The natural history of MFACs in children is variable and their treatment remains controversial. Some cysts remain static incidental findings on imaging studies or even spontaneously disappear, whereas others may enlarge and exert local or global mass effects, promote hydrocephalus, rupture, or hemorrhage. The extent of communication between the cyst and the ventricles or subarachnoid space, which is similarly variable, is often difficult to assess preoperatively. This explains, in part, the poor results noted in some patients who merely undergo CSF diversion via shunting or limited fenestration of the cyst into the sylvian fissure and basal cisterns. In addition, it has been suggested that variability in outcome may be partially attributed to differences in operative technique, specifically the aggressiveness of cyst fenestration.

We retrospectively reviewed our experience with 40 patients with MFAC treated during a 16-year period and compared the results among patients presenting with hydrocephalus, nonspecific macrocephaly (craniocephaly without ventriculomegaly), and normocephaly. Hydrocephalus in this population was defined as abnormal CSF circulation resulting in an accumulation of CSF and progressive ventriculomegaly and symptomatology. Although nonspecific macrocephaly has not been previously reported as a risk factor for subsequent hydrocephalus, children with this condition may harbor a latent derangement of CSF circulation that significantly impairs their chances of a successful fenestration.

**KEY WORDS** • arachnoid cyst • fenestration • shunt • macrocephaly • middle fossa

**Object.** The optimal management of children with middle fossa arachnoid cysts (MFACs) remains controversial. In this study the authors evaluated the relationship between two preoperative variables, hydrocephalus and nonspecific macrocephaly, in children undergoing fenestration of temporal arachnoid cysts and hydrocephalus-related shunt placement.

**Methods.** During a 16-year period, 40 children (30 boys and 10 girls) underwent treatment of MFACs. All but one patient experienced either worsening symptoms or progressive serial imaging–documented cyst enlargement. Hydrocephalus was present in six patients and nonspecific macrocephaly in another nine. The mean age at surgery was 66 months (range 1–201 months, median 36 months), and the mean follow-up duration was 54 months (range 6–83 months, median 39 months).

All patients presenting with hydrocephalus required placement of a ventriculoperitoneal (VP) shunt as well as cyst fenestration, regardless of which procedure was performed first. Five patients with macroencephaly undergoing initial fenestration required subsequent VP shunt insertion. Complications of cerebrospinal fluid (CSF) diversion were typical.

**Conclusions.** Patients with hydrocephalus or macrocephaly are likely to require VP shunt placement in addition to cyst fenestration. Children with nonspecific macrocephaly may harbor a latent derangement of CSF circulation.

**Abbreviations used in this paper:** CSF = cerebrospinal fluid; CT = computerized tomography; CVP = cystoventriculoperitoneal; MFAC = middle fossa arachnoid cyst; VP = ventriculoperitoneal.

**Clinical Material and Methods**

The 40 patients comprising this report were identified retrospectively from records of the Division of Neurosurgery at the Childrens Hospital of Los Angeles during a 16-year period. All patients undergoing surgery for an MFAC were included.

The study was composed of 30 boys and 10 girls, a ratio consistent with previous reports documenting a higher prevalence of MFAC in males. The mean age at surgery was 66 months (range 1–201 months, median 36 months). The mean follow-up duration was 54 months (range 6–183 months, median 39 months).
Cysts were graded using the scale established by Galassi, et al. All patients but one experienced either worsening symptoms or serial imaging–documented progressive enlargement of the cyst from Grade I to Grade II or III; in the remaining patient a large Grade III cyst was incidentally found in utero and, because of the lesion’s size, was treated shortly after the patient was born. Furthermore, all cysts were of considerable dimension and demonstrated significant displacement of normal brain structures, midline shift, or other radiological evidence of mass effect. The most common presentation in infants was irritability or lethargy accompanied by a tense fontanel and bulging sutures. In older children, headaches, vomiting, and lethargy predominated. Seizures were the second most common symptom. In three patients who presented with hemiparesis, two exhibited weakness on the side contralateral to the cyst; in the third case, the ipsilateral weakness presumably resulted from compression of the contralateral cerebral peduncle. The remainder of the presenting signs and symptoms are summarized in Table 1.

Cysts were located on the right side in 17 patients, on the left in 21, and bilaterally in two. This distribution is similar to that reported in other studies involving MFACS. The cyst extended across the incisura to the suprasellar region in three patients who presented with endocrine symptoms due to disruption of the hypophyseal–pituitary axis or with visual compromise due to interruption of the optic pathways.

