Endoscopic third ventriculostomy for shunt malfunction: What to do with the shunt?

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Abstract

Background: Endoscopic third ventriculostomy (ETV) is an effective surgical option for the treatment of shunt malfunction. The role of postoperative cerebrospinal fluid (CSF) diversion is not clearly understood at this time. We compare the effects of shunt-removal/ligation, shunt externalization or external ventricular drain placement, and no treatment to the indwelling shunt at the time of ETV.

Methods: We retrospectively reviewed the records of 20 consecutive patients treated at our institution for shunt malfunction with ETV. Patient data were retrospectively evaluated for the effect that the fate of the shunt plays on ETV success rates.

Results: In our series of 20 patients we had an overall success rate of 70% with using ETV for shunt malfunction. Patients who had their shunts ligated at the time of surgery had a success rate of 88%, in comparison to those whom the shunt was left untouched who had a success rate of 60%, or patients who had a perioperative external ventricular drain placed the success rate was 50%.

Conclusions: This series of ETV for shunt malfunction performed at a single center by a single surgeon shows a success rate similar to the published literature range of 67 to 80 percent success whether the shunt is ligated or left undisturbed. It is not necessary to ligate the in situ shunt at the time of ETV; however, there may be a trend toward an improved success rate with shunt ligation. Further studies with a greater numbers of patients are warranted.

Key Words: Endoscopic third ventriculostomy, hydrocephalus, shunt malfunction, shunt

INTRODUCTION

Endoscopic third ventriculostomy (ETV) is a widely accepted treatment for obstructive hydrocephalus. The procedure was first performed by William Mixter, a urologist in 1923 using a ureteroscope to fenestrate the floor of the third ventricle. The advent of valve-regulated shunting technology proved to be an effective method for treating hydrocephalus of multiple causes. Third ventriculostomy remained a stagnant procedure with percutaneous and stereotactic alterations to the procedure requiring more highly specialized techniques and equipment limiting interest in developing the technique. There has been a resurgence in interest...
of applying ETV to broader pathologies as modern endoscopes with their fiber-optics and advanced light sources that allow for excellent resolution of ventricular anatomy and control the safe fenestration of the floor of the third ventricle.

To this end, ETV has been applied in the setting of shunt malfunction in patients whose imaging characteristics suggest obstructive hydrocephalus. Given shunt survivals of 61% in 1 year and 47% in 2 years in well controlled pediatric studies, the possibility of shunt independence following successful ETV makes it worthy of consideration in patients presenting with shunt malfunction. Studies evaluating the effectiveness of ETV in the treatment of shunt malfunction or infection have reported success rates of 67 to 80% (67%, 80%, 76.7%, and 70%). Reported complications from ETV in these series include infection, bleeding, need for repeat procedure, weight gain, diabetes insipidus, precocious puberty, infantile, and cranial nerve palsy with rates from 4.8 to 31% depending upon complications tracked and follow up.

ETV is thus a safe and effective treatment in appropriately selected patients for shunt malfunction. There is no consensus as to the fate of the previously placed ventriculoperitoneal or ventriculoatrial shunt in this patient population. In various studies, the shunt has been addressed either by removal, ligation, externalization, or removal and replacement with an external ventricular drain. Studies evaluating cerebrospinal fluid (CSF) dynamics following ETV for shunt malfunction suggest that there is a gradual shift from a shunt-dependent to a shunt-independent state over the course of 1 week. The role of continued CSF diversion is a controversial issue as there is a gradual conversion from shunt-dependence to shunt-independence. There is also a theoretical risk that with CSF diversion there is decreased flow through the ventriculostomy and potentially a decreased success rate. Some believe that continued flow is necessary for the maturation of the fistulous connection. The flip side of the argument is that unnecessary early ventricular dilation can compress the subarachnoid CSF spaces leading to extraventricular hydrocephalus and failure of the ETV on these grounds. The goal of the current study is to evaluate a single institution’s experience with third ventriculostomy and assess the effects of continued CSF diversion on ETV success rates.

