Hydrocephalus is one of the most common pathologies requiring neurosurgical intervention in children. Managing hydrocephalus due to pediatric CNS tumors presents a complex and challenging problem. Postresection hydrocephalus occurs in approximately 30% of children with posterior fossa tumors, and obstructive hydrocephalus is present in approximately 80% of pediatric patients with posterior fossa tumors. Surgical options for addressing hydrocephalus have traditionally included ventriculoperitoneal shunting (VPS) when tumor resection or medical management alone are ineffective. Endoscopic third ventriculostomy (ETV) has emerged as an attractive treatment strategy for tumor-associated hydrocephalus because it offers a lower risk of infection and hardware-related complications; however, relatively little has been written on the topic of ETV specifically for the treatment of tumor-associated hydrocephalus. Here, the authors reviewed the existing literature on the use of ETV in the treatment of tumor-associated hydrocephalus, focusing on the frequency of ETV use and the failure rates in patients with hydrocephalus secondary to CNS tumor.

METHODS The authors queried PubMed for the following terms: “endoscopic third ventriculostomy,” “tumor,” and “pediatric.” Papers with only adult populations, case reports, and papers published before the year 2000 were excluded. The authors analyzed the etiology of hydrocephalus and failure rates after ETV, and they compared failure rates of ETV with those of VPS where reported.

RESULTS Thirty-two studies with data on pediatric patients undergoing ETV for tumor-related hydrocephalus were analyzed. Tumors, particularly in the posterior fossa, were reported as the etiology of hydrocephalus in 38.6% of all ETVs performed (984 of 2547 ETVs, range 29%–55%). The ETV failure rate in tumor-related hydrocephalus ranged from 6% to 38.6%, and in the largest studies analyzed (> 100 patients), the ETV failure rate ranged from 10% to 38.6%. The pooled ETV failure rate was 18.3% (199 failures after 1087 procedures). The mean or median follow-up for ETV failure assessment ranged from 6 months to 8 years in these studies. Only 5 studies directly compared ETV with VPS for tumor-associated hydrocephalus, and they reported mixed results in regard to failure rate and time to failure. Overall failure rates appear similar for ETV and VPS over time, and the risk of infection appears to be lower in those patients undergoing ETV. The literature is mixed regarding the need for routine ETV before resection for posterior fossa tumors with associated hydrocephalus.

CONCLUSIONS Treatment of tumor-related hydrocephalus with ETV is common and is warranted in select pediatric patient populations. Failure rates are overall similar to those of VPS for tumor-associated hydrocephalus.

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KEYWORDS tumor-associated hydrocephalus; endoscopic third ventriculostomy; pediatric

ABBREVIATIONS ETV = endoscopic third ventriculostomy; ETVSS = ETV Success Score; VPS = ventriculoperitoneal shunting.

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infection risk, but the overall reoperation rates for recurrent hydrocephalus are equivalent between ETV and VPS. However, relatively little has been written specifically about the use of ETV in the pediatric CNS tumor population.

Here, we review the existing literature on ETV in the treatment of tumor-associated hydrocephalus, paying specific attention to the frequency of ETV use in this clinical scenario and the failure rates of preresection ETV in patients with hydrocephalus secondary to CNS tumors.

Methods

We queried PubMed in August 2019 for the following terms: “endoscopic third ventriculostomy,” “tumor,” and “pediatric.” We excluded papers that evaluated only adult (age 18 years and older) populations, but we included mixed adult and pediatric population studies if analysis of the pediatric population was reported separately. We also excluded papers published before 2000 and case reports. Results were filtered for English-language literature. Even if the primary aim of the study in question was not tumor-related hydrocephalus, we analyzed subpopulations in papers that reported tumor as a cause for hydrocephalus.

Data were collected on hydrocephalus etiology or indication for ETV, study type (prospective cohort, retrospective cohort, or systematic review/meta-analysis), year of data collection, ETV failure rates, mean postoperative follow-up, and comparison of outcomes between ETV and VPS, if reported. Linear regression was used to analyze the effect of study publication year on ETV failure rate. Pooled calculations for frequency of ETV for tumor-associated hydrocephalus etiology and for ETV failure rates over multiple studies (excluding systemic reviews and meta-analyses) were performed.

