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Selecting patients for endoscopic third ventriculostomy

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At the annual meeting of the American Association of Neurological Surgeons Meeting in 1995, Professor Fred Epstein was called to discuss our paper on the use of endoscopic third ventriculostomy (ETV) as the ultimate treatment for the "slit ventricle syndrome" (SVS) as presented by Dr Jonathan Baskin [1]. Dr Epstein was surprised to learn that we were viewing all previously shunted patients with shunt problems as potential candidates for the procedure and wondered aloud whether patients with normal pressure hydrocephalus (NPH) might not be considered candidates for ETV.

The thought process leading to the following presentation regarding who is and who is not a candidate for ETV was largely generated by that discussion. Can we know beforehand who will and who will not benefit from an ETV? What are the absolute contraindications for performing the procedure? What are the relative contraindications? How can we analyze the risk-benefit ratio for ETV in various clinical settings? For the most part, defining the role of ETV in the overall management of hydrocephalus must await large, multicenter, randomized, prospective trials. In the interim, I will attempt to define a rational approach to the problem in partial answer to the posed questions.

Anatomy and biophysics of the cerebrospinal fluid as it relates to third ventriculostomy

The credit for defining the basic pathophysiologic mechanisms in hydrocephalus goes to Walter Dandy and his colleagues at Johns Hopkins University for work done between the second and fourth decades of the twentieth century. There had been many previous pathologic descriptions of the effects of hydrocephalus on cadaveric material. Dandy and Blackfan [2] studied laboratory animals and patients with hydrocephalus by injecting supravital dyes into the lateral ventricles of the subjects. They then performed lumbar punctures to determine whether the dye could be recovered in the spinal subarachnoid spaces (SSASs) [3]. Based on this information, Dandy and Blackfan classified hydrocephalus into communicating (the dye was recoverable) and obstructive or noncommunicating (the dye was not recoverable). As a result of these findings, they recommended attempting to create a communication between the third ventricle and the subarachnoid spaces by performing an open craniotomy and initially resecting one of the optic nerves.

By applying Dandy's classification, patients with noncommunicating hydrocephalus would be considered candidates for ETV to create an internal bypass for the treatment of hydrocephalus. Over the years, there has been a tendency to equate noncommunicating hydrocephalus with triventricular hydrocephalus, which could be diagnosed by air studies and, subsequently, by CT and MRI. In 1960, Ransohoff and Epstein [4] voiced their objection to the Dandy classification. They believed that all hydrocephalus was obstructive and preferred to classify the condition into intraventricular obstructive hydrocephalus and extraventricular obstructive hydrocephalus. The latter would be consistent with Dandy's communicating hydrocephalus. Based on this discussion, many patients with communicating hydrocephalus are good candidates for ETV.

With current imaging technologies, it is usually possible to determine the actual site of the obstruction and to plan treatment to address the specific pathophysiologic mechanisms involved. In

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our laboratory, we have measured the amount of resistance that can be anticipated at each point of potential obstruction. Because of the presence of multiple ventricular catheters, we have also attempted to measure intraventricular pressures in various components in human beings. Fig. 1 is a schematic diagram of the anatomy of the cerebrospinal fluid (CSF) pathways in a patient with aqueductal stenosis as a hydraulic analogue to an electrical circuit. In normal animals, pressure differentials can be measured only at the level of flow of CSF from the cortical subarachnoid spaces (CSASs) into the superior sagittal sinus (SSS) [5,6]. For CSF to be absorbed at the level of the SSS, intracranial pressure (ICP) must be 5 to 7 mm Hg higher than the pressure in the SSS. Fig. 2 is a schematic diagram of the physical effect of an ETV. Essentially, the procedure is an internal bypass between the third ventricle and the interpeduncular cistern in the CSASs.

Given this paradigm, all patients with obstructions between the third ventricle and the CSASs are potential candidates for ETV. Not only can ETV treat hydrocephalus caused by aqueductal stenosis, but it is at least of theoretic benefit in obstruction of the outlet foramina of the fourth ventricle and to blockage of CSF flow at the level of the basal cisterns between the SSASs and the CSASs. Only when CSF flow is obstructed at the level of the arachnoid villi or venous flow from the sagittal sinus is restricted do patients fail to benefit from ETV. These forms of distal obstruction represent absolute contraindications to ETV.

With contemporary neuroimaging techniques, it may or may not be possible to determine the actual site of obstruction to the flow of CSF. The more definitive the diagnosis of the site of obstruction, the better is the physician's ability to predict the outcome of ETV when used to treat patients with hydrocephalus. Some patients with hydrocephalus may have an obstruction at more than one point along the CSF pathway. In spina bifida, hydrocephalus caused by a Chiari II

