Endoscopic third ventriculostomy in adult patients

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Endoscopic third ventriculostomy (ETV) has gained popularity and has become the treatment of choice for certain pediatric and adult hydrocephalic conditions. The authors report their experience with 36 adult patients and evaluate the long-term outcome and safety of ETV. They discuss several improvements to the surgical techniques that they have developed based on their experience, including the use of intraoperative Doppler imaging before fenestration to trace the location of vessels underlying the floor of the third ventricle. They also report the use of a Rickham reservoir and endoventricular stent in selected cases and discuss the indications for their use. In cases of obstructive hydrocephalus due to congenital or acquired aqueductal stenosis in adults, the success rate of ETV in avoidance of shunt placement is 72%. Twenty-two percent of the patients in this series in whom ETV was initially successful later experienced closure of the fenestration and recurrent symptoms at a mean interval of 3.75 years. Thus, in patients who undergo this treatment, long-term periodic follow-up review should be performed.

KEY WORDS • endoscopic third ventriculostomy • adult chronic hydrocephalus • microvascular Doppler ultrasonography • stent

OVERVIEW

Prior to the modern age of shunts, third ventriculostomy through an open craniotomy was the main treatment modality for hydrocephalus. As detailed in Hellwig, et al.,²¹ Mixer first reported performing third ventriculostomy in 92 patients with hydrocephalus. Early high mortality and morbidity rates and the subsequent development of shunts decreased the use of third ventriculostomy. As the limitations of shunts for chronic hydrocephalus became more obvious, third ventriculostomy resurfaced as a plausible treatment for this condition. In 1973, Guiot²⁰ described 20 patients with hydrocephalus in whom he had performed camera-guided third ventriculostomy, with a 75% success rate in control of the hydrocephalus. Today, ETV is a common practice in tertiary care neurosurgical facilities, and the list of indications for this procedure has grown.

Many factors have contributed to the attractiveness and popularity of ETV. Foremost is the fact that this procedure, when successful, eliminates reliance on mechanical shunts with all their limitations, and returns CSF dynamics to essentially normal status in patients with obstructive hydrocephalus. In addition, the simplicity and elegance of ETV and the measurable good outcome and high success rate among the appropriate patient population contribute to its wide popularity.

Abbreviations used in this paper: AS = aqueductal stenosis; BA = basilar artery; CSF = cerebrospinal fluid; ETV = endoscopic third ventriculostomy; ICP = intracranial pressure.

Initially, ETV was used as an aqueductal bypass procedure for the treatment of obstructive hydrocephalus. Early indications for ETV included hydrocephalus due to congenital and acquired AS secondary to brainstem and tectal tumors. Although some believe that congenital AS should not be treated with ETV because of the insufficiently developed subarachnoid space,^{21,41} others have successfully treated congenital hydrocephalus with this procedure.¹⁸

Today, many patients with hydrocephalus may be considered candidates for ETV. Currently, the surgical indications for the procedure include all of the following: primary AS,^{6,10,11,17,19,23,29} isolated lateral ventricular hydrocephalus in combination with endoscopic septal fenestration,^{2,26,27,34,37} idiopathic stenosis of the foramina of Magendie and Luschka,^{31,50} Dandy–Walker malformation,^{9,36} and hydrocephalus due to intraventricular hematoma in combination with aspiration of the hematoma.¹⁴ In addition, ETV may be performed for hydrocephalus caused by mass effect from intraventricular tumors,^{6,15,16,54} pineal and tectal tumors,^{7,39} posterior fossa tumors,^{42,44} suprasellar arachnoid cysts,⁴² pineal cysts,^{16,51} or quadrigeminal cistern cysts.²⁵ This procedure has also been reported to yield some limited success in the treatment of postinfectious/posthemorrhagic hydrocephalus,⁴⁹ slit ventricle syndrome,^{5,40} and communicating and normal-pressure hydrocephalus.^{32,35}

With the dramatic rise in indications for ETV, we are faced with new neurosurgical complications and even more numerous technical difficulties. In this review, we present our experience with a series of adult patients and further

discuss the surgical technique, complications, and possible solutions to some of these problems.

