

INTRACRANIAL ARACHNOID CYSTS: TREATMENT ALTERNATIVES AND OUTCOME IN A SERIES OF 25 PATIENTS

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A series of 25 patients with intracranial arachnoid cysts is analyzed retrospectively. There were 14 males and 11 females ranging in age between a few days and 58 (mean 10) years. Seventy-six percent of patients were children below the age of 15 years. Most of the patients presented with symptoms and signs of long-standing raised intracranial pressure, while localizing signs were rather uncommon. The clinical manifestations were often mild relative to the large size of the cyst. Associated hydrocephalus was present in three patients: one with suprasellar cyst and two with posterior fossa cysts. Seven patients with mild symptoms and small cysts were treated conservatively, while the remaining 18 patients underwent surgical treatment. The initial surgical procedure consisted of craniotomy and fenestration of the cyst in three patients, and cystoperitoneal shunting in the other 15. Of the three patients who underwent craniotomy, one improved postoperatively, while the remaining two developed complications consisting of wound infection and postoperative epilepsy in one and permanent severe neurological deficit in the other. In contrast, six of the 15 cysts treated by shunting resolved completely, eight were smaller, and one remained unchanged. Radiological regression of the cyst after shunting was associated with various degrees of clinical improvement in 13 patients (87%). Two (13%) of 15 shunted patients developed complications in the early postoperative period, consisting of wound infection in one and early shunt failure in the other. Three patients (20%) with shunts had late complications during the follow-up period, consisting of recurrent shunt failure in the first, subdural hematoma in the second, and perforation of the peritoneal catheter into the hepatic bile ducts in the third. These findings, as well as recent data from the literature, suggest that in the management of intracranial arachnoid cysts, cystoperitoneal shunting was more effective and had fewer serious complications than craniotomy and cyst fenestration, and therefore, it is recommended as the treatment of first choice. *Ann Saudi Med* 1997;17(3):288-292.

Arachnoid cysts are non-tumorous intra-arachnoid fluid collections that account for about 1% of all intracranial space-occupying lesions.¹ They may develop throughout the cerebrospinal axis, with a predominance in the sylvian region.^{2,3} Because of their benign nature and slow expansion, arachnoid cysts may remain symptomatic or produce only subtle symptoms and signs. On the other hand, they sometimes give rise to focal neurological deficits, raised intracranial pressure, and/or epileptic seizures. The question of when these lesions should be operated upon is, therefore, not always easy to answer. Moreover, the choice of the most appropriate surgical approach is still debated.^{2,4-8}

In the following report, the author describes the clinical and radiological findings in 25 patients with intracranial arachnoid cysts. In addition, the treatment results are analyzed and compared with those published in

the literature with the goal of determining the advantages and disadvantages of the various surgical approaches.

Material and Methods

The hospital records of 25 consecutive patients with intracranial arachnoid cysts who were treated in the Neurosurgical Units at King Khalid University Hospital (15 cases) and Security Forces Hospital (ten cases) between 1985 and 1995, were analyzed retrospectively. The diagnosis of arachnoid cyst was based on computerized tomography (CT) scan findings demonstrating a well-circumscribed and non-enhancing cystic lesion that had attenuation values similar to those of the cerebrospinal fluid (CSF) and did not communicate with the ventricular system.⁹⁻¹¹

There were 14 males and 11 females, ranging in age from a few days to 58 years, with an average of ten years. Nineteen (76%) patients were children below the age of 15 years.

A summary of the presenting clinical symptoms and neurological status of the patients on admission are shown in Table 1. In four patients, intracranial arachnoid cysts had already been suspected or detected *in utero* during

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TABLE 1. *Presenting symptoms and signs.*

Neurological findings	Number of patients*
Normal neurological status	10
Macrocephaly	9
Headache	6
Cranial asymmetry and local skull bulging	4
Mental and motor retardation	4
Muscular hypotonia	3
Blurring of vision	3
Seizures	2
Dizziness	2
Unsteady gait	1
Papilledema	1
Squint	1
Sunset phenomenon	1
Spastic hemiparesis	1
Failure to thrive	1
Behavioral abnormality and learning difficulty	1
Deafness**	1
Hand tremor**	1

*Many patients had more than one neurological finding; **these findings were probably unrelated to the arachnoid cyst.

routine ultrasound examinations and three of these patients were macrocephalic, while the fourth had normal head size.

