Original Article

Secondary Endoscopic Third Ventriculostomy for Shunt Malfunction

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ARTICLE INFO

ABSTRACT

Background: The efficacy of endoscopic third ventriculostomy (ETV) following cerebrospinal fluid shunting procedures has been disputed and claimed as having a less favorable outcome. Objective: The aim of this study is to evaluate the success of ETV in the treatment of obstructive hydrocephalus in patients primarily treated by ventriculoperitoneal shunt (VPS). Patients and Methods: A retrospective analysis of the medical records of fifteen patients managed by secondary ETV for obstructive hydrocephalus after VPS malfunction was performed. Patients younger than 1 year of age and those with communicating hydrocephalus were excluded. In case of favorable clinical outcome, Computed tomography (CT) brain and Magnetic Resonance Imaging (MRI) proved functioning stoma, the VPS was initially ligated or removed 3 months after ETV. the patients clinical data were analyzed in relation to outcome. Results: The mean age of the patients was 5.6 ± 4.8 years (range 2 – 18 years). ETV was successful in ten patients (66.6%). No major morbidity or mortality was recorded; only four patients had a minor morbidity. Shunt revision was done in five patients, within the first month. We had no delayed ETV failure for an average twelve months follow up period. None of the contributing factors were statistically correlated to the outcome in our series. Conclusion: Secondary ETV is a safe and effective technique in the management of patients with non-communicating hydrocephalus with malfunctioning shunts. Failures manifest early in the first month, though long-term follow up is mandatory.

INTRODUCTION

Endoscopic third ventriculostomy (ETV) is considered a routine and safe technique in the management of obstructive hydrocephalus; the success rate of this technique has been reported to be as high as 90% especially in patients with proven MRI obstruction of the aqueduct or fourth ventricular outlets as the cause of hydrocephalus.

Despite ETV’s proven efficacy as a primary surgery for obstructive hydrocephalus, a substantial number of patients with obstructive hydrocephalus are still being treated by ventriculo-peritoneal shunt (VPS) insertion; principally, due to the unavailability of either the endoscopic instruments or trained personnel to perform it; especially if the patient presented by progressive deterioration of the level of consciousness or radiological evidence of acute hydrocephalus, necessitating rapid intervention.

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The potential advantages of ETV over VPS include a lower rate of infectious complications, the avoidance of the great variety of shunt-related mechanical complications, and lower long-term morbidity and mortality rates. For these reasons endoscopic third ventriculostomy (ETV) is gaining wide acceptance and popularity as a surgical alternative for patients with hydrocephalus.

The two main factors that affect the outcome of ETV are the patient’s age and the etiology of hydrocephalus; interestingly, a different outcome to these factors were observed, when ETV was performed after shunt failure (secondary ETV) than when it was 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.

In this study we are evaluating the possibility of ETV in treating V-P shunt malfunction, and making patients independent from V-P shunt drainage.
PATIENTS AND METHODS

We retrospectively studied fifteen cases with obstructive hydrocephalus that were primarily treated with ventriculo-peritoneal shunt and presented later with shunt malfunction (symptomatic persistent failure of CSF drainage) were treated by secondary ETV. Only cases of obstructive hydrocephalus that were treated with VPS and older than one year of age at time of cases of obstructive hydrocephalus that were treated by secondary ETV. Only cases of obstructive hydrocephalus that were treated with VPS and older than one year of age at time of shunt malfunction were included in our study. In all patients, MRI proved non-communicating variety of hydrocephalus; the obstruction was in aqueduct or the outflow of the fourth ventricle.

ETV was performed with patients under general anesthesia and in the supine position. We used semilunar skin incisions for the elevation of periosteal flap and to help in closure of the burrhole site after the procedure.

Frontal burrhole was planned for ventricular access (1-2 cm anterior to the coronal suture and 2.5-3 cm lateral to the midline). All the cases were performed by a free-hand technique, utilizing a GAAB rigid neuro-endoscopic system (Storz GmbH & Co., Tuttingen, Germany).

The floor of the third ventricle in front of the mammillary bodies was perforated using the blunt tip of the bipolar electrode, when the floor was opaque and tough, bipolar current was used to open the membrane. In cases where there was unilateral obstruction to the CSF flow in one of the lateral ventricles, septum pellucidotomy was performed to connect both lateral ventricles.

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. After retrieving the endoscope, the corticotomy was closed with a large triangular piece of gel foam, which was tightly compressed to form a wedged plug and soft tissues were sutured in layers over it.

We did not insert an external drainage catheter (which could decrease the success rate of ETV) and postoperatively closely observed the patient to detect any signs and symptoms of raised intracranial pressure. Antibiotics were administered 10-14 days post-operatively.

Our protocol was to perform ETV and to ligate the distal end of the shunt while leaving it in its place, the shunt was later removed once the ETV proved its success except in patient were the shunts was adherent, it was left in place to minimize hemorrhagic complications. Before discharge, CT or MRI CISS were performed in addition to the clinical picture to confirm a functioning ETV (Fig.1, O).