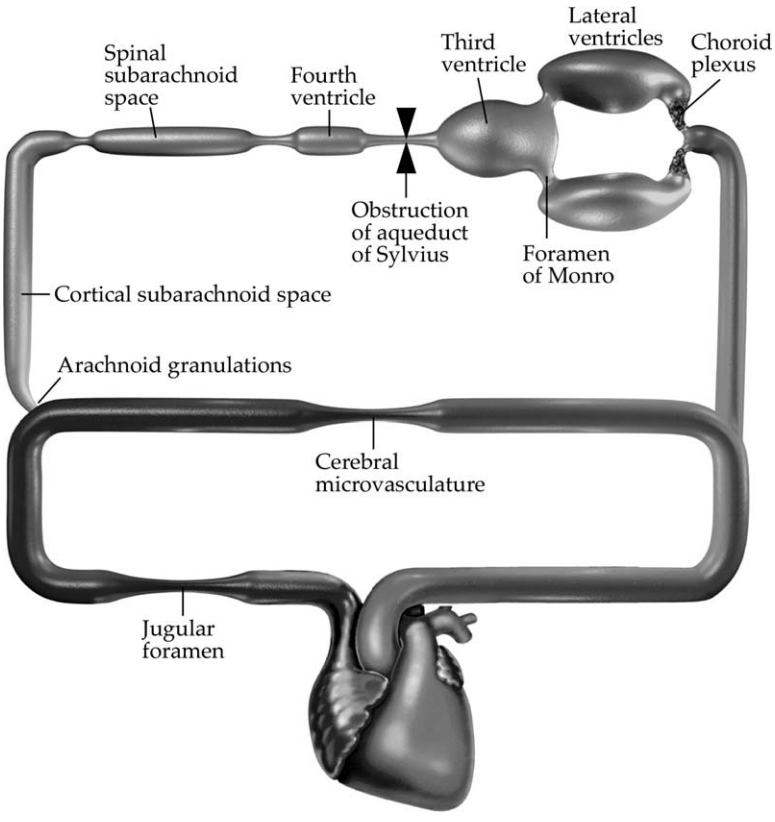

Fig. 1. Schematic hydraulics of the cerebrospinal fluid system with aqueductal stenosis. (Courtesy of Barrow Neurological Institute.)

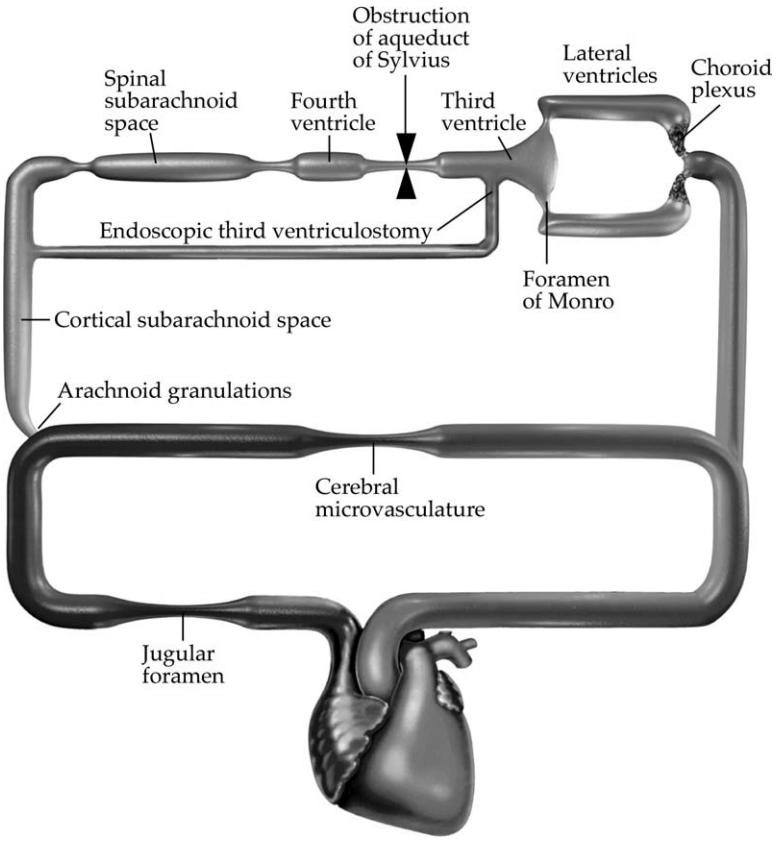

Fig. 2. Schematic diagram of the effect of performing an endoscopic third ventriculostomy on the circuit diagram of cerebrospinal fluid flow. (Courtesy of Barrow Neurological Institute.)

malformation is associated with as many as four different sites of potential obstruction to the flow of CSF [7]. Based on patients' clinical manifestations at shunt failure, all these potential sites of obstruction actually occur and any patient may have more than one site of obstruction.

Patients with hydrocephalus caused by intraventricular or subarachnoid hemorrhage (SAH) for whatever cause (prematurity, trauma, or aneurysmal bleeding) usually have a CSF absorptive difficulty at the level of the basal cisterns (SSASs to CSASs). They could have an obstruction at the level of the arachnoid granulations, however, which would not respond to ETV. Patients who suffer significant infections may develop hydrocephalus. The most common location is also the basal cisterns. Patients with an infection may have an obstruction at the level of the outlet foramina of the fourth ventricle or at the level of the arachnoid granulations, however.

Absolute contraindications for endoscopic third ventriculostomy

"Communicating hydrocephalus": obstruction to the absorption of cerebrospinal fluid

ETV should not be performed if CSF is already flowing unimpeded between the ventricles and the interpeduncular cisterns. It also should not be done if it is technically impossible to manipulate the endoscope within the lateral and third ventricles to a position that allows the floor of the third ventricle to be visualized. These are the only two absolute contraindications for performing the procedure. In the first instance, flow between the ventricles and the CSASs is not impeded. Bypassing the aqueduct of Sylvius, outlet foramina of the fourth ventricle, and the basal cisterns offers no advantage, because the point of obstruction is "downstream" from the internal bypass. As can be seen from a review of Fig. 2, this condition occurs only if the arachnoid

granulations are obstructed or the pressure of the SSS is increased markedly.

