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# Long-term follow-up of endoscopic third ventriculostomy performed in the pediatric population

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**OBJECTIVE** Endoscopic third ventriculostomy (ETV) is an effective treatment for obstructive hydrocephalus and avoids the risk for foreign-body infection associated with ventriculoperitoneal (VP) shunts. The short-term failure rate of ETV strongly depends on the indications for its use but is generally thought to be lower in the long term than that of VP shunts. However, few studies are available with long-term follow-up data of ETV for hydrocephalus in children. The authors reviewed the long-term success of ETV at their institution to investigate the rate of any late failures of this procedure.

**METHODS** Between April 1998 and June 2006, 113 children (including neonates and children up to 16 years old) had primary or secondary ETV for different causes of hydrocephalus. The patients' medical records and the authors' electronic operation database were reviewed for evidence of additional surgery (i.e., repeat ETV or VP shunt insertion). These records were checked at both the pediatric and adult neurosurgical hospitals for those patients who had their care transferred to adult services.

**RESULTS** The median length of follow-up was 8.25 years (range 1 month to 16 years). Long-term follow-up data for 96 patients were available, 47 (49%) of whom had additional ETV or VP shunt insertion for ETV failure. Twenty patients (21%) had a second procedure within 1 month, 17 patients (18%) between 1 and 12 months, 7 patients (7%) between 1 and 5 years, and 3 patients (3%) between 5 and 8 years.

**CONCLUSIONS** In the authors' series, ETV had an initial early failure rate for the treatment of pediatric hydrocephalus as reported previously, and this rate significantly depended on patient age and hydrocephalus etiology. Once stabilized and effective, ETV appeared to be durable but not guaranteed, and some late decline in effectiveness was observed, with some ETV failures occurring many years later. Thus, successful ETV in children cannot be guaranteed for life, and some form of follow-up is recommended long term into adulthood.

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**KEY WORDS** hydrocephalus; ETV; long term; follow-up; CSF disorders

Over the last few decades, endoscopic third ventriculostomy (ETV) has become a common treatment for pediatric hydrocephalus. This development is due to improved safety of the procedure enabled through advances in MRI and endoscopic technology.

Endoscopic third ventriculostomy has a high rate of early failure, which is influenced largely by the indications used for the procedure and also by the technical abilities and experience of the surgeon. Many studies have reported short-term success (≤ 6 months) and intermediate-term success (≤ 3 years) of ETV, both of which depend on age, the etiology of the hydrocephalus, and the presence of a preexisting VP shunt. However, few data are available demonstrating longer-term success of ETV or indicating late surgical failure, if it indeed occurs at all.[1,3,5,9,10]

Ventriculoperitoneal (VP) shunting is the main alter-

**ABBREVIATIONS** ETV = endoscopic third ventriculostomy; ETVSS = ETV Success Score; VP = ventriculoperitoneal.

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native to ETV for treating pediatric hydrocephalus. Although the failure rate of VP shunts is lower than that of ETV in the short term,[8] VP shunts are associated with a steady failure rate that continues over a longer period.[7,11]

We present a single-institution, retrospective review of long-term ETV success in the treatment of pediatric hydrocephalus. Late failures (≥ 5 years) were rare and occurred less frequently than would be expected if the patients had been treated with a VP shunt. Nonetheless, the occurrence of these occasional failures warrants long-term follow-up of children with hydrocephalus treated successfully with an ETV.

## Methods

We re-reviewed the data from a cohort of patients who the senior author had previously included in a multicenter study that investigated the risk factors of ETV at 6 months.[10] We identified any late failures of ETV treatment that had required an additional ETV or VP shunt since the original follow-up date.

We reviewed patient notes, electronic operative logs, hospital attendance logs, and all imaging shared within our regional network of hospitals. We looked for evidence of additional surgery for hydrocephalus. For those patients who were older than 16 years by the time of this study, this review was repeated at our corresponding adult neurosciences center that serves a similar population.

A time-to-event analysis was performed with a commercial statistical software package (SPSS), and data were presented as a Kaplan-Meier survival plot. An event was considered a repeat ETV, new VP shunt, or VP shunt revision if a nonfunctioning shunt was already in situ. Those patients who did not require additional surgery were censored at the time of their last follow-up.

ETV operative technique is now fairly standard and has been reported in our previous investigations related to the present study.[4,6,10] A routine follow-up included an MRI examination 6 weeks after ETV. This follow-up was repeated only if symptoms of ETV failure developed. Clinical follow-up continued throughout childhood with annual examinations, followed by transition to adult services when the patient was 16 years old.

