Endoscopic third ventriculostomy for shunt dysfunction in occlusive hydrocephalus: long-term follow up and review

JÜRGEN BOSCHERT, M.D., DIETER HELLWIG, M.D., AND JOACHIM K. KRAUSS, M.D.

Department of Neurosurgery, Inselspital, University of Bern, Switzerland; Department of Neurosurgery, Philipps University, Marburg; and Department of Neurosurgery, University Hospital, Mannheim, Germany

Object. Endoscopic third ventriculostomy (ETV) is the treatment of choice for occlusive (noncommunicating) hydrocephalus. Nevertheless, its routine use in patients who have previously undergone shunt placement is still not generally accepted. The authors’ aim was to investigate the long-term effects of ETV in a group of prospectively chosen patients.

Methods. Patients who underwent ETV and had previously undergone shunt placement for occlusive hydrocephalus were followed prospectively for at least 3 years (range 36–103 months, mean 63.6 months). Nine female and eight male patients ranging from 8 to 54 years of age (mean 32 years) had undergone shunt placement 0.7 to 23.5 years (mean 8.1 years) before ETV. Fifteen patients were admitted with underdrainage and two with overdrainage. In six cases, ETV was performed as an emergency operation. The origin of hydrocephalus was aqueductal stenosis in 12 cases and aqueductal compression by a tumor in two cases. Three patients suffered from a fourth ventricle outlet syndrome, and in two patients an additional malresorptive component was suspected. Thirteen patients underwent ETV with shunt removal and insertion of an external drain in one session. The drain served as a safety measure; it could be opened if raised intracranial pressure or ventricular dilation was observed on postoperative imaging studies. In the other four patients the shunt was initially ligated and then removed during a second operation.

Fourteen patients (82%) have remained shunt free. The other three patients, including the two with an additional malresorptive component, needed shunt reimplantation 3 days, 2 weeks, or 7 months after ETV.

Conclusions. Use of ETV is safe and effective for the treatment for shunt dysfunction in patients with obstructive hydrocephalus.

Key Words • aqueductal stenosis • hydrocephalus • endoscopic third ventriculostomy • shunt
Endoscopic third ventriculostomy for shunt dysfunction

several shortcomings: studies are retrospective, some reports do not differentiate the results of ETV according to whether or not patients received shunts, study populations are small and heterogeneous, there is a strong pediatric bias, and follow-up duration frequently is limited. We routinely use ETV instead of shunting in patients with occulsive hydrocephalus. In this study, we report on the long-term follow-up of ETV in a series of 17 predominantly adult patients with occulsive hydrocephalus who had previously undergone shunt placement. This is the first prospective investigation in which long-term follow up is provided for all patients in the study.

Clinical Material and Methods

Patient Population

Data for all patients with shunt dysfunction and occulsive hydrocephalus who underwent ETV were collected prospectively at three different centers. To be included in this open-label pilot study, patients had to fulfill the following criteria: an unequivocal history of occulsive hydrocephalus, shunt dysfunction at the time of presentation, they had to be at least 6 years of age, and they had to be available for long-term follow up. During the period of enrollment for the study, all patients who fulfilled these criteria underwent ETV. To achieve the primary goal of the study, that is, long-term follow up, only data for patients who were treated before July 1999 were analyzed for this report; follow-up data were available for all of them.

There were nine female and eight male patients whose ages when ETV was performed ranged from 8 to 54 years (mean 32 years). Shunts had been present in these patients for 8 months to 23.5 years before ETV (mean 8.1 years). Five patients had VA shunts and 12 had VP shunts. Demographic data, clinical findings, and imaging studies obtained in the individual patients are shown in Table 1. The origin of hydrocephalus was aqueductal stenosis in 12 patients (Cases 3–7, 9–11, 13–15, and 17), aqueductal compression by a tumor in two (Cases 1 and 8), and fourth ventricle outlet syndrome in three (Cases 2, 12, and 16). In two patients ventricle outlet syndrome occurred after resection of a cerebellar astrocytoma (Cases 2 and 16), and in two patients a malresorptive component was suspected in addition to occulsive hydrocephalus. The patient in Case 6 had a history of bacterial meningitis prior to diagnosis of occulsive hydrocephalus, and the patient in Case 7 had a history of perinatal toxoplasmosis.

