Severe intracranial hypertension in slit ventricle syndrome managed using a cisterna magna–ventricle–peritoneum shunt

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Object. Severely increased intracranial pressure (ICP) can be life threatening in patients who had previously undergone shunt treatment but who do not experience ventricular enlargement. The authors analyzed the utility of placing shunts into the cisterna magna concurrently with ventricular shunts in patients who were not candidates for lumbo-peritoneal (LP) shunt placement.

Methods. Ten patients treated with cisterna magna–ventricle–peritoneum (CMVP) shunts for complex problems of shunt function were reviewed retrospectively. All patients had documented increases in ICP and ventricles that did not expand despite life-threatening increases (> 80 mm Hg in one case) in ICP.

Between 1995 and 2003, 10 patients (four males and six females, age range 4–32 years) were identified as having life-threatening increases in ICP despite small or slit-like ventricles on imaging studies. Each episode was documented with intraparenchymal pressure monitoring. All patients had documented ventricular catheter failures at the time of the intervention, and all had undergone at least one previous attempt to treat the condition with a valve upgrade and replacement of the ventricular catheter. Three patients had achondroplasia, four had spina bifida, and three had a pre-existing Chiari malformation Type I.

All patients improved after the procedure, and none suffered permanent complications. For at least 48 hours after surgery, all patients underwent intraparenchymal monitoring of ICP (an intraparenchymal monitor was used that documented normal ICP).

Conclusions. The CMVP shunts are an excellent option for patients who are not candidates for LP shunts but who have high ICP and ventricles that do not enlarge at shunt failure. The ability to access the spinal fluid in the cortical subarachnoid space presumably accounts for this success.

KEY WORDS • hydrocephalus • cisterna magna • subarachnoid space • slit ventricle syndrome • achondroplasia • Chiari malformation • pediatric neurosurgery

The term SVS has been applied to a diverse group of pathological entities that share the common features of severe intermittent headaches and small ventricles evident on imaging studies. The most troubling form of this condition, termed NVH, involves severely increased ICP without ventricular dilation at the time of shunt failure. Normal-volume hydrocephalus does not occur when hydrocephalus develops in adulthood, and it is limited to late complications of shunt placement in patients who are treated in infancy. Authors of recent studies have emphasized the value of LP shunt treatment for this difficult condition. Some patients with the most refractory forms of NVH are not candidates for LP shunt placement because of their unique anatomy. This group includes patients with hydrocephalus related to achondroplasia due to severe spinal stenosis, those with spina bifida related to the CM-II and a tethered spinal cord, and those with preexisting CM-I, all of which increase the probability of worsening symptoms. We discuss the use of concurrent ventricular and cisterna magna–peritoneum shunts in the management of severe forms of SVS.

Clinical Material and Methods

Patient Selection

All patients discussed in this retrospective study were treated by the senior author (H.L.R.) between 1995 and 2003. Ten patients (four males and six females, age range 4–32 years) were referred with intermittent severe headaches...
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or recurrent failure of their ventricular catheter (Table 1). One of these patients has been reported on elsewhere. In all patients, very high ICP had been documented at shunt failure with no radiographic evidence of ventriculomegaly or increased ICP. Their ICP was measured using an intraparenchymal transducer (Codman Corp., Raynham, MA) inserted through a separate bur hole. When the ventricular catheter is obstructed, monitoring ICP through the shunt reservoir is not feasible because of the likelihood of intermittent plugging of the ventricular catheter or restriction of proximal flow to the reservoir. In all patients, several attempts to treat the condition by upgrading the opening pressure of the valve and using devices that retard siphoning had been unsuccessful.

Seven patients had conditions that we thought precluded placement of an LP shunt (four with spina bifida and three with achondroplasia). Three had conditions that we considered to be relative contraindications to placement of an LP shunt due to the concurrence of NVH with a significant CM-I. Two of these patients had undergone previous attempts at CM decompression, but their increased ICP persisted after the decompression.

Surgical Technique

Our surgical technique has evolved over time. Patients were placed prone in a three-pin headholder oriented vertically so that the occipitocervical region and the shunt system could be incorporated into the same operative draping. Their bodies were taped firmly to the table to allow maximum rotation of the table from side to side.

