Endoscopic third ventriculostomy for tumor-related hydrocephalus in a pediatric population

PULAK RAY, M.D., GEORGE I. JALLO, M.D., R. Y. H. KIM, M.P.H., BONG-SOO KIM, M.D., SEAN WILSON, M.D., KARL KOTHBAUER, M.D., AND RICK ABBOTT, M.D.

Division of Pediatric Neurosurgery, The Johns Hopkins Hospital, Baltimore, Maryland; Department of Neurosurgery, Temple University Hospital, Philadelphia, Pennsylvania; Division of Neurosurgery, Kantonsspital Lucern, Switzerland; and Department of Neurosurgery, Montefiore Medical Center, New York, New York

Object. Endoscopic third ventriculostomy (ETV) has become a common alternative for managing hydrocephalus in select patients. Nevertheless, there is still controversy regarding the indications for ETV as the primary procedure, given its variable success rates. The purpose of this study is to review the authors’ experience with ETV for a variety of patients.

Methods. A total of 43 children underwent ETV between July 1992 and June 2003. Their medical records, operative reports, and imaging studies, when available, were retrospectively reviewed with regard to outcome, complications, and patency rate. Treatment failure was defined as the need to place a shunt within 4 weeks of performing ETV in the patient.

There were 20 male and 23 female patients with a mean age of 9.6 years (range 8 weeks–21 years). The overall success rate was 69.8%, and the mean follow-up duration was 24.6 months. Six patients underwent eight repeated ETVs at a mean interval of 25 months, with a patency rate of 62.5% after the second procedure. Only two surgeries were aborted for anatomical reasons. The highest success rates (100% in each instance) were achieved for obstructive hydrocephalus resulting from midbrain/tectal tumor (four patients) and pineal tumor (three patients).

Conclusions. The ETV procedure is an effective management tool for obstructive hydrocephalus in children. It should be considered the primary procedure, rather than ventriculoperitoneal shunts, in carefully selected children. The success rate is dependent on the origin of the hydrocephalus.

KEY WORDS • endoscope • third ventriculostomy • hydrocephalus • outcome

Endoscopic third ventriculostomy is now a popular alternative to ventricular shunt placement for the treatment of obstructive hydrocephalus in children, because there have been significant advances in this field, such as the use of endoscopes, fiberoptic devices, and other instruments.1–3,10 In particular, ETV has become the procedure of choice for the management of triventricular hydrocephalus from aqueductal stenosis at most centers because this procedure provides a more physiological and anatomical diversion pathway for CSF. It obviates the needs to place a foreign body such as a ventricular shunt, thus avoiding shunt-related complications such as malfunction, infection, and overdrainage. Several authors have recommended ETV for the treatment of hydrocephalus arising from various causes such as brain tumors, myelomeningoceles, meningitis, hemorrhage, and infection. Nevertheless, the outcome of ETV was reported to depend on the origin of each case of hydrocephalus and the institution in which it was treated. In this study we evaluate the outcomes and efficacy of ETV in our population of children with intracranial tumors.

Abbreviations used in this paper: CSF = cerebrospinal fluid; ETV = endoscopic third ventriculostomy; ICP = intracranial pressure; MR = magnetic resonance; VP = ventriculoperitoneal.

CLINICAL MATERIAL AND METHODS

Patient Population

A total of 43 patients underwent ETV for hydrocephalus related to tumors during an 11-year period between July 1992 and June 2003 at our institution. Of these patients, 20 were male and 23 were female, and their total mean age was 9.6 years (median age 9.8 years). The medical records, operative reports, and imaging studies, when available, were retrospectively reviewed to evaluate the cause of hydrocephalus, the patient’s age at the time of the procedure, sex, number of previous ventricular shunts, number of ETVs, outcome of ETV, complications related to ETV, and time to follow-up evaluation.

