Endoscopic aqueductoplasty and placement of a stent in the cerebral aqueduct in the management of isolated fourth ventricle in children

GIUSEPPE CINALLI, M.D., PIETRO SPENNATO, M.D., LUCIANO SAVARESE, M.D., CLAUDIO RUGGIERO, M.D., FERDINANDO ALIBERTI, M.D., LORENZO CUOMO, M.D., EMILIO CIANCIULLI, M.D., AND GIUSEPPE MAGGI, M.D.

Departments of Pediatric Neurosurgery and Pediatric Neuroradiology, Santobono Children’s Hospital, Naples, Italy

Object. In this study the authors conducted a retrospective evaluation of the effectiveness of endoscopic aqueductoplasty, performed alone or accompanied by placement of a stent, in the treatment of an isolated fourth ventricle (IFV) in seven patients afflicted with loculated hydrocephalus after a hemorrhage or infection.

Methods. Seven children with symptomatic IFV and membranous aqueductal stenosis underwent endoscopic aqueductoplasty alone or combined with placement of a stent in the cerebral aqueduct. The mean age of the patients at the time of surgery was 10 months. The mean duration of follow up was 26 months. In all patients a supratentorial shunt had already been implanted, and in five patients neuroendoscopy had already been performed because other isolated compartments had been present inside the ventricular system. Aqueductoplasty alone was performed in three patients and aqueductoplasty and aqueductal stent placement in four. A precoronal approach was performed in five patients and a suboccipital approach in two. Signs and symptoms of intracranial hypertension resolved in all cases. Stent placement was successful in all five cases, resulting in clinical and neuroimaging-confirmed improvements in the IFV. Restenosis of the aqueduct occurred in two patients in whom stents had not been placed. In one of these patients restenosis was managed by an endoscopic procedure, during which the aqueduct was reopened and a stent implanted; in the other patient a shunt was placed in the fourth ventricle. Hydrocephalus was controlled by a single shunt in six cases (86%) and by a double shunt in one case.

Conclusions. Endoscopic placement of a stent in the aqueduct is more effective in preventing the repeated occlusion of the aqueduct than aqueductoplasty alone and should be indicated as the initial treatment in each case of compatible anatomy.

KEY WORDS • hydrocephalus • isolated fourth ventricle • aqueductal stenosis • neuroendoscopy • aqueductoplasty • pediatric neurosurgery

ISOlatioN of the fourth ventricle usually occurs as the result of an obstruction of CSF at the level of the cerebral aqueduct in patients in whom a shunt has already been placed for postmeningitic or posthemorrhagic hydrocephalus.3,7,8,12,16,18 Although in some cases IFV is caused by overdrainage of the supratentorial shunt and can be considered reversible with correction of this overdrainage,13 most forms are irreversible (due to inflammatory changes) and, if symptomatic, require surgical treatment.5,18 Several treatment options have been described, including the following: placement of a shunt in the fourth ventricle;6,8,12,16 microsurgical aqueduct canalization;3 microsurgical fenestration of the outlets of the fourth ventricle;1,21 and endoscopic procedures.5,17,20 Placement of a shunt in the fourth ventricle is the most commonly used procedure, even though it is associated with a high rate of dysfunction and dislocation.18 Moreover, it exposes patients to the risks of several complications directly related to fourth ventricular shunt insertion, such as new cranial nerve dysfunction and embedding of the catheter tip in the floor of the fourth ventricle.2,9 Overdrainage from the fourth ventricle may be responsible for obstructions of the catheter, paralysis of cranial nerves,19 and secondary penetration of the catheter tip in the floor of the fourth ventricle following a reduction in the size of the ventricle.2,9

Endoscopic reopening of the aqueduct (aqueductoplasty), may be considered the ideal treatment of IFV, because it reestablishes communication between the fourth ventricle and the supratentorial ventricular system, equilibrating the transtentorial pressure and allowing a single supratentorial shunt to control the patient’s hydrocephalus. Endoscopic aqueductoplasty cannot be performed in all cases, however; it can only be performed if a membranous occlusion of the aqueduct is present on preoperative MR images. The standard precoronal approach can be used only in cases in
which the supratentorial ventricular system is also dilated; in other cases a suboccipital approach should be used. We report our experience in the treatment of seven children affected by an IFV, in which endoscopic aqueductoplasty was performed as an alternative to placement of a shunt in the fourth ventricle.