**MATERIALS AND METHODS**

We retrospectively reviewed the medical records of all patients treated with third ventriculostomy for shunt malfunction presenting to our institution’s neurosurgical service between January 2004 and June 2009. Patient data were collected with the approval of the local institutional review board. All patients had a minimum of 6 months of follow up as it has been noted that in 95% of patients treated with ETV for shunt malfunction presented with failure within 1 month.

Patients presenting with shunt malfunction and imaging consistent with obstructive hydrocephalus were offered ETV in lieu of a shunt revision. Shunt malfunction was based upon history (including vomiting, progressive decreased level of consciousness, seizures, upgaze paralysis), physical examination (fontanel bulging, shunt refill), imaging findings (computed tomography or magnetic resonance imaging), and shunt tapping. The choice to proceed with ETV was based upon discussion of the risks and benefits of the procedure with the patient and surrogate. Depending upon the patient’s situation the shunt was left untouched, removed, ligated, or removed and replaced with an external ventricular drain.

All ETV’s were performed by the senior author using our standard technique. An incision was made over Kocher’s point and craniostomy was performed using a perforator drill. A ventricular catheter was then used to cannulate the lateral ventricle. This track was then followed under direct visualization with a zero degree scope. The floor of the third ventricle was then perforated and dilated with a four French Fogarty catheter, bipolar cautery, and irrigation were used as necessary for hemostasis. The scope was then removed, the craniostomy plugged with gel foam, and a layered closure was subsequently performed.

Patients were stratified into three groups. Group 1 patients underwent ETV and had removal or ligation of their shunts at the time of their initial surgery. Group 2 patients had their shunts left in place. Group 3 had their shunts ligated or removed and an external ventricular drain placed at the time of surgery.

All statistical analyses were performed using SPSS 17.0 (Chicago, IL). Means and standard deviations or medians were reported for continuous variables. Frequency and percentages were reported for categorical variables. Fisher’s exact test was conducted to examine categorical variables. Mann–Whitney testing was used to evaluate the association with continuous variables. In all analyses, a P value <0.05 was considered statistically significant.

**RESULTS**

**Study population**

Over the study period, 20 patients underwent ETV for the treatment of shunt malfunction. Of these patients, 11 were male and 9 were female. The average age of patients in this study was 12.9 years with a range from 7 months to 29 years. The causes of hydrocephalus were aqueductal stenosis in 30%, spinal dysraphism or spina bifida in 35%, germinal matrix hemorrhage in 15%, and other or unclear causes in 20% of patients. For this entire cohort the overall success rate was 70%. Patient characteristics are shown in Tables 1 and 2.
Operative complications and death
In our cohort of patients, there were no complications noted with the exception of a 30% third ventriculostomy failure rate determined by the need for shunt insertion.

Analysis of endoscopic third ventriculostomy failures
We had six endoscopic third ventriculostomies fail requiring the reinsertion of a shunt for continued CSF diversion [Table 3]. Four of the six failures were within the first 30 days of third ventriculostomy. The presentation of endoscopic third ventriculostomy failure in the subacute setting was wound leakage in half of the early failures. The remaining cases of early failure presented with the same symptoms as the patients’ previous shunt malfunctions leading to clinically symptomatic ventriculomegaly. There were two late failures of third ventriculostomy in this series; the first occurred at 5 months and presented with clinically symptomatic ventriculomegaly. The final case presented at 1 year and had extraventricular hydrocephalus manifesting as a pseudomeningocele.

Variables affecting endoscopic third ventriculostomy failure
In addition to the presenting symptoms associated with failure, we evaluated a number of patient and technical factors for significant effect on endoscopic third ventriculostomy failure. The P value of Fisher’s Exact test showed that sex did not significantly effect ETV success. For the status of the shunt at the time of surgery, Fisher’s Exact test was conducted only on the shunt ligated and shunt intact groups. It showed that there is no significant effect of the shunt status on the rate of ETV success. For the analysis of the effect of pathology on ETV success, our small number precluded our ability to detect a statistically significant effect [Table 4]. We did note a rate of 88% success for the ETV when the shunt was ligated or removed at the time of surgery, 60% when the shunt was not addressed at the time of surgery, and 50% when the shunt was replaced by an external ventricular drain (EVD). Of the patients undergoing EVD placement after ETV, one failed. In this patient the drain was clamped immediately postoperatively. The one successful