## Results

Between April 1998 and June 2006, 113 children had an ETV at the Royal Liverpool Children's Hospital, Alder Hey, under care of 4 neurosurgeons; 17 patients had no record of a follow-up and were therefore excluded from the analysis. The demographics of the remaining 96 patients are shown in Table 1. Both primary ETVs and secondary ETVs (after a VP shunt) were included.

The median length of follow-up of patients whose condition did not require further surgery was 8.25 years (range 1 month to 16 years). Twenty patients (21%) had additional surgery within the 1st month, 17 (18%) between 1 and 12 months, 7 (7%) between 1 and 5 years, and 3 (3%) after more than 5 years. The cumulative proportions of patients with revision-free survival are shown in Table 2; treatment success (i.e., no repeat surgery was needed) at 6 months was 61%. This corresponded to a predicted ETV Success Score (ETVSS)[9] of 62% (p = 0.88, chi-square test indicating no statistically significant difference between the predicted and observed success of ETV at 6 months). The revision-free survival data are presented as a Kaplan-Meier curve in Fig. 1. Their cases are described in the following.

**TABLE 1. Demographics and patient characteristics of 96 children undergoing primary or secondary ETVs between 1998 and 2006**

| Variable | No. of Patients (%) |
|---|---|
| Age when ETV performed | |
| <1 mo | 10 (10) |
| 1 to <6 mos | 21 (22) |
| 6 mos to <1 yr | 4 (4) |
| 1 to <10 yrs | 37 (39) |
| 10 to <16 yrs | 24 (25) |
| M/F ratio | 58:38 (60:40) |
| Etiology of hydrocephalus | |
| Aqueductal stenosis | 35 (36) |
| Tectal/pineal tumor | 8 (8) |
| Other tumor | 14 (15) |
| Intraventricular hemorrhage | 19 (20) |
| Chiari malformation/spina bifida | 13 (14) |
| Infection | 3 (3) |
| Other | 4 (4) |
| Type of ETV | |
| Primary | 63 (66) |
| Secondary | 33 (34) |

Delayed failure (≥ 5 years after ETV) was noted in 3 patients (3%). Patient 1 was a premature twin with an intraventricular hemorrhage who had had a primary VP shunt at age 4 months and an additional ETV when she was 5.5 months old. At age 8.5 years (8 years after the ETV), she developed headaches and unsteadiness. An MRI examination including CSF flow studies indicated flow through the ventriculostomy, but intracranial pressure monitoring revealed a raised intracranial pressure. She underwent a new VP shunt placement and did not require additional surgery.

Patient 2 was a term child with spina bifida and a Chiari malformation Type II. She had a nonfunctioning VP

**TABLE 2. Kaplan-Meier estimate of cumulative proportion 15-year revision-free survival after primary or secondary ETV***

| Time Point (yrs) | Revision-Free Survival |
|---|---|
| 0.5 | 61 (51–70) |
| 1 | 55 (45–65) |
| 3 | 47 (37–57) |
| 5 | 47 (37–57) |
| 10 | 42 (31–52) |
| 15 | 42 (31–52) |

* Data represent percentages of revision-free survival (median [95% CI]); a revision was a repeat ETV or VP shunt placement, and patients were censored at their last follow-up date.