As an isolated event, obstruction of the arachnoid granulations is quite rare. Pathologically, congenital absence of these structures is the cause of at least some cases of benign familial megalencephaly or external hydrocephalus. Radiologically, this type of obstruction shows mild to moderate ventriculomegaly and distention of the CSASs [8–10]. The terminal absorptive mechanisms can be clogged by blood or by inflammatory or tumor cells. This clogging is usually accompanied by thickening and scarring of the arachnoid in the basal cisterns. Although the CSF absorptive failures in acute SAH and meningitis may be related to an obstruction at this level, the hydrocephalus usually results from an obstruction between the SSASs and CSASs, a condition that is amenable to ETV.

Hydrocephalus caused by obstruction of absorption to CSF at the level of the arachnoid villi is never an all-or-none phenomenon. It is pressure dependent in that the presence of the particulate matter changes the pressure flow characteristics of the valvular mechanism already present. Under normal conditions, the arachnoid villi (microscopic structures) contained within the arachnoid granulations (macroscopically visible) function as a differential pressure valve between the CSASs and SSS with a closing pressure of 5 to 7 mm Hg (70–100 mm H_2O). In adults with closed sutures and fontanels, clogging of the arachnoid villi usually increases ICP, with a minimal increase in ventricular size. In the context of traumatic or aneurysmal SAH, the usual indication for shunting is the failure to be able to wean the patient from an external ventricular drain in the presence of minimal ventriculomegaly. The shunt can usually be removed later in the course of a patient's life, but ventricular size may increase significantly with symptoms at the time of shunt failure [11]. In this case, the point of obstruction usually is at the level of the basal cisterns and no longer at the level of the arachnoid granulations.

In infants with neonatal intraventricular hemorrhage or meningitis, clogging of the arachnoid villi increases both the CSAS and ventricular volume. In this situation, the open fontanels and distensibility of the sutures allow CSF to accumulate, because ICP cannot increase to the point that it will overcome the valvular mechanisms in the arachnoid villi. This condition responds to increasing ICP by wrapping the head with an ace bandage. ICP then increases to a point where CSF can be absorbed [12].

The second point of obstruction leading to the type of "communicating hydrocephalus" that cannot be affected by ETV involves obstruction to outflow of venous blood from the SSS. In adults, marked increases in pressure in the SSS lead to pseudotumor cerebri rather than to hydrocephalus [13,14]. In infants with open fontanels and distensible sutures, marked increases in pressure in the SSS lead to marked ventriculomegaly associated with concomitant increases in the volume of the CSASs. At presentation, it may be difficult, if not impossible, to discern that the hydrocephalus is caused by increased venous pressure. This form of hydrocephalus can lead to secondary obstruction of the aqueduct as the brain stem is displaced inward and the temporal horns of the lateral ventricles displace the midbrain medially [15]. This form of hydrocephalus has been well studied in the context of achondroplasia and craniofacial syndromes [16–20]. In the case of Crouzon's and Pfeiffer's syndromes, the hydrocephalus may not be evident until a formal cranial remodeling operation has been performed. These patients share the same syndrome in that their ventricles do not dilate to abnormally large volumes at the time of shunt failure. They show signs of overtly increased ICP without ventriculomegaly, a condition that has been referred to as "normal volume hydrocephalus" [21].

Unresponsive ventricles: ventricles too small or too distorted to manipulate an endoscope safely

Performing an ETV requires that the endoscope be placed in the lateral ventricle and manipulated through the foramen of Monro to the floor of the third ventricle, where the hole can be made. The smaller the ventricles are, the more difficult it becomes to perform the manipulations needed to fenestrate the floor of the third ventricle. If the size of the ventricles is normal or smaller than normal after chronic shunting, it may be impossible to manipulate the endoscope safely to the appropriate location to perform the fenestration. There are two important circumstances in which third ventriculostomy is contraindicated in this context. Marked enlargement of the massa intermedia of the third ventricle is a well-recognized concomitant of the Chiari II malformation in the context of spina bifida. In these situations, the walls of the third ventricle are often closely opposed because of the size of the massa intermedia despite the

