Third Ventriculocisternostomy for Shunt Failure

Virakpagna Chhun, Oumar Sacko, Sergio Boetto, Franck-Emmanuel Roux

BACKGROUND: **Our objective was to analyze the relevance, potential prognostic factors, and complications of endoscopic third ventriculostomy (ETV) in patients with shunt failures.**

METHODS: **Among 721 ETVs performed between 1999 and 2013, we studied 53 patients with shunts (31 men, 21 less than 18 years of age) who had an ETV performed for shunt failures as the result of various causes. We included all initial causes of hydrocephalus except adult chronic (i.e., "normal pressure") and pediatric communicant hydrocephalus. The mean duration between initial shunting for hydrocephalus and the ETV procedure was more than 11 years (137 months; range, 1 month to 34 years). Successful ETV procedure was defined as clinical improvement and shunt independence extending until the last follow-up visit.**

RESULTS: **The success rate of the ETV procedure was 70% (37 of the 53 cases) with a mean follow-up of 51 months (from 3 to 157 months) and was not related to the age of the patient ($P = 0.922$), to the cause of hydrocephalus ($P = 0.622$), or to the number of shunt failures ($P = 0.459$). We also found no statistical difference ($P = 0.343$) between patients whose shunt had been in place for less than 5 years and those shunted more than 5 years. The presence of an infected shunt was not predictive of ETV failure ($P = 0.395$). No significant intraoperative or postoperative complications were noted.**

CONCLUSION: **This study confirms that ETV should be considered as the first therapeutic option before shunt revision in cases of initial obstructive hydrocephalus.**

INTRODUCTION

Ventriculoperitoneal (VP) shunting of the cerebrospinal fluid has been the most common technique for the treatment of patients with hydrocephalus and is still the first therapeutic option for hydrocephalus in many countries (3, 24, 34, 45); however, it may be associated with several complications. Despite technological advances in their design, shunts still have significant rates of failure and complications (17, 25, 27, 35, 42). Endoscopic third ventriculostomy (ETV) has been developed as an alternative to shunts for treating obstructive hydrocephalus (12, 26, 38). More recently, it has been suggested (21, 30) that other etiologies of hydrocephalus could be managed by ETV, such as some cases of hydrocephalus associated with infection (20, 40) or overdrainage (1, 36). Finally, because of its increasing popularity and minimal complications (9, 13, 15), it has been suggested that ETV also could be considered as a good option for some cases of shunt failure (10, 31, 32).

The possibility for some patients with shunt failures to be "shunt free" after a single ETV can be considered as a major advantage. In this study, our objective was to report and analyze our experience concerning the role of ETV as an alternative to revision of VP shunts for patients with shunt failures. We analyzed the relevance of ETV in cases of shunt failure and its potential prognostic factors and complications.

METHODS

Between July 1999 and July 2013, 721 ETVs were performed in 654 patients with hydrocephalus (316 male and 338 female) at the University Hospital of Toulouse, France. Among these procedures, 54 ETVs were performed in patients previously fitted with various types of shunts to manage hydrocephalus and who had a shunt failure. One of these patients was excluded from the study because his clinical notes were incomplete. Thus, the ETV outcome was

Key words
- Aqueductal stenosis
- Endoscopic third ventriculostomy
- Hydrocephalus
- Shunt failure

Abbreviations and Acronyms
CSF: Cerebrospinal fluid
ETV: Endoscopic third ventriculostomy
VP: Ventriculoperitoneal

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analyzed in 53 patients. The information recorded included age, sex, clinical notes, etiology of hydrocephalus, any previous operations (including previous VP shunts), previous cerebrospinal fluid (CSF) infection or hemorrhage, intraoperative hemorrhage, and operative complications.

As suggested by Drake et al. (12), failure of ETV was defined as any subsequent surgical procedure for CSF diversion or death related to hydrocephalus management. In this study, criteria for success were: 1) disappearance of initial clinical signs and 2) independence from shunt extending until the last follow-up visit. The duration of a patient's follow-up was measured as the time to either ETV failure or the last follow-up news. Whenever possible, patients were interviewed by phone in Winter 2013. Overall, 73% of the patients (or the parents of patients who were young children) were contacted for a phone interview. (Questions included the following: No more clinical signs related to the shunt failure? Do you have any problem related to your ETV procedure? Did you return to the activities you did before shunt failure? For children, did you return to school?)

