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Clinical Article

Endoscopic Third Ventriculostomy in Patients with Shunt Malfunction

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Objective : This paper presents data from a retrospective study of endoscopic third ventriculostomy (ETV) in patients with shunt malfunction and proposes a simple and reasonable post-operative protocol that can detect ETV failure.

Methods : We enrolled 19 consecutive hydrocephalus patients (11 male and 8 female) who were treated with ETV between April 2001 and July 2010 after failure of previously placed shunts. We evaluated for correlations between the success rate of ETV and the following parameters : age at the time of surgery, etiology of hydrocephalus, number of shunt revisions, interval between the initial diagnosis of hydrocephalus or the last shunt placement and ETV, and the indwelling time of external ventricular drainage.

Results : At the time of ETV after shunt failure, 14 of the 19 patients were in the pediatric age group and 5 were adults, with ages ranging from 14 months to 42 years (median age, 12 years). The patients had initially been diagnosed with hydrocephalus between the ages of 1 month 24 days and 32 years (median age, 6 years 3 months). The etiology of hydrocephalus was neoplasm in 7 patients; infection in 5; malformation, such as aqueductal stenosis or megacisterna magna in 3; trauma in 1; and unknown in 3. The overall success rate during the median follow-up duration of 1.4 years (9 days to 8.7 years) after secondary ETV was 68.4%. None of the possible contributing factors for successful ETV, including age (p=0.97) and the etiology of hydrocephalus (p=0.79), were statistically correlated with outcomes in our series.

Conclusion : The use of ETV in patients with shunt malfunction resulted in shunt independence in 68.4% of cases. Age, etiology of hydrocephalus, and other contributing factors were not statistically correlated with ETV success. External ventricular drainage management during the immediate post-ETV period is a good means of detecting ETV failure.

Key Words : Shunt failure · Secondary endoscopic third ventriculostomy · Hydrocephalus.

INTRODUCTION

Shunt placement which has been a standard treatment for patients with hydrocephalus, widely used as a first line treatment, and also has shown major progress. It has broad indications for both communicating and obstructive types of hydrocephalus and for various etiologies of hydrocephalus, including infection; congenital malformations, such as aqueductal stenosis, congenital cysts, megacisterna magna, and Arnold-Chiari malformation; hemorrhage; and tumor. The risk of shunt malfunction is relatively high : 25 to 40% in the first year after shunt placement, 4 to 5% per year thereafter, and 81% of shunted patients require revision after 12 years. Therefore, shunt failure is almost inevitable during a patient's life[23,26]. Given this near inevitability of shunt failure, endoscopic third ventriculostomy (ETV) for hydrocephalus was an important advancement for patients with hydrocephalus. Patient age and the etiology of hydrocephalus have been reported to be associated with outcomes after ETV[4,5,7,20,22,23,25,26,30]. Interestingly, the results are different when ETV is performed after shunt failure (secondary ETV) than when ETV is performed as an initial treatment modality for hydrocephalus (primary ETV). Primary and secondary ETV outcomes for post-hemorrhagic hydrocephalus are 27% and 71%, respectively, and for post-meningitic hydrocephalus they are 0% and 75%, respectively[23]. Recently, Marton et al.[22] reported outcomes of secondary ETV in pediatric patients and described a post-operative protocol. In this paper, we present our data from a retrospective study of secondary ETV, and we propose a simple and reasonable post-operative protocol that can detect ETV failure. The approach and management of hydrocephalus recurrence after secondary ETV are also discussed.

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MATERIALS AND METHODS

In this retrospective study, we enrolled 19 consecutive hydrocephalus patients (11 male and 8 female) who were treated with ETV between April 2001 and July 2010 after experiencing failure of previously placed shunts. All available clinical and neuroimaging findings were retrospectively reviewed, focusing on patient demographics, the etiology of hydrocephalus, initial treatment modalities, number of shunt revisions prior to ETV, presenting symptoms for ETV, interval between the initial diagnosis of hydrocephalus or the last shunt placement and ETV, post-ETV management, and outcome of secondary ETV.

