Minimally Invasive Neurosurgery for Cerebrospinal Fluid Disorders

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KEYWORDS

• Cerebrospinal fluid • Neuroendoscopy • Hydrocephalus
• Endoscopic third ventriculostomy

The general goals for treating disorders of cerebrospinal fluid (CSF) circulation are restoration of the balance between CSF production and clearance, and elimination of pressure gradients between CSF compartments. In most cases, this translates to fluid diversion via membrane fenestration or shunting. The first implantable shunt valve, invented by Nulsen and Spitz[1] in the 1950s, revolutionized the treatment of hydrocephalus, a previously devastating disease. Unfortunately, despite attempts to make shunt dynamics more physiologic, complications related to shunting have continued to plague patients with hydrocephalus. Advances in shunt technology have failed to improve outcomes or circumvent shunt complications.

As technology continues to modify techniques used to treat neurosurgical conditions, there has been a trend toward techniques that achieve minimal invasiveness with access and visualization provided through the smallest opening possible, with greatest action limited to the point of interest, thus minimizing retraction of normal brain tissue. Advances in minimally invasive neurosurgical techniques, including endoscopy and frameless image guidance, have been part of a major driving force toward development of a superior means to achieve CSF diversion. Paralleling this has been innovation and evolution in the training, experience, knowledge sharing, and familiarity with the endoscope by both practicing neurosurgeons and residents. Applications are rapidly growing and this has had a dramatic effect on the treatment of nearly all disorders of CSF circulation.

This article focuses on minimally invasive approaches to address disorders of CSF circulation. The endoscope has played the major role in changing the way many of these disorders are treated. If studied carefully, these new approaches may also help us better understand the nature of some of these complex dynamic physiologic disorders. To date, the most prevalent use of the endoscope in neurosurgery has been in removing obstructions of CSF flow and/or diverting flow, with little disruption of normal brain tissue. As such, neuroendoscopy has gained most attention in the treatment of focally obstructive entities such as aqueductal stenosis, tumoral hydrocephalus, isolated ventricles, arachnoid cysts, multiloculated hydrocephalus, and fourth ventricular outlet obstructions. Investigative work has also been done using some endoscopic techniques in CSF disorders thought not to be obstructive in nature, including normal pressure hydrocephalus, and hydrocephalus secondary to hemorrhage or infection. The endoscope can also play a key role in treating these disorders by other means, including choroid plexus coagulation, endoscopic shunt placement, and endoscopic assistance of open techniques for CSF diversion.

Using minimally invasive techniques, in many cases shunts can be avoided or removed and craniotomies can be circumvented. For many

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patients this may translate to less pain, shorter hospital stays, and most importantly, improved outcome. The emphasis of investigations to come will need to focus on outcomes, and statistical evaluations to prove superiority of endoscopic procedures over traditional shunting operations.

DISORDERS OF CSF CIRCULATION AMENABLE TO MINIMALLY INVASIVE NEUROSURGERY

Disorders of CSF circulation most amenable to endoscopic treatment are those that are caused by an identifiable, focal obstruction. **Table 1** lists some conditions that are often successfully treated using endoscopy. In general, obstructions causing disruption of normal CSF flow result in dilatation of the ventricular system proximal to the obstruction, often resulting in compression of brain structures, raised intracranial pressure (ICP), and associated symptoms. Shunting the dilated fluid space has previously treated many of these conditions. Unfortunately, in addition to typical problems that accompany shunting, including infection, misplaced catheter, and malfunction, shunting an isolated fluid space creates a pressure differential across the membranous obstruction that is not physiologic. Endoscopic techniques, on the other hand, can effectively and more physiologically treat most cases of CSF obstruction by either membrane fenestration or removal of mass lesion, which can result in restoration of normal CSF flow or creation of a bypass into the ventricles or subarachnoid spaces distal to the obstruction.

The classic obstructive condition most commonly treated successfully by endoscopic means is aqueductal stenosis (**Fig. 1**A). Other lesions obstructing CSF flow include: fourth ventricular outlet obstruction (including Chiari Malformation), isolated lateral ventricle due to foramen of Monro stenosis (see **Fig. 1**B), tumors that block CSF flow (see **Fig. 1**C), multiloculated hydrocephalus (see **Fig. 1**D), isolated fourth ventricle due to both aqueductal obstruction and fourth ventricular outlet obstruction (commonly from adhesions) (see **Fig. 1**E), and arachnoid cysts (see **Fig. 1**F). Other conditions that have been investigated as possibly amenable to endoscopic treatment are those classically considered to be forms of communicating hydrocephalus, including hydrocephalus secondary to subarachnoid hemorrhage, normal pressure hydrocephalus, or hydrocephalus secondary to infection. Results with endoscopic treatments of these conditions have been varied and these mixed results are likely to due to an unclear (and possibly heterogeneous) etiology

Table 1
Common disorders of CSF circulation amenable to minimally invasive neurosurgical treatment

Condition	Point of Obstruction	Treatments
Aqueductal stenosis	Cerebral aqueduct	1. Endoscopic third ventriculostomy 2. Aqueductoplasty 3. Removal of mass lesion 4. Endoscopic lamina terminalis fenestration
Isolated lateral ventricle	Foramen of Monro	1. Septum pellucidotomy 2. Foraminoplasty 3. Removal of mass lesion
Isolated fourth ventricle	Cerebral aqueduct and fourth ventricular outlet	1. Aqueductoplasty + lysis of fourth ventricular outlet adhesions 2. Lysis of fourth ventricular adhesions + endoscopic third ventriculostomy 3. Removal of mass lesion
Fourth ventricular outlet obstruction (Chiari)	Fourth ventricular outlet	1. Endoscopic third ventriculostomy 2. Removal of mass lesion
Multiloculated hydrocephalus	Intraventricular adhesions	1. Adhesion lysis
Arachnoid cyst	Multiple locations	1. Cyst fenestration

