Ventriculocystostomy and endoscopic third ventriculostomy/shunt placement in the management of hydrocephalus secondary to giant retrocerebellar cysts in infancy

Report of 3 cases


1Department of Neurosurgery, The Royal Melbourne Hospital, Melbourne, Australia; 2Department of Neurosurgery, Children’s Hospital, University of California, San Francisco, and Children’s Hospital, Oakland, California; and Divisions of 3Pathology and 4Neurosurgery, The Hospital for Sick Children, University of Toronto, Canada

Hydrocephalus secondary to giant retrocerebellar cysts in infancy is a challenging condition and many treatment options exist. The authors report on 3 consecutive cases involving infants under the age of 6 months treated successfully with ventriculocystostomy in combination with direct hydrocephalus treatment (endoscopic third ventriculostomy or shunt placement). They describe the operative procedure, the surgical morbidity, and outcome in each case and review the literature regarding surgical approaches to this condition. (DOI: 10.3171/2009.10.PEDS09208)

KEY WORDS • retrocerebellar cyst • Blake pouch cyst • hydrocephalus • pediatric neurosurgery • ventriculocystostomy

Case Reports

We identified 3 cases of giant retrocerebellar cysts and hydrocephalus in infants treated consecutively with ventriculocystostomy and ETV/shunt placement between July 2007 and August 2008.

Case 1

This 3-month-old patient presented with rapidly increasing head circumference and clinical evidence of raised intracranial pressure. Magnetic resonance imaging revealed a giant retrocerebellar cyst with hydrocephalus (Fig. 1). The cerebral aqueduct appeared patent, and there were no other CNS abnormalities.

We performed an endoscopic cyst fenestration into the lateral ventricle and ETV, both via a left frontal ap-
proach, using intraoperative ultrasound, BrainLab neuronavigation, and the DORO Headrest System (Pro Med Instruments) for fixation. The patient made an uneventful recovery and was well at follow-up 20 months postoperatively when MR imaging demonstrated that the ventricular system and the cyst were significantly reduced in size (Fig. 1). The head circumference measured 45 cm (> 98th percentile) at 3 months of age and 51 cm (at 98th percentile) at 2 years of age.

Case 2

This child was diagnosed with a retrocerebellar cyst on antenatal ultrasound. There was no evidence of symptomatic hydrocephalus until the child was 4 months of age when she developed progressive macrocephaly, a bulging anterior fontanel and so-called sun-setting eyes. Repeated MR imaging revealed progressive hydrocephalus in the setting of a giant retrocerebellar cyst (Fig. 2). The cerebral aqueduct appeared patent, and there were no other CNS abnormalities.

We initially performed a posterior fossa craniotomy and open fenestration of the cyst into the basal cisterns. Pathological evaluation identified an epithelial-lined cyst with the morphological and immunohistochemical profile of choroid plexus (Fig. 3). Because of wound CSF leak, a right occipital medium-pressure VP shunt was placed 19 days after the original procedure. The child made an uneventful recovery and was discharged from the hospital.

The patient presented 2 months later with increasing lethargy, and MR imaging revealed adequate decompression of the ventricles but enlargement of the posterior fossa cyst. This time, we performed an endoscopic fenestration of the cyst into the lateral ventricle from a left frontal entry point, using intraoperative ultrasound, BrainLab neuronavigation, and the DORO Headrest System. Postoperative MR imaging performed after 17 months showed a decrease

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**Fig. 1.** Case 1. Preoperative sagittal (A), coronal (B) and 20-month postoperative sagittal (C) FIESTA MR images.

**Fig. 2.** Case 2. A: Sagittal FIESTA MR image obtained before open cyst fenestration and VP shunt insertion. B and C: Sagittal FIESTA and coronal T2-weighted MR image acquired before endoscopic ventriculocystostomy. D and E: Postoperative sagittal FIESTA MR images obtained at 4 and 17 months.
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in both ventricle and cyst size (Fig. 2). The head circumference measured 48 cm (> 98th percentile) at 4 months of age and 50.5 cm (just < 98th percentile) at 2 years of age. The child continues to achieve normal developmental milestones at 22 months since endoscopic fenestration.