Clinical Material and Methods

Between September 1999 and December 2004 at Santobono Children’s Hospital, Naples, Italy, seven children affected by symptomatic IFV underwent neuroendoscopic procedures during which endoscopic aqueductoplasty was performed alone or was accompanied by placement of a stent in the aqueduct. We retrospectively reviewed the charts of all these patients and the pertinent data we obtained are listed in Table 1. All patients, who ranged in age between 3 months and 2 years, had been afflicted with postmeningitic or posthemorrhagic hydrocephalus at birth and had received shunts for their conditions. Five children had already undergone endoscopic treatment for the presence of septations inside the supratentorial ventricular system (three cases) or isolation of one lateral ventricle (two cases). The IFVs in these patients developed at different times after the shunts had been implanted (mean 12 months postimplantation; range 1–17 months) and were discovered during routine follow-up CT studies. In all cases the serial CT scans demonstrated enlargement of the fourth ventricle. In two patients symptoms and signs of increased intracranial pressure developed despite the fact that the supratentorial ventricles were small; in one patient the IFV was considered symptomatic because the patient did not progress to expected developmental milestones. In four cases the IFV became symptomatic in children in whom shunts had to be externalized because of infection: three children became lethargic and one, a 3-month-old baby, suffered respiratory distress that required treatment in a neonatal intensive care unit.

Preoperative MR imaging, performed in all patients, revealed membranous occlusion of the aqueduct. In one case IFV was associated with the presence of septations inside the fourth ventricle.

Surgical Technique

We performed aqueductoplasty through two different endoscopic approaches. If the supratentorial ventricular system was large enough to admit the neuroendoscope, a precoronal approach was selected; in all other cases a suboccipital approach was preferred. All the procedures were performed using a 3.5-mm rigid fiberscope (Channel Neuroendoscope; Medtronic, Inc., Minneapolis, MN) after general anesthesia had been induced in the patient.

Precoronal Approach. Our technique is similar to the one introduced by Schroeder and Gaab as an alternative to third ventriculostomy for the treatment of hydrocephalus caused by short-segment aqueductal stenosis (< 5 mm) and

TABLE 1

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Age (mos)</th>
<th>Cause of IFV</th>
<th>Shunt History</th>
<th>Previous Endoscopy</th>
<th>Presentation</th>
<th>Surgery</th>
<th>Results</th>
<th>Follow Up (mos)</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>22</td>
<td>PMH</td>
<td>VP shunt (10 shunt revisions), EVD</td>
<td>no</td>
<td>lethargy, shunt infection</td>
<td>aqueductoplasty via suboccip approach; fenestration of septations inside the 4th ventricle</td>
<td>restenosis (25 days); Y-shaped shunt placed in 4th ventricle &amp; connected to supratent (2 revisions)</td>
<td>50</td>
</tr>
<tr>
<td>2</td>
<td>16</td>
<td>PMH</td>
<td>VP shunt (12 shunt revisions), EVD</td>
<td>intrav septa fenestration</td>
<td>lethargy, vomiting, shunt infection</td>
<td>1) aqueductoplasty via precoronal approach; 2) aqueductoplasty &amp; stent placement</td>
<td>1) early restenosis (16 days); 2) good single VP shunt placed, no shunt revision signs resolved, single VP shunt placed, no shunt revision</td>
<td>62</td>
</tr>
<tr>
<td>3</td>
<td>8</td>
<td>PHH &amp; PMH</td>
<td>VP shunt (3 shunt revisions), double EVD (supratent &amp; 4th ventricle)</td>
<td>septostomy</td>
<td>lethargy, vomiting, shunt infection</td>
<td>aqueductoplasty &amp; stent placement</td>
<td>aqueductoplasty &amp; stent placement via precoronal approach</td>
<td>33</td>
</tr>
<tr>
<td>4</td>
<td>7</td>
<td>PHH</td>
<td>VP shunt (4 shunt revisions)</td>
<td>intrav septa fenestration</td>
<td>lethargy, vomiting</td>
<td>aqueductoplasty &amp; stent placement via precoronal approach</td>
<td>signs resolved, single VP shunt placed (catheter through aqueduct shunted to peritoneum), no shunt revision</td>
<td>19</td>
</tr>
<tr>
<td>5</td>
<td>5</td>
<td>PHH &amp; PMH</td>
<td>VP shunt (1 shunt revision)</td>
<td>intrav septa fenestration</td>
<td>lethargy, vomiting</td>
<td>aqueductoplasty &amp; stent placement via suboccip approach</td>
<td>signs resolved, single VP shunt placed, no shunt revision</td>
<td>14</td>
</tr>
<tr>
<td>6</td>
<td>12</td>
<td>PHH</td>
<td>VP shunt</td>
<td>septostomy</td>
<td>shunt malfunction, diplopia, developmental delay</td>
<td>aqueductoplasty &amp; stent placement &amp; fenestration of supratent loculations via precoronal approach</td>
<td>signs resolved, single VP shunt placed (catheter through aqueduct shunted to peritoneum), no shunt revision</td>
<td>3</td>
</tr>
<tr>
<td>7</td>
<td>3</td>
<td>PHH &amp; PMH</td>
<td>VP shunt (1 shunt revision), EVD</td>
<td>no</td>
<td>shunt infection, respiratory distress</td>
<td>aqueductoplasty &amp; stent placement via precoronal approach</td>
<td>signs resolved, single VP shunt placed (catheter through aqueduct shunted to peritoneum), no shunt revision</td>
<td>3</td>
</tr>
</tbody>
</table>

