Endoscopic third ventriculostomy for hydrocephalus: A review of indications, outcomes, and complications

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Abstract

Endoscopic third ventriculostomy (ETV) has been in vogue for the past two decades, as a tool in the armamentarium of the neurosurgeon, for the management of hydrocephalus. Its utility has been proven consistently in congenital / acquired aqueductal stenosis, although the outcomes in communicating hydrocephalus as well as hydrocephalus secondary to other etiologies have not been as impressive. It is a relatively safe procedure with the appropriate selection of patients with a low rate of permanent morbidity. This review aims to define the current indications, management outcomes, and complications of ETV.

Key words: *Endoscopy, hydrocephalus, outcome, shunt*

Introduction

An endoscope was first used in neurosurgery in the early 1930s when a ureteroscope was used to coagulate the choroid plexus of the lateral ventricle. Since Dandy's classification of hydrocephalus into communicating and non-communicating types and further refinements of the same, bypass procedures to overcome obstruction of the cerebrospinal fluid (CSF) pathways have been established. Ventriculoperitoneal and ventriculoatrial shunts have played a major role in the successful management of patients with hydrocephalus. Insertion of a shunt is associated with a life-long risk of infection, as it is a foreign body, and despite technological advancements, there is a risk of shunt obstruction as well as overdrainage that can result in significant morbidity, needing frequent shunt revisions.[1-4] The reported rate of shunt malfunction in the first year of placement is 30%, and thereafter it is about 10% per year. The cumulative risk of infection is about 20% per patient, with most centers reporting rates between 5 to 10%.[1,3] As an alternative to a shunt, endoscopic third ventriculostomy (ETV) has been in routine neurosurgical practice for the past two decades, with long-term outcomes being reported in the recent literature. To perform an ETV, the third ventricular floor (and the Lillequist membrane) is fenestrated between the mamillary bodies and infundibular recess in the midline, to communicate the ventricular system with the basal cisterns.[1-3]

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Indications

All patients with obstruction between the third ventricle and the cortical subarachnoid spaces are potential candidates for ETV, while obstruction at the level of the arachnoid villi or the venous flow in the superior sagittal sinus is an absolute contraindication.[4]

Aqueductal stenosis

Endoscopic third ventriculostomy has been most consistently reported to have good outcomes in patients with aqueductal stenosis. This may be congenital or acquired, as in the case of tumors involving the posterior third ventricle and tectal gliomas.[1,5-10] It has been reported that preoperative observation of a closed aqueduct is a significant predictor of the success of ETV.[6] The reported success rates for ETV in aqueductal stenosis range from 70% to more than 90% in the long term.[5,8,11-18] Even in infants, those with aqueductal stenosis have higher success rates than those with other etiologies for hydrocephalus.[2,19] The success rates for ETV in infants with aqueductal stenosis are, however, less than those reported in older children and adults.[2,7,19]

Endoscopic third ventriculostomy has been combined with biopsy of the posterior third ventricular and thalamic lesions, thus achieving the goals of *cerebrospinal fluid* (CSF) diversion, as well as obtaining definitive diagnosis in cases of tumor-related aqueductal stenosis. The opinion is divided on whether ventriculostomy should be performed prior to or after the biopsy of the tumor.[14,20]

Posterior fossa tumors and cysts with hydrocephalus

Endoscopic third ventriculostomy has been attempted with good success rates ranging up to > 90% in patients with posterior fossa tumors and brain stem gliomas with hydrocephalus.[21-24] ETV has been of proven value in patients with posterior fossa tumors, who have persistent hydrocephalus after tumor resection.[22] No technical difficulties were encountered in patients with brain stem tumors in one series.[24] However, routine performance of ETV prior to posterior fossa tumor surgery or in postoperative hydrocephalus may not be necessary.[23] Dandy–Walker complex malformations, retrocerebellar cysts, mega cisterna magna, and hydrocephalus secondary to fourth ventricular outlet obstruction have all been managed successfully with ETV, with or without aqueductal stenting, as well as choroid plexus coagulation.[13,25-27]

Postinfectious hydrocephalus

Warf *et al.*[28,29] have reported the outcomes in a large number of pediatric patients with postinfectious hydrocephalus. The success rates in these patients are lower than in patients with hydrocephalus, due to aqueductal stenosis of non-infectious etiology. In their experience, the success rate in children older than one year of age was 81% in patients with postinfectious hydrocephalus and 91% in non-postinfectious hydrocephalus. The success of ETV in infants with postinfectious hydrocephalus was considerably less at 59%. In their analysis, the presence of obstruction at the level of the aqueduct was associated with higher success rates in infants. The five-year survival post ETV or the shunts performed in infants with postinfectious hydrocephalus was around 70% with about one-third of the survivors having significant disability.[29] The low level of success post ETV in this subgroup of patients is likely to be related to adhesions and scarring in the basal cisterns, secondary to the infection.[30] These patients are also likely to have multiple loculations within the ventricular system, rendering them difficult to treat without shunts.

