Hydrocephalus in children with posterior fossa tumors: role of endoscopic third ventriculostomy

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Object. Controversy exists regarding the indication for endoscopic third ventriculostomy (ETV) in children with hydrocephalus caused by posterior fossa tumors. The authors present their treatment modalities and discuss the role of ETV in the management of hydrocephalus.

Methods. The authors retrospectively reviewed the cases of 58 children who were admitted to their clinic consecutively with posterior fossa tumors between January 1999 and December 2003. Fifty-two patients presented with hydrocephalus. The mean age at the time of admission was 6 years and 3 months. The mean follow-up period was 25 months. The authors evaluated how many children required a cerebrospinal fluid (CSF) draining procedure (external ventricular drain [EVD], ventriculoperitoneal [VP] shunt, or ETV) prior to or following tumor removal.

Only six patients (11.5%) required permanent treatment for hydrocephalus. Four patients received a VP shunt and two patients underwent ETV. A temporary EVD was placed in five patients (two required a shunt). Forty-six patients (88.5%) did not require a permanent CSF draining procedure.

Conclusions. For children with posterior fossa tumors, ETV is not indicated as a standard operation either prior to or following tumor removal. Only six of 52 children presenting with hydrocephalus required a permanent CSF draining procedure. Endoscopic third ventriculostomy may be suitable for patients with fourth ventricle outflow obstruction and persisting or progressive hydrocephalus following tumor removal.

Key Words • posterior fossa tumor • hydrocephalus • endoscopic third ventriculostomy • pediatric neurosurgery

Discussions are ongoing in the neurosurgical literature regarding the indication for ETV in children with hydrocephalus caused by a posterior fossa tumor. Recommendations have been made to perform ETV as a routine procedure prior to tumor removal. The conclusion was made that ETV reduces the incidence of hydrocephalus following tumor removal. Further studies have been published that emphasize the use of ETV prior to posterior fossa tumor removal in pediatric patients.

In reviewing our patients’ cases, we concluded that the incidence of hydrocephalus following tumor removal is too low to justify routine ETV prior to tumor removal. We retrospectively analyzed our patient data in light of the following four questions: 1) What is the incidence of persisting hydrocephalus following posterior fossa tumor removal? 2) What is the rationale supporting the indication for ETV prior to tumor removal? 3) What are the risks of ETV prior to tumor removal? 4) If hydrocephalus persists or progresses, what is the role of ETV following tumor removal?

Abbreviations used in this paper: CSF = cerebrospinal fluid; CT = computerized tomography; ETV = endoscopic third ventriculostomy; EVD = external ventricular drain; MR = magnetic resonance; VP = ventriculoperitoneal.

Clinical Material and Methods

We retrospectively reviewed the cases of 58 children who were admitted consecutively to our department with posterior fossa tumors between January 1999 and December 2003. Five patients with brainstem tumors were included in this series. Fifty-two patients presented with symptomatic hydrocephalus on admission. Patients were considered to be symptomatic from hydrocephalus if they presented with the following criteria: headache, nausea, and vomiting; impaired consciousness; and an MR image revealing enlarged ventricles, together with obstruction of the fourth ventricle. The mean age at the time of admission was 6 years and 3 months (range 6 weeks–16 years). The mean follow-up period was 25 months (range 12–70 months).

All patients underwent MR imaging pre- and postoperatively and yearly thereafter. Posterior fossa surgery was performed by board-certified neurosurgeons, and ETV was performed by the two senior authors (M.J.F. and H.M.M.).