**Table 2: Characteristics of patients included in the study**

| Patient | Sex | Age (years) | Underlying pathology | Shunt status at time of surgery | ETV success? |
|---------|-----|-------------|----------------------|-------------------------------|--------------|
| 1       | F   | 22          | Spina bifida         | Ligated                       | Yes          |
| 2       | F   | 16          | Known                | Ligated                       | No           |
| 3       | M   | 16          | Aqueductal stenosis  | Ligated                       | Yes          |
| 4       | M   | 22 mo       | Aqueductal stenosis  | Intact                        | No           |
| 5       | F   | 12          | Spina bifida         | Intact                        | Yes          |
| 6       | F   | 27 mo       | Spina bifida         | Ligated                       | Yes          |
| 7       | M   | 3           | GMH                  | EVD placed                    | No           |
| 8       | F   | 8           | Spina bifida         | Ligated                       | Yes          |
| 9       | M   | 5           | GMH                  | Ligated                       | Yes          |
| 10      | F   | 25          | Aqueductal stenosis  | Removed                       | Yes          |
| 11      | M   | 21          | Tectal glioma        | Ligated                       | Yes          |
| 12      | M   | 7 mo        | Aqueductal stenosis  | Intact                        | No           |
| 13      | F   | 4           | Aqueductal stenosis  | Intact                        | Yes          |
| 14      | M   | 15          | Tectal glioma        | Intact                        | Yes          |
| 15      | M   | 29          | Spina bifida         | Intact                        | Yes          |
| 16      | M   | 9           | FVOO                 | Intact                        | No           |
| 17      | F   | 18          | Spina bifida         | Intact                        | Yes          |
| 18      | F   | 11          | Aqueductal stenosis  | Intact                        | Yes          |
| 19      | M   | 28          | Spina bifida         | EVD placed                    | Yes          |
| 20      | M   | 11          | GMH                  | Intact                        | No           |

GMH: Germinal matrix hemorrhage, FVOO: Fourth ventricular outlet obstruction

**Table 4: Analysis of patient factors associated with failure**

|                     | Successful ETV | Failed ETV | Success rate (%) |
|---------------------|----------------|------------|------------------|
| Overall             | 14             | 6          | 70               |
| Gender              |                |            |                  |
| Male                | 6              | 5          | 50               |
| Female              | 8              | 1          | 89               |
| Pathology           |                |            |                  |
| Aqueductal stenosis | 4              | 2          | 67               |
| Spina bifida        | 7              | 0          | 100              |
| Germinal matrix hemorrhage | 1  | 2      | 33               |
| Other               | 2              | 2          | 50               |
| Shunt Fate          |                |            |                  |
| Shunt ligated       | 7              | 1          | 88               |
| Shunt intact        | 6              | 4          | 60               |
| EVD                 | 1              | 1          | 50               |

Table 3: Analysis of ETV failures

| Patient | Time from ETV to failure | Symptoms at the time of failure |
|---------|--------------------------|-------------------------------|
| 2       | 12 Days                  | CSF leak from wound           |
| 4       | 8 Days                   | CSF leak from wound           |
| 7       | 15 Days                  | Symptomatic ventriculomegaly  |
| 12      | 1 Year                   | Pseudomeningocele             |
| 16      | 5 Months                 | Symptomatic ventriculomegaly  |
| 20      | 20 Days                  | Symptomatic ventriculomegaly  |
case of EVD after ETV had the EVD “weaned” over the course of 3 days. The limited number of patients in this grouping precludes analysis of the effect of the EVD weaning upon ETV success.

**Effect of age**

Age was found to be a statistically significant (\(P = 0.04\)) factor associated with shunt success or failure [Table 5]. The median age of patients who had a successful ETV was 15.5 years. The median age of patients who required revision of their third ventriculostomies to shunts was 6.0 years. These data demonstrate a significant effect with older patients having better outcomes with third ventriculostomy to treat shunt malfunction.