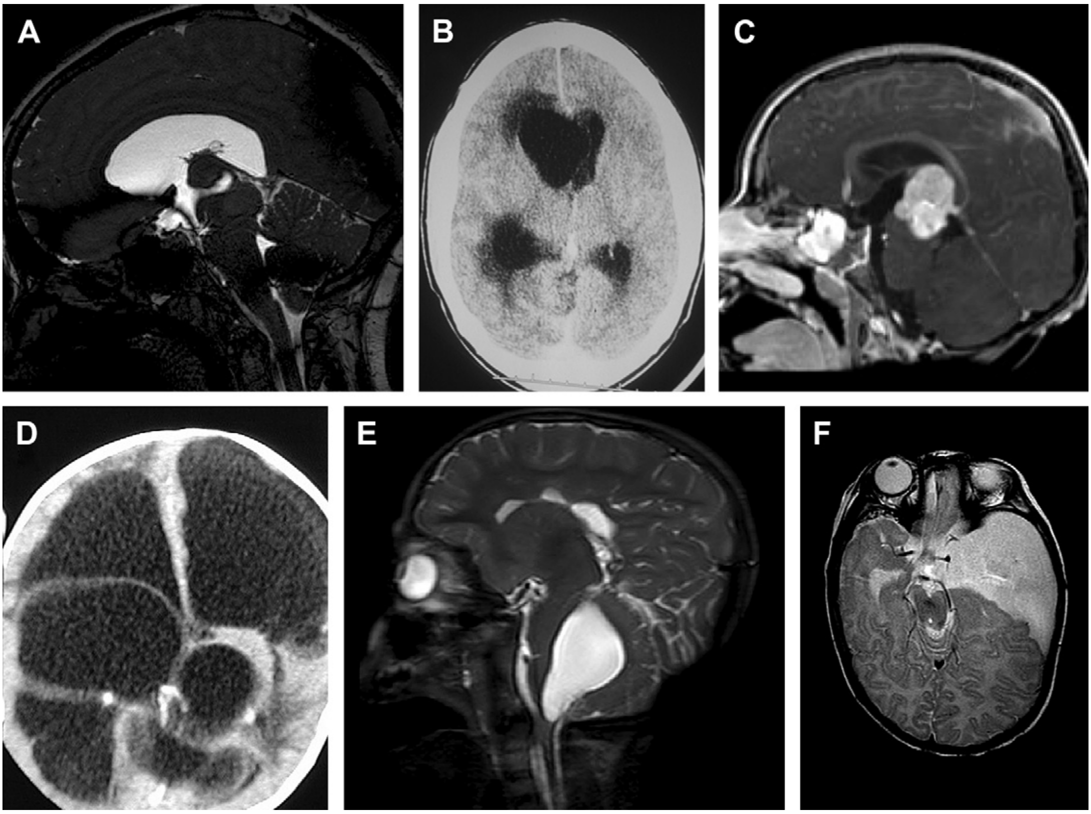

Fig. 1. Disorders of CSF circulation amenable to endoscopic treatment. Disorders most responsive to endoscopic treatment are those that are caused by an identifiable obstruction. Aqueductal stenosis (*A*) leads to dilatation of the third and fourth ventricles with a relatively normal-sized fourth ventricle. Isolated lateral ventricle (*B*), caused by blockage at the level of the foramen of Monro, leads to dilatation of one lateral ventricle with a normal or compressed contralateral lateral ventricle. Tumors involving the pineal region (*C*), posterior third ventricle, or posterior fossa lead to triventricular hydrocephalus, similar to aqueductal stenosis. In multiloculated hydrocephalus (*D*), the presence of multiple intraventricular membranes can lead to expansion of multiple isolated fluid spaces, causing compression of surrounding structures. Isolated fourth ventricle (*E*) can cause dilatation of all 4 ventricles, but will not respond to diversion of only supra- or infratentorial CSF spaces because of lack of communication. Arachnoid cysts (*F*) can directly compress brain structures and may also lead to obstruction of CSF pathways.

behind the mechanism and pathway of CSF flow in these conditions.

APPROACH TO WORKUP OF CSF DISORDER

In general, endoscopic membrane fenestration is most likely to be successful if a clinically significant membranous obstruction can be clearly demonstrated on preoperative imaging. Often, the nature of the obstruction will be obvious on simple preoperative imaging studies such as computed tomography (CT) scan because of significant ventricular dilatation proximal to an obstruction, with normal-sized or compressed ventricles distal. This view has been most commonly demonstrated in the case of aqueductal stenosis, with dilatation of the lateral and third ventricles in conjunction with a relatively normal-sized fourth ventricle and small-convexity subarachnoid spaces (**Fig. 2**A). Other forms of obstructive hydrocephalus easily diagnosed using only CT include isolated lateral ventricle (see **Fig. 1**B) and isolated fourth ventricle (see **Fig. 1**E). Although CT can demonstrate gross dilatation of isolated fluid spaces relative to normal spaces, in some cases it will be necessary to more closely define the nature of the obstruction, either functionally or anatomically. The 3 imaging studies most useful in this regard are: (1) high-resolution magnetic resonance imaging (MRI) using a constructive interference in the steady-state

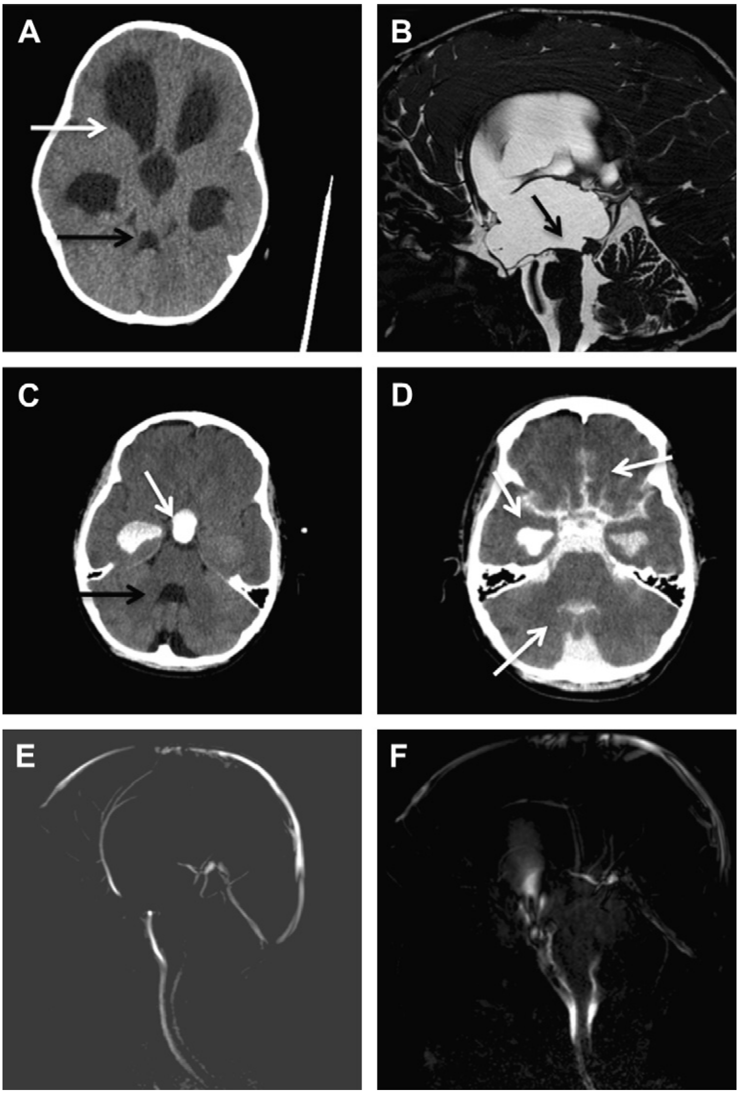

Fig. 2. Imaging workup of CSF disorders. In aqueductal stenosis (*A*), a head computed tomography (CT) scan shows significant lateral and third ventricular dilatation proximal to the obstruction, with a normal-sized fourth ventricle. High-resolution magnetic resonance imaging (MRI) using a constructive interference in the steady-state (CISS) sequence (*B*) produces an image with excellent CSF-to-brain contrast demonstrating the branches of the basilar artery and abnormal aqueductal membrane. Head CT following intraventricular instillation of iodinated contrast, in this patient with obstructive hydrocephalus due to aqueductal stenosis (*C*), demonstrates entrapment of contrast material within lateral and third ventricles with no contrast within fourth ventricle or subarachnoid CSF spaces. Following endoscopic third ventriculostomy (ETV) in the same patient, head CT following intraventricular injection of iodinated contrast shows presence of contrast within all CSF spaces, including the prepontine cistern and subarachnoid space (*D*). Before ETV, cine phase contrast MRI showed absence of flow in the aqueduct (*E*). Following ETV (*F*), the cine sequence shows flow through the third ventricular floor, indicating ventriculostomy patency.

