30th YEAR ANNIVERSARY SUPPLEMENT

SURGICAL MANAGEMENT OF ADULT HYDROCEPHALUS

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THE MANAGEMENT OF adult hydrocephalus spans a broad range of disorders and ages. Modern management strategies include endoscopic and adjustable cerebrospinal fluid shunt diversionary techniques. The assessment and management of the following clinical conditions are discussed: 1) the adult patient with congenital or childhood-onset hydrocephalus, 2) adult slit ventricle syndrome, 3) multicompartmental hydrocephalus, 4) noncommunicating hydrocephalus, 5) communicating hydrocephalus, 6) normal pressure hydrocephalus, and 7) the shunted patient with headaches. The hydrodynamics of cerebrospinal fluid shunt diversion are discussed in relation to mechanisms of under- and overdrainage conditions. A rationale for the routine implementation of adjustable valves for adult patients with hydrocephalus is provided based on objective clinical and experimental data. For the condition of normal pressure hydrocephalus, recommendations are offered regarding the evaluation, surgical treatment, and postoperative management of this disorder.

KEY WORDS: Adult hydrocephalus, Endoscopic third ventriculostomy, Headache, Normal pressure hydrocephalus, Slit ventricle syndrome

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For neurosurgeons treating adult patients, hydrocephalus is a commonly encountered disorder either as a primary condition or as a secondary phenomenon after intracranial hemorrhage, brain tumor, and/or meningitis. In many cases, the diagnosis is straightforward and the management is relatively simple (placement of a cerebrospinal fluid [CSF] shunt). Other cases, however, may constitute some of the most complex and challenging clinical conditions to face a neurosurgeon (11, 21). Over the past decade, there has been increasing interest in the diagnosis and treatment of normal pressure hydrocephalus (NPH), a disorder that represents a relatively small percentage of hydrocephalus cases encountered in a neurosurgeon's routine practice. Hydrocephalus occurring in adults is a heterogeneous group of disorders that spans a wide range of ages (teenagers to nonagenarians), severity and chronicity of symptoms, and physiological states (low versus high brain compliance). As such, each patient requires an individualized diagnostic and treatment approach.

Historically, the management approaches offered to adult patients have been extrapolations of the experiences obtained from treating pediatric patients. Aside from NPH, there is a relative paucity of literature and clinical studies pertaining specifically to the surgical management of adult hydrocephalus conditions. Given the scarcity of Class I and II evidence, reports such as this one are, by default, limited to an analysis and interpretation of published clinical reports that are of variable quality, plus an interjection of personal clinical experience. We openly acknowledge that there is more than one acceptable way to treat any of the clinical disorders discussed herein, but offer the information and recommendations in the hope that they are useful in aiding the often challenging management of patients with hydrocephalus.

Adult Patient with Pediatric-onset Hydrocephalus

The transition of care from a pediatric neurosurgeon to an "adult" neurosurgeon can bring great anxiety to patients and their families and present tremendous challenges to the recipient neurosurgeon. Many patients will have undergone multiple hydrocephalus-related operations, and those with congenital hydrocephalus may have significant baseline cognitive and physical disabilities. Assuming the care of these patients is facilitated by the acquisition of a detailed summary of the vari-

ous valve types, pressure settings, shunt types, and other operations the patient may have had, particularly over the most recent 5 to 10 years. In addition, it is imperative that the patient provide as many computed tomographic/magnetic resonance imaging (MRI) scans that are available. Patients who have had repeated shunt operations are a particular challenge, especially if the brain has multiple ventricular catheters, including some that are orphans and of various ages. It may not be clear which catheter is functioning. Obviously, the most recently placed shunt should be the one of primary interest if the question of shunt malfunction is considered.

It is often wise to carefully listen to and trust the intuition of the patient and/or their caring family member with regard to signs and symptoms of shunt malfunction. Many have learned the subtleties of their own unique presentations of shunt failure. A common pitfall is to interpret a computed tomography (CT) or MRI study showing small ventricles as proof of a functioning shunt (see Adult Slit Ventricle Syndrome) and therefore discount the complaints of the patient. Comparison with older imaging studies is important. The signs and symptoms of shunt failure in this group of patients can vary greatly and are not limited to headache and nausea. This is particularly true for patients with myelomeningocele, who can present with signs and symptoms seemingly related to spinal cord dysfunction.

Evaluation of shunt obstruction should entertain the possibility that the patient no longer requires CSF diversion (18). Because endoscopic third ventriculostomy (ETV) did not gain popularity until the mid-1990s, many pediatric patients with noncommunicating hydrocephalus were not offered endoscopic management originally (23). The "knee-jerk response" to proceed directly with shunt revision may deny the patient the opportunity to gain shunt independence. For patients demonstrating ventriculomegaly, an excellent screening test for noncommunicating hydrocephalus is a high-resolution, thin-slice sagittal MRI study using the constructive interference in steady state (CISS) sequence (*Fig. 1*) (1, 64).

Patients presenting with possible shunt malfunction and small/slit ventricles can also be considered for evaluation of shunt independence. Patients with noncommunicating hydrocephalus appear to be more prone to developing slit/collapsed ventricles, possibly as a result of the differential drainage of ventricular instead of cisternal CSF. O'Brien et al. (87) reported that ETV was 70% effective in the setting of shunt failure, whereas Woodworth et al. (120) reported a 2.5-fold lower efficacy of ETV in previously shunted patients.

If a revision is needed, assessing an acceptable site for distal catheter placement requires a detailed history of prior operations (shunt- and nonshunt-related). It is important to estimate the likelihood of encountering peritoneal adhesions, particularly in patients with a history of ventriculoperitoneal shunt infection, peritonitis, or extensive abdominal surgery. If peritoneal adhesions may pose problems for surgical access to the peritoneal cavity, conversion to a ventriculoatrial (VA) shunt may need to be considered (66). Our preferred method is ultrasound-guided percutaneous placement (37) of the catheter into the internal jugular vein and advancement to the superior vena cava-right atrial junction. Patients shunted in infancy with VA shunts, however, may have had the internal jugular vein ligated, which is critical to note if there is consideration for a new VA shunt.

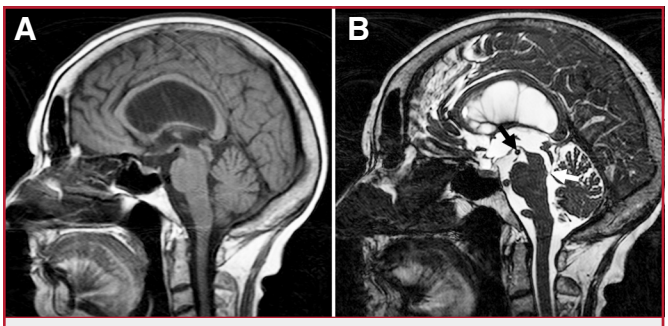

FIGURE 1. *Imaging was performed for aqueductal stenosis.* **A**, *midsagittal noncontrast T1-weighted magnetic resonance imaging (MRI) scan showing enlarged lateral ventricle and bowing of the corpus callosum and normal-sized fourth ventricle. The cerebral aqueduct appears patent.* **B**, *corresponding constructive interference in steady state (CISS) MRI scan reveals a distal cerebral aqueduct occlusive membrane (*white arrow*). The premesencephalic cistern has favorable anatomy, with a posteriorly placed basilar artery bifurcation (*black arrow*), for endoscopic third ventriculostomy (ETV).*

For patients presenting with multiple ventricular shunt catheters, complex shunting configurations who require continued drainage, considerations should be made to simplify the shunt. This may require the removal of orphan catheters and/or unifying multicompartmental hydrocephalus using endoscopic techniques (see Multicompartmental Hydrocephalus).

If shunt revision is the only or best option, consideration should be made to changing the valve to an adjustable ("programmable") valve regardless of the point of malfunction. It is our experience that at least 40% of (non-NPH) patients require at least one valve adjustment secondary to either over- or underdrainage conditions. This, coupled with the fact that the final ("optimal") valve opening pressure ranges from 10 to 400 mmH_2O, indicates that any given fixed-pressure valve (even multistaged) will either over- or underdrain in many patients. The exact percentage of patients who benefit from an adjustable valve is not known and, based on our experience, increases with age up to an estimated 40% in the elderly patient with hydrocephalus. There is no clear advantage for routine use of adjustable valves in children with hydrocephalus (91).

In symptomatic patients who have a patent shunt, continuous intracranial pressure (ICP) monitoring with an intraparenchymal microsensor may help in the management. In some cases, clear underdrainage (ICP values less than −10 mmHg with prolonged sitting or standing) or overdrainage (sustained ICP values greater than 15 mmHg when lying flat in bed) may be discovered and then rectified with either valve reprogramming or revision to an appropriate valve. In many cases, however, normal ICP values in both the supine and upright positions will be documented, thereby

avoiding an unnecessary shunt revision or adjustment. The medical management of headaches can then be more aggressively pursued.

The young adult presenting with shunt malfunction after a several-year period of good health can be a particularly challenging clinical scenario. These patients present with symptoms that are a blend of pediatric and elderly hydrocephalus features, including abnormalities in gait (70% of patients), cognition (70% of patients), urinary urgency (48% of patients), plus headaches (56% of patients) (28). It is our experience that young adults are more prone to CSF overdrainage resulting in ventricular collapse and accompanying shunt malfunction. Again, an evaluation for shunt independence should be entertained. For those requiring a shunt revision, very high valve opening pressures (up to 400 mmH_2O) may be needed. Flow-limiting valve designs may be an alternative.