Surgical Techniques

All operations were performed after induction of general anesthesia. Patients were positioned supine with the head slightly flexed. The ETV was performed through a coronal burr hole by using a standard method. The following types of endoscopic equipment were used: 1) MINOP, 6-mm trochar with 0° and 30° optics (Aesculap, Tuttingen, Germany); 2) Neuroscope I, 6.5 mm with 0° optics; 3) Cranioscope I, 5 mm with 30° optics; 4) a 5.5-mm ventriculoscope with 0° optics; and 5) a 6.5-mm hematoscope with 35° optics (Zeppelin, Pullach, Germany).

In 13 patients ventriculostomy and shunt removal were performed during the same session (Cases 3–6 and 9–17). By the end of the procedure an external ventricular drain was inserted via the burr hole through which the endoscopy had been performed. The drain was used as an additional safety measure, to be opened in case of symptoms of raised ICP or an increase in ventricular size on postoperative imaging studies. The drain remained closed in all except one patient (Case 6); in general, it was removed 3 days postsurgery. In the other four patients the shunt was ligated during ETV, and was then removed in a second operation performed 2 to 6 weeks later (Cases 1, 2, 7, and 8).

Two patients (Cases 2 and 8) presented initially with an overdrainage syndrome and ventricular collapse on imaging studies. In these two patients an antisiphoning device (ShuntAssistant; Miethke, Berlin, Germany) was implanted before ETV. The resistance was purposely set relatively too high so that the ventricles would become sufficiently large to introduce the endoscope. Endoscopy then could be performed in these two patients 7 days and 1 month later, when the ventricles had increased in size.

All patients received follow up for a period of at least 3 years.

Results

Clinical Outcome

Fifteen patients were admitted with symptoms related to shunt obstruction and two suffered from an overdrainage syndrome. In six cases ventriculostomy was performed as an emergency operation. No complications related to ETV occurred in our series. In fourteen of 17 patients the findings were unremarkable for follow up ranging from 36 to 103 months (mean 63.6 months) after ventriculostomy and shunt removal. Neuroimaging studies obtained in a representative patient are shown in Figs. 1 and 2.

The other three patients needed shunt reimplantation (3 days after ETV in Case 6, after 2 weeks in Case 17, and after 7 months in Case 7). Two of these patients had a history of intracranial infection (Cases 6 and 7). The clinical condition of the patient in Case 6 deteriorated rapidly due to acute intracranial hypertension. In this patient, the ventricular drain was connected to a drainage system for immediate relief of ICP. After reimplantation of a shunt system, he recovered completely and was discharged without neurological deficits. Findings on further follow-up review remained unremarkable. The patient in Case 7 suffered from progradient headache after ETV and shunt removal; her symptoms resolved after shunt reimplantation. In the patient in Case 17, symptoms of intracranial hypertension gradually developed. When she was readmitted to the hospital, no neurosurgeon with special expertise in neuroendoscopy was available. Therefore, a VP shunt was implanted to replace the original VA device. Retrospective analysis of the ETV on videotape revealed that the stoma was of insufficient diameter.

When the patients in whom ETV failed were compared with those who had an unremarkable course, the following differences were found: the mean age of patients in whom ETV failed was older (39.3 ± 8.3 years compared with 30.7 ± 11.6 years [mean ± standard deviation]); the interval between shunt placement and ETV was longer (17.7 ± 4.4 years compared with 6.1 ± 6.1 years [mean ± standard deviation]); and two patients had a history of in-
Infection and a suspected malresorptive component (66% compared with 0%). Because of the small number of failures, however, these findings did not reach statistical significance.

