

Results of endoscopic third ventriculostomy

Mark R. Iantosca, MD^{a,*}, Walter J. Hader, MD, FRCS(C)^b,
James M. Drake, FRCS(C)^c

^a*Connecticut Children's Medical Center, 100 Retreat Avenue, Suite 705,
Hartford, CT 06106–2565, USA*

^b*Division of Neurosurgery, University of Calgary, Alberta Children's Hospital,
Calgary, Alberta, Canada*

^c*Hospital for Sick Children, 555 University Avenue, Toronto,
Ontario M5G 1X8 Canada*

The recent resurgence of interest in ventriculostomy has arisen out of dissatisfaction with the complications and long-term outcomes of conventional cerebrospinal fluid (CSF) shunting systems [1,2]. Initial attempts at ventriculostomy via open craniotomy and subsequent percutaneous fluoroscopic and CT-guided techniques have largely been replaced by modern endoscopic procedures with reduced morbidity [3,4]. Unfortunately, sufficient long-term follow-up data necessary for direct comparison with outcomes of conventional shunting (CS) procedures are currently unavailable. Clinical decisions regarding use of endoscopic third ventriculostomy (ETV) are based on the results of studies with mean follow-up intervals of less than 5 years. Results of ETV are most closely associated with the etiology of hydrocephalus encountered as well as with the clinical and radiographic features of the individual patient.

Factors affecting outcome

Hydrocephalus etiology

Third ventriculostomy is designed to treat noncommunicating hydrocephalus with patent subarachnoid spaces and adequate CSF absorption. It is not surprising, therefore, that the results

of this procedure are most strongly influenced by the etiology of hydrocephalus. **Box 1** depicts hydrocephalus etiologies divided according to reported success rates of ETV. Patients with acquired aqueductal stenosis or tumors obstructing third ventricular outflow have demonstrated the highest success rates, exceeding 75% in carefully selected series of patients [4–10]. Previously shunted patients with or without myelomeningocele, tumors, or cystic abnormalities leading to fourth ventricular outflow obstruction (ie, arachnoid cyst, Dandy-Walker malformations) and patients with congenital aqueductal stenosis have shown an intermediate response [6,7,9,11–13]. Further study of this intermediate group is likely to identify subgroups with higher success rates. Infants developing hydrocephalus after hemorrhage or infection or with associated myelomeningocele (without prior shunting procedure) have demonstrated a poor response to ventriculostomy [3,5,13–16]; despite limited reports of success in such patients [7,11,12,17–20], they are more controversial candidates for this procedure. The procedure is not advisable in patients who have undergone prior radiation therapy because of the extremely poor response rates, altered anatomy (ie, thickened third ventricular floor), and increased risk of bleeding [3,7,11].

Aqueductal obstruction by stenosis or tumor

Blockage to CSF flow at the level of the cerebral aqueduct by anatomic or postinflammatory septations/membranes or by extrinsic compression

* Corresponding author.

E-mail address: miantos@ccmckids.org
(M.R. Iantosca).

Box 1. Endoscopic third ventriculostomy success rates by hydrocephalus etiology

High success rates (>75%)

Acquired aqueductal stenosis
Tumor, cyst, or infectious lesion obstructing third ventricular outflow
Tectal, pineal, thalamic, or intraventricular tumor
Arachnoid cyst
Toxoplasmosis
Shunt malfunction in patient with obstructive etiology

Intermediate success rates (approximately 50%)

Tumor obstructing fourth ventricular outflow
Myelomeningocele (previously shunted, older patients)
Congenital aqueductal stenosis
“Cystic” abnormalities obstructing fourth ventricular outflow
Arachnoid cysts
Dandy-Walker malformation
Previously shunted patients with “difficulties”
Slit ventricle syndrome
Recurrent or intractable shunt infections
Recurrent or intractable shunt malfunctions

Low success rates (<50%)