(CISS) sequence, (2) contrast ventriculography, and (3) cine MRI.

MRI scans are performed to localize an obstruction and to provide detailed information needed for surgical planning. In some cases, the planning MRI can be performed at the same time as a scan to be used for frameless stereotactic neuronavigation. Routine T1- and T2-weighted images are obtained in the axial, coronal, and sagittal planes. In addition, a CISS sequence can be performed to identify thin membranes that might interfere with CSF circulation (see **Fig. 2**B). CISS uses fast imaging with steady-state free precession to compensate for slow flow, thereby enhancing the cisternographic effect,[2] producing an image with excellent CSF-to-brain contrast. CISS can be especially useful in imaging the floor of the third ventricle and the contents of the prepontine cistern when contemplating endoscopic third ventriculostomy (ETV), because Liliequist's membrane, the branches of the basilar artery, and abnormal intraventricular or intracisternal membranes can be visualized (see **Fig. 2**B).

In patients harboring an external ventricular drain placed after removal of infected shunt, or to control acute hydrocephalus, there is an opportunity to evaluate for the presence of functional obstructions by contrast ventriculography. Iodinated contrast (approximately 3 mL), can be carefully instilled into the ventricular catheter after removing an identical quantity of CSF; this is followed by instillation of enough CSF or preservative-free normal saline to flush the contrast agent from the ventricular drain into the ventricular system. The drain is then clamped and ICP is transduced. The patient typically undergoes a head CT immediately following instillation of ventricular contrast, 1 hour later, and in some cases several hours later if there is interest in documenting slow CSF transit. This method is an ideal means to evaluate true functional ventricular obstructions. In some ways it is superior to MRI, as membranes may be observed on high-resolution CISS sequences that may still allow flow due to partial autofenestration not easily interpreted on MRI. Contrast ventriculography will demonstrate entrapment of contrast material within the ventricles proximal to the obstruction (see **Fig. 2**C) or presence of contrast within all CSF spaces including prepontine cistern, and subarachnoid space in the setting of communication of CSF spaces (see **Fig. 2**D). In cases of relative stenosis or narrowing of CSF passageways, a delayed scan may demonstrate eventual progression of contrast to distal CSF spaces, which is often seen in the setting of multiple arachnoid adhesions that have developed after either infection or hemorrhage. The obvious disadvantage of this invasive technique over MRI is that it requires access to CSF spaces.

Because most disorders of CSF circulation are caused by an anomaly in the dynamics of CSF flow, measurements of CSF flow are ideal for diagnosis of CSF flow disorders and also for monitoring in follow-up. Two MRI techniques can reliably measure flow: (1) a cine phase contrast (PC) sequence in which the flow is coded on a gray scale, and (2) a cine phase contrast flow (PCF) sequence in which the velocity is coded on a gray scale but also on a quantitative scale.[3] Velocity measurement is possible by means of flow analysis, with the time parameter given by a signal synchronized on the heartbeat. Cine PC and PCF are now routinely used to assess obstructions in the normal CSF pathways, and also to evaluate patency following treatments (see **Fig. 2**E, F). In obstructive hydrocephalus, cine PC MRI can objectively assess the absence of flow in a given structure such as the aqueduct or foramen of Monro (see **Fig. 2**E, F). Following treatment such as ETV, the cine PCF sequence allows measurement of the flow through the third ventricular floor (see **Fig. 2**E, F).

GENERAL APPROACH TO ENDOSCOPIC MANAGEMENT OF CSF DISORDERS

When it has been determined that a condition is amenable to endoscopic treatment, careful preparation can increase the chance of success and decrease complication rate. This aim usually requires careful study of preoperative imaging, thoughtful evaluation of treatment options, and consideration of the need for frameless neuronavigation.

When planning membrane fenestration, the overall objective is to restore normal CSF balance by creating openings in obstructive membranes that will remain patent, while avoiding trauma to functional brain tissue and/or blood vessels. A straight line from the entry point should traverse the membrane(s) that require fenestration. In many cases, a prior burr hole can be used. At other times, use of multiple burr holes is necessary to adequately and safely fenestrate the important obstructive membranes. The fenestrations should be as large as possible, and there should be as many as possible. Frameless navigation, with stereotactic tracking of the endoscopic tip, can be very useful because disorientation is a frequent encumbrance.

The repertoire of endoscopic equipment for management of complex disorders of CSF circulation should include rigid and in some cases flexible endoscopes, with high-resolution video camera systems, irrigation method, and ability for navigational tracking of the endoscope. The endoscope collection often includes a rigid rod-lens endoscope with good optical quality, variations of viewing angles (typically 0° and 30°), and easy handling. Various mechanical instruments of different sizes such as scissors, grabbers, hooks, puncture needles, biopsy and grasping forceps, bipolar and unipolar coagulation instruments, and Fogarty balloon catheters are useful for safely creating fenestrations.

General techniques for endoscopic approaches have been described.[4–8] In general, endoscopic techniques are performed under general anesthesia. The patient's head rests on a horseshoe headrest or 3-pin fixation when frameless navigation is used. The entry point is determined with the goal of approaching the target with a straight approach, to avoid injury to important structures between the entry point and target (such as the fornix). Side-to-side movement of the endoscope should be avoided. Proper entry can sometimes be accomplished by drawing a line on the preoperative MRI, or the frameless navigation station, from the target through a safe channel (such as the foramen of Monro) and extending this to cross the calvarium. The intersection with the calvarial surface and skin is the entry point for burr hole and incision, respectively. In addition to avoiding important neural structures, care is taken to avoid important vascular structures such as major arterial branches and venous sinuses.

The incision can be linear, but in general should be curvilinear if there is a chance a shunt may be required if the endoscopic procedure fails. A burr hole is created that is large enough to accommodate the working sheath of the endoscope (in some cases up to 1 cm in diameter). A funnel-shaped burr hole, with the narrow portion near the outer skull table, allows endoscopic movement while minimizing the outer defect. After opening the dura, a ventricular needle is inserted into the ventricle initially, if using a free-hand technique, to safely find the ventricle. Some describe placing the operating sheath directly into the ventricle after dural opening; however, the author finds it is safer to first cannulate the ventricle using a smaller diameter ventricular needle. If using image guidance, the operating sheath/trocar can be directly inserted into the ventricle using navigation while tracking the tip of the sheath/trocar. In the case of small or slit-like ventricles, or in cases with distorted anatomy, image guidance improves safety. Image guidance is also beneficial for determination of the ideal entry point and navigation toward target membranes.

Once the endoscope is inserted, the anatomy is inspected to confirm correct location using anatomic landmarks and, if using navigation, this is correlated to landmarks on the workstation. The author does not use continuous irrigation. Instead, lactated Ringer solution is used to flush through the irrigation ports of the endoscope using a hand-held syringe technique only as needed for visualization. This method offers the advantage of tactile feedback regarding ICP. If significant backpressure is encountered, this may indicate a problem with CSF egress, which must be addressed to avoid increases in ICP. One can also leave an external ventricular drain in place during the procedure for continuous measurement of ICP.