Discussion

Our study shows that ETV is an effective treatment in patients with occlusive hydrocephalus who had undergone previous shunting procedures. Because it allows permanent shunt removal and shunt independence in the majority of patients, it provides a cure for this group of patients with often difficult-to-manage shunt dysfunction. In our experience, ETV does not provide long-term relief in patients with concomitant malresorptive hydrocephalus, and those patients should be monitored closely after shunt removal or ligation. No long-term failures were observed during the minimum follow-up period of 3 years in our study.

After Nitze had described the construction of his cystoscope in 1887, as related by Davis, in 1910, L’Espinasse, a urologist from Chicago, was the first to remove the choroid plexus in two children with hydrocephalus by using such an instrument. According to his account, L’Espinasse was the first to introduce endoscopy into neurosurgery. In 1922, Dandy reported his experience with this new technique and created the terms “ventriculoscope” and “ventriculoscopy.” One year later, Mixter successfully performed

---

**TABLE 1**

Clinical features and long-term follow up in 17 patients with previously shunted occlusive hydrocephalus who underwent ETV for shunt dysfunction

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Age (yrs), Sex</th>
<th>Origin of Hydrocephalus &amp; Shunt Type</th>
<th>Time (yrs) Between SP &amp; ETV</th>
<th>Symptoms &amp; Imaging Findings on Admission</th>
<th>Preop Status</th>
<th>FU Post-ETV Status</th>
<th>Current Status</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>14, F</td>
<td>quadrigeminal plate glioma; VP</td>
<td>3.2</td>
<td>HA, somnolence, Parinaud syndrome; shunt dysfunction w/ acute hydrocephalus on CT</td>
<td>emergency</td>
<td>44 mos unremarkable</td>
<td></td>
</tr>
<tr>
<td>2</td>
<td>21, M</td>
<td>obstruction of 4th ventricle outlet after removal of cerebellar astrocytoma; VP</td>
<td>1.2</td>
<td>postural HA; overdrainage w/ ventricular collapse on CT</td>
<td>elective, ventricular dilatation w/ ASD</td>
<td>39 mos unremarkable</td>
<td></td>
</tr>
<tr>
<td>3</td>
<td>30, F</td>
<td>aqueductal stenosis; VP</td>
<td>14</td>
<td>slowly progressive somnolence, gait disturbance, urinary incontinence &amp; seizures; shunt dysfunction w/ 3rd ventricle hydrocephalus on MRI</td>
<td>elective</td>
<td>36 mos unremarkable</td>
<td></td>
</tr>
<tr>
<td>4</td>
<td>30, M</td>
<td>aqueductal stenosis; VA</td>
<td>14.8</td>
<td>HA, somnolence, vomiting; shunt dysfunction w/ 3rd ventricle hydrocephalus on CT</td>
<td>emergency</td>
<td>61 mos unremarkable</td>
<td></td>
</tr>
<tr>
<td>5</td>
<td>31, M</td>
<td>aqueductal stenosis; VA</td>
<td>17.2</td>
<td>HA, somnolence; shunt dysfunction w/ 3rd ventricle hydrocephalus on CT</td>
<td>emergency</td>
<td>61 mos unremarkable</td>
<td></td>
</tr>
<tr>
<td>6</td>
<td>35, M</td>
<td>aqueductal stenosis &amp; malresorptive component (meningitis); VA</td>
<td>16.5</td>