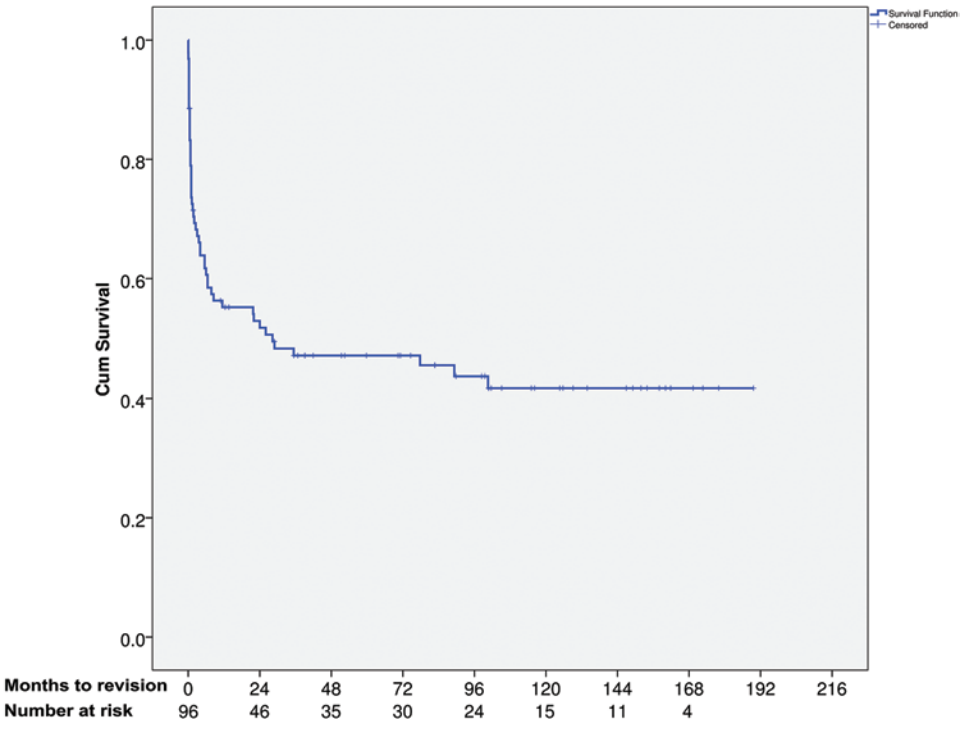


**FIG. 1.** Kaplan-Meier survival curve showing 15-year event-free survival after primary or secondary ETV. An event was a repeat ETV or VP shunt. Patients were censored at their last follow-up date. Number at risk denotes the number of patients remaining at risk at each time point (patients not yet censored or who have not had a repeat ETV or VP shunt). Cum = cumulative. Figure is available in color online only.

shunt in situ when an ETV was performed when she was 5.5 years old. She remained well until age 13 years (7.5 years after the ETV), when she developed headaches and unsteadiness. Another VP shunt was inserted, and she did not require any additional surgery.

Patient 3 was a 12-year-old boy who had an intraventricular hemorrhage resulting from premature birth and an entrapped fourth ventricle. He had a VP shunt in situ, which became blocked, so an ETV was performed. He presented with headaches and unsteadiness 6.5 years later. A revision ETV was performed successfully. During the procedure, thick membranes were noted that were divided anteriorly to the basilar artery.

## Discussion

### Short-Term Success

Short-term success (i.e., ≤ 6 months after the operation) of ETV has been described extensively in the literature and has been measured by Kulkarni and colleagues with the ETVSS.[1,3,9,10] Our cohort had a comparable but slightly lower revision-free survival rate than that reported in the literature[1,3] at 6 months (61%) and 3 years (47%). This lower rate was likely due to a high proportion of patients in our cohort both receiving secondary ETVs and having spina bifida. This is reflected in the 6-month predicted ETVSS of 62%. The similarity of our observed ETV success to that predicted by the ETVSS further validates the ETVSS tool.

### Long-Term Failures

Some authors have assumed that if an ETV has successfully controlled a patient's hydrocephalus for 5 years, the patient will not have to undergo additional surgery. Indeed, in a retrospective review of 119 children undergoing primary ETV for triventricular hydrocephalus, Cinalli et al. found no delayed failures beyond this 5-year period.[2]

The results of our study show that this was not the case in our cohort. As was expected, a small number of intermediate failures were observed between 1 and 5 years (affecting 7% of the patients), and a small number of late failures also occurred between 5 and 15 years (3% of the patients). The occurrence of late failures is supported by a retrospective review by Kadrian et al. of 203 patients undergoing both primary and secondary ETVs with a follow-up length of up to 25 years. These authors reported cases of ETV failure requiring surgery at 5–12 years postoperatively. However, unlike our cohort, their cohort included only patients with triventricular hydrocephalus and comprised both adults and children.[6] Similarly, a retrospective review by Vogel and colleagues of data from 100 adults and children undergoing primary and secondary ETVs for a variety of etiologies reported delayed failure requiring surgery up to 16 years after these ETVs.[12]

The 3 patients in our cohort who developed delayed failure (≥ 5 years) had all undergone secondary ETVs. They also presented with similar clinical symptoms including headaches and unsteadiness. Other than the similarities in these 2 symptoms, the 3 patients were not similar to each

other. The age at which they had an ETV ranged from 4 months to 12 years. Two patients had posthemorrhagic hydrocephalus, and the third patient had a Chiari malformation Type II and spina bifida. After review of MRI scans and CSF flow study results, it was noted that the ventriculostomy of Patient 1 was functioning, despite a raised intracranial pressure suggesting a failure of CSF absorption in the subarachnoid space. Patient 3 had macroscopic obstruction of the stoma with gliotic adhesions visible on revision endoscopy images. Patient 2 did not undergo MRI examination, CSF flow studies, or revision ETV, so it is unclear if her ventriculostomy remained patent at the time of clinical failure. Although few similarities could be found among these patients, their number was small, so any trends could not be determined.