* EVD = external ventricular drainage; intrav = intraventricular; PHH = posthemorrhagic hydrocephalus; PMH = postmeningitic hydrocephalus; suboccip = suboccipital; supratent = supratentorial.
to the techniques used by Teo and colleagues and Fritsch and associates for the management of an IFV. Briefly, the patient is positioned supine with his or her head lying in a horseshoe-shaped headrest. A bur hole is created just behind the hairline, at least 4 cm anterior to the coronal suture and over the midpupillary line. The right lateral ventricle is cannulated with a peel-away sheath. The endoscope is then inserted and a careful inspection of the frontal horn usually allows easy identification of the foramen of Monro (Fig. 1 left). In cases of multiloculated hydrocephalus, the anatomy of the lateral ventricle can be dramatically modified and significant attention should be paid to the safe identification of two anatomical landmarks, the foramen of Monro and the third ventricle (Fig. 1 right). The endoscope is advanced through the foramen of Monro (Fig. 2 upper left) into the third ventricle, where the obstructed aqueduct inlet is iden-

![Fig. 1. Left: Magnetic resonance image demonstrating the tip of the endoscope, which lies in the lateral ventricle and is aimed at the posterior half of the third ventricle. Right: Endoscopic view of the right foramen of Monro and the posterior portion of the third ventricle. Labeling for Figs. 1 through 5: 1, fornic; 2, septal vein; 3, thalamostriate vein; 4, mamillary bodies; 5, interthalamic adhesion (massa intermedia); 6, third ventricle floor; 7, aqueduct inlet; 8, posterior commissure; 9, suprapineal recess; 10, ventricular catheter used for the stent; 11, Fogarty balloon catheter; 12, endoscope.](image1)

![Fig. 2. Magnetic resonance image (upper left) and endoscopic views (upper right and lower left and right). Upper Left: The endoscope is advanced into the third ventricle and stopped in front of the aqueduct entrance. Upper Right: View of the interthalamic adhesion (massa intermedia) and the suprapineal recess. Lower Left: As the endoscope is advanced, the aqueduct comes into view behind the interthalamic adhesion (massa intermedia). Lower Right: The inlet of the aqueduct occluded by a membrane.](image2)
tified. The aqueduct should not be confused with the suprapineal recess, which is located above the aqueduct and behind the posterior commissure (Fig. 2 upper right, lower left and right). The membrane occluding the inlet of the aqueduct is usually broken by simply probing the aqueduct with the aid of the smooth tip of a Fogarty balloon catheter (Fig. 3); this allows visualization of the cavity of the fourth ventricle. After the membrane has been perforated, the endoscope is withdrawn into the lateral ventricle and a multiperforated ventricular catheter is inserted into the lateral ventricle (Fig. 4). We prefer to insert this multiperforated catheter through a second bur hole created 1 cm away from the initial bur hole. In fact, if the catheter is inserted into the same track used for the endoscope, the catheter can be placed in conflict with the endoscope itself and can be pushed along an axis other than the optic axis of the endoscope, if a 0° optic is used, making it difficult to visualize the catheter and driving it into the aqueduct. The tip of the ventricular catheter is observed within the lateral ventricle with the aid of the endoscope, and the catheter is first advanced into the third ventricle and then into the aqueduct. After the tip of the catheter has been introduced into the aqueduct, the stylet is removed to minimize the risk of harming periaqueductal structures with a semirigid instrument, and the catheter is advanced to the desired distance into the fourth ventricle. The endoscope is then withdrawn while checking the surface of the catheter to ensure the presence of additional holes into the third or lateral ventricle (Fig. 5). After the endoscope has been removed, the catheter is connected to a subcutaneous reservoir (if a working shunt is already present) or to a VP shunt. The position of the catheter is easily verified on a postoperative CT scan (Fig. 6).