Adult Slit Ventricle Syndrome

The adult slit ventricle syndrome is an ill-defined disorder, but the key components are a symptomatic, shunted patient with "slit" or collapsed ventricles seen on CT or MRI (*Fig. 2*). The incidence is unknown but represents approximately 5% of the non-NPH evaluations in our clinic. Although relatively few in number, these patients represent a disproportionate amount of clinical effort expended, with frequent emergency room visits and requests for office visits. The syndrome occurs more commonly in patients who have been shunted for many years either as an adult or during childhood. Additionally, it is our observation that a significant proportion of patients with adult slit ventricle syndrome have previously unrecognized noncommunicating hydrocephalus.

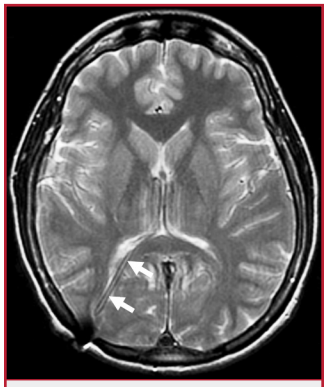

FIGURE 2. *Slit ventricle is shown. Axial T2-weighted MRI scan demonstrating collapse of the lateral ventricles in a patient with slit ventricles. A portion of the shunt ventricular catheter is seen* (white arrows). *The finding of slit ventricles does not indicate a properly functioning shunt.*

Common symptoms of adult slit ventricle syndrome include intermittent headaches that become more frequent and intense over time. The etiology of these intermittent headaches has been unclear but is thought to be related to periods of insufficient CSF drainage. In addition, collapse of the ventricular system lowers intracranial compliance, further amplifying elevations in ICP during shunt underdrainage. At shunt revision, the typical intraoperative finding is near total, but not complete, obstruction of the ventricular catheter (typically only one or two holes are patent). Left untreated, the symptoms may progress to more continuous headaches, presumably as a result of completed mechanical obstruction of the shunt system.

The neurological examination is often unremarkable. In patients harboring a valve with a pumping chamber or reservoir, the finding of very slow refilling of the chamber after depression is consistent but not diagnostic (90) of this syndrome.

Why a patient with nearly complete shunt obstruction does not present with ventriculomegaly may seem counterintuitive. However, one explanation for the lack of ventricular enlargement is contingent on the presence of a marginally flowing shunt system. A balloon analogy is helpful in understanding this clinical scenario. As dictated by Laplace's law, a high intraluminal pressure is initially required to expand an uninflated balloon. Once inflated, however, less pressure is required as the balloon enlarges (the force is translated to tension forces on the surface).

The existence of the slit ventricle state occurs as a result of chronic shunt overdrainage. Once the ventricles are collapsed, Laplace's law suggests that a high intraventricular pressure needs to be generated for ventricular enlargement to occur. In patients with adult slit ventricle syndrome, presumably this amplitude of pressure does not occur because the shunt allows enough, albeit intermittent, CSF flow. If the shunt is externalized and the flow is occluded, gross ventricular enlargement is usually observed within 24 hours, indicating that ventriculomegaly (hydrocephalus) is still possible.

Patients who are asymptomatic but are found to have slit ventricles (either ipsilateral to the ventricular catheter or bilaterally) on computed tomographic or MRI scans are a special, and more common, circumstance. Apposition of the ependymal wall and/or choroid plexus to the ventricular catheter likely increases the risk of developing ventricular catheter obstruction. If a patient has an adjustable valve, consideration should be given to incrementally increasing the valve opening pressure over several weeks with the hope of normalizing the ventricular size. Elective surgical shunt revision, however, is not indicated in an asymptomatic patient with slit ventricles.

We recommend that the management of adult slit ventricle syndrome be primarily directed at assessing the possibility of shunt independence. An attempt should be made to acquire prior MRI studies demonstrating ventricular enlargement to screen for obvious noncommunicating hydrocephalus. The best screening examination, however, is a high-resolution, thin-slice sagittal MRI study using CISS (*Fig. 1*). A standard T1-weighted sagittal MRI study does not have the resolution necessary to detect membranous aqueductal webs. If the patient has slit ventricles, MRI screening for noncommunicating hydrocephalus is typically of little or no value. In selected cases, our preferred management approach is to iatrogenically produce ventriculomegaly and then assess candidacy for ETV by imaging. If the patient has an adjustable valve, a noninvasive approach is to reprogram the valve to its highest setting and then closely monitor the patient for the development of hydrocephalus. This should not be performed in the outpatient setting in a cognitively impaired patient or if there is no responsible caregiver. A computed tomographic scan should be obtained 2 to 4 days after the adjustment and then a week later to assess for ventriculomegaly.

Inpatient Protocol for Shunt-dependence Assessment

For patients with adult slit ventricle syndrome, the iatrogenic induction of ventriculomegaly typically requires complete cessation of CSF flow through the shunt (7, 97). In our practice, the first stage is shunt externalization and placement of an ICP monitor. We perform this step in the operating room, under general anesthesia, to optimize sterility and minimize patient discomfort. The peritoneal catheter is connected to a sterile external ventricular drainage system. Postoperatively, the patient is monitored in the intensive care unit or an equivalent setting, and CSF drainage is completely stopped. We have determined that the alternative approach, increasingly raising the ventricular drainage level (20), unnecessarily prolongs the time required to expand the ventricles.

The intensive care unit management must be tailored for the individual patient, but in general, a computed tomographic scan of the brain is obtained at 24 hours and daily thereafter until ventricular enlargement is documented (if it develops). The clinical progression of acute hydrocephalus is quite variable. Some patients experience a decreased level of consciousness within hours of shunt occlusion and, therefore, CSF drainage is reinstituted to regain consciousness. CSF is drained judiciously because overdrainage of CSF will defeat the goal of enlarging the ventricles. Interestingly, ICP may not be elevated ($<$12 mmHg) in these cases. In other patients, significant intracranial hypertension can be encountered with or without the development of ventriculomegaly. This too may prompt CSF drainage for sustained elevation of ICP even if the patient has no change in neurological status.

If ventriculomegaly is documented on a computed tomographic scan, the patient undergoes a sagittal CISS MRI study to assess for aqueductal obstruction as well as to study the premesencephalic anatomy for ETV safety evaluation. Suitable candidates then undergo an ETV procedure, and the shunt is removed. If ventricular enlargement cannot be attained, then a shunt revision is needed. We favor placement of a ventriculocisternoatrial shunt with an adjustable valve, typically in conjunction with limited suboccipital decompression (97). The rationale is to equalize the ventricular and subarachnoid/cisternal CSF pressure, thereby eliminating transmantle pressure gradients that promote collapse of the ventricular system. We prefer an atrial configuration because it can be accomplished in a single operative setting (in the lateral position) without the need for repositioning the patient. Some have reported successful treatment of slit ventricle syndrome with conversion to a lumboperitoneal shunt (106).

Over the past 2 years, we have treated nine patients with slit ventricle syndrome using the previously described shunt independence assessment protocol. Four underwent successful ETV operations, are clinically doing well, and remain shunt-free after a mean follow-up of 2 years. One patient clinically failed ETV (progressive ventriculomegaly at 3 d postoperatively) and subsequently underwent a ventriculocisternoatrial shunt. Three patients proceeded directly to ventriculocisternoatrial shunts, in two patients as a result of inability to dilate the ventricular system and in the other, the MRI demonstrated communicating hydrocephalus. Among these four patients with ventriculocisternoatrial shunts, normalization of the ventricular system size was achieved in all cases. Clinical resolution of headaches, however, was achieved in only two of the four patients. On stopping CSF flow through the externalized shunt in the ninth patient, the ventricles normalized, and the ICP values remained low (normal). This patient had her shunt removed and did well for approximately 3 weeks, and then returned with symptomatic hydrocephalus. She opted for a simple ventriculoperitoneal shunt with an adjustable valve, and she has improved with a higher valve pressure setting. Together, seven out of nine patients were symptomatically improved using this protocol. Similar promising results were reported by Reddy et al. (95). All patients treated with ETV should be followed clinically because late failure, including rare late rapid deterioration (33) and even death (48, 71), have been reported.

Multicompartmental Hydrocephalus

The phenomena of compartmentalized hydrocephalus and isolated fourth ventricle were described by Dandy (31) more than 80 years ago. Multicompartmental, or multiloculated, hydrocephalus can occur after ventriculitis or, more rarely, after ventricular hemorrhage. The most common anatomic variant is the isolated fourth ventricle. To add to the complexity of management, many of the patients we have treated were first treated elsewhere with multiple shunt catheters and/or valves. Several patients had incompletely treated ventriculitis and/or continued shunt infection, a condition that must be objectively assessed and treated before definitive management can progress.

In terms of general principles, the first is to simplify. If possible, endoscopic techniques should be considered to fenestrate dividing membranes and create a unicompartmental system. We have identified axial and coronal CISS MRI imaging to be helpful in preoperative planning, and frameless stereotactic neuronavigation is important for choosing the optimal initial endoscopic trajectories. Existing catheters should be removed, if possible, and one catheter should be positioned endoscopically.