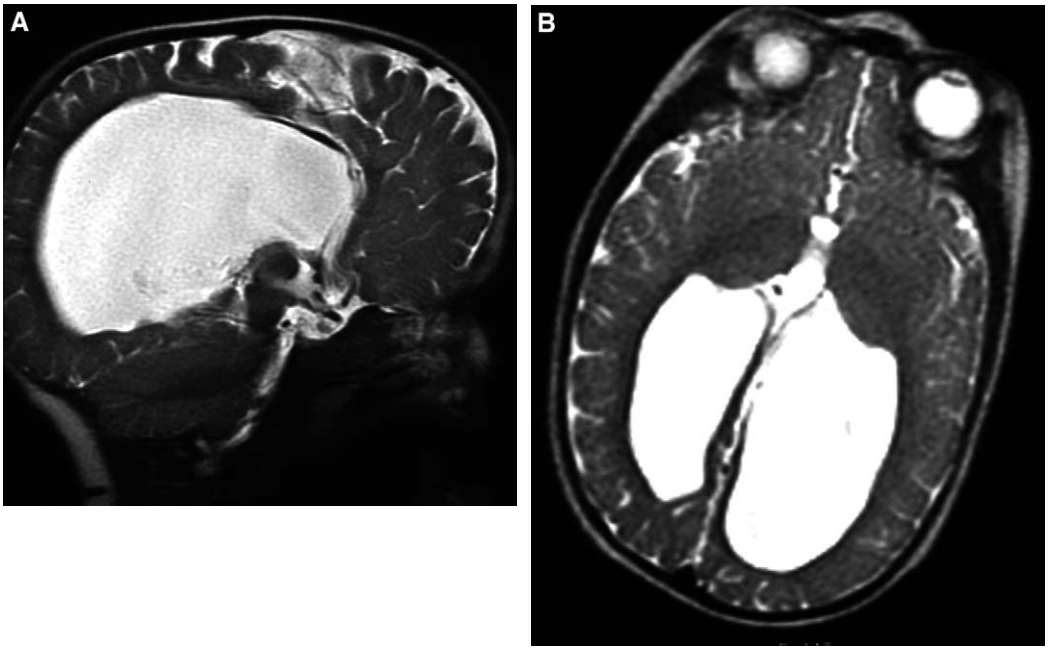

Fig. 3. MRI of a patient with spina bifida showing enlargement of the massa intermedia and a small third ventricle despite marked enlargement of the lateral ventricles. (*A*) Sagittal T2-weighted MRI demonstrating the enlarged massa intermedia in a small third ventricle despite massive enlargement of the lateral ventricles. (*B*) Axial T2-weighted MRI demonstrating the difference in size between the lateral and third ventricles.

enlargement of the lateral ventricles (Fig. 3). The third ventricle can be markedly distorted in patients with a Chiari II malformation. Even when the third ventricle can be cannulated, the anatomic landmarks can be so distorted that it may be impossible to find a point on the floor of the third ventricle to make the fenestration.

The second context in which ETV is contraindicated because it would be unsafe to manipulate the endoscope occurs in patients with normal volume hydrocephalus as described previously. These patients have smaller than normal ventricles, and the ventricles do not expand at the time of shunt failure. These patients should not undergo ETV for two reasons. First, the procedure is associated with a high risk of damaging the columns of the fornix, mamillary bodies, cerebral peduncles, or hypothalamus. Second, ETV in this context is unlikely to be of benefit to the patient. We have performed iohexol ventriculography in 31 such patients, and 28 showed free communication from the ventricle to the interpeduncular cistern [22]. In the 3 patients who did not show such communication, treatment of their hydrocephalus was complicated by a significant infectious process after the original shunt had been placed.

Some nonresponsive ventricles have resulted in surprising observations. In one patient whose hydrocephalus was associated with a pineal tumor originally treated in infancy, overt shunt failure occurred despite no change in ventricular volume. Ventriculography revealed that the dye injected through the shunt into the lateral ventricle rapidly flowed into the interpeduncular cistern area (Fig. 4). The hydrocephalus of patients with normal volume hydrocephalus is related to increased venous pressures, and they are not candidates for ETV for both reasons.

Selection of candidates for endoscopic third ventriculostomy as an initial procedure in the treatment of hydrocephalus

Late occlusion of the aqueduct of Sylvius

Hydrocephalus caused by occlusion of the aqueduct of Sylvius is most common in infants who manifest overt hydrocephalus as newborns or whose head circumference rapidly increases after birth. This indication would seem to be ideal for performing an ETV, because occlusion of the aqueduct obstructs CSF flow between the third

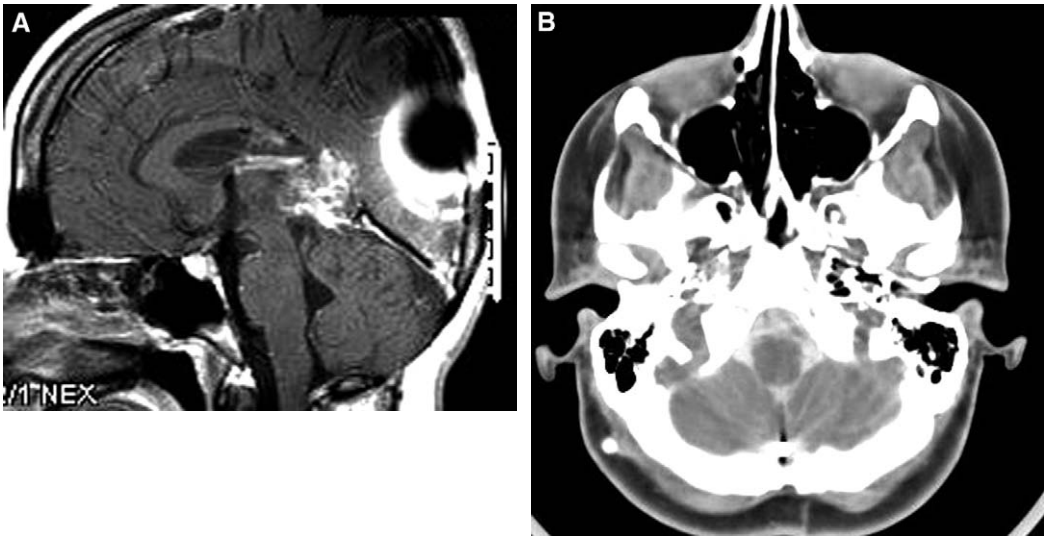

Fig. 4. (*A*) Sagittal MRI of a child with hydrocephalus associated with a pineal region tumor showing communication between the lateral ventricles and spinal subarachnoid spaces despite the presence of the tumor. (*B*) CT of the basal cisterns after iohexol has been injected into the lateral ventricle showing the flow of contrast into the basal cisterns, thus confirming communication in the cerebrospinal fluid pathways.

and fourth ventricles. Theoretically, CSF should be left in the interpeduncular cistern in communication with the arachnoid granulations, which should be normal. Creating this internal bypass between the third ventricle and the cistern should normalize CSF dynamics.