Results

The initial search query yielded 179 study results. Of these, 166 studies were published in 2000 or later. Of these, 32 of which met the inclusion criteria (Fig. 1).

Thirty-two studies with data on pediatric patients undergoing ETV for tumor-related hydrocephalus were analyzed, including 3 systematic literature reviews, 3 international prospective cohort studies, 2 multiinstitutional prospective cohort studies, and 24 retrospective or prospective single-institution studies.

Frequency of ETV for Hydrocephalus Secondary to CNS Tumors

Table 1 displays a summary of the literature on frequency of ETV use for tumor-associated hydrocephalus. The cause of hydrocephalus was neoplasia in 29% to 55% of ETV cases analyzed. In the largest studies (> 1000 patients), the cause of hydrocephalus was neoplasia in approximately 35% of ETV cases. The overall pooled frequency from all primary studies (excluding review articles) was 38.6%, whereas the mean (± SD) frequency was 39.5% ± 9.0%. Few studies reported the percentage of tumor by subtypes, but in the studies that did so, posterior fossa tumors and midbrain/tectal tumors were the most common subtype.

ETV Failure Rates in Tumor-Related Hydrocephalus

Table 2 displays a summary of the literature that reported ETV failure rates for tumor-related hydrocephalus. In the largest studies analyzed (> 100 patients undergoing ETV for tumor-related hydrocephalus), the failure rate of ETV ranged from 10% to 38.6%. Overall, failure rates
TABLE 1. Frequency of ETV use for tumor-associated hydrocephalus

<table>
<thead>
<tr>
<th>Authors &amp; Year</th>
<th>Design, Level of Evidence</th>
<th>Pt Population</th>
<th>Frequency</th>
<th>Tumor Subtype/Location</th>
</tr>
</thead>
<tbody>
<tr>
<td>Madsen et al., 2018</td>
<td>Systematic literature review (130 studies), level III*</td>
<td>11,952 ETVs in children w/ any-cause hydrocephalus</td>
<td>Pooled mean: 34.1% (SD 17.2%)</td>
<td>NR</td>
</tr>
<tr>
<td>Bouras &amp; Sgouros, 2011</td>
<td>Systematic literature review (34 studies), level III*</td>
<td>2985 ETVs in 2884 adult &amp; pediatric pts for any-cause hydrocephalus</td>
<td>37.6%</td>
<td>NR</td>
</tr>
<tr>
<td>Kulkarni et al., 2016</td>
<td>International prospective cohort, level II</td>
<td>336 pts who had ETV for any-cause hydrocephalus</td>
<td>137 (41.0%)</td>
<td>21.2% midbrain; 14.4% posterior fossa; 5.4% supratentorial</td>
</tr>
<tr>
<td>Lam et al., 2014</td>
<td>National database retrospective review, level III</td>
<td>501 pts who had ETV for any cause</td>
<td>209 (41.7%)</td>
<td>NR</td>
</tr>
<tr>
<td>Kulkarni et al., 2010*</td>
<td>International prospective cohort, level II</td>
<td>1209 pts who had ETV (n = 489) or VPS (n = 720) for any-cause hydrocephalus</td>
<td>175 (35.8% of all ETVs)</td>
<td>NR</td>
</tr>
<tr>
<td>Naftel et al., 2011</td>
<td>Single-institution consecutive case series, level IV</td>
<td>151 pts who had ETVs btwn 1995 &amp; 2009</td>
<td>72 (47.7%)</td>
<td>5 (6.9%) supratentorial; 46 (63.9%) midbrain; 21 (29.2%) posterior fossa</td>
</tr>
<tr>
<td>Kulkarni et al., 2009</td>
<td>International cohort from 12 institutions, level II</td>
<td>618 pts who had ETV for any cause</td>
<td>182 (29.4%)</td>
<td>62 (34.1%) midbrain tectal; 120 (65.9%) nontectal</td>
</tr>
<tr>
<td>Beuriat et al., 2017</td>
<td>Single-institution retrospective review, level III</td>
<td>975 pts treated for hydrocephalus; 280 pts who had ETV</td>
<td>155 (55.4%)</td>
<td>NR</td>
</tr>
<tr>
<td>Feng et al., 2004</td>
<td>Single-institution retrospective review, level III</td>
<td>58 pts who had ETV for any-cause hydrocephalus</td>
<td>21 (36.2%)</td>
<td>NR</td>
</tr>
<tr>
<td>Furlanetti et al., 2012</td>
<td>Consecutive case series, level IV</td>
<td>114 pediatric pts who had ETV for any-cause hydrocephalus</td>
<td>33 (28.9%)</td>
<td>NR</td>
</tr>
</tbody>
</table>