Tuberculous meningitis with hydrocephalus

Tuberculous meningitis (TBM) is characterized by the presence of exudates in the basal cisterns and the site of obstruction could be at the level of the aqueduct, fourth ventricular outlet or at the basal cisterns.[31-37] Traditionally, placement of shunts has been the treatment of choice in TBM-related hydrocephalus, but high rates of malfunction necessitating revisions have been reported.[34]

There is a role for ETV in patients who are in the chronic phase of the disease, where hydrocephalus is believed to be due to an aqueductal block on account of a small tuberculoma or ependymitis or fourth ventricular outlet block due to basal exudates, which prevents CSF flow over the cortical surface.[32-34] In the acute phase of the disease, the presence of inflammation in the ependymal lining, presence of tubercles within the ventricle, as well as the subarachnoid spaces, and the basal exudates render performing ETV technically difficult and decrease the chance of success. During ETV, the basal exudates can be washed out.[35,36] With adequate medical therapy, the tubercles tend to heal and the basal exudates disappear to a large extent. ETV may be attempted in this phase of the disease rather than in the acute phase.[35-37] Even in the chronic phase, the third ventricular floor is opaque and thickened, making ETV a technical challenge. The overall success rate for ETV in patients with TBM associated hydrocephalus, as reported in large series, is around 70%.[33,34,36] Higher success rates of up to 87% have been reported in well-nourished patients with a thin-to-transparent third ventricular floor, without any exudates in the cisterns.[33,37] Following ETV, patients in better clinical grades at presentation had better outcomes.[34,36,37] In this issue of the Journal, Yadav *et al*,[37] have concluded that ETV failures in TBM-related hydrocephalus could be managed with serial lumbar punctures and lumboperitoneal shunts. In their series, 58% of the patients improved with ETV alone, while 80% of the patients improved with ETV and lumboperitoneal shunt.

Hydrocephalus associated with myelomeningocoele and Chiari malformations

Hydrocephalus associated with myelomeningocele may be of the obstructive or communicating type. As these patients require treatment in infancy, ETV does not provide as good results as in older patients, with failure rates of up to 50% being reported in patients with myelomeningocoele.[2,27,30] However, in a recent series, Warf *et al*,[38] have demonstrated good outcomes in up to 78% of infants with myelomeningocoele associated hydrocephalus treated with ETV and choroid plexus cauterization. There are reports on the successful management of hydrocephalus associated with occipital encephalocoele with ETV, although there is a chance of delayed failure necessitating shunt placement.[39]

Endoscopic third ventriculostomy remains a viable option in the management of hydrocephalus in patients with Chiari I malformation and syringomyelia, with shunt independence being achieved in up to 94% of the patients and resolution of the syrinx being observed in a significant number of patients.[40,41] In children with Chiari II malformation, the cause of hydrocephalus is unlikely to have an obstructive etiology and ETV has been proposed to play a very limited role in this indication.[4]

Hydrocephalus secondary to intracerebral / intraventricular hemorrhage

Post hemorrhagic hydrocephalus of prematurity has been reported to have poor outcome with ETV and choroid plexus cauterization, with only 40% of the patients needing no further procedures. Scarring of the prepontine cistern was correlated with poor outcome.[18,30,42] It has been suggested that in patients with neonatal intraventricular hemorrhage, the obstruction is likely to be in the basal cisterns and the arachnoid villi, in addition to the aqueduct, rendering them poor candidates for an ETV.[4]

Fukuhara *et al.*[43] have reported that ETV may not be the optimal treatment option in the long term for patients with hydrocephalus, secondary to aneurysmal subarachnoid hemorrhage (SAH), but may be employed as a temporary intervention during the waiting period for the clearance of aneurysmal SAH. Oertel *et al.*[44] have reported that ETV may be an alternative to the placement of an external ventricular drain in patients with hydrocephalus secondary to intraparenchymal / intraventricular bleeds. During ETV, intraventricular clots can be evacuated. However, the outcome in these patients may be related to the primary etiology of the hemorrhage and its natural course. Performing ETV with a blurred field of vision in the presence of intraventricular blood and distorted ventricular anatomy can be technically challenging. In this issue of the Journal, Chen *et al*., from Taiwan, have reported on the endoscopic removal of intraventricular blood in patients with spontaneous intraventricular hemorrhage. They did not perform ETV due to the poor visibility of the third ventricular floor caused by the blood in the CSF. Data on patients in whom this procedure was not performed, such as, in historical controls, has not been provided and no definite conclusion on its benefit can be drawn from their experience.[45]