Aqueductal stenosis is often diagnosed by MRI only when triventricular hydrocephalus is present and the size of the fourth ventricle is normal. Triventricular hydrocephalus does not always mean that the underlying cause of the hydrocephalus is aqueductal stenosis. The phenomenon of communicating hydrocephalus causing dilatation of the temporal horns of the lateral ventricle and compression of the midbrain from both sides leading to a secondary occlusion of the aqueduct of Sylvius has been discussed previously. In these patients, shunting opens the aqueduct of Sylvius to CSF flow again [15]. Depending on the initial site of obstruction, the patient may or may not be a candidate for ETV.

Regardless of the site of obstruction to CSF flow, infants are usually poor candidates for a third ventriculostomy, presumably because the ventricles fail to respond after third ventriculostomy despite the blockage at the aqueduct. In these cases, the ventricles are quite large; unlike shunting, which literally sucks CSF from the brain, ETV normalizes CSF absorption. The latter process requires intraventricular pressure to increase 5 to 7 mm Hg greater than atmospheric pressure, which may be impossible in babies with open anterior fontanels. Some authors suggest that these infants should not be candidates for ETV because of their poor response rate [23,24]. Other authors quote low rates of shunt independence after ETV but believe that the benefit is great enough and the risks of the procedure are low enough to justify ETV in infants in an attempt to avoid placing a shunt [25].

Aqueductal stenosis is diagnosed in older children and adults in two contexts. It is difficult to establish treatment criteria for and to assess the outcome of treatment in patients who develop symptomatic hydrocephalus later in life and have extremely large heads. Obviously, these patients have had severe ventriculomegaly from infancy. Oi et al [26] have termed this condition *long-standing overt ventriculomegaly of the adult* (LOVA). These patients usually have no overt symptoms after undergoing an imaging study for a seizure or minor head injury. Frequently, they have no symptoms related to their hydrocephalus. When asked, some patients identify daily headaches, clumsiness, or poor memory that may or may not have changed recently.

How to treat these patients and, indeed, whether to treat these patients are controversial

issues. Some authors strongly believe that these patients will benefit from intervention, usually ventricular shunting [27]. Data showing improvement in these patients are hard to find, and such patients seldom have high ICP. My approach to these patients is to obtain formal neuropsychologic studies to determine whether they exhibit problems with higher cognitive function. If so, it is important to determine whether the deterioration is recent or long standing. I then provide patients with the pertinent information and attempt to help them reach a treatment decision. A significant decrease in the size of the lateral ventricles after shunting or ETV is unusual. Shunts can be shown to be working manometrically, but it may be difficult to determine whether an ETV has produced the best outcome. Imaging studies, at least cine MRI through the floor of the third ventricle and preferably the injection of iodinated contrast material into the lateral ventricles to trace its flow into the basal cisterns, is needed to confirm that CSF dynamics have been maximized.

There is one important caveat if the decision is made to attempt to treat LOVA with ETV. Occasionally, the head is so large that some neuroendoscopes are too short to reach the floor of the third ventricle. The distance from the skull to the floor of the third ventricle can be measured on the console of the CT or MRI scanner. A selection of endoscopes should be available to obviate this problem.

In the second clinical syndrome, older patients develop hydrocephalus caused by aqueductal stenosis from the presence of severely increased ICP. Patients often seek treatment from an ophthalmologist for some form of ophthalmoplegia, such as bilateral sixth nerve palsies or Parinaud's syndrome. On examination, the patient is found to have bilateral papilledema. This phenomenon can occur as an isolated event with obstruction caused by fusion of the walls of the aqueduct of Sylvius as occurs in some cases of neurofibromatosis 1. This overt form of late aqueductal stenosis is more likely to result from small benign tumors (tectal gliomas) of the aqueduct. These are true tumors, but their prognosis in terms of propensity to grow or disseminate is usually good [28]. A direct attack on these tumors, even for a biopsy, is seldom warranted.

This acute adult form of hydrocephalus from aqueductal stenosis is perhaps the most compelling and most satisfying condition to treat with ETV. The success rate for this particular subset of patients is 75% to 80%. The rate of significant morbidity, including endocrinopathy, memory deficits, and hemiparesis, has been reported to be 3%. After 1 year, less than 1% are still troubled by complications of ETV [24]. In this situation, the risk-benefit ratio favors ETV over internal shunting.

Hydrocephalus associated with newly diagnosed brain tumors, particularly of the posterior fossa

This area of study is evolving, and the use of ETV before tumor removal is discussed more than it is reported in the literature. Posterior fossa tumors usually manifest with hydrocephalus and acutely increased ICP. The blockage of the CSF flow meets the criteria for ETV in that CSF flow between the third ventricle and interpeduncular cistern is obstructed. Conceptually, performing an ETV in this context is similar to placing an internal shunt before tumor resection. It relieves symptoms in most patients and thus allows a direct attack on the tumor at a later stage. It shares with the placement of an internalized shunt the possibility, or even the probability, that it will not be needed after the tumor has been removed. It shares with shunts the real possibility of leading to an upward herniation syndrome—the herniation of the superior cerebellar vermis through the tentorium, leading to compression of the midbrain. Typically, contemporary pediatric neurosurgeons begin patients on high doses of dexamethasone at the time of diagnosis to stabilize their condition. When the tumor is attacked directly, an external ventricular drain is placed before the lesion is removed.