NR = not reported; pt = patient.
* Data presented from systemic reviews were not included in the overall calculations of pooled/mean frequencies of ETV for tumor-associated hydrocephalus. The overall pooled/mean frequency calculations are derived from primary studies only.

ranged from 6% to 38.6%. The overall pooled failure rate from all primary studies (excluding review articles) was 18.3%, whereas the mean (± SD) failure rate was 17.4% ± 8.6%. Postoperative follow-up ranged from 6 months to 8 years. Timing of ETV (pre- vs postresection ETV) and tumor subtype by study are shown.

Figure 2 displays failure rates by publication year of each study. No difference in failure rate by study year was seen on linear regression analysis (R² = 0.016, p = 0.61).

Comparison of ETV and VPS Failure Rates for Tumor-Associated Hydrocephalus

Table 3 compares failure rates and time to failure for ETV versus VPS in pediatric patients with tumor-associated hydrocephalus. There were only 5 studies with direct comparison of ETV versus VPS specifically for tumor-associated hydrocephalus. Two studies (those by El-Ghandour and Beuriat et al.) reported a significantly lower failure rate with ETV than with VPS. Regarding time to failure, Dewan et al. reported a significantly shorter time to failure for ETV than for VPS (p = 0.03), whereas El-Ghandour did not find a significant difference in time to failure.

Discussion

As enthusiasm for the use of ETV in the treatment of hydrocephalus as a whole has grown, understanding the benefits of ETV in tumor-associated hydrocephalus may improve overall outcomes. Here, we have reviewed the literature on ETV in tumor-related hydrocephalus, including the relative frequency of ETV use in patients with this disorder and the failure rates of ETV in this population. Tumors, particularly in the posterior fossa, were reported as the etiology of hydrocephalus in 29% to 55% of all ETVs performed. The ETV failure rate ranged from 10% to 38.6% in the largest studies analyzed. No differences were observed for failure rate by study year. Only 5 studies directly compared ETV with VPS for tumor-associated hydrocephalus, and they reported mixed results regarding superiority in regard to failure rate and time to failure.

Frequency of ETV for Tumor-Related Hydrocephalus

Tumor was the etiology of hydrocephalus in 29%–55% of ETV cases in the studies reported. Two systematic literature reviews, one by Bouras and Sgouros and the other by Madsen et al., demonstrated that 34.1% ± 17.2% (pooled mean ± SD) and 37.6% of pediatric ETVs, respectively, were performed for tumor-associated hydrocephalus. These findings are similar to those that Kulkarni et al. found in an international prospective cohort (35.8% of all ETVs were for tumor-related hydrocephalus). Slightly higher rates were reported by Lam et al. (41.7% of patients in the cohort), Beuriat et al. (55.4%), and Naftel et al. (47.7%). Generally, the rates of ETV use for tumor-related hydrocephalus have not increased over time. The
## TABLE 2. Failure rates of ETV for tumor-related hydrocephalus

