Childs Nerv Syst (2008) 24:423–430
DOI 10.1007/s00381-008-0579-7

REVIEW PAPER

Shunt-related headaches: the slit ventricle syndromes

Harold L. Rekate

Received: 13 November 2007 / Published online: 8 February 2008

Abstract
Purpose The purpose of this work is to review the pathophysiology and treatment of severe headache disorders in patients having a shunt for hydrocephalus.
Materials and methods The literature on the management of the slit ventricle syndrome is reviewed as well as an assessment of personal experiences over a 30-year period in the management of severe headache disorders in shunted patients.
Results If the slit ventricle syndrome is defined as severe, life-modifying headaches in patients with shunts and normal or smaller than normal ventricles with ventricular shunts for the treatment of hydrocephalus, there are five different pathophysiologies that are involved in the process. These pathologies are defined by intracranial pressure measurement as severe intracranial hypotension analogous to spinal headaches, intermittent obstruction of the ventricular catheter, intracranial hypertension with small ventricles and a failed shunt (normal volume hydrocephalus), intracranial hypertension with a working shunt (cephalocranial hypertension), and shunt-related migraine. The treatment of these conditions and identifying patients with each condition are facilitated by attempting to remove the shunt.
Conclusions Following the analysis of attempts to remove shunts, there are three possible outcomes. In about a quarter of patients, the shunt can be removed without having to be replaced. This is most common in patients treated in infancy for post-hemorrhagic hydrocephalus or patients shunted early after or before brain tumor surgery. Another half of patients have increased intracranial pressure and enlarged ventricles. In these patients, there is an 80% success rate for endoscopic third ventriculostomy. Finally, the most severe form of the slit ventricle syndrome involves intracranial hypertension without ventriculomegaly, which is managed optimally by shunt strategies that emphasize drainage of the cortical subarachnoid space such as lumboperitoneal shunts or shunts that include cisterna magna catheters.

Keywords Shunt · Headaches · Hydrocephalus · Slit ventricle syndrome

H. L. Rekate
Pediatric Neurosciences, Barrow Neurological Institute,
St. Joseph's Hospital and Medical Center,
Phoenix, AZ, USA

H. L. Rekate (✉)
Neuroscience Publications, Barrow Neurological Institute,
350 West Thomas Road,
Phoenix, AZ 85013-4496, USA
e-mail: harold.rekate@bnaneuro.net

Introduction

Headaches are one of the most common afflictions of mankind. Based on large series of patients, about 4% of the adults in the world suffer headaches everyday, with a female-to-male ratio of 2.5:1 [1]. A larger but inestimable number of individuals have occasional incapacitating headaches [2]. Not surprisingly then, patients with shunts have headache disorders. The presence of the shunt in patients with headaches always leads to the assumption that something is wrong with the shunt. This assumption can lead to large numbers of expensive and possibly dangerous imaging studies, long waits in emergency rooms, and the expenditure of considerable money when the headaches are a chronic condition. There is always the possibility that patients could die or develop severe neurologic dysfunction from high intracranial pressure (ICP) at the time of shunt failure. Medically, therefore, it is reasonable to ascertain

that their shunt is working. Doing so, however, is not always straightforward.

Since their development in the 1950s, valve-regulated shunts have probably saved more lives and more cognitive function for more years than anything else neurosurgeons have ever done [3, 4]. However, shunts have led also to a great many problems that were not easy to predict from the beginning. The most common and most chronic of these newly recognized conditions is the association of chronic headaches with the presence of a shunt. Severe headache disorder in patients with shunts and small ventricles has been called the "slit-ventricle syndrome" (SVS) [5]. SVS is not a single condition; rather, several different pathophysiologies can underlie this constellation of findings [6]. This article defines the various causes of severe headaches in shunted patients, suggests an algorithm for the diagnosis and treatment of this common condition, and suggests a management approach to patients with shunt-related problems.

Classification

As shunting of the ventricles to the jugular vein or peritoneum became standard treatment for hydrocephalus, problems related to overdrainage were recognized with increasing frequency [7]. Before valve regulation of cerebrospinal fluid (CSF) flow was developed, drainage of the lateral ventricles in children with hydrocephalus was precluded by the routine collapse of the brain with associated lethal subdural hematomas. The development of valve regulation made it possible to treat severe hydrocephalus successfully with much less likelihood of this dreaded complication.