### Comparison With VP Shunts

Although our findings demonstrated late failure of ETV for the treatment of hydrocephalus, it should be considered in the context of the alternative: VP shunting. Ventriculoperitoneal shunts are associated with a higher rate of infection than ETV but have lower immediate complication and failure rates.[5,7,11] While the failure rate of ETV fell significantly after 3 years postoperatively, VP shunts continued to fail at a steady rate. The revision-free survival of our cohort at 5 years was 47%, compared with a survival of 35% at 5 years in the Shunt Design Trial of 344 patients treated with primary VP shunts for pediatric hydrocephalus.[7] Similarly, a retrospective review of the records of 64 children treated with primary VP shunts and followed up for 18 years indicated a 5-year revision-free survival of 29%, with only 15% of the patients not having surgery for shunt failure during the study. The Kaplan-Meier plot of this group's survival to revision surgery has been reproduced in Fig. 2 with permission of the authors for comparison with the ETV results.[11]

## Limitations

As a retrospective review, our study has certain limitations. We attempted to limit selection bias by including every consecutive patient undergoing an ETV in the study selection period. A wide range of etiologies for hydrocephalus was included, and the sex and age at which ETV was performed were similar to those reported in other retrospective studies.[1,3] Both primary and secondary ETVs were included, but they were not analyzed separately, as the numbers of patients receiving these ETVs were too small. To avoid underestimating the rate of ETV failure, we censored follow-up duration at the last clinical encounter with sufficient information to ascertain whether further surgery had been performed. As a result, a proportion of patients (15%) with no recorded follow-up was excluded from the analysis.

## Conclusions

Consistent with previous reports by other authors, ETV for the treatment of pediatric hydrocephalus had an initial early failure rate in our series. After 3 years, successful outcomes of ETV seem to be durable but not guaranteed, with failures of ETV occurring as late as 8 years postoperatively. Delayed failure typically resulted in patients presenting with headaches and unsteadiness, but also occurred in patients with a variety of primary etiologies, varying patient age at the time of the ETV, and in the presence of an open or closed ventriculostomy detected on MRI or endoscopy examination on re-presentation. Thus, successful ETV in children cannot be guaranteed for life, and some form of long-term follow-up or patient education is recommended into adulthood.

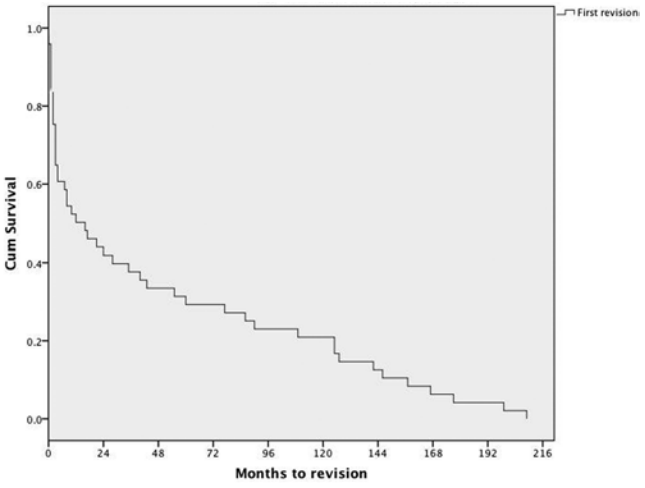


**FIG. 2.** Kaplan-Meier survival curve indicating 20-year cumulative revision-free survival after initial VP shunt placement. Reproduced with permission from Stone et al: *J Neurosurg Pediatr 11:*15–19, 2013.

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## Disclosures

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

## Author Contributions

Conception and design: Stovell, Mallucci. Acquisition of data: Stovell, Gallagher, Hayhurst. Analysis and interpretation of data: Stovell, Mallucci. Drafting the article: Stovell, Gallagher. Critically revising the article: Stovell, Zakaria, Ellenbogen, Jenkinson, Mallucci. Reviewed submitted version of manuscript: Stovell. Approved the final version of the manuscript on behalf of all authors: Stovell. Statistical analysis: Zakaria. Study supervision: Jenkinson, Mallucci.

## Supplemental Information

### Previous Presentations

Portions of this work were presented in abstract form for an oral presentation at the Sixth Meeting of the International Society for Hydrocephalus and CSF Disorders, Bristol, UK, September 6–8, 2014.

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