<table>
<thead>
<tr>
<th>Authors &amp; Year</th>
<th>Design, Level of Evidence</th>
<th>Pt Population</th>
<th>ETV Failure Rate &amp; Follow-Up Duration</th>
<th>Pre- or Post-Resection ETV</th>
<th>Tumor Subtype/Location</th>
</tr>
</thead>
<tbody>
<tr>
<td>Lam et al., 2014</td>
<td>National database retrospective review, level III</td>
<td>501 pts who had ETV for any etiology; 209 pts w/ tumor</td>
<td>24.9% (52/209), 1 yr; 38.6%, 5 yrs</td>
<td>NR</td>
<td>NR</td>
</tr>
<tr>
<td>Kulkarni et al., 2009</td>
<td>International cohort from 12 institutions, level II</td>
<td>618 pts who had ETV for any etiology; 133 pts w/ tumor</td>
<td>27.8% (37/133), 6 mos</td>
<td>NR</td>
<td>88 nontectal tumors; 45 tectal tumors</td>
</tr>
<tr>
<td>Beuriat et al., 2017</td>
<td>Single-institution retrospective review, level III</td>
<td>975 pts treated for hydrocephalus; 280 pts treated w/ ETV; 155 pts w/ tumor</td>
<td>10.3% (16/155), 1 yr</td>
<td>NR</td>
<td>NR</td>
</tr>
<tr>
<td>Sainte-Rose et al., 2001</td>
<td>Single-institution consecutive series, level IV</td>
<td>206 pts who had surgery for posterior fossa tumor; 67 pts treated w/ ETV</td>
<td>7.5% (5/67), mean 2.2 yrs</td>
<td>Pre</td>
<td>46% medulloblastoma; 24% ependymoma; 2% astrocytoma; 3% ganglioglioma; 3% PNET; 3% other</td>
</tr>
<tr>
<td>Dewan et al., 2017</td>
<td>Time-to-failure analysis via a systematic review &amp; single-institution retrospective review, level III*</td>
<td>408 pts who had ETV &amp;/or VPS for posterior fossa tumor-related hydrocephalus</td>
<td>21% cumulative ETV failure for tumor, median 26 mos</td>
<td>NR</td>
<td>49.5% medulloblastoma; 24.7% astrocytoma; 23.3% ependymoma; 2.6% other</td>
</tr>
<tr>
<td>Morelli et al., 2005</td>
<td>Single-institution retrospective review, level III</td>
<td>160 pts w/ posterior fossa tumors; 22 pts treated w/ ETV for hydrocephalus</td>
<td>18% (4/22), 12 mos</td>
<td>Mixed pre &amp; post</td>
<td>26% pilocytic astrocytoma; 19% ependymoma; 17% medulloblastoma; 14% WHO II astrocytoma; 5% WHO III astrocytoma; 19% other</td>
</tr>
<tr>
<td>Frisoli et al., 2019</td>
<td>Single-institution retrospective review, level III</td>
<td>74 pts w/ hydrocephalus who had posterior fossa tumor resection; 38 pts treated w/ ETV</td>
<td>16% (6/38), mean 5 yrs</td>
<td>Pre</td>
<td>44.7% medulloblastoma; 39.5% WHO I astrocytoma; 7.9% ependymoma; 5.3% ATRT</td>
</tr>
<tr>
<td>O'Brien et al., 2006</td>
<td>Single-institution series, level III</td>
<td>42 pts w/ hydrocephalus &amp; midline tumors</td>
<td>32% (13/41; 1 pt excluded from follow-up), mean 32 mos</td>
<td>Intra-resection</td>
<td>24 pineal tumors; 9 tectal tumors; 3 midbrain tumors; 3 thalamic tumors; 1 pontine tumor; 1 3rd ventricular tumor</td>
</tr>
<tr>
<td>El Beltagy et al., 2010</td>
<td>Single-institution retrospective review, level III</td>
<td>40 pts w/ hydrocephalus who had posterior fossa tumor resection &amp; preresection ETV</td>
<td>35.0% (14/40), mean 13 mos</td>
<td>Pre</td>
<td>21 medulloblastoma; 9 WHO I astrocytoma; 4 WHO II astrocytoma; 6 ependymoma</td>
</tr>
<tr>
<td>Bhatia et al., 2009</td>
<td>Single-institution retrospective review, level III</td>
<td>37 pts w/ hydrocephalus secondary to posterior fossa tumors who had preresection ETV</td>
<td>13.5% (5/37), mean 56 mos</td>
<td>Pre</td>
<td>42% medulloblastoma; 41% WHO I astrocytoma; 7% ependymoma; 10% other</td>
</tr>
<tr>
<td>El-Ghandour, 2011</td>
<td>Single-institution retrospective review, level III</td>
<td>53 pts w/ obstructive hydrocephalus due to posterior fossa midline tumors; 32 pts who had ETV</td>
<td>6.2% (2/32), mean 27 mos</td>
<td>NR</td>
<td>62.5% medulloblastoma; 37.5% ependymoma</td>
</tr>
<tr>
<td>Feng et al., 2004</td>
<td>Single-institution retrospective review, level III</td>
<td>58 pts who had ETV for any-cause hydrocephalus; 21 pts w/ tumor</td>
<td>19% (4/21); w/in 9 mos</td>
<td>NR</td>
<td>8 pineocytoma; 4 hemangioblastoma; 3 thalamic astrocytoma; 2 ependymoma; 2 medulloblastoma; 2 other</td>
</tr>
<tr>
<td>Tamburini et al., 2008</td>
<td>Prospective single-institution cohort, level II</td>
<td>30 w/ persistent postresection hydrocephalus who had ETV; 104 pts treated w/ posterior fossa tumor resection</td>
<td>10.0% (3/30), NR</td>
<td>Post</td>
<td>12 WHO I astrocytoma; 10 medulloblastoma; 8 ependymoma</td>
</tr>
<tr>
<td>Furfantecci et al., 2012</td>
<td>Consecutive single-institution case series, level IV</td>
<td>114 pediatric pts who had ETV for any-cause hydrocephalus; 33 pts w/ posterior fossa tumors</td>
<td>9.1% (3/33), mean 65 mos</td>
<td>NR</td>
<td>NR</td>
</tr>
</tbody>
</table>