Hydrocephalus and underlying pathology were diagnosed based on computed tomography (CT) scans, magnetic resonance image (MRI) findings, or pathologic confirmation. Based upon clinical and radiological data, shunt failure was diagnosed when there were symptoms associated with ventriculomegaly on CT scans.

ETV procedures were performed under general anesthesia with patients in the supine position. After linear skin incisions, the approach was through a frontal burr hole (1-2 cm anterior to the coronal suture and 2.5-3 cm lateral to the midline) to ensure an adequate trajectory, when needed, to reach the posterior part of the third ventricle or the aqueduct and fourth ventricle. All intraventricular procedures were performed with a rigid Storz endoscope (2.9-mm outer diameter) using a free-hand technique through a 5.6 mm working channel with a high-resolution (HD) camera. Intraventricular manipulations started with observation of the lateral and third ventricles. On the third ventricle floor, landmarks for ETV, including the mamillary bodies, tuber cinereum, and infundibular recess, were identified and the tissue was penetrated using bipolar coagulation. To establish communication between the lateral ventricles, the septum pellucidum was perforated as needed. During the procedure, we connected an intraventricular catheter to a reservoir of Ringer's lactate solution for intraventricular inflation and irrigation. Hemostasis was attained either by bipolar coagulation or by local continuous irrigation over several minutes. After confirmation of both CSF flow through the stoma and complete hemostasis, prolonged irrigation with Ringer's lactate solution was performed to remove any free tissue debris. Intraventricular inflation with Ringer's lactate solution was continued up to the final stages of the procedure to prevent ventricular collapse or the entrance air. After retrieving the endoscope, an external ventricular drainage (EVD) catheter was inserted through the same trajectory the endoscope had made. The openings of the dura were covered with gel foam, and soft tissues were sutured in layers. After ventriculostomy, the ventricular catheter of the shunt was removed gently. Usually, the ventricular catheter showed obstruction of the holes located distally in the catheter and adhesions to adjacent vessels and the ventricle wall that required adhesiolysis with the bipolar coagulator after ventriculostomy (Fig. 1).

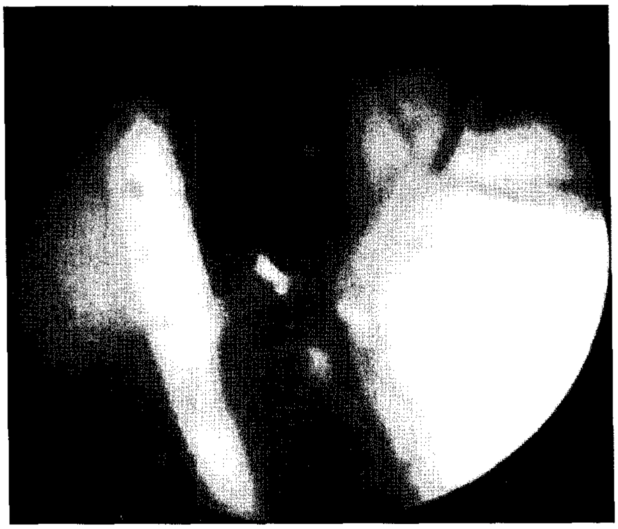

Fig. 1. Endoscopic image of the right lateral ventricle in patients with shunt failure. The ventricular catheter shows obstruction of holes distally in the catheter and adhesion to adjacent vessels and the ventricle wall.