Surgical Technique

The standard surgical technique for ETV has been well described^{4,22,28,30,33,38,43,52,53} and will not be detailed step by step in this paper. Several refinements of surgical technique that we believe are valuable and lead to either improved outcome or improved safety have been introduced over time. We will focus our discussion on these refinements.

Preoperative Evaluation

It is important to note that this technique is appropriate only for patients who have sufficiently large ventricles in which to introduce the endoscope to navigate into the third ventricle. Generally, the width of the third ventricle should be approximately 5 mm or greater. If a patient has slit ventricles caused by overshunting of CSF, it will first be necessary to externalize the shunt to control drainage until an adequate working diameter of the ventricles is achieved. Although stereotactic guidance can be used as a surgical adjunct, it is typically not necessary and was not used for any of our adult cases.

We find it beneficial to perform ETV with patients placed in a semisitting position with the head flexed further forward so that the entry burr hole is positioned superiorly. This helps to minimize loss of CSF and air entrapment into the ventricles or the subdural space, which can be a cause of protracted postoperative headache, nausea, and vomiting. Loss of CSF may also be a risk factor for postoperative subdural hematoma. Closing the burr hole incision under a constant stream of saline infusion also helps to minimize any air entrapment.

Trajectory and Access to the Ventricular System

The optimal working trajectory into the third ventricle is usually achieved with an entry burr hole placed at or just anterior to the coronal suture (2.5–3 cm lateral to the midline). In most cases, this will provide a direct trajectory from the entry site through the foramen of Monro into the third ventricle. Once entry into the ventricle has been obtained with a conventional ventriculostomy catheter, we prefer to replace the catheter with a peel-away sheath (Neuroview Introducer Sheath; Neuronavigational Corp., Costa Mesa, CA) with a diameter just slightly larger than the endoscope to provide a working porthole into the anatomy. This enables easy insertion of the endoscope into the ventricle without repeated trauma to cerebral cortex and epidural vessels. It also allows for convenient egress of saline irrigation, eliminating any risk of overpressurizing the ventricle. If any significant hemorrhage occurs in the ventricle as a result of the procedure, the peel-away sheath also facilitates copious irrigation until the hemorrhage is cleared.

Identifying the BA Complex

We have previously described a technique of using a microvascular Doppler probe inserted through the endoscope to locate precisely by insonation the position of the BA complex below the floor of the third ventricle.⁴⁵ Injury to the BA complex or its perforating vessels has been reported in multiple cases and can result in catastrophic and

often fatal hemorrhage or brainstem infarction. Thus, precise knowledge of the location of this artery is essential before performing fenestration of the ventricle, and the fenestration should always be made anterior to the BA complex. In the ideal situation, this artery complex lies at the posterior edge of the third ventricle floor at the level of the mammillary bodies.

In our series of 36 adult patients, however, we found that variations in the relationship of the BA complex to the third ventricular floor occur frequently, and anomalies were noted in 14 (39%) of our patients. These anomalies included anterior translocation of the BA complex, so that it lies underneath the midportion of the third ventricular floor, and ectatic anterior and upward bowing of an enlarged BA trunk, in which the vessels lie at risk underneath the midportion of the third ventricular floor. If the floor of the ventricle is translucent, these vascular anomalies can be readily detected and a safe entry site determined. In our series of patients, however, 19 patients (53%) had thickened and opaque third ventricular floors, which rendered the underlying vascular anatomy optically invisible. The combination of an aberrantly located BA and an opaque third ventricular floor occurred in 19% of our patients, placing them at increased risk for vascular injury.

We have found that microvascular Doppler ultrasonography can quickly and easily trace the exact position and layout of the underlying vessels when they are not optically visible, and this should help to minimize the risk of vascular injury. No cases of vascular injury occurred in our series. Learning the technique of microvascular Doppler examination of the floor of the third ventricle is facilitated in cases in which the vascular anatomy is visible (47% of the patients in our series) and thus increases operator confidence in cases in which the ventricular floor is opaque (53% of the patients in our series).