The role of ETV in the overall management of children with brain tumors affecting the CSF pathways is evolving. Soon a series of publications will support its more or less routine use. We await the results of those studies before applying this technique generally. If neurosurgeons elect to perform an ETV before removing a posterior fossa brain tumor, they must carefully review the anatomy as it appears on sagittal MRI scans. In this situation, the brain stem is often pushed anteriorly against the clivus. The distance between the clivus and basilar artery is short, which makes the procedure more risky than it would be otherwise. The procedure should be performed by an experienced neuroendoscopist and can be made significantly safer by using frameless stereotaxy.

If tumor resection is not essential to the patient's outcome, the neuroendoscope becomes

a much more exciting tool. Adolescent male patients with a pineal tumor that enhances diffusely, hydrocephalus, and no markers of malignancy (elevated levels of α-fetal protein or human chorionic gonadotrophin) are likely to have a pineal germinoma. Ideally, these patients should be managed by using an endoscope to perform a third ventriculostomy to obtain a biopsy of the tumor. Definitive treatment can then be managed using conformal radiation therapy. This approach is associated with minimal rates of morbidity and high rates of success.

Hydrocephalus associated with intraventricular and subarachnoid hemorrhage

Whether SAH results from diffuse craniocerebral trauma or from the rupture of an aneurysm or arteriovenous malformation (AVM), patients are often left with a significant CSF absorption problem. Treatment is usually improved substantially by the widespread use of external ventricular drains. In these contexts, the most common indication for shunting is failure to wean the patient from the ventriculostomy without high levels of ICP. The ventricles do not dilate. ETV is contraindicated based on the two reasons stated previously. The ventricles are small enough to make performing ETV problematic. The presumed point of obstruction is probably the arachnoidal granulations distal to the interpeduncular cisterns. Therefore, these patients are unlikely to benefit from the procedure. They respond to treatment using either ventricular shunting or lumbar shunting. If it is possible to temporize long enough, they may not need a shunt at all.

If the inflammatory process associated with SAH is allowed to continue long enough, the primary point of obstruction becomes the basal cisterns. CSF flow is then occluded between the SSASs and CSASs. This process also follows bacterial or fungal meningitis. Under the Dandy classification system, these forms of hydrocephalus would be considered "communicating hydrocephalus." Consequently, patients would be considered candidates for ventricular shunting or lumboperitoneal shunting. The obstruction to flow is upstream from the interpeduncular cistern, however, and patients may indeed respond to ETV. In fact, the success of the procedure has been highest in this group.

This point of obstruction causes most cases of NPH [29]. In a small series, three of four patients with NPH had excellent outcomes after treatment [30]. This approach represents an exciting area for future research.

Special case: hydrocephalus in infancy

ETV is much less likely to be successful when used to treat infants than when used to treat hydrocephalus later in life. Consequently, a number of authors have concluded that an age less than 6 or 12 months should be considered a contraindication to the use of ETV in the management of hydrocephalus. A number of reasons may underlie this failure. The first relates to the ability to define the point of obstruction in these babies with severe hydrocephalus. Because of the presence of open fontanels and unfused sutures, the degree of hydrocephalus is greater than when hydrocephalus occurs later in life. Furthermore, it is difficult to interpret the scans to determine the actual point of obstruction. The same patient also may have multiple points of obstruction, especially when hydrocephalus is associated with the Chiari II malformation (spina bifida cystica). In the latter case, there are four different points of obstruction, and more than one site can be obstructed at once [7]. When hydrocephalus occurs later in life, obstruction is less likely to occur at more than one site. It is also easier to determine where the actual point of obstruction is based on imaging studies.

The second reason relates to the actual physics of the system. When the anterior fontanel is opened widely and the sutures are splayed, the intracranial compartment is essentially in communication with and has the same pressure as atmospheric pressure. Natural absorption of CSF depends on ICP being at least 5 mm Hg greater than sagittal sinus pressure [5,31,32]. It is often easier for the size of the head to expand than to maintain ICP greater than 5 mm Hg. In addition, Laplace's law implies that the larger ventricles are, the less distending force is needed to maintain them at that size.

Although ETV is less likely to be successful in these children than in older children, good outcomes are obtained in some babies. The procedure also may be safer to perform in infants than in older children or adults. The floor of the third ventricle is more diaphanous in infants than it is likely to be in older children. The basilar artery can be identified with certainty and thus protected. The ventricular membrane is so fragile that it can usually be opened with normal irrigation.

Theoretically, it is better to have communicating than noncommunicating hydrocephalus. When a patient with hydrocephalus related to aqueductal stenosis experiences a shunt failure, no mechanisms are available to compensate for the increased ICP that accompanies failure of the shunt. If the point of obstruction is distal, a significantly longer time is likely to elapse before the situation becomes critical.

Endoscopic third ventriculostomy for programmed shunt removal

Chronically shunted patients may suffer from severe and incapacitating headaches caused by SVS. The percentage of patients suffering from this problem is controversial, and some investigators doubt its existence. If the threshold for making this diagnosis includes not only the classic triad of severe intermittent headaches lasting 10 to 90 minutes, smaller than normal ventricles on imaging studies, and a slowly refilling flushing mechanism but the need to discontinue activity or be brought home from school at least two times per month, about 15% of chronically shunted older children and adults suffer from this condition [33]. Based on chronic monitoring of ICP, we have identified four different pathophysiologic mechanisms responsible for SVS: intermittent proximal obstruction, intracranial hypertension with smaller than normal ventricles and a failed shunt (normal volume hydrocephalus), intracranial hypertension with a working shunt (cephalocranial disproportion), and migraine in shunted patients [34].