We obtained CT scans of all patients upon awakening from anesthesia post-EVD. If there were no contraindications, we kept the EVD catheter closed. Immediate post-operative contraindications to closure of the EVD catheter included signs or symptoms of increased intracranial pressure (ICP), a significant amount of blood mixed with CSF from the EVD catheter, and aggravated hydrocephalus on post-operative CT scans. After 2-3 days of observation in the ICU with hourly ICP monitoring measured using the EVD catheter, the decision of whether or not to remove the EVD was made. During this period, we raised the chamber gradually to close the EVD catheter in order to assess how the patient would tolerate having the ETV without the EVD; we called this process EVD weaning. When this was well tolerated, the EVD was removed and the patient was transferred to a general ward. Otherwise, after 3-4 days of additional observation and attempts at EVD weaning, we determined whether or not secondary ETV had failed. The duration of observation varied for each case. Antibiotics were administered 2-7 days post-operatively during the period when EVD catheter was indwelling. We did not perform post-operative MRI on a routine basis. We also did not perform lumbar punctures, though it is reported to have a positive effect on facilitating flow through the ventriculostomy[22]. In all of the patients who were determined to have secondary ETV failure, shunt placement was performed.

The correlation between the success of ETV and patient age at surgery, etiology of hydrocephalus, number of shunt revisions, interval between the first diagnosis of hydrocephalus or the last shunt placement and ETV, and EVD indwelling time were tested with the Wilcoxon two-sample test or Fisher's exact test, with $p<0.05$ indicating statistical significance.

RESULTS

At the time of ETV after shunt failure, 14 out of 19 patients were in the pediatric age group and 5 were adults; ages ranged from 14 months to 42 years (median age, 12 years). Among the five adult patients, two had been diagnosed with hydrocephalus when they were children. Ages at first diagnosis of hydrocephalus ranged from 2 months to 32 years (median age, 6 years 3 months). The etiology of hydrocephalus was neoplasm in 7 patients, including anaplastic ependymoma, pilocytic astrocytoma, germinoma, epidermoid cyst, tectal tumor, and 2 cases of medulloblastoma; infection in 5; malformation in 3, including 2 cases of aqueductal stenosis and 1 megacisterna magna; trauma in 1; and unknown in 3. Initial treatment modalities for hydrocephalus were shunt placements, except in one patient who received ETV. He had medulloblastoma, and 1 month after ETV he developed aggravating ventriculomegaly and a shunt was placed. After two revisions for shunt malfunctions, ETV was performed again and the patient was free of symptoms of hydrocephalus. The average number of shunt revisions before ETV was 0.84 (range, 0 to 3 times), and these revisions occurred 40 days to 17.8 years (median 4.8 years) before the ETV. The presenting symptoms at the time of secondary ETV were the following : headache in 5 patients; mental change in 4; vomiting in 4; cranial nerve palsy in 2; and other symptoms, including seizure, gait disturbance, increased head circumference, and lethargy. The median interval between the first diagnosis of hydrocephalus and ETV was 4.8 years (range, 40 days to 17 years), and the median interval between last VPS and ETV was 3.4 years (range, 17 days to 13 years). After ventriculostomy, the shunt was removed in all patients except one. As it had been 13 years since this patient's ventriculoperitoneal shunt operation, we anticipated a high risk of complications during removal. Most cases had the EVD catheter inserted at the end of the operation, except for one patient. The EVD was left in place for a minimum of 0 to a maximum of 14 days, and the mean duration was 4.3 days. All of the successful secondary ETV patients showed ICP below 10-15 cmH_2O post-operatively, whereas failed patients showed ICP over 20 cmH_2O, CSF leakage, or neurological deficits. The overall success rate during the median follow-up duration of 1.4 years (9 days to 8.7 years) after secondary ETV was 68.4%. The success rate according to each etiology was as follows; 71% in neoplasm, 80% in infection, 33% in malformation, 100% in trauma, and 67% in idiopathic. The rest of the patients, who had secondary ETV failure, went on to receive shunt placement. Mean time to secondary ETV failure was 13.2 days (range, 5 days to 30 days). There was no morbidity associated with the neuroendoscopic procedure and no late ETV failure (Table 1). None of the possible contributing factors for successful ETV, including age (p=0.97) and etiology of hydrocephalus (p=0.79), were statistically correlated with outcomes in our series (Table 2).