<td>intermittent HA &amp; vomiting; no obvious signs of shunt dysfunction on CT</td>
<td>elective</td>
<td>shunt reimplemented after 3 days</td>
<td>unremarkable</td>
</tr>
<tr>
<td>7</td>
<td>51, F</td>
<td>aqueductal stenosis &amp; malresorptive component (perinatal toxoplasmosis); VP subependymoma of the aqueduct; VP</td>
<td>23.5</td>
<td>HA; shunt dysfunction w/ chronic hydrocephalus on CT</td>
<td>elective</td>
<td>shunt reimplemented after 7 mos</td>
<td>unremarkable</td>
</tr>
<tr>
<td>8</td>
<td>54, F</td>
<td>subependymoma of the aqueduct; VP subependymoma of the aqueduct; VP</td>
<td>3.6</td>
<td>postural HA, Parinaud syndrome; overdrainage w/ ventricular collapse on CT</td>
<td>elective, ventricular dilatation w/ ASD</td>
<td>40 mos unremarkable</td>
<td></td>
</tr>
<tr>
<td>9</td>
<td>25, F</td>
<td>aqueductal stenosis; VP</td>
<td>3</td>
<td>HA, vomiting, visual disturbance; shunt dysfunction w/ 3rd ventricle hydrocephalus on CT</td>
<td>elective</td>
<td>103 mos unremarkable</td>
<td></td>
</tr>
<tr>
<td>10</td>
<td>37, F</td>
<td>aqueductal stenosis; VP</td>
<td>2</td>
<td>vertigo, gait disturbance; shunt dysfunction w/ 3rd ventricle hydrocephalus on CT</td>
<td>elective</td>
<td>67 mos unremarkable</td>
<td></td>
</tr>
<tr>
<td>11</td>
<td>41, M</td>
<td>aqueductal stenosis; VP</td>
<td>1.1</td>
<td>vomiting, drop attacks, loss of consciousness; shunt dysfunction w/ 3rd ventricle hydrocephalus on MRI</td>
<td>emergency</td>
<td>102 mos mild occasional HA, vertigo</td>
<td>unremarkable</td>
</tr>
<tr>
<td>12</td>
<td>8, M</td>
<td>obstruction of 4th ventricle outlet; VP</td>
<td>0.7</td>
<td>vomiting, gait disturbance; shunt dysfunction w/ dilation of all ventricles on CT</td>
<td>emergency</td>
<td>83 mos unremarkable</td>
<td></td>
</tr>
<tr>
<td>13</td>
<td>40, F</td>
<td>aqueductal stenosis; VP</td>
<td>16</td>
<td>HA, loss of consciousness; SVS evolving to 3rd ventricle hydrocephalus on CT/MRI</td>
<td>emergency</td>
<td>42 mos unremarkable</td>
<td></td>
</tr>
<tr>
<td>14</td>
<td>38, M</td>
<td>aqueductal stenosis; VA</td>
<td>2</td>
<td>HA, neck rigidity; shunt dysfunction w/ 3rd ventricle hydrocephalus on CT</td>
<td>elective</td>
<td>79 mos unremarkable</td>
<td></td>
</tr>
<tr>
<td>15</td>
<td>39, M</td>
<td>aqueductal stenosis; VP</td>
<td>5</td>
<td>vertigo, somnolence, adynamia; 3rd ventricle hydrocephalus w/ multiple septations on CT/MRI</td>
<td>elective</td>
<td>83 mos mild occasional HA, vertigo</td>
<td>unremarkable</td>
</tr>
<tr>
<td>16</td>
<td>22, F</td>
<td>obstruction of 4th ventricle outlet after resection of cerebellar astrocytoma; VP</td>
<td>1.3</td>
<td>HA, somnolence; shunt dysfunction w/ dilation of all ventricles on CT/MRI</td>
<td>elective</td>
<td>51 mos unremarkable</td>
<td></td>
</tr>
<tr>
<td>17</td>
<td>32, F</td>
<td>aqueductal stenosis; VA</td>
<td>13</td>
<td>gait disturbance, vertigo; shunt dysfunction w/ 3rd ventricle hydrocephalus on CT/MRI</td>
<td>elective</td>
<td>shunt reimplemented after 2 wks†</td>
<td>unremarkable</td>
</tr>
</tbody>
</table>