A total of 19% had been seen for evaluation in the previous 6 months and, for the remaining 8%, the last follow-up date was established as the last clinical follow-up on record. Etiologies of hydrocephalus were defined as follows: primary aqueductal stenosis; Chiari II hydrocephalus; hydrocephalus attributable to posterior fossa tumors; hydrocephalus attributable to pineal, tectal, and posterior third ventricle tumors; postinfectious hydrocephalus; and other causes of hydrocephalus (one suprasellar arachnoid cyst; one posterior fossa hemorrhage). No shunt failure in the context of 1) adult chronic hydrocephalus or 2) pediatric communicating hydrocephalus was managed by ETV. Furthermore, one neurosurgeon of our group did not perform ETV, and the patients he received in emergency for shunt failure were only managed by shunt revision. Finally, during these years 2 patients who were shunted for a long time refused the ETV procedure and preferred to be managed by shunt revision.

We used bifrontal ventricular size and third ventricle diameter to evaluate ETV success (comparing a computed tomography scan or MRI before the ETV procedure with the last available follow-up computed tomography scan or MRI). Both measures (bifrontal and third ventricle diameter) were added and compared pre- and postoperatively. A difference of more than 5% between pre- and postoperative measures was considered as a significant diminution of the ventricular size.

Endoscopic Technique

The technique used was based on the procedure described by Sainte-Rose (39). Since 1999, we have followed the same procedure (38), in which we used a rigid endoscope: Neuroendoscope (Aesculap, Tuttlingen, Germany), diameter 2.7 mm, 0° with 6.0 mm trocar, during the years 1999–2005; then a single-use, rigid endoscope (Medtronic Channel Neuroendoscope, ref 2233-002, Goleta, California, USA) until 2009; and thereafter the Storz rigid endoscope (Karl Storz GmbH & Co. KG, Tuttlingen, Germany). The operating sheath was inserted freehand and was never fixed during the procedure. The floor was perforated just behind the clivus, halfway between the infundibulum and the mammillary bodies in the midline, by the use of a thermal technique with a monopolar electrode, followed by dilatation with an inflated 2- or 4-French balloon catheter (7CB-D10; Integra NeuroSciences, Sophia Antipolis, France). The endoscope was threaded gently into the prepontine cistern through the stoma to confirm the perforation of Liliequist's membrane. From a technical point of view, the procedure was considered successful if Liliequist's membrane was opened. We did not leave extraventricular drainage after ETV, except in cases of significant bleeding during the procedure. Teo and Jones (43) noted that, in general, extraventricular drainage was not necessary after an ETV. In addition to its possible infectious complications, it could reduce the success rate of ETV by decreasing the pressure gradient in the ETV orifice (31).

Statistical Analysis

We performed Statistical analysis by using the Sigma Stat 3.5 software package (Systat Inc., Point Richmond, California, USA). Qualitative variables were compared using a χ^2 or Fisher's exact test, and variance analysis or nonparametric tests were used for continuous variables. The relationships between risk of ETV failure and the initial etiology of hydrocephalus, the presence of infected shunt material, the number of shunt failures before the ETV procedure and, finally, and the number of years the patient had been using a shunt before the ETV procedure were analyzed with logistic regression. Cumulative survival rates were estimated by the use of Kaplan-Meier methods. A P value < 0.05 was considered significant.

RESULTS

Patients' Characteristics

The study included 31 men (58%) and 22 women (42%). Fifty-one of the patients had VP shunts and 2 ventriculo-cardiac shunts. Table 1 shows the age distribution of the 53 patients included. The median age was 26 years (range, 2 months to 66 years, SD: 12 years). Twenty-one patients (40%) were younger than 18 years of age, and 5 (8%) were younger than the age of 2 years. The main initial etiology of hydrocephalus was aqueductal stenosis (19 cases). Other etiologies were hydrocephalus caused by posterior fossa or tectal plate tumors, infection, hemorrhage, or spina bifida (Table 2).