Several techniques are available for the management of unexpected bleeding. Small hemorrhages usually cease spontaneously after a few minutes of waiting, or with irrigation. If this fails, a bipolar diathermy probe can be used to achieve hemostasis while using irrigation to improve visualization of the bleeding point. Other techniques include tamponade either with the scope itself, or by placing a small cottonoid down the working sheath and holding pressure using a grabbing instrument. In extreme circumstances, air can be injected into the ventricular system to replace CSF, to transform the ventricular space from liquid to air medium; this aids visualization of bleeding. CSF should not be removed using suction because this can lead to ventricular collapse, potential extra-axial hemorrhage, and worsening of visualization. In extreme circumstances, though very rare, the endoscope can be removed and the operation converted to an open procedure for hematoma evacuation.

SPECIFIC ENDOSCOPIC TECHNIQUES

Endoscopic Third Ventriculostomy

ETV has gained nearly worldwide acceptance as a safe and effective treatment for many causes of pediatric and adult hydrocephalus. Despite the increasing use of this technique, no clear guidelines are available and controversy continues over which patients are appropriate candidates for the procedure. This controversy largely exists due to an overabundance of retrospective case series suggesting relative safety and efficacy of endoscopic procedures over more traditional CSF diversion procedures, and a lack of well-designed prospective randomized trials comparing ETV with shunting.

There is a consensus that ETV is generally indicated as a treatment for any type of hydrocephalus that is caused by obstruction proximal to the fourth ventricular outlet.[9–14] A typical example is shown in **Fig. 3**. ETV has also been successful in the management of CSF obstructions distal to the fourth ventricle, such as Dandy-Walker malformation and Chiari malformations.[15–21]

By studying outcomes, indications for ETV have become more precisely defined.[4,22–38] One of the most controversial issues has been the age at which ETV is likely to be effective. Most reports support that improved outcome after ETV is associated with older age.[12,39–41] Different studies have shown improved outcome in older patients with the cut-off in different studies varying from older than 6 months of age,[11] older than 12 months,[42] and older than 2 years.[12] A few studies, on the other hand, suggest that outcome is not dependent on age.[43,44]

The Canadian Pediatric Neurosurgery Study Group conducted a collaborative study to address the important determinants of outcome after ETV, including age, by pooling 368 patients who underwent ETV from 9 Canadian centers.[45] By multivariate analysis, only age had a significant effect on outcome, with younger patients failing at higher rates, particularly neonates and infants, leading to the conclusion in this study that the only statistically significant variable influencing outcome is age.

Despite this result, some believe that ETV may be a reasonable method of initial CSF diversion in children even younger than 6 months.[46] In a study of 14 patients younger than 6 months with obstructive hydrocephalus treated with ETV, success was obtained in 8 patients (57%). It could be reasoned that ETV should be recommended because it gives an opportunity for shunt independence in a significant proportion of patients, potentially avoiding the long-term complications

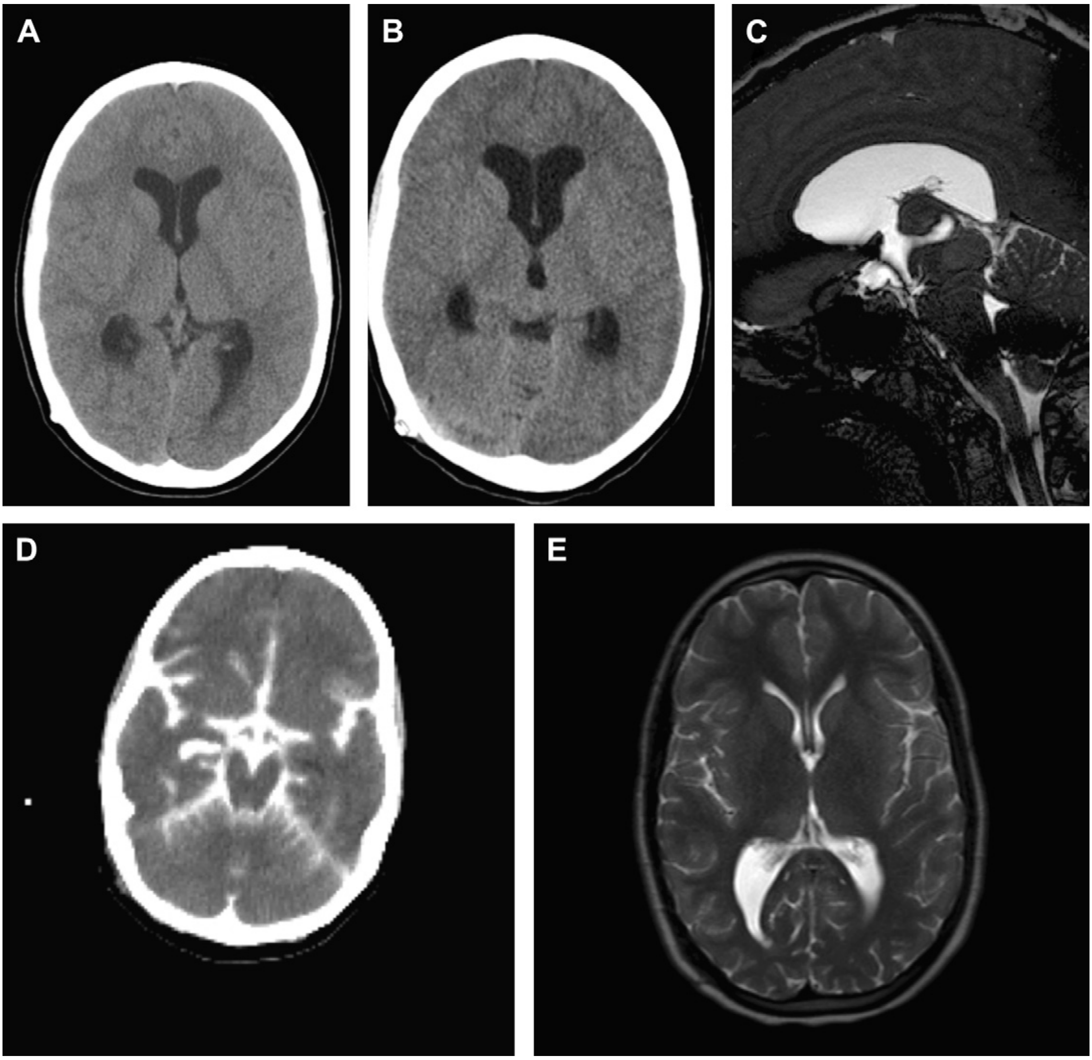

Fig. 3. Case example: ETV with removal of malfunctioning ventriculoperitoneal shunt. This 12-year-old girl was shunted at birth for triventricular hydrocephalus. She presented 1 month after proximal revision with headache, nausea, vomiting, decreased level of consciousness, and bradycardia. Head CT on presentation after removal of 20 mL CSF from shunt reservoir (*B*) showed enlarged lateral and third ventricles compared with baseline scan (*A*). High-resolution sagittal MRI CISS sequence (*C*) showed evidence of obstruction at the level of the aqueduct, and suggested she would be a good candidate for ETV. After ETV, an external ventricular drain was placed and transduced ICPs were normal. Instillation of ventricular iodinated contrast showed communication of all CSF spaces (*D*). MRI scan obtained 3 months later (*E*), when she was feeling "better than baseline," showed ventricles that were smaller than prior well baseline, suggesting partial obstruction or nonideal valve pressure on her original shunt.

associated with a shunt. Conversely, it can be argued that it is equally reasonable to initially shunt these patients, reserving ETV for consideration in the face of a shunt malfunction occurring when older than 2 years.