Twenty patients had supratentorial cysts distributed as follows: 12 sylvian, two suprasellar, one interhemispheric, one holohemispheric, one temporal, one frontal, one bifrontal, and one parietoccipital. The remaining five patients had infratentorial cysts, of which two were supracollicular, one posterior to the vermis, and two lateral to cerebellar hemisphere. Two sisters in this series had bilateral arachnoid cysts of the sylvian regions associated with glutaric aciduria type 1.¹²

In 18 (72%) patients, the cysts exhibited local pressure effects, predominantly compression of the ipsilateral ventricle with (n=7) or without (n=11) shift of the midline structures. Three patients (one with a suprasellar and two with post-fossa cysts) had associated obstructive hydrocephalus, and in a fourth case, the cyst resulted in sequestration and isolated dilatation of the ipsilateral posterior horn.

Eighteen patients were treated surgically. The initial surgical procedure consisted of craniotomy and fenestration of the cyst wall in three patients and placement of a low pressure cystoperitoneal shunt in 15 patients. The remaining seven patients received no specific treatment. They all had small cysts and their symptoms and signs were mild or seemed not to be related to the cysts.

The outcomes were expressed as improved, unchanged, or worse, based on how the preoperative symptoms and signs were affected by surgical treatment. In addition, the postoperative change in the size of the cyst on CT scan or MRI was classified as complete resolution, reduction in size, and unchanged.

The postoperative complications were divided into early (occurring during the same period of hospitalization up to 30 days postoperatively), and late complications (occurring after discharge from the hospital or later than 30 days postoperatively). The efficacy of the different surgical treatment methods was further assessed by estimating the re-operation ratio for each procedure. This ratio is defined as the number of complication-related operations divided by the number of patients with complications who received the same initial treatment.⁸

All patients were followed up for six months to 7.5 years (average 38±14 months) after the initial diagnosis.

Results

All seven patients with untreated cysts remained clinically stable, and the size of their cysts did not change on CT scan during follow-up periods ranging from six to 39 (mean 27) months.

In the three patients treated initially by craniotomy and cyst fenestration, the cyst resolved completely in one, was smaller in another, and remained unchanged in the third. In the first patient who had a suprasellar arachnoid cyst, the preoperative visual symptoms disappeared and six months after surgery, there was no recurrence of the cyst. The second patient developed postoperative wound infection and required two more operations: one for removal of the bone flap and evacuation of a subdural empyema, and the other for cranioplasty eight months later. He ultimately made a fairly good recovery without neurological deficit, but suffered occasionally from new generalized epileptic seizures. The third patient underwent an uneventful fenestration of a suprasellar arachnoid cyst and at the end of the operation, was awake and could be smoothly extubated. However, while in the intensive care unit, he was found three hours later apneic and deeply cyanosed. His arterial pO₂ was 40 torr and pCO₂ was 70 torr. Emergency CT scan revealed congested brain without intracranial hemorrhage. The lateral ventricles were dilated, as before surgery, and contained a lot of air. He underwent immediate placement of an external ventricular drainage (EVD), which was converted a few days later into a ventriculoperitoneal shunt. The patient survived with severe neurological disability and frequent convulsions until he died seven years later following a status epilepticus. The re-operation ratio in this patient group was 2.0.