By the mid-1970s, problems related to the ventricles becoming too small, leading to intermittent or recurrent obstruction of the ventricular catheter, was recognized as a severe problem. Several strategies, including subtemporal decompression for the management of this condition, were introduced [8–10]. In an early report on our experiences with this condition, we defined SVS as a triad involving intermittent headaches lasting 10 to 90 min, small ventricles on imaging studies, and slow refilling of the pumping mechanism of the valve [5]. In this same article, we demonstrated that it was possible to increase the volume of the lateral ventricles by using valve upgrading and a device that retards siphoning (DRS). We also recommended that the use of low-pressure shunts should be abandoned unless dictated by specific indications.

As experience grew, especially with the ability to track changes in ventricular size using contemporary neuroimaging such as computed tomography (CT) and magnetic resonance imaging (MRI), it became obvious that this view of the SVS was too simplistic and that there were multiple different forms of the problem. Reports dealing with SVS were not describing the same condition [11]. In 1993, we published our experiences with ICP monitoring in patients with small ventricles and severe headaches. We identified five distinct pathophysiologies for this condition. There is some overlap, but each condition requires specific treatment paradigms [6]. Based on our previous study, it has been our policy to upgrade the valve and include a DRS in all patients. All patients studied had previously undergone the procedure [5].

All shunt operations are associated with some risk, especially the risk of infection. Therefore, we require a significant discussion with all patients and if children, their parents, to determine whether to intervene. We intervene surgically only in patients whose headaches significantly interfere with normal life. If children have to leave school or adults need to leave work or discontinue working more than twice a month, we believe that surgical intervention is justified.

There is little consensus about the definition or causes of headaches in shunted patients. Most such patients undergo a valve upgrade and incorporation of a DRS. About one in five of these patients, however, does not improve or improves only temporarily. Before further intervention is pursued, the causes of the patient's headaches and the relationship of the headaches to shunt function and ICP must be understood fully. To understand the causes of the headaches, it is essential to define the relationship of ICP and shunt function to the headaches. Based on chronic monitoring of ICP in these patients with headaches, we have defined five syndromes of shunt-related headaches. Each syndrome leads to specific treatment strategies [6].

Intracranial hypotension

These patients develop severe headaches that are not present while they are reclined in bed. The headache develops later in the day and gets worse with time as patients maintain an erect position. The headache improves rather rapidly if patients can lie down. Monitoring shows significantly subnormal ICP. We have recorded ICP from −25 to −30 mmHg. These headaches are analogous to postlumbar puncture headaches and may be associated with enhancement of the meninges on contrast-enhanced imaging studies.

This condition implies that the DRS has failed and that the patient will respond to replacement of the valve mechanism with insertion of an effective DRS and possibly a valve upgrade. Since the publication of the article [6], we now use a programmable valve. We prefer to use the Codman Hakim Programmable Valve with Siphonguard™ (Codman Corporation, Raynham, MA). This valve does not

depend on a diaphragm mechanism. Therefore, it can be placed anywhere along the course of the shunt. The skin does not have to move freely. Other programmable or adjustable valves are commercially available.

Intermittent proximal obstruction

This problem is probably the most common of the five conditions and represents patients who were originally described as having SVS. Monitoring shows that patients with this condition have normal to low ICP most of the day, but their ICP increases suddenly with activity. As their ICP increases, the headache worsens until the ventricular catheter reopens and the pressure reverts to normal again. These patients are also managed by placement of a DRS and valve upgrade. In chronically shunted individuals, proximal shunt failure is the most common form of mechanical failure of shunts. For decades, my policy has been to assume that this condition is a result of over-drainage of CSF and collapse of the ventricular walls around the ventricular catheter. Therefore, it is a sign that the back pressure or opening pressure of the valve is inadequate to maintain CSF within the ventricular system. I believe that the valve should be upgraded and a DRS should be incorporated into the system any time a proximal obstruction occurs.

Shunt failure without ventricular enlargement

Engel et al. [12] originally described intracranial hypertension with nondistending ventricles, so-called normal volume hydrocephalus (NVH). This enigmatic condition has been studied intensively. It is the most important and difficult to manage of the subtypes of SVS. In the series of Engel et al. [12], the patients were found to have signs and symptoms of increased ICP with no enlargement of the ventricular system. They recommended exploration of these shunts, which were routinely found to be nonfunctional.