The Isolated Fourth Ventricle

The isolated fourth ventricle has been variously referred to as the trapped, sequestered, or occluded fourth ventricle and as an aspect of "double compartmental hydrocephalus" (25, 32, 39, 72, 86, 122). It has garnered multiple publications for pediatric patients (24–26, 32, 35, 41, 50, 52, 53, 68, 72, 85, 103, 104, 107, 111) as well as adults (5, 27, 39, 51, 56, 80, 85, 105, 115, 116). The incidence and prevalence have not been reported for the adult hydrocephalus population. Symptomatic patients with isolated fourth ventricle present variously with headache, nausea, vomiting, anorexia, ataxia, cranial nerve palsies, and coma (35).

In general, surgical treatment of isolated fourth ventricle requires the placement of a shunt catheter into the fourth ven-

tricle. Various approaches for placement of fourth ventricular shunt catheters by means of a suboccipital craniectomy have been described, including a transvallecular approach through the foramen of Magendie and a transcerebellar cannulation of the fourth ventricle (52). In our opinion and experience, the anatomy of the fourth ventricle is poorly suited for ideal catheter placement from a posterior fossa approach. The trajectory angle from a posterior or inferior approach tends to leave the catheter tip touching or even impinging on the floor of the fourth ventricle. This can lead to disabling neurological deficits as well as direct irritation of the emesis center (35, 68).

Alternative treatments for isolated fourth ventricle have been devised, including ETV, aqueductal reconstruction (85), aqueductal stent placement (24, 105), retrograde aqueductoplasty and stenting (102), and endoscopic interventriculostomy connecting the lateral or third ventricle to the fourth ventricle (41). Depending on the individual case, each has potential advantages and shortcomings. Obviously, an ETV alone does not address an isolated fourth ventricle. Aqueductoplasties without stenting have high restenosis rates, whereas a separate stent adds complexity to a shunt system. In many cases, direct catheter drainage of the fourth ventricle is required.

An endoscope-assisted placement of a fourth ventricle catheter using a single, frontal burr hole, transaqueductal approach has been described by us (116) and by Torres-Corzo et al. (115). Our fourth ventricle shunt system consisted of a panventricular catheter modified by perforations corresponding to the lateral, third, and fourth ventricles. This multiperforated, panventricular catheter allows simultaneous drainage of the lateral, third, and fourth ventricles, acting as a drain pipe that equalizes the ventricular pressures and permits the ventricular system to be served by one shunt. In our early experience with four patients, this fourth ventricular shunt system resulted in good outcomes, assessed both clinically and radiographically, in each case.

Newly Diagnosed Noncommunicating Hydrocephalus

There are two distinct clinical presentations of newly diagnosed noncommunicating hydrocephalus. The first are the patients who present with a longstanding "compensated" hydrocephalus state, whereas the second are those with newly acquired noncommunicating hydrocephalus secondary to tumor mass effect, intraventricular neurocysticercosis, or other mass lesions.

Late-onset Idiopathic Aqueductal Stenosis

Fukuhara and Luciano (42) reported a series of 31 symptomatic patients with late-onset idiopathic aqueductal stenosis (LIAS). Clinical presentation was most commonly headache with subsequent gait disturbance, memory disturbance, incontinence, blurred vision, tremor, seizure, swallowing difficulty, and rarely, Parinaud syndrome. A similar pattern of presentation has been reported by others (42, 114). The patients were treated with ETV with an overall success rate of 83.9%. The mean follow-up was 26.2 months. Tissell (112) and Tissell et al. (113) reported a similarly good response rate after ETV.

Over the period of 1995 to 2006, we have treated 52 patients with LIAS at the University of California–Los Angeles Medical Center. With a median follow-up of 8 months, symptomatic relief was accomplished in 86.5% of patients (34). These patients must be followed after ETV because late failures do occur (34, 96).

LIAS can present with signs and symptoms indistinguishable from NPH (114). In our experience, these NPH–LIAS patients present 10 to 20 years younger than idiopathic NPH (iNPH). It is important that for all patients being evaluated for NPH, aqueductal stenosis be considered because these patients can respond favorably to an ETV procedure.

The management of asymptomatic aqueductal stenosis is controversial. Traditionally, these patients have been managed conservatively with a watch-and-wait approach. Presumably, this approach is a carryover of the pre-ETV era in which shunting procedures were the only treatment option. The historical complication rate, at a time preceding the availability of adjustable valves, was exceedingly high. Although the natural history of asymptomatic aqueductal stenosis is not well established, we believe that converging evidence supports a prophylactic management strategy. First, we have encountered only a few patients older than age 60 years with asymptomatic aqueductal stenosis. Assuming that incidental aqueductal stenosis is discovered equally across all age groups, this would suggest that few persons with asymptomatic aqueductal stenosis remain so in old age. Second, as noted previously, symptomatic NPH-like patients undergoing LIAS present at a younger age than their iNPH counterparts. Lastly, once a patient becomes symptomatic, it may be too late to effectively intervene. Another factor is that ETV, as a treatment option, carries a much lower overall risk compared with shunting procedures. We therefore give asymptomatic patients with aqueductal stenosis the option of prophylactic surgical intervention with an ETV. Whether this prevents or delays the onset of hydrocephalus symptoms is unknown without a prospective, randomized trial. Naturally, the risk to (theoretical) benefit ratio is only favorable if the ETV complication rate is very low. For asymptomatic patients choosing to forego or delay surgical intervention, we obtain a baseline neuropsychological testing battery and then repeat the battery yearly along with MRI studies. If there is any unexplained worsening of the test scores, interval enlargement of the ventricular system, or the earliest onset of gait or bladder control symptoms, surgical intervention is recommended. If an ETV operation is not technically possible or refused, we recommend the use of an adjustable valve with an initial very high-pressure setting in the shunt system.

Secondary Noncommunicating Hydrocephalus

Although a randomized trial has not been performed comparing ETV versus shunting, ETV should be considered as the primary treatment for noncommunicating hydrocephalus secondary to a defined, obstructive lesion such as an intra- or periventricular tumor (*Fig. 3*) (3, 59). This of course pertains to cases in which CSF pathways cannot be reestablished by removal of the offending lesion. Patients with secondary noncommunicating

hydrocephalus typically present with more acute symptomatology and accompanying imaging findings such as periventricular "edema" that are best seen on fluid-attenuated inversion recovery MRI imaging. Pineal region tumors are the prototypical lesions causing secondary noncommunicating hydrocephalus, although a multitude of other possibilities exist, including thalamic masses, tectal gliomas, and even multiple sclerosis plaques (6).

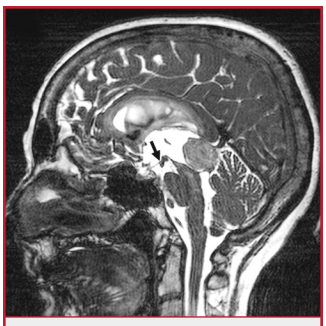

FIGURE 3. *Secondary noncommunicating hydrocephalus is demonstrated in this midsagittal CISS MRI scan of a cerebral aqueduct obstruction secondary to pineal region tumor. The premesencephalic cistern has favorable anatomy for endoscopic third ventriculostomy. The basilar artery bifurcation location (*black arrow*) is posteriorly placed, allowing ample room for creation of the stoma.*

It is important to note that ETV treatment is not appropriate for all patients with secondary noncommunicating hydrocephalus. First, it is our standard of care to obtain a sagittal CISS (or fast imaging employing steady-state acquisition) MRI study to assess the premesencephalic anatomy (1, 54, 65). This is especially important for larger mass lesions, which may displace the midbrain and pons anteriorly, pushing the basilar artery against the clivus. Although successful ETV procedures have been reported on patients with this unfavorable anatomic arrangement (6, 99), certainly the risk of fatal basilar artery injury is higher in such cases. There is no defined criteria as to what constitutes an acceptable premesencephalic distance for ETV; however, in our practice, we prefer to see a minimum of 3 mm of anteroposterior "working" space (*Fig. 4*).

Another consideration of ETV pertains to its timing relative to endoscopic tumor biopsies. One of the recognized adverse events after endoscopic biopsy of intraventricular tumors is CSF seeding of the tumor. This appears to occur with higher frequency in germinomas, pineoblastomas, and higher-grade glial tumors. If the ETV procedure is performed at the same time as the endoscopic biopsy, the tumor is potentially disseminated throughout the cranial–spinal compartments, thereby possibly relegating the patient to craniospinal radiation rather than a ventricular field.

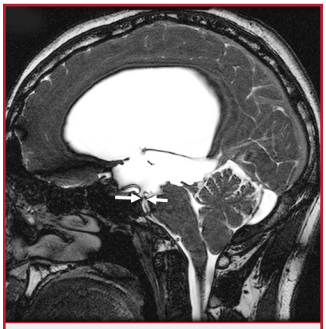

FIGURE 4. *High-risk ETV case is shown. A midsagittal CISS MRI scan reveals a proximal cerebral aqueduct occlusive membrane. The premesencephalic cistern has unfavorable anatomy for ETV. The pons is anteriorly displaced with the basilar artery bifurcation occupying nearly the entire prepontine cistern (*white arrows*).*

In our, and others', experience, patients with chronic meningitidis such as coccidiomycosis who present with a component of noncommunicating hydrocephalus respond poorly to ETV (38, 43). Presumably, this is attributable to concomitant obstruction of the cisternal and subarachnoid spaces.