All patients underwent early posterior fossa craniotomy and tumor removal on the day of admission or on one of the 2 following days. The histopathology revealed medulloblastoma (17 lesions), pilocytic astrocytoma (12 lesions), anaplastic astrocytoma (nine lesions), ependymoma (eight lesions), and other rare entities, including plexus papilloma, plexus carcinoma, and teratoma (12 lesions).
Endoscopic third ventriculostomy in post fossa tumors

**TABLE 1**

<table>
<thead>
<tr>
<th>Type of Tumor</th>
<th>Procedure(s)</th>
<th>Tumor Location</th>
<th>Extent of Removal</th>
<th>Time Interval From Op to ETV/VP Shunt</th>
<th>Comments</th>
</tr>
</thead>
<tbody>
<tr>
<td>ependymoma</td>
<td>EVD, VP shunt</td>
<td>4th ventricle</td>
<td>complete</td>
<td>10 days</td>
<td>none</td>
</tr>
<tr>
<td>medulloblastoma</td>
<td>EVD, VP shunt</td>
<td>vermis, 4th ventricle</td>
<td>complete</td>
<td>36 mos</td>
<td>diffuse tumor</td>
</tr>
<tr>
<td>spread</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>ependymoma</td>
<td>VP shunt</td>
<td>4th ventricle, left CPA</td>
<td>complete</td>
<td>21 days</td>
<td>36 mos after tumor op</td>
</tr>
<tr>
<td>plexus papilloma</td>
<td>VP shunt</td>
<td>4th ventricle</td>
<td>subtotal</td>
<td>27 mos</td>
<td>meningitis 3 days</td>
</tr>
<tr>
<td>medulloblastoma</td>
<td>ETV</td>
<td>vermis, 4th ventricle</td>
<td>complete</td>
<td>7 days</td>
<td>none</td>
</tr>
<tr>
<td>astrocytoma</td>
<td>ETV</td>
<td>4th ventricle, brainstem</td>
<td>subtotal</td>
<td>5 days</td>
<td>reop 4 yrs later</td>
</tr>
</tbody>
</table>

* CPA = cerebellopontine angle.

**Results**

Five patients received an EVD prior to tumor removal. In three patients the EVD was removed without complication within 10 days. Two patients required placement of a VP shunt: one patient (with an ependymoma) 10 days later and one patient (with a medulloblastoma) 36 months after the tumor surgery.

Two patients (with ependymoma/plexus papilloma) required a VP shunt after tumor removal. Two other patients (with medulloblastoma/astrocytoma [World Health Organization Grade I]) underwent ETV at Days 5 and 7 following posterior fossa surgery. Further details on the six patients who required VP shunts or ETVs are displayed in Table 1.

In summary six patients (11.5%) required a permanent CSF draining procedure, either a VP shunt or an ETV. None of the six patients who had hydrocephalus postoperatively was free of hydrocephalus on admission. Forty-six patients (88.5%) did not require a permanent CSF draining procedure. No complications (infection or neurological deficit) were related to any of the CSF draining procedures (EVD, VP shunt, or ETV). The overall long-term incidence of hydrocephalus in the total of 58 patients was 10.3%.

**Discussion**

In 2001, the results of a retrospective study in which the effectiveness of ETV was evaluated in the management of hydrocephalus before and after posterior fossa tumor removal in children were published.7 The 196 patients were categorized into three groups. Group A consisted of 67 patients with hydrocephalus in whom ETV was performed prior to tumor removal. The mean time interval between ETV and tumor removal was 40 days. Group B consisted of 82 patients with hydrocephalus who underwent early tumor removal and EVD when needed. Group C, 47 patients, had no hydrocephalus on admission. Four patients (6%) in Group A compared with 22 patients (27%) in Group B required further treatment (ETV or VP shunt) following tumor removal because of progressive hydrocephalus. The conclusion was made that ETV, when performed prior to posterior fossa surgery, reduces the incidence of hydrocephalus and should be performed as a routine procedure for the treatment of hydrocephalus in symptomatic patients.

In 2003, Bognar, et al.,1 published a retrospective analysis of the cases of 180 children with posterior fossa tumors. Hydrocephalus was present in 137 patients on admission. All patients underwent early posterior fossa surgery for tumor removal and transient EVD if needed. The overall long-term treatment rate for hydrocephalus following tumor removal was 15.5% (28 patients). The authors concluded that the routine application of preoperative ETV is not indicated because of the small number of patients requiring definite treatment for hydrocephalus.

These two publications exemplify the controversy regarding the role of ETV in the management of hydrocephalus secondary to a posterior fossa tumor in children. Obviously, several different established and accepted means of patient management exist; however, the following questions and answers outline our method and speak to why we do not see an indication for routine ETV prior to tumor surgery.