These patients become symptomatic with progressive symptoms of increased ICP with morning headaches progressing to all-day headaches, papilledema, visual loss, and diplopia. If the condition is not treated early, neurologic deterioration is possible and blindness is likely. This problem occurs rarely, if at all, in patients who develop hydrocephalus beyond infancy, but it is a common problem in cases of congenital hydrocephalus [13].

These patients begin life with hydrocephalus but when older have pseudotumor cerebri. We performed retrograde venographic measurements of sagittal sinus pressure in five such children. All had elevated venous sinus pressure, as have all patients with pseudotumor cerebri that we have tested [14]. The management of this condition is considered below. In general, however, a shunting strategy that incorporates drainage of the subarachnoid space, such as lumboperitoneal shunts or shunts from the cisterna magna, is needed to manage these patients adequately [15–17].

Increased ICP with a working shunt: cephalocranial disproportion

Based on shunt flow studies and surgical exploration, these patients have working shunts but show significant signs of increased ICP. In my experience, this problem has been universally associated with hindbrain herniation (Chiari I malformation) and found exclusively in patients with craniofacial disorders such as oxycephaly, Crouzon's and Pfeiffer's syndrome (Fig. 1). Other authors have postulated that sutural closure results from decreasing ICP and from insufficient room for the growing brain [18].

With neither a significant abnormality of the shape of the skull and face nor the presence of hindbrain herniation, the problem in these patients is likely NVH and not cephalocranial disproportion. It is best managed with shunts that incorporate the subarachnoid space. Patients with true cephalocranial disproportion need a cranial expansion procedure or large subtemporal decompression. Further manipulation of the shunt is of no benefit. Hindbrain herniation in these patients can be dealt with effectively by enlarging the posterior hemicranium [19].

Shunt-related migraine

Headaches are common in the general population. Patients with shunts can have migraines or other headache disorders that are unrelated to their shunt. Shunt-related migraine usually occurs in the context of a strong family history of

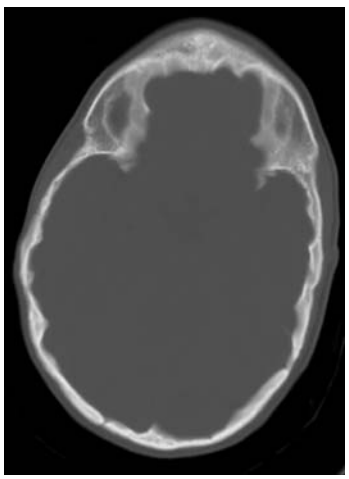

Fig. 1 CT scan of a patient with syndromic craniosynostosis and hydrocephalus showing scalloping of the inner table of the bone due to cephalocranial disproportion

migraine and is episodic. As small children, these patients usually suffer from seasonal allergies. Their descriptions of their headaches may or may not be typical of migraine.

These patients often improve briefly after shunt manipulation, but the same problems return rather quickly after intervention. Because of the potential medicolegal problems associated with failure to diagnose a shunt malfunction, these patients have had many visits to the emergency room and a large number of CT scans (because these episodes are usually considered emergencies) and other diagnostic studies. Management of these patients is complicated and frustrating. The patient, family, and neurology staff are often reluctant to believe that the headaches are unrelated to the shunt. Therefore, considerable energy and financial resources are necessary to treat these patients. Documentation by ICP monitoring is often needed to prove that the headaches are unrelated to the shunt before a commitment to medical management can be reached [6].

Shunt-removal protocol

Fortunately, most shunted patients do well for long periods. After the first few years, they may live decades with little or no need for intervention regardless of the type of shunt used or the pressure setting. Unfortunately, this statement is not true for a small percentage of shunted patients who require frequent shunt revision and who experience major interference with their function in daily life. Although most shunted patients have radiographic slit ventricles after years of shunting, only a small number have the symptoms of SVS [20].

My practice includes a large number of patients referred from outside my normal catchment area. Therefore, it is difficult for me to predict the actual incidence of symptomatic SVS. Based on my practice, however, I would predict that a third of infants followed for more than 5 years will have a severe chronic headache disorder that requires intervention. At least 20% will have ventricles that do not expand at the time of shunt failure (NVH). This second percentage is supported by information from the Division of Neurosurgery of the Children's Hospital of Los Angeles (McComb, JG, personal communication, 2000). At the time of this writing, I am working with the Hydrocephalus Association, which is maintaining a database of shunted patients, to define the scope of the problem. Regardless, severe and activity-limiting headaches are common among shunted patients.

