a shunt developed a Chiari malformation. The VAD was exposed in 4 patients with infection or wound breakdown. The LP shunt was revised in 2 patients who developed a pseudomeningocele. In 1 patient, a small bowel obstruction responded to conservative management. Seven patients had headaches despite documented normal ICP. That is, the headaches were unrelated to shunt function, and these patients were referred to a pain management clinic.

Conclusions. Lumboperitoneal shunts with programmable valves effectively controlled the outflow of lumbar cerebrospinal fluid to ameliorate the symptoms of PTC. The VAD permitted assessment of ICP and thus, indirectly, LP shunt function, and benefits outweighed risks. The programmable valve permitted cerebrospinal fluid flow to be adjusted based on patients’ clinical status and ICP to be measured by the VAD. (DOI: 10.3171/PED/2008/2/7/019)

KEY WORDS • benign intracranial hypertension • intracranial pressure monitoring • lumboperitoneal shunt • programmable valve • pseudotumor cerebri • ventricular access device

PSEUDOTUMOR cerebri is a poorly understood condition characterized by high ICP and the absence of focal neurological deficits (except for diplopia caused by abducens palsy, a nonlocalizing sign of increased ICP). Imaging studies show normal or smaller than normal ventricles and no mass lesion.10 Elevated intracranial venous pressure may be a universal mechanism underlying PTC from different causes.11

The condition demands treatment because of the danger of blindness due to papilledema. Resistance to CSF outflow is increased in patients with PTC,10 and the cerebral subarachnoid spaces are widely patent.3 Little is known about the natural history of PTC. In patients whose signs and symptoms are resistant to medical treatment, CSF diversion may be an effective treatment. Patients with PTC have been reported to respond to LP shunt treatment.25

Despite the efficacy of LP shunts in controlling PTC, there are concerns about their use in clinical practice. It is difficult to assess the function of these shunts. They work well in most patients, but a significant subset requires many shunt revisions.7,12,25 Whether the need for revision represents true shunt failure or difficulty assessing the functioning of the shunt itself can be unclear. Shunt malfunction may be associated with low-pressure headaches and may precipitate hindbrain herniation.5,17
The strategy proposed in the present paper evolved from our experiences with the management of complex shunt problems related to SVS, which we have managed using LP shunts. Both subsets of patients are notoriously difficult to assess and treat. They undergo many imaging studies and are frequent visitors to emergency rooms. The results of lumbar puncture are difficult to assess in anxious patients and particularly in obese patients who are generally the most likely to have PTC. Furthermore, accurate assessment of papilledema in these patients is complicated by preexisting optic atrophy associated with pale optic discs. We therefore evaluated the outcomes of such patients when treated using an LP shunt with a programmable valve and with a concurrent VAD.

Methods

We reviewed the records of 40 patients with PTC (31 females and 9 males; mean age 32 years, range 6–78 years) who were treated by the senior author (H.L.R.) with the concurrent use of an LP shunt with a programmable valve and with a VAD. All patients treated with shunts for PTC were included in this series. Other patients were evaluated for PTC but were found to have either obesity-related PTC and were referred for bariatric surgery (4 patients) or normal CSF dynamics and did not require treatment for PTC. All shunt-treated patients had papilledema and increased lumbar spinal pressure at the onset of treatment. The patients complained of headaches, visual obscurations, and occasional diplopia. All patients underwent retrograde venography, and venous pressure was measured in the sagittal sinus, both transverse sinuses, both sigmoid sinuses, the jugular veins, and the right atrium to determine the causes of the PTC.

Shunt Valve

The Codman Hakim Programmable Valve with Siphonguard (Codman Corp.), specially modified for this purpose, was used in this study. When used for ventricular shunt treatment, the valve is cylindrical. When we first used the valve to treat patients with SVS, we encountered 2 problems related to setting adjustment. The valve tended to rotate in the soft tissues of the abdomen, and it was often placed too deep in these tissues to allow reprogramming. The valve was therefore modified to include a flat-bottomed stage to prevent it from rotating (Fig. 1). The valve must be placed superficially to allow it to be reprogrammed. The Codman Hakim valve programmer (Codman Corp.) was used to adjust the valve. A radiograph was obtained to confirm the setting on the valve.

The Codman VPV System (Codman Corp.) permits the valve to be adjusted in an acoustic mode. Because the valve setting is displayed on a liquid display screen, a radiograph is unnecessary. In 20 patients, the valve was adjusted using the acoustic programmer.