Success was defined as shunt independence for at least 1 month until the last follow-up evaluation. The follow-up evaluation consisted of clinical examination and neuroimaging studies including MR imaging and/or computerized tomography scanning. If a patient exhibited worsening clinical symptoms and/or signs during the follow-up period, an MR imaging with CSF flow study was performed to verify patency of the ventriculostomy. If the stoma of the ventriculostomy was confirmed as closed, a repeated ETV.
was performed. In cases in which the CSF flow study showed a patent third ventriculostomy, however, a VP shunt was inserted.

**RESULTS**

Fifty ETVs were attempted in 43 patients (Table 1). Twenty-seven of the 43 patients were shunt independent at the time of last follow-up evaluation (mean 24.6 months). Thirteen of them required VP shunt placement following ETV. Two procedures were abandoned because of anatomical distortion. Both of these patients had obstructive hydrocephalus arising from a thalamic tumor.

Of the 32 patients who underwent a successful ETV, 12 had VP shunt placement prior to undergoing ETV (Table 2). The shunt was usually externalized 1 day before ETV and the height of the drain was progressively elevated with or without ICP monitoring to increase the size of the ventricles. The shunt was usually removed at the time of the ETV procedure. An external ventricular catheter was placed if patients were experiencing moderate to severe symptoms or a significant increase in ICP the day before the procedure.

Outcomes were compared in patients who had undergone previous VP shunt insertion and in those who had not, regardless of tumor location. Seven of 12 patients who had previously received a VP shunt achieved long-term successful outcomes, compared with 21 of 31 patients who had not undergone this treatment previously. Although patients who had undergone a previous shunt placement had lower long-term success rates after ETVs compared with those who had no history of shunt treatment (58% compared with 68%), the difference was not statistically significant (p > 0.05).

There were eight repeated ETVs in six patients (Table 3). This included three patients with posterior fossa tumors, two with midbrain/tectal tumors, and one with a third ventricular tumor. The mean interval to repeated ETV was 25 months (range 2.3–48 months). Among the six patients who underwent repeated ETV, all were shunt independent at the last follow-up visit. An MR imaging/CSF flow study was performed in these patients before repeating the ETV.

**Follow-Up Evaluation**

Clinical follow-up data were available to evaluate outcome in all 43 patients. The follow-up period ranged from 3 to 78 months (mean 24.6 months). The overall success rate for patients with brain tumors was 69.8%. All patients in whom treatment failed presented with symptoms and signs related to increased ICP. Of the 13 patients who needed a ventricular shunt after ETV, five required the device within 7 days after undergoing ETV. The median interval to placement of a ventricular shunt was 29.5 days (range 3–187 days).

**Procedure-Related Complications**

For the 43 ETVs performed, there were no deaths related to the procedure. Nine patients (20.9%) in this series had intra- and/or postoperative complications (Table 4). Three cases of intraoperative venous bleeding were controlled by continuous irrigation during surgery. Two patients experienced a temporary decrease in their level of consciousness with no pathological finding on postoperative imaging studies. Other complications that the authors observed in this series included temporary diabetes insipidus, infection, and temporary limitation of ocular palsy.

**DISCUSSION**

Authors of several clinical studies have described the effectiveness of ETV in select patients with obstructive hydrocephalus. The ETV was initially considered to be an alternative procedure to ventricular shunt insertion for triventricular hydrocephalus arising from aqueductal stenosis. It has also been recommended as the initial management for hydrocephalus from other causes. Nevertheless, despite these optimistic reports, the outcomes for ETV vary from 23 to 100% success rates, depending on the institution, definition of failure, origin of hydrocephalus, indication, and follow-up period.

