Intracranial arachnoid cysts in children

A comparison of the effects of fenestration and shunting

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The best operative intervention for children with arachnoid cysts remains the subject of controversy. Recent reports stress that craniotomy for cyst fenestration is associated with a low incidence of morbidity and mortality and may leave the child shunt-independent. The cases of 40 pediatric patients with arachnoid cysts treated between 1978 and 1989 are reported. Five children with mild symptoms and small cysts that remained stable on follow-up studies have not required surgical intervention. Of 15 patients with cysts initially treated by fenestration, 10 (67%) showed no clinical or radiographic improvement postoperatively and have undergone cyst-peritoneal (eight patients) or ventriculoperitoneal (VP) shunting (one patient), or revision of a VP shunt placed for hydrocephalus before cyst fenestration (one patient). Two other patients with existing VP shunts required no further procedures. Thus, only three (20%) of 15 patients initially treated by fenestration remain shunt-independent after a median follow-up period of 8 years. The 20 other patients were initially treated by cyst shunting and all improved postoperatively; shunt revision has been necessary in six (30%) of these 20 patients because of cyst recurrence. Cyst location influenced the success of shunt treatment; none of the seven middle cranial fossa cysts treated by shunting have required revision, but results with cysts in other locations were less favorable. In all locations, though, shunting was more successful than fenestration. It is concluded that cyst-peritoneal or cyst-VP shunting is the procedure of choice for arachnoid cysts in most locations, including those in the middle cranial fossa.

KEY WORDS • cyst, arachnoid • fenestration • shunt • craniotomy • children

ARACHNOID cysts are intra-arachnoid collections of cerebrospinal fluid (CSF). By compressing adjacent neural tissue or by obstructing CSF flow, they produce symptoms such as craniomegaly, developmental delay, seizures, and headaches. In 75% of patients, the congenital origin of these cysts is manifest in the early appearance of signs and symptoms, usually before 6 months of age. Cranial ultrasonography, computerized tomography (CT), and magnetic resonance (MR) imaging have facilitated the diagnostic workup of patients with these cysts. Indeed, cysts are frequently discovered incidentally during radiographic evaluation for an unrelated complaint. Many of these cysts are asymptomatic, requiring no immediate treatment. They should, however, be periodically assessed with CT or MR imaging.

In patients whose cysts are symptomatic, surgical intervention is called for, but which is the best method of treatment remains controversial. Options include needle aspiration, cyst-peritoneal shunting, ventriculocystostomy, and craniotomy for partial or complete cystectomy or for marsupialization into the subarachnoid space, basilar cisterns, or ventricle. Of these, cyst-peritoneal shunting and craniotomy for fenestration are the most common. Many authors advocate fenestration of all middle cranial fossa cysts, which averts the risks involved in placing a permanent shunt. Fenestration, however, often fails to obliterate the cyst and therefore does not eliminate the need for shunting. If complications from shunting can be minimized, then shunting as an initial procedure may be preferable to fenestration.

In this paper we update our original report published in 1986. Forty pediatric patients with intracranial arachnoid cysts were treated between 1978 and 1989; their clinical and radiographic presentation and the
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results of their surgical treatment are presented. In particular, we compare the effectiveness of fenestration and shunting.

Summary of Cases

Since 1978, we have treated 40 patients for intracranial arachnoid cysts; 16 of these patients have been reported previously. There were 26 boys and 14 girls, whose ages at the time of clinical presentation ranged from birth to 15 years (median 2.2 years, mean 4.3 years). All 40 were referred to the neurosurgical service because of refractory headaches, neurological dysfunction, or rapidly increasing head size. Seventeen patients had symptoms and signs of elevated intracranial pressure (vomiting, lethargy, bulging fontanelles, or split sutures), 15 had craniomegaly, 12 had developmental delay, 11 had seizure disorders, and 10 patients had headaches.

Diagnostic Workup

Computerized tomography or MR imaging showed intracranial cystic masses that contained fluid of CSF density and, in 35 of the 40 patients, compressed surrounding neural or ventricular structures. In the other five patients, the cysts were smaller than 5 cm and caused no compression of surrounding structures (Fig. 1). Most of the cysts were supratentorial: 16 cysts were in the middle cranial fossa, six were interhemispheric or diffuse, five were suprasellar, and one cyst was in the quadrigeminal cistern. The other 12 cysts were in the posterior fossa. Ventriculomegaly was present in 12 of 28 patients with supratentorial cysts and in seven of 12 patients with posterior fossa cysts but in only three of 16 patients with middle cranial fossa cysts. Patients with symptoms referable to elevated intracranial pressure were usually found to have cysts in the posterior fossa, whereas seizures and headaches more often indicated middle cranial fossa cysts (Table 1).