Patients were followed on weekly basis for a month, then every 2 weeks for the following two months to exclude early failure of ETV.

In cases with good clinical status, functional stoma, and reduction of ventricular enlargement according to MRI performed 3 months after ETV, the procedure was considered successful.

The correlation between the success of ETV and patient symptoms at first clinical presentation, etiology of hydrocephalus, age at initial shunt, number and date of shunt revisions, symptoms at late clinical presentation, age at ETV, time for ETV to fail, complications, post-ETV management, and outcome of secondary ETV and subsequent shunting procedures performed were tested with the Fisher’s exact test, with p<0.05 indicating statistical significance.

RESULTS

The study included nine males and six females. All patients were previously treated by V-P shunting for hydrocephalus. The age at which the patients were first diagnosed with hydrocephalus and at which V-P shunt was inserted ranged from 2 months to 11 years (mean age, 2.5 years ± 3.7). The average number of shunts revisions before ETV was 0.93±1.4 (range, 0 to 5 times).

The cause for which the patients were initially operated for by V-P shunting was as follows: seven patients had hydrocephalus due to congenital aqueductal stenosis, three patients had post-hemorrhagic hydrocephalus, two patients had post-infectious hydrocephalus, , a patient had a tectal tumor , a patient had traumatic cerebellar hemorrhagic contusions causing hydrocephalus and another patient had a Chairi malformation which was shunted elsewhere and the parents had refused posterior fossa decompressive surgery.

The presenting symptoms at the time of secondary ETV were: headache in eight patients; vomiting in four; mental change in three; abducent nerve palsy in two and other symptoms, including seizure, gait disturbance, increased head circumference, and lethargy.

The mean age at the time of ETV was 5.6 ± 4.8 years (range 2 – 18 years). The mean duration from the last shunt placement to ETV was 2.4 ± 2.9 years (ranged from 4 months to 12 years). Follow up ranged from 4 months to 24 months with an average of 12 months.

The overall success rate for ETV was 66.6 % was successful where in ten out of Fifteen patients, the V-P shunt drainage was removed and no subsequent need of drainage or surgery for hydrocephalus throughout the follow-up.

Patients who failed to improve with ETV presented with CSF leak from the site of the wound, a disturbed conscious level and fits. CT brain proved signs of high pressure. Those patients had V-P shunt placement. The
Mean time to secondary ETV failure was 13.2 days (range, 5 days to 25 days).

Overall, we were not successful in five patients (33.3%). Subgialial CSF bogginess was detected in 5 patients of which 3 had CSF leak; all five patients eventually have failed the procedure and required V-P shunt placement. Minor hemorrhage was observed in 2 patients (A thin rim of subdural hematoma in one case and intraventricular hemorrhage in the other case) both patients were managed conservatively and their postoperative period went uneventful. There were no mortalities 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.

All the patients who had a successful ventriculostomy, had their shunts removed; except in one patient where the shunt had been inserted 13 years earlier for fear of hemorrhagic complications due to adherence of the ventricular end to the choroid plexus.

Illustrative case 1 (Fig. 1)
A 3.5 years old poly-traumatized boy presented by rapid deterioration of the level of consciousness; due to acute hydrocephalus caused by a cerebellar contusion obstructing the fourth ventricle as observed by CT. The patient was initially managed by VP shunt insertion that nine months later malfunctioned. ETV and shunt ligation were performed, after which the patient showed full recovery.

Illustrative case 2 (Fig. 2)
A 15 year old male patient presented by severe headache, persistent vomiting and unsteady gait. The patient had been diagnosed as a Chiari malformation grade 0 for which he was operated for through VPS five years earlier at another institute. The patient had five shunt revisions prior to his presentation. MRI revealed pan-ventriculomegaly and extensive cervico-dorsal syrinx. Posterior fossa decompression was proposed, but it was refused by the parents thus secondary ETV was suggested as a possible alternative. Patient had complete resolution of his preoperative symptoms with normal neurological examination till the last recorded follow up one year following the ETV.

Fig. 2 A-D: Illustrative case 2 of Chiari malformation: A,C: Axial and sagittal MRI images of a Chiari malformation patient with pan ventricular dilatation and extensive syrinx in the cervical region due to malfunction of VPS  B, D: One year post ETV axial and sagittal images showing marked reduction of both the ventriculomegaly and syrinx with patency of the ETV ostium.