* ASD = antisiphoning device; FU = follow up; HA = headache; MRI = magnetic resonance imaging; SP = shunt procedure.
† Insufficient third ventricular stoma.
the first ETV. It was not until the last two decades, however, that endoscopy gained widespread acceptance for the treatment of occlusive forms of hydrocephalus, especially if they were associated with aqueductal stenosis. This belated acknowledgment is due to the great progress that has been made in the refinement of endoscopic optics and in video technology during the last 20 years. As a result, nowadays we benefit from high-performance, yet small-sized video cameras. Thus, handling of the equipment has become much simpler and easier in the sterile environment of an operating theater, while simultaneously, image resolution and color performance have increased dramatically.

The use of percutaneous ventriculostomy in patients with hydrocephalus who have previously undergone shunt placement was first described by Sayers and Kosnik in 1976.44 They performed percutaneous third ventriculostomy under stereotactic and fluoroscopic guidance in 46 children with shunts. Prior to ventriculostomy, these children had undergone a total of 163 shunt-related operations. Subsequent to ventriculostomy, only three shunt revisions were necessary during follow up. In 1990, Jones, et al.,29 were the first to publish a series of patients treated with ETV, and they included 12 who had previously received shunts. Since then, various groups have reported on their experience. In Table 2 we have summarized the data available from the literature. Because most reports did not concentrate on ETV in patients who had previously received shunts, we analyzed the data for all patients in those reports and, whenever possible, we recalculated the numbers for those who had previously undergone shunt placement. When data were not stated directly in a paper, we were often able to recalculate or reconstruct them on the basis of the given facts. Analysis of the publications renders a rather nonuniform collection of single-center experiences with ETV. Although there are some publications in which large numbers of patients are included,16,17,26,28,50 the published data are not suitable for a metaanalysis.

Successes, Failures, and Abandoned Procedures

Overall, successful ETV in patients who had previously undergone shunt placement has been reported in 42 to 100% of patients in different studies. The rate of 82% of the adult population in our study who became shunt independent compares well with that of 76% described by Cinalli and colleagues8 in a pediatric population. There have been controversial findings regarding predictors for success or failure of ETV in the treatment of shunt malfunction. In some centers similarly good outcomes were not achieved in patients who had previously received shunts compared with those who had never undergone shunt placement.47,53 In others no difference was found between these groups.4,16 Moreover, there are reports in which previous shunt insertion is asserted to be a positive predictive factor for the success of ETV.26,50,51 This effect is most obvious in the data published by Teo and Jones51 in 1996 and by Teo50 in 1998. The patients in both of their studies were predominantly children. The authors gave a possible explanation for their findings: in general, children who had previously undergone shunt placement were older than those who received ETV as the first treatment for their hydrocephalus, and in older children the CSF resorption sites may be more mature than in younger ones. Additionally, the shunt may lower the pressure gradient between the ventricles and the subarachnoid space (transmantle pressure), thus allowing the subarachnoid space to open and again to mature. Both factors may lead to a lower risk for a malresorptive component in the origin of hydrocephalus in patients who had previously undergone
TABLE 2