Initial Treatment

Because the five small cysts (including one posterior fossa cyst) caused no symptoms and no compression, they have not required surgical intervention. The patients remain clinically stable, and their MR images have not changed during follow-up periods ranging from 1½ to 3½ years.

For 15 patients, cysts were first treated by craniotomy in which the cyst cavity was widely fenestrated into the subarachnoid space. Three of these 15 patients had previously undergone ventriculoperitoneal (VP) shunting for hydrocephalus. The cysts of the other 20 patients were initially treated by shunting; 14 underwent cyst-peritoneal shunting, and six underwent cyst-VP shunting in which separate cyst and ventricular catheters were joined to the peritoneal tubing with a Y-shaped connector.

Operative Results and Follow-Up Treatment

The follow-up period in these 40 patients ranged from 3 months to 12 years (mean 5.0 years, median 8.0 years).

Cysts Initially Treated by Fenestration. In five of the 15 patients whose cysts were treated initially by fenestration, the cyst decreased in size postoperatively and there was at least some clinical improvement (Table 2); in one of these patients, symptoms not referable to the cyst were unchanged. Nine cysts were unchanged by primary fenestration and one continued to enlarge; in these 10 patients, symptoms either remained the same or grew more severe, and in one case diminished but returned 9 years after fenestration.

After fenestration (in one case after a second fenestration) eight patients had persistent symptoms and underwent cyst-peritoneal or cyst-VP shunting (Fig. 2). Of these, three have subsequently received cyst-peritoneal shunt revisions (three revisions each in two pa-
patients and one revision in the other patient). In a ninth patient, a suprasellar cyst recurrent 6 years postoperatively; fenestration was repeated and a VP shunt was placed. A 10th patient, who had undergone VP shunting for hydrocephalus before fenestration of a quadrigeminal cistern cyst, required a VP shunt revision after fenestration. Thus, of the 15 patients whose cysts were widely fenestrated, nine required new shunts and one required revision of an existing shunt after fenestration. The other two patients with existing VP shunts required no further procedures after fenestration. Only three patients had no other operative procedures before or after fenestration, and remain shunt-independent.

In one patient, fenestration failed, but a cyst-peritoneal shunt obliterated the cyst. The shunt was removed 5 years later during a temporal lobectomy for refractory seizures. In the absence of a shunt, a subgaleal fluid collection developed, and the cyst recurred, but a new cyst-peritoneal shunt alleviated both problems.

**Cysts Initially Treated by Shunting.** All 20 patients initially treated by cyst shunting showed some clinical and radiographic improvement postoperatively (Fig. 3). Fourteen of the 20 patients have not required any further procedures. Six patients, however, have undergone shunt revisions (two cyst-peritoneal shunt revisions in each of two patients, a cyst-VP and a VP shunt revision in another patient, and another cyst-peritoneal shunt revisions in the other three patients). Despite the necessity for these revisions, shunting had a higher success rate than fenestration.

**Summary of Results.** Overall, 34 of the 35 treated cysts are reduced in size on follow-up studies; the quadrigeminal cistern cyst, after unsuccessful fenestration and VP shunting for hydrocephalus, has remained stable. Only 10 cysts, however, have been completely obliterated (seven of these were shunted, and three were fenestrated). Signs and symptoms of intracranial hypertension were most readily improved, whereas longstanding developmental delay responded poorly to surgical intervention of both types.

**Table 2**

<table>
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<tr>
<th>Procedure</th>
<th>With Hydrocephalus</th>
<th>Without Hydrocephalus*</th>
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<tr>
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</table>

*ND = procedure not done.
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Complications

Cysts Treated by Fenestration. Six of the 15 patients with fenestrated cysts suffered complications after the procedure. Two patients treated by craniotomy developed aseptic meningitis after fenestration. Another patient developed new grand mal seizures, and her neurological deficit increased. One patient, who underwent fenestration of a small arachnoid cyst in the choroidal fissure, developed a third cranial nerve palsy. Another patient developed a third nerve palsy and hemiparesis after repeat fenestration. The sixth patient developed a subdural hemorrhage and required reoperations for drainage and, subsequently, repeat cyst fenestration.

