A population-based study of intracranial arachnoid cysts: clinical and neuroimaging outcomes following surgical cyst decompression in children

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Object. If, when, and how children with arachnoid cysts should undergo surgery has been a matter of debate. In the present study the authors describe long-term clinical and neuroimaging results in children with intracranial arachnoid cysts, treated in accordance with the authors’ policy of performing surgery in most of these cases.

Methods. The study included 48 pediatric patients (age < 16 years of age) who underwent surgery for treatment of an arachnoid cyst at Haukeland University Hospital between January 1987 and September 2004. Forty-one patients underwent cyst fenestration to the basal cisterns and posterior fossa. Long-term results were assessed retrospectively on the basis of medical and neuroimaging records. Additional information was obtained by means of a check-box questionnaire completed by the patients and their parents.

At their follow-up examinations, 82% of the patients had no or insignificant complaints, 14% reported no improvement, and 4% noted a worsening of symptoms. In 56% of the patients with appropriate imaging for analysis, the cyst was no longer visible on neuroimaging studies. In 23%, the postoperative fluid volume was reduced to less than 50% of the original cyst volume, and in another 19%, the volume was reduced but was larger than 50% of the original. The cyst was unchanged in only 2%. There was a significant association between a volume reduction greater than 50% and clinical improvement. Three patients (6%), all with temporal cysts, had minor complications that led to additional surgery in one patient. No complications caused permanent disability. Eight patients (17%) underwent additional surgery due to suspected or demonstrated treatment failure.

Conclusions. Most children who underwent cyst fenestration via a craniotomy experienced a good long-term outcome with no severe complications.

KEY WORDS • arachnoid cyst • decompression surgery • long-term follow up • pediatric neurosurgery

Modern neuroimaging techniques have made the diagnosis of arachnoid cysts easier and have resulted in an increasing number of patients of all age groups presenting to neurosurgical departments with such cysts. Although children with arachnoid cysts may experience symptoms of intracranial hypertension or lateralizing symptoms, many present with only mild and rather unspecific symptoms such as headache, learning deficits, or behavioral disturbances. In infants, increasing head circumference may be the only indication. Nevertheless, in previous studies it has been documented that even cysts that cause only mild and unspecific symptoms may affect the function of neighboring cerebral tissue, causing impaired cognition, and that such cognitive deficits tend to normalize after surgery. We previously reported similar reversible cognitive deficits in adults.

In the majority of publications concerning symptomatic arachnoid cysts in children, the authors have advocated surgical intervention for these lesions. Nevertheless, some researchers favor a more conservative approach. Cysts in infants and older children have been reported to grow to a substantial size. Such observations would lend support to treatment by surgical decompression.

Not only is it a matter of debate whether these children should undergo surgery, there are also diverging views on how such surgery should be performed. Some surgeons prefer fenestration through a craniotomy whereas others advocate shunt insertion or fenestration through an endoscopic procedure.

By presenting the results from our department, it is our aim to contribute to the discussion of the questions raised earlier. On the basis of our increasing experience, we have gradually adopted more liberal indications regarding surgery for patients with arachnoid cysts. However, a policy of treating a relatively benign condition with surgery is justified only when a clear clinical benefit and a low risk of
complications can be demonstrated. In the present study we describe clinical and neuroimaging outcomes in children who underwent surgery for an arachnoid cyst in our department between January 1987 and September 2004.

Clinical Material and Methods

Hospital Structure and Study Population

During the study period, Norway had a three-level, hierarchical hospital structure, in which local community hospitals served as primary referral centers. As a secondary referral center, most counties had a central county hospital with a pediatric department that could refer patients to one of the five university clinics, each serving a health region. Only university clinics have a neurosurgical department. A health region is a geographic area consisting of several counties with a well-defined population, and there is little or no overlap between these regions. Haukeland University Hospital is located on the southwestern coast of Norway; it serves three counties with a total population of 930,000 (as of 2003). During the study period, a hospital in the southern part of our health region referred some of its pediatric neurosurgical patients to another university hospital in Norway, thus reducing the population served at the time of the study to approximately 700,000.