CONTINUED ON PAGE 5 »
rates of ETV reported in these studies for various locations or tumor subtypes varied substantially.

ETV Failure in Tumor-Related Hydrocephalus

In the largest studies analyzed (> 100 patients undergoing ETV for tumor-related hydrocephalus), the failure rate of ETV ranged from 10% to 38.6%. Overall, failure rates ranged from 6% to 35%. ETV failure generally occurs quickly (within the first 3 months) but then the failure rate plateaus. These failure rates are overall unsurprising and provide a frame of reference with which to counsel patients and families regarding the expected outcomes of ETV surgery.

Lam et al. analyzed the national MarketScan database from 2003 to 2011 and determined that of the 209 patients with tumor-associated hydrocephalus who underwent ETV, 157 (75.1%) of the procedures were successful at 1 year. Furthermore, the 5-year success rate for ETV in this population of patients was 61.4%. The 5-year success rate for ETV in children with tumor-associated hydrocephalus was also found to be similar to that for other causes of hydrocephalus (65.5% with myelomeningocele, 52.3% with congenital etiologies).

Kulkarni et al. reported data from an international cohort from 12 institutions. The authors included 618 patients who underwent ETV for any cause, 29.4% of whom (182 patients of 618 total) underwent ETV for tumor-related hydrocephalus. Ninety-six (72.2%) of 133 patients who underwent ETV for tumor-related hydrocephalus had treatment success at 6 months. Tectal tumors were associated with a higher ETV success rate at 6 months than nontectal tumors (82.2% vs 67.0%).