Performing Ventriculostomy

Because of the risk of vascular injury, the third ventricular floor should always be penetrated bluntly and never with laser energy or electrocautery. We prefer to use a 1-mm blunt glass fiber to make the penetration; this is accomplished using a slight back-and-forth twisting motion. The initial fenestration is then dilated up to approximately a 4-mm opening by using a No. 2 French Fogarty catheter. In some cases, an imperforate membrane of Liliequist can be identified lying beneath the floor of the third ventricle, which would obstruct CSF outflow into the prepontine cistern. If such a membrane is present, it can be opened under direct endoscopic visualization with the glass fiber or balloon catheter.

General Considerations and Closure

We prefer to perform the endoscopic procedure with a steady flow of saline irrigation through the endoscope, using a gravity feed as opposed to pressure feed to avoid any barotrauma to the brain or ventricles. If any hemorrhaging is encountered during the procedure, copious warm saline irrigation should be used until all bleeding is visibly stopped and the ventricular CSF is clear. Once the endoscope and peel-away sheath are removed, we keep the ventricle inflated with a steady infusion of saline through a ventricular catheter while the burr hole incision is being closed.

Endoscopic third ventriculostomy in adult patients

In recent years, it has become our custom in the majority of patients to leave a Rickham reservoir in place at the time of closure. As will be discussed later, this greatly facilitates management and diagnosis if patients are suspected to have persistent elevation of ICP because of deficient resorption of CSF or if the ventriculostomy site becomes closed over. In patients who are acutely ill at presentation with symptomatic elevated ICP, postoperative monitoring of the ICP and drainage of CSF can be accomplished through a needle ventriculostomy inserted percutaneously into the reservoir.

In patients in whom a Rickham reservoir has been left in place, we instill a combination of 10 mg preservative-free vancomycin and 4 mg preservative-free gentamicin into the ventricle at the time of closure. In a recent study, we have found that this regimen decreases the incidence of shunt infection 10-fold, to a rate of 0.5% (BT Ragel, et al., personal communication, 2005).

In two of our patients we found it expeditious to place a ventricular catheter stent through the third ventricular fenestration to maintain its patency (Fig. 1). In one of these patients, the stent was desirable because of early closing over of the ventricular fenestration caused by an ependymal cyst within the anterior half of the ventricle. In the second patient, the third ventricular floor was so stretched and patulous that, after fenestration, the redundant tissue tended to fold over on itself, causing concern for potential obstruction of the opening.

In both cases, stent placement was accomplished by inserting a ventricular catheter of appropriate length through the opening under endoscopic observation so that the distal catheter fenestrations lay within the prepontine cistern. A second set of fenestrations was created in the shaft of the ventricular catheter at the level of the lateral ventricle, and the catheter was secured in position with a Rickham reservoir. Long-term patency of the ventriculostomy has been achieved in both patients.

We prefer to keep our patients overnight in the neurosurgical intensive care unit for observation, and most patients can be discharged home the following day if they do not require postoperative ventricular drainage or ICP monitoring. If patients are doing well clinically, we defer postoperative imaging with computerized tomography or magnetic resonance imaging studies for 6 to 8 weeks to allow for stabilization of the ventricle size. After 6 to 8 weeks, an image is obtained for use as the baseline for subsequent follow-up evaluations. Even in those with clinically successful outcomes, and especially in adult patients with chronic compensated hydrocephalus, the ventricle size may decrease only minimally compared with its preoperative size.^{8,47,48}

Outcome and Complications

The incidence of complications with ETV has been reported ranging from 0 to 20%,^{17,22,29,46} and the mortality rate has been reported as high as 1%.^{46,54} The mortality rate appears to be directly related to the learning curve of the surgeon.⁴⁶ The most devastating intraoperative complication is injury to the BA and its branches lying underneath the floor of the third ventricle.^{1,24} Common technical complications include bleeding at the level of the choroid plexus, ventricular wall, ventriculostomy, or interpeduncular cistern. Intracranial or subdural bleeding and infection are less common complications of ETV.¹⁷

Fig. 1. Lateral x-ray film showing an intraventricular stent passing through the lateral ventricle, the foramen of Monro, and the floor of the third ventricle into the prepontine cistern. The proximal end is secured to a Rickham reservoir.