Outcome is also strongly related to the etiology of hydrocephalus, with success of ETV being ideally suited to those with primary and secondary aqueductal stenosis.[8,22,23,28,31,33,37–39,41,47–77] In the best candidates, the distal CSF pathways are patent and the basal cisterns are free from adhesions that occur after bleeding or infection. MRI sequences, as described earlier, can delineate the nature of the obstruction. Downward bulging of the third ventricular floor into the prepontine cistern, dilatation of the suprapineal recess into the quadrigeminal cistern, flattening of the mesencephalon, funneling of the upper part of the aqueduct, and normal or even small size of the fourth ventricle are all frequently associated with aqueductal stenosis, and are rarely found with communicating hydrocephalus.[78–80]

In addition to obstructive types of hydrocephalus, ETV has been investigated in the study of communicating types of hydrocephalus, such as normal pressure hydrocephalus, with mixed results.[26,81] Some studies document clinical improvement in up to 70% of patients.[26,29,81,82] Although the mechanism of improvement cannot be explained by the bulk CSF flow theory, the mechanism may become clear in the future as CSF hydrodynamics become better understood.

Relative contraindications to ETV include structural anomalies, such as a very large massa intermedia, a thickened third ventricular floor, or the presence of extensive scar tissue involving the third ventricular floor, prepontine cistern, and basilar artery apex. Other anatomic variations, such as a very narrow third ventricle, an effaced interpeduncular cistern, or slit ventricles, are not in themselves contraindications, but require experience and sometimes frameless navigation for safety and success.[36]

Variations on this technique have been extensively described.[8,22,26,30,31,59,64,71,80,83–86] The patient lies supine with head in slight flexion (approximately 15°) with the head positioned on a horseshoe headrest. If stereotactic guidance is required, as in the case of extremely small ventricles or abnormal anatomy, the head is fixed in a 3-pin headrest. Some frameless navigational systems, which use electromagnetic guidance, can avoid placement of pins, and this can be extremely beneficial in the case of small infants. The monitor should be placed in front of the surgeon to allow the surgeon to operate in a neutral position. A frontal curvilinear incision is preferred over a straight incision to prepare for the possibility of ETV failure and need for shunt placement. The burr hole is typically made approximately 3 cm lateral and 1 cm anterior to the coronal suture, which should always be identified before drilling. These coordinates are based on retrospectively obtained imaging data from 27 patients undergoing ETV.[87] A ventricular needle is first inserted into the right lateral ventricle. Following its removal, the endoscope can be navigated along the path directly into the ventricular system. One should avoid attempting to find the ventricle using the endoscope, even in the case of markedly dilated ventricles, as devastating vascular and neurologic complications can occur. If frameless stereotactic navigation is used, the tip of the endoscope/trocar can be tracked as it enters the ventricular system. Once in the right lateral ventricle, the endoscope is navigated through the foramen of Monro into the third ventricle. The floor of the third ventricle with landmarks including mammillary bodies and infundibular recess is identified. The location of the fenestration depends on the patient's specific anatomy. The placement should avoid damage to the basilar artery and its branches, as well as the infundibulum. The typical location is made in the midline, halfway to two-thirds the distance from the infundibular recess to the mammillary bodies.

The floor can be perforated using one of several described techniques including use of the endoscope itself,[14,88] use of a semirigid stylet,[89] bipolar or monopolar coagulation,[90,91] suction cutting devices, and a spreading instrument.[47,92] There are advantages and disadvantages to each method. Perforation using the scope itself is thought to decrease the risk of vascular injury; however, the force required to penetrate the thickened floor can potentially cause hypothalamic injury. Use of a rigid instrument, such as closed biopsy forceps or Decq forceps,[93] which can be opened under direct vision, is less traumatic and allows direct vision during perforation; however, there is a risk of vascular injury if the forceps are opened after penetration (with avulsion of perforators) or if they are closed before withdrawal. Typically, the opening is enlarged by inflating a balloon of a 3F Fogarty catheter to achieve an adequate fenestration size of 3 to 6 mm. The interpeduncular and prepontine cisterns are inspected through the ventriculostomy and, if Liliequist's membrane is present, it is fenestrated. After withdrawing the endoscope, the brain tract is filled with Gelfoam (Upjohn, Kalamazoo, MI) to prevent egress and potential accumulation of subdural fluid.

The outcome after ETV is clearly related to patient selection.[12,23,31,51,59,63,67,73,94–106] In patients with obstruction at the level of the cerebral aqueduct, success rates are generally high and have been reported at up to 90%.[9,51,63] Outcomes are generally poorer in those with communicating types including postinfectious or post-hemorrhagic hydrocephalus. In a retrospective analysis of the operative success and long-term reliability of ETV in 203 patients with follow-up for up to 22.6 years,[12] the overall probability of success (failure defined as shunt insertion, ETV revision, or death) was 89%. Compared with the Canadian study, this study supported an association between the surgical success and the individual operating surgeon (odds ratios for success, 0.44–1.47 relative to the mean of 1.0, $P = .08$). As mentioned, the only statistically significant factor associated with long-term reliability was age, with the statistical model predicting the following reliability at 1 year after insertion: at 0 to 1 month of age, 31% (14%–53%); at 1 to 6 months of age, 50% (32%–68%); at 6 to 24 months of age, 71% (55%–85%); and at more than 24 months of age, 84% (79%–89%). In this study, there was no support for an association between reliability and the diagnostic group (n = 181, *P* = .168) or a previous shunt. In the Canadian Pediatric Neurosurgery Study Group study,[45] the overall 1- and 5- year success rates for ETV were 65% and 52%, respectively. These slightly lower outcomes results can be explained by use of a different measure of success and failure: in this study, success was strictly defined by no further CSF diversion procedures, whereas others have used placement of a shunt as the definition of failure. In a study of 131 patients undergoing ETV,[107] at 1 year follow-up, 82.5% of 86 primary ETV and 80% of 45 secondary ETV were shunt free.

In the case of ETV for treatment of obstructions distal to the fourth ventricle, results have been mixed, which may be due to a variety of factors causing distal obstruction. Fourth ventricular outlet obstruction is most commonly associated with prior intraventricular hemorrhage or infection. In a retrospective analysis of patients with fourth ventricular outlet obstruction, ETV was successfully performed in 20 of 22 patients with a follow-up period 1 to 8 years (Mean 4.2 years).[19] There was a high failure rate in those younger than 6 months.