Cysts Treated by Shunting. Twenty-eight patients underwent shunting procedures, 20 as the initial treatment and eight after failed fenestration. Although shunt revisions have been required in nine of these patients, there have been no shunt infections and no permanent operative complications. Symptoms, such as nausea, vomiting, and postural headache, prolonged hospitalization for six patients. One patient developed small bilateral subdural fluid collections that resolved spontaneously, and another patient with shunt placement developed aseptic meningitis. One patient developed a left sixth cranial nerve palsy after shunting of a right middle cranial fossa cyst. The palsy resolved over the course of 3 months. These last three were the only patients for whom shunting led to complications.

One patient died during the follow-up period. This 3-year-old patient with a large posterior fossa cyst was successfully treated with a cyst-peritoneal shunt. He recovered, and follow-up CT scans showed that the cyst had decreased in size. Four months after the shunt was placed, the patient fell while climbing in a tree and became apneic. He failed to respond to cardiopulmonary resuscitation and never regained signs of brainstem function. Although a postmortem examination failed to reveal the cause of death, it showed that the posterior fossa cyst had been obliterated.

Discussion

Patient Selection

Not all arachnoid cysts require surgical intervention. Asymptomatic cysts and those producing minor skull defects should be followed clinically, with annual CT or MR imaging, and with neuropsychiatric testing. When patients develop symptoms or show deteriorating school performance or other evidence of cognitive impairment, surgical intervention should be considered. Such treatment is undoubtedly required for children with symptoms of intracranial hypertension, intractable seizures, and focal neurological deficits. We believe, furthermore, that arachnoid cysts that produce direct or indirect brain compression warrant treatment as well. In children, the potential for impaired development and function of brain tissue or for rapid neurological deterioration caused by intracystic or subdural hemorrhage outweighs the risks of operative intervention.

Cyst Fenestration

Controversy continues regarding which surgical treatment is best. One of the greatest advantages of fenestration is that it can leave the patient shunt-independent. In addition, it allows direct inspection and excision of a portion of the cyst wall by biopsy to confirm the diagnosis, and coagulation of the fragile arachnoidal blood vessels under direct inspection. For quadrigeminal cistern cysts and suprasellar cysts, craniotomy remains the preferred surgical treatment. Because of their proximity to the brain stem, small cysts in these locations are more safely approached under direct visualization, and fenestration into adjacent cisterns is usually curative. Large suprasellar cysts may be approached transcallosally, allowing fenestration into the lateral ventricle and subsequent VP shunting.

For patients without hydrocephalus, several authors advocate cyst fenestration as the initial treatment. In their series of 77 adult and pediatric patients with middle cranial fossa cysts initially treated by fenestration, Galassi, et al. reported only one death, caused by bacterial meningitis, and two major postoperative complications (one extradural and one intracystic hematoma). The rest (95%) of their patients improved clinically or radiographically after fenestration. These good results may reflect the fact that adults generally suffer fewer complications and that middle cranial fossa cysts are among the least difficult to treat. Poorer results are seen in studies that involve only children or include patients with cysts outside the middle cranial fossa. Raffel and McComb reported that 11 (73%) of 15 patients without hydrocephalus had cysts successfully fenestrated, and remain shunt-independent. Lange and Oeckler reported a series of 35 patients in which 13 (68%) of 19 patients treated by craniotomy showed significant clinical improvement. Four of the six patients who did not improve after craniotomy required subsequent shunting procedures. Menezes, et al., reported a series of 10 children with arachnoid cysts in whom eight (89%) of nine fenestrations were free of complications. None of these studies, however, used CT or MR imaging in all patients to document cyst resolution after craniotomy. Despite the clinical success indicated in these studies, fenestration for us has not been as effective. In our series, only two (33%) of six patients without hydrocephalus remain shunt-independent after fenestration.