A standard occipitocervical decompression was performed, and the C-I arch was removed. The bone removal was continued 2 cm above the foramen magnum in patients with a CM-I or achondroplasia. In patients with a CM-II (that is, those with spina bifida), no bone was removed from the occiput, and the entire procedure was performed in the cervical spine, usually with laminectomies that extended to C-3.

The dura mater was opened vertically beginning in the cervical canal. The opening was continued proximally to ensure control of the marginal sinus. A patch of bovine peri-cardium was used to create a copious new cisterna magna. To maximize the size of the cisterna magna for the drainage procedure, we placed a titanium plate across the occipital defect (expansion cranioplasty) and used central tuck-up sutures to hold the patch against the expanded roof of the posterior fossa (Fig. 1). The lack of bone removal in the patients with spina bifida precluded this degree of expansion of the cisterna magna. In the first patient suffering from achondroplasia, the posterior fossa expansion cranioplasty was performed using split skull from the craniotomy. Since that procedure, we have favored the use of the titanium plates because they are easy to insert.

In the first two patients, we inserted ventricular catheters into the newly created cisterna magna. In each case, we found that the stiff catheter caused pain because it rested on a cervical nerve root. After these experiences and because we had positive experiences using LP catheters to place shunts in isolated fourth ventricles, we used pieces of the small-diameter tubing to cannulate the new cisterna magna. Once the catheter had pierced the patch, a stepdown connector was used to splice the small tubing to standard shunt tubing. The ventricular shunt was then explored. A Y or T connector was used to splice the cisterna magna catheter to the existing shunt system proximal to the valve. This system ensured that pressure in the ventricle and cortical subarachnoid spaces was balanced.

Results

A few weeks after the procedure, one patient needed a revision of both proximal catheters due to an infection. Three patients required a single revision of the cisterna catheter because of nerve root irritation. In the first two patients, the problem was related to the use of standard ventricular catheters. In the third patient, a previous infection and arachnoiditis had caused the lumbar catheter to curl onto a cervical nerve root, causing shoulder pain. In the last seven patients, this problem has not occurred (range of follow up 6 months–5 years). All patients have working shunts, and their increased ICP resolved without further need for revision. There were no permanent complications related to the procedure.

Two of the 10 patients continue to have severe headaches after successful placement of the CMVP shunt. In these cases, a shunt tap was performed to guarantee the free flow of CSF into the tapping reservoir. An intravenous butterfly needle was inserted into the reservoir, affixed to the scalp, and attached to a fluid-coupled pressure transducer to record ICP overnight. Once the cisternal catheter is in place, the free flow of CSF into the reservoir is ensured. The brain does not collapse around this catheter. In contrast to the situation before the cisternal catheter is added, the fluid-coupled monitoring of ICP is reliable. During the recording, iohexol-180 (3–5 ml) was injected into the reservoir and the patient underwent CT scanning within an hour. If the ventricles and upper cervical subarachnoid spaces were in communication with the shunt system and pressure was within normal limits, no further intervention was indicated. Normal ICP was assumed to be between 5 and 15 mm Hg in the recumbent position and between −5 and 5 mm Hg while standing. We use a programmable valve system (Codman-Hakim programmable valve with Siphonguard; Codman Corp.) that allows multiple steps so that this level of pressure is attainable in almost all patients.

Illustrative Cases

Case 1

This 20-year-old woman was born with a thoracic myelo-
meningocele, CM-II, and hydrocephalus that was treated at birth. She also had undergone multiple shunt revisions and syringosubarachnoid shunt and orthopedic spinal stabilization procedures. On this occasion, shunt failure was associated with progressively increasing headaches and pain along the shunt track. On examination, she was obese, paraplegic, and wheelchair bound, with severe scoliosis. She had tenderness and swelling along the shunt track. A CT scan showed mild ventricular dilation. At surgery, the shunt tubing was found to be disconnected, and the entire shunt was replaced. Her symptoms were relieved immediately.