**DISCUSSION**

ETV is a safe and effective procedure for the treatment of appropriately selected patients with shunt malfunction with reported success rates in the 67-80% range. Our overall success rate of 70% falls within this published range. Our patient population, as in published studies, is a heterogeneous group in terms of age, gender, and underlying pathology.

To the best of our knowledge, there have been no studies looking at the effect of the fate of the shunt catheter in this population of patients. Dr. Teo notes that the presence of an external ventricular drain following ETV may lead to a reduced pressure gradient through natural CSF pathways if an external ventricular drain is left open at the time of surgery.\(^{[7]}\) This reasoning is the rationale for the routine removal or ligation of indwelling malfunctioning shunt. The theory is that the previous shunt may not necessarily be entirely occluded but may simply be too sluggish to allow the full treatment of hydrocephalus. This approach subjects patients to a second procedure, with the associated complications and longer anesthesia time. The counter argument to this reasoning is that ventriculomegaly in the early post-ETV period may lead to compression of the cisternal subarachnoid space and may be equally detrimental to ultimate long term patency of the ETV in treated patients. To the best of our knowledge, no formal study of the effect of an indwelling CSF diversionary device has been undertaken in the published literature.

We have presented here a small series of patients in whom the previous shunt was left unfettered except in the setting of infection or ventriculoatrial shunting. In either of these settings the goal of surgery was both to treat the underlying hydrocephalus and to remove the applicable hardware. In this population of patients we found a success rate of 60% for ETV in the setting of shunt malfunction in whom the existing hardware was left untouched. There was a trend toward increasing ETV failure without ligating the shunt, but the sample size precluded meaningful statistical significance testing. Likewise a success rate of 60% in this setting is near the published success rates for ETV in the setting of shunt malfunction. We have demonstrated that leaving the shunt in place following the fenestration of the floor of the third ventricle is safe and efficacious in treating hydrocephalus in previously shunted patients and preventing the need for shunt re-insertion. Interestingly, we had an even lower rate of ETV long-term viability (50%) in patients who had an extraventricular drain placed at the time of surgery. Admittedly, this is a small subset of our patients, but there appears to be a trend that with more efficacious extra-fenestration CSF flow there is decreasing durability of the iatrogenically-created fistula.

These data demonstrate that outside of the setting of active shunt infection, it is safe to simply perform a third ventriculostomy to treat shunt malfunction. The additional operative time and time under general anesthesia to remove a “clean” shunt system may be an additional risk for the patient. This additional operative time may be trivial in the setting of a relatively freshly placed shunt. However, the option to not remove the previously placed shunt may be most beneficial in the setting of the old calcified non-infected shunt. In this setting the additional procedure may require multiple incisions to attempt the removal. There is also the possibility that the proximal catheter may be adherent to the underlying brain, making attempts to remove it all the more dangerous at the time of third ventriculostomy.

Our experience is similar to the published literature with the majority of ETV failures occurring within the first 30 days following the procedure. These early failures usually present with CSF leakage from their wounds. The late (>30 days) failures of ETV tended to present more often with signs and symptoms of hydrocephalus and their previous shunt malfunctions. For this reason long-term follow up of these patients is necessary.

**CONCLUSIONS**

ETV is a safe procedure with relatively few complications and a reasonably high success rate in the setting of shunt malfunction. Failure in the setting of ETV tends to be within the first 30 days, however delayed failure can occur requiring long-term follow up for patients undergoing this procedure. A second procedure at the time of ETV to remove the previously malfunctioning shunt outside of the

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**Table 5: Effect of age upon ETV success**

| ETV success | Number of patients | Mean age (Years) |
|-------------|--------------------|-----------------|
| No          | 6                  | 6.33            |
| Yes         | 14                 | 12.29           |

*The mann–whitney test was used to evaluate the effect of patient age upon ETV success rates, yielding a \(P = 0.04\). This finding was statistically significant.*
setting of infection is not necessary on statistical grounds; however, there may be a theoretical benefit in terms of CSF dynamics and a trend toward improved outcomes. Further study regarding the fate of implanted hardware at the time of third ventriculostomy is warranted.

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