Case 3

This 9-week old child was being followed for an antenatally diagnosed posterior fossa cyst and subsequently developed macrocephaly and a bulging fontanel. Magnetic resonance imaging confirmed progressive hydrocephalus and a giant retrocerebellar cyst (Fig. 4). The cerebral aqueduct appeared patent, and there were no other CNS abnormalities.

We performed an endoscopic fenestration of the cyst into the lateral ventricle assisted by intraoperative ultrasound, BrainLab neuronavigation, and the DORO Headrest System. An ETV was not performed. At surgery, there was grossly visible evidence of thinned-out choroid plexus tissue incorporated into the cyst wall. The patient initially did well but returned 2 months after the original procedure, presenting with irritability, vomiting, and a bulging fontanel. Brain CT scanning demonstrated evidence of progressive enlargement of the ventricles and posterior fossa cyst. After performing a frontal endoscopic approach we observed that the ventriculocystostomy was still widely patent. Therefore, an ETV was performed. The patient made an uneventful recovery and was discharged from the hospital. At 21 months, MR imaging revealed a reduction in the size of both the ventricles and the posterior fossa cyst (Fig. 4). Twenty-one months after the second operation, the patient remained clinically well, with a soft fontanel. The head circumference measured 42 cm (> 98th percentile) at 1 month of age and 49 cm (75th percentile) at 2 years of age.

Discussion

Classification of Retrocerebellar Cysts

Posterior fossa cysts can be difficult to classify, and to be inclusive, we have referred to the cysts in our cases by the generic term “retrocerebellar cyst.” In general, one can subcategorize posterior fossa cysts as either part of the broad Dandy-Walker complex or arachnoid cysts. The Dandy-Walker complex includes Dandy-Walker malformation/variant, Blake pouch cysts, and mega cisterna magna. The Dandy-Walker malformation is believed to be due to dysgenetic development of the roof of the rhombencephalon. A Blake pouch cyst is described as the persistence of the posterior midline evagination (area membranacea caudalis) of the roof of the embryonic fourth ventricle, which would normally thin out and rupture to form the foramen of Magendie. The cyst may or may not communicate with the fourth ventricle. A key differentiating feature is that examination of a Blake pouch cyst wall will reveal choroid plexus elements, unlike a Dandy-Walker lesion. Arachnoid cysts are thought to arise from a developmental variation of the meninx primitiva that surrounds the neural tube during the differentiation of the mesenchyme, and the cyst wall demonstrates arachnoid alone. Table 1 lists the radiological and pathological features that help distinguish these retrocerebellar cysts.

Classification of the cysts in our group of patients is difficult (Table 1). We believe that the striking uniformity of these 3 cases suggests a common etiology. In 2 of our cases, we found evidence of choroid plexus in the

Fig. 3. Case 2. Photomicrograph of a surgical specimen from an undulating fibrovascular membrane that is covered by cuboid epithelium. The epithelium exhibits the morphology and immunohistochemical profile characteristic of choroid plexus. H & E.

Fig. 4. Case 3. Preoperative sagittal (A), coronal (B), and 21-month postoperative sagittal (C) FIESTA MR images.
cyst wall (at pathological examination in Case 2 [Fig. 3] and by intraoperative visual inspection in Case 3), which strongly favors a Blake pouch cyst. In the absence of definitive histology in all cases we have referred to them collectively as retrocerebellar cysts. Regardless, we agree with Strand et al. who have stated that “…it is the identification of the retrocerebellar cystic (RCC) anomaly that is significant…It is more important to describe the morphological findings in detail…than to label the RCC with an eponym,” as for the purposes of treatment the nomenclature is less important.