Myelomeningocele (previously unshunted neonatal patients)
Posthemorrhagic hydrocephalus
Postinfectious hydrocephalus (excluding aqueductal stenosis of infectious etiology)

secondary to tumor, benign cyst, or vascular lesions in a surrounding structure (tectum, pineal, posterior thalamus, and superior cerebellum) is currently the best-studied and most successful indication for ETV. Success rates generally exceed 75% for this select group of patients [4–10]. In a recent large clinical series, including a wide age range of patients (3 weeks to 77 years, $n = 95$) Hopf et al [9] reported an ETV success rate greater than 80% for postinflammatory and idiopathic aqueductal stenosis and a 95% success rate for aqueductal stenosis caused by benign

space-occupying lesions. ETV was less successful in patients with progressive neoplastic lesions, primarily posterior fossa tumors and posthemorrhagic (intraventricular or subarachnoid) cases (64% and 63%, respectively) [9]. Similar results have recently been reported in a series analyzing 213 children with similar etiologies by Cinalli et al [4] encompassing stereotactic ventriculostomy and ETV. The less favorable outcomes associated with posthemorrhagic and posterior fossa tumors and cysts likely reflect the multifactorial causes of hydrocephalus in these patients [4,9]. Studies of ETV for other hydrocephalic etiologies are limited to smaller series and require further investigation to establish overall success rates.

Clinical features

Box 2 depicts preoperative clinical and radiographic criteria frequently cited for improving outcome and limiting morbidity of ETV. A recent study by Gangemi et al [10] offers the only detailed assessment of the contribution of presenting symptoms and signs to procedure success. Clinical presentations indicative of elevated intracranial pressure (ICP) were associated with the highest rate of ETV success (83%), followed by Parinaud’s syndrome (77%) and macrocephaly (71%). Other presentations (memory disturbances, incontinence, and focal deficits with associated tumors) exhibited success rates of approximately 50% or less. Patient age and history of prior shunting procedure are the most frequently cited clinical features associated with outcome.

Age

There seems to be a significant association between increasing patient age and a more favorable outcome of ETV in multiple pediatric studies [6–9,11,12,15,16,21]. These studies frequently implicate a decreased pressure gradient across the arachnoid granulations (secondary to open sutures) or age-related immaturity of patient CSF absorptive capacity as reasons for this phenomenon. Evidence suggests that reduced success rates are associated with the age at which hydrocephalus initially developed as well as with the age at the time of ventriculostomy [7,21]. The series by Jones et al [21] reports that only 8 of 25 ventriculostomies were successful in patients less than 6 months of age, with 8 of 17 successful procedures in patients developing hydrocephalus

Box 2. Favorable clinical and radiographic features for endoscopic third ventriculostomy*Clinical*

Etiology of hydrocephalus in high or intermediate success group (see Box 1.)

Age greater than 6 months at time of hydrocephalus diagnosis

Age greater than 1 year at time of procedure (controversial)

No prior radiation therapy

No history of hemorrhage or meningitis

Previously shunted, particularly for high success group etiology (see Box 1.)

Radiographic

Clear evidence of ventricular noncommunication

Obstructive pattern of HCPQ

Aqueductal anatomic obstruction

Absence of aqueductal flow on T2-weighted MRI or cine study

Favorable third ventricular anatomy

Width and foramen of Monro sufficient to accommodate endoscope

Rigid > 6 mm

Flexible > 4 mm

Thinned floor of third ventricle

Downward bulging floor draped over clivus

Basilar posterior to mamillary bodies

Absence of structural anomalies impeding procedure

Arteriovenous malformation (AVM) or tumor obscuring third ventricular floor

Enlarged massa intermedia

Insufficient space between mamillary bodies/basilar and clivus

Basilar artery ectasia

before this age. Most of the successes in these groups were in previously shunted patients [21]. Several studies show success rates at or below 50% in patients less than 2 [6,7,9] or 1 [8,15,16] year of age, regardless of etiology. Results are even poorer in patients less than 6 months of age [12,21]. Teo and Jones [12] demonstrated statistically significant ($P = 0.005$) lower success rates in patients less than 6 months of age versus older patients (12.5% vs. 80%). Open third ventriculostomy has also been shown to have a dramatically decreased success rate in patients less than 6 months of age [18,22]. The results of these studies are seemingly contradicted by the recent report of Cinalli et al [4], who demonstrated no difference in failure rates of ETV between patient groups older or younger than 6 months, concluding that patient age should no longer be considered a contraindication to the procedure.