Data on our case series of 36 adult patients who underwent ETV for hydrocephalus are presented in Table 1. Our overall success rate in avoiding the need for a ventricular shunt was 61%. In only one patient were we unable to perform ventricular fenestration because of unfavorable anatomy at the floor of the third ventricle. Ventricular fenestration was performed in all other patients, but 37% of them later required implantation of a ventricular shunt.

In our case series, 56% of the patients had congenital AS, and of this subgroup, 70% presented with adult onset of symptoms (chronic compensated hydrocephalus) and had not received a shunt previously. Seventy-two percent of these patients without shunts had a successful outcome after an ETV and did not require shunt placement. Of those patients who had a shunt in place, 57% became shunt free after ETV. Forty-three percent of patients presented with acquired obstructive hydrocephalus and 58% of these had AS secondary to a tectal plate glioma. Other causes included higher-grade tumors of the posterior fossa or pineal region, ventricular cysts, and one case of cysticercosis. Among this group of patients with acquired obstructive hydrocephalus, 58% overall were able to avoid shunt placement after ETV.

No surgically related deaths or major morbidity occurred in our case series. Complications included transient short-term memory difficulty in one patient. There was one case of bacterial ventriculitis, and one patient suffered an intraventricular hemorrhage from a tumor biopsy procedure that was performed simultaneously with ventriculostomy. In one patient CSF leakage developed through the burr hole incision site because of persistently high ICP; she required treatment with a ventricular shunt.

TABLE 1
Results in a series of 36 adult patients
with hydrocephalus who underwent ETV

Factor	No. of Patients (%)
diagnosis	
congenital AS	20 (56)
acquired AS	12 (33)
posterior fossa tumor	3 (8)
cysticercosis	1 (3)
ventricular floor	
translucent	17 (47)
opaque	19 (53)
vascular anatomy	
normal	22 (61)
abnormal	14 (39)
opaque & abnormal	8 (22)
outcome	
shunt free	22 (61)
shunt dependent	14 (39)
complications	
memory problems	1 (3)
unsuccessful ETV	1 (3)
infection	1 (3)
minimal hemorrhage	1 (3)
CSF leak	1 (3)
overall	5 (14)

The literature on ETV describes various treatment-related complications that we did not find in our case series. Such complications have included endocrinopathy caused by hypothalamic or pituitary stalk injury, which typically manifests as diabetes insipidus.^{12,18} Third and sixth cranial nerve palsies have been encountered and may be caused by direct nerve injury, barotrauma from overpressurization of the ventricles with irrigation fluid, or injury to one or more perforating arteries at the BA apex. Direct vascular injury to the BA apex or the proximal posterior cerebral artery has been described as a cause of either fatal hemorrhage or a traumatic aneurysm, but fortunately this complication is rare.^{1,24} Injury to the fornix can be caused by direct trauma from the endoscope while navigating through the foramen of Monro, and it typically leads to postoperative cognitive problems. Injury can also occur to the caudate, the thalamus, or the thalamostriate venous complex, especially if the endoscope is blindly overinserted into the ventricle.¹⁸ Cardiac arrhythmias or respiratory arrest have been encountered and typically result from manipulation or irritation of the hypothalamus.^{3,13} Subdural hemorrhage is a risk, especially in patients with very large ventricles who experience excessive CSF loss during the procedure.^{17,18} All of these potential complications should be taken into consideration when performing an ETV. The technical recommendations described in the preceding section have been developed to minimize the chance of these various complications occurring and appear to be effective; that is, we have avoided these problems.