Four days later, the patient presented with an altered sensorium. A CT scan showed no significant change in the size of the ventricles. An intraparenchymal pressure monitor placed through a right frontal bur hole showed that her ICP was as high as 60 mm Hg. The ventricular catheter was replaced with an external ventricular drain. Despite evidence of CT scans that the ventricular catheter was positioned appropriately, her ICP remained high, and drainage of CSF was minimal. A CT cisternogram showed that ventricular CSF flowed freely into the subarachnoid space of the upper cervical spine (Fig. 2). Magnetic resonance imaging confirmed slit-like ventricles. A CM-II was associated with brainstem compression at the foramen magnum. The hindbrain hernia extended to C-3. Magnetic resonance venography showed the absence of both transverse sinuses, the termination of the superior sagittal sinus, and its connection to the vertically oriented straight sinus at or below the foramen magnum.

The patient underwent decompression of the CM-II through a C1–3 laminectomy. All adhesions in this area were lysed. The brainstem was detethered from the dura. The fourth ventricle was opened widely by excision of a piece of cerebellar vermis. A lumbar catheter was passed into the spinal subarachnoid space or newly created cisterna magna. Duraplasty was performed using bovine pericardium. A concurrent shunt was placed, and her ICP normalized immediately. The patient recovered uneventfully and has done well for the past 3 years.

**Case 2**

This 16-year-old boy suffering from achondroplasia, hydrocephalus, and a seizure disorder had undergone multiple shunt revisions and surgeries related to his achondroplasia, including four previous cranial expansion operations. An LP shunt placed after an earlier thoracolumbar decompression for spinal stenosis had failed. Venography showed severe bilateral stenosis of the jugular foramen, and the microcatheter could not be negotiated into the intracranial circulation. Expansion of the jugular foramen through a transmastoid approach was attempted. An attempt to place an endovascular stent to bypass the vascular constriction failed, as well as an attempt to bypass the obstruction using a saphenous vein graft. Severe spinal stenosis and multiple spinal corrective procedures precluded managing this severe case of SVS with an LP shunt. On at least four occasions, the boy’s ICP was confirmed to be higher than 50 mm Hg at the time of shunt malfunction.

The foramen magnum was expanded with a split skull cranioplasty to create a cisterna magna where none had previously existed. A lumbar catheter was inserted into this space and spliced into the existing ventriculoperitoneal shunt proximal to the valve. This system has functioned without revision for 5 years.

**Discussion**

Increased pressure in the dural venous sinuses leads to a condition called PC or to benign intracranial hypertension. The mechanism may be universal in the development of PC.3 This increase in sinus pressure has two effects on the dynamics of ICP. First, it increases the volume of intrapar-
enchymal veins, increasing the turgor of the brain and decreasing its compressibility. Second, it increases the pressure that must exist within the intracranial compartment to absorb CSF, leading to distension of the CSF compartments, particularly the cortical and spinal subarachnoid spaces. As described here, PC is often treated by placement of an LP shunt. This shunt accesses all CSF compartments, including the spinal and cortical subarachnoid spaces, and has the advantage that the thecal sac is a fairly fixed volume that cannot collapse around the catheter.

The situation in infants is quite different. Infants have open fontanelles and sutures that are capable of expanding. Decreasing cerebral venous return in infants does not lead to PC, which would probably be difficult if not impossible to identify. Rather, it leads to hydrocephalus. The increase in sagittal sinus pressure impedes CSF absorption. Intracranially, however, pressure cannot rise significantly because of the open sutures. Consequently, CSF backs up in both the cortical subarachnoid spaces and the ventricles. In this context, the condition may or may not be distinguishable from hydrocephalus related to intraventricular obstruction of CSF flow or from intraventricular obstructive hydrocephalus. After a shunt is inserted, the ventricles return to a normal or subnormal size, the fontanelles close, and the skull loses its distensibility. What remains is a drained ventricular system, a brain with increased stiffness or turgor, and distended cortical subarachnoid spaces.

There are only two ways that CSF in the cortical subarachnoid spaces can be absorbed. It can flow upstream through the ventricular system and out through the ventricular catheter and the shunt. Alternatively, it can be absorbed by bulk flow into the venous sinuses or through other pathways of CSF absorption. For CSF to be absorbed into the dural venous sinuses, the ICP must be 5 to 7 mm Hg higher than the sinus pressure. If sagittal sinus pressure is greater than the opening pressure of the ventricular shunt valve, the ventricle containing the shunt must empty of CSF completely. The ventricular catheter is thus blocked before CSF pressure can become high enough to lead to absorption from the cortical subarachnoid spaces, which distend and push the brain inward, causing further malfunction of the shunt system. Attempting to manage PC as it occurs in adults or the related condition found in older children or adults who received a ventricular shunt in infancy requires that the opening pressure of the shunt system remain higher than the absorption pressure of the cerebral venous sinuses.