**TABLE 1**

<table>
<thead>
<tr>
<th>Tumor Location</th>
<th>No. of Patients</th>
<th>% Success</th>
<th>No. of Txs Failed</th>
<th>No. of Txs Abandoned</th>
</tr>
</thead>
<tbody>
<tr>
<td>midbrain</td>
<td>4</td>
<td>100</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>posterior fossa</td>
<td>9</td>
<td>44</td>
<td>5</td>
<td>0</td>
</tr>
<tr>
<td>brainstem</td>
<td>14</td>
<td>79</td>
<td>3</td>
<td>0</td>
</tr>
<tr>
<td>thalamus</td>
<td>6</td>
<td>67</td>
<td>0</td>
<td>2</td>
</tr>
<tr>
<td>pineal</td>
<td>3</td>
<td>100</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>3rd ventricle</td>
<td>2</td>
<td>100</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>(incl arachnoid cyst)</td>
<td>1</td>
<td>100</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>other (diffuse, frontal, CPA)</td>
<td>5</td>
<td>80</td>
<td>1</td>
<td>0</td>
</tr>
<tr>
<td>total</td>
<td>43</td>
<td>74</td>
<td>9</td>
<td>2</td>
</tr>
</tbody>
</table>

* CPA = cerebellopontine angle; incl = including; Tx = treatment.
† Includes two in whom the operation was abandoned.

**TABLE 2**

<table>
<thead>
<tr>
<th>No. of Patients (%)</th>
<th>No. of Patients</th>
<th>Success</th>
<th>Failure</th>
</tr>
</thead>
<tbody>
<tr>
<td>Tx</td>
<td>Total No. of Patients</td>
<td></td>
<td></td>
</tr>
<tr>
<td>VP shunt</td>
<td>12</td>
<td>7 (58)</td>
<td>5 (42)</td>
</tr>
<tr>
<td>non-VP shunt</td>
<td>31</td>
<td>21 (68)</td>
<td>10 (32)</td>
</tr>
</tbody>
</table>

**TABLE 3**

<table>
<thead>
<tr>
<th>No. of Repeated ETVs</th>
<th>Tumor Location</th>
<th>Patients</th>
<th>Ops</th>
<th>No. w/ Final Success</th>
</tr>
</thead>
<tbody>
<tr>
<td>midbrain</td>
<td>2</td>
<td>2</td>
<td>2</td>
<td></td>
</tr>
<tr>
<td>posterior fossa</td>
<td>3</td>
<td>5</td>
<td>2</td>
<td></td>
</tr>
<tr>
<td>3rd ventricle (incl arachnoid cyst)</td>
<td>1</td>
<td>1</td>
<td>1</td>
<td></td>
</tr>
<tr>
<td>total</td>
<td>6</td>
<td>8</td>
<td>5</td>
<td></td>
</tr>
</tbody>
</table>

* Interval between failure and repeated ETV was 25 months (751 days).
Some authors, however, believe that CSF flow from the third ventricle is blocked. Neverthe-
less, these results support the theoretical basis of ETV.

TABLE 4
Complications in nine patients who underwent ETV for hydrocephalus

<table>
<thead>
<tr>
<th>Complication</th>
<th>No. of Patients</th>
</tr>
</thead>
<tbody>
<tr>
<td>venous bleeding</td>
<td>3</td>
</tr>
<tr>
<td>decreased conscious</td>
<td>2</td>
</tr>
<tr>
<td>diabetes insipidus</td>
<td>1</td>
</tr>
<tr>
<td>EOM limitation</td>
<td>2</td>
</tr>
<tr>
<td>infection</td>
<td>1</td>
</tr>
<tr>
<td>total (rate)</td>
<td>9 (20.9%)</td>
</tr>
</tbody>
</table>

* EOM = extraocular muscle.

Patient Age

Some authors have suggested that younger children may have less favorable outcomes than older patients after undergoing ETV to manage hydrocephalus. Other articles support our outcomes for ETV performed to treat these causes of hydrocephalus.