Six patients presented with hydrocephalus (Table 2). Nine other patients exhibited nonspecific macrocephaly, with a head circumference greater than the 98th percentile but no radiographic evidence of ventriculomegaly (Table 3). Compared with normocephalic patients, those with macrocephaly tended to harbor larger cysts or were more likely to have complicating lesions such as subdural effusions. Their age and sex distributions, however, were similar to those of the group as a whole.

Four patients presented with an associated subdural

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**TABLE 1**

Summary of clinical presentations in 40 pediatric patients with hydrocephalus and MFACS

<table>
<thead>
<tr>
<th>Symptoms &amp; Signs</th>
<th>No. of Cases</th>
<th>Presenting</th>
<th>Resolved</th>
</tr>
</thead>
<tbody>
<tr>
<td>headache</td>
<td></td>
<td>12</td>
<td>10*</td>
</tr>
<tr>
<td>progressive cyst growth</td>
<td></td>
<td>10</td>
<td>10</td>
</tr>
<tr>
<td>seizures</td>
<td></td>
<td>7</td>
<td>5†</td>
</tr>
<tr>
<td>developmental delay</td>
<td></td>
<td>5</td>
<td>1</td>
</tr>
<tr>
<td>visual disturbance</td>
<td></td>
<td>3</td>
<td>1</td>
</tr>
<tr>
<td>hemiparesis</td>
<td></td>
<td>3</td>
<td>2</td>
</tr>
<tr>
<td>papilledema</td>
<td></td>
<td>3</td>
<td>3</td>
</tr>
<tr>
<td>6th cranial nerve palsy</td>
<td></td>
<td>2</td>
<td>2</td>
</tr>
<tr>
<td>endocrinopathy</td>
<td></td>
<td>2</td>
<td>0</td>
</tr>
<tr>
<td>ataxia</td>
<td></td>
<td>2</td>
<td>1</td>
</tr>
<tr>
<td>behavioral change</td>
<td></td>
<td>2</td>
<td>1‡</td>
</tr>
<tr>
<td>incidental</td>
<td></td>
<td>1</td>
<td>1</td>
</tr>
</tbody>
</table>

* In 10 of 12 patients presenting with headache resolution occurred after surgery.
† Resolution of seizures was defined as independence from related medication.
‡ Of five patients with developmental delay, evidence of improvement was observed in one following fenestration.

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**TABLE 2**

Summary of data in cases involving hydrocephalus requiring CSF diversion

<table>
<thead>
<tr>
<th>Age (mos.), Sex</th>
<th>Treatment Indications</th>
<th>Initial Treatment</th>
<th>ShuntFU Treatment</th>
<th>Complication</th>
<th>Shunt FU</th>
<th>No. of Revisions</th>
<th>FU Result (no. of mos)</th>
</tr>
</thead>
<tbody>
<tr>
<td>1, M</td>
<td>progressive cyst enlargement</td>
<td>VP shunt</td>
<td>craniotomy</td>
<td>none</td>
<td>VP shunt</td>
<td>0</td>
<td>no hydrocephalus or cyst; intact (155)</td>
</tr>
<tr>
<td>2, M</td>
<td>progressive cyst enlargement</td>
<td>VP shunt</td>
<td>craniotomy</td>
<td>none</td>
<td>VP shunt</td>
<td>0</td>
<td>no hydrocephalus or cyst; intact (109)</td>
</tr>
<tr>
<td>22, F</td>
<td>progressive cyst enlargement</td>
<td>VP shunt</td>
<td>craniotomy</td>
<td>none</td>
<td>VP shunt</td>
<td>0</td>
<td>no hydrocephalus or cyst; intact (65)</td>
</tr>
<tr>
<td>26, M</td>
<td>progressive cyst enlargement</td>
<td>VP shunt</td>
<td>craniotomy</td>
<td>none</td>
<td>VP shunt</td>
<td>0</td>
<td>no hydrocephalus or cyst; intact (155)</td>
</tr>
</tbody>
</table>

AF = anterior fontanel; delay = developmental delay; FU = follow-up; IVH = intraventricular hemorrhage; PO = pars oralis; POD = postoperative Day; SDH = subdural hematoma; SPS = shunt for subdural fluid collection.
## TABLE 3