Study Design and Data Extraction

The present study is a questionnaire-based, retrospective study that includes all children (age <16 years) who underwent surgery for an arachnoid cyst at Haukeland University Hospital between January 1987 and September 2004. Patients with an arachnoid cyst were identified on the basis of information from the hospital’s computer bank, and each patient with a diagnostic code of 348.0 (ICD 9) or G 93.0 (ICD 10) was registered in the study. Only patients with a verified arachnoid cyst were included. A cross-check was performed with hand-written operating room protocols from the time period as well as with typed surgical records kept by the senior author (K.W.) to ensure that all children who underwent surgery to treat an arachnoid cyst were included.

All the patients underwent a preoperative computed tomography or magnetic resonance imaging examination and a postoperative neuroimaging study within 72 hours. The patients were routinely readmitted 3 to 6 months after surgery for a postoperative visit that included clinical examination and a computed tomography or magnetic resonance imaging study. Patients in whom any postoperative abnormalities (for example, subdural hematoma or hygroma) appeared on neuroimages were followed up further until there was evidence of spontaneous remission or remission after surgical treatment of the complication.

The following data were extracted from patients’ medical records: demographic information; presenting complaints; clinical findings; location and side of the cyst; Galassi type for the temporal cysts;‡ postoperative change in cyst size; surgical method; and possible adverse effects of the operation identified by clinical or neuroimaging examination, such as subdural hygromas or hematomas, or new neurological symptoms or deficits.

During autumn 2004, a personal letter containing an explanation of the purpose of the investigation was sent to the parents or to the child if he or she was considered old enough. A check-box questionnaire, to be completed by the patient, the parents, or both, was enclosed together with a preaddressed, prestamped envelope. The questionnaire solicited information regarding postoperative changes in the presenting complaints, grading of the present level of complaints and level of function in everyday activities (shown on visual analog scales), any regrets parents or patients may have had about choosing a surgical option, and any new complaints that might have resulted from the surgery. There was also ample free space after each question for the respondents to give supplementary information.

Patient Characteristics

A total of 48 patients, 38 boys and 10 girls, were included in the study. The mean age of the patients at surgery was 6.3 years (range 1 month–15 years, median 5.7 years), and the mean follow-up duration was 84 months (range 6–180 months, median 90 months). The cysts were located in the temporal fossa in 29 patients (21 cysts on the left side, six on the right, and two bitemporal), overlying the frontal convexity in five patients (two cysts on the left side and three on the right), and in the posterior fossa in six patients (three cysts on the left side and three at the midline). Eight patients had cysts in other locations. The main preoperative complaints are summarized in Table 1.

In addition to the included patients, two boys underwent surgery for bitemporal arachnoid cysts. When children present with bitemporal cysts, there is a risk that they may have GAT 1, a congenital metabolic disorder that makes any kind of surgery or catabolic state a hazard to the patient. These two patients were found to have GAT 1 and were therefore not included in this study. Their cases have been described in detail elsewhere.26

Surgical Procedures

Forty-one patients (85.4%) underwent primary surgery via craniotomy after induction of general anesthesia. During this operation, the parietal cyst membrane covering the inside of the dura mater was removed from its attachment to the surrounding cortex up to the tentorial slit in cases of middle fossa cysts. The arachnoid membrane that covered the brain surface (the visceral membrane) was not removed.

TABLE 1

<table>
<thead>
<tr>
<th>Symptom</th>
<th>No. of Patients (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>headache</td>
<td>15 (31)</td>
</tr>
<tr>
<td>impaired cognition</td>
<td>13 (27)</td>
</tr>
<tr>
<td>seizures</td>
<td>10 (21)</td>
</tr>
<tr>
<td>behavioral problems</td>
<td>6 (13)</td>
</tr>
<tr>
<td>increasing head circumference</td>
<td>6 (13)</td>
</tr>
<tr>
<td>cerebellar symptoms</td>
<td>6 (13)</td>
</tr>
<tr>
<td>motor symptoms</td>
<td>5 (10)</td>
</tr>
<tr>
<td>oculcar symptoms</td>
<td>4 (8)</td>
</tr>
<tr>
<td>craniofacial anomaly</td>
<td>3 (6)</td>
</tr>
<tr>
<td>speech disorder (including dyslexia)</td>
<td>3 (6)</td>
</tr>
<tr>
<td>posttraumatic subdural hematoma†</td>
<td>2 (4)</td>
</tr>
</tbody>
</table>

* Some patients presented with more than one symptom.
† Cysts discovered by means of imaging investigation after head trauma. The patients had minimal symptoms from the cysts prior to injury.