In our experience of 47 cases (1995–2006) of ETV for secondary noncommunicating hydrocephalus (34), the two most common causes were pineal region tumors (14 cases) and midbrain or diencephalic gliomas or cysts (13 cases). The success rate, defined as shunt independence, was 77%. This success rate is consistent with that of other published studies (46, 55, 101). We determined that compared with nontumoral ETV cases, the complication rate was higher in patients with intracranial malignancies, including two mortalities. This risk of significant morbidity or mortality among patients with brain tumor treated with ETV, particularly if a biopsy is performed, was described previously by Beems and Grotenhuis (7a) in a large pediatric population. In their series, although the overall rate of complication was only 7.7%, they identified a 19.5% complication rate in patients who had a biopsy at the time of their ETV. Their only mortality was in such a patient.

Placement of a ventriculoperitoneal shunt should always be discussed as an alternative to ETV. The shunt carries a lower operative complication risk, but the long-term complication rate may be cumulatively higher. It is our experience that patients with secondary noncommunicating hydrocephalus are more prone to ventricular collapse after shunt procedures, and therefore, we typically incorporate adjustable differential pressure valves with very high opening pressure settings (300 mmH_2O).

The management of secondary noncommunicating hydrocephalus caused by intraventricular neurocysticercosis deserves special consideration. Endoscopic removal of the intraventricular cyst is the treatment of choice when technically feasible (4, 12, 29, 58, 92). Our experience with fourth ventricular cysticercal cysts suggests that ETV may be a valuable adjunct to the direct endoscopic removal of the cyst (9).

Non-NPH Communicating Hydrocephalus

Communicating hydrocephalus is more common than noncommunicating hydrocephalus among patients diagnosed in adulthood. The symptomatology of communicating hydrocephalus, in general, varies with age. Younger patients are most likely to present with headaches, nausea, sixth nerve palsies, and declining consciousness. Conversely, elderly patients typically present with signs and symptoms of NPH. Headache is a very uncommon complaint in the elderly.

Just as symptomatology varies with age, our experience with continuous ICP monitoring suggests a transition of intracranial pressure dynamics with age as well. Younger patients with communicating hydrocephalus are more likely to have sustained intracranial hypertension, which is uncommonly encountered in the elderly. The parallel trend between headaches and ICP, however, is not necessarily a cause-and-effect relationship. Our continuous ICP monitoring rarely discloses a clear correlation with instantaneous ICP and headache severity.

Many cases of communicating hydrocephalus occur after subarachnoid and/or ventricular hemorrhage, particularly as a consequence of aneurysmal rupture. Other cases are sequelae of meningitis or ventriculitis. In younger patients (<60 yr of age) presenting with communicating hydrocephalus, it is important to seek an instigating etiology rather than simply proceeding to a shunt. This should include a contrast-MRI study and CSF analysis. Failure to diagnose certain conditions such as tuberculous meningitis can result in a deleterious delay in appropriate treatment.

The treatment of communicating hydrocephalus has traditionally been a CSF shunting procedure (such as a ventriculoperitoneal shunt). This has been based on the concept that communicating hydrocephalus occurs in conjunction with an elevation in CSF outflow resistance (as a result of blockage of the arachnoid granulations) and that the shunt normalizes the CSF outflow resistance by providing an alternative CSF egress. The validity of this bulk flow concept of hydrocephalus pathophysiology has increasingly been questioned over the last several years. From a practical standpoint, the traditional dichotomy of communicating versus noncommunicating hydrocephalus, proposed by Dandy (31), has diminishing relevance. Although seemingly nonintuitive, ETV appears to be an effective treatment for some patients with communicating hydrocephalus (44). The role of ETV for communicating hydrocephalus, at this point in time, remains poorly studied and understood.

NPH

Clinical Presentation

A comprehensive review of the diagnosis of iNPH can be found in the published Guidelines for the Diagnosis and Management of Idiopathic Normal Pressure Hydrocephalus (10, 61, 77, 78, 98). The following represents an overview of a single center's practical application of these guidelines.

The disorder of NPH, first recognized in 1965 (49), is not a distinct disease. It is better thought of as a mode of clinical presentation of hydrocephalus in the elderly. The classic triad of gait, bladder, and cognitive disturbance differs from that of younger patients with hydrocephalus, who may present with severe headaches, nausea, cranial palsies, or coma. The clinical presentation of NPH is also not limited to the classic triad, with some patients presenting with predominantly parkinsonian symptoms (30, 40, 62, 63, 84) or even findings suggestive of progressive supranuclear palsy (2).

The gait abnormality of NPH has been difficult to characterize in part because it varies from patient to patient. In our experience, "magnetic" apraxia may best represent the typical shunt-responsive gait. Patients initially have difficulty initiating gait, taking small, nonadvancing steps. This typically manifests during turns as well (*Video 1*). This freezing phenomenon, which shares features with parkinsonian gait, often can be overcome by verbal cuing. This suggests that the subconscious, automatic gait pathways are disrupted with NPH. After successful treatment, improvement in gait can be remarkable (*Video 2*), and can return a patient to independence.

More commonly, NPH evaluations are of patients presenting with varying degrees and/or features of truncal ataxia and slowed gait. It is our observation that patients with a significant degree of periventricular white matter ischemic changes commonly use the term "imbalance" to describe the gait problem. This dysequilibrium often fails to improve with treatment, although other components of gait dysfunction may improve (19).

Although incontinence is commonly stated as the second triad element, bladder urgency is the earlier symptom. This may be accompanied by urinary frequency complaints as well. Patients who exhibit incontinence without awareness, so-called incontinence sans gêne, commonly have advanced dementia. Fecal incontinence is rarely encountered with NPH.

The most common complaint related to cognition with NPH is forgetfulness. Patients are often, if not typically, largely unaware of the degree of their short-term memory problems. When asked directly, patients with NPH deny or minimize memory problems, whereas accompanying family members contradict the patient's impression. Clearly, multiple studies have demonstrated that NPH affects cognitive domains other than memory (98).

Neuroimaging Findings

The imaging criteria for hydrocephalus are rather straightforward (98). As a general rule, an Evans index of 0.3 is a minimal threshold, although we evaluate a small number of patients with lesser degrees of ventriculomegaly if the gait abnormality reveals a strong magnetic quality. Focal enlargements of the convexity subarachnoid space (*Fig. 5*) should not be automatically assumed to be *ex vacuo* changes. Additionally, evidence of cerebrovascular disease should not be used as an exclusionary criterion, although its existence lowers the probability of significant improvement (17). Last, patients with triventriculomegaly should be evaluated with a sagittal CISS MRI study to rule out an aqueductal web, because these patients may be candidates for ETV.

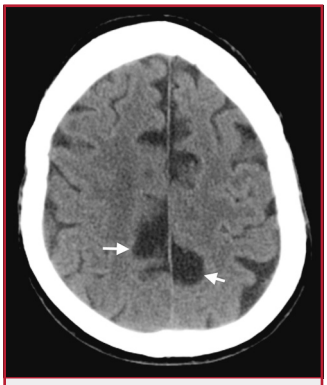

FIGURE 5. *Focal subarachnoid space enlargements are demonstrated in this axial noncontrast computed tomographic scan of a patient with panventriculomegaly (not shown). Focal enlargements of the subarachnoid sulcal spaces are evident* (arrows). *Focal cerebral atrophy may not be the etiology of this finding.*

Supplemental Diagnostic Testing

Over the past several decades, most of the attention toward NPH has been focused on identifying or improving supplemental diagnostic studies. These studies, however, are more prognostic (predicting shunt responsiveness) than diagnostic, primarily because they do not address the pri-

mary pathophysiology and, in addition, there is no gold standard by which a definitive diagnosis can be made.

There remains no consensus as to what the "best" supplementary prognostic test is for iNPH. As detailed in the guidelines (77), there are many diagnostic/prognostic tests that have been proposed for iNPH. The validation of individual tests, however, is largely lacking as a result of a dearth of well-designed, prospective studies. Most studies have significant methodological weaknesses, including retrospective analysis, treating only those patients with "positive" testing results, and inconclusive end point determinants. With regard to the latter, it is commonly assumed that the "diagnosis" of NPH can be confirmed or excluded based on whether a patient improves after a shunt procedure. Although a positive response strongly supports the diagnosis of iNPH, a lack of response to a shunt procedure cannot be assumed to be attributable to misdiagnosis of iNPH. What constituted a true shunt nonresponder versus an undertreated (underdrained) NPH is rarely addressed in most NPH-related studies. The experience with the Codman programmable valve (Codman/Johnson & Johnson, Raynham, MA (14, 121) supports the concept that iNPH is a heterogeneous, hydrodynamic disorder, with some patients requiring very low valve opening pressures (20 mmH_2O), and some requiring very high pressures (240 mmH_2O). Accordingly, the results of studies in which a fixed-pressure (nonadjustable) valve was used to treat patients may have an unknown proportion of undertreated subjects.

Currently, the most accurate test to predict shunt responsiveness is the external CSF lumbar drainage trial (77). In essence, this test constitutes a trial shunt in that a relatively large amount of CSF is drained for a sufficient period of time to detect an improvement in the greatest number of patients. The prospective study of 151 patients with iNPH by Marmarou et al. (79) demonstrated a 90% accuracy of this test. The patients were treated using adjustable valves, resulting in a low complication rate and, presumably, a low undertreatment rate. The negative predictive value of this test remains unknown, although studies suggest that a small percentage of patients with a negative test may still respond favorably to a shunting procedure (79, 88, 118).