Summary of data in cases involving macrocephaly requiring CSF diversion*

<table>
<thead>
<tr>
<th>1st Op</th>
<th>2nd Op</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Age (mos)</td>
</tr>
<tr>
<td>Sex</td>
<td>At Imaging</td>
</tr>
<tr>
<td></td>
<td></td>
</tr>
<tr>
<td>F yes</td>
<td>18</td>
</tr>
<tr>
<td>F yes</td>
<td>5</td>
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<tr>
<td>M yes</td>
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<tr>
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<td>10</td>
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<tr>
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<td>M no</td>
<td>11</td>
</tr>
<tr>
<td>M no</td>
<td>14</td>
</tr>
<tr>
<td>M no</td>
<td>5</td>
</tr>
</tbody>
</table>

*CN = cranial nerve; HC = head circumference; HCP = hydrocephalus; NA = not applicable; PM = pseudomeningocele.
Results

Three of six patients presenting with ventriculomegaly as well as temporal arachnoid cysts underwent initial attempts at CSF diversion. In two cases a VP shunt was placed and in one a CVP shunt was placed. All three required subsequent craniotomy and cyst fenestration because of progressive symptoms associated with continued growth of their cysts. The remaining three patients initially underwent combinations of cyst fenestration and/or shunt therapy. In all three cases a VP shunt was subsequently required because of continued symptomatic progression.

Nine patients presented with nonspecific macrocephaly in the absence of ventriculomegaly, and all underwent initial cyst fenestration. Five patients subsequently required CSF diversion for continued symptom progression and progressive ventriculomegaly (Case 1, Figs. 1–3) and in the remaining four patients imaging studies demonstrated subdural fluid collections related to craniocerebral disproportion. With regard to those patients requiring CSF diversion, all underwent pre- and postoperative imaging studies. Initial diagnostic images were obtained at a mean of 9 ± 5 months (range 1–18 months). In patients with macrocephaly who required subsequent shunt placement, shunt insertion was performed 7 ± 6 months after the initial surgery (range 2–15 months). With regard to the patients presenting with macrocephaly, all underwent postoperative imaging studies at a mean of 4 ± 1.5 months (range 2–6 months). Postoperative radiological studies were obtained at 3 months (all cases), 10 months (all cases), and 37 months (90% of cases). In those five patients with macrocephaly who subsequently required CSF diversion, postoperative imaging studies were obtained at 3, 10, and 21 months in all cases and at 37 months in 80% of cases.

In the 25 patients with normocephaly undergoing cyst fenestration, one required VP shunt therapy postoperatively. Four patients presented with an associated subdural hygroma: two with subdural hematoma and two with hemorrhage within the cyst.

The operative data may be summarized as follows. All patients presenting with hydrocephalus required shunt placement as well as craniotomy and fenestration, regardless of which procedure was performed first. Five patients with macrocephaly undergoing initial fenestration required insertion of a shunt.

The surgery-related outcome, in terms of symptom resolution, is summarized in Table 1. In general, seizures, headaches, and cranial nerve palsies responded favorably to surgery, whereas visual disturbances, developmental delay, and endocrinopathies were likely to persist despite adequate treatment of the cyst, as has been noted in previous studies.5,8,17,33 The resolution of seizures was defined as independence of the child from medication. All children presenting with seizures were weaned from anticonvulsant agents during a 6-month period while being monitored with serial electroencephalography. Recurrence of seizure activity or electrical evidence of foci resulted in the continuation of anticonvulsant agents.

We avoided shunt placement when possible to limit the complications potentially due to shunts. In our series, shunt-related complications included a high incidence of malfunction requiring at least one revision. This was true not only in cases involving a VP shunt (67%), but also a CVP shunt (72%), and subdural fluid collections due to craniocerebral disproportion (28%). The maximal number of shunt revisions in one patient is 12 to date. Two patients suffered complications secondary to overshunting, with the development of contralateral subdural hematomas requiring evacuation and a separate shunt to treat subdural fluid collections. Two patients experienced shunt-related infections, an incidence consistent with our overall shunt infection rate of approximately 3 to 4%, and one patient with hemophilia developed an abdominal hemorrhage requiring a laparotomy after insertion of a CVP shunt.

Other complications included one case of transient hemiparesis, two transient seizures within 6 hours following surgery, two transient third cranial nerve palsies, one case of intracerebral hemorrhage requiring evacuation in a patient with coagulopathy and subsequent brain abscess formation, and one case of a subdural hematoma requiring removal 6 weeks after fenestration in a patient with macrocephaly due to craniocerebral disproportion. No permanent neurological defects were sustained in any patient.