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The study included 408 patients, 284 of whom were patients with tumor-related hydrocephalus. Numerous studies have compared ETV and VPS for treatment of pediatric hydrocephalus generally, but few have specifically compared the two treatments for tumor-related hydrocephalus. Dewan et al. conducted time-to-failure analysis of ETV and VPS for hydrocephalus secondary to posterior fossa tumors via a systematic review and single-institution retrospective review. The study included 408 patients, 284 of whom were from the authors’ institution. The cumulative failure rates were statistically similar for ETV and VPS, but the median time to failure was earlier for ETV than for VPS (0.82 [IQR 0.2–1.8] months vs 4.7 [IQR 0.3–5.7] months, p = 0.03). In the time-to-failure analysis, the ETV survival curve dropped sharply and then plateaued about 2 months postoperatively, whereas the VPS survival curve fell gradually but then crossed below the ETV curve at 5.7 months postoperatively (p = 0.21, log-rank test). The authors concluded that ETV failure occurred before VPS failure, but long-term success may be similar or higher for ETV. El-Ghandour also compared ETV and VPS for posterior fossa tumor–related hydrocephalus in 53 pediatric patients, finding ETV to be superior to VPS because of the shorter length of surgery (15 minutes vs 35 minutes), lower morbidity (9.3% vs 38%), reduced mortality (0% vs 4.7%), and lower incidence of failure (6.2% vs 38%). The ETV failure rate at a 27-month mean follow-up was 6.2% versus a VPS failure rate of 38% at a 25-month mean follow-up (p = 0.003). Sainte-Rose et al. reported an ETV failure rate of 7.5% at follow-up of 2.2 years versus a VPS failure rate of 12.5% at the same follow-up duration (p = 0.033). In patients with hydrocephalus secondary to posterior fossa tumor. Given the dearth of literature in this subpopulation of patients undergoing ETV, future prospective studies investigating efficacy and safety of ETV versus VPS in patients with tumor-related hydrocephalus are warranted.

### TABLE 3. Comparison of failure rates of ETV and VPS for tumor-associated hydrocephalus

<table>
<thead>
<tr>
<th>Authors &amp; Year</th>
<th>Design, Level of Evidence</th>
<th>Pt Population</th>
<th>Length of Follow-Up</th>
<th>Failure Rate</th>
<th>Time to Failure (mos)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Dewan et al., 2017</td>
<td>Systematic review &amp; single-institution retrospective review, level III</td>
<td>408 pts undergoing ETV &amp;/or VPS for posterior fossa tumor–related hydrocephalus</td>
<td>Median 26 mos</td>
<td>ETV: 21%, VPS: 29%</td>
<td>p = 0.105</td>
</tr>
<tr>
<td>Bauriat et al., 2017</td>
<td>Retrospective review of single-institution cohort, level III</td>
<td>975 pts treated for hydrocephalus; 280 pts treated w/ ETV (155 for tumor-related hydrocephalus); 695 pts treated w/ VPS (160 for tumor-related hydrocephalus)</td>
<td>1 yr</td>
<td>ETV: 10.3% (16/155), VPS: 38.8% (62/160)</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>El-Ghandour, 2011</td>
<td>Single-institution retrospective review, level III</td>
<td>53 pts w/ obstructive hydrocephalus due to posterior fossa midline tumors</td>
<td>ETV: mean 27 mos; VPS: mean 25 mos</td>
<td>ETV: 6.2% (2/32), VPS: 38% (8/21)</td>
<td>0.003</td>
</tr>
<tr>
<td>Sainte-Rose et al., 2001</td>
<td>Single-institution consecutive series, level IV</td>
<td>206 pts who had posterior fossa tumor resection; 67 undergoing ETV; 16 undergoing VPS</td>
<td>Mean 2.2 yrs</td>
<td>ETV: 7.5% (5/67), VPS: 12.5% (2/16)</td>
<td>0.61</td>
</tr>
<tr>
<td>Ruggiero et al., 2004</td>
<td>Single-institution retrospective review, level III</td>
<td>63 children w/ posterior fossa tumors; 20 undergoing ETV; 4 undergoing VPS</td>
<td>Mean 21 mos</td>
<td>ETV: 20% (4/20), VPS: 0% (0/4)</td>
<td>NR</td>
</tr>
</tbody>
</table>

ETV Versus VPS in Tumor-Related Hydrocephalus

Numerous studies have compared ETV and VPS for treatment of pediatric hydrocephalus generally, but few have specifically compared the two treatments for tumor-related hydrocephalus. Dewan et al. conducted time-to-failure analysis of ETV and VPS for hydrocephalus secondary to posterior fossa tumors via a systematic review and single-institution retrospective review. The study included 408 patients, 284 of whom were...
up of 264 days. The failure rates they observed in children undergoing VPS for tumor-associated hydrocephalus are relatively similar to the ETV failure rates for children undergoing ETV for tumor hydrocephalus reported in the largest studies found in our current review (10%–38.6%).

