A rachnoid cysts are frequently encountered pathological conditions. They account for 1% of all intracranial lesions, with an estimated incidence of 0.5%–2.6%. The optimal treatment for symptomatic ACs remains controversial. Cystoperitoneal shunt placement is believed to be a safe and effective surgical therapy for symptomatic ACs in children, allowing rapid symptom relief and cyst reduction, especially in younger children. But it also presents complications, such as early shunt failure, overdrainage, and lifelong shunt dependence. In this paper, we review long-term follow-up data on the treatment of ACs in children via CP shunt placement performed at our institution.

Methods
To collect enough long-term follow-up data, we retrospectively studied 62 children with ACs who had undergone CP shunt placement at our institute between January 2003 and January 2005. From each patient’s file we recorded age, sex, presenting symptom, modality of treatment, complications, outcome, and follow-up. All patients were preoperatively evaluated via CT and/or MRI studies. Postoperatively, all patients underwent regular clinical and radiological follow-up evaluations.

Results

Patient Characteristics
The age of the patients ranged from 1 to 13 years, and the mean age at the time of diagnosis was 6.5 years. Clinical manifestations on presentation were characterized by signs of increased ICP (headache, vomiting, drowsiness, bulging fontanelle, and an enlarged head) in 26 patients, seizures in 15, hemiparesis in 5, and huge asymptomatic ACs in 15. As regards the children with seizures, the surgical candidates in our group were those who had not responded to antiepilepsy drugs for at least 1 year. One child presented with spontaneous subdural hematoma and hygroma as a complication of the AC. Forty-six cysts (74%) were within the sylvian fissure, 8 (13%) were infratentorial. A CP shunt was placed in all patients. Follow-up imaging studies showed that 59 (95%) of 62 ACs reduced in size during a mean postoperative follow-up period of 6.5 years (range 6–8 years). Although a CP shunt was effective in achieving early obliteration, shunt dependency occurred within the patient group (13%). Shunt revision for various reasons was performed in 16 patients (26%).

Conclusions
Shunt placement is a safe and effective surgical treatment for symptomatic ACs in children, although efforts should be made to decrease complications in the procedure.

Abbreviations used in this paper: AC = arachnoid cyst; CP = cystoperitoneal; ICP = intracranial pressure; VP = ventriculoperitoneal.
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Patient Outcome

The mean follow-up period was 6.5 years (range 6–8 years). Hemiparesis gradually resolved in 4 of 5 patients. Among 7 patients with macrocrania and fontanel bulging, the size of the head was reduced, fontanel bulging decreased, and satisfactory neurological results were obtained in all except 1 patient. As regards the children who presented with seizures, there was complete seizure control in 9 patients, partial control in 3, and no improvement in 3. Follow-up imaging studies showed that 59 (95%) of 62 ACs reduced in size during the postoperative follow-up period. In 3 patients, the cysts showed no change in size.

Surgical Complications

Several complications were specific to the shunt, and shunt revisions were performed for various reasons in 16 patients (26%). There were 2 cases of skin breakdown and 14 cases of hardware erosion. Among 26 patients (26%), there were 2 cases of skin breakdown and shunt revisions were performed for various reasons in 16 patients (26%).

Discussion

It is widely agreed among neurologists and pediatric neurosurgeons that symptomatic ACs must be surgically treated, including those causing increased ICP or focal neurological deficits. Surgical intervention can improve clinical signs, decrease cyst volume, and prevent cranial deformations and psychological problems when patients reach adolescence. Numerous surgical procedures for effective decompression of ACs have been recommended, including stereotactic aspiration, cyst excision, cyst fenestration, endoscopic cystocisternostomy, CP shunting, and combined surgeries. However, we observed a reduction in cyst size in 95% of cases, and symptom relief was also observed.

Although shunting is an effective method, more attention should be paid to the related complications, such as early shunt failure and lifelong shunt dependence. Other studies, even those supporting shunting as the treatment of choice, have documented a high rate of shunt-related complications. The main problem relates to shunt dependence. In our study, there were 8 patients who showed symptoms of pseudotumor cerebi with a nearly obliterated cyst and 1 patient who demonstrated Chiari malformation Type 1 as the result of overdrainage. This finding reflects an inherent issue with shunts, because this rate is similar to the revision rate when treating other pathologies with a shunt. The reason that shunt dependence happens after CP shunt insertion is still unclear. But CSF hypotension seems to be the triggering factor in overdrainage. Under low ICP, the venous system becomes distended, leading to meningeal swelling, which in turn contributes to dural and suture fibrosis. The low ICP also leads to a deceleration in cranial growth and ultimately craniofacial disproportion. Moreover, a decrease in cerebral pulse pressure probably contributes to the decreased reabsorption of CSF as

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Patient Age (yrs)</th>
<th>Occurrence of Shunt Malfunction (yrs after surgery)</th>
<th>Cyst Location</th>
<th>Treatment</th>
</tr>
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<tr>
<td>1</td>
<td>2</td>
<td>4</td>
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<td>lt frontotemporal</td>
<td>VP shunt</td>
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<td>7</td>
<td>4</td>
<td>1</td>
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<td>shunt revision</td>
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<tr>
<td>8</td>
<td>7</td>
<td>1</td>
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<td>shunt revision</td>
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* Valve type at the initial shunt insertion was low in all 8 patients. All patients had increased ICP after shunt insertion.
well as subependymal gliosis or glial scar formation.\textsuperscript{12,21} Ahn et al.\textsuperscript{1} proposed a “chronic idling” of the CSF absorption mechanism. They suggested that before insertion of the shunt, the CSF drains via the CSF absorption mechanism, but after shunting, this absorption mechanism becomes unnecessary and later regresses, which might lead to shunt dependency. We are still unable to identify which factor could predict which children with shunts would or would not become shunt dependent. However, shunt dependency may be closely related to several factors such as a younger age at shunt insertion, lower-pressure valve type, longer follow-up, and larger initial cyst size.\textsuperscript{27}