<table>
<thead>
<tr>
<th>Authors &amp; Year</th>
<th>Total No.</th>
<th>Mean Age (range)</th>
<th>Mean FU (range)</th>
<th>No. W/ Successful ETV (%)</th>
<th>No. W/ ETV</th>
<th>Mean Age (range)</th>
<th>Presentation: UD/OD/Inf (other)</th>
<th>Mean Time Brunt SP &amp; ETV (range)</th>
<th>No. W/ Successful ETV (%)</th>
<th>Mean FU (range)</th>
<th>No. W/ Shunt Re- &amp; Removed (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Jones, et al., 2003</td>
<td>24†</td>
<td>8.4 yrs*</td>
<td>3.3 yrs*</td>
<td>15*</td>
<td>5*</td>
<td>12*</td>
<td>11.6 yrs*</td>
<td>NA</td>
<td>NA</td>
<td>10*</td>
<td>3.2 yrs*</td>
</tr>
<tr>
<td>Kelly, 1993</td>
<td>16</td>
<td>24 yrs</td>
<td>3.5 yrs</td>
<td>15*</td>
<td>4*</td>
<td>11</td>
<td>26 yrs</td>
<td>8/0/6</td>
<td>7.3 yrs*</td>
<td>9/0/31</td>
<td>100*</td>
</tr>
<tr>
<td>Dalrymple &amp; Kelly, 1992</td>
<td>85</td>
<td>NA</td>
<td>1–66 mos</td>
<td>25 mos</td>
<td>NA</td>
<td>27</td>
<td>4*</td>
<td>5 yrs*</td>
<td>(3–8 yrs)*</td>
<td>(3–14 yrs)*</td>
<td>100*</td>
</tr>
<tr>
<td>Jones, et al., 1993</td>
<td>95</td>
<td>NA</td>
<td>NA</td>
<td>54</td>
<td>(56.8)</td>
<td>36*</td>
<td>NA</td>
<td>NA</td>
<td>NA</td>
<td>NA</td>
<td></td>
</tr>
<tr>
<td>Yamamoto, et al., 1994</td>
<td>69</td>
<td>11 yrs</td>
<td>32 mos</td>
<td>50</td>
<td>4</td>
<td>55</td>
<td>NA</td>
<td>NA</td>
<td>NA</td>
<td>NA</td>
<td></td>
</tr>
<tr>
<td>Goumnerova &amp; Frim, 1992</td>
<td>23</td>
<td>11.2 yrs</td>
<td>17 mos</td>
<td>16</td>
<td>13*</td>
<td>4</td>
<td>25 yrs*</td>
<td>4/0/0</td>
<td>3*</td>
<td>(75)*</td>
<td></td>
</tr>
<tr>
<td>Baskin, et al., 1998</td>
<td>97</td>
<td>8.1 yrs</td>
<td>24.2 mos</td>
<td>35</td>
<td>18</td>
<td>36*</td>
<td>NA</td>
<td>NA</td>
<td>NA</td>
<td>NA</td>
<td></td>
</tr>
<tr>
<td>Cinalli, et al., 1999</td>
<td>38</td>
<td>57 mos</td>
<td>NA</td>
<td>11</td>
<td>0/2</td>
<td>11</td>
<td>8.7 yrs</td>
<td>3/0/0</td>
<td>10.8 mos*</td>
<td>6/0/0</td>
<td>10</td>
</tr>
<tr>
<td>Hopf, et al., 2000</td>
<td>95</td>
<td>30.5 yrs</td>
<td>24 mos</td>
<td>60</td>
<td>NA</td>
<td>25</td>
<td>NA</td>
<td>NA</td>
<td>NA</td>
<td>NA</td>
<td></td>
</tr>
<tr>
<td>Fukuhara, et al., 2000</td>
<td>89</td>
<td>3.8 yrs</td>
<td>3–6 mos</td>
<td>67 (4.7)</td>
<td>6/0/0</td>
<td>32</td>
<td>NA</td>
<td>NA</td>
<td>NA</td>
<td>NA</td>
<td></td>
</tr>
<tr>
<td>Hayashi, et al., 2000</td>
<td>10</td>
<td>NA</td>
<td>NA</td>
<td>9</td>
<td>5*</td>
<td>4</td>
<td>NA</td>
<td>NA</td>
<td>NA</td>
<td>NA</td>
<td></td>
</tr>
<tr>
<td>Murshed, et al., 2000</td>
<td>27</td>
<td>3.9 yrs</td>
<td>24 mos</td>
<td>22</td>
<td>14*</td>
<td>9*</td>
<td>3.3 yrs*</td>
<td>9/0/7</td>
<td>(88.8)*</td>
<td>8*</td>
<td>11*</td>
</tr>
<tr>
<td>Scarfow, et al., 2000</td>
<td>54</td>
<td>10.8 yrs</td>
<td>13 mos</td>
<td>40/6</td>
<td>32/0</td>
<td>14</td>
<td>NA</td>
<td>NA</td>
<td>NA</td>
<td>NA</td>
<td></td>
</tr>
<tr>
<td>Tisell, et al., 2000</td>
<td>18</td>
<td>48 yrs</td>
<td>NA</td>
<td>9</td>
<td>3*</td>
<td>36.3 yrs*</td>
<td>2/10</td>
<td>NA</td>
<td>NA</td>
<td>NA</td>
<td>NA</td>
</tr>
<tr>
<td>Buxton, et al., 2001</td>
<td>63</td>
<td>37.5 yrs</td>
<td>3.1 yrs</td>
<td>31</td>
<td>30*</td>
<td>24</td>
<td>NA</td>
<td>NA</td>
<td>NA</td>
<td>NA</td>
<td>19*</td>
</tr>
<tr>
<td>Present study</td>
<td>17</td>
<td>32 yrs</td>
<td>(8–54 yrs)</td>
<td>15/0/2</td>
<td>8.4 yrs</td>
<td>4/1</td>
<td>14 (20 mos–35 yrs)</td>
<td>63.6 mos</td>
<td>14</td>
<td>(3–8 yrs)</td>
<td></td>
</tr>
</tbody>
</table>