As evident from the above classification of the pathophysiology underlying headaches in these patients, their management is time-consuming, frustrating, expensive, and potentially dangerous. As stated, the first step is to use programmable shunts that incorporate a DRS. In my opinion, the prevalence of this problem justifies the routine use of these devices. As Aschoff noted, avoiding one shunt revision would justify implanting a shunt that cost $50,000 [21].

What are the alternatives for patients with a valve upgrade and incorporation of a DRS who are still incapacitated by headaches? Of the patients described in the discussion of the classification, a specific therapy is available only for those with cephalocranial disproportion: cranial expansion. Before undertaking this procedure, we would always recommend a trial of ICP monitoring to document the problem. MRI often shows a hindbrain herniation in such patients (Fig. 2). These patients require cranial expansion. We agree with Di Rocco that occipital expansion is likely to treat both the cephalocranial disproportion and the hindbrain herniation if the bone around the foramen magnum is removed at the same time [19].

If adequate decompression cannot be achieved from above, occasionally, it is necessary to explore the craniovertebral junction directly. The patients with complex craniofacial abnormalities often have an associated set of abnormalities of venous outflow from the intracranial venous sinuses and compression of the jugular foramen. It is essential to obtain either angiographic images or MR venograms to ascertain that this flow is not interrupted (Fig. 3). It is possible that a very large percentage of venous flow is through emissary veins and may need to be saved [22, 23]. These venous anomalies can lead to a pseudotumor-like picture, and they can coexist with NVH as discussed below.

We have developed an algorithm for diagnosis and treatment of all other patients with SVS who do not improve after a valve change. This strategy is an attempt to improve understanding of the pathophysiology and treatment of debilitating headaches in individual patients (Fig. 4). After prolonged discussions with patients and family, if appropriate, patients are offered the "shunt removal protocol [24]." This procedure has evolved over time.

In general, after an informed consent is obtained, the patients undergo surgery to have their entire shunt system

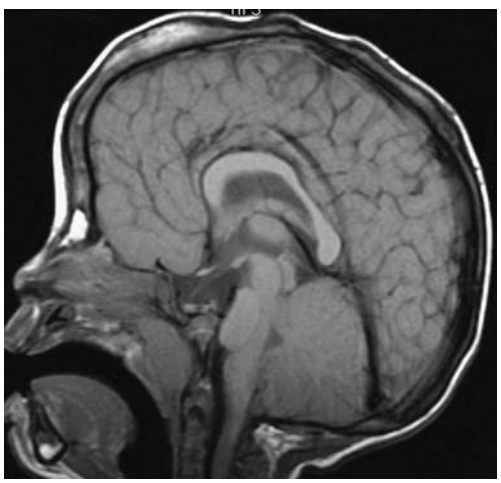

Fig. 2 MRI of brain showing hindbrain herniation

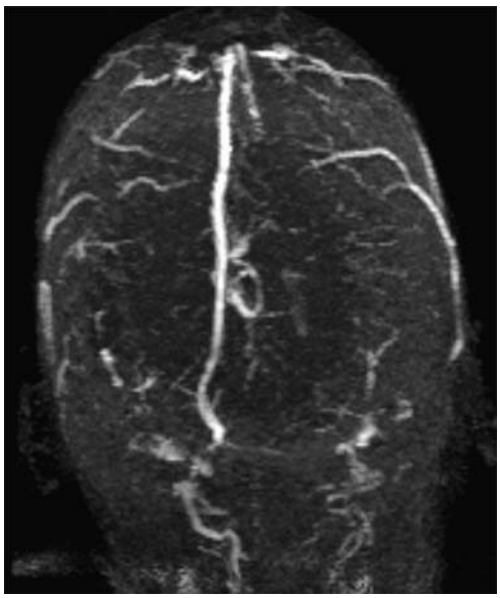

Fig. 3 MR venography of patient with normal volume hydrocephalus showing abnormalities of venous drainage leading to late pseudotumor syndrome in NVH

removed and replaced with an external ventricular drain (EVD). After recovering from anesthesia, the patients are taken to the intensive care unit where the EVD is used to monitor ICP and to drain CSF if needed. Initially, the drain is left open at 25 cm H_2O above the midposition of the head as long as the setting is tolerated. All participants, the patient, the nurses caring for the patient in the ICU, the family, and the neurosurgical residents are informed of the goals of the procedure and of the parameters for management of the EVD.