In this situation, a ventricular shunt will remain open as long as flow from the cortical subarachnoid space to the ventricle is unimpeded. Two important pieces of information imply that retrograde flow may be impaired from the subarachnoid spaces to the ventricle. The first relates to the pharmacokinetics of antibiotic and chemotherapeutic agents that are injected intrathecally. It is often difficult to obtain bacteriocidal levels of these antibiotics within the ventricles. Consequently, some pharmacologists suggest that some of these drugs should be injected intraventricularly. Second, the difficulty of flow upstream into the ventricle relates to a phenomenon called “postshunt ventricular asymmetry.” If the septum pellucidum is intact, drainage of one lateral ventricle can cause it to collapse against the head of the caudate nucleus, closing the foramen of Monro and leading to measurable pressure changes between the two sides.

We cannot say with certainty that all patients with NVH who exhibit this severe form of SVS harbor intracranial venous hypertension; however, when we have looked for evidence of venous hypertension by retrograde venographic pressure recording in these patients, we have always found it. Furthermore, the severity of the condition has related to the severity of the increase in venous pressure. In the patient in Case 1, who had a CM-II, pressure in the transverse sinuses was related to the low position of the torcular herophili, as is common in this situation. Intracranial venous hypertension, as a cause of hydrocephalus in infants, has been studied thoroughly by several authors. Sainte-Rose, et al., successfully treated infantile hydrocephalus using a bypass from the transverse sinus to the jugular vein. Likewise, other authors have shown that the cause of hydrocephalus in achondroplastic dwarfs is stenosis of the jugular foramina. We have reported the same situation in the context of complex craniofacial anomalies such as Crouzon syndrome.

Some authors have recommended cranial expansion in...
such patients. Between 1985 and 1995, we used this strategy to treat seven patients. In none of them was the procedure effective for more than 24 months, and a new intervention eventually followed. One of the current patients with achondroplasia had been referred to our center after undergoing four craniotomies to increase the size of the head. Each procedure was successful for a limited period. After the insertion of a CMVP shunt, however, his symptoms resolved completely and his condition improved permanently.

Based on this analysis, severe cases of SVS and NVH are best managed by shunt systems that access the cortical subarachnoid spaces. Insertion of a shunt leading directly to the cortical subarachnoid spaces is feasible, but these shunts are seldom reliable because the brain’s cortical surface will collapse around the ventricular catheter. Lumboperitoneal shunts are seldom reliable because the brain’s cortical surface will collapse around the ventricular catheter. Lumboperitoneal shunts are ideal as long as there is no element of intraventricular obstructive hydrocephalus (which is exceedingly rare, except in the context of severe late shunt infections). Direct shunts to the pleura and the peritoneum have been described by other authors. There is no compelling reason to place shunts in the ventricle as well as the subarachnoid spaces when there is free communication among those spaces, as documented by iohexal cisternography in seven of our 10 patients. Nonetheless, some practical concerns remain. Placing catheters in the cisterna magna, within the fourth ventricle, or within cysts of the posterior fossa leaves the connection to the valve mechanism below the nuchal line. This configuration places considerable stress on the shunt when the neck is moved. Repeated fractures of shunt systems in this location are common. For patients with PC who require a cisternal shunt (that is, with a coexistent CM-I and no hydrocephalus), we recommend tunneling the tubing from the cisterna magna to the skull well above the nuchal line and the use of a reservoir in that location to test the system. The valve should then be added distally to ensure that it and all connectors are located above the nuchal line.

Conclusions

In patients with severe problems of increased ICP in whom the ventricles do not expand, shunt management often requires drainage of CSF from the cortical subarachnoid space. Experience with PC has shown that LP shunts are the best solution. In patients in whom this option is unavailable, CMVP shunts may be the ideal solution.

References


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