Despite the lower success rates and increased morbidity that they report in this younger age group, several authors still advocate an initial attempt at ventriculostomy in these patients so as

to avoid the lifelong complications of shunting [8,15,16]. These authors argue that the “successes” are spared the long-term morbidity, mortality, and economic burden of repeated evaluations and hospitalizations for shunt evaluation. Barlow and Ching [23] recently published an analysis of anticipated cost reduction comparing ETV and CS for treatment of obstructive hydrocephalus at their institution, estimating a reduction of 74 hospital days and nine surgeries annually. This study, however, assumes a success rate of 80%, with no subsequent admissions or procedures for complications or failures among the successful procedures. Insufficient long-term data currently exist to support this argument in younger patient groups, particularly given the often-cited reduced success rates and increased rates of complications.

ETV in this younger age group has also been advocated as a primary treatment to remove intraventricular hemorrhage or purulent CSF, assuming that this will result in a more suitable environment for shunt insertion [15,24,25]. Comparison of this procedure with standard

“temporizing measures,” such as ventriculostomy, ommaya placement, and ventriculosubgaleal shunting is needed to determine comparative efficacy and safety.

Interestingly, a recent report by Tisell et al [26] demonstrates an increased failure rate of ETV in older adults with aqueductal stenosis. These delayed failures were hypothetically attributed to decreasing CSF absorptive capacity as a result of age-related loss of arachnoid villi and decreasing resistance to transependymal CSF resorption over time. Only 50% of ETVs were successful in this series, with almost all failures occurring after an initially favorable response lasting between 1 month and 1 year [26]. An earlier report by Kelly [5] demonstrated a 94% success rate in a population of adults with the same etiology treated with stereotactic CT-guided ventriculostomy. Interestingly, most patients in Kelly’s series were previously shunted (11 of 16), whereas only 3 of Tisell et al’s 18 patients had existing shunts. Clearly, further delineation of the pathophysiology of age-related changes in CSF absorption, and development of studies to quantify this capacity, would greatly benefit clinicians attempting to identify appropriate patients for ETV.

Previous shunting

Multiple studies have demonstrated a trend toward more successful ventriculostomy outcome in patients with existing shunt systems [6,12,21]. Teo and Jones [12] demonstrated a statistically significant difference in success rates for previously shunted patients with myelomeningocele (84%) versus those never shunted (29%; $P = 0.002$). In fact, third ventriculostomy has been found to be useful in the treatment of intractable shunt infections and malfunctions and even slit ventricle syndrome refractory to other treatments [5,11,24,27–30]. Other series of ventriculostomy in previously shunted patients have been less promising [13,31]. These contradictory results may reflect the mix of patients in small series. High success rates in patients with aqueductal obstructive etiologies present a strong argument in favor of ETV for patients in this group who present with shunt malfunction. Improved outcome in previously shunted patients with other etiologies is commonly attributed to increased CSF absorptive capacity; however, the effect of the shunt itself on CSF absorption is difficult to distinguish from the effect of increased age in these patients.

Preoperative assessment of CSF absorptive capacity has been advocated by some authors [32,33], but these techniques have not been widely accepted [7,21]. Because the patency of CSF pathways is one of the assumptions on which the procedure is based, a preoperative CSF absorption study would be invaluable in identifying patients who would likely benefit from ventriculostomy. The observed delay between operation and decrease in ventricular size (see section on assessing outcome) suggests that CSF absorption may increase slowly after ventriculostomy, however. Therefore, preoperative assessment of CSF absorptive capacity may fail to identify all appropriate patients [7]. Until an accurate functional predictor of this capacity is practical, radiographic assessment of the obstructive pattern of hydrocephalus by MRI is likely to remain the preoperative diagnostic procedure of choice.