The next morning, a scan is obtained to determine whether the ventricles have enlarged. If so, this form of treatment is continued another 24 h. If not and the patient is only mildly ill from increased ICP, the drain is closed under careful observation in an attempt to increase the size of the lateral ventricles. If the ventricles have enlarged significantly on the third hospital day, the patient returns to the operating room for an endoscopic third ventriculostomy.

We have identified three potential outcomes associated with closure of the drain. In the best scenario, the ventricles enlarge only slightly, ICP normalizes, and the patient is essentially asymptomatic. This hoped-for result has occurred in about 25% of our patients. These patients have usually undergone resection or treatment of a brain tumor or have experienced a subarachnoid or intraventricular hemorrhage. In these cases, ICP is monitored for 48 h and the drain is removed. The patient remains in close contact with our service and undergoes follow-up scans 6 weeks and 1 year after the procedure. These results support

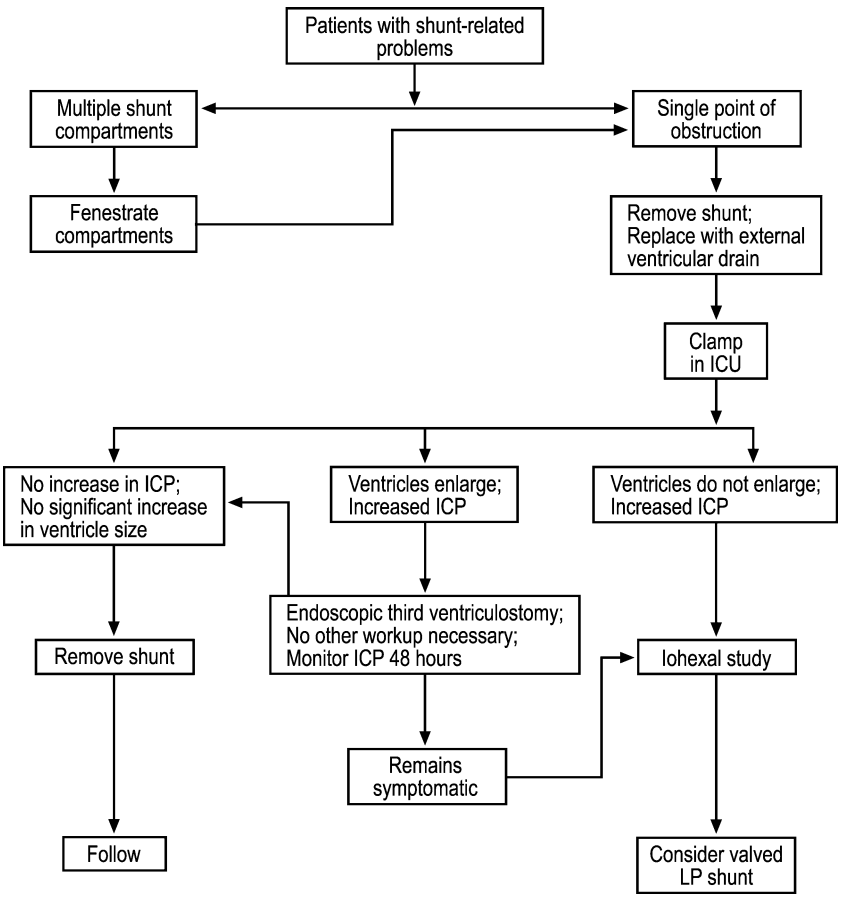

Fig. 4 Algorithm for managing shunt-related difficulties with a shunt removal protocol (with permission from Barrow Neurological Institute)

findings from our previous study on the possibility of shunt-independent arrest of hydrocephalus [25].

The second possibility is that the ventricles expand and the patient becomes ill. If the patients with a coexistent myelomeningocele (Chiari II malformation) are excluded, these patients are excellent candidates for endoscopic third ventriculostomy. The success rate for shunt-independent arrest of the hydrocephalus is 80%.