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Arachnoid cysts in children

Any bridging veins coursing for some distance along the parietal membrane before they drained into the basal sinuses were coagulated and removed. This was done because we regularly observed oozing of blood from the site where these veins drained into the basal sinuses when manipulating the parietal membrane and the vein. Large bridging veins near the sphenoid ridge were not extirpated, as they coursed for only a very short distance along the membrane before draining and were believed to be supported by surrounding tissue in such a way that they would not move and cause bleeding. In cases of temporal cysts, the remaining medial membrane covering the basal structures (the tentorial slit, the oculomotor nerve, the carotid artery, and the optic nerve) was fenestrated, thus creating communication to the basal cisterns and the posterior fossa. When accessible, the arachnoid covering the sylvian fissure was also opened, thus creating communication to the subarachnoid space surrounding the carotid and middle cerebral arteries.

In three patients (6.25%), an internal shunt was inserted from the cyst to the subdural space through a trephine hole after induction of local anesthesia. This method has been described in detail elsewhere.36 One patient received a cysto-toventricular shunt.

In two patients, primary surgery consisted of the insertion of a CP shunt. In one patient, a VP shunt was inserted as the primary measure to treat hydrocephalus. This patient’s condition included multiple cerebral malformations in addition to an arachnoid cyst (dysmorphism of the cerebellum, posterior meningocele, bilateral schizencephaly, and a probable septooptic dysplasia).

Scoring of Clinical Results

The clinical results were categorized into one of four COGs based on information from medical reports and the questionnaire: in COG 1, the preoperative complaints had disappeared entirely or were negligible; in COG 2, the preoperative complaints were clearly reduced but still present; in COG 3, the preoperative complaints were unchanged; and in COG 4, the patient had more complaints after the operation.

Scoring of Neuroimaging Results

The neuroimaging results of the decompression, which are defined as the measured change in cyst volume on the neuroimaging follow-up study obtained after 3 to 6 months, were categorized into one of four possible NOGs: in NOG 1, the cyst had disappeared and was no longer visible; in NOG 2, fluid volume amounting to less than 50% of the preoperative cyst volume was still visible at the site where the cyst had been; in NOG 3, fluid volume was also found, but the residual volume was greater than 50% of the preoperative cyst volume; and in NOG 4, no change in cyst volume could be observed. When the exact categorization was difficult to determine, the postoperative neuroimaging result was always assigned to the less favorable group.

Answers to Letters

Forty-eight letters with questionnaires were mailed to patients. Forty-four patients or parents (92%) responded by filling out and returning the questionnaire.

Statistical Analysis

The statistical analyses were performed using SPSS (version 12.0 for Windows; SPSS Inc., Chicago, IL). An alpha level of 0.05 was used for all statistical calculations. Contingency tables were analyzed using Fisher exact tests or chi-square tests.

Results

Clinical Outcomes

The clinical results for different cyst locations are summarized in Table 2. Four patients did not respond and are therefore not included in the analyses of clinical results.

Twenty-one patients (48%) were completely symptom free at their follow-up examination (COG 1). Another 15 patients (34%) reported a significant reduction in the preoperative complaint (COG 2), whereas six patients (14%) reported no reduction in their complaints (COG 3).

Two patients experienced a worsening in their symptoms after surgery (COG 4), despite an improvement shown on neuroimaging. Both had impaired cognition as the presenting complaint. One of these patients had a left temporal Galassi Type II cyst, and the other, a right frontal cyst. The frontal cyst was not visible on postoperative neuroimages (NOG 1), whereas the temporal cyst was reduced but was still greater than 50% of its original volume (NOG 3).

There was no difference in clinical outcome on the basis of cyst location. When all locations were considered, patients who underwent craniotomy and fenestration had a better outcome than those in whom shunts were placed ($p = 0.014$; Fig. 1). Subgroup analyses for different cyst locations could not be performed because of the small numbers in the groups.