Illustrative Case

Case 1

This 3-month-old patient with a history of progressive macrocephalus and a Grade III cyst developed hydrocephalus after cyst fenestration. Six serial axial CT scans of the brain demonstrated the initial presence of a large Grade III left-sided arachnoid cyst. After cyst fenestration, a pseudomeningocele developed at the site, and imaging and clinical characteristics were consistent with hydrocephalus. At 9 months after placement of a right-sided VP...
Hydrocephalus and middle fossa arachnoid cysts

shunt, ventricle size was normal and the preexisting arachnoid cyst had resolved.

Discussion

Pathogenesis of Arachnoid Cysts

Arachnoid cysts consist of congenital CSF collections that occur between layers of the arachnoid. They likely arise from anomalous splitting and duplication of the endomine during development.32 Although they can arise throughout the central nervous system, they are most commonly localized in the middle cranial fossa.9

The natural history of MFAC in children is variable. Some lesions remain static incidental findings on imaging studies or may even spontaneously disappear, whereas others may expand and exert local or global mass effects, promote hydrocephalus, rupture, or hemorrhage.2,9,29,30,35,39 Controversy exists regarding the pathophysiology of arachnoid cyst enlargement. Some investigators have proposed a unidirectional ball-valve mechanism that progressively traps increasing amounts of CSF.9,16 Alternatively, analysis of morphological and enzyme cytochemical evidence had indicated that cyst wall cells actively secrete fluid.15 Both theories account for the tendency of some cysts to expand with time. Enlarging cysts produce symptoms by compressing nearby neural structures or through their own mass effect. In other cases, MFACs may obstruct CSF flow.9,31,37,39

Indications for Treatment

In this series, all patients but one experienced either worsening symptoms or progressive cyst enlargement documented on serial imaging. Because of the age of the children in this study, long-term evidence of progressive symptoms is difficult to assess. Children with symptoms potentially related to the cysts and with an obvious increase in cyst size on serial imaging studies, or those harboring an extremely large cyst (Galassi Grade III), underwent fenestration. After treatment, most seizures or focal neurological deficits resolved (Table 1), although visual disturbances, developmental delay, and endocrinopathies tended to persist despite cyst fenestration. The persistence of various symptoms has also been noted in previous reports.2,9,17,33 Other investigators have confirmed seizure reduction7,16,23,37 and improved neurological status10,13,15,18,20,31,37,38 following fenestration.

The benefit of cystoperitoneal shunt therapy on seizures is less clear, given evidence that seizures may persist after treatment, indicating that the arachnoid cyst was an incidental finding in patients with epilepsy.6,23,37 Seven patients with previously documented cysts underwent clinical and radiographical observation prior to becoming symptomatic. Status in one of these patients, who presented with optic atrophy, was stabilized after surgery but never improved. In another case a sixth cranial nerve palsy developed.

Large cysts (Grades II and III) that do not produce focal neurological deficits may still exert pressure on the brain and promote developmental delay.2,9,15,31,40 and evidence of improved cognitive function postoperatively has been documented.16,34,39 Positron emission tomography and single-photon emission computerized tomography studies have demonstrated hypometabolism in speech areas and global reductions in cerebral blood flow34,40 in patients with large cysts, with postoperative improvement noted in perfusion parameters and vocabulary.16,34

Children with untreated cysts have been reported to develop intracystic or subdural hemorrhage spontaneously or after minor head injury.2,9,11,28,30,33,35 Patients with an MFAC have also been reported to be at risk of developing a seizure disorder that may not respond to subsequent treatment.27 Evidence of clinically silent prior hemorrhage detected on magnetic resonance imaging may be another indication for surgery in asymptomatic patients.9 Finally, findings in recent reports of attention deficit–hyperactive disorder associated with MFACs have indicated that we do not fully understand this entity, which may not be as innocuous as once thought.16,34

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**Treatment Options**

The treatment options for intracranial arachnoid cysts include either craniotomy with fenestration or shunting of the cyst contents. Endoscopy has been used as an adjunct for both procedures. We consider avoidance of shunt therapy a major objective of treatment. Shunts are associated with a high incidence of malfunction, possibly due to reexpansion of the brain, which forces the tip of the catheter into the cerebral parenchyma potentially causing its obstruction. Cystoperitoneal shunts are also associated with potential infection, slit-ventricle syndrome, shunt dependency, and other complications to which patients are susceptible for a lifetime. In contrast, treatment failures or successes after fenestration tend to manifest soon after surgery. Thus, only the latter procedure can be truly considered a cure.