It is essential to ascertain that patients who have had chronic headaches for a long time are safe and that they have no recurrent increase in ICP. Consequently, over time we have tended to leave a ventricular access device or tapping reservoir in place after the ETV has been inserted. We affix a butterfly needle into the reservoir for ICP monitoring for 48 h. If patients return with headaches, the needle remains in place for later assessment of ICP and to inject contrast to ensure that the stoma is open. Using this protocol, we have had only one late failure, which occurred 1 year after the procedure. Reexploration of that patient revealed that the basilar artery had herniated through and sealed the stoma. A second hole was made anterior and lateral to the artery, and the patient's symptoms resolved.

The third possibility is that the ventricles do not expand, ICP increases, and patients seem ill. Such patients have NVH and are not candidates for ETV. There are two reasons why ETV is inappropriate in this condition. The first reason is practical. Even with the use of frameless stereotaxy, it is difficult to manipulate an endoscope within such small ventricles without potential damaging important structures.

The second reason relates to the underlying origin of the hydrocephalus. In our experience, all patients in this series obviously had hydrocephalus during early infancy, when their first shunts were placed. Based on retrograde venous manometry performed in several of these patients, their sagittal sinus pressure is higher than normal. The ventricles have failed to expand in more than 30% of patients undergoing the protocol. It also has occurred in at least 20% of patients with hydrocephalus related to a Chiari II malformation. Consequently, we no longer consider these patients as candidates for the shunt-removal protocol.

Several scenarios frequently result in NVH and respond in this way to attempts at shunt removal. The first group begins life as premature infants and is in the neonatal ICU for prolonged periods. Their hydrocephalus has been attributed to intraventricular hemorrhage, but venous studies reflect abnormal venous drainage either from congenital anomalies or chronic central lines associated with stenosis of the jugular veins or superior vena cava.

The other group is usually diagnosed with congenital aqueductal stenosis. Triventricular hydrocephalus is diagnosed when their heads are discovered to be enlarging and crossing percentile lines. Developmental assessments in these babies are normal, except for some delays in gross motor behavior related to the relatively large head. Later in life at the time of shunt failure, these children develop the signs and symptoms of increased ICP but without ventriculomegaly. At this point, MRI shows open flow through the aqueduct of Sylvius. In such cases, the hydrocephalus has actually caused the closure of the aqueduct [26]. In all of these patients and particularly in the two classes of patients discussed here, ETV offers no advantage because communication between the third ventricle and intrapeduncular cistern enables CSF to flow briefly. Intraventricular injection of Iohexal confirms free communication among the ventricles, cortical subarachnoid spaces, and cisterns except in children who experienced severe ventriculitis after their original shunt procedure [17, 24]. For these patients, it is essential to develop treatment strategies that access the cortical subarachnoid spaces [16].

NVH: pathophysiology and treatment

Increased ICP associated with small ventricles that can be managed by draining CSF is called pseudotumor cerebri. Older children and adults whose ventricles do not expand at shunt failure no longer have hydrocephalus. Rather, they have a form of pseudotumor cerebri that may be more difficult to treat and more dangerous than pseudotumor cerebri that begins in adulthood.

Increased pressure in the dural venous sinuses is likely to be a universal mechanism in the pathogenesis of pseudotumor cerebri in adults [14]. As can be seen in the context of a radical bilateral neck dissection, bilateral ligation of the jugular veins increases ICP and is associated with high venous pressure, increased turgor of the brain, and increased CSF in the subarachnoid spaces (i.e., pseudotumor cerebri). If bilateral radical neck dissection is performed in an infant, CSF in the cortical subarachnoid space, ventricular size, ventricular volume, or hydrocephalus would increase with increases in ICP. After shunting, the sutures close and the skull can no longer expand. The result is a shunted patient with pseudotumor cerebri. The clinical situation is the same as would have occurred at the time of shunt failure.

Clinically, these patients are at great risk because the level of recognition of this common condition among general neurosurgeons, radiologists, and emergency physicians is relatively low. Almost all of these patients have been in emergency rooms where imaging studies (almost always CT scans) have been interpreted as "no evidence of hydrocephalus" or "no evidence of shunt failure." The patients with complete and irrevocable shunt failure often have prolonged periods of suffering and occasional blindness with no evidence of ventricular dilatation. Such patients have almost universally been shunted during

infancy, and some measurement of ICP may be the only way to determine if their shunt is working. In one patient with NVH in the context of spina bifida, ICP was recorded at 70 mmHg with no increase in ventricular size [15, 16].