Clinical outcomes for the most common symptoms are summarized in Table 3. No difference was found in the comparison of clinical outcomes (COG 1 and COG 2 compared with COG 3 and COG 4) for the different symptoms.

Neuroimaging Results

The neuroimaging outcomes for different cyst locations are summarized in Table 4. Either pre- or postoperative imaging studies were missing in five patients, and these patients are therefore not included in the neuroimaging analysis. In 24 (56%) of the remaining 43 patients, the cyst was no longer visible (NOG 1) on imaging studies. In another 10 patients (23%), the postoperative fluid volume was reduced to less than 50% (NOG 2). In eight patients (19%), the volume was reduced but was still larger than 50% of the preoperative cyst volume (NOG 3). In only one patient was there no change in postoperative volume (NOG 4).

No difference in neuroimaging outcomes was identified

<table>
<thead>
<tr>
<th>TABLE 2</th>
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<tbody>
<tr>
<td><strong>Clinical outcome in 44 patients with different cyst locations</strong>*</td>
</tr>
<tr>
<td><strong>Cyst Location</strong></td>
</tr>
<tr>
<td>(no. of patients)</td>
</tr>
<tr>
<td>temporal fossa (25)</td>
</tr>
<tr>
<td>frontal region (5)</td>
</tr>
<tr>
<td>posterior fossa (6)</td>
</tr>
<tr>
<td>other (8)</td>
</tr>
</tbody>
</table>

* Data were not available for four patients.
on the basis of cyst location or between the two most frequently used surgical techniques.

**Association Between Clinical and Neuroimaging Results**

In 42 patients, both the clinical and neuroimaging outcomes could be assessed. There was no strong association between postoperative volume and clinical outcome. However, after patients were dichotomized according to their good or poor clinical (COG 1 and 2 compared with COG 3 and 4) and neuroimaging (NOG 1 and 2 compared with NOG 3 and 4) outcomes, there was a significant association between a good clinical outcome and a good neuroimaging outcome ($p = 0.05$).

**Self-Reported Level of Function.** A total of 28 patients (67%) reported better function after surgery. Eight patients (19%) reported no change, and six patients (14%) reported a worse level of overall function after the operation. There was a slight discrepancy between the patients/parents’ estimates of the postoperative functional level and the COG classification based only on the clinical complaints. Of the six patients who reported a worsening of overall function after surgery, two reported complete relief of the preoperative complaint (COG 1), two reported significant relief (COG 2), and two experienced a postoperative increase in the presenting complaint (COG 4). The symptoms of these patients were impaired cognition in two, behavioral disturbances in two, and epilepsy in one. The last of these children was diagnosed with an arachnoid cyst on the basis of increasing head circumference without any other clinical symptoms; in this child postoperative hydrocephalus developed and required insertion of a VP shunt. According to the postoperative neuroimaging results, three of the patients with worsening function were in NOG 1, two were in NOG 2, and one was in NOG 3.

**Complications and Treatment Failures**

Complications are defined in the following sections as additional pathological conditions caused by the treatment, whereas treatment failures are defined as a lack of effect of the treatment. None of the complications encountered were severe, nor did they lead to any permanent sequelae.

**Complications.** Three patients (6%) experienced complications, all of whom had a temporal fossa cyst (Galassi Type II in two and bitemporal in one). The association between complications and cyst location was not significant. One patient had a subdural hygroma and one had a chronic subdural hematoma. In the third patient, who had bitemporal cysts, a postoperative hydrocephalus developed and was treated with a VP shunt. That patient was screened for GAT 1, but this was not found. There was no difference in the complication rate between the different surgical techniques.

**Treatment Failures.** Eight patients (seven boys and one girl) underwent additional surgery because of suspected or demonstrated treatment failure (three temporal fossa, one posterior fossa, one suprasellar, and three in other locations). Four of these patients underwent fenestration through a craniotomy as their primary surgery; in the remaining four, a shunt (cystosubdural, CP, VP, and cysstventricular) was inserted as the primary procedure. Cyst location did not seem to influence the failure rate ($p = 0.132$ for temporal cysts compared with other locations); however, patients who had undergone craniotomy and fenestration as their primary surgery had a significantly lower risk of treatment failure than patients in whom a shunt was placed ($p = 0.01$). Table 5 shows the relationship between the primary and subsequent procedures.