Shunting Procedures

Patients with hydrocephalus rarely exhibit ventricular decompression after fenestration alone. In our series, only one of nine patients with hydrocephalus was successfully treated with fenestration. Patients often require cyst-peritoneal shunting for cyst recurrence or VP...
shunting for persistent ventriculomegaly.\textsuperscript{15,16,21} Because VP shunting is usually necessary to treat hydrocephalus, a cyst-VP shunt appears to be a more prudent surgical option than fenestration when hydrocephalus is present. The patient will need the VP shunt regardless, and an additional shunt from the cyst to the peritoneal tubing does not substantially increase the risk. In our study, of the nine shunts that required revision, six were cyst-peritoneal whereas only three were the more complex cyst-VP shunts. Thus, two joined shunts do not appear to malfunction at a greater rate than a single shunt.

Arachnoid cysts frequently recur after fenestration,\textsuperscript{1,9,13} either because of reclosure of the cyst wall after incomplete resection or because of inadequate CSF flow through the subarachnoid space into which the cyst is opened.\textsuperscript{7} In these cases, a shunt must be placed to eliminate the cyst permanently. Shunting is also necessary\textsuperscript{16,23} when a cyst is successfully treated by fenestration, but an obstructive or communicating hydrocephalus accompanying the cyst does not improve.\textsuperscript{7} Raffel and McComb\textsuperscript{26} treated 29 cysts by craniotomy for fenestration. Thirteen of the 29 patients remain shunt-independent, but 16 have required cyst-peritoneal shunting for persistent cysts or VP shunting for hydrocephalus. In our series, only three of the 15 patients initially treated by fenestration remain without a shunt.

Considering the complications of craniotomy, which include aseptic meningitis, increased neurological deficits, and delayed intracranial hemorrhage,\textsuperscript{1,3,5,12,16} and the large number of patients who need shunts even after fenestration, shunting should be considered as the initial surgical procedure for most arachnoid cysts. When initial fenestration or excision has failed, shunting is effective as a secondary treatment to bring about decompression of arachnoid cysts.\textsuperscript{1,6,15,16,31} Because it deals directly with the pathophysiology of cystic CSF loculation and with the associated abnormalities of CSF flow and absorption, shunting is a simpler and more effective procedure than fenestration.\textsuperscript{1,5,17,20,21,25,26}

A drawback of shunting is that a shunt must usually remain in place throughout the patient's life. Despite its permanence, however, shunting seems to cause few complications and to impose few limitations on the patient. Complications such as proximal or distal shunt malfunctions and infection rarely occur,\textsuperscript{15,17,20} and if the arachnoidal leaves are reapproximated after shunting, fluid may not reaccumulate even if the shunt is later occluded. Although symptoms such as nausea and headache sometimes occur after shunting, they are not dangerous and invariably diminish with time.

All patients should undergo postoperative CT or MR imaging to assess cyst size and to confirm catheter placement. Patients who show no radiographic improvement on follow-up scans require close clinical and radiographic observation and may warrant radionuclide evaluation to rule out shunt malfunction. Periodic radiographic evaluation is also prudent after successful shunting and is imperative if clinical symptoms recur or if symptoms of shunt malfunction or infection develop.

Cyst location appears to influence the success of shunt therapy (Table 3). In our series, seven children with middle cranial fossa cysts were treated initially with cyst-peritoneal shunting, and four received a cyst-peritoneal shunt after failed fenestration; all improved after shunting. In contrast, cyst shunting was successful in only about half of the children with posterior fossa cysts, whether shunting was used as the first mode of treatment (five of nine cases) or after failed fenestration or shunting (three of five cases). Cysts in both locations, however, were more successfully treated by shunting than by fenestration.

**Conclusions**

Arachnoid cysts create a serious potential for permanent severe neurological deficit as a result of progressive expansion or hemorrhage. When mass effect is demonstrable on radiographic studies, the availability of a simple, safe, and effective method of therapy argues for surgical intervention, even in asymptomatic cases. Cyst location appears to play an important role in the success of shunt treatment; all of our middle cranial fossa cysts were successfully shunted, whereas results with cysts in other locations were less favorable. Regardless of cyst location, however, shunting was more successful than fenestration in our hands. Only in our few patients with suprasellar cysts were both forms of intervention equally efficacious. Cyst-peritoneal shunting, or cyst-VP shunting if hydrocephalus is present, is the procedure of choice for the treatment of intracranial arachnoid cysts in children.

**References**


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**TABLE 3**

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