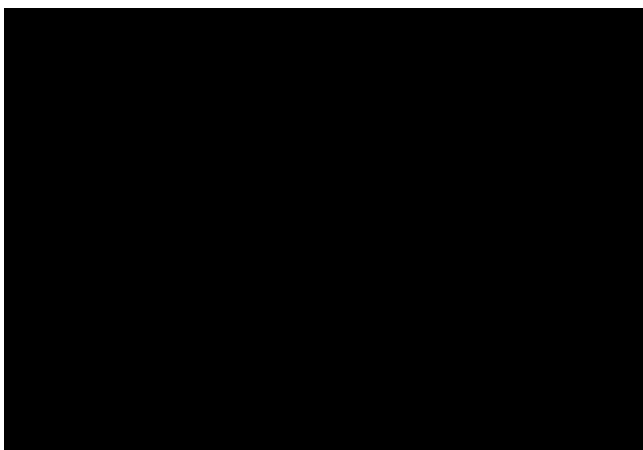


FIGURE 1. A) CT scan showing a large right frontotemporal arachnoid cyst in an 11-month-old boy with delayed psychomotor development. B) Follow-up CT scan 3.5 years post-cystoperitoneal shunt demonstrating complete resolution of the cyst.

Among the 15 patients treated initially by cyst shunting, the cyst resolved completely in six (Figure 1), was markedly smaller in six, slightly smaller in two, and remained unchanged in one. Reduction of the cyst size was associated in 13 (87%) cases with clinical improvement of different degrees. Focal neurological deficit, and symptoms and signs of raised intracranial pressure were more readily improved than other less specific manifestations, such as developmental delay. Two patients did not benefit from cyst shunting, but no patient was made worse by the procedure.

Among the 12 patients in whom the cyst resolved or decreased markedly after shunting, three patients developed asymptomatic ipsilateral subdural hygroma which resolved spontaneously on subsequent CT scans without re-expansion of the cyst. Three more patients had persistent dilatation of the ipsilateral ventricle, suggesting the presence of focal brain atrophy.

Five (33%) of the 15 patients treated by cyst shunting had complications that required additional treatment. The complication occurred early in the postoperative period in two patients (13%), one of whom developed shunt infection with *Staphylococcus aureus*, necessitating removal of the infected shunt and external drainage of the cyst, followed later by placement of a new cystoperitoneal shunt. Both patients made excellent recoveries, however, with good reduction of the cyst size. The remaining three patients (20%) developed the following late complications: one infant with a posterior fossa cyst suffered from recurrent shunt failure for which he underwent two shunt revisions and finally craniotomy and cyst fenestration. His cyst decreased in size, but he remained in poor neurological condition with generalized hypotonia and poor sucking reflex until he died of aspiration pneumonia at the age of two years. The second patient bled into a

residual cyst following minor head injury two months after shunting. Because his neurological status was normal and there was no cyst enlargement on CT scan, he was managed conservatively. The third patient presented with right hypochondrial tenderness two years after cyst shunting. Shuntography revealed a most peculiar perforation of the peritoneal catheter into the bile tree. As this patient had not benefited clinically from the shunting procedure and the cyst was only slightly smaller in size, the shunt system was removed completely. Six months later, the patient was clinically unchanged and the cyst did not increase in size. The re-operation ratio in this group of patients was 1.4.

A summary of the results of both treatment methods is shown in Table 2.

Discussion

Intracranial CSF-containing cysts may be caused by a variety of mechanisms, including trauma, hemorrhage, and inflammation.^{2,13,14} True arachnoid cysts are, however, widely accepted to be developmental anomalies in which splitting or duplication of the primitive arachnoid membrane in the early embryonal life leads to intra-arachnoid fluid collection.^{3,15-18} These lesions may remain small and clinically silent, or enlarge and produce symptoms mainly by virtue of their mass effect and pressure on the surrounding neural structures. The cyst enlargement seems to be the result of secretory activity of the arachnoid cells lining the cyst,¹⁹ but other mechanisms such as infiltration across osmotic gradient and unidirectional flow through a ball-valve arrangement have also been considered.²⁰

The clinical manifestations of arachnoid cysts are variable and often unspecific.^{14,21-26} The most common presenting symptoms and signs are those of raised intracranial pressure, craniomegaly, and developmental delay.^{7,10} They are usually seen in cases of large supratentorial cysts, but may also be caused by smaller suprasellar or posterior fossa cysts associated with obstructive hydrocephalus.²¹⁻²⁴ Focal neurological deficits and epilepsy are present in less than 30% of patients with middle fossa arachnoid cysts.^{2,7} A similar percentage of patients with suprasellar cysts may show visual impairment.^{2,6,21,24} In children, arachnoid cysts often produce mild neurological impairment relative to their large size, as seen in many of the patients in this series who had large sylvian cysts but no focal neurological deficits.