The 72-hour CSF lumbar drainage trial has several inherent strengths. Compared with the high-volume lumbar puncture test, the continuous lumbar drainage trial is less likely to be affected by transient motivation/concentration-related clinical improvements. Patients (or families) typically report that the improved state continues for a mean of 8 days after removal of the lumbar CSF catheter. Such a sustained positive response is more likely to be physiologically related. Second, CSF drainage trials are of clinical benefit in that the patient and/or family experiences the improvement firsthand, and therefore, the preoperative conversation of risk-to-benefit ratio has a more concrete foundation. This is a drawback of other supplemental tests such as CSF outflow resistance measurement, in which the abstract results must be taken on faith by the patient. Third, the highest degree of improvement achieved after the lumbar drainage trial can serve as the minimal treatment goal after the shunt procedure. After a shunt procedure, a patient who achieves partial improvement will always want to know whether further improvement is possible. If an adjustable valve was implanted, the valve pressure can judiciously be lowered until the patient at least achieves the postlumbar drainage clinical response.

The external CSF lumbar drainage test has drawbacks, limitations, and risks. To date, it is primarily performed at major teaching hospitals because of the level of skilled nursing required. In the original description of the technique, Haan and Thomeer (47) reported a 25% complication rate. The two major risks (complications) are infection and overdrainage. We immediately remove the catheter if there is any external CSF leak such as a disconnected or broken catheter. Monitoring patients for at least 12 hours after removal of catheters should be considered because rare gram-negative meningitis can be fulminant. Overdrainage complications range from "spinal headaches" to subdural hematoma. Spinal headaches are easily treated with reduction in the drainage rate (such as 5 mL/h). Narcotics are ineffective and may result in delirium in elderly cognitively impaired patients. We have reduced inadvertent overdrainage by using intermittent drainage (~10 mL at the beginning of each hour and then turning the drainage off for the remaining part of the hour) (70).

The external CSF lumbar drainage test may not be appropriate for all patients. For a small percentage of patients who have very minimal symptomatology that spontaneously waxes and wanes, it may be hard to detect a true response. One question that is frequently asked is: what constitutes a positive response to drainage? From a scientific/academic standpoint, a positive response should be based on objective improvement on a reproducible test (of gait, bladder control, and/or memory). From a pragmatic clinical standpoint, however, a *meaningful* response may be more important than a *definable* response. For example, a patient who scores slightly better on a neuropsychological test after drainage may appear no different to his or her family. If you have to convince the patient and/or family that improvement occurred after the test, the likelihood of a disappointing result after shunting is much greater. Patients who experience an obvious response after external lumbar drainage will often return in follow-up and request (demand) a shunt operation. Lastly, patients should be screened for significant lumbar stenosis because lumbar catheter placement may evoke catheter-induced sciatica.

Patients presenting with classic NPH findings—clear ventriculomegaly, magnetic gait, urinary incontinence, and mild cognitive impairment—comprise a small minority of the greater cohort referred for evaluation. For such patients, one might question the necessity of performing "confirmatory" supplemental tests. Given the ubiquitous false-negative rate of all of the tests, it could be argued that performing a supplemental test is superfluous, and potentially dangerous, if the patient will proceed to a shunt operation regardless of the results of the test. An important consideration, however, relates to the perceived community standard of care definition. Given the historical uncertainty in making the diagnosis of

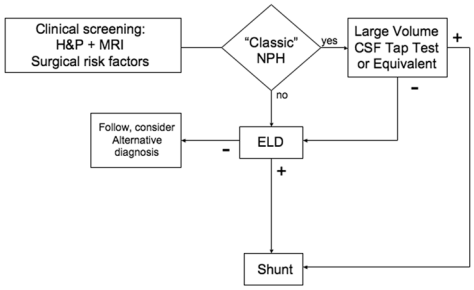

FIGURE 6. *Simplified version of the University of California–Los Angeles idiopathic normal-pressure hydrocephalus (iNPH) diagnostic evaluation algorithm. The patient undergoes initial evaluation of the clinical history and physical (H&P) and the MRI and/or CT findings. Patients with a "classic" iNPH presentation may be considered for a low-risk prognostic supplemental test such as a high-volume lumbar puncture. If the results of this test are not supportive of the diagnosis of iNPH, then the patient should be considered for the external lumbar cerebrospinal fluid (CSF) drainage (ELD) trial. Most patients evaluated, however, will have any one or multiple clinical features that bring uncertainty to the clinical diagnosis of iNPH; therefore, proceed directly to the more definitive external lumbar CSF drainage trial.*

iNPH, many might argue that some type of supplemental testing should be performed in all cases. In our current NPH diagnostic evaluation algorithm (*Fig. 6*), such classic patients have the option to undergo a lower risk alternative study to the external lumbar drainage trial. The choice of test is clinician-, patient-, and facility-specific. Although these alternative tests generally have positive predictive rates no higher than approximately 70% (77), it is reassuring to both the patient and physician that objective testing was performed. A comprehensive overview of the relative value of supplemental testing is located in the published guidelines (77). If the supplemental test does not indicate shunt-responsive iNPH, then the patient should be considered for the more definitive external CSF lumbar drainage trial.

Surgical Risk Assessment

Aside from routine preoperative medical concerns in the elderly, careful attention needs to be placed on lowering the risk of postoperative subdural hematoma. It is our routine to stop aspirin administration at least 10 days before surgery and then reinstate it 1 week afterward if there are no bleeding-related complications after surgery. Warfarin should be completely reversed at the time of surgery and may be reinitiated as early as 3 days after surgery (45). Clopidogrel (Plavix; Bristol-Myers Squibb/Sanofi Pharmaceuticals, New York, NY) administration should be stopped at least 14 days before surgery and strong consideration should be given to converting to aspirin or warfarin after surgery. It is our anecdotal experience that combination therapy of clopidogrel and aspirin exposes the shunted patient with iNPH to a subdural hematoma rate as high as 30%.

Shunt-related Management Issues in iNPH

As documented in the published guidelines (10), there is a scarcity of Class I and II evidence from which to base definitive recommendations regarding the surgical management of NPH. There is clearly more than one successful management strategy for iNPH. The following describes our center's approach to the surgical management of iNPH based on a large clinical experience, the results of clinical studies performed at our institution, and a critical review of the literature.

Once the diagnostic phase has been completed, surgical options must be discussed with the patient. In addition to discussing the shunt operation and possible risks, it is important to establish reasonable expectations with the patient and family. This relates to the degree of clinical improvement anticipated as well as how long any improvement experienced may last. Patients often assume that a CSF shunt procedure is "curative" of their condition. In one study of 42 patients with iNPH, Malm et al. (76) reported that improvement was observed in 64% of patients at 3 months but only 26% at 3 years follow-up. The relative risk of death among patients with iNPH compared with a general elderly population was 3.3-fold higher. In a 5-year follow-up study reported by Kahlon et al. (60), 37% of patients had died of non-NPH related causes. Of those who had improved initially after shunting, sustained improvement in gait was only 40% of the initial improvement level. Fewer than 10% of patients had an improvement in cognitive tests at 5 years follow-up. More patients (64%) improved if younger than 75 years, whereas only 11% patients older than 75 years had remained improved.

Historically, the surgical management of iNPH has been fraught with a high complication rate, the most troubling of which was subdural hematoma formation (8, 15, 57, 67, 89, 93, 94, 117). The reasons elderly patients with hydrocephalus (iNPH) are more prone to subdural hematoma collections are multifactorial; some are preventable and others are not. We believe that the high rates of subdural hematoma in early series were in part a consequence of excessive, and too abrupt, CSF drainage after shunt implantation, specifically as a result of the use of lower pressure shunt valves. Although the reason that iNPH was treated with low-pressure valves was not explicitly stated, it is our assumption that clinicians reasoned that, for CSF to flow through a shunt, the valve opening pressure must be lower than the ICP.

Inherent to this line of thought is the (incorrect) assumption that ICP is low with iNPH. There is a general consensus that *normal pressure* hydrocephalus is a misnomer. Whereas it is true that elderly patients with hydrocephalus are much less likely to have elevated ICP compared with their pediatric counterparts, multiple studies have documented various degrees of intracranial hypertension with iNPH. The studies of Malm et al. (74, 75) as well as our own data (14) suggest that only a minority of patients with iNPH have consistently low ICP values. Using

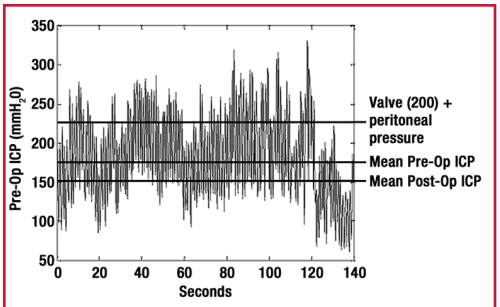

FIGURE 7. *Mechanism of intracranial pressure (ICP) reduction after shunt placement. Raw ICP waveform was measured, using an intraparenchymal microsensor, in a patient with iNPH in the recumbent, supine position before any CSF removal procedure. The mean preoperative (*Pre-Op*) ICP was approximately 170 mmH_2O. The patient underwent placement of ventriculoperitoneal shunt with an initial valve opening pressure of 200 mmH_2O and also measurement of the peritoneal pressure (14). Note that even with a combined valve opening pressure plus peritoneal pressure of approximately 230 mmH_2O, peak ICP values likely exceed this value. Because shunt valves contain a one-way valve, this results in obligatory CSF egress, the result of which is a reduction in mean ICP. This may explain how the postoperative (*Post-Op*) ICP exceeds the Pre-Op ICP despite the use of a valve with an opening pressure higher than the Pre-Op mean ICP.*