ETV may also be a safe and effective means of treating hydrocephalus in the older (greater than age 2 years) spina bifida population and offers the hope of long-term shunt independence for selected patients.[14] In one study, ETV was performed on 69 patients with hydrocephalus and myelomeningocele. Most of the patients had been previously shunted, although in 14 patients ETV was the initial treatment. Patient selection was based on preoperative imaging studies suggesting noncommunicating hydrocephalus. There were no serious complications and the overall success rate was 72%, although selecting only patients who had been previously shunted or who were older than 6 months at the time of endoscopy increased success to 80%. ETV success in the myelomeningocele population has clinical importance, as approximately 90% of those with myelomeningocele develop hydrocephalus requiring diversion.[108]

The reasons for failure after ETV are still poorly understood and likely multifactorial. In some cases, there is presumably a communicating component to the hydrocephalus, with clearance issues that cannot be addressed by bypassing an interventricular obstruction. These patients require shunt placement. Ventriculostomy closure has been identified for failure in some cases, offering the possibility for a second ETV procedure over shunting. This question was addressed in a retrospective analysis of 482 ETVs in pediatric patients from 2 Italian centers.[109] In the 40 patients undergoing a second operation after initial ETV failure, the stoma was found to be closed in 28 patients without underlying adhesion, to be open but with significant prepontine arachnoid adhesion in 8 patients, to be open without adhesions in 2 patients, to have a pinhole orifice in 1 patient, and to be closed with underlying adhesions in 1 patient. A second procedure allowed reopening of the stoma or lysis of adhesions in 35 of 40 patients, and this was effective 75% of the time. Of importance, age younger than 2 years at the time of the first procedure and arachnoid adhesions in the subarachnoid cisterns observed during the second procedure were the main negative prognostic factors for the success of a second ETV. This finding may support the strategy to shunt children younger than 2, and reserve ETV in the case of shunt failure after the age of 2 years. In the article by Kadrian and colleagues[12] that retrospectively analyzed 203 patients, there was no support for an association between reliability and the diagnostic group (n = 181, *P* = .168) or a previous shunt. Sixteen patients had ETV repeated but only 9 were repeated after at least 6 months. Of these, 4 procedures failed within a few weeks, and 2 patients were available for long-term follow-up. In the future, stent placement may decrease the incidence of stoma closure.

In developing countries where a dependence on shunts is considered dangerous, ETV combined with choroid plexus coagulation (CPC) may be the best option for treating hydrocephalus in infants, particularly for those with non-postinfectious hydrocephalus and myelomeningocele. Warf[104,105,110,111] prospectively studied bilateral CPC in combination with ETV in 266 patients, and ETV alone in 284 patients. Of importance, 81% were younger than 1 year. The hydrocephalus was postinfectious in 320 patients, non-postinfectious in 152, and associated with myelomeningocele in 73. Overall, the success rate of ETV-CPC (66%) was superior to that of ETV alone (47%) among infants younger than 1 year (*P*<.0001). The ETV-CPC combined procedure was superior in patients with a myelomeningocele (76% compared with 35% success, *P* = .0045) and those with non-postinfectious hydrocephalus (70% compared with 38% success, *P* = .0025). For patients at least 1 year old, there was no difference between the 2 procedures (80% success for each, *P* = 1.0000).

Aqueductoplasty

In the treatment of aqueductal stenosis, anatomic variations can often render ETV a dangerous or impossible option. Cerebral aqueductoplasty has gained popularity as an effective treatment for membranous and short-segment stenosis of the cerebral aqueduct.[33,53,56,112–115] This procedure can be performed via a precoronal approach (burr hole placed 8 cm from nasion and 3 cm lateral to midline), passing through the lateral ventricle, foramen of Monro, and third ventricle into the aqueduct, or via a suboccipital foramen magnum trans-fourth ventricle approach. The floor of the third ventricle and proximal aqueduct can be inspected with a 0°, 30°, or 70° endoscope. If the stenosis is located distally in the aqueduct and this cannot be visualized using a rigid scope, a steerable fiberscope can be used to inspect the aqueduct. Aqueductoplasty can be performed with the aid of a 2F or 3F Fogarty balloon catheter, which is gently passed into the stenosis and dilated. The tip of the catheter should be bent to enable passage through the distal aqueduct and fourth ventricle because the aqueduct is not straight, but curved (see **Fig. 2**B), and a straight catheter may damage the tectum.[115]

A foramen magnum trans-fourth ventricular approach may offer advantages compared with the more traditional anterior approach. The fourth ventricular approach does not traverse brain tissue, does not depend on ventricular dilatation, and is a straight approach with no pressure on structures of the foramen of Monro.[114]

The decision to treat aqueductal stenosis by ETV rather than aqueductoplasty depends on the anatomy of the individual patient and the experience and comfort of the endoscopist.

To address the benefit of an intra-aqueductal stent to ensure continued patency of the aqueduct following this procedure, Cinalli and colleagues[116] conducted a retrospective evaluation of the effectiveness of endoscopic aqueductoplasty performed alone or accompanied by placement of a stent in the treatment of isolated fourth ventricle in 7 patients with supratentorial shunts and loculated hydrocephalus. These investigators found placement of a stent to be more effective than aqueductoplasty alone in preventing the repeated occlusion of the aqueduct. Aqueductal stenting is also a good procedure for treatment of isolated fourth ventricle.[18,19,56,113] Stenting is indicated when an increased risk of restenosis is expected. The stent should be at least 6 cm or longer to prevent migration. An alternative fixation option is suturing the stent to the dura at the entry point.

Lamina Terminalis Fenestration

If abnormal anatomy makes ETV and/or aqueductoplasty too risky, lamina terminalis fenestration can be an option for the treatment of any obstruction proximal to the fourth ventricular outlet. This procedure is performed in the setting of a translucent lamina terminalis to enable identification of the anterior cerebral arteries. The burr hole is usually placed in the same location as an ETV. If the foramen of Monro is large, the lamina terminalis can be visualized with a rigid 0° scope. If the foramen is narrow, a flexible scope must be used to avoid fornix damage. The lamina terminalis is perforated in the same manner as ETV and the perforation enlarged as well by inflating a Fogarty balloon catheter. Care is taken to preserve the chiasm and anterior cerebral arteries.[33,117]

Septum Pellucidotomy

Fenestration of the septum pellucidum is indicated for treatment of a blocked foramen of Monro, leading to dilatation of an ipsilateral lateral ventricle (**Fig. 4**).[33,117,118] This procedure is successful only if the contralateral foramen of Monro is patent and there is no further distal obstruction. It is also indicated in the event of bilateral obstruction of both foramina of Monro (see **Fig. 4**B and C), in conjunction either with communication of one lateral ventricle with the third ventricle (foraminoplasty), or with placement of single ventriculoperitoneal shunt; this avoids the need to place either 2 shunts, or a "Y" connection of 2 ventricular catheters.

The entry point is typically 5 to 6 cm paramedian, and 1 cm anterior to the coronal suture. Some surgeons prefer to enter on the side of the smaller lateral ventricle, to avoid injury to the contralateral head of caudate nucleus following fenestration. Others advocate entering on the side of the dilated ventricle,[33,117,118] allowing easier initial access to the ventricular system. When using a free-hand technique, entering on the side of the dilated ventricle is easier. However, if the contralateral ventricle is compressed or slit-like, there is a possibility of injury to the contralateral head of the caudate nucleus when creating an opening in the septum. This possibility is minimized if the smaller ventricle is entered initially, as the fenestration is made into a large fluid space. The disadvantage to placing a burr hole on the side of the smaller ventricle is that with a free-hand technique, it is possible that the contralateral ventricle will inadvertently be entered. This possibility can be avoided by using navigation. For