With these wide variations in the natural course and clinical manifestations, there has been considerable controversy regarding the indications for surgical treatment of arachnoid cysts. Galassi et al.² concluded from

their experience with 25 cases of middle fossa arachnoid cysts that all arachnoid cysts should receive surgical treatment because of their frequent complications, including acute cyst enlargement, subdural effusion following rupture of the cyst, and subdural or intracystic bleeding. This view was supported by Sato et al.³ based on their observation of intracystic CSF flow disturbances.

In this series, the author elected, in agreement with other authors,^{8,27} a more selective approach, with surgical treatment being recommended only for patients with steady progression of neurological symptoms and patients with large space-occupying cysts, whereas asymptomatic cysts and cysts producing only vague symptoms or minor skull deformity were followed clinically and radiologically with CT scan or MR imaging. None of the seven patients in this series who were not operated upon had any clinical progression of symptoms or radiological enlargement of lesion during a mean follow-up period of 27 months. Patients presenting with seizures were treated initially with anticonvulsants, reserving surgery for those with intractable epilepsy.

The goals of treatment are to drain the cyst and to prevent the recollection of fluid, allowing for re-expansion of the compressed neighboring neural tissue. The optimal treatment method is still a debated subject. Basically, two surgical approaches have been used: 1) Craniotomy with cyst excision or marsupialization into the subarachnoid space, basilar cisterns, or ventricles, and 2) cystoperitoneal shunt (and/or ventriculoperitoneal shunt in cases of associated hydrocephalus). The main advantages of the first approach are that it allows for histopathological diagnosis and it can leave the patient shunt-independent.

In this small series, the results of the three cysts treated by craniotomy and fenestration were modest, with only one patient improving after surgery, while the other two became worse due to early postoperative complications.

A review of the literature confirms the high incidence of complication and recurrence after this type of treatment. Stein⁵ quoted a postoperative mortality rate as high as 20%, while the postoperative complications in two recent publications were 38% and 40%, including aseptic meningitis, subdural hemorrhage, increased neurological deficit, and new epilepsy.^{8,27} And between 31% and 60% of the cysts treated by craniotomy and wall excision recurred, necessitating additional surgical treatment, mostly a shunting procedure.^{6,8,10} Moreover, coexisting hydrocephalus is rarely controlled by craniotomy and cyst excision, and ventriculoperitoneal shunt will be necessary in almost all these cases.²⁷ These results raise considerable doubt regarding the justification for this form of treatment as the primary treatment. Modern neuroimaging techniques, such as CT scan and MR imaging, allow accurate differentiation of arachnoid cysts from other intracranial cystic lesions, whether neoplastic, inflamma-

TABLE 2. Summary of the treatment results: a comparison between craniotomy with cyst fenestration and cystoperitoneal shunting.

	Craniotomy and cyst fenestration	Cystoperitoneal shunting
Number of patients	3	15
Outcome		
Improved	1	13
Unchanged	1	2
Worse	1	0
Complications	2 (67%)	5 (33%)
Re-operation ratio	2.0	1.4

tory, or parasitic, so that histopathological confirmation of diagnosis is rarely necessary in most of the cases.^{28,29}

The effectiveness of cystoperitoneal shunts in the treatment of arachnoid cysts was first reported by Geissinger et al.³⁰ in 1978 and later confirmed by several authors.^{4,5,23,25,31} In two recent studies, primary cystoperitoneal shunting of arachnoid cysts was found more effective and associated with fewer complications than craniotomy and cyst fenestration.^{8,27} Ciricillo et al.²⁷ reported that all 20 patients treated initially by cystoperitoneal shunt were improved postoperatively, whereas ten of 15 (67%) patients treated initially by craniotomy and cyst fenestration showed no clinical or radiographic improvement. Lawton et al.⁸ found that 91% of their 11 patients treated initially with cystoperitoneal shunts were improved postoperatively, compared with 69% of the 13 patients treated with cyst fenestration.