ETV was performed for the first shunt failure in 20 patients (38%) and for at least second shunt failure in 15 (28%); 18 patients (34%) had had more than 3 shunt failures before ETV. The mean duration between the placement of the shunt for initial hydrocephalus and the ETV procedure was more than 11 years (137 months, range: from 1 month to 34 years; SD: 124 months). The

Table 1. Age Distribution in the 53 Patients of This Study

No. Patients (%)	Age
3 (6%)	0–6 months
4 (8%)	6–24 months
14 (26%)	2–18 years old
32 (60%)	> 18 years old

Table 2. Causes of Initial Shunting and Number of Cases

Initial Reason for Shunting	No Cases (%)
Sylvius aqueductal stenosis	19 (36%)
Chiari II malformation	7 (13%)
CSF Infections	5 (9%)
Posterior fossa tumors	9 (17%)
Third ventricular, tectal plate tumors	11 (21%)
Other	2 (4%)
Total	53 (100%)

CSF, cerebrospinal fluid.

clinical signs of shunt failure are detailed in Table 3 and were mainly headaches (19 patients; 36%) and altered states of consciousness (12 patients; 23%).

The causes of shunt failure were mainly infected material (13 patients), obstruction of the catheter (13 patients) or disconnection (4 patients). No cause was found for shunt failure in the remaining 23 patients (43%). The mean length of hospitalization of all 53 patients was 9.60 days (from 2 days to 40 days; SD: 8.20).

ETV Outcome

The overall success rate was 70% (37/53 cases) with a mean follow-up of 51 months (from 3 to 157 months, SD: 41 months). This follow-up comprised a total of 1972 patient-months of observation. ETV success was defined as complete disappearance of the clinical signs and no placement of further VP shunt (Figure 1). No deaths occurred in this series. The mean length of hospitalization for patients with successful ETV was 9.35 days (range, 2–40 days), duration globally similar to that of patients with unsuccessful ETV (mean hospital stay: 10.12 days, range, 4–21 days).

Ventricular Size After ETV

Overall, all 53 patients had at least one pre- and postoperative imaging study available. In our 37 cases of successful ETV, ventricular size decreased in 21 patients and remained unchanged in 16 patients (43%) at the last brain imaging follow-up. In this study, the ventricular size was not associated with ETV outcome ($P = 0.43$).

Table 3. Shunt Failure: Clinical Signs in Patients of This Series (Some Patients Had More Than 1 Clinical Sign)

Clinical Signs	Number of Cases
Headaches	19
Altered state of consciousness	12
Nausea, vomiting	9
Signs of infection	9
Intracranial hypertension	6
Visual troubles	6
Balance, walking difficulties	3
Bulging fontanelle	3
Epilepsy	1

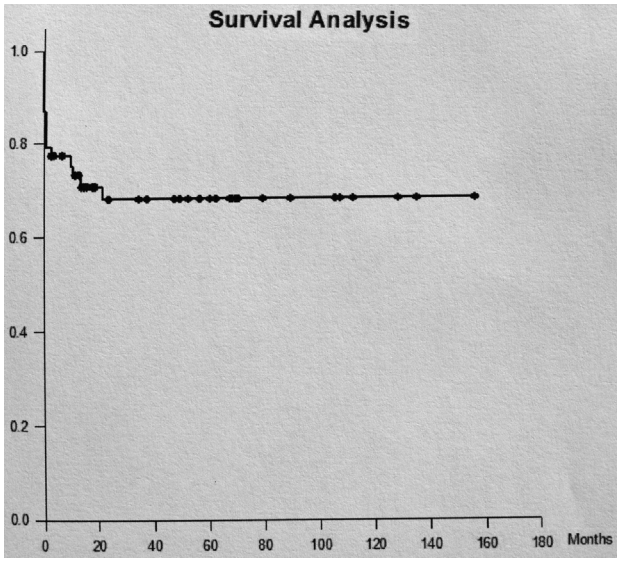

Figure 1. Endoscopic third ventriculostomy (ETV) success rate and follow-up in the 53 patients of the study. This figure shows that, in our group of patients, ETV failures were detected in 2 years of follow-up and mainly within a few days of the procedure.