Originally, these patients were managed with shunt revision using higher resistance valves and devices that retarded siphoning. Recently, we have offered these patients the opportunity to assess the possibility of becoming shunt independent [1]. These patients are admitted to the hospital for externalization of their shunt, or their shunt is removed and replaced with an external

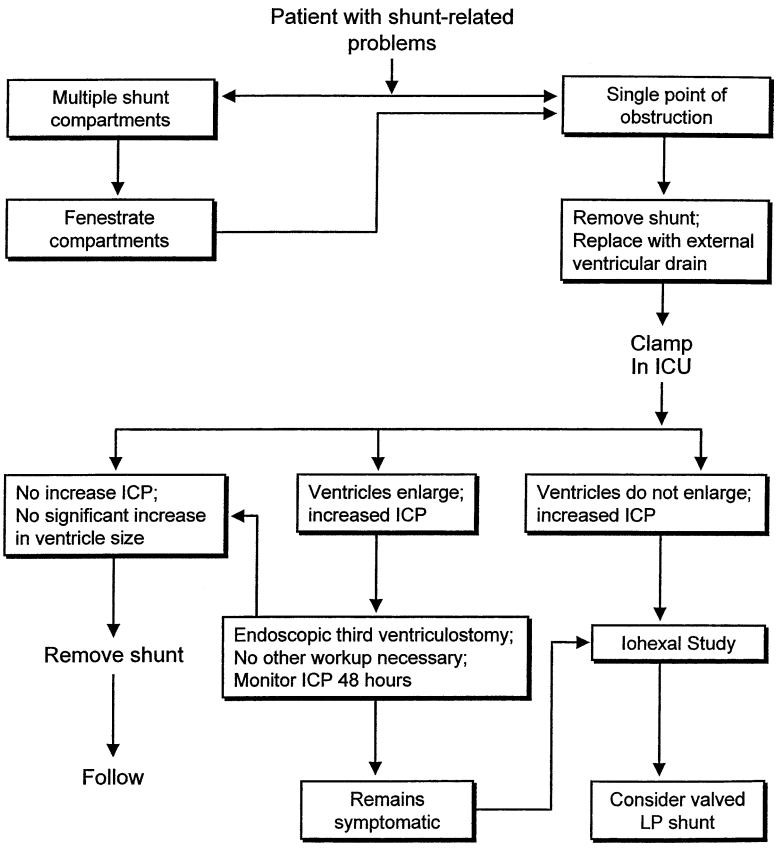

Fig. 5. Algorithm for managing shunt-related difficulties with a shunt removal protocol. (Courtesy of Barrow Neurological Institute.)

Box 1. Contraindications to endoscopic third ventriculostomy

Absolute contraindications

1. Third and lateral ventricles too small to allow safe manipulation of the endoscope within the ventricular system
2. Tumor or other mass lesion obstructing the surgeon's access to the floor of the third ventricle
3. Proven communication between the CSF that is in the third ventricle with CSF within the interpeduncular cistern

Relative contraindications

1. Multicompartment hydrocephalus
2. Thickened floor of third ventricle
3. Chiari II malformation
4. Age less than 1 year
5. Distortion of third ventricular anatomy

CSF = cerebrospinal fluid.

ventricular drain. Under controlled conditions in the intensive care unit, their shunt is occluded or raised to a higher level and a scan is obtained. About two thirds of these patients become symptomatic with marked ventricular dilatation. These patients undergo ETV, and 70% tolerate shunt removal. Patients who develop marked intracranial hypertension without ventricular distention undergo cisternographic assessment. In my practice, 28 of 31 patients have been shown to have communicating hydrocephalus and have been treated with lumboperitoneal shunts employing valve systems. In chronically shunted patients, ventricular distention at the time of shunt failure is probably sufficient to recommend ETV (Fig. 5).

Summary

ETV using contemporary instrumentation has been used for more than 50 years, but its use has become widespread only in the last 10 to 15 years. Randomized prospective trials comparing ETV with shunts are needed before definitive statements can be made about the role of the former in managing the many forms of hydrocephalus. The absolute and relative contraindications for the use of ETV in the management of hydrocephalus are shown in the Box 1 on this page. It is important not to presume that a specific radiographic or clinical feature would prevent a patient from responding to this rather new procedure without testing the hypothesis. Patients should be given as much information as possible regarding the risks and benefits of ETV so they can participate in the decision-making process.

When should the role of ETV in the management of hydrocephalus be discussed with a patient? At the initial diagnosis of hydrocephalus, the patient or family should be informed of this potential alternative to shunting for the management of hydrocephalus. I also believe that patients with working shunts who are being followed chronically should be informed about ETV as a potential treatment option when their shunt fails. Every shunt failure or infection should be viewed as an opportunity to explore the possibility that the patient could become shunt independent.

References

[1] Baskin JJ, Manwaring KH, Rekate HL. Ventricular shunt removal: the ultimate treatment of the slit ventricle syndrome. J Neurosurg 1998;88:478–84.
[2] Dandy W, Blackfan K. An experimental and clinical study of internal hydrocephalus. JAMA 1913;61:2216–7.
[3] Dandy W, Blackfan K. Internal hydrocephalus. An experimental, clinical and pathological study. Am J Dis Child 1914;8:406–82.
[4] Ransohoff J, Epstein F. Proceedings: avoidance of shunt dependency. J Neurol Neurosurg Psychiatry 1975;38:410–1.
[5] Olivero WC, Rekate HL, Chizeck HJ, et al. Relationship between intracranial and sagittal sinus pressure in normal and hydrocephalic dogs. Pediatr Neurosci 1988;14:196–201.