The management of hydrocephalus associated with posterior fossa tumors has been a longstanding controversy. Use of ETV in patients with posterior fossa tumors has been reported before or after tumor resection, and its successful outcome has been between 50 and 85%. In the past, some authors advocated preoperative placement of a permanent ventricular shunt before the definitive tumor resection because this decreased the morbidity and mortality rates associated with the tumor resection. Nevertheless, the value of a precraniotomy shunt was questioned by others because some patients’ postoperative courses were complicated by upward herniation, intratumoral hemorrhage, and infection. In addition, early surgery and the use of corticosteroid agents have decreased the morbidity and mortality rates with no need for a precraniotomy shunt placement.

Our data showed that six (66.6%) of nine of our patients with posterior fossa tumors required shunt placement after ETV. This supports our approach of early surgery rather than initial ETV followed by surgery in children with posterior fossa tumors. We then perform an ETV in symptomatic patients, but cautiously observe them for signs that they need shunt placement. Total resection of posterior fossa tumors should restore the CSF pathway from the third ventricle to subarachnoid space through the sylvian aqueduct and the fourth ventricle; however, secondary adhesions around the sylvian aqueduct or outlets of the fourth ventricle may cause obstructive hydrocephalus.

In our series, six patients who underwent repeated ETVs were finally shunt independent (100%), and the mean interval between the first and second ETV was 25 months. Others have reported a 35% success rate with repeated ETV. The most common cause of failed ETV was occlusion of the stoma by scar tissue, and the mean interval between the first and second ETV was 12.8 months in a multicenter evaluation.
Effect of Previous Ventricular CSF Shunt Therapy

The role of previous ventricular shunt placement in the outcome of ETV is controversial. Teo and Jones\textsuperscript{20} had reported statistically significantly higher success rates in the outcome of patients with previous ventricular shunt placement than in those without a previous shunt insertion (84% compared with 29%). They proposed a hypothesis that the shunt decreased the transmantle pressure (the pressure gradient between the ventricles and the subarachnoid space) and thereby allowed subarachnoid space to open and become mature. In addition, patients tend to be older because they have gone through a period of CSF shunting.\textsuperscript{20} In contrast, the opposite outcome was observed in other reports.\textsuperscript{17}

Postoperative Complications

Although ETV is generally considered a safe procedure, complication rates between 6 and 20% have been reported in several series. The majority of published instances were clinically insignificant, although several serious and fatal complications have been reported.\textsuperscript{1,3,8,12,18,20} Complications related to the ETV procedure included intraoperative venous/arterial bleeding, seizures, CSF leakage, infection, oculomotor nerve palsy, bradycardia, temperature regulation disturbance, diabetes insipidus, syndrome of inappropriate antidiuretic hormone, basilar artery injury due to formation of a traumatic aneurysm or perforation, and subdural hematoma.\textsuperscript{1,3,7,8,12,18,20} Schroeder, et al.,\textsuperscript{18} reported the first death associated with ETV. A severe perimesencephalic–prepontine subarachnoid hemorrhage was identified on the postoperative computerized tomography scan. A review of the operative video showed that the Fogarty catheter had slipped posteriorly, leading to perforation of the floor of the third ventricle and injury to the basilar artery or perforating vessel. Therefore, to avoid this vascular injury, perforation of the floor of the third ventricle should be performed in the midline, halfway between the infundibular recess and the mammillary bodies, just behind the dorsum sellae.\textsuperscript{7}

CONCLUSIONS

Due to significant advances in technique and equipment, ETV is now a popular alternative to ventricular shunts for the treatment of obstructive hydrocephalus secondary to tumors in children. It obviates the placement of a foreign body such as a ventricular shunt, thus avoiding shunt-related complications such as malfunction, infection, and over-drainage.

References


Manuscript received October 10, 2005. Accepted in final form November 11, 2005.
Address reprint requests to: George I. Jallo, M.D., Division of Pediatric Neurosurgery, The Johns Hopkins Hospital, 600 North Wolfe Street, Baltimore, Maryland 21287. email: gjallo1@jhmi.edu.