Although the shunt malfunction rate ranged between 30% and 45%, revision of the shunt usually resulted in good recovery and serious complications were rare.^{8,27} In this series, postoperative reduction of the cyst size was achieved in 93% of the patients, with 87% of them also showing various degrees of clinical improvement. The overall complication rate was 33%, of which 20% were due to shunt malfunction, and 7% due to shunt infection. While these results confirm the experiences of other authors concerning the efficacy and relative safety of cystoperitoneal shunting in the management of arachnoid cysts, they demonstrate some of the shunt problems, which are basically the same as those known from the shunting of hydrocephalus.³² A major drawback of shunting is that a shunt must remain in place throughout the patient's life, which may impose a few limitations on the patient. Nevertheless, considering the lower rate and severity of the postoperative complications, cystoperitoneal shunting should be the primary treatment of arachnoid cysts. Only patients with recurrent shunt failure should be considered for craniotomy and cyst excision.

In recent years, treatment of arachnoid cysts by endoscopic fenestration have been described by various authors.^{33,34} This treatment combines the advantages of minimal invasiveness and shunt independence.

Nevertheless, considering the ability of an arachnoid cyst to reconstitute its wall after craniotomy and wide excision, long-term follow-up results should be awaited before a final conclusion regarding the usefulness of this technique can be made.

In conclusion, this series of intracranial arachnoid cysts shows that these benign developmental lesions usually present in children with unspecific symptoms of raised intracranial pressure and less commonly with focal signs suggestive of their location. CT scan and MR imaging are diagnostic in the majority of cases, alleviating the need for histopathological confirmation. Small cysts with minimal symptoms should be treated conservatively with regular clinical and radiological follow-up examinations at six-month to one-year intervals. Large space-occupying cysts and those causing neurological impairment require surgical treatment. At present, cystoperitoneal shunting is the treatment of first choice. Craniotomy and excision of the cyst should be reserved for those cases with recurrent shunt failure. The role of endoscopic cyst fenestration is still to be determined.