Shunt dysfunction

Endoscopic third ventriculostomy is an effective option in patients with hydrocephalus who present with shunt dysfunction due to underdrainage or overdrainage. The overall rate of shunt failure in patients shunted during infancy is reported to be 48, 52, and 63% at one, two, and five years, with 20% of the patients having more than three revisions in five years.[3] ETV in patients with shunt dysfunction was successful in about 80% of the patients.[3,46,47] Patients with aqueductal stenosis had the best outcome, while postinfectious and myelomeningocele patients had a poor outcome. Ligation of the existing shunt system after ETV has been recommended, as an intermittent or remaining flow of CSF through a malfunctioning shunt could result in decreased flow through the ventriculostomy, predisposing to its closure.[46] Other series have reported that up to 70% of the patients become shunt-free after ETV for shunt dysfunction.[48,49] Woodworth *et al*.[50] have reported that after ETV for shunt dysfunction, only 25% remained symptom-free at the two-year follow-up and have suggested ETV combined with concurrent CSF shunting in patients with multiple shunt revisions and complications. In their experience, there was a 2.5 times higher risk of ETV failure when it was performed in patients with prior shunts than when it was performed in shunt naïve patients. A higher rate of procedure-related complications has been reported when ETV is performed in patients with prior shunt placement.[51]

Communicating hydrocephalus

Endoscopic third ventriculostomy has been performed on patients with idiopathic normal pressure hydrocephalus, with reported success rates of 65 to 72%. Patients with gait disturbance as the predominant symptom, age < 65 years, and minimal dementia have shown the highest rates of improvement. This is comparable with the outcome following shunt placement.[52,53] In a series with pediatric patients, communicating hydrocephalus secondary to meningitis or hemorrhage have been reported to have lower rates of success with ETV, as opposed to children with obstructive hydrocephalus.[30]

Endoscopic third ventriculostomy in patients less than one year of age

Infants presenting with hydrocephalus are not appropriate candidates for ETV as they have poorly developed absorptive surfaces in the subarachnoid spaces and have an open anterior fontanelle, with a soft skull.[4] Nevertheless, ETV has been attempted by several authors in infants with hydrocephalus, due to varying etiologies. Ogiwara *et al*.,[54] reported an overall success rate of 34.8% in infants less than six months of age. Even as success rates have been satisfactory in isolated aqueductal stenosis, there is a high rate of failure with other etiologies as well as in preterm infants.[2,4,19,30,55-60] Sukianov *et al*.,[61] have reported favorable outcomes of up to 75% in children who manifest symptoms of hydrocephalus at more than one month of age and in those with a thinned-out third ventricular floor. Combining choroid plexus cauterization with ETV in infants presenting with hydrocephalus has been reported to improve the success rates from 47% (with ETV alone) to 66%.[62] On analyzing the mechanisms of failure of ETV in infants, an immature CSF absorption capacity in infants, reclosure of the stoma, as well as formation of new arachnoid membranes in the basal cisterns, in the presence of patent stoma, were identified.[4,63]

Complications of Endoscopic Third Ventriculostomy

In a review of complications following ETV in 2884 patients, permanent morbidity was 2.38% and permanent neurological complications (hemiparesis, gaze palsy, memory disorders, and altered sensorium) occurred in 1.44%. The overall complication rate was 8.5%. The other complications related to ETV included intraoperative hemorrhage from the ependymal veins, choroid plexus or basilar artery and its branches (3.7%), and permanent diabetes insipidus, weight gain, and precocious puberty. Early postoperative mortality due to sepsis and hemorrhage was 0.21%.[64] Complication rates of around 8 to 9% were reported in other series.[9,60] Intraoperative complications included intraventricular hemorrhage, cardiovascular changes, such as, bradycardia during fenestration and inflation of the balloon of the Fogarty catheter, and damage to the hypothalamus and fornix.[60] Within the first month following ETV, the complications reported included a CSF leak, ventriculitis, subdural fluid collection, and restenosis of the stoma.[30,60,64,65] The risk of complication was higher with repeat ETV procedures and in patients with prior shunts.[51]

Misplacement of the fenestration in the third ventricular floor usually accounts for most of the intraoperative hemorrhagic as well as cranial nerve-related complications. This can be avoided by recognizing the mamillary bodies, foramen of Monro, infundibular recess, dorsum sellae, and staying in the midline. It is also important to not go ahead with performing the ETV if the anatomy is not suitable.[66]

Bradycardia has been reported in up to 41% of the cases during fenestration of the third ventricular floor and the mechanisms postulated include stimulation of the preoptic area, at which time there may be associated hypotension. Stimulation of the posterior hypothalamus causes tachycardia with hypertension.[60,67] Use of normal saline instead of Ringer's Lactate for irrigation could reduce the risk of hyperkalemia-induced bradycardia.