Surgical Options

It has been suggested the management of the hydrocephalus associated with Blake pouch cysts is similar to that of Dandy-Walker malformation, although no large series on the management of these cysts has been published. We have described a procedure involving ventriculocystostomy in combination with an ETV or shunt placement as a successful surgical management option for hydrocephalus in infants with giant retrocerebellar cysts. Our cases highlight the need for treatment of both the cyst and the hydrocephalus, with either treatment in isolation unlikely to be successful, despite the presence of a patent aqueduct. In Case 2, VP shunt therapy with cisternal cyst fenestration was unsuccessful until the addition of ventriculocystostomy. In Case 3, the ventriculocystostomy was unsuccessful until we also performed an ETV. Only in Case 1, in which ventriculocystostomy and ETV were performed simultaneously, was definitive successful treatment achieved in one sitting.

Open cyst fenestration has been reported to be an effective treatment in posterior fossa arachnoid cysts, and it can be hypothesized that this experience could be translated to treating Blake pouch cysts. In the largest series of surgically treated posterior fossa arachnoid cysts, Marin-Sanabria et al. reported a 64% success rate with open fenestration. Endoscopic cyst fenestration of posterior fossa arachnoid cysts has been performed via a retrosigmoid bur hole but not via a precoronal bur hole, as we have used in our series. In the Marin-Sanabria series, the endoscopically treated group of 3 patients had no treatment failures. Endoscopic third ventriculostomy alone or in addition to cyst fenestration has been described in the treatment of Dandy-Walker malformation.

Cystoperitoneal shunting is a well-established treatment for symptomatic Blake pouch and arachnoid cysts and is highly effective, but it is associated with the inherent problems of shunting—namely, infection and recurrent malfunction. There are conflicting reports, however, of the outcomes of shunt therapy compared with open fenestration for arachnoid cysts, with 1 study suggesting a 20% success rate of fenestration at 8 years postsurgery and another documenting 76% success with fenestration. Our favored therapeutic approach involves endoscopic fenestration of the retrocerebellar cyst into the lateral ventricle via a precoronal bur hole, preferably with ETV performed in the same sitting. This procedure can theoretically treat both problems through one incision without the need for shunt hardware. We aim to perform a ventriculocystostomy of at least 1 cm in diameter to minimize the risk of failure, and we believe that this is feasible with modern endoscopic instruments. Given the issue of CSF leakage in infants after neuroendoscopy, we advocate pre-

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Table 1: Pathological and imaging criteria used in the differentiation of retrocerebellar cysts

<table>
<thead>
<tr>
<th>Pathological Feature</th>
<th>Dandy-Walker Malformation</th>
<th>Blake Pouch Cyst</th>
<th>Arachnoid Cyst</th>
<th>Case 1</th>
<th>Case 2</th>
<th>Case 3</th>
</tr>
</thead>
<tbody>
<tr>
<td>Arachnoid</td>
<td>Present</td>
<td>Present</td>
<td>Present</td>
<td>NA</td>
<td>Present</td>
<td>NA</td>
</tr>
<tr>
<td>Ependyma</td>
<td>Present</td>
<td>Present</td>
<td>Absent</td>
<td>NA</td>
<td>Present</td>
<td>NA</td>
</tr>
<tr>
<td>Choroid Plexus</td>
<td>Absent</td>
<td>Present</td>
<td>Absent</td>
<td>NA</td>
<td>Present</td>
<td>NA</td>
</tr>
<tr>
<td>Glia</td>
<td>Present</td>
<td>Absent</td>
<td>Absent</td>
<td>NA</td>
<td>Present</td>
<td>NA</td>
</tr>
<tr>
<td>Neurons</td>
<td>Present</td>
<td>Absent</td>
<td>Absent</td>
<td>NA</td>
<td>Absent</td>
<td>NA</td>
</tr>
<tr>
<td>Imaging Feature</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Posterior fossa size/level of torcular Herophili</td>
<td>Norm to very large</td>
<td>Usually norm to large</td>
<td>Usually norm to large</td>
<td>Very large</td>
<td>Large</td>
<td>Very large</td>
</tr>
<tr>
<td>Presence &amp; Degree of hindbrain deformity due to mass effect</td>
<td>Significant to nil</td>
<td>Nil to significant</td>
<td>Significant</td>
<td>Significant</td>
<td>Significant</td>
<td>Significant</td>
</tr>
<tr>
<td>Degree of Vermian &amp; Hemispheric Dysgenesis</td>
<td>Severe</td>
<td>Mild</td>
<td>Absent</td>
<td>Mild</td>
<td>Mild</td>
<td>Mild</td>
</tr>
<tr>
<td>4th ventricle choroid plexus</td>
<td>Absent</td>
<td>Norm to thinned</td>
<td>Norm to thinned</td>
<td>Norm to thinned</td>
<td>Unclear</td>
<td>Displaced</td>
</tr>
<tr>
<td>Occipital bone</td>
<td>Norm to thinned</td>
<td>Usually present</td>
<td>Usually present</td>
<td>Present</td>
<td>Present</td>
<td>Present</td>
</tr>
<tr>
<td>Falc cerebelli</td>
<td>Usually absent</td>
<td>Rare</td>
<td>Rare (13%)</td>
<td>None</td>
<td>None</td>
<td>None</td>
</tr>
<tr>
<td>CNS Malformations</td>
<td>14%^a</td>
<td>Absent^a</td>
<td>26%^a</td>
<td>None</td>
<td>Absent</td>
<td>Absent</td>
</tr>
<tr>
<td>Aqueductal Stenosis</td>
<td>86–94%^1^b</td>
<td>100%^1^b</td>
<td>26%^1^b</td>
<td>Present</td>
<td>Present</td>
<td>Present</td>
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<tr>
<td>4th Ventricular Dilatation</td>
<td>Absent</td>
<td>Absent</td>
<td>Absent</td>
<td>Absent</td>
<td>Absent</td>
<td>Absent</td>
</tr>
</tbody>
</table>