Lumboperitoneal shunts do not siphon. The word “siphon” relates to a tubular structure with 2 arms of different lengths. The fluid-filled tube drains fluid between the higher (shorter) limb into the lower (longer) limb. Negative pressure “drains” or “sucks” the fluid out. Lumboperitoneal shunts are basically tubular structures at the base of a fluid-filled column (the intracranial compartment and the spinal subarachnoid space upstream from the insertion of the lumbar catheter). They respond to a head of hydrostatic pressure upstream from the shunt itself. The hydrostatic pressure depends on the height of the fluid column. The fluid column is much greater above the valve when the patient is standing. The Siphonguard add-on device with the valve responds to rapid changes in hydrostatic pressure by restricting CSF flow. It therefore effectively prevents overdrainage when used with LP shunts. Figure 2 illustrates the functioning of the Siphonguard device. It consists of a central chamber that
Lumboperitoneal shunt and ventricular access device

allows unrestricted flow as long as it is open. A ruby ball with a spring mechanism is proximal to the opening into this chamber. As the rate of flow increases, the ball is pushed into the channel. It then becomes occluded, forcing the CSF to flow into the high-resistance secondary pathway. In the patent it is described as a "flow-restricting device." It prevents overdrainage with little or nothing to do with siphoning. It can be placed anywhere within a shunt system and requires no specific orientation to function appropriately.

Operative Technique

The LP shunts were inserted with patients in the lateral decubitus position after induction of general endotracheal anesthesia. A midline lumbar incision was made over L3–4. A Tuohy spinal needle was introduced into the thecal sac, where the brisk flow of CSF was encountered. The lumbar catheter introduced into the spinal canal for 20 cm was directed toward the cranium. The spinal needle was withdrawn. The CSF flow was confirmed at the distal end of the lumbar tubing. Three patients with stenosis of the lumbar spinal canal required a small laminotomy for insertion of their lumbar catheter. The lumbar catheter was then connected to the modified programmable valve. An anchoring stitch was placed at the step-down connector site to prevent the shunt from migrating. The distal end of the valve was connected to the peritoneal catheter.

Before surgery the patient underwent either stereotactic MR imaging or CT scanning for frameless stereotaxy. The patient’s head was fixed in a Mayfield headholder in a lateral position. Using surface markers the patient was registered with the workstation of the frameless stereotactic unit (StealthStation Treon Plus, Medtronic Navigation, Medtronic Corp.). An insertion site was chosen in the precoronal area. The appropriate trajectory for placing the catheter into the frontal horn of the ventricle was selected using frameless stereotaxy. The Rickham reservoir (9.5-mm diameter, Codman Corp.) was affixed after the free flow of CSF was confirmed. Despite being small, the ventricle was accessed in all patients.

Follow-Up

Postoperatively, patients were followed to assess the resolution of PTC. When necessary, headaches and shunt function were evaluated by assessing ICP via the VAD. Because ventricular size does not change in these individuals, CT scans are of little or no value except immediately after surgery to assure that the catheter is positioned appropriately. Late MR imaging was performed in all patients 3–12 months after surgery and whenever the new onset of symptoms suggested that hindbrain herniation may have occurred. No patient has been noted to have developed hindbrain herniation.

During follow-up when patients complained of headaches that suggested shunt malfunction due to failure or overdrainage, the VAD was tapped with a 23-gauge needle and attached to a manometer to assess ICP. The reservoir incorporated into the valve mechanism of the system may be accessed to measure ICP. However, it is difficult to assess the possibility of diagnosing shunt overdrainage or to assess ICP in an upright position. The follow-up period ranged from 3 months to 72 months (mean follow-up 18 months).

Results

Intracranial pressure was assessed 49 times in 21 patients: once in 6 patients, twice in 6 patients, 3 times in 5 patients, and 4 times in 4 patients.

Seven patients with severe intractable headaches that did not respond favorably to shunt valve adjustments underwent chronic monitoring of ICP by a butterfly needle sewn into the reservoir and connected to a transducer.

Depending on the measured ICP, the valve was redialed in 14 patients on 28 occasions. The valve position was confirmed by obtaining a radiograph. An ICP of −5 to 5 mm Hg in the erect position and of 5 to 15 mm Hg in the recumbent position was considered normal. The valve was redialed once in 6 patients, twice in 5 patients, 3 times in 1 patient, and 4 and 5 times in 1 patient each. In patients with postural headaches, valve pressure was increased to reverse overdrainage of the CSF. When patients complained of early morning or constant headaches, valve pressure was decreased. Such patients usually had documented increases in ICP. In 3 patients exposed to a magnetic field, the valve had to be redialed due to resetting of the mechanism. Two of these patients had undergone MR imaging for an unrelated disorder, and 1 patient had undergone wand surveillance at the security gate at the airport.