Predictors of Good Outcome After Preresection ETV

In 2009, Riva-Cambrin et al.\(^3\) published a scoring system to predict those pediatric patients who would require CSF diversion after posterior fossa tumor resection. The score consisted of 10 points: 3 points for age < 2 years, 3 points for cerebral metastases, 2 points for moderate or severe hydrocephalus, 1 point for papilledema, and 1 point for preoperative radiographic features consistent with an ependymoma, dorsally exophytic brainstem glioma, or medulloblastoma. High-risk patients were those with scores > 4 points, and low-risk patients were those with scores of 0–2. The rate of postoperative CSF diversion was 73% for high-risk patients and 25% for low-risk patients. Foreman et al.\(^9\) published a modified version of the success score published by Riva-Cambrin et al.\(^3\) in a cohort of 76 patients, finding that 4 variables were significant in predicting postresection hydrocephalus: age < 2 years, moderate/severe hydrocephalus, preoperative tumor diagnosis, and transependymal edema.

The original ETV Success Score (ETVSS) published by Kulkarni et al.\(^7\) was developed to predict which pediatric patients would have the highest chance of hydrocephalus treatment success with ETV and was later validated in a multicenter cohort of 489 patients who underwent ETV.\(^6\) The ETVSS is an estimate of the percentage probability of ETV success, ranging from 0% to 90% probability. There are 3 primary components: age, etiology, and history of previous shunt. Tumors are considered within the etiology component, with nontectal tumors adding 20% to the ETVSS, and tectal tumors adding 30% to the ETVSS. Overall, tumors portend a favorable increase in the ETVSS compared with other etiologies.

Complications of ETV

In the meta-analysis by Madsen and colleagues,\(^23\) complications related to ETV were reported, although those specifically related to tumor-associated hydrocephalus were not analyzed. Overall perioperative mortality was 0.2% ± 0.1%, and the mean ETV failure rate was 34.7% ± 18.0%.

Texakalidis et al.\(^3\) performed a meta-analysis comparing ETV and VPS for pediatric hydrocephalus including 8419 patients (not specifically for tumor-associated hydrocephalus), finding that infection was significantly lower in the ETV group (OR 0.19, 95% CI 0.07–0.53) but that mortality, postoperative CSF leakage, and reoperation rates were similar between groups.

Bouras and Sgouros\(^1\) performed a systematic literature review including 2985 ETVs performed in 2884 patients from 34 studies. Importantly, the population included for this review was mixed (adult and pediatric). The early postoperative mortality rate was 0.21%. The overall complication rate was 8.5%. The rate of intraoperative hemorrhage was 3.7%, and the rate of severe intraoperative hemorrhage was 0.6% (including a 0.21% rate of basilar rupture).

Macarthur et al.\(^22\) published a detailed report of complications encountered in 61 neuroendoscopic procedures for pediatric CNS tumors, 47 of which were ETVs for tumor-associated hydrocephalus. Only 1 death, 6 intraoperative hemorrhages, 4 CSF fistulas, and 1 deep infection occurred, and 9 extraventricular drains were required for temporary CSF diversion postoperatively.

El-Ghandour\(^1\) analyzed complications related to ETV in a posterior fossa tumor cohort, reporting no mortality in 32 patients who underwent ETV. 1 case (3.1%) of CSF leak, and 2 cases (6.2%) of bleeding. Ray et al.\(^30\) also analyzed ETV complications in a pediatric tumor-related hydrocephalus cohort, finding a total complication rate of 20.9% in 43 ETVs, including 1 infection and 3 cases of venous bleeding.

Patient Selection for ETV in Tumor-Related Hydrocephalus

Controversy exists regarding the necessity of default preresection ETV in posterior fossa tumor–related hydrocephalus.\(^5\) Fritsch et al.\(^11\) reported that in a single-institution series of 58 patients with posterior fossa tumors, 52 presented with hydrocephalus, but only 6 patients (11.5%) required permanent treatment for hydrocephalus at a mean follow-up of 25 months. Of those 6, 4 patients received a VP shunt and 2 patients underwent ETV. Morelli et al.\(^26\) posed a similar question of the necessity of ETV in posterior fossa tumor–related hydrocephalus, finding that routine postoperative ETV may be beneficial in the treatment of persistent hydrocephalus, but not prophylactically, given that only 22 (13.8%) of 160 patients with posterior fossa tumors required treatment for hydrocephalus (ultimately with ETV) in the cohort.