Factors Associated with ETV Outcome

No significant difference ($P = 0.922$) in ETV outcome was detected in this series when we compared ETV success in adults (23 of 32) and in children younger than 18 years old (14 of 21), nor was there any significant difference ($P = 0.361$) regarding the success of the ETV procedure detected in children younger than 2 years old (5 successful ETV in the 7 performed) compared with other children (9 of 14 successful ETV). Nevertheless, the number of children in each group was small.

In this study, the success of the ETV procedure was not related to the cause of hydrocephalus ($P = 0.622$) or to the number of shunt failures before the ETV procedure ($P = 0.459$; Table 4). Regarding outcome of ETV, we also found no statistical difference ($P = 0.343$) in patients shunted for less than 5 years (16 ETV successful of 20 performed) compared with those shunted for more than 5 years (21 ETV successful of 33). Of the 13 patients with infected shunts, 11 were successfully treated by ETV. Compared with other ETV results, the presence of an infected shunt was not predictive of ETV failure ($P = 0.395$).

Shunt Removal

In our 37 patients with successful ETV, we tried to remove the shunt in 17 patients. The shunt was actually removed 15, but in 2 other patients, the proximal catheter was blocked. We decided to not remove it, fearing possible intraventricular hemorrhage. These 2 proximal catheters were stitched and left in position; only the distal catheter was removed. Other shunts were not removed because their

Table 4. ETV Success Rates According to the Number of Previous Shunt Failures

Previous Shunt Failures	Number of Cases, n (%)		ETV Success Rates, n (%)	
1	20	37.74%	15	75.00%
2	15	28.30%	11	73.33%
3	8	15.09%	6	75.00%
≥4	10	18.87%	5	50.00%
Total	53		37	69.81%

ETV, endoscopic third ventriculostomy.
ETV was less successful (50%) in patients who had had more than 4 shunt failures. However, compared with other results, this rate was not significantly different.

removal was judged risky (especially for shunts in place for a long time) or useless. In the remaining 20 patients, the shunt was checked 6 times (but not removed) and found not functioning. Other 14 patients did not have their shunt checked because we assumed that checking their shunt was useless. No complication related to the remaining shunt was noted in these patients.

ETV Procedure and Complications

In 3 children (1, 6, and 11 years) who had had multiple shunt revisions (3 or 4 shunt revisions), intraoperative difficulties were experienced during the ETV because of anatomic changes. One 6-year-old child had a third ventricle floor that was anatomically very modified, descending on to the lower part of the pons. Another had no mammillary bodies and the location of the basilar artery was modified. In these 2 cases, the ETV was finally performed and was successful (157 and 22 months of follow-up). In the last case, attempted ETV failed; there were membranes in the third ventricle and no anatomic landmarks were visible on its floor. The ETV procedure was stopped. This was the only case in this series of failed attempted ETV. Finally, there was one case of intraventricular bleeding after insertion of the endoscope in the third ventricle in a 6-year-old patient. An extraventricular drain was placed, which was removed 2 weeks later when a second ETV was performed with no problems. The patient had no clinical consequences of this hemorrhage. This ETV was finally successful (12 months of follow-up).

Postoperatively, we had 3 complications linked to the ETV procedure: a 42-year-old patient with meningitis (with no germ found) and a 26-year-old woman with a frontal abscess (staphylococcus aureus) on the ETV brain path. These 2 complications were treated successfully by antibiotics. Finally, a 38-year-old patient had a granuloma on the ETV skin scar. This granuloma was painful and was removed 2 months after the ETV procedure. In these 3 patients, the ETV procedure was successful at 157, 36, and 12 months of follow-up.

DISCUSSION

Between June 1999 and July 2013, we performed on 53 ETVs for shunt failures. The overall success rate was almost 70% (37 of 53) over a mean follow-up of more than 4 years (51 months). Very few complications were observed. The initial cause of hydrocephalus leading to shunt placement, the number of previous shunt failures, or the cause of shunt failure in this study did not significantly affect the ETV success rate. This study, involving one of the largest groups of patients of the literature on this topic, confirms the important role of ETV in cases of shunt failure.

CSF shunts are associated with many shunt malfunctions (27, 42). Patients with hydrocephalus can have several shunt failures and operations during their lives (35, 42). Several authors (7, 23, 33, 37) have noted high rates of shunt failures over the years, with 20% of the cases needing more than 3 revisions in 5 years. The risk of subsequent shunt failure may also increase with the number of revisions (29). With the advent of endoscopic techniques, ETV has become a procedure of choice for the treatment of obstructive hydrocephalus (2, 12, 26, 38). ETV also is considered as a good option for some cases of CSF infection (20), overdrainage (1, 36), and also shunt failures (11, 22).