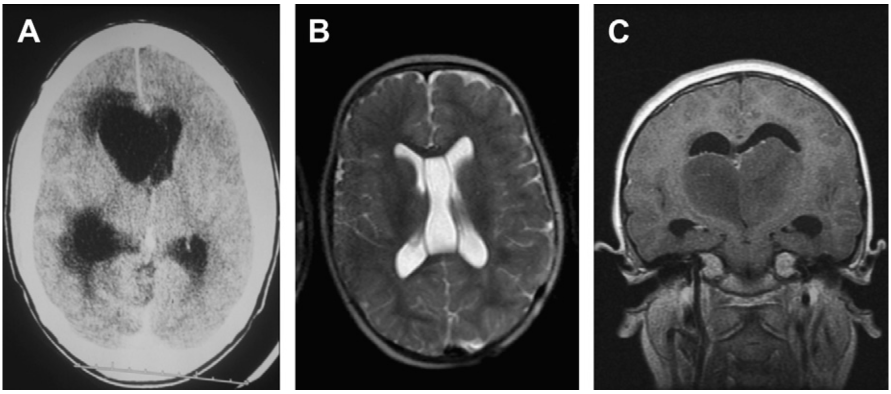

Fig. 4. Indications for septum pellucidotomy. Septum pellucidotomy is indicated in the setting of obstruction at the level of the foramen of Monro. Examples include unilateral foramen of Monro stenosis (*A*) treated with septum pellucidotomy alone, enlarged cavum septum pellucidum leading to bilateral foramen of Monro stenosis (*B*) treated with fenestration of both cavum membranes, and bithalamic tumor with obstruction of both foramina of Monro (*C*), which can be treated with septum pellucidotomy, and placement of unilateral shunt.

septum pellucidotomy, navigation is useful for determining the optimal entry point as well as the optimal point of septum fenestration.

The point of fenestration often depends on the individual anatomy. In some cases, there is a thin and avascular portion of the septum suitable for fenestration. Fenestration can be performed using monopolar coagulation with a hook instrument, or using blunt perforation.[118] The perforation can be enlarged with the aid of a Fogarty balloon catheter and scissors, or with monopolar, or bipolar diathermy. The fenestration size should be at least 1 cm in diameter; this is especially true in cases of thick septum, so as to avoid risk of ostomy closure.

Foraminoplasty

An alternative technique for treatment of isolated lateral ventricle is foraminoplasty.[119] This technique aims to restore the normal drainage pathway from the lateral ventricle into the third ventricle. The entry point is similar to that for ETV. The foramen of Monro is found by following the choroid plexus. The ependyma is coagulated and a spreading instrument inserted into the third ventricle anterior to the plexus, avoiding the fornix. Navigation improves safety. Dilatation with a Fogarty balloon is helpful.

Lysis of Intraventricular Adhesions

Neuroendoscopic management is an effective treatment for complex multi- and uni-loculated hydrocephalus, which has more traditionally been treated with multiple shunts, multiperforated catheters, stereotactic aspiration, or shunt placement and craniotomy with lysis of intraventricular septations.[12,21,117,120,121] Multiple shunts are often unsuccessful due to increased risk of infection, obstruction, and hemorrhage associated with their removal.

Endoscopic treatment is extremely valuable in treating loculated ventricles and for communication of isolated CSF compartments. The goal is communication of all CSF spaces. Membrane lysis/fenestration is often combined with other traditional fenestration techniques including foramen of Monro reconstruction, septum pellucidotomy, septal wall removal, cyst wall fenestration, ETV, stenting between lateral and third ventricle, fourth ventriculostomy, and endoscopic shunt placement.

In an attempt to simplify cases of complex hydrocephalus using minimally invasive endoscopic techniques, Kadrian and Teo and colleagues[121] studied 114 patients treated endoscopically who presented with either more than one shunt and/or multiloculated hydrocephalus (47 patients), isolated lateral ventricle (25 patients), isolated fourth ventricle (20 patients), arachnoid cyst (15 patients), slit ventricle syndrome (4 patients) or cysticercosis (3 patients). The endoscopic procedures performed included cyst or membrane fenestration, septum pellucidotomy, ETV, aqueductal plasty with or without stent, endoscopic shunt placement, and retrieval and removal of cysticercotic cysts. Reduction to one shunt was possible in 72%, shunt independence in 28%, and only 11% required shunt revisions long-term.

There are numerous benefits to this approach. Multiloculated hydrocephalus is most commonly secondary to infection. Establishing CSF flow (a nidus for infection) and removal of shunts may be beneficial. With just one shunt, there are fewer opportunities for obstruction, malfunction, disconnection, and infection. Moreover, in patients with multiloculated hydrocephalus, it is easier to identify the source of shunt malfunction as compared with multiple shunts.

Practically speaking, the following principles should direct surgical approach: (1) the burr hole should be made in a position to make the cortical passage as short as possible, the endoscope will enter the largest cavity, and the trajectory will take the endoscope to the membrane that separates the 2 cavities that need to be joined; (2) if there is a pre-existing ventricular catheter that does not drain an isolated portion of the ventricle, then its tract can be used to enter the ventricle; (3) the trajectory should try to communicate as many cavities as possible; (4) as many fenestrations as possible should be made using a sharp technique.

Arachnoid Cysts

Optimal treatment of arachnoid cysts remains controversial. Options include no treatment, shunting, or fenestration via craniotomy with resection of cyst wall, stereotactic cyst fenestration, or endoscopic fenestration (**Fig. 5**). When cyst wall fenestration into the basal cisterns is contemplated, endoscopy is valuable due to its better visualization, magnification, angled perspective, and ability to maintain cyst distention during the procedure. Endoscopic management of arachnoid cysts has been extensively reported.[12,122–136] Greenfield and Souweidane[128] analyzed a prospectively generated database of 33 patients who underwent endoscopic fenestration of arachnoid cysts. Fenestration was successful as judged by cyst decompression and symptom resolution in 97%, with only one initial failure that was successful after repeated endoscopic fenestration. Suprasellar cysts, which represent less than 10% of all intracranial arachnoid cysts, can be successfully treated endoscopically with good clinical outcome and low surgical morbidity. Ventriculo-cysto-cisternostomy should be attempted, but when the communication between the cyst and the cistern is considered too dangerous, ventriculo-cystostomy is acceptable.

Tumor-Related CSF Obstruction

Neuroendoscopy plays several roles in the surgical management of brain tumors and their occasional interference with CSF circulation.[33,102,117,137–146] Often, the CSF obstruction can be fully treated via endoscopic removal of the tumor causing the obstruction. Colloid cysts, benign lesions that arise from the velum interpositum or choroid plexus of the third ventricle, can produce hydrocephalus by obstruction of the foramina of Monro. Colloid cysts represent the ideal tumor for purely endoscopic excision (small with minimal vascularity).

Obstructive hydrocephalus is common with presentation of posterior fossa tumors. The incidence of hydrocephalus requiring CSF diversion following tumor resection remains at between 10% and 62%.[16,51,147–153] Due to the lack of prospective clinical trials, there is no consensus on proper perioperative management of this condition.[153] Pre-resectional hydrocephalus can be managed with an initial CSF diversional procedure such as ventricular drain or CSF shunt, or by resection alone. Postoperative shunt placement has historically been the management strategy for definitive treatment of communicating