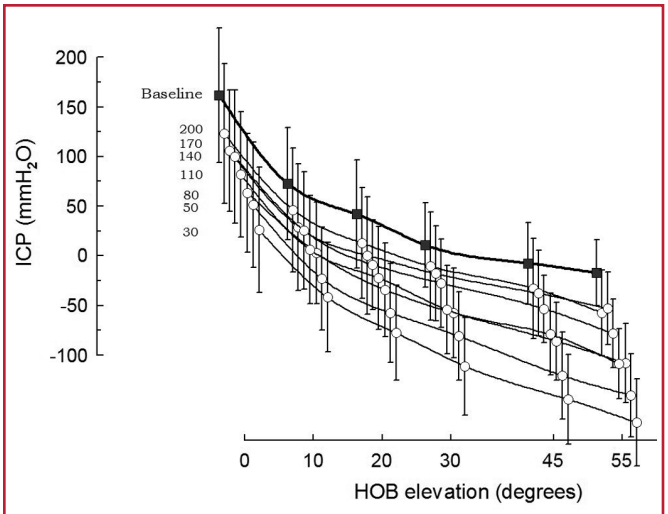

FIGURE 8. *ICP (mean ± standard deviation) versus head-of-bed (*HOB*) elevation curves through the full range of differential pressure opening pressures (200 mmH_2O, 170, 140, and so on) were measured from patients with iNPH who were treated with a ventriculoperitoneal shunt (*from, *Bergsneider M, Yang L, Hu X, McArthur DL, Cook SW, Boscardin WJ: Relationship between valve opening pressure, body position, and intracranial pressure in normal pressure hydrocephalus: Paradigm for selection of programmable valve pressure setting.* ***Neurosurgery*** *55:851–859, 2004 [14]). The preshunt baseline curve (*thicker line, solid square*) was obtained from the same group of patients. Note that the pre- and postoperative curves roughly parallel one another, demonstrating the limited role of siphoning as the cause of overdrainage in patients with iNPH.*

continuous intraparenchymal ICP monitoring, we documented a mean ICP of 164 ± 64 mmH_2O among patients with iNPH (14) with the mean ICP of some patients exceeding 240 mmH_2O. Even among patients with "normal" mean ICP values of less than 200 mmH_2O, more than 50% of patients exhibit vasogenic (Lundberg B) waves during sleep (110).

Our studies demonstrate that the assumption that the valve opening pressure must be lower than the preoperative ICP for CSF to flow through the shunt is unfounded and incorrect. We conducted a prospective study of iNPH in which ICP was measured both pre- and postoperatively using a parenchymal microsensor (14). In all patients, an adjustable valve initially set at 200 mmH_2O was used as part of a ventriculoperitoneal shunt. Despite the mean preoperative ICP of 164 ± 64 mmH_2O, the mean postoperative ICP was 125 ± 69 mmH_2O ($P = 0.04$).

This finding, that an ICP reduction occurs in iNPH even with a very high valve opening pressure, may appear to be counterintuitive and physiologically untenable. This misconception, however, arises from a perpetuated oversimplification of ICP and CSF flow hydrodynamics. The concepts of CSF opening pressure (which by default is a mean pressure) and bulk CSF flow have been the standards of hydrocephalus pathophysiology teaching for decades. In reality, the ICP waveform is pulsatile, with significant elevations of ICP occurring as a result of coughing, Valsalva maneuvers, as well as intrinsic vasomotor changes. The interaction between pulsatile ICP and the one-way valve mechanism (inherent to differential pressure valves) is poorly studied. Our continuous ICP recordings demonstrate that *peak* ICPs often exceed 200 mmH_2O among patients with a *mean* ICP of 164 mmH_2O (*Fig. 7*). Even taking into account distal intra-abdominal pressure, one-way CSF egress occurs during these peaks, thereby lowering the mean ICP. This one-way flow "check valve" phenomenon results in the shunt draining CSF even with opening pressures exceeding the mean ICP. This demonstrates that use of a low-pressure valve setting is not necessary and, instead, results in excessive CSF drainage in many patients.

For many years, the cause of subdural hematoma has been attributed to "siphoning." It is well known that negative ICP values are generated by gravity-dependent drainage (22). Multiple valve designs have been developed through the years including antisiphon devices (119), flow-limiting designs (100), and gravitational valves (108, 109) to counteract siphoning, none of which have been shown to prevent (or even reduce the incidence of) subdural hematomas. Our studies, in which ICP was measured 3 days after implantation of a shunt for iNPH, suggest that the degree of intracranial hypotension achieved in the upright (60-degree head-of-bed elevation) is not excessive. Although negative ICP is generated by a ventriculoperitoneal shunt in the upright position, the degree of reduction in ICP at the 60-degree head-of-bed elevation was not significantly out of

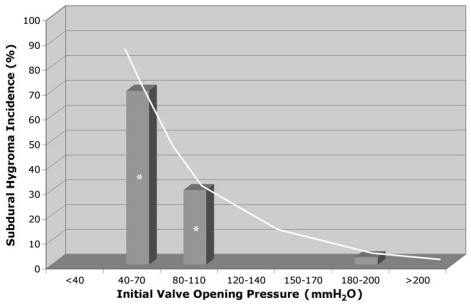

FIGURE 9. *Estimated risk of subdural hygroma formation with iNPH. The Dutch Normal Pressure Hydrocephalus Study (16) documented a subdural hygroma (effusion) incidence of approximately 70% and approximately 30% with low- and medium-pressure differential pressure valves, respectively (data signified with* asterisk*). We encountered a 4% incidence among patients with an initial valve setting of 200 mmH_2O. Combining these data sets results in a near linear relationship between valve opening pressure and subdural hygroma incidence. The hygroma incidence for other valve designs and arrangements has not been well documented.*

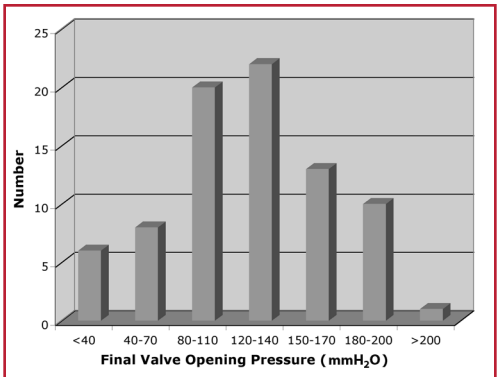

FIGURE 10. *Range of valve opening pressures in the treatment of iNPH. Histogram of final differential valve opening pressure values showing a Gaussian distribution centered at approximately 140 mmH_2O. The wide range of valve opening pressures required indicates that no single valve opening pressure is appropriate for the treatment of iNPH.*

proportion to the ICP reduction at the zero-degree head-of-bed elevation for any given valve opening pressure (*Fig. 8*). It was our conclusion that siphoning played a lesser role in the generation of overdrainage complications with iNPH.

The formation of subdural effusions (hygromas) is thought to be a precursor to and/or risk factor for subdural hematoma formation. Clearly, most stable small subdural effusions do not transition into frank subdural hematomas. However, expanding and/or large (>8 mm) subdural effusions are at higher risk of hemorrhagic conversion. Therefore, it is logical to correlate the risk of subdural hematoma formation with the incidence of subdural effusions after shunt procedures. Here again, clinical evidence supports the relationship between lower-pressure valve settings and CSF overdrainage. The Dutch Normal-Pressure Hydrocephalus Study (16) documented that subdural effusions occurred in 71% of patients with low-pressure valve shunts and in 34% of patients with medium-pressure valve shunts. The analysis of our series of 114 consecutive patients with iNPH, each treated with an initial valve opening pressure of 200 mmH_2O, revealed a subdural effusion incidence of 4%. As shown in *Figure 9*, combining the results of the Dutch Normal-Pressure Hydrocephalus Study with our experience suggests a near linear relationship between subdural effusion and valve opening pressure. Extrapolating to subdural hematoma, this suggests that the high subdural hematoma rates reported with early NPH reports (117) were largely the result of the use of too-low valve opening pressures, not necessarily the result of a lack of antisiphon (or similar) devices. More recent prospective studies, using adjustable valves with valve pressures initially set at higher pressures, reveal subdural hematoma rates as low as 2% (79).

Valve Selection

On the basis of these observations, it is our current practice to use an adjustable valve (Codman Hakim programmable valve) for the treatment of every patient with iNPH. With the primary goal of minimizing the subdural hematoma rate, the valve is initially set at 200 mmH_2O. It is explained to each patient that clinical improvement may not be experienced immediately after the operation, and that, in most cases, a couple of adjustments will be necessary.

Our justification for the routine use of an adjustable valve is based on the range of final valve opening pressures that optimally treat patients with iNPH. In our retrospective evaluation of 114 consecutive patients with iNPH surgically treated with a CSF shunt, the histogram distribution of the final valve opening pressure reveals a roughly Gaussian distribution with most patients in the 120 to 140 mmH_2O range (*Fig. 10*). This finding is in agreement with that of Zemack and Romner (121) noted previously. The wide distribution of final valve pressures shown in *Figure 10* (from <40 to >200 mmH_2O) is *prima facie* evidence that use of a fixed-pressure valve will either under- or overdrain for a significant proportion of patients.