* Not shown in original publication, but recalculated according to the given data. The ETV was considered successful if patients remained shunt independent postprocedure. Abbreviations: gwks = gestational weeks; inf = infection; IS = intact; lig = ligated; med = median; NA = not available (recalculation not possible: insufficient data); OD = overdrainage; res = reservoir; UD = underdrainage.
† In four of these patients ETV was abandoned; two had previously received shunts.
‡ All ventriculostomies were performed stereotactically under endoscopic guidance.
§ The ETV was combined with aqueductoplasty in two of the three cases.
|| Only patients with myelomeningocele were included. Shunts were removed only if placed frontally (two cases), and were ligated if their status was questionable.
|| All ventriculostomies were performed under fluoroscopic control, and in the remaining 23 it was done endoscopically.
†† In seven patients third ventriculostomy was performed under fluoroscopic control, and in 11 it was done endoscopically.
‡‡ Interval between last shunt placement and ETV.
§§ All patients in this study had previously received shunts; seven did not undergo ETV.
|| All patients in this study had previously received shunts; seven did not undergo ETV.
Endoscopic third ventriculostomy for shunt dysfunction

It remains unclear whether a longer interval between shunt placement and ETV yields a higher risk for subsequent failure of ETV.

Different authors have reported that they had to abandon ETV sometimes, for a variety of reasons (for example, distorted or unsuitable anatomy, blurred vision, bleeding). A systematic analysis of these events, however, is not possible due to insufficient data.

Fukuhara and colleagues noted a history of shunt infection at presentation, three or more previous shunt revisions, and post-ETV meningitis were independent risk factors for the failure of ETV. These findings are in accordance with our experience: two of our ETV failures occurred in patients who had a history of intracranial infection. It is possible that patients with apparently primarily occlusive hydrocephalus who do not respond to ETV might have an additional malresorptive component. It is noteworthy, however, that ETV has also been successfully used to cure recurrent shunt infections.

Removal of Shunt Hardware

Keeping in mind that infection is one of the major risks in patients who receive shunts, we do not find it useful to perform an ETV without also removing the shunt. Foreign material is always prone to infection. Moreover, it has been argued that the intermittent or remaining flow through a malfunctioning shunt may cause reduced flow through the stoma itself, and consequently may promote its closure. Hence, we believe that all shunt hardware should be removed after ETV whenever possible. In patients in whom a shunt system has been present for a long period, however, removal may seem too risky and is sometimes not feasible. The remaining shunt should then at least be ligated in such cases. We were able to remove the shunt systems completely in all of our patients, either in the same ETV session or during a second session after ETV had proven to be effective. Presently, we prefer to remove the shunt hardware directly after ETV and insert an occluded external ventricular drain, which remains in place for some days.