Repeated Closure and Operation

Over the long term, three of our patients died of progression of malignant brain tumors. One patient with a tectal plate glioma died of recurrent hydrocephalus at a remote hospital 3 years after a successful third ventriculostomy. Au-

topsy demonstrated closing over of the ventricular fenestration. Including this one, five of our 22 patients with initially successful ETVs experienced reclosure of their ventriculostomy. In one patient, as described previously, early obstruction by a third ventricular cyst developed, and the lesion was successfully treated with insertion of a stent. Four other patients presented again between 3 and 5 years after the initial operation, with a mean follow-up duration of 3.75 years. Except for the patient who died at another facility, these patients all underwent successful reoperation at our facility and remain shunt free.

In the patient with the longest follow-up duration (6 years), no reclosure has occurred. Because the overall incidence of ventriculostomy reclosure is relatively frequent (22%), we recommend that all patients with initially successful ETVs be followed on a regular basis. Moreover, these patients, their families, and their physicians should be clearly warned about the possibility of recurrent hydrocephalus. We do not yet have enough experience with endoventricular stents to know if this is a potential long-term solution to the problem of ventriculostomy reclosure.

Determining whether reclosure of the fenestration and recurrent hydrocephalus has developed can be clinically difficult, especially in the adult population presenting with chronic compensated hydrocephalus. In these patients, a return of symptoms may be fairly subtle and slow in onset. Because the ventricle size may change only minimally in these patients, this is not always a reliable indicator. If obstructive hydrocephalus has recurred, a diagnostic lumbar puncture may not reflect pressure within the ventricles and could also be potentially unsafe. Cine-flow magnetic resonance imaging may be helpful to demonstrate CSF flow through the ventricular fenestration; however, technical difficulties with such studies may yield false-negative results. If the return of clinical symptoms is compelling and diagnostic studies yield equivocal results regarding the status of the ventriculostomy, endoscopic reexploration is recommended.

We have performed such endoscopic reexploration in two of our 22 patients and demonstrated a patent ventricular opening. Implanting a Rickham reservoir at the time of initial surgery greatly simplifies the problem of determining the status of a patient's hydrocephalus and the question of reclosure of the ventriculostomy. Assessment is done by percutaneous needle tap or, if necessary, by contrast ventriculogram. Moreover, the implanted reservoir can provide an avenue for emergency treatment if a patient presents with acute decompensation.

Success Rate

The overall shunt-free success rate of ETV has been reported to be between 50 and 90%. Beems and Grotenhuis⁶ reported a complication rate of 7.7% and a success rate of 76% in a large series of 339 pediatric patients. In another series of adult and pediatric patients, Hopf, et al.,²³ reported that their success rate with ETV was 76% and concluded that ETV is most successful in occlusive hydrocephalus. Gangemi, et al.,¹⁷ presented follow-up results in 125 adult and pediatric patients who underwent ETV for obstructive hydrocephalus. They reported overall shunt independence rates of 86.4%. Most of the reported series involve either pediatric patients or a mixed population. Our series in-

cludes adult patients exclusively and provides long-term follow-up results. Our success rate, which is defined by shunt independence, was 61%, and this is comparable to previously reported outcomes. The overall complication rate was 14%; none of these complications led to any permanent sequelae. As has been reported in other studies, we had a high success rate in patients with chronic compensated hydrocephalus and tectal plate glioma, suggesting that ETV should be the treatment of choice in these patients.

CONCLUSIONS

Our case series confirms that ETV is an effective method of treatment for adult patients with hydrocephalus, especially in those with chronic compensated hydrocephalus and brainstem tumors. To avoid some of the reported complications, we recommend regular use of a peel-away sheath and microvascular Doppler ultrasonography studies to avoid injury to the cerebral cortex, the ependymal vessels, and the BA complex. Although long-term follow-up results are very encouraging, patients with successful ETV remain at risk of reclosure of the fenestration, which can lead to a fatal outcome if not promptly recognized and treated. Therefore, we recommend that all patients with initially successful ETV receive follow-up care on a regular basis. Moreover, these patients, their families, and their physicians should be clearly warned about the possibility of recurrent hydrocephalus. Evaluation of patency of the ETV remains difficult. We have had encouraging experience with placement of a Rickham reservoir, but long-term follow-up review is needed to study its long-term efficacy. Although we have had successful results with stent placement, our experience is very limited and a firm conclusion is premature.

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