Although our results and those of Zemack and Romner (121) suggest that initially setting the valve at 140 mmH_2O would result in the high statistical likelihood of improvement, the subdural effusion incidence would likely be above 15% based on *Figure 9*. As noted previously, our management strategy is to minimize the incidence of subdural hemorrhage complications at the possible clinical expense of delaying neurological improvement.

Other valve designs (other than simple differential pressure valves) and/or initial pressure settings have been successfully used for iNPH. Valve designs incorporating a siphon-control device, such as the Medtronic Strata valve (Medtronic, Goleta, CA), may lower the incidence of over-

drainage complications; however, as noted previously, our data suggest that siphoning may play a lesser role in the etiology of postshunt subdural hygroma formation. Additionally, antisiphon devices predispose some patients to a low-pressure hydrocephalus syndrome (13). So-called gravitational valves, in which the opening pressure varies based on body position, are another approach to decreasing the hydromechanical effect of siphoning (83). Again, the use of a low-pressure valve setting with gravitational valves in the recumbent position would still likely result in overdrainage of CSF (14) and therefore not eliminate the risk of subdural hematomas. Whatever valve is chosen, it is likely more important to understand the benefits and disadvantages of each and select an appropriate management scheme.

Postoperative Management

Our management algorithm places high priority on reducing the morbidity associated with overdrainage complications, specifically, subdural hematoma formation. As noted previously, we begin with an initial valve pressure setting of 200 mmH_2O and then lower it every 2 to 3 weeks by 30 mmH_2O increments if clinically indicated. The philosophy is that any inconvenience associated with the possible delay in clinical improvement is offset by the avoided morbidity associated with a subdural hematoma. The management algorithm is summarized as follows:

- If there is no significant clinical improvement, the patient is seen at 2- to 3-week intervals and consideration is given to lower the valve setting by 30 mmH_2O. As noted previously, the aim should be to achieve at least the same level of improvement experienced after the temporary lumbar drainage. It is our observation that clinical improvement with iNPH tends to occur as if a toggle switch was turned on rather than in slow, incremental steps. In other words, if a patient experiences a significant improvement, there is little to no use in further decreasing the valve pressure with the hope of gaining further improvement.
- In addition to managing the valve conservatively, we proactively scan for subdural fluid collections even in asymptomatic patients. Most subdural effusions, and even hematomas, that develop in patients with iNPH are clinically silent. Seemingly paradoxical, patients with iNPH with subdural effusions or small subdural hematomas often report clinical improvement. Therefore, we obtain a noncontrast computed tomographic scan at 1 month postoperatively or after every other downward valve adjustment. The computed tomographic scans are obtained for two reasons: to assess for subdural effusion or subdural hematoma formation and to determine whether there has been a significant reduction in ventricular size.
- The decision of whether to lower the valve setting is based on several considerations. First, there should be a lack of, or insignificant, clinical improvement. The second consideration relates to the CT findings, if a scan was obtained. The valve pressure should not be lowered if the computed tomographic scan reveals a subdural effusion or subdural hematoma (see the discussion under Management). If there is no subdural effusion and no significant change in ventricle size (based on <2-mm change in the maximal bifrontal ventricular distance), then lowering the valve setting by 30 mmH_2O has minimal risk. We proceed in this incremental manner all the way down to the valve's lowest setting.
- If the ventricles are identified to be significantly reduced in size, it is our experience that patients who are clinically improved will have experienced near maximal improvement. On the other hand, patients who have not experienced any improvement are unlikely to do so, and therefore, likely represent true nonresponders.

Management of Subdural Fluid Collections

Asymptomatic Subdural Effusion. If a subdural effusion is less than or equal to 8 mm in diameter, we generally leave the valve at its existing setting and repeat the CT in 7 to 14 days to make sure the effusion is stable in size. If the subdural effusion is between 8 and 15 mm thick, the valve pressure is raised by 30 mmH_2O and a repeat scan is obtained 1 week later. At that time, the valve is again raised by 30 mmH_2O, and the CT is repeated 1 week later, and so on. For large subdural effusions (>15 mm), the valve is immediately reset to 200 mmH_2O and a repeat CT is obtained in 1 week.

Symptomatic Subdural Effusion. The most common symptom related to a subdural effusion is new-onset headaches, although focal neurological deficits can occasionally be present. If the degree of morbidity is minimal, the management can be similar to that described previously. Patients with neurological deficits generally require a more expedited management strategy. For smaller subdural effusion collections, an abrupt change to 200 mmH_2O may be appropriate. In some cases, placement of a temporary subdural catheter drain or a subdural-to-peritoneal shunt may be necessary. If a surgical drainage approach is chosen, it is important to increase the valve opening pressure accordingly so as to aid with the resolution of the subdural fluid collection.

Subdural Hematoma. Patients with hemorrhage conversion of a subdural effusion, or a frank subdural hematoma, should be treated on an urgent basis, and sometimes even admitted them to the hospital. Anticoagulation should be reversed and/or antiplatelet medication stopped. A consideration should be made regarding prophylactic anticonvulsants. Smaller, asymptomatic subdural hematoma collections can be often resolved after increasing the valve pressure to 200 mmH_2O. Larger and/or symptomatic subdural hematoma collections typically require surgical evacuation in addition to valve pressure adjustments.

If overdrainage problems, including subdural effusion and/or subdural hematoma, present or continue despite the valve set at its maximal pressure setting (200 mmH_2O for the Codman Hakim programmable valve), we typically revise the shunt system by adding a fixed-pressure differential pressure valve (for example, a 70-mmH_2O valve) in series with the programmable valve. Another choice is to substitute with another adjustable valve with a higher valve setting.

The determination of shunt underdrainage in iNPH can be another clinical challenge. For example, based on the previously described algorithm, a question arises as to what to do if there remains no clinical improvement despite the valve being at its lowest setting (30 mmH_2O for the Codman Hakim programmable valve). This state has been referred to as "low-pressure hydrocephalus" and it occurs with higher incidence in patients with antisiphon devices (13, 73, 81, 82). As noted previously, if the computed tomographic scan reveals a reduction in ventricular size, this scenario defines a nonresponder. If there has been no change in ventricular size, then we consider one of the following two options:

1) If the valve contains an antisiphon device, a siphon control device, or other mechanism to limit drainage, we replace the valve with an adjustable differential pressure valve (Codman Hakim programmable valve). At the same time, we consider changing the shunt to a VA shunt. It is our general experience that VA shunts drain more CSF than ventriculoperitoneal shunts in patients with iNPH.
2) Progressive subatmospheric ventricular drainage is an option that we typically reserve for patients whom we think have a higher probability of clinical improvement potential. Our protocol (13) has been described previously but, in general, involves the placement of a separate ventriculostomy catheter and progressive lowering of the drainage level well below the external auditory meatus. The goal is to achieve a reduction in ventricular size, at which time a dramatic clinical improvement is observed in responders. Drainage levels as low as −40 cm below the external auditory meatus may be required.

Management of the Shunt Patient with Headaches

The evaluation and treatment of headaches (or other subjective complaints, such as dizziness) in a shunted patient can be challenging. Headache is a common symptom of hydrocephalus, except for patients with NPH. The diagnosis of an obstructed shunt, with overt acute ventriculomegaly, is not difficult in younger patients. A symptomatic shunted patient with slit ventricles poses a different situation, and can be managed as described previously (see the discussion under Adult Slit Ventricle Syndrome).

Positional headaches in the shunted patient require special consideration. Within the first month after shunt placement, revision, or downward valve adjustment, consistently occurring headaches in the upright position that are relieved in the recumbent position are indicative of overdrainage headaches (akin to post-lumbar puncture headaches). If a patient has an adjustable valve, overdrainage headaches are usually easily addressed unless the valve is at it highest setting. Surgical intervention may then be required with addition of a gravitationally assisted device or, alternatively, an antisiphon-type device.

The etiology of shunt-related overdrainage headaches is postural-induced intracranial hypotension. The converse, however, is not necessarily true. Intracranial hypotension causes headaches in only a minority of patients. This fact is evident based on the vast lumbar puncture experience. Less than 10% of patients undergoing lumbar puncture experience spinal "headaches," although presumably all are subjected to some period of intracranial hypotension after the procedure. It is probable that the same subset of "vulnerable" patients is susceptible to overdrainage headaches after a shunt procedure. This point is important with regard to the interpretation of ICP monitoring and MRI results. It has been suggested that the diagnosis of shunt-related overdrainage headaches can be confirmed by ICP monitoring. Although it is true that the lack of intracranial hypotension excludes the diagnosis, its presence is not diagnostic. As noted previously, shunt-related overdrainage headaches present acutely after an intervention that may have led to intracranial hypotension, and therefore, it is a clinical diagnosis. A shunted patient presenting with headaches that have some possible, and usually inconsistent, postural dependence in a delayed fashion after any intervention is highly unlikely to have shunt-related overdrainage headaches. Shunt systems spontaneously underdrain over time, not overdrain.