**Discussion**

In this report we show that most of this cohort of pediatric patients with an arachnoid cyst experienced clinical improvement after surgical cyst decompression as well as a significant postoperative reduction in complaints and a better level of overall function. Furthermore, this clinical ben-

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**TABLE 3**

<table>
<thead>
<tr>
<th>Symptom</th>
<th>COG 1</th>
<th>COG 2</th>
<th>COG 3</th>
<th>COG 4</th>
</tr>
</thead>
<tbody>
<tr>
<td>headache</td>
<td>7 (47)</td>
<td>6 (40)</td>
<td>2 (13)</td>
<td>0 (0)</td>
</tr>
<tr>
<td>impaired cognition</td>
<td>3 (23)</td>
<td>6 (46)</td>
<td>2 (15)</td>
<td>2 (15)</td>
</tr>
<tr>
<td>epilepsy†</td>
<td>4 (44)</td>
<td>3 (33)</td>
<td>2 (22)</td>
<td>0 (0)</td>
</tr>
</tbody>
</table>

* Some patients did not experience any of these symptoms.
† Follow-up data were missing in one patient.
Arachnoid cysts in children

The optimal treatment for arachnoid cysts in children has yet to be determined. The most common procedures—shunt placement and fenestration of the cyst—each have their benefits and drawbacks. Some authors advocate endoscopic techniques in the treatment of arachnoid cysts. In previous published comparisons of the results of fenestration and shunt insertion in the treatment of arachnoid cysts, the postoperative volume reduction seen on neuroimages has rarely been described accurately. In papers in which such information is given, a greater degree of volume reduction has been credited to the shunt placement procedure. In our study, however, no difference in volume reduction was found between surgeries involving shunt placement and fenestration. Furthermore, clinical outcome was generally better in the patients who underwent fenestration, and the number of subsequent surgeries for treatment failures was significantly lower.

Cyst decompression through a fenestration also seems more physiologically sound than insertion of a CP shunt. A fenestration balances the pressure gradients that may exist between the cyst interior and the rest of the intracranial cavity, much in the same way as achieved by an endoscopic ventriculostomy. A CP shunt, on the other hand, creates a new intracranial pressure gradient, without equilibrating the intracystic and intracranial pressures. This has been reported to cause headache, which probably is related to the pressure inequality between the intracranial and the intracystic compartments.

Although the numbers in our series are small, we consider the avoidance of shunt insertion in children a major treatment objective, and thus recommend fenestration through a craniotomy as the first treatment choice in children with arachnoid cysts.

Conclusions

Surgical decompression of arachnoid cysts in children yields good long-term outcomes in the majority of patients, with a low risk of complications or additional impairment. We believe that once the surgical option has been chosen, it should be performed as early as possible to avoid a skull–brain mismatch. An additional advantage of early surgery, at least in theory, is that it prevents pressure from the cyst on the underlying, developing brain. Fenestration of the cyst through a craniotomy seems the treatment of choice because of better outcome and a lower rate of treatment failure.

References


Table 5

<table>
<thead>
<tr>
<th>Repeated Op Method</th>
<th>Primary Op Method</th>
</tr>
</thead>
<tbody>
<tr>
<td>craniotomy (no. of patients)</td>
<td>1</td>
</tr>
<tr>
<td>shunt (no. of patients)</td>
<td>3†</td>
</tr>
<tr>
<td>revision of shunt</td>
<td>1</td>
</tr>
<tr>
<td>total no. of interventions</td>
<td>5</td>
</tr>
</tbody>
</table>

* Two craniotomies were performed in one patient.
† A VP shunt for hydrocephalus required two revisions; an endoscopic third ventriculostomy was also required.
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33. Raffel C, McComb JG: To shunt or to fenestrate: which is the best surgical treatment for arachnoid cysts in pediatric patients? Neurosurgery 23:338–342, 1988

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