Anatomy

Preoperative MRI optimally demonstrates all relevant anatomic features and should be obtained for all proposed third ventriculostomy patients. Initially, confirmation of noncommunicating hydrocephalus of favorable etiology should be established by the pattern of ventricular dilatation. Anatomic obstruction of CSF pathways between the aqueduct and the fourth ventricular outflow foramina may be visible on T2- or T1-weighted contrast images. Additionally, T2-weighted and cine images should reveal no evidence of the normally present aqueductal CSF flow pattern.

Once the patient’s suitability for the procedure has been established, the neurosurgeon must clarify the details of third ventricular anatomy that are likely to affect morbidity. First, the width of the third ventricle and diameter of the foramen of Monro must be sufficient to accommodate the endoscope of choice. Additionally, the thickness of the third ventricular floor and anatomy of the proposed puncture site must be assessed in relation to vital structures, particularly the basilar artery and its branches. A downward bulging third ventricular floor draped over the clivus has been cited as a prerequisite for success of this procedure in the past, but others have not found this to be necessary [3,7]. Ultimately, the surgeon must be satisfied that there is no structural lesion (ie, tumor, AVM) or anatomic variation that would render the procedure unduly difficult or

hazardous. In cases of doubt, it is reasonable to visualize the floor of the third ventricle and to abandon the procedure if the floor is unsuitable. Reported procedure abort rates as a result of unfavorable anatomy or intraoperative bleeding range from 0% to 26% (Table 1).

Assessing outcome

ETV has yielded a higher success rate with lower morbidity and mortality than earlier methods of third ventriculostomy. Mortality rates for open ventriculostomy procedures varied between 5% and 27%, with success rates from 37% to 75% [3,5,18,31,34,35]. Percutaneous radiographic and, later, CT-guided techniques reduced this mortality rate to 2% to 7%, with a 44% to 75% rate of shunt independence [3,5,14,19,31,36]. Most recent studies using modern endoscopic techniques and equipment, with or without stereotactic CT or MRI guidance, have reported low morbidity (3%–12%) and extremely rare mortality, with success rates greater than 75% for carefully selected patient groups. Table 1 depicts the results of recent ETV studies with success rates by etiology where these data are available. The current challenge is to define appropriate indications and devise objective measures for pre- and postoperative assessment of these patients.

The goal of ETV and, to date, the best objectively quantifiable measure of a successful outcome is shunt independence. Vague outcome measures, such as “successful,” “improved,” or “favorable,” are used in a number of studies [12,21]. Only two reports have critically compared modern ETV techniques with CS [37,38]. Sainte-Rose [38] reported no statistically significant differences in measures of neurologic, endocrinologic, social, or behavioral outcomes in a group of 68 patients treated for aqueductal stenosis with either ETV ($n = 30$) or CS ($n = 38$). Tuli et al [37] demonstrated no difference in failure rate between ETV ($n = 32$, 44% failure rate) and CS ($n = 210$, 45% failure rate) in a prospectively followed group of patients with aqueductal stenosis or tumor. The long-term outcomes of CS, including multiple procedures for obstruction, infection, and overdrainage, have been well documented. Failure rates range from 30% to 47% over 1 year to 63% to 70% over 10 years [39–42]. Additional long-term studies of ETV and analytic comparison to established CS outcomes are necessary to clarify clinical decisions in these patients.

One of the most confusing aspects of outcome evaluation in ETV patients is the failure of the ventricles to return to normal size. Many reports detail a gradual decrease in the ventricular size over months to years after surgery, with resolution of periventricular edema and increased extracerebral spaces coinciding with clinical improvement [5–8,31,38]. One series of patients treated with either CSF shunts or ETV showed no difference in intellectual outcome despite enlarged ventricles in the ETV group [38]. Radiographic evaluation of suspected closure has been confounded by the presence of persistently enlarged ventricles.

Studies detailing the serial measurements of multiple radiographic indices of ventricular size after surgery show that the third ventricular size responds more quickly (usually within 3 months) than the lateral ventricular size (up to 2 years) [43,44]. Additionally, third ventricular size seems to correlate most closely with outcome in these patients [8,43,44]. A recent study limited to adult patients showed no relation between ventricular size reduction and outcome [26]. Kulkarni et al [45] have demonstrated a greater reduction in ventricular size after successful procedures versus treatment failures in 29 children.