In some populations of patients with CNS tumors, the overall survival prognosis brings into question the ethics of hydrocephalus treatment. For instance, Roujeau et al.\(^33\) reported that in patients with hydrocephalus secondary to brainstem gliomas, the overall 1-year survival rate was 33%, and the survival rate of patients with obstructive hydrocephalus was not significantly different from that of patients who did not develop hydrocephalus despite treatment. Given the overall poor prognosis in these patients, the need for CSF diversion as a whole must be counterbalanced with overall patient outcome and quality of life.

ETV in Non–Posterior Fossa Tumors

Most of the studies analyzed in this review investigated patients who underwent ETV for posterior fossa tumors; however, several studies reported results from ETV for those with lesions outside the posterior fossa. For example, Macarthur et al.\(^22\) reported a consecutive series of 61 neuroendoscopic procedures in 53 children with tumors, 47 of which were ETV for tumor-related hydrocephalus. Sixteen of the 53 tumors were cerebellar (30.2%), whereas 13 were brainstem (24.5%), 12 were third ventricular (22.6%), 5 were thalamic or hypothalamic (9.4%), 4 were pineal (7.5%), and 3 were classified as “other” location. Morgenstern et al.\(^\) determined that none of the 15 patients who underwent ETV for pineal region tumors in their study required repeat CSF diversion procedures at the 6-month follow-up. Hayhurst et al.\(^\) reported a small series of 11 patients who underwent ETV for hydrocephalus second-
ary to cerebellopontine angle tumors, with 7 (63.6%) of the patients remaining shunt-free at follow-up. Li et al.29 published a consecutive case series of 31 patients with tectal glioma and hydrocephalus (cohort age range 6 weeks–20 years), 18 of whom were treated with ETV, and 2 (11%) of those 18 experienced ETV failure at long-term follow-up (median 8 years).

Molecular Diagnostics and Tumor-Related Hydrocephalus

New tools of molecular analysis have shed light on how different tumor subgroups may be more susceptible to developing hydrocephalus. For instance, Schneider et al.30 reported that in a cohort of 143 medulloblastoma pediatric patients, no patient with the Wnt tumor subtype required CSF diversion procedures (ETV or VPS), whereas 29% of Shh, 29% of group 3, and 43% of group 4 patients required CSF diversion. Therefore, molecular diagnostic techniques may aid in stratifying the need for CSF diversionary procedures for patients with tumors harboring a variety of molecular signatures.

Limitations

This study has several limitations. We only searched the PubMed database and may have excluded contributory literature contained in other sources. Most of the studies reported herein are single-center retrospective studies, which limits the generalizability of results and also implies significant heterogeneity in patient management and surgical decision-making. We did not perform a meta-analysis, which limits how the data presented can be quantified or interpreted with any statistical significance. Furthermore, we did not assess patient characteristics (e.g., age differences, tumor subtype), hospital size/case volume, or technical differences in ETV operations between studies (e.g., use of Fogarty balloons for wider fenestration), which may affect failure rates.

Conclusions

The use of ETV in tumor-associated hydrocephalus is largely heterogeneous with regard to indications, timing, and tumor subtypes. The failure rate of ETV for tumor-associated hydrocephalus in the studies with the largest patient populations ranges from 10% to 38.6%. Although ETV has been reported to fail more than VPS in tumor-associated hydrocephalus in the short term, overall rates of success/failure may be similar over time, and the risk of infection appears to be lower in those patients undergoing ETV. Molecular tumor profiles may help predict those patients at higher risk of a CSF diversionary requirement associated with tumor resection. Nonetheless, treatment of tumor-associated hydrocephalus with ETV is relatively common (approximately 30% to 55% of all ETVs performed), and future studies are warranted to elucidate the role of ETV in this complex patient population.

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References


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