Some surgeons have advocated CSF diversion on the basis of reported recurrence after only partial resection of the cyst wall or limited fenestration, the latter confirmed by postoperative metrizamide cisternography in some cases. Proponents of CSF diversion have also contended that it is a smaller less risky operation with fewer complications than fenestration; however, we found in this study that fenestration of MFACs can be performed with minimal morbidity. In addition, only craniotomy provides the appropriate management of cysts in patients who present after an episode of associated hemorrhage. We suggest that simple fenestration of the cyst contents into the basilar cisterns greatly enhances the likelihood of avoiding a need for shunt placement, despite isolated anecdotal evidence from previous series in which partial excision of the cyst wall alone has been claimed to be an insufficient treatment of MFACs. Fenestration of the cyst membrane also appears to be the most rational treatment because of the potential secretory properties of the cyst wall.

**Subsequent Treatment Strategies**

In the present series, the requirement for subsequent procedures was determined by a patient’s clinical status and the radiographic appearance of the cyst after the initial operation. In patients who had previously undergone attempted CSF diversion, subsequent craniotomy and cyst fenestration were mandated either by progressive symptoms or by the imaging-documented progression of the cyst despite adequate deflation of the ventricles. Conversely, shunt therapy following previous attempts at cyst fenestration was inevitably undertaken when bulging and/or leakage at the surgical site was observed within a few weeks, indicating a derangement in the CSF circulation, and frequently accompanied by recrudescence of presenting symptoms.

**Significance of Hydrocephalus and Nonspecific Macrocephaly**

A successful outcome is unlikely in patients with hydrocephalus in whom a single shunt has been placed; separate treatment of the ventricles and the cyst will be required. Patients with nonspecific macrocephaly (cranio-megaly in the absence of hydrocephalus) will likely require placement of a shunt, regardless of fenestration. Symmetrical or asymmetrical macrocephaly without ventriculomegaly is a presenting sign in numerous patients with an MFAC, however, it has not been previously reported as a risk factor for treatment failure in other studies.

Children with nonspecific macrocephaly may experience a latent derangement of CSF circulation that significantly impairs the prospect for a successful fenestration. Alternatively, patients may suffer from craniofacial disproportion due to brain atrophy or the presence of complicating space-occupying lesions (for example, hygromas or subdural hematomas) that are removed during craniotomy. For these reasons, the cyst may persist despite attempted fenestration. Nonetheless, a cystoperitoneal shunt alone may be insufficient in macrocephalic patients. Some surgeons have advocated combining the shunt procedure with excision of the outer cyst wall to prevent this membrane, which easily detaches from the dura mater following shunt insertion, from occluding the shunt catheter. This adjunctive measure might also minimize the hazard of blindly passing a catheter across the outer cyst wall, which may have fragile arteries and veins coursing over it.

**Conclusions**

Based on our experience, we suggest that patients with macrocephaly or hydrocephalus associated with temporal arachnoid cysts are likely to require eventual CSF diversion following initial cyst fenestration. All patients in our series with progressive ventriculomegaly required subsequent CSF diversion. It has been our practice to follow these patients radiologically after cyst fenestration until such time that progressive ventriculomegaly defines the need for shunting. All patients presenting with hydrocephalus required shunt therapy in addition to fenestration, regardless of which procedure was performed first. Five patients with macrocephaly who underwent initial fenestration required insertion of a subsequent shunt. Given the potential complications associated with long-term shunt therapy, it was our practice to fenestrate the cyst and follow the patient with serial imaging for evidence of progressive ventriculomegaly. Cyst fenestration remains an important component of treatment to avoid the need for two shunt systems. In normocephalic patients, treatment consists of craniotomy and fenestration alone. The timing of shunt placement in these patients is more difficult to define: should the device be placed at the time of fenestration or should patients be followed for several weeks to identify the small subset of patients who will not require CSF diversion of any type? In any scenario, symptomatic temporal arachnoid cysts in the absence of ventriculomegaly or macrocephalus have all been treated successfully following fenestration alone.

**Disclaimer**

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