MRI and Doppler ultrasound studies documenting CSF flow through a patent ventriculostomy are currently the most useful and widely used postoperative radiographic indices. MRI detection of T2-weighted flow void around the ventriculostomy has been correlated with clinical outcome in ETV [46–48]. This observation has proven most helpful in confirming ventriculostomy patency after surgery, particularly in patients with persistent ventriculomegaly [47,48]. Kulkarni et al [45] have recently demonstrated a statistically significant relation between evidence of postoperative aqueductal CSF flow on MRI and clinical success. Multiple studies have reported long-term patency of third ventriculostomy by means of cine phase contrast (PC) MRI studies. A report by Cinalli et al [4] included 15 patients with confirmed long-term (>10 years) patency of their ventriculostomy on cine PC MRI (median = 14.3 years, range: 10–17 years). Unfortunately, several recent studies in adults and children have demonstrated a lack of correlation between postoperative MRI findings and outcome [15,16,26,37]. Quantification of flow velocity through ventriculostomies has also been achieved by PC MRI and Doppler ultrasound [49,50]. Hopefully, methods of quantifying ventriculostomy function will allow neurosurgeons to refine

Table 1
Outcome of endoscopic third ventriculostomy by etiology

Author/Year	Age mean (range)	Morbidity	Procedure abort rate	Follow-up mean (range)	Success rate (%) by etiology (n)			
					Tumor/AS	Dysraphic	PHH/PIH	Other (n-etiology)
Jones et al / 1990 [11]	(4 M–17 Y)	8%	16%	N/R	59% (17)	40% (5)		
Jones et al / 1992 [6]	(PNB–78 Y)	7%	9%	27 M (3 M–7 Y)	68% (19)	40% (10)	0% (2)	
Jones et al / 1994 [7]	N/R	5%	6%	N/R	80% (25)	52% (21)		
Sainte-Rose and Chumass / 1996 [8]	5.3 Y	N/R	N/R	1.8 Y	81% (82)			
Teo and Jones/ 1996 [12]	11 Y (1 W–32 Y)	3%	9%	32 M (1 μ –17 Y)		72% (69)		
Goumnerova and Frim / 1997 [47]	11.2 Y (2 D–36 Y)	9%	N/R	17 M (7 μ –44 M)	70% (20)		0% (1)	100% (2-IDO)
Baskin et al / 1998 [30]	17.3 Y (1.5–49 Y)	12%	N/R	(21.4 M)				64% (22-SVS)
Brockmeyer et al / 1998 [13]	8.1 Y (1 D–29.5Y)	6%	26%	24.2M (15 μ –69 M)	59% (34)	50% (16)	0% (7)	
Buxton et al / 1998 [16]	3.7 M (0 M–10 M)	15%	0.4%	N/R (6 μ –42 M)	57% (7)	0% (2)	10% (10)	20% (5-IDO)
Buxton et al / 1998 [15]	2 M (0–11 M)* All PNB	21%	11%	(6 μ –42 M)	43% (7)		30% (10)	
Tuli et al / 1998 [37]	8.1 Y (0 M–18 Y)	N/R	N/R	N/R (1 Y–11 Y)	66% (32)			
Gangemi et al / 1999 [10]	31 Y (7 D–81 Y)	12%		28 M (12 μ –54 M)	91% (110)			
Cinalli et al / 1999 [4]	NR (1 M–18 Y)	N/R	6%	2.1 Y (4 D–9 Y)	86% (119)			
Hopf et al / 1999 [9]	36 Y (3 W–77 Y)	6%	2%	26 M (3 μ –71 μ)	83% (82)		63% (8)	
Tisell et al / 2000 [26]	48 Y (17–80 Y)	N/R	N/R	37 M (3 M–5 Y)	50% (18)			

Abbreviations: AS aqueductal stenosis; D, days; IDO, idivpathie hydrocephalus; m, months; N/R, not reported; **MMC**, myelomeningocele, **PHH** post hemorrhagic hydrocephalus; **PIH** post infectious hydrocephalus; **PNB**, Premature newborn, **SC**; shunt complications (intractable shunt infection/malfunction); **SVS**, slit-ventricle syndrome; **W**, weeks; **Y**, year's.