On 16 occasions 10 patients required revision for shunt failure. Five patients required 1 revision, 4 patients required 2 revisions, and 1 patient required 3 revisions. The causes for revision were obstruction in 12 patients, shunt migration in 3, and valve dysfunction in 1 patient. In the latter case, the valve could not be dialed and it had to be replaced. The failure rate is comparable to the failure rate of LP shunts in general and points to the many frustrations associated with dealing with PTC. Eggengerber et al. reported that 44% of their patients with PTC did not require shunt revision, but 3 patients required a total of 35 shunt revisions. The complication rate in their study was relatively high. In our study we found a lower incidence of need for shunt revision than in other series: shunt failure was documented in 10 patients, and 4 (10%) suffered a shunt infection. However, fewer shunt revisions were necessary than in other reports. These
observations do not establish whether this technique results in fewer complications. The technique, however, does make it easier to diagnose problems. There are no longer reasons to operate on patients for overdrainage or “suspected” failure of their LP shunt.

In 4 patients in our study, the VAD had to be removed due to infection or wound dehiscence. The shunt had to be revised in 2 patients because they developed a lumbar pseudomeningocele. One patient developed an acute abdomen due to a small bowel obstruction that responded to conservative management.

After treatment of their PTC and continuous monitoring of ICP, headaches in 7 patients were found to be unrelated to ICP or shunt function. These patients were referred to our headache clinic for further assessment and treatment of chronic daily headaches. All of these patients had presented with documented intracranial hypertension and were found to have high venous pressure in the dural venous sinuses at the time of their original treatment. They required treatment for PTC but also suffered from chronic daily headaches. Chronic daily headaches, which occur in 4% of the normal population, have many causes, including migraine and medication overuse. In these patients, it is extremely important to be able to diagnose shunt failure accurately. The neurosurgeon must work with the patients’ other health care providers managing the headache disorder to prove that the headaches are unrelated to the shunt function and that the patient’s condition is not worsened by excessive use of analgesic medication.

Discussion

Pseudotumor cerebri is a syndrome of increased ICP when no intracranial mass lesion is present. It has been associated with obesity, obstruction of cerebral venous drainage, endocrine disorders, exogenous agents, infections, and other medical conditions. It is characterized by headaches, transient visual obscurations, visual loss, intracranial noises, and cognitive decline. The hallmark of PTC is papilledema, which may be asymmetric. The accepted value of CSF pressure for diagnosing PTC is >250 mm H2O in the obese and >200 mm H2O in the nonobese.

Venous hypertension has been suggested as the universal mechanism underlying PTC from different causes. The elevated sinus pressure increases the resistance to CSF absorption. The CSF pressure subsequently increases to restore the pressure gradient necessary for CSF to flow across the arachnoid villi and into the venous system. The CSF production continues despite the increasing ICP, further exacerbating intracranial hypertension. A second effect of increased resistance to venous outflow is stiffening of the brain, or an increase in brain turgor. In the presence of closed sutures, intracranial venous hypertension leads to PTC instead of hydrocephalus. This reasoning derives from observations based on the senior author’s mathematical model of ventricular volume regulation. It is also consistent with the observations of other researchers in this field.

Cerebrospinal fluid from both the ventricles and cortical subarachnoid spaces is accessed by LP shunts through the spinal subarachnoid space. Therefore, LP shunts represent an appropriate choice to ameliorate PTC, and they have been used successfully to treat PTC. However, they are associated with a high failure rate and complications that include low-pressure headaches and tonsillar herniation. It is also difficult to assess the function of LP shunts.

Patients with PTC frequently have other types of concurrent headaches that are not necessarily related to increased ICP. In 1 report of 82 patients, 68% had headaches even after their PTC was treated. Of these, 30% had episodic tension-type headaches and 20% had migraines without an aura. Migraine is an important part of the differential diagnosis of idiopathic intracranial hypertension, especially when no papilledema is present. It is a diagnosis of exclusion that requires measuring ICP. Thus headaches may persist in patients treated for PTC even after their ICP normalizes. The use of a VAD facilitates accurate assessment of ICP and thus allows rational decisions to be made about the treatment of this complicated and confusing group of patients. Because headaches in 7 of our patients were unrelated to ICP, they were referred to our headache clinic for assessment.

Disadvantages of LP Shunts

Papilledema may be difficult to assess, especially in fair-skinned individuals or in patients in whom chronically increased ICP has resulted in optic atrophy from previous episodes. The CSF pressure determined by lumbar puncture may be unreliable and is unpleasant for patients. If a patient is anxious or in pain during the procedure, which may require several attempts, ICP increases. This situation is especially likely in the morbidly obese who represent most of the PTC patients in most series. The inability to assess LP shunt function is considered a major disadvantage of their clinical use. Tapping of the reservoir incorporated into the valve mechanism allows the diagnosis of intracranial hypertension if there is no proximal obstruction. It does not allow pressure to be recorded in different positions upright and supine. We thus propose the use of a VAD to assess ICP in patients with LP shunts.