In two cases the catheter was left in place for use as a stent without connecting it to an extracranial draining sys-

**Fig. 3.** Upper: Magnetic resonance image demonstrating how the membrane obstructing the inlet of the aqueduct is perforated using the smooth tip of a No. 3 French Fogarty balloon catheter. Inflation of the balloon is seldom necessary and should be avoided if stent placement is planned. Lower: Endoscopic view of a noninflated No. 3 French Fogarty balloon perforating the loose membrane occluding the aqueduct.

**Fig. 4.** Upper: Magnetic resonance image. After the catheter has been withdrawn into the lateral ventricle, it is inserted through a separate bur hole and can be observed in the third ventricle. Lower: Endoscopic view of the ventricular catheter inserted through a separate bur hole into the foramen of Monro.
tem; this method was used when a supratentorial shunt was already in place and working properly without the need of revision.

**Suboccipital Approach.** The patient is placed prone with his or her head lying in a horseshoe-shaped headrest; the head is flexed as much as possible to create the best trajectory to the aqueduct. An infratentorial bur hole is drilled 2 to 3 cm from the midline over the right cerebellar hemisphere. The dilated fourth ventricle is cannulated with the aid of a peel-away sheath and then the endoscope is inserted. After the aqueduct has been identified, the obstructing membrane is perforated using a Fogarty balloon catheter while carefully avoiding any damage to the periaqueductal gray matter. The inspection of the aqueduct permits the operator to visualize the third ventricle. In one case multiple membranes were fenestrated. The limit of this approach is the lack of landmarks available for orientation in the unusual and distorted anatomy of a trapped fourth ventricle, where recognition of the inlet of the aqueduct can be very difficult.

**Results**

The follow-up period ranged from 3 to 62 months (mean 26 months). Details of symptoms at presentation are listed in Table 1. The endoscopic procedure was uneventful in all cases. Postoperatively, one patient experienced dysconjugate ocular movements, but these completely resolved by 3 weeks after surgery.

Signs and symptoms of raised intracranial pressure resolved in all cases following the operations. In five cases, in which the supratentorial ventricular system was also dilated, a precoronal approach was used. In two of these cases fenestration of intraventricular supratentorial septations was also performed. In two cases aqueductoplasty was performed through a suboccipital approach; in one of these cases, septations inside the fourth ventricle were also fenestrated during the same endoscopic procedure.

Aqueductoplasty alone was performed in three cases and aqueductoplasty followed by placement of a stent in four cases (Fig. 3). In two patients in whom stents had not been placed, symptoms recurred 10 and 21 days, respectively, after the procedures and MR images demonstrated restenosis of the aqueduct. Both patients had undergone multiple shunt revisions before endoscopy because of shunt infection. One patient was treated with a Y-shaped shunt, which was placed in the fourth ventricle and connected to the supratentorial shunt; the other patient was treated by endoscope-aided reopening of the aqueduct and placement of a stent. Thereafter, stents were placed in five patients (in four as the initial procedure and in one following closure of the aqueductoplasty). In two patients the stents consisted of a “cut” ventricular catheter, and in three cases an “uncut” ventricular catheter was inserted and connected to a valve and a peritoneal catheter, constituting the only shunt system in these patients.
Stent placement appeared to have been successful in all cases; this was reflected in both clinical and neuroimaging-confirmed improvements in IFV, even though in two cases the duration of follow up was too short for a fair estimate. Hydrocephalus was controlled by a single shunt in six cases (86%) and by a double shunt in one case.