Slit Ventricle Syndrome

A rare but extremely vexing condition for both patients and physicians, SVS is very difficult to handle. Because it is entirely secondary to the placement of a ventricular shunt, a protocol that includes shunt removal and an alternative pathway for intracranial CSF diversion is most feasible, particularly in these patients. Although small or slit ventricles may not seem to be quite apt for endoscopic procedures, Baskin and coworkers showed encouraging results with ETV even under such circumstances. They included 22 patients in a protocol that required ICP monitoring after shunt removal, or externalization and blocking of shunt systems. Sixteen patients who demonstrated a need for further CSF drainage thereafter underwent ETV. Ten (62.5%) of these 16 patients were no longer shunt dependent during a mean follow-up period of 18.8 months (range not stated). Treatment failures occurred in two patients with congenital aqueductal stenosis, in two other patients with tumors, in one with intraventricular hemorrhage, and in one with hydrocephalus related to myelomeningocele.

Patients suffering from SVS tend to be very sensitive toward elevated ICP. After removal or occlusion of their shunt they are at high risk of becoming seriously ill. Therefore, a more gradual way to increase ventricle size prior to endoscopy would be preferable. In both of our patients with SVS we implanted an antisiphoning device in which the resistance was purposely selected to be relatively too high. This made the ventricles grow slowly in size. Instead of developing symptoms caused by intracranial hypertension, both patients improved, and the symptoms caused by overdrainage were no longer present. Because of the long history of shunt dysfunction, the request of the patients to become shunt independent, and our previous experience, however, we decided to perform ETV as planned earlier. The ventricles were sufficiently large, and ETV could be performed without additional risks. Both patients remain free of their hydrocephalic and shunt-related symptoms to date. Alternatively, when a programmable valve has been implanted, the valve setting could be switched to a higher opening pressure to induce ventricular dilation prior to endoscopy.

Complications of ETV

There were no complications associated with ETV in our series. Although endoscopy seems to be performed with ease in general, it can be difficult and hazardous in certain situations. This holds true particularly for patients previously treated with shunts, in whom the typical anatomy can be distorted and the landmarks are not readily recognizable. Recently, Schroeder, et al., published a detailed analysis of the complications associated with ETV. Several authors have stressed the steep learning curve of neuroendoscopy and reported substantial, sometimes even life-threatening complications. In series in which both patients with and those without previous shunts were included, complication rates often were not detailed with regard to whether patients had shunts in place or not.

It has been stated over the decades that long-term diversion of CSF will result in the reduction or even the extinction of CSF resorption capacity. The dogma “once a shunt, always a shunt” was based on such pathophysiological hypotheses. These hypotheses, however, are nullified by the experience of successful ETV performed in patients who have previously undergone shunt placement. Many patients have become shunt independent after living for years, sometimes even for more than a decade, with a shunt in place. It has been argued that the CSF absorption sites could have been reopened. It is much more likely, however, that the CSF resorption sites were never really occluded in these patients.

Conclusions

In experienced hands ETV is a safe and effective alternative for the treatment of shunt dysfunction in patients with occlusive hydrocephalus. It can be performed as an emergency operation, and should be the treatment of choice in all such cases. Even in the difficult situation of SVS, ETV can be the definitive remedy. The long-held dogma “once a shunt, always a shunt” is no longer valid.
References


J. Boschert, D. Hellwig, and J. K. Krauss
Endoscopic third ventriculostomy for shunt dysfunction


Address reprint requests to: Jürgen Boschert, M.D., Neurochirurgische Universitätsklinik, Universitätsklinikum Mannheim, Theodor-Kutzer-Ufer 1-3, D-68167 Mannheim, Germany. email: juergen.boschert@nch.ma.uni-heidelberg.de.