* All patients born prematurely: mean age at birth was 31.9 weeks (range: 26–36 weeks).

the techniques necessary to improve outcome further. Radiographic confirmation may also help to define indications with more subjective outcomes, for example, the observation of less fulminant shunt malfunctions in patients after ventriculostomy [11].

The authors use postoperative MRI imaging with sagittal T2-weighted images or cine MRI to confirm ventriculostomy flow and assess ventricular size 3 to 6 months after surgery. The absence of a flow void, although occasionally present in patients with good clinical outcome, was present in 12 of 13 patients presenting with delayed treatment failure in the series by Cinalli et al [4]. Increasing head circumference, signs and symptoms of elevated ICP, or evidence of CSF egress at the ventriculostomy incision site generally heralds early failures. Because of the risk of delayed failure of ETV, and the potential mortality if this is unrecognized, we recommend that these patients use medical alert bracelets or wallet identification cards. A low threshold of suspicion for reimaging these patients should be followed, especially for the first 5 years after ETV. Families and caregivers are given the same education as for patients with shunts regarding symptoms of failure. Education and regular follow-up are important to counteract the false sense of security that may arise in the absence of a shunt.

Failure of endoscopic third ventriculostomy

Studies of ETV are currently limited to studies with an average of less than 5 years of follow-up. These studies have documented early (<6 months) and late (>1 year) postoperative failures. Early failures are more frequently reported in patients less than 6 months of age [4,15,16]. Buxton et al [15,16] have reported a mean time to failure in patients younger than 1 year of age of 1.36 months. Multiple authors have reported “late failures,” where the ventriculostomy closes, sometimes years after surgery [4,7,9,24,26]. Obstruction of the ventriculostomy by a gliotic scar or arachnoid membranes has been described [31,51]. Rare cases of sudden death after late failure of ETV have been reported [52].

Although the long-term results of ETV are largely unknown, studies that include long-term follow-up on stereotactic ventriculostomy cases are likely applicable to ETV. Cinalli et al’s recent report [4] included 94 cases of stereotactic ventriculostomy with an average of 6.32 years of follow-up (range: 20 days to 17.4 years) and 119

case of ETV with an average of 2.1 years of follow-up (range: 4 days to 9 years). The overall functional ventriculostomy rate for the study was 72% at 6 years, without significant differences in the long-term results between the stereotactic and endoscopic groups [4]. Cine PC MRI studies on 15 patients up to 17 years after stereotactic ventriculostomy confirmed long-term patency (range: 10–17 years, median = 14.3 years) [4]. No failures were reported in either group after 5 years of follow-up. Additionally, 7 of 9 patients failing ventriculostomy with radiographically proven obstruction of the stoma had successful repeat ventriculostomies. Long-term data on ETV outcomes are only now beginning to emerge, and comparison to known outcomes of standard shunting procedures will be instrumental in defining the indications for initial treatment with ETV and the management of treatment failures.

Summary

ETV is emerging as the treatment of choice for aqueductal stenosis caused by anatomic, inflammatory, and selected neoplastic etiologies. The technique has also proven useful in the pathologic diagnosis and treatment of these conditions [53–56]. Long-term results of this procedure and comparison to standard shunting procedures are necessary to define indications for patients with pathologic findings in the intermediate response groups. Development of new studies for preoperative assessment of CSF absorptive capacity and quantitative postoperative measures of ventriculostomy function would be invaluable additions to our ability to assess candidates for this procedure and their eventual outcome. Further study and technical refinements will, no doubt, lead to many more potential uses for these procedures in the treatment of hydrocephalus and its associated etiologies. The challenge for neurosurgeons will be to define the operative indications and outcomes, while refining techniques for safely performing these useful procedures.

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