Discussion

The goal of treatment of an IFV is to equalize transtentorial pressure by draining the excess CSF trapped in the fourth ventricle. Management strategies for the treatment of trapped fourth ventricle have been reviewed by Harter in a recent paper. Among the therapeutic options—placement of a shunt in the fourth ventricle, open surgery, and endoscopy—endoscopic aqueductoplasty performed in cases of compatible anatomy (in which there is a short segment, or < 5 mm of aqueductal stenosis) seems to be the ideal treatment of this rare condition, because it restores a more physiological CSF pathway and eliminates the need for placement of multiple extracranial ventricular shunt catheters and the risk associated with open posterior fossa surgery. Despite these findings, Oi and associates performed two endoscopic aqueductoplasties, one via the rostral approach and the other through the caudal approach, with poor results. Better results have been achieved by Teo and colleagues. They performed four aqueductoplasties (one through the suboccipital approach) and in three operations they did not insert a stent. These authors observed two cases of restenosis in their group, which they managed with repeated aqueductoplasty and stent implantation. Fritsch, et al., treated 13 patients with endoscopic aqueductoplasty; there were no treatment failures in patients in whom a stent was placed, whereas among the eight patients in whom aqueductoplasty was performed alone, six experienced restenosis (Table 2). Our results confirm these data, indicating that aqueductoplasty should always be followed by placement of a stent, even if the patient has a history of CSF infection. This appears to be appropriate because in patients affected by IFV, all of whom have a history of intraventricular hemorrhage or meningitis, it is very unlikely that an aqueductoplasty will remain patent if no shunt is present. It is always preferable to connect the stent to a preexisting shunt system or to some subcutaneous reservoir to allow easy removal in case of infection.

The choice of both the approach and the stent design is dictated mainly by the size of the ventricular system and by the eventual need of revision of the supratentorial shunt. The prerequisite of the precoronal approach is sufficient dilation of at least one lateral ventricle with an open foramen of Monro and the third ventricle. If the ventricles are slit, they can be dilated by upgrading the valve opening pressure (in cases of externally adjustable valves) or by externalizing the distal catheter and slowly elevating the CSF drainage bag. Not all patients tolerate this procedure, however, because symptoms can occur before sufficient dilation of the ventricles is accomplished (Fig. 7).

### TABLE 2

<table>
<thead>
<tr>
<th>Authors &amp; Year</th>
<th>No. of Cases</th>
<th>Aqueductoplasty Alone</th>
<th>Restenosis</th>
<th>Aqueductoplasty &amp; Stent Placement</th>
<th>Failure</th>
</tr>
</thead>
<tbody>
<tr>
<td>Oi, et al., 1999</td>
<td>2</td>
<td>2</td>
<td>2/2</td>
<td>1 (initial procedure)</td>
<td>0/3</td>
</tr>
<tr>
<td>Fritsch, et al., 2004</td>
<td>13</td>
<td>8</td>
<td>6/8</td>
<td>5 (initial procedure)</td>
<td>0/9</td>
</tr>
<tr>
<td>present series</td>
<td>7</td>
<td>3</td>
<td>2/3</td>
<td>4 (initial procedure)</td>
<td>0/5</td>
</tr>
</tbody>
</table>

Fig. 7. Proposed algorithm of management of an IFV in children.
Endoscopic aqueductoplasty in patients with IFV

A suboccipital approach can be used when slit ventricles do not allow navigation in the supratentorial system. This approach has rarely been performed because of the higher risk of severe consequences if a hemorrhagic complication occurs and because the rarity of the condition makes this procedure less natural even for experienced neuroendoscopists. Nevertheless, slitlike lateral and third ventricles with an abnormally large fourth ventricle constitute by far the most frequently encountered condition in everyday clinical practice, and aqueductoplasty performed through a suboccipital approach appears to be the most logical and appealing procedure. With the rapid spread of neuroendoscopy associated with neuronavigation, this latter technique has the potential to become the most widely used surgical procedure for a trapped fourth ventricle. Neuronavigation is extremely helpful for placing the bur hole on the ideal trajectory into the axis of the aqueduct to be cannulated, so that the risk of entering the periaqueductal gray matter can be minimized. It is also very helpful for identification of the inlet of the aqueduct, which can be difficult to recognize in the unusual and distorted anatomy of a trapped fourth ventricle. Finally, solid training in more basic neuroendoscopic procedures is preferable before attempting any aqueductoplasty procedure and aqueductal stent placement, given the attendant serious potential complications.

Conclusions

In precoronal surgery and in special situations in which the suboccipital approach is used, endoscopic aqueductoplasty followed by placement of a stent in the aqueduct appears to be a safe and effective method of restoring the CSF pathway in cases of IFV and should be indicated as the initial treatment whenever the anatomy appears suitable on preoperative MR images.

References


Address reprint requests to: Giuseppe Cinalli, M.D., Via Gennaro Serra No. 75, 80132 Naples, Italy. email: giuseppe.cinalli@fastwebnet.it.