Table 1. Patient characteristics

	No. of patients and others
Age at secondary ETV	12.0 years (1-42 years)
Sex (male/female)	11/8
Etiology of hydrocephalus	
Neoplasm	7
Infection	5
Malformation	3
Trauma	1
Idiopathic	3
Average number of shunt revision before ETV	0.84 (range, 0-3)
Presenting symptom for ETV	
Headache	5
Mental change	4
Vomiting	4
Cranial nerve palsy	2
Etc.*	4
Median interval between	
The first diagnosis of hydrocephalus and ETV	4.8 years (40 days-17 years)
The last shunt placement and ETV	3.4 years (17 days-13 years)
Mean EVD indwelling time	4.3 days (range, 0-14 days)
Overall success rate of secondary ETV	68.4%
Mean time to secondary ETV failure	13.2 days (range, 5-30 days)

*Seizure, gait disturbance, increased head circumference, and lethargy. ETV : endoscopic third ventriculostomy, EVD : external ventricular drainage

Table 2. Univariate analysis of contributing factors for secondary ETV success

	p-value
Age at secondary ETV	0.97
Etiology of hydrocephalus	0.79
Number of shunt revisions before ETV	0.43
Interval between	
The first diagnosis of hydrocephalus and ETV	0.90
The last shunt placement and ETV	0.42
EVD indwelling time	1.00

DISCUSSION

ETV has progressed greatly through the refinement of neuroendoscopic techniques and endoscopic optics since Sayers and Kosnik (1976) first described the use of percutaneous ventriculostomy in patients with hydrocephalus who had previously undergone shunt placement and Jones et al.[5,17] published a series of patients treated with ETV for the first time.

ETV has proven to be effective in the management of hydrocephalus regardless of the etiology, patient age, and other contributing factors, though there are reported differences in success rates. However, the timing of ETV, that is whether it is primary or secondary ETV, affects results. At present, there are many patients with shunts; therefore, there is good reason to understand secondary ETV and its implications.

Herein, we focused on the outcome of ETV in patients with shunt malfunction. Representative studies have reported overall success rates of 52%[7], 82% in an adult obstructive hydrocephalus population[5], and 76.7% in a pediatric obstructive hydrocephalus population[8]. As post-operative failure usually occurs early within the first month after surgery, strict follow-up, especially in the first year after surgery, is mandatory. Follow-up should be maintained in these patients as delayed failures have been described, which can cause acute malfunction of the ETV and can even result in sudden death[23]. In our series, the patients were predominantly children, and the overall success rate was 68.4%. All failures of secondary ETV occurred within the first month after operation. No delayed failure occurred during a median of 1.4 years of follow-up. The outcomes of secondary ETV were analyzed with respect to various possible contributing factors, and none of them showed statistical significance.

Particular considerations on age and the etiology of hydrocephalus have been discussed. Data from the literature suggest that the clinical response to ETV in adult patients with obstructive hydrocephalus is different from that in children, and the differences are secondary to the age at onset of hydrocephalus, CSF dynamics, and changes in brain viscoelastic properties[8,11,22]. Rates of success reported for ETV in patients 2 years old and younger vary from 0 to 83.3% with a mean of 47.8%, which is significantly lower than the success rate in older children[2,3,6,9,10,12-14,16,18-21,24,28,29]. However, when evaluating success rates of secondary ETV, Marton et al.[22] reported that age does not have a statistically significant effect, which is also reflected by our data.