The VAD permits an easy, acceptable, and reliable assessment of ICP at the bedside, even in the pediatric population. These ICP measurements can guide the adjustment of the LP shunt valve and provide information on shunt function. Sikorski et al. reported that an adjustable shunt valve can even be reprogrammed at home in appropriately selected patients. However, comprehensive patient education and close patient-physician communication are necessary.

Most LP shunts lack valves that regulate drainage. Such shunts depend on the resistance of the draining tube and a slit at the end of the peritoneal catheter to control CSF flow. This configuration may lead to postural overdrainage. Low-pressure headaches and hindbrain herniation are directly related to such overdrainage. Wang et al. reported that overdrainage symptoms were observed in 11 of their 74 patients. Seven of these patients had LP shunts with no valve, and 4 had an HV valve (Integra Neurosurgical). Important- ly, 9 of these 11 patients’ symptoms resolved after a valve was inserted or after their valve was set to a higher pressure.

In the present study, the LP shunt was inserted with a programmable valve that permitted a controlled, individualized adjustment of outflow resistance tailored to the patient’s clinical response. The shunt also contains a device to retard siphoning; in this case, the Siphonguard. This add-on device diverts the flow of CSF into a high-resistance pathway in
response to rapid flow and therefore prevents overdrainage. Low-pressure headaches were reversed by increasing valve resistance, and high-pressure headaches were reversed by decreasing valve resistance. Thus CSF drainage was individualized, thereby effectively addressing the issues of overdrainage and low-pressure headaches.

Chumas et al.6 and Payner et al.17 alluded to the high incidence of tonsillar herniation after LP shunt treatment in the pediatric population. They studied their patients between 1974 and 1991 when T-tube and percutaneous shunts were used. These studies primarily considered the late outcomes of children who had received LP shunts at birth and in whom further brain growth could be expected.

However, none of the other studies alluding to the use of LP shunts have reported such a high incidence of herniation.4,7,22,27 Aoki2 reported a Chiari malformation in only 2 of 207 patients, including 28 pediatric patients undergoing LP shunt treatment. No patient with an HV valve system acquired a Chiari malformation or subdural hematoma.27 Wang et al.27 reported that hindbrain herniation might occur in patients treated with LP shunts. However, this complication is exceedingly rare when valve mechanisms are used. Overall, the therapeutic efficacy of an LP shunt far outweighs the risk of hindbrain herniation. We believe that this regulation of CSF flow prevents creation of the pressure gradient that is responsible for overdrainage, low-pressure headaches, and hindbrain herniations. During the mean follow-up of 18 months, none of our patients have developed hindbrain herniation.

Most revisions were related to catheter obstruction or migration. Lumboperitoneal shunts are prone to migrate from the spine or abdomen. Of our 40 patients, 10 needed shunt revisions on 16 occasions (12 obstructions and 4 migrations). Of the 74 patients reported by Wang et al.,27 44 had revisions: 27 patients had 1 revision, 10 had 2 or 3 revisions, and 1 patient had 5 revisions. The HV valve was responsible for shunt malfunctions in 9 patients. In the series reported by Eggenberger et al.,7 of 15 patients requiring revisions underwent 35 (53%) of the overall 66 shunt revisions performed in their patients. These data suggest that a subset of patients undergoing LP shunting needs multiple revisions, but the reason is unclear.

Ventriculoperitoneal shunts that access the cerebral ventricles are placed into small ventricles. When drained, the shunted ventricle becomes smaller than the contralateral ventricle. The brain and septum pellucidum tend to coapt around the catheter, leading to intermittent or complete obstruction of the ventricular system.23 No studies have compared the reliability of LP to that of VP shunts. However, intermittent or complete proximal obstruction of the ventricular catheter is a common finding in patients with SVS.20 The LP shunts are placed in an area with a reliable reservoir of CSF. Their caliber is small, and they may have a tendency to fracture, to pull out, or to obstruct with kinking. However, nothing is present in the lumbar theca to collapse around the catheter. The CSF does not need to flow upstream into the ventricle to be removed through the shunt system. Compared with VP shunts, LP shunts are associated with a lower incidence of serious complications such as subdural hematomas and infection.27 Lumboperitoneal shunts maintain the patency of all CSF spaces. In contrast, VP shunts occasionally collapse the drained ventricle and thereby lead to shunt malfunction.

Conclusions

The concurrent use of an LP shunt with a programmable valve with a VAD is a safe and effective procedure for the treatment of PTC. When a programmable valve is used, the rates of shunt failure and of overdrainage of CSF are low. A VAD permits a reliable assessment of ICP that reflects not only control and resolution of PTC but also function of the LP shunt. A similar approach with the insertion of a VAD may be used to treat conditions other than PTC that warrant the use of an LP shunt.

Disclosure

Dr. Rekate occasionally consults with Codman Corporation on the development of devices for the treatment of hydrocephalus. Dr. Rekate consulted on the design of the valve system but receives no reimbursement for its use.

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