Usually, ventricular collapse around the catheter leads to intermittent failure of the shunt and to intermittent severe intracranial hypertension and symptoms. That these patients actually have NVH and nonresponsive ventricles may not be obvious. Although patients undergo many imaging studies (usually CT scans with the attendant risks of radiation), the diagnosis is elusive. Only when papilledema or constant agony leads to a shunt tap, ICP monitoring, or shunt exploration is the condition finally diagnosed.

Goals of management

As stated, 4% of humans have chronic daily headaches. Neurosurgeons need to do whatever is possible to "normalize" the dynamics of ICP. Medical management is complicated and frustrating and requires the cooperation of neurologists, internists, and mental health specialists to improve the quality of life for patients with normal ICP dynamics. A review of the management of such headaches, which must be individualized, is beyond the scope of this discussion. Based on reviews of management strategies that treat such headaches, however, narcotic medication must be avoided if possible. Narcotics are widely recognized as leading to the well-defined entity classified as "medication overuse headaches."

How does the physician decide that all has been done to "normalize" ICP dynamics? In normal patients, all CSF compartments, including the ventricles, spinal subarachnoid, cortical subarachnoid spaces and basal cisterns, freely communicate with each other. When multiple CSF compartments are isolated from each other, rapid changes in the dynamics of these compartments occur. In response, the brain, which is a viscoelastic substance, shifts. Even if all the compartments are shunted, this rapid shifting can occur unless all CSF catheters are spliced together proximal to the valve mechanism. Such rapid shifts at the time of Valsalva maneuvers or sudden positional changes cause distortion of the pain sensitive intracranial structures such as the basal dura. Ventricular shunting in the context of very large ventricles is also problematic. It is difficult to drain the cortical subarachnoid spaces because retrograde flow through the foramen of Monro is restricted [27].

Conclusions

The role of neurosurgeons in the management of shunt-related headaches or SVS must be to ensure that ICP dynamics are normalized as much as possible. Doing so requires ascertaining that all CSF compartments communicate without resistance or obstruction. Neuroendoscopic procedures can be used to fenestrate membranes that lead to compartmentalization or by splicing multiple ventricular or subarachnoid space catheters proximal to a single programmable valve containing a DRS. The final step is to monitor ICP over time to ascertain that ICP remains normal during all positions and during sleep.

What should be done if patients return complaining of reexacerbation of their headaches? First, the above shunt system must be identified as working as planned. If the ventricles do not expand at the time of shunt failure, which is the likely scenario in these cases, performing CT scans is futile. If it is essential to image the brain, rapid sequence MRIs are preferred. Typically, these patients have undergone many CT scans. At best, patients' risk of cataracts increases. At worst, there is a theoretical possibility that the risk of developing induced malignancies is increased over the ensuing decades.

In NVH patients with worsening symptoms, a reservoir tap is placed. ICP is measured while the patient is recumbent. The patient is asked to sit up to ensure that their ICP falls to a maximum of 5 cm H_2O or lower.

At this point about 5 cc of Iohexal is injected. The patient is scanned within an hour. The cortical subarachnoid and spinal subarachnoid spaces of the upper cervical spine are analyzed carefully. If there are no obstructions, the system is working as hoped, and the headaches are unrelated to the ICP. These patients can then be managed medically with the full understanding that all pressure manipulations that can be done have been done. These systems would greatly benefit by the incorporation of a telemetered monitoring device that allows the accurate noninvasive measurement of ICP in all positions.

Conflict of interest statement Dr. Rekate serves as a consultant of Codman Corporation in the development of improved shunt systems.

References

1. Wang SJ, Fuh JL, Lu SR, Juang KD (2006) Chronic daily headache in adolescents: prevalence, impact, and medication overuse. Neurology 66(2):193–197
2. Strine TW, Chapman DP, Balluz LS (2006) Population-based U.S. study of severe headaches in adults: psychological distress and comorbidities. Headache 46(2):223–232
3. Nulsen FE, Becker DP (1967) Control of hydrocephalus by valve-regulated shunt. J Neurosurg 26(3):362–374
4. Nulsen FE, Spitz EB (1951) Treatment of hydrocephalus by direct shunt from ventricle to jugular vein. Surg Forum 2:399–403
5. Hyde-Rowan MD, Rekate HL, Nulsen FE (1982) Reexpansion of previously collapsed ventricles: the slit ventricle syndrome. J Neurosurg 56(4):536–539
6. Rekate HL (1993) Classification of slit-ventricle syndromes using intracranial pressure monitoring. Pediatr Neurosurg 19 (1):15–20