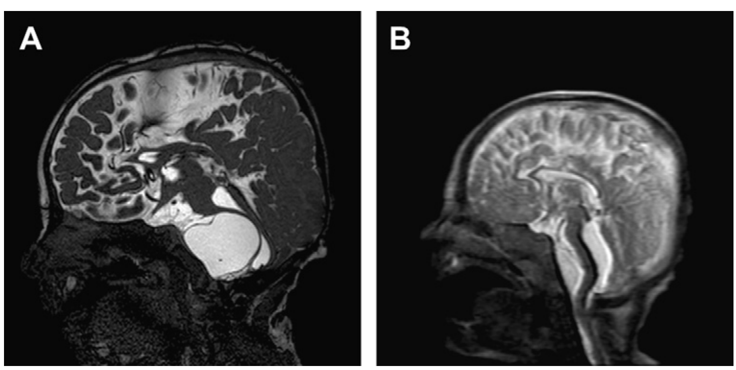

Fig. 5. Case example: large prepontine arachnoid cyst. A 26-week preterm female infant with grade 4 intraventricular hemorrhage developed post-hemorrhagic hydrocephalus and underwent shunt placement at age 4 weeks. She presented at age 7 months with pneumonia, respiratory distress, apnea, and poor feeding. MRI showed a large cyst anterior to the brainstem with severe thinning of her pons and medulla (*A*). She underwent endoscopic membrane fenestration through existing shunt tract, with improvement of symptoms within 24 hours. MRI scan 48 hours later (*B*) showed significant improvement of brainstem compression.

hydrocephalus. ETV has emerged as a compelling treatment strategy for CSF diversion both before and after resection. Tamburrini and colleagues[154] found that postoperative ETV placement in the setting of high ICP following removal of posterior fossa tumor was successful in 90% (27/30) of the patients. In a retrospective analysis of 59 children presenting with posterior fossa tumors and no (n = 16), mild,[49] moderate,[95] or severe[155] hydrocephalus, 37 (63%) underwent pre-resectional ETV with a high success rate for up to 7.5 years, with only 5 of 37 being failures.[156] Although no direct comparisons have been made, evidence appears to support ETV as a method for treatment of hydrocephalus associated with posterior fossa tumors, even after resection.

Intracranial Frameless Navigation

The combination of neuronavigation with neuroendoscopy improves safety and accuracy with such things as tumor biopsy, tumor resection, ETV, cyst wall fenestration, and ability to access small or slit ventricles. The usefulness of navigation in intracranial endoscopy has been extensively studied.[6,79,115,128,131,135,155,157–161] Navigation can be used to plan the correct entry point and trajectory, minimizing the potential for damage to the foramen of Monro and other brain tissue by reducing back-and-forth and side-to-side motion of the endoscope. The technique can be performed using a simple rigid endoscope through predetermined entry points and trajectories. Navigational tracking is most helpful in entering small ventricles, in approaching the posterior third ventricle when the foramen of Monro is narrow, and in selecting the best approach to colloid cysts. It is essential in some cystic lesions lacking clear landmarks, such as intraparenchymal cysts or multiloculated hydrocephalus.

COMPLICATIONS

Although neuroendoscopy has revolutionized the treatment of CSF disorders, specific complications occur due to risks inherent in the procedure and, sometimes, inexperience. Attempts should be made to avoid these problems. These complications have been extensively reviewed by experienced endoscopists, with suggestions made for their avoidance.[4,8,12,14,33,40,51,55,63,73,85,98,100,107,162–178]

Complications specific to ETV are best understood, and include bradycardia and asystole during manipulation of the third ventricular floor,[179] damage to the fornices with scope manipulation or poorly placed burr holes, hypothalamic pituitary axis damage,[173,176] damage to structures adjacent to the floor of the third ventricle including cranial nerves,[173] injury to major vessels resulting in subarachnoid hemorrhage[170] or ischemic stroke,[180] subdural hygroma or hematoma,[143,181] herniation,[51,181] remote intracranial hemorrhage,[42,169] infections,[42,173] and severe cognitive[182] and psychiatric sequelae.[163] Deaths associated with ETV have been reported acutely due to hemorrhage[4,42,45,54,68,165,173,183–186] and delayed from infection,[173] and even late, after sudden closure of the third ventriculostomy many months or years after a successful procedure. In a retrospective review of 131 patients undergoing ETV with a minimum follow-up duration of 1 year, serious complications after ETV occurred more frequently in patients who presented at the time of shunt malfunction (14 of 45 patients, 31%) compared with patients who underwent primary ETV (7 of 86 patients, 8%), (P = .02).[107] In a retrospective study of 203 patients for up to 22.6 years, infections were observed in 4.9%, transient major complications in 7.2%, and major and permanent complications in 1.1%.[12] Complications specifically related to aqueductoplasty include transient vertical diplopia or upgaze weakness.[114]

Complications in endoscopic surgery are not negligible even in experienced hands. Cinalli and colleagues[165] recently analyzed the complications recorded in a prospectively collected database of pediatric patients undergoing neuroendoscopic procedures. Complications occurred in 32 of 231 (13.8%) procedures performed for the management of obstructive hydrocephalus (137 patients), multiloculated hydrocephalus (53 patients), arachnoid cysts (29 patients), and intraventricular tumors (12 patients). Among complications, subdural hygroma occurred in 11 cases, CSF infection in 11, CSF leak in 9, intraventricular hemorrhages in 2, technical failures in 7, subcutaneous CSF collection in 1, thalamic contusion in 1, and transient akinetic mutism in 1 patient who died 6 months following the procedure. Three patients developed permanent disability as a consequence of surgical complication (1.3%).

Many of these complications can be avoided with experience and careful technique. As a general rule, the endoscope should not be used to find the ventricle. Even though the ventricle may be grossly dilated, inaccurate placement of an endoscope, unlike a fine brain needle, is not well tolerated. Side-to-side movements of the scope can tear bridging veins or stretch neural structures, and should be minimized. One should always check to see that there is an adequate outflow mechanism to allow for egress of the irrigation fluid. One should make sure that the edges of the rigid scope are blunt and rounded. Alternatively, when using a flexible scope one should check to ensure that the scope is in the neutral position before removal. One should use a peel-away sheath if the procedure requires multiple passages of the endoscope. Fornix damage can be avoided with optimal burr hole placement, and if the contralateral ventricle is entered, it should not be used for ETV. If the foramen of Monro is small, it should be enlarged before the procedure. Hypothalamic damage can be avoided by penetrating the third ventricular floor away from infundibular recess in the midline. Cranial neuropathies can be avoided by staying in the midline and recognizing and identifying the relevant anatomy before making any definitive maneuvers. Subdural hygromas can be avoided by expanding ventricle before removal of scope, plugging the cortical hole with Gelfoam, and avoiding ventricular drainage. CSF should be drained from cysts and ventricles slowly, and once the procedure has been accomplished, the cavities should be refilled to minimize the risk of subdural collections.

SUMMARY AND FUTURE DIRECTIONS

When faced with the patient with hydrocephalus, the ideal goal is to restore normal CSF hydrodynamics to a near physiologic state, with a safe procedure that is likely to not fail or require revision. To meet this goal, minimally invasive techniques, particularly neuroendoscopic procedures, have emerged as a therapy that is superior to shunting in appropriately selected patients. The potential applications for endoscopic techniques are rapidly growing, and this has had a dramatic effect on the treatment of nearly all disorders of CSF circulation. The main role of the endoscope in neurosurgery continues to reside in removing obstructions of CSF flow and/or diverting flow, with little disruption of normal brain tissue. Using these minimally invasive techniques, shunts can often be avoided or removed. For the patient this may mean less pain, shorter hospital stays, and improved outcome. Future investigations in this area should focus on better elaborating postoperative patient outcomes, providing a more thorough understanding about patient selection and factors that predict treatment failure, and undertaking formal, prospective evaluations of the comparative efficacy of endoscopic procedures compared with traditional shunting and open craniotomy operations.

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