As mentioned earlier, secondary ETV outcomes are better compared to primary ETV regardless of the etiology of hydrocephalus. Siomin et al.[27] reported an increase in ETV efficacy in post-hemorrhagic hydrocephalus from 60.9% for primary ETV to 100% for secondary ETV. O'Brien et al.[23] reported success rates for primary and secondary ETV as 27% and 71%, respectively, in post-hemorrhagic hydrocephalus and 0% and 75%, respectively, in post-meningitic hydrocephalus. These results are probably related to the fact that post-hemorrhagic and post-meningitic hydrocephalus initially result in communicating hydrocephalus because of obstruction of the subarachnoid spaces and arachnoid granulations with hemorrhagic or infective debris. The procedure may also induce an acquired aqueductal stenosis through continuous CSF diversion, increasing the likelihood of ETV success after a shunt malfunction. Similarly, the failure of primary ETV in infective or hemorrhagic hydrocephalus can be explained by the blockade of the CSF subarachnoid spaces and arachnoid granulations by debris and membranes, which counteract the mechanism of the ETV[22]. In an effort to unveil the mechanism behind this phenomenon, we tested whether the interval between the first VPS and the ETV affected the outcome, and we found no statistically significant effect. Further investigation should seek to understand the mechanism of secondary ETV.

Additionally, studies should be organized according to the type of hydrocephalus. Many former studies compared outcomes between obstructive and communicating hydrocephalus. However, communicating hydrocephalus carries its own uncertainty in nomenclature and pathophysiology[15,22]. Secondary ETV is performed at the time of induced or acquired aqueductal stenosis, which is obstructive hydrocephalus, and it is meaningless to categorize initial hydrocephalus as communicating or obstructive. For further study and understanding of ETV, it seems reasonable to categorize hydrocephalus, as in our series, according to etiology, including neoplasm, infection, trauma, malformation, and other causes. Though our data showed no statistically significant effect of hydrocephalus etiology on ETV outcome, likely because of the small number of enrolled patients, we could not definitively conclude that there is no correlation between the etiology of hydrocephalus and outcomes of secondary ETV.

Previous studies from various institutions have outlined their own pre- and post-ETV management strategies. Most of them performed MRI when shunt malfunction was suspected. One study selected patients for ETV based on sagittal T2-weighted MRI images, evaluating the patency and morphology of the prepontine cisterns and the anatomical features of the third ventricle floor[22]. We did not perform MRI routinely because, practically, there was no time to do it for patients who presented with acute hydrocephalus. In a previous study, MRI was performed after EVD insertion[22], but this may distort the patient's original anatomy. As shown in a study by Aydin et al.[1], in most cases with ventriculomegaly the basilar artery gets closer to the third ventricle floor. This allows direct visualization of the pulsations of the basilar artery and its silhouette. Patients who present to hospitals with shunt failure are considered urgent cases, and most of them receive EVD rather than shunt placement initially. ETV and EVD have a common entry point and trajectory. If the ventricular floor status allows, ETV can be performed; otherwise, only EVD is performed initially and then shunt placement is considered instead of ETV.

Aside from these considerations, means of augmenting the effect of ETV, such as lumbar puncture, and post-secondary ETV failure management strategies need to be discussed. We do not think it is necessary to perform MRI after shunt failure. In cases of secondary ETV failure, MRI should be performed to determine if another ETV is possible. If the failure is not due to closure of the ETV stoma, shunt placement should be considered.

The major weakness of our study was the small number of patients, which limited our ability to reach conclusive results. But our study, combined with results of previous studies, suggests that further categorization of hydrocephalus patients will lead to better decision-making and treatment of patients with hydrocephalus.

CONCLUSION

In this study, the use of ETV in patients with shunt malfunction resulted in shunt independence in 68.4% of cases. Age, etiology of hydrocephalus, and other contributing factors were not statistically correlated with outcome of secondary ETV. External ventricular drainage management during the immediate post-ETV period is a good means of detecting ETV failure. Future studies with larger groups of patients that investigate the mechanism of ETV and the factors that contribute to successful outcomes will elucidate the proper clinical pathway for the treatment of patients with hydrocephalus.

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