Subdural hygromas as well as chronic subdural hematomas have been reported following ETV, attributable to the sudden excessive release of CSF or a large cortical puncture allowing egress of CSF into the subdural space.[60]

In this issue of the Journal, Singh *et al.*,[68] have reported the perioperative complications during neuroendoscopic procedures, including ETV. Tachycardia was the most frequently encountered intraoperative complication, while fever was the most common immediate postoperative complication. Massive intraoperative bleeding was the most serious complication. As their study was a retrospective one, involving patients with varied etiologies, they could only speculate on the causes of the complications and suggest possible preventive measures.[68]

Failed Endoscopic Third Ventriculostomy and its Management

Endoscopic third ventriculostomy failure occurred, with recurrence of symptoms, in about 10 to 20% of the patients with aqueductal stenosis and in up to 50% of the patients in whom ETV was performed for other indications. In a literature review, the failure rate was reported to range from 8 to 69% (8 to 47% when infants were excluded).[69] It was acknowledged that most ETV failures occurred early within a few days to two weeks following the procedure, with very few failures reported after six months. The cumulative probability of a failure presenting itself during the first 16 days after ETV was found to be 90%.[2,8,9,15,17,38,46,60,70] However, there are reports of delayed failures after many months, leading to rapid deterioration and death.[65] The predictors of failure include, age less than one year, with failures being higher in younger infants, posthemorrhagic and postinfectious indications, hydrocephalus with repeated shunt malfunctions, intraoperative recognition of a thickened third ventricular floor, patent aqueduct, scarring within the basal cisterns, and routine placement of external ventricular drains.[6,30,50,54,60,70]

When failure of ETV is suspected, cine MR and two millimeter sagittal TSE T2 sequences to detect the flow across the stoma may be performed.[36,70,71] In patients with no flow, endoscopic re-exploration may be done to reopen the stoma in cases of closure. In several cases, newly formed arachnoid membranes may be detected and these may have to be reopened. In case the stoma is patent, shunt insertion must be considered.[70] Shunt placement has also been the option when late failures occur.[9] In this issue of the Journal, the role of repeat ETV in patients with closure of a previously performed ETV was analyzed, and poor patient selection was noted to be the most important cause for early failure of ETV. The authors chose to shunt 45 of 51 patients with early

ETV failures (within seven days of the ETV), while repeat ETV was performed in 26 of 32 patients, with delayed failure. The success rate of repeat ETV was 50% in the early failure group and 78% in the late failure group, further reiterating that the poor choice of patients in whom ETV was performed initially could contribute to a high rate of initial failures.[72]

Following ETV, the intracranial pressure tends to remain high in a subgroup of patients for up to nine days, while in others it normalizes within 24 hours. These patients have an 'adaptation period' for establishment of flow through the ventriculostomy stoma. Up to three lumbar punctures in the interim period have been recommended to promote flow through the stoma (before terming it as a failure) for ensuring success of the ETV in asymptomatic as well as symptomatic patients.[73-75] It has also been noted that the CSF absorptive capacity as well as CSF circulation through the subarachnoid spaces shows further improvement only several months following ETV.[74]

Endoscopic Third Ventriculostomy Versus Shunt

Even as ETV may score over a shunt in terms of: (a) Avoidance of a foreign body implantation, and (b) establishment of 'physiological' CSF circulation, shunts still have a role in the management of hydrocephalus in certain situations, such as, infantile hydrocephalus, particularly in resource-constrained environments.[1,4,30,33,60,76] Shunts are technically easier to perform than ETV. The risk of intraoperative complications is slightly higher with ETV than with a shunt.[21,76] In the long term, shunts have a slightly higher rate of malfunction and infection. In a review of literature, the percentage of pediatric patients who were revision-free following ETV was 52% at two years and 30% at five years compared to 55 and 24% of the patients, following a ventriculoperitoneal shunt.[69] Infective complications in children are significantly lower following ETV vis-à-vis shunts and infections following ETV have a more benign course, being amenable to antibiotic treatment alone.[76] However, the quality of life estimates in children undergoing ETV or shunt, one year following the procedure, have been found to be similar.[77]

Conclusions

Endoscopic third ventriculostomy has been found to be an effective alternative strategy to shunt placement in obstructive hydrocephalus secondary to aqueductal stenosis, with good long-term results. Use of ETV in other situations must be judicious, with an appropriate review of the patient's ventricular anatomy as well as etiology of the hydrocephalus. Ultimately, the management of hydrocephalus needs to be individualized with both the options of ETV and shunt being appropriate in individual circumstances.

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