Table 1. Patient characteristics

<table>
<thead>
<tr>
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<th>No. of patients and others</th>
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<tbody>
<tr>
<td>Age at secondary ETV</td>
<td>5.6±4.8 years (4mo-12 years)</td>
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<tr>
<td>Sex (male/female)</td>
<td>9/6</td>
</tr>
<tr>
<td>Etiology of hydrocephalus</td>
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</tr>
<tr>
<td>Tectal tumor</td>
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<tr>
<td>Post-infectious hydrocephalus</td>
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<tr>
<td>Posthemorrhagic hydrocephalus</td>
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<tr>
<td>Chiari malformation</td>
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<tr>
<td>Congenital aqueduct stenosis</td>
<td>7</td>
</tr>
<tr>
<td>Trauma</td>
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<tr>
<td>Average number of shunt revision before ETV</td>
<td>0.93±1.4 (range,0-5)</td>
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<td>Presenting symptom for ETV</td>
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<tr>
<td>Headache</td>
<td>8</td>
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<tr>
<td>Mental change</td>
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<tr>
<td>Vomiting</td>
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<tr>
<td>Abducentpalsy</td>
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</tr>
<tr>
<td>Others*</td>
<td>4</td>
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<tr>
<td>The last shunt placement and ETV</td>
<td>2.4±2.9 years (4month-12 years)</td>
</tr>
<tr>
<td>Overall success rate of secondary ETV</td>
<td>66.6 %</td>
</tr>
<tr>
<td>Mean time to secondary ETV failure</td>
<td>13.2 days (range, 5-25 days)</td>
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</tbody>
</table>

*Seizure, gait disturbance, increased head circumference, and lethargy.
ETV: Endoscopic Third Ventriculostomy, EVD: External Ventricular Drainage
DISCUSSION

The V-P shunt or alternatively ventriculo-atrial shunt insertion were the most commonly used methods for the management of hydrocephalus, including obstructive hydrocephalus. In the past decade ETV has been established as the optimum treatment of most forms of obstructive hydrocephalus aided by the marked improvement of neuro-diagnostic imaging and neuro-endoscopy.6,12,15

High success rates of up to 90% have been reported with ETV with the advantage of restoration of the physiological CSF pathway without the use of any foreign material. The lower incidence of late complications in comparison to CSF shunt procedures, have endorsed the superiority of ETV.5

The original assumption that CSF resorption capacity would be lowered in the subarachnoid space after a few years with a working V-P drainage has not been confirmed. This assumption could not be substantiated nor refuted from the statistical point of view, despite achieving a success of ETV in patients who had longstanding CSF diversion by V-P shunts. Also literature review have demonstrated that the success rates of secondary ETV were comparable to primary ETV.4,13,14,15,26 In the current study the success rate of secondary ETV was 66.6%.

Secondary ETV is still a matter of controversy where an increased liability of complications has been reported by some studies8 while others reported that secondary ETV was safe and as successful as primary ETV14. No mortality was reported in the present series and minor complications related to the procedure were met in two patients (13.3%), which is not a significantly higher complication rate compared to primary ETV. Both secondary ETV complication (13.3%) and failure (33.3%) rates in the present study were comparable to those published in literature.11,12,15,26

Woodworth et al27, point out a 2.5 times greater risk of ETV failure in patients with previous V-P drainage implantation. We had Five ETV failures in our group. Failure of ETV is usually observed in the first postoperative month13, and for this reason the patients were kept under close observation for relatively longer period to deal with early ETV failure. In the current study secondary ETV failure had a mean of 13.2 days following surgery.

Some authors advocate the use of temporary external ventricular drainage insertion simultaneously with secondary ETV in previously V-P shunted patients presenting with acute hydrocephalus, in order to deal with early failure and record the intraventricular pressure.6,11,22 This strategy was not implemented in the current study, for fear of reduction of intraventricular tension and diversion of the CSF flow away from created ETV ostium leading to its early closure. Instead the patients were instructed to remain in vicinity of the hospital for the first postoperative month after their discharge, for close observation for early detection of signs and symptoms of raised intracranial pressure.

The majority of patients of the current study were operated on for either acute V-P drainage malfunction or surgical revision necessity, mostly due to insufficient abdominal drainage catheter length. The decision to perform secondary ETV was taken after thorough discussion of the various surgical options with the patient’s parents. Those who gave their consent for ETV were aiming to achieve shunt independence, which is reported to be as high as 70% or more in literature reviews of patients with shunt malfunction who underwent secondary ETV1,3,4,11,12,26.

Secondary ETV was mostly successful amongst congenital aqueduct stenosis cases, while patients with obstructive post-hemorrhagic hydrocephalus had the highest frequency of failure probably due to diminished CSF resorption capacity in the subarachnoid space. The small population of the study made it impossible to detect a statistically significant correlation between the success and failure of secondary ETV in relation to the different etiologies. Due to the fact that, even after the ETV, late failure can occur with the incidence of 2–15%,13 we recommend performing regular MRI examinations every second year in all patients, even without clinical problems.

CONCLUSION

Secondary ETV proved to be a viable option in the management of shunt malfunction with a success rate of 66% which is comparable to primary ETV. As it is a safe and a rapid treatment option even in emergency conditions, it is worth performing this procedure in previously shunted children. The presence of a malfunctioning shunt in situ should not be considered a contraindication to this procedure. Since most failures manifest early in the postoperative course, those who achieve successes will have a long lasting one. But long-term follow up is still mandatory.

REFERENCES