Some authors (1, 4-6, 8, 10, 14, 16, 19, 32, 44) have pointed out that ETV can be effective in patients with shunt failure, with rates of success varying from 38 to 88%. Series including more than 50 patients are rare. Buxton et al. (8) studied 88 patients for shunt failure and ETV (including communicating hydrocephalus). Their overall success rate was 52% but was 73% in patients with noncommunicating hydrocephalus. O'Brien et al. (32), in a series of 63 patients, reported a success rate of 70%, similar to ours. Considering the relationships between initial hydrocephalus etiology and ETV prognosis, Bilginer et al. (4) studied 21 patients in the context of aqueduct stenosis and shunt failure. They reported an ETV success rate of 85.7% (18 of 21 patients). The result of our study on this specific etiology was similar, with a success rate close to 80% (15 of 19). Moreover, hydrocephalus resulting from aqueduct stenosis is generally well treated by first-line ETV (38). Bilginer et al. (4) performed ETV on patients (mean age 15.8 years) with tumors of the tectal plate and the pineal region who had been shunted previously. Their group of patients all had an optimal success rate (100%), a rate quite close to the 82% of our study, with 9 of 11 of patients shunt free. Curiously, in patients with myelomeningocele and Chiari II malformation, results seem to be better in patients who have an ETV for shunt failure (84% shunt free) than in patients treated in their infancy by first-line ETV (44). Bilginer et al. (4) suggested that the development of the subarachnoid space and of granulations of Pacchioni could promote the success of the ETV. In patients who had a shunt failure for intraventricular hemorrhage, other authors (10, 28, 32) also obtained rather high success rates (between 60% and 100% of cases) for ETV. Finally, we observed that infected shunt failures could also be managed by ETV (20, 40, 41). In our series, 13 patients had an infection of the shunt. Eleven of them were successfully treated by ETV with a mean follow-up of 63 months. In these cases, all shunts were removed after the ETV procedure. A CSF infection is not a contraindication to performing an ETV (8, 41) a result also found by Cinalli et al. (10) in 13 patients.

It is worth to remember that our results concerned selected patients with shunt failure. We excluded adult and pediatric patients with communicating hydrocephalus. The role of ETV in

communicating hydrocephalus is debated; we considered that ETV was not suited to treat shunt failures in the context of communicating hydrocephalus.

Despite the relative success of ETV in shunt malfunction, some complications can occur. As in our study, some authors have pointed out anatomical changes and thickening of the floor of the third ventricle (18, 41) in some previously shunted patients. Brockmeyer et al. (6) reported that ETV in shunted patients increased the risk of abandon of the procedure to avoid complications. Overall, the risks associated with the ETV procedure in patients having an initial shunt can vary from 0% to 13.3% depending on the authors (1, 5, 8, 11, 14, 32, 44). In their largest series of patients undergoing ETV for shunt failure (88 patients of all ages), Buxton et al. (8) reported a complication rate of only 5.6%, with one death. Among 63 patients, O'Brien et al. (32) had 2 patients (1.6%) who suffered from an infection and 6 from intraoperative bleeding (4.8%).

Should We Remove the Shunt After ETV?

Concerning the removal of the shunt after ETV, Boschert et al. (5) preferred to remove the shunt in all cases of their series. They pointed out the risk of infection attached to leaving the shunt. In contrast, in the study by Neils et al. (31) on the management of the shunt after ETV, the policy was not to remove the shunt. In our study, we did not have a systematic attitude. We completely removed the shunt in 16 cases only (13 cases of infection). In the other cases, we left the shunt after ETV, thus avoiding the risk of bleeding from a ventricular catheter located close to or in the choroid plexus. Some shunts also can be very calcified subcutaneously and difficult to remove.

In conclusion, the current study describes one of the largest series on this topic. The overall success rate in our study was almost 70% (37 of 53), which is consistent with the results in the literature (4, 32). In patients with shunt failure, ETV should be considered as the first therapeutic option, before shunt revision.

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