*Partly derived from the following studies: Nelson et al., Barkovich et al., and Strand et al. Abbreviations: NA = not assessed; norm = normal; pst = posterior.

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ventitative measures such as a semicircular skin incision that does not directly overlie the cortical opening, a linear dural opening, the placement of Duragen to plug the cortical endoscope tract with extension of the Duragen over the cortical surface to prevent migration, watertight primary dural closure, meticulous skin closure, and a skin adhesive such as Dermabond.

Suitability for ventriculocystostomy and ETV requires specific anatomical criteria, including the presence of a large retrocerebellar cyst with significant herniation through the tentorial hiatus, gross thinning of the intervening parenchyma, and a favorable configuration of the venous anatomy in this area. High-quality MR imaging, therefore, is imperative prior to contemplating such intervention. This will detail the anatomy of the cyst, the patency of the aqueduct, the position and suitability of the floor of the third ventricle for ETV, as well as the location of the important deep venous structures in relation to the cyst and ventricular walls. The axial and sagittal CISS (constructive imaging in the steady state) or FIESTA images are particularly useful in delineating the anatomy of the cyst and the presence of additional membranes that can require additional fenestration.

Additionally within these anatomical constraints we believe that this procedure can be used in selected pediatric cases involving multiculated hydrocephalus and an entrapped fourth ventricle.

Although we did not perform CT cisternography, this may assist in surgical decision making, as clear communication between the cyst and the CSF pathways may prompt the surgeon to perform a single procedure aimed at treatment of the hydrocephalus alone. The behavior of our patients following treatment, in particular the need for both cyst and hydrocephalus treatment even in the setting of a patent aqueduct, suggests that these are all non-communicating cysts.

Conclusions

This report describes a number of surgical management strategies for infants with giant retrocerebellar cysts and hydrocephalus. We have described ventriculocystostomy in conjunction with ETV through the same precoronal bur hole as our favored treatment option.

Disclosure

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

Author contributions to the study and manuscript preparation include the following. Conception and design: AV Kulkarni, JAJ King, KI Auguste. Acquisition of data: JAJ King, KI Auguste, J Drake. Analysis and interpretation of data: JAJ King, KI Auguste. Drafting the article: JAJ King. Critically revising the article: AV Kulkarni, J Drake. Final approval of the article: AV Kulkarni. Administrative/technical/material support: W Halliday. Study supervision: AV Kulkarni.

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