References

- Robinson RG. Congenital cysts of the brain: Arachnoid malformations. *Prof Neurol Surg* 1971;4:133-73.
- Galassi E, Piazza G, Giast G, Frank F. Arachnoid cysts of the middle fossa: a clinical and radiological study of 25 cases treated surgically. *Surg Neurol* 1980;14:211-19.
- Sato K, Shimoji T, Yagushi K, Sumie H, Kuru Y, Ishii S. Middle fossa arachnoid cyst: clinical, neuroradiological, and surgical features. *Child's Brain* 1983;10:310-16.
- Kato M, Nakada Y, Ariga N, Kakubo Y, Makino H. Prognosis of four cases of primary middle fossa arachnoid cyst in children. *Child's Brain* 1980;7:195-204.
- Stein SC. Intracranial developmental cysts in children: treatment by cystoperitoneal shunting. *Neurosurgery* 1980;8:647-50.
- Rappaport ZH. Suprasellar arachnoid cysts: options in operative management. *Acta Neurochir (Wien)* 1993;122:71-75.
- Artico M, Cervoni L, Salvati M, Fiorenza F, Caruso R. Supratentorial arachnoid cysts: clinical and therapeutic remarks on 46 cases. *Acta Neurochir (Wien)* 1995;132:75-8.
- Lawton MT, Golfinos JG, Harrington T, Sonntag VKH, Spetzler RF. Surgical management of intracranial arachnoid cysts. *BNI Quarterly* 1995;11:14-8.
- Banna M. Arachnoid cysts on computed tomography. *Am J Roentgen* 1976;127:979-82.
- Anderson FM, Segall HD, Caton WL. Use of computerized tomography scanning in supratentorial arachnoid cysts. *J Neurosurg* 1979;50:333-38.
- Leo JS, Pinto RS, Hulvat GF, Epstein F, Kricheff II. Computed tomography of arachnoid cysts. *Radiology* 1979;130:675-80.
- Jamjoom ZAB, Okamoto E, Jamjoom AHB, Al-Hajery O, Abu-Melha A. Bilateral arachnoid cysts of the sylvian region in female siblings with glutaric aciduria. *J Neurosurg* 1995;82:1078-1081.
- McCullough DC, Herbert JC, Manz HJ. Large arachnoid cysts at the cranial base. *Neurosurgery* 1980;6:76-81.
- Menezes AH, Bell WE, Perret GE. Arachnoid cysts in children. *Arch Neurol* 1980;37:168-72.
- Starkman SP, Brown TS, Linell EA. Cerebral arachnoid cysts. *J Neuropath Exp Neurol* 1958;17:484-500.
- Choux M, Rayboud C, Pinsard N, Hassoun J, Gambarelli D. Intracranial supratentorial cysts in children excluding tumor and parasitic cysts. *Child's Brain* 1978;4:15-32.
- Go KG, Houthoff HJ, Blaauw EH, Stokross I, Blaauw G. Morphology and origin of arachnoid cysts: scanning and transmission electron microscopy of three cases. *Acta Neuropath* 1978;44:57-62.
- Rengachary SS, Watanabe I, Brackett CE. Pathogenesis of intracranial arachnoid cysts. *Surg Neurol* 1978;9:139-44.
- Smith RA, Smith WA. Arachnoid cysts of the middle cranial fossa, surgical considerations. *Surg Neurol* 1976;5:246-52.
- Go KG, Houthoff HJ, Blaauw EH, Havinga P, Harsuiker J. Arachnoid cyst of the sylvian fissure. Evidence of fluid secretion. *J Neurosurg* 1984;60:803-10.
- Raimondi AJ, Shimoji T, Gutierrez FA. Suprasellar cysts: surgical treatment and results. *Child's Brain* 1980;7:57-72.
- Di Rocco C, Caldarelli M, di Trapani G. Infratentorial arachnoid cysts in children. *Child's Brain* 1981;8:119-33.
- Vaquero J, Carrilo R, Cabezedo JM, Nombela L, Bravo G. Arachnoid cysts of the posterior fossa. *Surg Neurol* 1981;16:117-21.
- Hoffman JH, Hendrick EB, Humphreys RP, Armstrong EA. Investigations and management of suprasellar arachnoid cyst. *J Neurosurg* 1982;57:597-602.
- Harsh IV GR, Edwards MSB, Wilson CB. Intracranial arachnoid cysts in children. *J Neurosurg* 1986;64:835-42.
- Dei-Anang K, Voth D. Cerebral arachnoid cysts: a lesion of the child's brain. *Neurosurg Rev* 1989;12:59-62.
- Ciricillo SF, Cogen PH, Harsh GR, Edwards MSB. Intracranial arachnoid cysts in children. A comparison of the effects of fenestration and shunting. *J Neurosurg* 1991;74:230-35.
- Mayr U, Aichner F, Bauer G, Mohsenipour I, Pallua A. Supratentorial extracerebral cysts of the middle cranial fossa. A report of 23 consecutive cases of the so-called temporal lobe agenesis syndrome. *Neurochirurgia* 1982;25:51-6.
- Pomeranz S, Wald U, Amir N, Shalit M. Arachnoid cysts: unusual aspects and management. *Neurochirurgia* 1988;31:25-8.
- Geissinger JD, Kohler WC, Robinson BW, Davis FM. Arachnoid cysts of the middle fossa: surgical considerations. *Surg Neurol* 1978;10:27-33.
- Kaplan BJ, Mickle JP, Parkhurst R. Cystoperitoneal shunting for congenital arachnoid cysts. *Child's Brain* 1984;11:304-11.
- Hirsch J-F, Hoppe-Hirsch E. Shunts and shunt problems in childhood. In: Symon L, ed. *Advances and Technical Standards in Neurosurgery*. Vol. 16. New York: Springer Verlag, 1988:177-96.
- Caemert J, Abdullah J, Calliauw L, Carton D, Dhooge C, van Coster R. Endoscopic treatment of suprasellar arachnoid cysts. *Acta Neurochi (Wien)* 1995;119:68-73.
- Schroeder HWS, Gaab MR, Niendorf W-R. Neuroendoscopic approach to arachnoid cysts. *J Neurosurg* 1996;85:293-8.