The finding of diffuse pachymeningeal enhancement of MRI imaging is likewise not diagnostic of shunt overdrainage. The fact that the finding is diagnostic in the clinical setting of *spontaneous* intracranial hypotension cannot be extrapolated to the shunted patient. It is our observation that the majority of patients shunted with differential pressure valves receiving contrast MRI studies exhibit diffuse pachymeningeal enhancement. Our data, as well as other reports (22), document the existence of negative ICP values in the upright position. Therefore, like ICP monitoring, clinical correlation is required. In an asymptomatic shunted patient, the finding of diffuse pachymeningeal enhancement might better be considered as confirmatory of a patent shunt.

A common complaint of patients treated with a standard differential pressure valve is transient headaches occurring after coughing, sneezing, or bending over. These are benign in nature and rarely require intervention. It is our anecdotal experience that valve designs incorporating flow limitation such as the Codman Siphonguard reduce this type of headache.

Most shunted patients who report headaches will not have a clear history of postural headaches. Many will have vague headache complaints, although a "pressure" sensation is common. For patients in which imaging does not indicate an anatomic explanation for headaches, and have disabling headaches despite a concerted pain management regimen, we consider continuous ICP monitoring. This is performed as an inpatient with a parenchymal monitor (Codman microsensor) over a 48- to 72-hour period. We have found that the data obtained from a one-time shunt tap, or lumbar puncture, to be incomplete and sometimes spurious. Although some have found a high correlation between ICP measured by lumbar puncture versus brain tissue (36, 69), the lumbar puncture technique used in these studies was highly specialized and controlled (including having the patient resting comfortably on a special bed with a hole to accommodate the supine position, which differs from the straining patient curled up laterally in the fetal position). Importantly, continuous ICP moni-

toring (recording) allows for instigating factors to be investigated such as exertion and postural changes. Although the patient has the ICP monitor in place, he or she is asked to keep a detailed hourly diary (when awake) of the intensity of these headaches, what they were doing at the time, and the position of their body. If possible, it is best if the patient cannot see his or her ICP values during this monitoring period. It is important to measure ICP values at different body positions, including prolonged sitting and standing. At the end of the 48-hour monitoring period, it is insightful to correlate ICP values with the headache intensities recorded in the diary. In the majority of cases, there will be no correlation. Reviewing these data with the patient will let them see that the "pressure" sensations they are experiencing have nothing to do with their actual ICP. Equally or more importantly, this information is useful to the pain management physician who often limits treatment as a result of fear that they are masking intracranial pathology.

If the diary complaints and ICP recordings do show a correlation, then these cases are often amenable to an appropriate shunt (usually valve) revision. We recommend that the ICP monitor be retained after the surgical intervention to allow confirmation that the ICP abnormality has been rectified.

SUMMARY AND FUTURE DIRECTIONS

The management of adult patients with hydrocephalus has changed significantly over the past two decades. Neuroendoscopic procedures have obviated the need for CSF diversionary shunts in many patients or, in other cases, allowed for the simplification of what would otherwise be complex plumbing arrangements. Advanced MRI imaging, particularly the CISS sequence, has allowed for the identification of a greater number of noncommunicating hydrocephalus cases that are amenable to endoscopic management. As such, the evaluation of every patient with hydrocephalus should begin with an assessment of whether a CSF shunt can be avoided or removed.

For patients requiring CSF diversion, it is our opinion that adjustable valves maximize the likelihood of both attaining a positive clinical response and avoiding or reversing complications. The fact that one patient may require a differential pressure valve opening pressure of 20 mmH_2O, whereas another needs 400 mmH_2O, is indicative of the wide range of hydrodynamics encountered with this disorder. To date, it is not possible to determine, preoperatively, the optimal valve type or pressure setting to select for any given patient. Fixed-pressure valve designs such as flow-limiting devices and multistaged valve mechanisms do not address heterogeneity across patients. Our studies, thus far limited to patients with iNPH, suggest that siphoning plays a lesser role in shunt overdrainage than previously assumed, and that we must be more cognizant of excessive CSF flow in the recumbent position. We need an improved understanding of the hydromechanics of hydrocephalus, both treated and untreated, to optimize therapy for individual patients.

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COMMENTS

Bergsneider et al. provide us with an excellent monograph of their work in the diagnosis and management of hydrocephalus and a thorough overview of their experience treating pediatric and adult patients. In some controversial areas, the discussion lacks hard data, and the authors emphasize that more work is needed. This is always the case when it is difficult to organize and fund randomized prospective clinical trials for answering these questions. It is particularly gratifying that attention is paid to the evidence-based international guidelines for diagnosis and management of idiopathic normal-pressure hydrocephalus (NPH), which have been a valuable aid in documenting the current state of the art. Certainly, there are differing points of view with regard to the etiology of NPH, and years of research at the bench and bedside are required to answer these questions. The management of patients developing a subdural hematoma subsequent to shunt placement by raising the valve pressure is advantageous; however, the NPH patient, with a pronounced gait disturbance and dizziness, is placed at risk for falls during the period that the valve pressure is increased. As Bergsneider et al. state, there is tremendous heterogeneity in the NPH patient cohort, and the tolerance of each patient to subdural effusion varies considerably. Therefore, the surgeon must walk a fine line between accepting this period of vulnerability, which may extend into weeks, as opposed to evacuating the subdural hematoma. In summary, this is a job well done and a valuable aid to all working in this field.

Anthony Marmarou
Richmond, Virginia

This is a nice summary of the current state of affairs of adult hydrocephalus. Bergsneider et al. outline their treatment strategies for a number of common circumstances including the adult patient with congenital or childhood-onset hydrocephalus, adult slit ventricle syndrome, multicompartment hydrocephalus, noncommunicating hydrocephalus, communicating hydrocephalus, NPH, and shunt patients with headaches. As the authors point out, there is little Class I or II evidence to support many of the treatments and diagnostic tests discussed. Nevertheless, the authors' experience is considerable, and this could be regarded as the best advice available.

James M. Drake
Toronto, Canada

This is a wonderfully comprehensive account of adult hydrocephalus that every neurosurgeon should read, internalize, and use in daily practice. It demonstrates that adult hydrocephalus includes much more than idiopathic NPH. On the contrary, it includes many conditions that may be complicated for the practicing neurosurgeon.

The patient who has had early hydrocephalus and now reaches adulthood is an example. This patient may have slit ventricle syndrome or other syndromes that make it very problematic to deal with shunt malfunction. An adult patient with the "stiff brain" of slit ventricle system can become sick as quickly as a child, and the adult neurosurgeon must be willing and able to deal with this problem. The use of ventriculoscopy for management of such patients is also an important new concept for neurosurgeons.

The management of idiopathic NPH is also an important topic not much dealt with in the neurosurgical literature. For a patient who has this condition and has a successful shunt, there are few interventions that are more successful in neurosurgery. A patient can be changed from a nonambulatory, incontinent nursing-home inhabitant to a self-sufficient member of society with a simple 1-hour procedure. The use of lumber drainage to predict shunt response and of a variable-pressure valve to optimize outcome are important components of the 21st century management of this condition. Bergsneider et al. have made a major contribution to the neurosurgical literature with this article.

Peter M. Black
Boston, Massachusetts

In this extensive review of the management of adult hydrocephalus, Bergsneider et al. offer recommendations in neurosurgical care drawn from published clinical series, scientific data, and personal experience. As they rightly point out, our care for these patients is rarely based upon the results of well-designed, prospective trials. There has been a recent trend in the care of hydrocephalic children that offers recommendations based upon multi-institutional, prospective trials, a trend that we hope will extend to encompass all age groups (1–3).

First, it is clear that an increased demand is being placed upon the neurosurgeons treating hydrocephalus, as more patients with this condition "transition" from childhood to adulthood. This phenomenon has not been adequately prepared for on many fronts, including residency training, institutional limitations, and government funding.

The second underlying thread delivered by Bergsneider et al. is the all-important adage of relying on the impression of the patient or family members with respect to probable shunt malfunction. I would propose that my diagnostic accuracy of shunt-related malfunction increases linearly, albeit with a small slope, with a more familiar interaction with the patient or family. There is no replacement for this insight, a component that is too often lost in our reliance on data, which governs our clinical decisions. I cannot foresee that the optimal care of the shunted patient can ever be completely reliant on a flow diagram.

I agree fully that endoscopic third ventriculostomy has improved the care of a substantial number of patients who would not otherwise be enjoying the benefit of shunt independence. However, accomplishing shunt independence in the previously shunted patient can be challenging. This is clearly evident in their reported failure in five out of nine patients with slit ventricles. Shunt externalization and clamping, in my experience, infrequently results in expanded ventriculomegaly without overt symptoms. Furthermore, at the time of endoscopic third ventriculostomy in this population, the procedure is usually hampered by a rapid decline in the functional workspace of the intraventricular compartment and a thickened floor. Last, it has not been my experience that a minimum "premesencephalic" interval is necessary for safe endoscopic third ventriculostomy. If a patient is a candidate as judged by noncommunicating criteria, I offer exploration with some modification in technique (i.e., stereotactic guidance). Thus far in this scenario, adequate fenestrations have been possible without complications. Of course, as stressed by the authors, relative risk is an important function of a surgeon's experience.

This summary offers a comprehensive overview of the diagnostic and therapeutic issues facing neurosurgeons caring for adults with

hydrocephalus. It will probably serve as a reference for the student and veteran clinician in years to come.

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Section view of a human head (c. 1493–1494), by Leonardo da Vinci. Windsor, Royal Library (RL 12603r; K/P 32r). (From: Pescio C, Crispino E (eds): *Leonardo: Art and Science*. Florence, Giunti, 2000).