[6] Rekate HL. Circuit diagram of the circulation of cerebrospinal fluid. 1989. Pediatr Neurosurg 1994; 21:248–52.
[7] Rekate H. Neurosurgical management of the newborn with spina bifida. In: Rekate H, editor. Comprehensive management of spina bifida. Boca Raton, FL: CRC Publishers; 1991. p. 1–26.
[8] Gilles F, Davidson R. Communicating hydrocephalus with deficient dysplastic parasagittal arachnoidal granulations. J Neurosurg 1971;35:421–6.
[9] Barlow CF. CSF dynamics in hydrocephalus—with special attention to external hydrocephalus. Brain Dev 1984;6:119–27.
[10] Akaboshi I, Ikeda T, Yoshioka S. Benign external hydrocephalus in a boy with autosomal dominant microcephaly. Clin Genet 1996;49:160–2.
[11] Rekate H, Nulsen FE, Mack H, et al. Establishing the diagnosis of shunt independence. Monogr Neural Sci 1982;8:223–6.
[12] Epstein F, Hochwald GM, Ransohoff J. Neonatal hydrocephalus treated by compressive head wrapping. Lancet 1973;1:634–6.
[13] Karahalios DG, Rekate HL, Khayata MH, et al. Elevated intracranial venous pressure as a universal mechanism in pseudotumor cerebri of varying etiologies. Neurology 1996;46:198–202.
[14] Pare LS, Batzdorf U. Syringomyelia persistence after Chiari decompression as a result of pseudomeningocele formation: Implications for syrinx pathogenesis: report of three cases. Neurosurgery 1998;43:945–8.
[15] Nugent G, Al-Mefty O, Chou S. Communicating hydrocephalus as a cause of aqueductal stenosis. J Neurosurg 1979;51:812–8.
[16] Francis PM, Beals S, Rekate HL, et al. Chronic tonsillar herniation and Crouzon's syndrome. Pediatr Neurosurg 1992;18:202–6.
[17] Saint-Rose C, LaCombe J, Pierre-Kahn T, et al. Intracranial venous sinus hypertension: cause or consequence of hydrocephalus in infants? J Neurosurg 1984;60:727–31.
[18] Pierre-Kahn A, Hirsch JF, Renier D, et al. Hydrocephalus and achondroplasia. A study of 25 observations. Childs Brain 1980;7:205–19.
[19] Cinalli G, Renier D, Sebag G, et al. Chronic tonsillar herniation in Crouzon's and Apert's syndromes: the role of premature synostosis of the lambdoid suture. J Neurosurg 1995;83:575–82.
[20] Nishihara T, Hara T, Suzuki I, et al. Third ventriculostomy for symptomatic syringomyelia using flexible endoscope: case report. Minim Invasive Neurosurg 1996;39:130–2.
[21] Engel M, Carmel P, Chutorian A. Increased intraventricular pressure without ventriculomegaly in children with shunts: "normal volume" hydrocephalus. Neurosurgery 1979;5:549–52.
[22] Rekate HL, Wallace D. Lumboperitoneal shunts in children. Pediatr Neurosurg 2003;38:41–6.
[23] Jones RF, Kwok BC, Stening WA, et al. Neuroendoscopic third ventriculostomy. A practical alternative to extracranial shunts in non-communicating hydrocephalus. Acta Neurochir Suppl (Wien) 1994;61:79–83.
[24] Teo C, Jones R. Management of hydrocephalus by endoscopic third ventriculostomy in patients with myelomeningocele. Pediatr Neurosurg 1996;25: 57–63.
[25] Buxton N, Macarthur D, Mallucci C, et al. Neuroendoscopic third ventriculostomy in patients less than 1 year old. Pediatr Neurosurg 1998;29:73–6.
[26] Oi S, Shimoda M, Shibata M, et al. Pathophysiology of long-standing overt ventriculomegaly in adults. J Neurosurg 2000;92:933–40.
[27] Larsson A, Stephensen H, Wikkelso C. Adult patients with "asymptomatic" and "compensated" hydrocephalus benefit from surgery. Acta Neurol Scand 1999;99:81–90.
[28] Pollack IF, Pang D, Albright AL. The long-term outcome in children with late-onset aqueductal stenosis resulting from benign intrinsic tectal tumors. J Neurosurg 1994;80:681–8.
[29] Di Rocco C, Di Trapani G, Maira G, et al. Anatomo-clinical correlations in normotensive hydrocephalus. Reports on three cases. J Neurol Sci 1977;33:437–52.
[30] Mitchell P, Mathew B. Third ventriculostomy in normal pressure hydrocephalus. Br J Neurosurg 1999;13:382–5.
[31] Ransohoff J, Shulman K, Fishman R. Hydrocephalus: a review of etiology and treatment. J Pediatr 1960;56:399–411.
[32] Cutler R, Page L, Galicich J. Formation and absorption of cerebrospinal fluid in man. Brain 1968;91:707–20.
[33] Hyde-Rowan MD, Rekate HL, Nulsen FE. Re-expansion of previously collapsed ventricles: the slit ventricle syndrome. J Neurosurg 1982;56:536–9.
[34] Rekate HL. Classification of slit-ventricle syndromes using intracranial pressure monitoring. Pediatr Neurosurg 1993;19:15–20.