1. **What is the Incidence of Persisting Hydrocephalus Following Tumor Removal?**

   The incidence of hydrocephalus requiring treatment in pediatric patients with posterior fossa tumors has been reported to be between 15.5 and 36%.1,5,7 Imielinski, et al.,4 reported shunt implantation in 62% (59 of 95) of children with posterior fossa tumors. The range of numbers indicates that definitions differ regarding hydrocephalus and the need for treatment. In our patient population, the percentage of patients that needed permanent CSF drainage (either VP shunt or ETV) following tumor removal was 11.5%. In our opinion, this number is too low to justify a standard additional surgical procedure that has certain risks.

2. **What is the Rationale Supporting the Indication for ETV Prior to Tumor Removal?**

   In the literature, several arguments favor an ETV prior to tumor removal. The first is that an ETV may reduce the incidence of hydrocephalus following tumor removal, yet no study has proved that the incidence of hydrocephalus can be reduced by an ETV procedure prior to tumor removal. The only retrospective study evaluating incidences compares a patient group that had already been treated with an ETV for obstructive hydrocephalus with a group that had not been treated for hydrocephalus.7 Patients in both groups underwent suboccipital craniotomy and tumor removal, the definitive treatment for hydrocephalus.
The question remains, however, regarding how many patients underwent an unnecessary ETV procedure prior to tumor removal. Recent studies and our own data have shown that the long-term incidence of hydrocephalus requiring treatment is only approximately 15%.1

The second argument favoring an ETV prior to tumor removal is that tumor surgery is elective and that deterioration from hydrocephalus during the waiting period can be prevented by ETV. It is often difficult to differentiate between symptoms related to hydrocephalus or to tumors. In patients who are mildly symptomatic and in whom imaging studies demonstrate free CSF flow through the aqueduct and the open fourth ventricle, the indication for ETV is questionable. On the other hand, symptomatic patients with aqueductal or fourth ventricle tumor obstruction are candidates for immediate or early surgical tumor removal. It is our policy to perform such surgery within 24 to 48 hours of admission. Tumor removal constitutes definite treatment, not only for relief from mass effect but also for relief from hydrocephalus in the majority of patients. There is no need for a staged procedure with two operations.

A third argument for an ETV prior to tumor removal is that patients are often admitted with CT scans but without MR images, and if the latter are not immediately available, it may be safer to perform an ETV while waiting for the MR imaging results.

Performing ETV on the basis of CT scans only, without knowing the anatomy of the basilar artery and the third ventricle floor, presents a certain risk in patients with posterior fossa mass effect. It is our policy to obtain MR images for all patients.

3. What are the Risks of ETV Prior to Tumor Removal?

The anatomy of the interpeduncular cistern may be distorted due to elevation of the brainstem and compression of the brainstem against the clivus. No complications have been reported so far resulting from ETVs in patients with posterior fossa mass lesions. On reviewing the MR images of our patients, however, we conclude that we assume an increased risk within this patient population compared with patients without posterior fossa mass effect.

4. If Hydrocephalus Persists, What is the Role of an ETV Following Tumor Removal?

If patients require a permanent CSF draining procedure following tumor removal, what is the indication for an ETV compared with a VP shunt? In our opinion, an ETV is preferable because of the cause of the hydrocephalus. Most patients suffer from obstruction at the level of the fourth ventricle outflow. In addition, an ETV would allow for intrathecal chemotherapy (if indicated, after insertion of a reservoir) without the risk of spreading chemotherapeutic agents into the peritoneal cavity. If chemotherapy and/or radiation therapy are indicated, the risk of shunt infection is eliminated.

Conclusions

Children with posterior fossa tumors should not routinely undergo ETV. Only six (11.5%) of 52 children in our series required a permanent CSF diverting procedure prior to or after tumor removal. A staged procedure (ETV, elective tumor removal) is not necessary in the majority of patients. We favor early, immediate tumor removal without a prior ETV. If indicated, an ETV should be performed in patients with fourth ventricle outflow obstruction following tumor removal.

References


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