7. Portnoy HD, Schulte RR, Fox JL, Croissant PD, Tripp L (1973) Anti-siphon and reversible occlusion valves for shunting in hydrocephalus and preventing post-shunt subdural hematomas. J Neurosurg 38(6):729–738
8. Epstein F, Marlin AE, Wald A (1978) Chronic headache in the shunt-dependent adolescent with nearly normal ventricular volume: diagnosis and treatment. Neurosurgery 3(3):351–355
9. Epstein FJ, Fleischer AS, Hochwald GM, Ransohoff J (1974) Subtemporal craniectomy for recurrent shunt obstruction secondary to small ventricles. J Neurosurg 41(1):29–31
10. Holness RO, Hoffman HJ, Hendrick EB (1979) Subtemporal decompression for the slit-ventricle syndrome after shunting in hydrocephalic children. Childs Brain 5(2):137–144
11. McLaurin RL, Olivi A (1987) Slit-ventricle syndrome: review of 15 cases. Pediatr Neurosci 13(3):118–124
12. Engel M, Carmel PW, Chutorian AM (1979) Increased intraventricular pressure without ventriculomegaly in children with shunts: "normal volume" hydrocephalus. Neurosurgery 5(5):549–552
13. Rekate HL (2002) Adults with hydrocephalus treated in infancy and childhood. In: Ellenbogen R (ed) Pediatric Neurosurgery for the General Neurosurgeon. New York, Thieme, pp 19–28
14. Karahalios DG, Rekate HL, Khayata MH, Apostolides PJ (1996) Elevated intracranial venous pressure as a universal mechanism in pseudotumor cerebri of varying etiologies. Neurology 46(1):198–202
15. Nadkarni TD, Rekate HL (2005) Treatment of refractory intracranial hypertension in a spina bifida patient by a concurrent ventricular and cisterna magna-to-peritoneal shunt. Childs Nerv Syst 21(7):579–582
16. Rekate HL, Nadkarni T, Wallace D (2006) Severe intracranial hypertension in slit ventricle syndrome managed using a cisterna magna-ventricle-peritoneum shunt. J Neurosurg 104(4 Suppl):240–244
17. Rekate HL, Wallace D (2003) Lumboperitoneal shunts in children. Pediatr Neurosurg 38(1):41–46
18. Albright AL, Tyler-Kabara E (2001) Slit-ventricle syndrome secondary to shunt-induced suture ossification. Neurosurgery 48 (4):764–769
19. Di Rocco C, Velardi F (2003) Acquired Chiari type I malformation managed by supratentorial cranial enlargement. Childs Nerv Syst 19(12):800–807
20. Walker ML, Fried A, Petronio J (1993) Diagnosis and treatment of the slit ventricle syndrome. Neurosurg Clin N Am 4(4):707–714
21. Oikonomou J, Aschoff A, Hashemi B, Kunze S (1999) New valves—new dangers? 22 valves (38 probes) designed in the 'nineties in ultralong-term tests (365 days)'. Eur J Pediatr Surg 9 (Suppl 1):23–26
22. Chumas PD, Drake JM, Del Bigio MR (1992) Death from chronic tonsillar herniation in a patient with lumboperitoneal shunt and Crouzon's disease. Br J Neurosurg 6(6):595–599
23. Thompson DN, Harkness W, Jones BM, Hayward RD (1997) Aetiology of herniation of the hindbrain in craniosynostosis. An investigation incorporating intracranial pressure monitoring and magnetic resonance imaging. Pediatr Neurosurg 26(6):288–295
24. Baskin JJ, Manwaring KH, Rekate HL (1998) Ventricular shunt removal: the ultimate treatment of the slit ventricle syndrome. J Neurosurg 88(3):478–484
25. Rekate HL (1982) Establishing the diagnosis of shunt independence. Monogr Neural Sci 8:223–226
26. Borti A (1976) Communicating hydrocephalus causing aqueductal stenosis. Neuropadiatrie 7(4):416–422
27. Rekate HL, Williams FC Jr, Brodkey JA, McCormick JM, Chizeck HJ, Ko W (1988) Resistance of the foramen of Monro. Pediatr Neurosci 14(2):85–89