Pseudotumor cerebri syndromes must be managed or they will cause neurological deterioration. In our study group, VP shunts were placed when the proximal catheter could not be removed or reinserted. Alternatively, other authors have inserted lumboperitoneal shunts.\textsuperscript{15,17} However, the lumoperitoneal shunt seems to increase the risk of hindbrain herniation, as it might increase the gradient of pressure between the cranial and spinal compartments.\textsuperscript{10}

Decreasing complications in AC shunting is a formidable challenge to neurosurgeons. Given the limitations of devices in the past, we selected a normal valve without an antisiphon device in the study period of this report. Now we try to attach an antisiphon device or implant a programmable valve. With a programmable shunt, it is possible to change the shunt pressure rate and modify the management of CSF problems, which decreases the rate of shunt revision but can’t completely avoid shunt dependence. In our study, the shunt system was successfully removed in 2 patients after complete disappearance of the cysts on follow-up radiological studies, which indicated the possibility of removing the shunt systems to avoid the complication of long-term shunt dependence. But the indicators for removing a shunt still need to be studied. We think that the indicators should not include complete disappearance of the cyst on neuroimaging studies, as it would be sufficient to achieve resolution of the clinical manifestations produced by the ACs. In a recent report in which SPECT scanning was used to define the perfusion defects caused by ACs, it was found that the defects disappeared even though the middle fossa cysts were not completely collapsed. This implies that the drive to collapse the cyst entirely with a CP shunt may represent an unnecessary overengineering of treatment.\textsuperscript{26} Further study is needed to support this observation.

Controversy still exists concerning the treatment of asymptomatic ACs. Al-Holou and associates\textsuperscript{2} followed up 111 patients who each presented with an asymptomatic AC (mean follow-up 3.5 years). They found that cyst enlargement occurred in only 11 patients. No patient older than 4 years demonstrated either clinical changes or cyst enlargement on imaging. These findings indicate that many asymptomatic cysts may not need surgical treatment. However, other investigators have suggested that the potential for hindering normal development and function of the adjacent brain in this group outweighs the risk of operative treatment.\textsuperscript{13,14} Furthermore, some asymptomatic ACs may expand or present significant clinical manifestations such as an acute or chronic subdural hematoma.\textsuperscript{22,23} In our study, there were 15 cases of huge asymptomatic ACs (diameter > 6 cm). These cysts all reduced in size after surgery. However, 4 patients in this group experienced shunt dependency. Therefore, further study is needed to find the most appropriate indication for the surgical treatment of asymptomatic ACs. More reliable criteria should be studied to distinguish between individuals who may benefit from surgical management and those who should only be observed.

Another problem relating to the treatment of ACs is the therapy for epileptic seizures. The relationship between ACs and epilepsy is controversial.\textsuperscript{3} One of the possible mechanisms is mass effect of the ACs, such as compression or irritation of the surrounding cortex, and disturbance of local CSF dynamics.\textsuperscript{6} Thus, for the treatment of epilepsy with ACs, cyst wall resection or CP shunt insertion has been performed.\textsuperscript{29} There are studies demonstrating seizure reduction after the removal of ACs.\textsuperscript{11,16} Cystoperitoneal shunts in some epileptic cases have also had a similar outcome in our experience. In other cases, however, seizures have continued to occur despite the decreased size of the postoperative cyst. Therefore, it is unlikely that mass effect of the AC is the cause of epilepsy in every case. In fact, the lesions are distant from the seizure focus in some epileptic cases. So, a comprehensive study, including neurological symptom assessment and PET studies, is important for conclusions regarding therapy in patients with ACs and intractable epilepsy. On the other hand, comparative studies must confirm the effect of surgical procedures in the future.

Since the advent of neuroendoscopy and its proven effectiveness and safety in many pathologies, several authors have recommended the minimally invasive endo-
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Endoscopic approach for the treatment of ACs as the first therapy of choice, especially for deeply located cysts. Mortolose et al.9 has suggested that the association of an endoscopic fenestration with a programmable valve seems to be a good strategy in treating ACs. They believe that this method can decrease shunt dependence and increase the real possibility of achieving a complete collapse of cysts. But a longer follow-up is necessary to confirm the results.

Conclusions

We believe that shunt placement remains a safe and effective surgical treatment for symptomatic ACs in children, allowing rapid symptom relief and cyst reduction. Nevertheless, given the possibility of shunt dependence, more rigid standards should be adopted in the choice of shunt treatment.

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. Analysis and interpretation of data: B Zhang. Drafting the article: B Zhang. Critically revising the article: all authors. Reviewed submitted version of manuscript: all authors. Approved the final version of the manuscript on behalf of all authors: B Zhang. Administrative/technical/material support: Y Zhang, Ma. Approved the final version of the manuscript on behalf of all authors: B Zhang. Reviewed submitted version of manuscript: all authors. Drafting the article: B Zhang. Critically revising the article: all authors. Reviewed submitted version of manuscript: all authors. Approved the final version of the manuscript on behalf of all authors: B Zhang. Administrative/technical/material support: Y Zhang, Ma.

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